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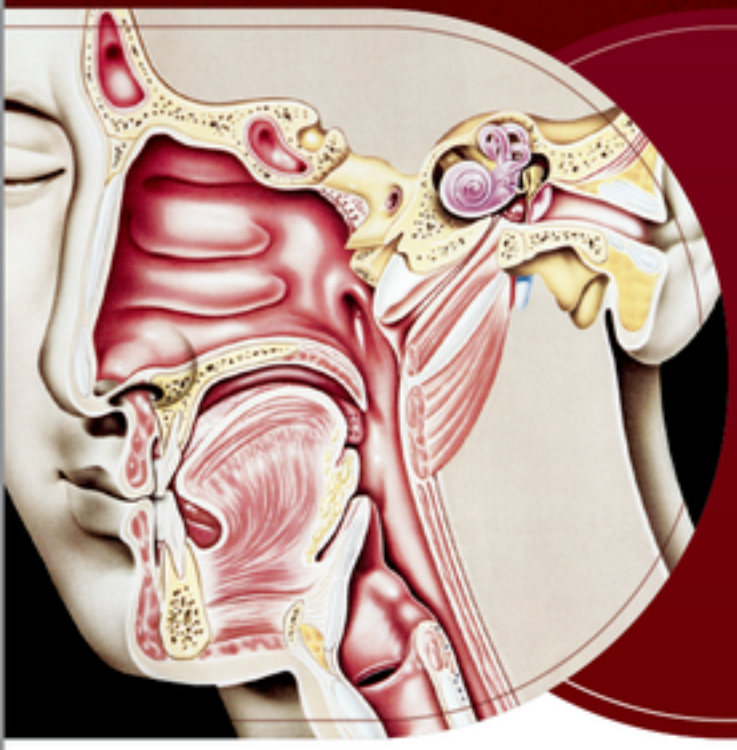

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I dedicate this work to my amazing parents: Martyn and Paula; my wonderful better-half: Beth-Anne; my inspirational children: Lydia and Benson; and most importantly: to Doron.
JSP

I dedicate this work to my beautiful wife and our children—thank you for your ongoing support, you are all the foundation upon which my life and career are built. X
RB

I dedicate this work to my wife, Koki, who picked up the slack from my reduced contribution to family commitments, patiently and gracefully, as I juggled a heavy post-COVID clinical workload, research commitments, and book editing. I can't thank you enough, dearest Koks.
VP

I dedicate this work to Dad, Carol, Kerry, and Mum and to my wonderful wife Christine and our amazing children Joseph, Grace, and Aidan. Thank you for all your love and support.
SMP

Foreword

The great attraction of otolaryngology has always been the breadth of its component parts, affecting all age groups and covering the widest range of pathologies and treatments from the commonest of conditions seen in primary care to some of the rarest, requiring tertiary investigation and state of the art treatments. This makes it difficult for those training and working in otolaryngology and its allied specialities to learn about and keep up to date with all its component parts. The *Oxford Textbook of Otolaryngology* provides the perfect solution to this problem, supporting trainees faced with their final professional examinations as well as established consultants wanting to refresh their knowledge across all aspects of the speciality. Or indeed is useful to anyone with an interest in an up-to-date synthesis of the current state of knowledge in this area.

Otolaryngology has for many years been at the forefront of innovation and technological advancement, both in diagnostics and in an expanding range of medical and surgical treatment modalities. It has benefited from refinements in imaging, anaesthesia, immunology, and chemotherapeutics while also driving technical and scientific advances in reconstructive and preservative approaches to the ear, nose, and throat, as well as the skull base and head and neck cancer. No other speciality impacts so many physiological functions vital to quality of life and our survival, including hearing, balance, airway, smell, swallowing, and voice.

To provide all of this information in one comprehensive and readable volume is quite a task, but to that end, the *Oxford Textbook of Otolaryngology* brings together an amalgam of UK and international experts in their fields, under the leadership of four excellent editors, to provide a distillation of practical as well as evidence-based management. As a consequence, some chapters are based on specific symptoms and signs while others deal with particular conditions or surgical procedures. Controversies are confronted and the ever-expanding guidelines presented, often by those involved in their authorship, together with a careful selection of further reading rather than extensive lists of references.

Importantly, chapters on good clinical practice, ethics, and the rights of the patient head up the text as well as an important section on COVID which impacted the practice of ear, nose, and throat surgeons and their patients more than many other areas of medicine. With the continued threats of airborne disease, pollution, and climate change, these issues are likely to be with us for the future.

This concise and accessible text comprehensively covers the problems and their solutions in paediatric and adult otolaryngology for anyone with an interest in the subject.

Professor Dame Valerie Lund DBE FRCS

Preface

A simple internet search for otolaryngology textbooks will reveal a vast array of material, ranging from simple, concise revision aids to comprehensive, authoritative texts covering multiple volumes. The challenge that I proposed to our team of section editors from the initial stages of conception of this book was to create a book that can be read cover to cover, is practical, and covers all the information that is essential for an otolaryngology trainee to pass key examinations, and for established specialists to remain up to date and to meet the necessary requirements for revalidation.

The *Oxford Textbook of Otolaryngology* covers all the knowledge that clinicians from all subspecialties of otolaryngology would require to keep abreast of current practice, with a strong emphasis on evidence-based practice. The book is first split into the expected sections related to otology and audiology, rhinology, head and neck surgery, and paediatric otolaryngology. These sections are then further split into two broad subsections: the first covering common presenting symptoms and signs, and the second covering specific conditions.

While this book has been conceived in the UK, the section editors have drawn upon their network of respected international authorities on the particular subject matter at hand, to allow the reader unrestricted access to expertise from around the world. While national guidelines may vary, the fundamental principles of diagnosis and management of common conditions should not differ. Where possible, dedicated emphasis on how to perform key procedures and what issues are essential for consent are clearly expressed; this is further supplemented with 'how to' diagrams. Like many areas of medicine, the practice of otolaryngology is based on appreciating where gaps in knowledge occur and where controversies need to be considered in order to offer our patients the best care available. The ability to reflect upon what we do not know, rather than what we do know, is often the greatest inspiration to develop flexible thinking and to promote further research into important areas. These difficult aspects of otolaryngology practice are fully acknowledged and explained throughout the main chapters of the book.

The introductory chapters of this textbook aim to provide insight into emerging aspects of medicine that are essential for the day-to-day practice of otolaryngology. The first chapter reviews the roles and responsibilities of the twenty-first-century otolaryngologist, not only for the provision of best care for our patients, but also as a role model to surgeons in training in how to practise 'good medicine'. The second chapter outlines some of the more common ethical issues that we might face when we need to act in the best interest of our patients. The third chapter considers the anaesthetic aspects of otolaryngology and has been authored by the current vice-president of the Royal College of Anaesthetists. While it was not our intention to provide a 'how to' chapter on providing a general anaesthetic, an appraisal of those aspects relevant to the otolaryngologist has been carefully detailed. This book was conceived pre-COVID, written during the COVID pandemic, and by the time you are reading this preface I very much hope that COVID will have been a distant memory (more so that the COVID pandemic has passed swiftly, rather than there being a significant delay in the book's submission and subsequent production). The final introductory chapter of this book outlines the challenges and implications of COVID for the practice of otolaryngology.

It has been a great honour to work alongside three highly respected colleagues who oversaw the production of the sections covering rhinology, head and neck surgery, and paediatric otolaryngology. We have had the opportunity to pick the best authors from around the world to contribute to this book. While exhausting, it has been great fun compiling this book and I very much hope you enjoy reading it.

John S. Phillips, PhD FRCS (ORL-HNS)

Contents

List of Contributors xv
List of Abbreviations xix

SECTION 1
Introduction

1. **Being a good doctor** 3
   Peter Chapman
2. **Rights of the patient** 7
   Martyn Patel
3. **Anaesthesia for otolaryngological surgery** 12
   Simon Fletcher
4. **Practising otolaryngology in the COVID era** 19
   Andrew Thamboo

SECTION 2
Otology and audiology

5. **Clinical and audiological assessment of hearing in adults** 27
   Gemma Crundwell
6. **Tinnitus and disorders of sound tolerance** 37
   Don McFerran
7. **Vertigo** 45
   Peter Rea and Joel A. Goebel
8. **Otalgia and otorrhoea** 59
   Philip J. Clamp
9. **Aural fullness** 69
   Holger H. Sudhoff
10. **The facial nerve** 72
    Richard M. Irving
11. **Sensorineural hearing loss** 77
    Ted A. Meyer
12. **Conductive (or mixed) hearing loss with an intact tympanic membrane** 81
    Paul Merkus and Thadé Goderie
13. **The external ear** 92
    Pablo M. Reyes Burneo and Miguel Vaca González
14. **Ménière's disease and vestibular migraine** 105
    Louisa Murdin
15. **Benign paroxysmal positional vertigo** 112
    Jeffrey D. Sharon and Danielle Gillard
16. **Audiovestibular implants** 119
    Neil Donnelly and Patrick Axon
17. **Noise-induced hearing loss** 125
    Peter Webber
18. **Lateral skull base disease** 131
    J. Walter Kutz Jr and Daniel E. Killeen
19. **Cholesteatoma** 141
    Neil Bailie and Colin Leonard
20. **Otosclerosis and stapes surgery** 148
    Christopher Aldren
21. **Ossiculoplasty** 154
    Christopher Aldren
22. **Tympanoplasty** 160
    Matthew P.A. Clark
23. **Temporal bone trauma** 166
    Nicola Quaranta and Vito Pontillo
24. **Necrotizing otitis externa** 176
    Emma Stapleton and Glen Watson

SECTION 3
Rhinology

25. **Clinical assessment of the nose and paranasal sinuses** 183
Andrew C. Swift

26. **Olfactory disorders** 191
Carl Philpott

27. **Epistaxis** 198
Rajiv Bhalla and John S. Phillips

28. **Nasal septal perforation** 204
Tim Woolford

29. **Rhinorrhoea** 208
Amin R. Javer

30. **Postnasal drip** 215
Isma Iqbal

31. **Headache and facial pain** 220
Iain Hathorn

32. **Epiphora** 227
Rohit D. Gohil and Ashok Rokade

33. **Nasal obstruction** 236
Peter Andrews and Alfonso Luca Pendolino

34. **Nasal crusting** 242
Catherine Rennie

35. **Rhinoplasty** 247
Fazil Apaydin

36. **Acute rhinosinusitis** 267
Isam Alobid

37. **Endoscopic sinus surgery** 274
Claire Hopkins

38. **Chronic rhinosinusitis** 281
Richard J. Harvey and Janet Rimmer

39. **Sinonasal tumours** 290
Carl H. Snyderman and Eric W. Wang

40. **Surgical facial rejuvenation** 297
Ozcan Cakmak and Mahmoud Daoud

41. **Non-surgical facial rejuvenation** 309
Alwyn R. D'Souza and Chew Lip Ng

42. **Allergy of the nose** 314
Richard Douglas

43. **Hereditary haemorrhagic telangiectasia** 319
Simon Gane

44. **Endoscopic approaches to the anterior skull base** 326
Haissan Iftikhar and Shahzada Ahmed

45. **Anterior skull base reconstruction** 333
Caroline Smith, Omar N. Pathmanaban, and Rajiv Bhalla

SECTION 4
Head and neck surgery

46. **Principles of clinical assessment and management of the head and neck patient** 343
Jemy Jose and B.Y. Winson Wong

47. **Dysphonia** 348
Yakubu Gadzama Karagama

48. **Dysphagia** 356
Somiah Siddiq

49. **Sore throat** 368
Jenny Montgomery

50. **Clinical assessment of neck lumps** 375
David Walker and Cameron Davies-Husband

51. **Snoring and sleep apnoea** 386
Khalid Ghufoor and Jahangir Ahmed

52. **Acute and subacute airway obstruction** 391
Navin Mani, Amy Teasdale, and Sofia Clegg

53. **Head and neck trauma** 397
Jaiganesh Manickavasagam and Kishore Shekar

54. **Oral ulceration: A pragmatic guide to diagnosis and management** 406
Stefano Fedele and Stephen Porter

55. **Laryngeal precancer and cancer** 411
David Hamilton and Anna Thompson

56. **Oropharyngeal cancer** 419
Zaid Awad and Dorothy M. Gujral

57. **Oral precancer and cancer** 428
Deepa Nair, Florida Sharin, and Sarbani Ghosh Laskar

58. **Salivary gland tumours** 438
Khanh K. Nguyen and Nitin A. Pagedar

59. **Thyroid cancer** 444
Hannah Nieto and Neil Sharma

60. **Hypopharyngeal cancer** 450
Laura Warner and Kee Wong

61. **Nasopharyngeal cancer** 456
Shreerang A. Bhide and Pablo Nenclares

62. **Adult deep neck space infections** 465
Zi-Wei Liu and Abdul Ahmed

63. **Parathyroid disorders** 470
Ekpemi Irune, Brian Fish, and Ruth Casey

64. **Principles of head and neck reconstruction** 477
Vijay V. Haribhakti

65. **Non-surgical treatments for head and neck cancer** 491
Muhammad Shahid Iqbal and Charles Kelly

66. **Rehabilitation of the head and neck cancer patient** 498
Sarah Stephen and Joanne Patterson

67. **Benign neck lumps** 508
Samantha Hassid, Georges Lawson, and Sébastien Van Der Vorst

68. **Head and neck pathology** 520
Max Robinson and Selvam Thavaraj

69. **Head and neck radiology** 527
Derfel ap Dafydd

70. **Tracheostomy** 540
Jack Faulkner, Aleix Rovira, and Asit Arora

71. **Phonosurgery** 547
Kate Heathcote

72. **Movement disorders of the larynx** 554
Julia Selby

SECTION 5
Paediatrics

73. **Assessment of the neonate, infant, child and adolescent** 561
Hasnaa Ismail-Koch and Andrea Burgess

74. **Acute airway management and paediatric stridor** 570
Sean E. Evans and Sanjay R. Parikh

75. **Snoring and obstructive sleep apnoea** 580
Kate A. Stephenson and Michael Kuo

76. **Sore throat in paediatric patients** 588
Alex Millington and Nico Jonas

77. **Rhinorrhoea and epistaxis** 596
Sujata De and Ravi Sharma

78. **Blocked nose** 608
Neil K. Chadha

79. **Paediatric dysphonia** 613
Eaodaoin O'Cathain and Scott Rickert

80. **Paediatric dysphagia** 624
Michelle Wyatt and Vicky Thorpe

81. **Paediatric neck masses, cysts, and sinuses** 630
Haytham Kubba and Tash Kunanandam

82. **Paediatric drooling and salivary glands** 638
Anna Harrison and Neil Bateman

83. **Paediatric hearing loss** 643
Jaya Nichani, Adam Walker, and Iain A. Bruce

84. **The painful and discharging ear in children** 650
Ricky Pal and Mark Felton

85. **Dizziness in children** 659
Alok Sharma and Gundula Thiel

86. **Acquired paediatric airway stenoses and congenital laryngotracheal disorders** 665
Ben Hartley and Claire Frauenfelder

87. **Tonsillectomy and adenoidectomy** 675
Keith G. Trimble

88. **Otitis media with effusion** 681
Mat Daniel

89. **Cleft and craniofacial abnormalities** 685
Louisa Ferguson

90. **The syndromal child** 691
Ravi Thevasagayam and Sanjeev Gupta

Index 705

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Abbreviations

4D	four-dimensional
5FU	5-fluorouracil
AAR	anterior active rhinomanometry
ABG	air–bone gap
ABI	auditory brainstem implant
ABR	auditory brainstem response
ABRS	acute bacterial rhinosinusitis
ACHA	air conduction hearing aid
AGMP	aerosol-generating medical procedure
AHI	apnoea–hypopnoea index
AICA	anterior inferior cerebellar artery
AJCC	American Joint Committee on Cancer
ALT	anterolateral thigh
AMEI	active middle ear implant
ANCA	antineutrophil cytoplasmic antibodies
AOM	acute otitis media
AP	action potential *or* anteroposterior
AR	allergic rhinitis
ARIA	Allergic Rhinitis and its Impact on Asthma
ARS	acute rhinosinusitis
AVM	arteriovenous malformation
BCHD	bone conduction hearing device
BMI	body mass index
BnNT	botulinum neurotoxin
BOR	branchio-oto-renal
BPPV	benign paroxysmal positional vertigo
BPVC	benign paroxysmal vertigo of childhood
BTE	behind the ear
CAEP	cortical audiological evoked potential
c-ANCA	cytoplasmic antineutrophil cytoplasmic antibodies
CBT	cognitive behaviour therapy
CCAD	central compartment atopic disease
CCCR	calcium creatinine clearance ratio
CHAOS	congenital high airway obstruction syndrome
CHARGE	coloboma, heart defects, atresia choanae, retardation of growth, genito-renal defects, ear anomalies/deafness
CHL	conductive hearing loss
CI	cochlear implant *or* confidence interval
CL±P	cleft lip and/or palate
CLB	Coles, Lutman, and Buffin
CLIA	chemiluminescent immunoassay
CN	cranial nerve
CO_2	carbon dioxide
CoV	coronavirus
COVID-19	coronavirus disease 2019
CRC	chronic refractory cough
CRS	chronic rhinosinusitis
CRSsNP	chronic rhinosinusitis without nasal polyps
CRSwNP	chronic rhinosinusitis with nasal polyps
CRT	chemoradiotherapy
CSF	cerebrospinal fluid
CT	computed tomography
CTV	clinical target volume
cVEMP	cervical vestibular evoked myogenic potential
CVST	cerebral venous sinus thrombosis
CWD	canal wall down
CWU	canal wall up
dB	decibel
DCR	dacryocystorhinostomy
DGMP	droplet-generating medical procedure
DISE	drug-induced sleep endoscopy
DNS	deviated nasal septum
DoLS	Deprivation of Liberty Safeguards
DVT	deep vein thrombosis
DWI	diffusion-weighted imaging
DXA	dual-energy X-ray absorptiometry
EAC	external auditory canal
EBRT	external beam radiotherapy
EBV	Epstein–Barr virus
ECochG	electrocochleography
EGPA	eosinophilic granulomatosis with polyangiitis
ELISA	enzyme-linked immunosorbent assay
EMG	electromyography
ENoG	electroneuronography
ENT	ear, nose, and throat
ENV	external nasal valve
EORD	extra-oesophageal reflux disease
EPOS2020	European Position Paper on Rhinosinusitis and Nasal Polyps 2020
ESR	erythrocyte sedimentation rate
ESS	endoscopic sinus surgery
ET	Eustachian tube
ETD	Eustachian tube dysfunction
EXIT	*ex utero* intrapartum treatment
FDG	fluorodeoxyglucose
FEES	fibreoptic endoscopic evaluation of swallow
FHH	familial hypocalciuric hypercalcaemia
FMT	floating mass transducer
FNAC	fine needle aspiration cytology
FRAFF	free radial forearm flap
FTSG	full-thickness skin graft

GA	general anaesthetic
GABHS	group A beta-haemolytic *Streptococcus*
GEN	gaze-evoked nystagmus
GORD	gastro-oesophageal reflux disease
GPA	granulomatosis with polyangiitis
GTV	gross tumour volume
H&N	head and neck
HA	hyaluronic acid
HB	House–Brackmann
HCW	healthcare worker
HHT	hereditary haemorrhagic telangiectasia
HIT	head impulse test
HIV	human immunodeficiency virus
HL	hearing loss
HNC	head and neck cancer
HPTH	hyperparathyroidism
HPV	human papillomavirus
HSV	herpes simplex virus
HTL	hearing threshold level
HZO	herpes zoster oticus
IAC	internal auditory canal
ICA	internal carotid artery
ICHD	International Classification of Headache Disorders
Ig	immunoglobulin
ILO	inducible laryngeal obstruction
IMCA	independent mental capacity advocate
IMRT	intensity-modulated radiotherapy
INV	internal nasal valve
IV	intravenous
JNA	juvenile nasopharyngeal angiofibroma
KO	keratosis obturans
LA	local anaesthetic
LD	laryngeal dystonia
LFIA	lateral flow immunoassay
LPR	laryngopharyngeal reflux
LTR	laryngotracheal reconstruction
MCA	Mental Capacity Act
MD	Ménière's disease
MDT	multidisciplinary team
MEN	multiple endocrine neoplasia
MERS	Middle East respiratory syndrome
MIP	minimally invasive parathyroidectomy
MIVAP	minimally invasive video-assisted parathyroidectomy
MRI	magnetic resonance imaging
MTD	muscle tension dysphonia
NAR	nasal airway resistance
NARES	non-allergic rhinitis with eosinophilia syndrome
NF	neurofibromatosis
NHS	National Health Service
NICE	National Institute for Health and Care Excellence
NIHL	noise-induced hearing loss
NOE	necrotizing otitis externa
NPA	nasopharyngeal airway
NPC	nasopharyngeal cancer
OA	oesophageal atresia
OAE	otoacoustic emission
OAR	organ at risk
OB	olfactory bulb
OE	otitis externa
OI	osteogenesis imperfecta
OME	otitis media with effusion
OPSCC	oropharyngeal squamous cell cancer
ORN	osteoradionecrosis
OS	overall survival
OSA	obstructive sleep apnoea
OSCC	oral squamous cell carcinoma
oSDB	obstructive sleep-disordered breathing
OSN	olfactory sensory neuron
oVEMP	ocular vestibular evoked myogenic potential
p-ANCA	perinuclear antineutrophil cytoplasmic antibodies
PCD	primary ciliary dyskinesia
PDA	patent ductus arteriosus
PEES	flexible endoscopic evaluation of swallowing
PET	positron emission tomography
PFAPA	periodic fever, aphthous stomatitis, pharyngitis, and adenitis
PFS	progression-free survival
PHPTH	primary hyperparathyroidism
PIFT	persistent idiopathic facial pain
PMD	potentially malignant disorder
PMMC	pectoralis major myocutaneous
PND	postnasal drip
PNIF	peak nasal inspiratory flow
PPE	personal protective equipment
PRS	Pierre Robin sequence
PSG	polysomnography
PTH	parathyroid hormone
PVOD	postviral olfactory dysfunction
RAS	recurrent aphthous stomatitis
RAST	radioallergosorbent test
RCT	randomized controlled trial
RLN	recurrent laryngeal nerve
RMM	rhinomanometry
RRP	recurrent respiratory papillomatosis
RT	radiotherapy
SARS-CoV-2	severe acute respiratory syndrome coronavirus 2
SCC	semicircular canal *or* squamous cell carcinoma
SCM	sternocleidomastoid
SIGN	Scottish Intercollegiate Guidelines Network
SLE	systemic lupus erythematosus
SLT	speech and language therapist/therapy
SMAS	superficial musculoaponeurotic system
SNAPS	specialized neonatal and paediatric surgeons
SNHL	sensorineural hearing loss
SNOT	Sino-Nasal Outcome Test
SNUC	sinonasal undifferentiated carcinoma
SP	summating potential
SPL	sound pressure level
SRBD	sleep-related breathing disorder
SSCD	superior semicircular canal dehiscence
SVR	surgical voice restoration
TM	tympanic membrane
TMJ	temporomandibular joint
TNO	transnasal oesophagoscopy
TOF	tracheo-oesophageal fistula
TOP	tracheo-oesophageal puncture

TORP	total ossicular replacement prosthesis
TORS	transoral robotic surgery
TSH	thyroid-stimulating hormone
UACS	upper airway cough syndrome
UPMC	University of Pittsburgh Medical Center
US	ultrasound
VAS	Visual Analogue Scale
VEMP	vestibular evoked myogenic potentials
VFSS	videofluoroscopic swallow study
vHIT	video head impulse test
VM	vestibular migraine
VNG	videonystagmography
VOG	video-oculography
VOR	vestibulo-ocular reflex
VPI	velopharyngeal insufficiency
VRA	visual reinforcement audiometry
VSD	ventricular septal defect
VT	ventilation tube
WHO	World Health Organization
WVA	wide vestibular aqueduct

SECTION 1

Introduction

1. **Being a good doctor** *3*
   Peter Chapman
2. **Rights of the patient** *7*
   Martyn Patel
3. **Anaesthesia for otolaryngological surgery** *12*
   Simon Fletcher
4. **Practising otolaryngology in the COVID era** *19*
   Andrew Thamboo

1

Being a good doctor

Peter Chapman

Introduction

It would be an omission to write this chapter without reference to the Hippocratic oath. Although of doubtful provenance in that it may be inaccurate to attribute it to Hippocrates himself, and despite being out of date in some respects, it does contain some core principles (e.g. non-maleficence and confidentiality) that remain very relevant to practitioners in the twenty-first century. A form of it is taken by new medical graduates in many countries and in the UK some form of oath continues to be used by 86% of institutions.[1]

The General Medical Council, the regulatory body that provides doctors with their registration and licence to practise in the UK, articulates the principles of what it means to be a good doctor in its publication 'Good Medical Practice'.[2] These principles are:

- make the care of your patient your first concern
- be competent and keep your professional knowledge and skills up to date
- take prompt action if you think patient safety is being compromised
- establish and maintain good partnerships with your colleagues and colleagues
- maintain trust in you and the profession by being open, honest, and acting with integrity.

It would also be an omission to write this chapter without reference to the COVID-19 pandemic which at the time of writing is at its peak in the UK and which has affected so many people, including healthcare workers around the world. Because of the oropharyngeal nature of the speciality, ear, nose, and throat (ENT) surgeons have been particularly at risk. Doctors are now faced with personal risks to their own health that have not been of such concern since the discovery of antibiotics or outside of conflict, and they may also now be required to practise in a clinical field which may be unfamiliar to them. In such a time, the core principles of what it means to be a good doctor are even more important.

Appraisal and revalidation

In the UK, following a series of high-profile national healthcare scandals and inquiries in the 1990s and 2000s culminating in the Shipman Inquiry, the need for a mandatory system through which a doctor's competence and ability to practise would be regularly reaffirmed was recognized by the public, government, the NHS, and the profession. Following widespread consultation with stakeholders, legislation was passed by parliament in 2010 for the current system of revalidation based upon annual professional appraisals over a 5-year cycle, and through which all doctors must demonstrate that the principles and values contained within 'Good Medical Practice' have been met.

The requirements of 'Good Medical Practice' are divided into four domains that describe what this means in the UK (**Table 1.1**).

Understanding roles and responsibilities

Every doctor must understand and be clear about the scope of their own practice. Different doctors even in the same speciality may have varied and sometime quite different roles. A doctor who works for a pharmaceutical company engaged in clinical research, while also working a day per week as an assistant general practitioner, will have a very different practice to an ENT surgeon in a district general hospital. The fundamental characteristics of a good doctor will be the same for each, but the specific evidence that may be required to demonstrate this will be different.

To take the example of the district general hospital ENT surgeon, they will have their clinical patient-facing role, perhaps with a subspecialty interest, both at their district general hospital but also perhaps at the local private hospital (where it is important that their practice should mirror that of their NHS post); they are likely to have a teaching role even if they are not formally involved in the training of junior doctors or of undergraduates; and they may be involved in some research so have a research role. They will necessarily be working in a managed environment and so have a responsibility for the proper use of resources and for complying with local as well as national guidance and policies. They will also have a clinical managerial role themselves even though they may not occupy a specific clinical managerial post with time allocated for it. All these roles need to be understood and accounted for during appraisal and the principles of 'Good Medical Practice' applied to each. A doctor's licence to practise is applicable to their practice within the declared scope, and not beyond.

Table 1.1 'Good Medical Practice' domains

Domain 1. Knowledge, skills, and development	Provide a good standard of practice and care, and work within your competence	Keep your knowledge and skills up to date	Record your work clearly, accurately, and legibly		
Domain 2. Patients, partnership, and communication	Respect every patient's dignity and treat them as an individual	Listen to patients and work in partnership with them, supporting them to make informed decisions about their care	Protect patients' personal information from improper disclosure		
Domain 3. Colleagues, culture, and safety	Work with colleagues in ways that best serve the interests of patients, being willing to lead or follow as circumstances require	Be willing to share your knowledge, skills, and experience with colleagues, whether informally or through teaching, training, mentoring, or coaching	Treat people with respect and help to create a working and training environment that is compassionate, supportive, and fair, where everyone feels safe to ask questions, talk about errors, and raise concerns	Act promptly if you think that patient safety or dignity may be seriously compromised	Take care of your own health and well-being needs, recognizing and taking appropriate action if you may not be fit to work
Domain 4. Trust and professionalism	Act with honesty and integrity, and be open if things go wrong	Protect and promote the health of patients and the public	Never unfairly discriminate against patients or colleagues		Never abuse patients' trust in you or the public's trust in your profession

Source data from https://www.gmc-uk.org/professional-standards/the-professional-standards/good-medical-practice

The importance of evaluation, reflection, and then action

A number of common themes run through all domains of 'Good Medical Practice' and the need for reflection and then taking action is one such theme. Doctors must be honest and even-handed about their own reflection. A good doctor will try and see the other point of view and then evaluate it with an open mind. The good doctor must also be realistic about what may be achieved in terms of actions in a particular timescale. Where patient safety is involved, an action may need to be immediate (e.g. the withdrawal of a particular service if it is deemed to be unsafe), whereas the development of a new service may be broken down into a series of achievable steps over a longer timescale.

The 'SMART' acronym is useful to consider and describe actions which are planned after the evaluation and reflection of an event or need, whether that be, for example, an educational meeting, a clinical audit, or a review of complaints made against the doctor. Where changes need to be made, they should be described as Specific, Measurable, Achievable, Relevant, and Time-bound. Having set such an objective based upon a perceived need, the doctor should achieve this within the set timescale unless there are identifiable reasons why this is not possible.

Keeping up to date

Doctors must comply with and therefore keep up to date with the law, and with local and national guidance and policies as they relate to all aspects of their work.

In the UK, the medical Royal Colleges provide and signpost educational resources to enable a doctor's individual practice to be kept up to date. The varied scope of an individual doctor's professional life makes it likely that the advice and requirements of a number of different institutions will need to be followed since, as previously discussed, that doctor must ensure that their educational activity covers the whole scope of their practice. Employers will provide clarity of local requirements for mandatory training, and compliance with these is also necessary. A doctor may of course have more than one employer.

It is important to recognize that clinical techniques and treatments will change sometimes quite markedly over an individual doctor's working life. An example would be the development of laparoscopic rather than open abdominal surgery. A doctor's practice must therefore change in order to provide patients with the safest and most effective up-to-date and evidence-based care. Doctors need to be prepared to learn new techniques and to be honest through reflection and discussion with peers as to their own competence in these new techniques before they treat patients on their own. Doctors must recognize their own limitations and cooperate with initiatives to introduce new techniques, such as initially only within a limited group of trained practitioners. Doctors will need to work in collaboration with others, submit their results to local and national audit and registries, and be open to feedback both directly and also through resources such as 360-feedback exercises.

Review and audit of practice

Within a publicly funded national health system such as the NHS, a patient should expect to receive the same high standard of medical care wherever they live and whichever institution they attend. Although hospitals are large and complex organizations, health systems across the world collect information to try and quantify the quality and cost-effectiveness of patient care in them.

This enables comparison of different institutions across an array of different metrics, such as mortality, length of stay, and use of resources. The Care Quality Commission in the UK incorporates these indicators into its regular reporting which rates hospitals against the domains of whether they are safe, effective, caring, responsive to the needs of patients and the local population, and whether they have effective leadership.[3]

Outcomes are also recorded and published at a specialty level and, where there is sufficient data, for individual doctors. The longevity and failure rates for joint arthroplasty and cardiac surgery outcomes are measured, and histopathologists participate in national benchmarking in relation to error rates.

National and local audits record compliance against recommended or commissioned pathways of care. The 'Getting It Right First Time' initiative from NHS England,[4] based upon an exercise conducted originally in trauma and orthopaedics specialties, seeks to review and compare specialty practice in all hospitals nationally against established evidence and best practices. Where needed, changes are required and supported through a regional support network.

Doctors have an obligation to both participate fully in these initiatives, to contribute to them, and to constructively address the outcomes as they pertain to them both individually and collaboratively within their team and in all of the institutions in which they work.

All doctors will at times receive a complaint, a legal claim against them, or be the subject of a significant incident report relating to their practice. A doctor must cooperate fully and openly with any enquiries that result and be willing to learn from them and actively engage with any changes or restrictions to their practice that may result. Mistakes should be acknowledged. Where a patient has experienced harm, whether or not a mistake has occurred, the doctor should tell the patient, apologize, offer an appropriate remedy, and explain fully any short- or long-term effect. Where the doctor is the consultant responsible for the action of a team, this remains their responsibility though the particular error may not have been theirs personally. In the UK this responsibility is called 'duty of candour'.

Doctors must therefore be open and honest with colleagues, employers, and relevant organizations and take part in reviews as requested. They must raise concerns where appropriate and encourage others to be open and honest as well so as not to stop others from raising their own concerns.[5]

Whereas a doctor will have a duty to make the care of their patient their first concern, doctors also have a duty to comply with processes which ensure the optimum usage of resources across the whole health economy. This may on occasions mean that a particular treatment that the doctor would recommend is not available where they work, sometimes for financial reasons. Whereas the doctor should act as the patient's advocate by petitioning for the availability of evidence-based treatment, they should ultimately comply with the locally agreed guidance and requirements where this cannot be provided. As the pace and cost of new drug developments and technological advances increase and further outstrip the budgets of all health systems, this will be a dilemma that doctors will face more and more.

Working with colleagues and patients

A doctor must have a collaborative relationship with a patient. The assumption that 'the doctor knows best' is no longer appropriate. Consultations need to be a discussion and a patient's view should be actively sought. Options with their consequences rather than just didactic opinions should be laid out and explained in a way consistent with the patient's understanding. The doctor can guide the patient in their decision but should not make the decision for them. The mental capacity of all patients must be understood and accounted for during all consent for treatment discussions. It is the patient who bears the consequences of the decision that is taken.

All doctors will work in teams and in order that the team functions well, all members must respect each other and be able to communicate well. Hierarchies should be abandoned—for instance, in the World Health Organization theatre checklist[6] team brief, the introduction of all members at the start of the operation session should serve to emphasize that everyone there, from the most junior medical student on their first attachment in theatres to the senior professor, should feel able to speak up if they have any concerns.

The multidisciplinary team is another area where a well-functioning team without hierarchies is essential to patient safety. It is vital that all members feel able to contribute and question and to have their contribution valued.

In every team, all members but especially senior ones have a particular and important role in setting an example and ensuring that the team functions well. The Association of Surgeons in Training survey into bullying and harassment in the workplace[7] demonstrated an unacceptable level of this in the workplace in the recent past and this undoubtedly continues in many areas still. Where teams do not function well then intervention is needed in the form of facilitated discussion, and perhaps individual and team profiling to understand better the interactions and personalities that are present. Doctors must cooperate with these initiatives.

Surveys can be helpful and doctors in the UK must for their revalidation participate in one 360 feedback of colleagues and one of patients in every 5-year revalidation cycle. For these to be useful though the doctor should be actively seeking constructive criticism and feedback rather than a reaffirmation of their ability. So, participants should be chosen wisely and from all different areas of a doctor's work, and patients should be surveyed randomly and without the doctor's knowledge. The doctor then needs to reflect properly on what has been said and commit to changes if required.

Probity

> You must make sure that your conduct justifies your patients' trust in you and the public's trust in the profession.[8]

The requirement for doctors to observe the highest standards of probity extends beyond the professional life of clinical work, research, teaching, and management and into a doctor's personal life of financial dealings, relationships, media communication, and interactions with the law.

Honesty is the cornerstone upon which a doctor's probity is built. Complete honesty is required in all financial dealings, in a doctor's curriculum vitae, and in all communications including, for instance, the writing of reports and the advertising of services. All claims must be factual, relevant, and able to be checked. Patient confidentiality should never be breached. Social media represents a particular risk

and doctors should remember that comments intended to be private may not stay so.

Doctors, as decision-makers where the purchasing of expensive medicines and equipment is involved, need to ensure that any potential conflict is properly understood and declared. A doctor may need to clearly exclude themselves from such a decision. Great care must therefore be taken when accepting invitations from companies. The distinction between the offer of training on a particular piece of equipment or technique necessary for the safe treatment of patients, and hospitality on its own must be clearly understood. The former may be reasonable, the latter not so.

The vulnerability of a patient must never be exploited whether this be in relation to pursuing a relationship with a patient or in relation to a doctor's own beliefs. Discussions of religious, political, or moral beliefs therefore have no place in a consultation.

Treatment must be based solely on clinical judgement and assessment of effectiveness and must be independent of any subjective view about a patient's lifestyle or beliefs.

Looking after yourself and others

Doctors do have a relatively high incidence of significant mental health illness related to their work.[9] At the time of writing, in relation to the COVID-19 pandemic, this is very much in the headlines, but it is true at other times as well. Such a health problem can develop with triggers that may not be obvious even after years of experience and practice. Some may describe it as 'burnout'. A doctor's capability may be affected by physical ailments as well as mental health problems. Experiencing illness whether it be mental or physical and being open about it is not an admission of weakness or failure—often it is quite the converse. It should be managed supportively, properly, and sensitively by a good employer.

It is true though that doctors may not seek the help they need as early as they should. Unfortunately, this can have significant consequences for their own health but also for their ability to practise and therefore to treat their patients effectively and safely.

Increased stress may lead to an increased use of alcohol and sometimes drug dependence, further damaging a doctor's own health and risking that of their patients.

A doctor experiencing illness that affects or may affect their ability to practise has a clear professional responsibility to seek help through their general practitioner or through their employer's occupational health service at the earliest opportunity, and must be open and honest with their regulator and employer.

The annual appraisal requires a doctor to make a statement in relation to their health and whether it may impinge upon their ability to care properly for their patients.

Summary

Despite differences in practice and experience across different generations, countries, and specialties, the fundamentals of what it means to be a good doctor have remained broadly the same.

These requirements are well articulated in the UK in the General Medical Council's guide 'Good Medical Practice'. In order to maintain a licence to practise (and therefore to continue to practise within the law), all UK doctors are required to demonstrate these requirements have been met every 5 years. Similar mechanisms exist in other countries to ensure that all practising doctors maintain their knowledge and skills, practise with regard to safety and quality, work well within their teams, communicate effectively, and maintain patients' and the public's trust in themselves and the profession.

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2

Rights of the patient

Martyn Patel

Introduction

As a geriatrician working in a large UK-situated university teaching hospital, I am often asked to consult on patients admitted primarily under the care of surgical teams. What follows is a distillation of the practical advice I have often meted out with specific regard to the rights of the patient. This chapter will focus on the law as applied to adults. For information on consent and safeguarding with respect to children please see Chapter 73.

Through each section is threaded a (hopefully) realistic series of scenarios that demonstrate how a clinician should respect the rights of the patient, and how those relate to the current legal landscape. The legal frameworks explicitly referred to are those that apply in England and Wales, and although other UK regions and countries may have broadly similar approaches, please check the details of your own national frameworks.

The Mental Capacity Act

The Mental Capacity Act (MCA) was enshrined in law in 2005[1,2] and established five key principles for clinicians to follow (**Box 2.1**) which will be drawn out in the following scenarios.

Box 2.1 The five core principles of the Mental Capacity Act: 'PLUMB'

1. **P**resumption of capacity—most people can make some decisions.
2. **L**east Restrictive—consider other ways to promote rights and freedom.
3. **U**nwise decisions—remember, unwise is not the same as unable.
4. **M**aximize capacity—consider time, pictures, and language and make adjustments as needed.
5. **B**est interests—if an individual lacks capacity, any decision made/actions taken must be made in their best interest.

Source data from Mental Capacity Act 2005, https://www.legislation.gov.uk/ukpga/2005/9/contents

Assessing mental capacity

CASE 1

John is 25 and has sustained a nasal injury and pronounced epistaxis playing rugby. Despite basic attempts to stem the bleeding in the emergency department, his situation has not settled overnight. You decide the next step in his management is operative, and discuss the pros and cons of doing so with John. He seems in clear agreement with this, asking only a few relevant questions about postoperative care. You ask him a few brief questions to check he has registered exactly what the procedure entails, which he does, and you proceed to fill out a consent form with him which he signs.

This is as easy as it gets. Don't worry, if you didn't notice, but you did assess John's mental capacity, or at least the first part of what is known as the 'two-stage' assessment (**Box 2.2**). Stage one is being alert to the possibility that someone may lack capacity, and recognizing that that trigger should lead you to stage two—assessing capacity. In this case, as in the majority of doctor–patient interactions, there was no stage one 'trigger' and therefore no explicit need to document or comment on it. We should start with a default assumption that everyone has mental capacity, until such time that a concern presents itself (one of the five key principles of the MCA—see **Box 2.1**). In Case 1, there was no triggering concern, and the conversation around the consent form contained an appropriate level of 'check and challenge' to ensure this, before signing the consent form. If you are reassured that there is no triggering concern, there would be no need to make a separate formal statement about capacity.

What constitutes a concern? It can take different forms. It could be a low or fluctuating conscious level, a diagnosis of historically impaired cognition, a current acute cognitive processing problem (typically delirium), or a patient that makes an unorthodox or unwise choice when presented with medical facts (but remember principle 3—unwise is not the same as unable, it is just a trigger to do the formal capacity assessment). So, while a triggering concern alone

Box 2.2 The two stages of mental capacity assessment

Stage one

Does your patient have a chronic impairment of, or acute disturbance in, the functioning of the mind or brain? Examples include a previous stroke, known dementia, acute delirium, significant learning disability, mental disorder, intoxication, etc.

If the condition is temporary or fluctuating, can the decision be put off until the person regains mental capacity?

Stage two

Consider: is your patient's impairment or disturbance sufficient that they lack the capacity to make the particular decision?

Can the person:

1. *Understand* information about the decision that is to be made?
2. *Retain* enough of that information; *recall* in sufficient detail what was discussed?
3. *Weigh* up the information as a part of the process of making the decision (the quoted risks and benefits)?
4. *Communicate* their decision using any means (whether by talking, using sign language, writing boards, or any other means)?

To *have capacity*, the person must be able to do all of the four components.

To *lack capacity*, the person must be deficient in any one (or more) of the four components.

Source data from Mental Capacity Act 2005, https://www.legislation.gov.uk/ukpga/2005/9/contents

does not directly imply the patient lacks capacity, it does mean a more formal assessment is required to ascertain the patient's capacity status—stage two of the MCA process.

CASE 2

Peter is 40. He has been involved in a road traffic accident, and has sustained many injuries, including nasal trauma and severe epistaxis. It is clear that he requires urgent surgical care for this and other injuries. At present he is intubated and sedated as on arrival he had a low conscious level and was not protecting his airway (Glasgow Coma Scale score of less than 8). Thankfully an urgent computed tomography scan has cleared any intracranial damage.

You wish to proceed to an operative solution to his bleeding. In your expert opinion there is not time to extubate and wait to see if Peter would regain a sufficient level of consciousness to discuss his care. You judge the treatment you would offer to be life-saving. Peter's wife, Jo, is available on the telephone; you explain the situation, including the fact Peter is currently unconscious, and the risks and benefits of the operation you propose, and stress the urgency of the timescale. Jo acknowledges what you say to her and asks you to do all you can; in her opinion Peter would wish any and all interventional care you can offer, and agrees this course of action is in his best interests. You document the conversation in the notes (and in the UK also complete a Consent Form 4—Consent for an Adult Patient Lacking Capacity), and make the necessary arrangements to proceed.

Case 2 draws out some more of the key features of mental capacity law. The unconscious patient always lacks capacity—they cannot communicate (equally they cannot understand, retain, or weigh up any information!). The two key principles engaged here therefore are principle 4—maximize capacity and principle 5—best interests (Box 2.1). Firstly, would it be reasonable to wait until Peter regains a higher level of consciousness and ask him about the proposed treatment? The answer to this depends on whether the treatment you propose is time critical and of sufficient importance that to delay incurs a risk to life or increases the risk of serious morbidity. Secondly, there is any reason to suppose that the patient would not agree that what you propose is in their best interests? Answering the first question is reliant upon your expert judgement. Answering the second is reliant upon what other sources of information are available. In this case, a clearly defined friend or relative who knew Peter well was available, and could reflect what they felt his wishes would be, and confirm that based on their knowledge of his past opinions and expressed wishes that your proposed treatment was in his best interests.

If we alter the variables, let us say the bleeding was less intense and a trial of conservative management was an option, then you may reasonably elect to choose to follow that more conservative route, particularly to allow Peter the chance to regain consciousness, and then make his own decisions about conservative versus interventional approach. Again, this would be down to your expert judgement on the risks of delaying the more interventional treatment.

CASE 3: PART 1

Andrea is 78 and has fallen in her care home, sustaining a nasal trauma and severe epistaxis. After a period of conservative management she is still bleeding profusely and you feel an operative approach is now warranted. Your hospital has a policy of screening for delirium utilizing the 4AT tool, on which Andrea has scored highly, indicating a high likelihood of the presence of delirium. Andrea's notes contain a historical diagnosis of vascular dementia. You review Andrea on the ward, she appears distracted, plucking at the bedclothes and while she does initially appear to follow what you say, she looks blankly at you when you ask her if she can recall the nature of the treatment you just proposed. Furthermore, she is unable to answer even very basic questions about her current health or situation. You contact Andrea's daughter (her only living relative) and discuss the situation with her; she agrees to come talk in person. Later that day, you discuss with the daughter your proposed treatment plan, and the risks and benefits of operative care versus further conservative management. You specifically ask if Andrea had ever taken out any power of attorney forms for either health or welfare decisions, or completed an advance care plan. (She hasn't.) You fill out a Consent Form 4, which the daughter is happy to sign, as in her opinion Andrea would want the operation.

In Case 3, your patient has two 'red flags' that should trigger stage one of a mental capacity assessment. Firstly, she presents with overt delirium and secondly, she has a historical diagnosis of dementia. You sensibly chose to conduct a stage two capacity assessment and it was clear Andrea could not weigh or retain the information required to make a decision, and therefore lacked capacity based on a demonstrated lack to carry out at least two of the four components required to demonstrate possession of mental capacity (Box 2.2). You were able to discuss the proposed treatment with a person who knows Andrea well, and determine from that conversation that your proposed treatment

was in her best interests. You asked specifically about the existence of any existent legal documents pertaining to health choices—typically lasting power of attorney and advance decisions. You confirmed that the relative answered with the patient in mind, and not with their own personal choice if they were in a similar situation.

To be clear, by no means will every patient with a diagnosis of dementia lack capacity when faced with a medical decision. It is, however, a reasonable stage one trigger, to propel you to stage two and take steps to assess capacity formally and to clearly document that you have done so. Where the patient lacks capacity, and they have a registered power of attorney for health decisions, you should ask to see a copy of the power of attorney, and take a copy for the medical notes. Every attempt must be taken to consult with the attorney, before proceeding to non-time-critical treatment. If no such power of attorney exists, you should consult with a friend or relative, typically starting with the registered first contact in the person's healthcare records.

Tip: where there is no registered power of attorney, it is sensible to establish if there are other family members that would wish to be involved in making the decision when you talk with the first friend or relative that you are able to contact. If so, where possible arrange to meet them jointly. Beware, not all family will reach the same conclusion, and while you could proceed having only sounded out one opinion, it can save you a lot of problems in the future to consult as widely as possible in the first instance.

Finally, what if there was no friend or relative? Not even a neighbour who knows Andrea well. In that case, presuming time is not critical, then you would need to contract the services of an independent mental capacity advocate (IMCA). Most NHS trusts will have a process in place for arranging this, with an agreed provider of IMCA services. This provider has to be completely separate from the body that is your employer.

Tip: the key to a good IMCA service is the quality of the information made available to the IMCA at the point of referral, and sufficient contact time allowed for them to meet with both the individual and then yourself as the lead clinician to form a best interests decision.

Deprivation of Liberty Safeguards

The Deprivation of Liberty Safeguards (DoLS) arose in 2009[3] as an amendment of the MCA 2005. It is now a specific legal requirement for NHS trusts (and other healthcare and social care providers) to apply for permission to detain someone who lacks the mental capacity to make an informed decision about their ability to leave the healthcare setting. A sentinel case reviewed by the Supreme Court in 2014[4] further clarified that to trigger a DoLS request a patient must be under supervision, that supervision is continuous, controlling, and that the individual would not be allowed to leave if they were to attempt to do so.

DoLS was created as a legal framework that allows for an explicit code of good practice for institutions to ensure they are not unlawfully detaining individuals. While the intent is laudable (see 'Controversy warning'), the nature of how it is implemented in practice is under active review to streamline the process, particularly for acute NHS trusts. Case 3 follows the current structure of the implementation of the MCA since 2009, but watch this space, as developments were planned with introduction of a new system called Liberty Protection Safeguards due in October 2020,[5] although this has been delayed by the COVID-19 pandemic.[6] As of June 2025 there is still no set date for when these changes might be implemented, although a government consultation is underway.

Controversy warning

DoLS as a topic has become for some akin to a red rag to a bull. The idea that health services need permission to detain people to provide appropriate and safe ongoing care will always be an anathema to some. To fully appreciate DoLS, one has to take a step back and realize that the MCA 2005 was written to encompass all settings, and is applicable equally to healthcare providers and social care providers alike. The nagging feeling among clinicians remains, however, that all of the DoLS requests they file end with a 'No further action' rubber stamp from their DoLS authorizing authority, so is it worth the effort? My answer (and I've been asked this a lot) is that how an organization deals with the legal frameworks it is beholden to such as MCA and DoLS is widely seen as an excellent surrogate marker of the care they deliver. It is not a coincidence that the Care Quality Commission has been so hot over the last 5 years on confirming that the MCA and DoLS implementation is up to speed in the NHS trusts they inspect, in much the same way one might check the ward areas for cleanliness. Those trusts that 'get' MCA and DoLS seem to also to 'get' how to deliver high-quality care. Also, it is the law.

CASE 3: PART 2

Andrea has sailed through surgery and is now recovering on your ward. It is essential, in your professional opinion, that she has an extended period of monitoring under your team, with access to rapid further treatment should she rebleed. However, on a daily basis Andrea attempts to leave the ward, and the nursing team, having had to call security to restrain her on the ward, are now providing her with advanced observations (sometimes called 'specialling' or 1:1 care'). You note in her medical notes that while you have attempted to explain to Andrea why she must stay in hospital, unfortunately due to her dementia (stage one—identify a trigger) she is unable to retain the information relevant to making an informed discharge decision (stage two—assess all four components of capacity). You document that it is Andrea's best interests to remain detained in hospital at present, and request that a DoLS request is put in. This DoLS application is completed for you by the senior ward sister (some hospitals have a 'flying squad' to complete DoLS applications, larger trusts tend to train a cadre of senior nurses to do this, and any health professional, yourself included, can complete the application). You consider how you can make the detention less restrictive, and offer open visiting to Andrea's family, and allow them to take her off the ward to the canteen for short periods accompanied by a trained member of staff. Several days later, after what you deem a sufficient period of monitoring, you judge that Andrea no longer requires to be in hospital and her discharge is arranged.

Once you recognize that you are subjecting a patient of yours who lacks capacity for discharge decisions to supervision that is continuous and controlling, such that they would not be allowed to leave and you would take steps to prevent them from leaving, then you must consider a DoLS application. It is hopefully likely that your

employer will have taken steps to make the actioning of this need as simple as possible for you, most NHS trusts having recognized it is not the best use of consultant surgeons' time to do this themselves. Indeed, well-run ward teams often will have recognized the need for a DoLS and requested it seamlessly for you in the background. However, as the named clinician in charge, it is wise to be aware that you need to signal this need for ongoing hospital care as a justification for deprivation of liberty.

DoLS requests are made to your local council, and typically one applies for both the urgent request (covers at most the next 7 days) and standard request (when requested simultaneously covers a period from day 8 onwards, but is not indefinite). Sadly, the vast majority of local council DoLS authorization teams are unable to respond in person and formally review the patient to authorize or reject the urgent application within the first 7-day timescale. In fact, very often the patient is already discharged long before any in-person review occurs as to whether the application was reasonable—which somewhat invalidates the original intent and purpose of the law. Happily, the whole process is under review with reforms set to be introduced soon. For now, one must still be alert to the need to request DoLS orders in the correct patient scenarios.

Adult safeguarding

Safeguarding is rightly a compulsory component of NHS trust mandatory training for all staff. From a wider societal viewpoint, it is a human right to be able to live a life in safety, free of abuse or neglect. Within the health and social care arena, the Care Act 2014[7] puts the onus on health and social care professionals to act when they have reason to believe a patient (health) or client (social care) may have been subject to any category of abuse (physical, sexual, psychological, financial, and so on). While any and all adults should have their right to live free from abuse or neglect safeguarded, particular attention is paid with respect to those patients that are termed 'vulnerable adults'. Namely, someone over the age of 18 who due to mental or other disability, age, or illness is unable to take care of themselves, or to protect themselves from abuse. There is a very significant overlap between adults who lack capacity for complex healthcare choices and adults at risk of abuse.

CASE 4

You have arranged an elective admission for Mike from his care home for a minor operative procedure. At preoperative checks Mike, who has vascular dementia, was found by your assessment to lack capacity for the decision to operate in his case, but you were able to converse with his wife Jane, who agreed the procedure was in his best interests, and hence Mike has been booked to come in. As part of the admission, a routine nursing check of skin integrity has found that Mike has a significant sacral sore, of at least grade 2, and possibly higher. This was not recorded in his hospital medical notes, and on contacting his care home, the nurse on duty is unaware of this sore. Mike is unaware of the sore and unable to provide you with any information. He does not retain information about pressure care and the risk of pressure sores to health, and you judge Mike as lacking capacity for a safe discharge decision. Your nursing team file a 'vulnerable adult at risk of abuse' notice or 'safeguarding referral' to the local multi-agency safeguarding hub. You judge it safe to proceed with the procedure, which goes well. You would at this stage normally discharge back to usual residence, but you are aware that with an unresolved 'safeguarding referral' you must not discharge a person lacking capacity back to their own existing care arrangement until that referral is resolved. You inform Jane of the reason for the delay in discharge—to ensure that Mike's care home can demonstrate they can safely look after Mike. A social worker is allocated to investigate the case, and on contacting the care home, and speaking to the manager, discovers that this is actually a known pressure sore, with a community tissue viability care plan and appropriate pressure care in place. The social worker closes the safeguarding, and Mike is discharged.

Noting of a 'new' or 'worsening' grade 2 or higher pressure sore is probably one of the commonest reason for a 'safeguarding referral' in the NHS.[8] Not all cases end as happily as Mike's, and the accumulation of safeguarding referrals and concerns by a care home can lead to a suspension of new admissions or even an outright closure at the most extreme end of sanction.

Tip: if an adult lacking capacity has an unresolved safeguarding referral you would not discharge them back to their previous care situation until either the safeguarding referral is resolved, or the case manager for the referral gives explicit written permission to do so.

Ethical approaches to medicine

There are many different ethical approaches to medicine, of which the 'Four principles of Bioethics' approach and Aristolean virtue ethics are probably the two I have most time for when browsing scholarly articles. However, a better question to ask me is, 'What is the approach that you teach your trainees?' The answer to that is what is sometimes known in geriatric medicine circles as the 'grandmother rule'. Put simply, if at the end of every working day, you can examine your conscience and say you have given all your patients the care you would wish your grandmother to receive, then you have probably done a pretty good job, and one that is 'ethical' to boot. This, of course, does assume you love your grandmother … but if one takes that as read, it forms a pretty good ethical safety check.

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3

Anaesthesia for otolaryngological surgery

Simon Fletcher

Introduction

This is not an anaesthetic textbook and thus the emphasis of this chapter will be to provide an overview of the relevant concerns and considerations when delivering perioperative anaesthetic care to the patient groups considered within. Thus, I will not be describing the anaesthetic options for a comprehensive list of surgical procedures—all but irrelevant, uninteresting, and repetitive. While it is essential within surgical practice for all professionals to cooperate for the mutual benefit of the patients, this is never more the case where the airway, if not directly shared, is usually not immediately accessible. I have always promoted a symbiotic relationship, understanding that there will be times when I need skilled help to get me out of trouble, and equally that surgery can be a very lonely business somewhat alleviated by appropriate interest, sympathetic support, and shared decision-making. Ear, nose, and throat (ENT) surgery is undertaken on neonates and, increasingly, elderly patients with complex comorbidities. The vast majority of this surgery is not urgent and thus allows adequate assessment and planning. Airway obstruction may be anticipated but often presents without warning as a consequence of an acute, or progression of a chronic, disease process. It is recognized that airway management is often suboptimal in these situations.

I thus plan to discuss the management of the airway in some detail before addressing other areas of mutual interest.

The airway

Anaesthetists are airway fixated and rightly so. The 'can't intubate, can't ventilate' scenario is a nightmare best avoided with a bad outcome for all, accounting for 25% of anaesthetic-related deaths.[1] Airway difficulties occur in all areas of anaesthetic practice but are most prevalent within ENT practice. This is the case at induction, intraoperatively, and at emergence through an extended recovery period. The vast majority of cases, though, are completed without difficulty. Surgery for the highest risk groups (head and neck, neonatal, and paediatric) has been largely cohorted over the last 20 years. Experience and planning should minimize, but will never eliminate, risk. This is not the case for the acute presentation. Traumatic airway compromise, foreign body obstruction, acute airway closure through inflammation, infection, or tumour present to all acute hospitals at any time of the day or night. The Royal College of Anaesthetists and the Difficult Airway Society conducted a year-long, comprehensive, UK-wide audit[2] that investigated major airway complications: 39% occurred in an 'acute or chronic disease process involving the head, neck or trachea'. Chapter XX discusses in detail these cases. In summary, of the 70 reported cases deemed relevant, ten were within critical care and tracheostomy-related problems predominated. Failure to intubate in a deteriorating patient post tumour resection was also reported. Fifteen were emergent presentations and included trauma, obstructing haemorrhage, tumour, anaphylaxis, and epiglottitis; five of these were in the emergency department and ten in an operating theatre. Forty-five of the reported cases occurred during the perioperative period; twenty-four at induction, eight during the surgical procedure, and thirteen postoperatively of which eight were in recovery.

It is acknowledged early in this report that evidence gained from retrospective case reports is not considered of academic high validity. However, in the real world, comprehensive analysis of these cases identified a number of recurring themes. Outside of medicine, critical incident reporting is deemed an invaluable part of safety and risk management. So, what do we learn through peer-reviewed analysis of the above? Issues fall into three main areas: planning, skills, and human factors.

It is not possible to directly plan for an acutely compromised airway, but it is now embedded in anaesthetic training and practice to rehearse relevant scenarios using simulation. This must be multi-professional and should then help to develop and maintain technical skills and, importantly, reinforce team working and specifically consider and manage human factors. Repeated attempts to intubate with progressive loss of the ability to oxygenate is cited in a significant number of the cases mentioned above. All are aware of this and will be able to vocalize a stepwise escalation plan, often lost in the intensity of reality.[3] An array of difficult airway devices will be tried and discarded, many just variations on a theme. Fibreoptic intubation may be attempted (perhaps the first time a particular device has been in a set of hands) in an increasingly blue patient; the outcome is now inevitable. Through simulation individuals will rapidly discover which aides they are most comfortable with and use these

exclusively. The rest of the team, including the ENT specialists, can and should manage the human factors, allowing a rapid switch to plan B, C, and D—or perhaps straight to 'D' if required.

It is possible, though, to plan for an elective procedure and the processes are well embedded in centres conducting head and neck procedures. Most cases will not involve significant airway difficulty and the majority (but not all) of the rest can be predicted. Symptomatic stridor is an obvious feature of a closing airway, but patients often significantly compensate for a slowly progressive process. The multidisciplinary team meeting(s) should include a formal, joint assessment of the airway, reviewing symptoms, signs, scans, and nasendoscopy, where obstruction lies at the base of the tongue or beyond. The site of the obstruction is clearly critical and large masses in the pharynx, larynx, and trachea may preclude all attempts at intubation via the upper airway.

The output should be an airway plan—or strategy. This will frequently be 'no difficulty anticipated'.

When difficulty is anticipated, a surgical airway will always be an option. The theatre team, operating surgeon, and anaesthetist (if different from those who reviewed at the multidisciplinary team meeting) should be aware of the potential issues and these should be discussed in detail at the team brief.

All necessary equipment should be present and checked. The team members should be present, and the surgeon scrubbed in anticipation. The airway should be secured in the operating theatre, not anaesthetic room. There is no clear evidence what the best anaesthetic approach is—whether intravenous (IV) induction, with or without paralysis, gas induction, or a fibreoptic approach, awake or asleep. All can fail and spectacularly, particularly when not part of an individual's regular practice. All should be prepared to challenge a plan which is clearly inappropriate. Human factors are again in play. There is no loss of face in abandoning a futile intubation attempt and moving to the definite plan. This may be a surgical airway, perhaps after allowing the patient to wake up (this may have been plan A). Occasionally, abandoning the procedure may be the only option.

Preoperative preparation and risk assessment

The Royal College of Anaesthetists publishes online a comprehensive standards document, 'Guidelines for the Provision of Anaesthetic Services'. This is updated yearly using National Institute for Health and Care Excellence methodology after reviewing the best available evidence. Chapter 2 in these guidelines addresses presurgical assessment and preparation.[4]

Preoperative preparation is just one aspect of the perioperative care of a patient and the complete process is best referred to as perioperative medicine. The emphasis is on placing the patient at the centre of a triangle involving primary care, surgeon, and anaesthetist. This facilitates[5]:

- shared decision-making
- fitness for the procedure and medical optimization
- risk stratification when appropriate.

Best practice strongly supports a preoperative anaesthetic assessment for all. This is one part of preparation for surgery which is stronger when there is multi-speciality involvement—general practitioner, surgeon, anaesthetists, and case-relevant allied professionals. There is no requirement for direct anaesthetic input in all cases and a nurse, physician associate, or anaesthesia associate will often deliver this service. The majority will have a face-to-face assessment but in fit patients for relatively minor procedures and where there are significant logistical issues, a telephone or virtual interview will suffice. Ideally the anaesthetic assessment should be temporally linked with surgical booking but, in major or complex surgery, particularly in the frail and elderly, it becomes part of a process. This process includes initiating medical optimization, reviewing investigations, and anaesthetic planning. Thus, the patient may be seen on more than one occasion.

Ideally, health and fitness should have been considered in primary care before referral.

The majority of ENT procedures are suitable for day-case surgery and are associated with a relatively small amount of physiological stress. Thus, preoperative interventions are likely to be minor. When this is not the case, the anaesthetist is best placed to coordinate medically optimizing these higher-risk patients (**Box 3.1**).

Generally, fit and young patients will survive very major surgery without much support and frail elderly patients tolerate procedures associated with minimal physiological stress—that is, most ENT procedures.

Head and neck surgery

This group often tick all the above: frail, elderly, malnourished, with multiple comorbidities and physiologically unfit.[6] Interestingly, despite extensive and lengthy surgery, the associated physiological stress in usually moderate. It is standard practice to cohort these patients in specialist centres and the ENT surgeon is one part of a surgical team involved in their care.

There is usually time (weeks) to prepare the patient for surgery as associated malignancies are often slow growing. Given that smoking and high alcohol intake are strongly associated, cardiorespiratory disease is frequently significant and neglected. Smoking cessation, even for a few weeks will improve respiratory function and reduce postoperative complications.[7]

Nutritional interventions in malnourished patients (weight loss >10% from baseline, body mass index <18.5 kg/m^2)[8] mitigate poor would healing and reduce postoperative infections.

Programmed physical activity will improve aerobic exercise tolerance and has been shown to improve outcomes from major surgery. There is, however, little evidence to support routine use of cardiopulmonary exercise testing in this group at present.

Patients with significant cardiac disease are at most risk and specifically those with New York Heart Association grade 3 or 4 and/or decompensated cor pulmonale.[9]

Box 3.1 Higher-risk patient and/or surgery

Surgical risk is independently linked to:
- age (due to physiological decline)
- type of surgery
- comorbidities
- physical (aerobic) fitness.

Brain natriuretic peptide levels are a valuable biomarker in assessing the degree of cardiac dysfunction and are routinely used for this purpose in the workup of patients for major vascular surgery.[10]

In general, most patient medication should be continued up to the time of surgery. Anticoagulant medication is often discontinued but aspirin use is not a significant risk.

Perioperative diabetic care should be formally considered and discussed and individually managed. Little adjustment is required in most patients having day-case procedures other than reducing or omitting medication during the preoperative fast.

Clear fluid (and a carbohydrate drink) is encouraged up to 2 hours prior to planned anaesthesia.

Shared decision-making, consent, and postoperative planning are complicated but essential and best made with involvement of all involved professionals, the patient, and close relatives.

Standard risk stratification is unhelpful in this group particularly as death from primary head and neck cancer is unpleasant. Surgery is often disfiguring, recovery protracted, complications frequent, cognitive decline likely, and dependency increased. Overall, many will experience a significant decline in quality of life. Thus, delivering fully informed consent (where patient not surgeon determines the information required) is challenging and requires an exploration of individual expectations and values especially around independence and quality. This is a good time to complete the ReSPECT (Recommended Summary Plan for Emergency Care and Treatment) process.

Immediate postoperative care is best delivered within an intensive care unit with agreed treatment limitations consistent with the above.

Paediatrics

A significant proportion of surgery undertaken in children falls under the domain of ENT and paediatric surgery is an equally large part of ENT workload. Excluding complicated neonatal procedures, surgery associated with congenital and genetic disorders, or complications of life-saving neonatal care, the majority of these procedures are relatively minor in relatively well children and are often managed as day cases. Notwithstanding this, perceptions and fears, driven by regulatory changes and competency demands, have, in recent years, pushed an increasing proportion of this workload from the community-serving district hospitals into tertiary centres. These offer specialized anaesthetic and surgical services as well as some form of augmented care facilities. This has not been a positive development. Children and their families are being required to travel long distances and this has rendered day surgery both impractical and perhaps inadvisable. Further deskilling in the base hospitals further increases the drift. While the tertiary centres might welcome the increased work and revenue, in reality the result has been volume overload with increased workload and a poor service.

Fortunately, in 2018 the presidents of ENT UK and the British Association for Paediatric Otolaryngology in partnership with the Royal College of Anaesthetists and Association of Paediatric Anaesthetist had the foresight to challenge this service change. A working party was formed to examine the drivers of this process and to establish an evidence-based national strategy for paediatric ENT surgery. The resulting document—'Safe Delivery of Paediatric ENT Surgery in the UK: A National Strategy' was published on 11 July 2019.[11]

This is a balanced, pragmatic, and sensible document. It is also short and a must for all with an interest. Data are presented, using Hospital Episode Statistics from across the UK, demonstrating significant shifts in care from secondary to tertiary care, for children undergoing tonsillectomy, particularly if under 3 years of age. Pragmatic recommendations are made for tonsillectomy following a comprehensive analysis of risks associated with this procedure. Essentially the authors conclude that, with a number of obvious caveats around facilities, governance, and training, this procedure (whatever the indication) should be undertaken in secondary care for children 2 years and over and as young as 1 year in high-acuity centres. Those with a number of significant risk factors (congenital, genetic, or degenerative) and at the extremes of body mass (0.4% at either end), should be managed in a tertiary centre.

Other common ENT procedures can safely be managed, beyond the neonatal period, in secondary care, again given appropriate skill mix and support. The management of emergencies will vary depending on local arrangements within operational delivery networks, but it is essential to remember that all hospitals that receive emergencies must be able to resuscitate and stabilize. Simple ENT emergences in otherwise fit children, can be safely managed in these centres (**Box 3.2**).

Anaesthesia

Children undergoing ENT procedures are usually medically fit and scheduled to have relatively minor interventions as day cases. They should be managed in a separate area from adults by experienced staff who all require appropriate safeguarding training. While preoperative medical assessment remains essential, the emphasis, particularly in younger children, lies around psychological preparation and presenting information in a suitable format for their age and ability to comprehend and process.

Generally, children do not tolerate procedures under local anaesthesia. Some adolescents, however, will be fine with minor, body surface procedures and simple external ear/tympanic membrane surgery using topical (EMLA) anaesthesia.

Pre-assessment

Nurse-led questionnaire-based pre-assessment is appropriate for the majority of normal children having minor ENT procedures. A direct anaesthetic review immediately prior to the surgery remains

Box 3.2 Controversy: day-case tonsillectomy

Day-case procedures can safely be undertaken in children older than 1 year. A target of 70% is widely quoted for tonsillectomy. This procedure has a particular additional risk of major haemorrhage and thus trusts across the UK have developed care pathways considering patient demographics and local geography.

Given the above, many of these guidelines are rather conservative. In my own trust, children under 4 years and those with symptoms of obstructive sleep apnoea are excluded.[12] This is not supported by the evidence. Distance from hospital is also relevant and we exclude all more than 30 minutes away. Consequently, targets are missed, and money is lost ('Payment by Results'). ENT departments should review their guidelines and better align with the evidence.

Box 3.3 Controversy: recent or active upper respiratory tract infection

There is no debate or controversy around cancelling surgery in children who present with active infection and who are systemically unwell. Airway hyperreactivity persists after symptom resolution and a 2-week delay is generally supported. If this policy were extended to all children with a runny nose or cough who are otherwise well, some would never come to theatres. Thus, most now accept an increased, but still small, risk of early and short-lived respiratory complications. Anaesthetic technique is relevant here. There is support for the use of total IV anaesthesia with propofol but IV induction and inhalational anaesthesia using sevoflurane or isoflurane is perfectly acceptable.[13]

mandatory. Children with chronic disease, symptoms of sleep disordered breathing, and extremes of weight will require more formal review (Box 3.3).

Pre-medication

I rarely use sedative premedication in children although we all recognize cases where it is almost impossible to get a child into the theatre suite, let alone induce anaesthesia. Parental cooperation is essential but the administration of midazolam (orally, nasally, or rectally) may be unavoidable and particularly in children with 'prior experience'.

It is increasingly popular to administer both paracetamol and ibuprofen to all but for very minor procedures. Fewer opioids are required postoperatively and there is less nausea and vomiting, particularly following tonsillectomy.

Choice of anaesthesia

The choices revolve around gas versus IV induction, IV versus gaseous-based maintenance, tracheal intubation versus supraglottic airway, and paralysis requiring mechanical ventilation versus spontaneous ventilation.

There is no one correct technique, and each will have their own preference for different procedures. Generally, less is more, and I try not to overcomplicate things.

IV induction is favoured by most but gas induction may be the most sensible option where IV access is difficult and the child uncooperative. Propofol is the preferred agent. It is safe with a wide therapeutic window, has antiemetic properties, and reduces airway reactivity. This holds even when anaesthesia is maintained using a volatile anaesthetic.

The avoidance of endotracheal intubation is of significant benefit and this can be safely achieved using a second-generation supraglottic airway. The smallest reinforced laryngeal mask airway (with narrow pilot tubes), particularly for tonsillectomy, is suitable for all but children under about 3 years as there is no evidence of increased surgical difficulty. Surgical perception sometimes leads to difficult conversations.

I still use nitrous oxide in all but middle ear surgery. The only sustainable case against its use (and this holds for all volatile anaesthetics, particularly desflurane) is environmental. The most frequent issue with propofol is not giving enough. Whenever possible, intubation should be avoided, as discussed above. Clearly there are cases where this will not be possible (congenital issues, morbid obesity for all but short procedures). Spontaneous ventilation seems to be preferred by most and allows removal of the airway while still anaesthetized. Even when intubating for tonsillectomy, I prefer spontaneous ventilation, and this is relatively straightforward with sevoflurane or isoflurane. The risks of administering succinylcholine are significantly overstated. Intra- and postoperative analgesia should reflect the needs of the surgery. Fentanyl is contextually short-acting and suitable for intraoperative analgesia and procedures with little postoperative pain such a myringotomy and intranasal surgery.

Dexamethasone (0.15 mg/kg) appears to be of significant benefit in reducing postoperative nausea and vomiting in children undergoing tonsillectomy.[14]

I usually administer morphine during tonsillectomy and prescribe postoperatively. Outside the early years of life, the anaesthetic requirements for different otolaryngology procedures are similar in adults and children and will be discussed together in the following sections.

Local and regional anaesthesia

Many procedures can and are carried out without general anaesthesia although the addition of sedation is sometimes useful in the anxious or where the procedure is not straightforward or likely to be prolonged. Axial and plexus (excluding superficial and deep cervical) blocks are not available for ENT procedures and thus topical, infiltrative, and individual nerve blocks must be used. It is self-evident that minor procedures on skin and soft tissues are routinely performed, as are nasal, pharyngeal, and laryngotracheal flexible endoscopies (including awake intubation). These are not well tolerated using rigid scopes, however. Operations on the auditory canal and tympanic membrane are fine using a combination of infiltrative and topical anaesthesia, as are procedures on the nasal cavity including turbinectomy and polyp removal. Tonsillectomy and other pharyngeal procedures can and are performed without general anaesthetic. The ability to perform awake tracheostomy is essential. In fact, a brief literature search uncovers a description, usually by an 'enthusiast', of almost all surgical procedures without the use of a general anaesthetic.

Just because something is possible does not mean it is a good idea. General anaesthesia is safe, effective, and has facilitated the development of modern surgical practice. There are very few patients who cannot have a general anaesthetic. It is a complete misnomer to say a patient is not fit for an anaesthetic as the issue is one of tolerating physiological stress and this is almost entirely triggered by surgery. The patient is still (usually) and kept safe. There is scope to manipulate the surgical conditions and the surgeon is freed from these responsibilities to concentrate on the procedure. Unfortunately, safe general anaesthesia is not available to a significant portion of the world population and here many surgeons have developed the skills to perform an extensive list of procedures quickly, using local anaesthesia. It is facile to compare outcomes in this context given the lack of options.

Procedural (general) anaesthesia

Most anaesthesia is induced with an IV injection of a rapidly acting hypnotic (usually propofol although others still exist). Gaseous induction is generally well tolerated and frequently used in small

children. There is an ongoing debate about gaseous versus IV induction in patients with upper airway obstruction where awake intubation is not feasible. The choice is essentially operator and context specific and there is no correct answer. A potent narcotic is usually part of the induction process as is a muscle relaxant when indicated. Propofol has a high plasma clearance, short elimination half-life not dependent on renal or hepatic function, is safe with a wide therapeutic index, suppresses airway reflexes, and has potent antiemetic properties. It is thus an excellent baseline agent for maintaining anaesthesia by infusion. Remifentanil is an ultrashort-acting synthetic opiate. Administration can be rapidly adjusted to match the changing surgical requirements.

Computerized infusion systems coupled with depth of anaesthesia (cerebral function monitoring) are reasonably (but not completely) reliable in ensuring the patients are actually asleep. Total IV anaesthesia is very popular with an increasing number of first-world anaesthetists. I do not use this technique, preferring to maintain anaesthesia with a combination of potent volatile agent and non-infusion opiate. At the end of the day it is very difficult to show any difference in patient outcome. The only significant argument against volatile anaesthetics is environmental. All are either greenhouse gases (nitrous oxide) or ozone depleters, or both. The standout 'bad boy' here is desflurane with a global warming potential of approximately 2500 times that of carbon dioxide. Given that desflurane is also incredibly stable in the atmosphere it has been estimated that it is responsible for at least 80% of the pollution from all volatile anaesthetics combined. The main issue with nitrous oxide is the volume released and anaesthesia contribution is significant. There is thus a logical argument to either find a non-polluting volatile anaesthetic (xenon) or capture at source or substantially reduce first-world use. Desflurane use has already stopped in many centres and their world did not end.[15]

Otology

Excluding minor surgery on the external auditory meatus and ear canal, most otological surgery involves either procedures to deal with infection (usually chronic), cholesteatoma, deafness, or any combination. Chronic middle ear conditions often develop in children and progress, variably, into adulthood.

Deafness may be congenital or acquired and a bone-anchored hearing aid is an alternative to a conventional hearing aid where middle ear transmission is disrupted. Cochlear implanted electrodes aim to stimulate the auditory nerve and are most useful in children who are profoundly deaf, usually placed early in life when communication skills are being developed.

Minor procedures, including myringotomy and myringoplasty can be performed under local (or topical anaesthesia) in compliant patients. Given that more complicated procedures often involve prolonged surgical time and require a still patient and bloodless field, the vast majority of these procedures are performed under general anaesthesia.

There are a number of relevant considerations:

- Preoperative:
  - Associated syndromes (Down, Klippel–Feil) with both potential airway and cardiovascular issues.
  - Age and frailty but minimal surgical stress so low risk.
- Lengthy procedure with difficult airway access once underway.
- Technically difficult procedure requiring good surgical field.
- Pressure equalization in a closed cavity.
- Proximity of facial nerve.
- Potential for postoperative nausea and vomiting (multifactorial).

The facial nerve runs across the medial and posterior wall of the middle ear, usually within bone. Middle ear pathology often distorts the anatomy and may also erode the bony covering. Facial nerve injury is disfiguring and psychologically damaging and thus the use of a nerve monitor is standard practice for many. This relies on motor stimulation and does not work in the presence of neuromuscular blockers. Their use can be completely avoided by securing the airway with a supraglottic device and allowing spontaneous ventilation. Excessive respiratory movement and elevated carbon dioxide has the potential to be an issue.

Intubation with a single dose of a short-acting relaxant is an option favoured by many.

Controlled ventilation is relatively straightforward in most without paralysis but does involve a deeper level of anaesthesia and respiratory suppression. The use of short- or ultrashort-acting opiates (fentanyl, alfentanil, remifentanil) will facilitate this. Bleeding is not usually a problem and infiltration with adrenaline-containing local anaesthetics helps. Controlled hypotension may not. There is some controversy concerning nitrous oxide and its differential diffusion. This is not an issue with an open middle ear but may result in a rise in pressure when initiated and fall when terminated. Tympanic graft displacement is a potential issue and thus, if used, nitrous oxide can be safely discontinued 15 minutes before closure.

Postoperative pain is not excessive and can often be managed with simple analgesics.

Nausea and vomiting are significant issues with middle ear surgery, particularly in children.

There is evidence that the use of a propofol infusion in place of a volatile anaesthetic and the avoidance of nitrous oxide is of some benefit. These are one of the few groups of patients I routinely give perioperative antiemetics to and this may include dexamethasone in addition to ondansetron.

Rhinology

Surgeries on the nose and nasal cavity are among the most frequently performed of all procedures. Most are suitable for ambulatory care and include simple bony manipulations following trauma, operations on the nasal septum, or to control haemorrhage. Rhinosinusitis is an unpleasant and debilitating condition, and many sufferers require repeated surgical interventions given the limited effectiveness of medical treatment. Precision endoscopic surgery has largely replaced directly visualized surgery. Functional endoscopic sinus surgery is thus the staple of this subspecialty. Patients are of all ages, shapes, and sizes and medical fitness. However, the vast majority remain suitable for a day-case procedure. Unlike surgery on the middle ear, procedures on the nasal cavities and sinuses have the potential to become a blood bath, rendering further intervention both dangerous and impossible. Both anaesthetist and surgeon act to mitigate this.

However probably the most important intervention is the application of topical vasoconstrictive agents both before and during surgery. It remains common practice to pack the nose with a topical solution immediately after induction. Cocaine is still widely used, and a 4% solution has proven safe and effective.[16] A maximum dose of 200 mg is widely quoted but without scientific basis. Adrenaline-soaked packing (1 in 1000) is probably equally effective but most frequently used intraoperatively. Many inject lidocaine and adrenaline (1 in 200,000) submucosally. It is possible to carry out this procedure without general anaesthesia; please feel welcome to try.

Not withstanding this sort of heroics, a short general anaesthetic is my choice for all.

The same arguments regarding IV versus inhalational maintenance, as rehearsed above, pertain. In reality, there is little difference. It is safe and reasonable to allow the patient to breathe spontaneously in all but a few cases and the use of a second-generation supraglottic airway provides excellent barrier protection, smother emergence, and less irritating minor postoperative complications. It is not universally accepted that hypotensive anaesthesia improves surgical conditions. Unless there is an obvious contraindication, however, I do aim to achieve this and am perfectly happy to couple with 15–20° head-up position. Tranexamic acid, 1000 mg IV, has been shown to reduce bleeding in these procedures (**Box 3.4**).

Laryngotracheal, head, and neck surgery

Most of these procedures involve a malignancy of the mouth, pharynx, larynx, and/or associated head and neck structures. Management is multiprofessional and often well planned. Oral resections are the domain of the maxillofacial surgeon while ENT surgeons deal with the pharynx, larynx, and neck structures. Plastic surgeons most frequently concentrate on reconstruction including microvascular procedures. Major head and neck procedures are lengthy and may well run late into the evening. Surgical roles change as the operation progresses but there remains an issue with a single anaesthetist conducting the whole procedure. Concentration is inevitably lost, and tiredness is a real problem, most evident at probably the riskiest time—emergence. An inevitable solution is a rotating anaesthetic team. Not all of this workload involves a major resection. Endoscopic examinations and limited surgery of the pharynx and larynx arguably demand more experience and skill from the anaesthetist than the above but also are usually limited to a small team. Not so airway emergencies in adults and children that may appear anywhere at any time. Simulation has a major role in maintaining skills and confidence in surgeons and anaesthetists who may only have to deal with this sort of issue every few years. I have already discussed preoperative management and airway issues in some detail and will not revisit these.

Box 3.4 Controversy: throat packs

A retained throat pack sits high on the list of never events. All never events, while rare, seem to occur randomly and are thus inevitable. The only way to avoid a retained throat pack is to not insert one. Given this, a review was conducted by the Difficult Airway Society in association with ENT UK and the British Association of Oral and Maxillofacial Surgery[17] to identify evidence supporting throat pack insertion. None was identified (perhaps because it has not been looked for) but there was plenty citing adverse events. I no longer insert a throat pack and it seems sensible and right that if a throat pack is *required*, it is part of the surgical procedure and thus included in the routine instrument/swab count.

Short procedures

I was tempted to call these 'minor procedures' but nothing involving the airway is without significant risk. Procedures include diagnostic endoscopic examinations including biopsies of the pharynx and larynx, emergency removal of a foreign body, and resections of tumour (at any level including subglottic). Airway and ventilatory management will depend on the surgical access required. Fortunately, the majority of procedures can be undertaken with the trachea intubated. Microlaryngeal tubes, flexometallic tubes, and laser-resistant tubes all allow a safe airway and conventional ventilation. Airway resistance is high and paralysis with assisted ventilation all but mandatory.

Tubeless anaesthesia

It is not possible to carry out some procedures with an endotracheal tube *in situ*. Oxygenation relies on some form of apnoeic ventilation, or through use of high-pressure (jet) ventilation, or allowing spontaneous ventilation. The patient population here rarely do well with apnoeic ventilation but use of high-flow nasal oxygenation is proving of benefit. There are a number of devices available for jet ventilation, some fixed and some variable pressure. Supraglottic delivery relies on attachment to a channel on a suitable endoscope. There is often a degree of entrainment and risk of gastric insufflation if the jet is not aligned with the glottis. There are two, laser-resistant, fine-bore catheters available for transglottic jet ventilation. The Hunsacker Mon-Jet has a basket distally designed to help centralize in the trachea while the LaserJet catheter is without. Both can be attached to automated jet ventilators. Both seem to do the job intended. I have no experience of transtracheal catheters but there is evidence of an increased complication rate. One wonders if formal tracheostomy might be a better option in this very small and clearly compromised group. The anaesthetic options are limited in this group. Inhalational anaesthetic cannot be used as delivery is not possible and even if it were, the surgeon would also be anaesthetized. An IV technique, either manual or automated, is thus used. I tend to try and avoid ongoing paralysis, beyond initial airway manipulation (surgical and anaesthetic), but this is not always possible. These patients have little postoperative pain and rarely need to remain in hospital overnight. A few find their way into critical care.

Long procedures

In our practice all elective major head and neck cancer resections are admitted to critical care postoperatively. Free flap viability is routinely monitored both by surgically attached Doppler and frequent nurse observations. We aim to extubate all patients at the end of the procedure, whatever the finish time. Surgical tracheostomies are sited electively when indicated and some more when swelling and airway compromise is evident at the end of the procedure. Many patients undergoing intraoral procedures have a patent airway but may be very difficult to reintubate in the immediate postoperative period. A transnasal airway exchange catheter is left *in situ* and removed the next day. Full consideration of patient positioning is imperative with meticulous attention to airway security, skin and

nerve compression, and temperature control. Direct arterial pressure monitoring is usually a good idea. It facilitates more reliable blood pressure manipulation and regular examination of metabolic status as an aid to fluid replacement. I very rarely undertake central venous cannulation in this group.

There are two distinct phases to the procedure: resection and reconstruction. While blood loss is not usually an issue, elective hypotension may be useful during resection and when raising tissue for reconstruction. Optimizing blood flow with normotension is the aim when reconstructing. Generally, a full, relatively dilated circulation is the aim at this stage. Given that this patient group may well have significant associated vascular disease and untreated hypertension it may be evident that flap perfusion is more pressure than flow dependent. There is little surgical stimulation during reconstruction and thus active intervention to increase blood pressure is a common requirement. Direct-acting vasopressors are most frequently used and metaraminol is often the first choice. Ephedrine is likely to be ineffective beyond a couple of doses. Noradrenaline is not recommended by some, but I think this is wrong. The increase in perfusion pressure usually outweighs any local vasoconstriction and short half-life facilitates immediate termination. Dobutamine rarely increases blood pressure while unhelpfully increasing heart rate. Adrenaline use is associated with a rise in lactate—indicative of worsening tissue perfusion—exactly the opposite of the requirement.

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4

Practising otolaryngology in the COVID era

Andrew Thamboo

Background

COVID-19: origin and source of transmission

Coronavirus disease 2019 (COVID-19) is a transmissible and pathogenic viral infection that is caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2).[1] Coronaviruses (CoVs) are a part of the *Coronaviridae* family and can cause respiratory illness in humans and gastrointestinal illness in animals.[2,3] Alpha (α), beta (β), gamma (γ), and delta (δ) coronaviruses are subdivisions of the *Coronaviridae* family.[4] Alpha-coronaviruses and β-coronaviruses have the capability to infect mammals; however, γ-coronaviruses and δ-coronaviruses usually infect birds.[4] In November 2002, the first outbreak of severe acute respiratory syndrome (SARS) disease in humans caused by SARS-CoV-1 occurred in Guangdong, China.[5] A decade later, the Middle East respiratory syndrome (MERS) disease caused by MERS-CoV (a β-coronavirus) was observed in Middle Eastern countries[6] and starting in December 2019, we are now dealing COVID-19 caused by SARS-CoV-2 (β-coronavirus), which was witnessed first in Wuhan, China.[7]

After SARS and MERS, COVID-19 is the third zoonotic disease experienced by mankind.[8] Genomic analysis has determined that bats are the primary reservoirs for coronaviruses, including the SARS-CoV-like viruses.[4] However, the transmission routes and intermediate hosts involved in animal-to-human transmission remain elusive.[4] In contrast, it is confirmed that the main cause of human-to-human transmission is through (healthy) humans being in close proximity with an infected individual.[4]

Infection mechanism

SARS-CoV-2 is an enveloped, non-segmented, positive-sense RNA virus consisting of a single-stranded RNA.[9] The main SARS-CoV-2 structural proteins are the spike, envelope, membrane, and nucleocapsid proteins.[10] The general infection mechanism for SARS-CoV-2 begins with its spike glycoprotein binding to the host cell's angiotensin-converting enzyme 2 (ACE2) receptor.[1] This results in a conformation change in the spike glycoprotein which enables viral envelope fusion with the cell membrane.[1] Afterwards, SARS-CoV-2's RNA is released into the host cell where it is translated into two polyproteins (pp1a and pp1b) that form a replication–transcription complex (RTC) inside a vesicle.[4] The RTC replicates and synthesizes RNAs that encode accessory and structural proteins which assemble along the endocytic pathway to form viral buds that then fuse with the plasma membrane and release the virus.[4]

This SARS-CoV-2 infection mechanism is closely related to SARS-CoV-1, as they both utilize the host cell's ACE2 receptor to infect human cells.[11,12] This ACE2 receptor is the main mediator of cross-species and human-to-human transmission, and is highly expressed in nasal tissue and on type I and II alveolar epithelial cells in normal human lungs.[4,13] This lung expression is drastically increased with the binding of SARS-CoV-2, which has a 10–20-fold higher ACE2 receptor binding affinity than SARS-CoV-1.[8,14] This results in SARS-CoV-2 causing severe damage to alveolar epithelial cells which prompts a series of systemic reactions that can lead to death.[8,14]

Anosmia—a COVID-19 symptom

It is vital to identify COVID-19 symptoms to enable swift screening, diagnosis, and self-isolation for infected patients.[6] Anosmia is one presenting symptom that has been associated with COVID-19 patients.[15] Initially, early reports from Wuhan, China, regarding COVID-19 did not have anosmia as a common symptom.[15] However, the World Health Organization has added anosmia to their list of official symptoms as a number of countries such as South Korea, Italy, Germany, the UK, Iran, and the US reporting anosmia cases related to COVID-19.[16,17] A case series conducted by Heidari et al. analysed the health condition of 23 COVID-19 confirmed patients with recent anosmia.[18] Of these 23 patients, 16 patients had anosmia only, three patients had anosmia and low-grade fever, and four patients had anosmia and mild myalgia/fatigue.[18] The fever, myalgia, and fatigue symptoms subsided in a few days, but anosmia symptoms persisted.[18] Interestingly, 19 patients (83%) from their 23-patient sample reported anosmia as their first symptom.[89] This smell impairment has been shown to be strongly associated with COVID-19 infection.[19] Lechner et al. demonstrated this in a 2020 study using three frontline healthcare professionals who displayed early anosmia. All three of these healthcare professionals subsequently tested positive for SARS-CoV-2.[17]

The association between COVID-19 and anosmia has been demonstrated but the pathogenesis is unconfirmed. One theory is that SARS-CoV-2 could use the high number of ACE2 receptors on nasal epithelial cells to enter and disrupt olfactory neuroepithelium cells.[20] This can lead to inflammation which can damage olfactory receptor neuron function resulting in anosmia.[21] Overall, anosmia is a good predictor for COVID-19; however, additional research is needed to confirm the pathogenic mechanism.

Testing for COVID-19

The diagnosis of this infection can occur through two different types of testing: molecular (direct) and serological (indirect) testing. The former, specifically nucleic acid amplification tests such as reverse transcriptase polymerase chain reaction, is the current gold standard while the latter is under development.

Molecular tests are currently recommended for diagnosing the SARS-CoV-2 infection.[22] They detect viral RNA from samples using nucleic acid amplification tests such as reverse transcriptase polymerase chain reaction, which targets gene sequences that code for components of the viral envelope, nucleocapsid, or RNA-dependent RNA polymerase.[22] The location of respiratory sample collection is important due to differences in viral load. Current guidelines from the US Centres for Disease Control recommend attaining nasopharyngeal specimens for swab viral testing. If this is not possible then alternative methods are available, such as throat and nasal swabs, with higher viral loads being detected in the latter and a combination of throat and nasal sampling having a similar sensitivity to nasopharyngeal samples.[23,24] Additional alternative methods include sputum swabs, oropharyngeal specimens, nasal mid-turbinate specimens, and nasal and nasopharyngeal wash or nasal aspirate specimens.[22] Titres of SARS-COV-2 can also be detected in blood and stool but this is less reliable than the aforementioned respiratory samples.[22] These direct tests have played large roles in controlling the current pandemic, but they have limitations, including potential false-negative results, changes in diagnostic accuracy over the course of the disease, and precarious availability of materials.[25] In contrast, serological tests are cheap and user-friendly, can greatly improve our understanding of the host immune response and cross-reactivity, and can help guide public health measures.[22,25]

Serological tests detect anti-SARS-CoV-2 immunoglobulins and can be divided into three main categories: lateral flow immunoassays (LFIAs), enzyme-linked immunosorbent assays (ELISAs) and chemiluminescent immunoassays (CLIAs). In terms of test performance, LFIA has a lower sensitivity than ELISA and CLIA methods and is at risk of false positives caused by other viral infections.[25] Therefore, similar to the previous two coronaviruses, LFIA serological tests are not recommended for the diagnosis of SARS-CoV-2.[22,25] In contrast, ELISA and CLIA are more suitable for clinical application due to costs, time to results, simplicity, and their ability to be scaled up to high-throughput testing.[22] To be viable diagnostic tests, ELISA and CLIA require a sensitivity of at least 90% which in their case is only achieved or trended towards 3 weeks after symptom onset.[25,26] As a result of their high false-negative rates during the first 2 weeks of symptoms, these serological tests should not be recommended as a point-of-care or in-hospital diagnostic test for SARS-CoV-2.[25–27] Despite this, they could play a role in detecting previous SARS-CoV-2 infection or protective immunity if used 15 or more days after symptom onset but additional studies based on peer-reviewed literature are needed to validate this statement.

Aerosol- and droplet-generating medical procedures in otolaryngology

Aerosol-generating medical procedures (AGMP) can create (<5–10 μm) aerosols that can travel more than 2 metres.[28] In contrast, droplets are larger particles (>5–10 μm) that travel less than 2 metres from the infected person and can be produced through coughing, sneezing, talking, singing, and droplet-generating medical procedures (DGMP).[28,29] Defining AGMPs and DGMPs is essential in determining the appropriate personal protective equipment (PPE) required to maintain the safety of healthcare professionals and patients. Given the close proximity of otolaryngologists with patients during the physical examination, it puts each individual at higher risk of infection. At the time of writing this chapter, the knowledge surrounding AGMPs and DGMPs in the field of otolaryngology is limited and knowledge surrounding COVID-19 is exponentially growing.

Nonetheless, we can extrapolate our understandings from the literature surrounding other pandemics and viral studies to keep our patients and healthcare providers safe during this pandemic. The most common examination tool used by otolaryngologists is endoscopy (rigid or flexible). Endoscopic examinations in areas of high viral load such as the nose, nasopharynx, oropharynx, and larynx are considered AGMPs and DGMPs.[28,30–33] This is potentially due to the increased risk of coughing and sneezing associated with endoscopic examinations.[28,34] Conversely, bronchoscopy is considered an AGMP likely due to the suctioning involved in the procedure.[34] Procedures that involve open suctioning of an airway are considered AGMPs and DGMPs, and the former could produce aerosols containing viable pathogens.[28,34,35] Labelling endoscopy as an AGMP has been controversial as the knowledge surrounding this topic is scarce; therefore, based on different health authorities and expert opinion, endoscopy has been labelled as either an AGMP or DGMP, or both. Nebulizers used in the clinic setting for anaesthetizing the nose for endoscopy are considered DGMPs.[28] In the management of epistaxis, it is considered a DGMP and a potential AGMP due to the coughing and sneezing that can be elicited.[28,30]

There are a variety of otolaryngology surgeries that can be labelled as AGMPs or DGMPs. Tracheostomy is considered an AGMP due to the high-pressure airflow originating from the endotracheal intubation that can cause aerosolization and an increased risk of acute respiratory infection transmission.[28,30,33,36] Intubation and extubation associated with tracheostomy and in general are considered AGMPs that carry an increased risk of acute respiratory infection transmission.[28,33,36] Endoscopic sinonasal and anterior skull base surgery are considered AGMPs due to the high-powered instruments such as the microdebrider and drill that are utilized.[28] These instruments can cause droplet and airborne transmission, and this is amplified when the suction on the microdebrider becomes plugged.[28] Such risks can be mitigated by placing another suction into the nasopharynx via the nasal or oral cavity. Additional tools such as carbon dioxide laser ablation and electrocautery can cause aerosolization, with the former creating small virus-containing aerosols in the

smoke plume.[28,32] Electrocautery generates blood aerosols and tissue particles whose concentration increases with higher electric current, but the presence of SARS-CoV-2 RNA has yet to be confirmed.[28,34] The likely explanation is that SARS-CoV-2 is inactivated by the electrocautery because the virus is sensitive to temperatures above 70°C which is greatly surpassed by electrocautery devices.[28]

Surgical procedures in the oral cavity and middle ear also pose a risk for aerosolization. Oropharyngeal surgery and dental procedures tend to use drills, ultrasonically activated devices, saws, and high-pressure water sprays which can cause aerosolization.[28,30,34] Therefore, procedures that use these instruments in the oral cavity or pharynx are considered AGMPs. Similarly, the high-speed bone drilling utilized during a mastoidectomy produces aerosolized bone dust, blood droplets, tissue (mucosa) particles, and irrigation fluid that increases the risk of disease transmission.[28,34] Overall, AGMPs and DGMPs increase the risk of COVID-19 transmission which emphasizes the importance of proper PPE and surgical protocols to keep everyone in the operating room safe.

Personal protective equipment

PPE is required to reduce the risk of contracting COVID-19. Different levels of PPE are worn based on the risk of SARS-CoV-2 infection. Recommendations of PPE should be based on local health authority rules and supersede statements made in this section but the following should provide guidance in places where recommendations are not available (Tables 4.1 and 4.2).[37,38] First-level PPE is recommended in various situations. During routine patient care and all non-AGMPs, patients who are asymptomatic and at low risk of community contact require only level-one PPE be worn by all healthcare workers (HCW).[38,39] During outpatient situations, patients should also be wearing surgical masks to help eliminate all detectable aerosol spread.[40] Additionally, staff who welcome patients should be wearing a surgical mask at the discretion of their workplace. A caveat to this is that they can discontinue use of level-one PPE and only practise social distancing if local COVID-19 prevalence is low and the patient is asymptomatic or tested negative.[38] In contrast, if patients are symptomatic but tested negative for SARS-Cov-2 they are at an intermediate risk of spread, thus level-one PPE can be used for non-AGMPs only when local COVID-19 prevalence is low.[38] If COVID-19 prevalence is high, HCWs should wear level-two PPE.[38]

Table 4.1 Levels of personal protective equipment

Level	PPE
Level 1	• Work clothes • Isolation gowns • Head cover • Normal surgical mask • Gloves • Face shield or goggles
Level 2	• Work clothes • Water-impermeable gowns and/or coveralls • Head cover with neck protection • N95 respirator or FFP2 (if N95 respirator is not available a surgical mask with goggles or a face shield is sufficient) • Double gloves • Shoe covers • Face shield or goggles
Level 3	• Gowns (coverall and second sterile surgical gown or a water-impermeable gown) • Head cover with neck protection • PAPR (if PAPR is not available, a N95/FFP2 mask with a surgical mask over top) • Double gloves • Goggles or a face shield

FFP2, filtering face piece 2; PAPR, powered air-supply filter respirator.

Source data from Cui C, Yao Q, Zhang D, et al. Approaching otolaryngology patients during the COVID-19 pandemic. *Otolaryngol Head Neck Surg*. 2020;163(1):121–131 and Lammers MJW, Lea J, Westerberg BD. Guidance for otolaryngology health care workers performing aerosol generating medical procedures during the COVID-19 pandemic. *J Otolaryngol Head Neck Surg*. 2020;49(1):36.

Level-two PPE should be worn in three scenarios: (1) when asymptomatic patients cannot undergo testing and require an urgent AGMP. (2) Also, when symptomatic patients test negative for SARS-CoV-2 HCWs should wear a minimum of level-two PPE when performing AGMPs, especially if local COVID-19 prevalence is high.[32,38] This is due to the potential for false-negative test results due to the sensitivity of current molecular tests.[38] (3) Lastly, patients who are symptomatic and are SARS-CoV-2 positive or unknown are at high risk of spreading COVID-19, thus all HCWs need level-two PPE when working close to these patients or when performing short-duration AGMPs.[38,41] If N95 respirator supply is low, HCWs can wear the same mask for 4–8 hours as long as they are not donned and doffed, but this comes at the risk of the mask's air tight seal leaking during a procedure.[32,34,39]

Table 4.2 Different levels of PPE and situations for use

Patient characteristics	Level 1	Level 2	Level 3
Asymptomatic patients	Patients with low risk of community contact: • Routine patient care • All non-AGMPs	Can't undergo testing and require urgent AGMP	
Symptomatic patients and COVID-19 negative	Non-AGMPs when local COVID-19 prevalence is low	Non-AGMPs when local COVID-19 prevalence is high AGMPs	Based on symptoms, risk of aerosol formation, COVID-19 prevalence, and local resources/protocols
Symptomatic patients and COVID-19 positive or unknown		Working close to patients Short-duration AGMPs	Long-duration AGMPs

Source data from: Cui C, Yao Q, Zhang D, et al. Approaching otolaryngology patients during the COVID-19 pandemic. *Otolaryngol Head Neck Surg*. 2020;163(1):121–131 and Lammers MJW, Lea J, Westerberg BD. Guidance for otolaryngology health care workers performing aerosol generating medical procedures during the COVID-19 pandemic. *J Otolaryngol Head Neck Surg*. 2020;49(1):36.

Level-three PPE should also be considered in two categories of patients. First, if the patient is symptomatic but tested negative for SARS-CoV-2, level-three PPE should be used based on the patient's symptoms, risk of aerosol formation, local COVID-19 prevalence, and local resources and protocols.[38] Second, level-three PPE should be worn if the patient is SARS-CoV-2 positive and a long-duration AGMP that is high risk for aerosol formation is being performed.[38,42] Furthermore, the decision on which level of PPE to use depends on the patient symptoms and COVID-19 status, the procedure being performed, local COVID-19 prevalence, resources, and protocols.

Future of otolaryngology

Otolaryngologists are at high risk of SARS-CoV-2 infection due to the high viral titres in the head and neck region; thus the future of otolaryngology must adapt to the threat of additional upper respiratory virus pandemics. Telemedicine has become common since the pandemic but limitations exist when it comes to the physical exam. Applicable telemedicine platforms include Quintree, ZOOM, Google Hangouts, and Apple Facetime.[43,44] Teleconsultations using these platforms are amendable to head and neck cancer, otological, and rhinological patients with the caveat of lacking physical examinations.[44–46] These teleconsultations allow for the patient to be heard, surgical sites to be assessed, pathology and imaging to be discussed, and proper triaging of patients.[44,46] For telemedicine to be permanently integrated into otolaryngology practices two steps are recommended: (1) explore secure telemedicine platforms, and (2) develop written policies and procedures for providing telemedicine services.

Without the ability to perform a physical examination, otolaryngologists have leaned more on investigations to facilitate in the diagnosis. Unfortunately, there is an inherent increase in healthcare dollars spent in making a diagnosis. Moreover, the physical examination cannot always be replaced by an investigation. There are a lack of studies comparing the efficacy and medical risks associated with using computed tomography over nasal endoscopy. Therefore, a low threshold for in-person consultations is needed if imaging abnormalities are found.[44]

In the operating room, surgeons may have initially shied away from elective procedures that are of higher risk of infection like sinus surgery but with increases in knowledge regarding COVID-19, availability of PPE, and mitigation strategies, surgeons have resumed doing all previous elective cases. The future for otolaryngology with respect to the breadth of surgery will likely be similar to the pre-COVID era. The changes in prevalence of community COVID-19 infections will dictate how hospitals and physicians proceed.

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8

Otalgia and otorrhoea

Philip J. Clamp

Introduction

Otalgia and otorrhoea are common otological symptoms with a broad range of aetiologies. Otorrhoea is the term given to any discharge from the ear and can be sub-categorized as mucopurulent, bloody, serous, or watery. Otalgia may be caused by inflammation of the outer ear, middle ear, or by referred pain.

The majority of cases of otalgia and otorrhoea are caused by acute ear infections such as otitis externa (OE; which predominates in adults) or acute otitis media (AOM; more common in children). However, a number of conditions may present with similar features (Table 8.1). Initial treatment of the acute infection with subsequent follow-up examination may be required in order to reveal underlying conditions. Cases of recurrent, atypical, or non-resolving infection should be viewed with suspicion and investigated for additional pathology.

Anatomy

Otorrhoea may originate from the external ear canal, tympanic membrane (TM), or from the middle and inner ear via a TM defect.

Primary otalgia is caused by conditions of the outer or middle ear (the inner ear being relatively insensate). A sensation of aural fullness or pressure may be present in inner ear disorders such as Ménière's disease (see Chapter 14), Eustachian tube disorders see (Chapter 9), or neurological conditions.

Secondary otalgia originates from surrounding structures (such as the temporomandibular joint (TMJ)) or is referred from distant sites that share innervation with the ear. The ear has a complex and overlapping sensory supply involving a number of cranial and cervical nerves. Pathology affecting any of these nerves can produce referred otalgia (Figure 8.1). A common example of cross-innervation occurs during aural cleaning; stimulation of vagal nerve fibres supplying the floor of the ear canal may induce a sensation of throat irritation and coughing. Examination and investigation for sources of referred otalgia should be undertaken in cases where no causative pathology is identified in the ear.

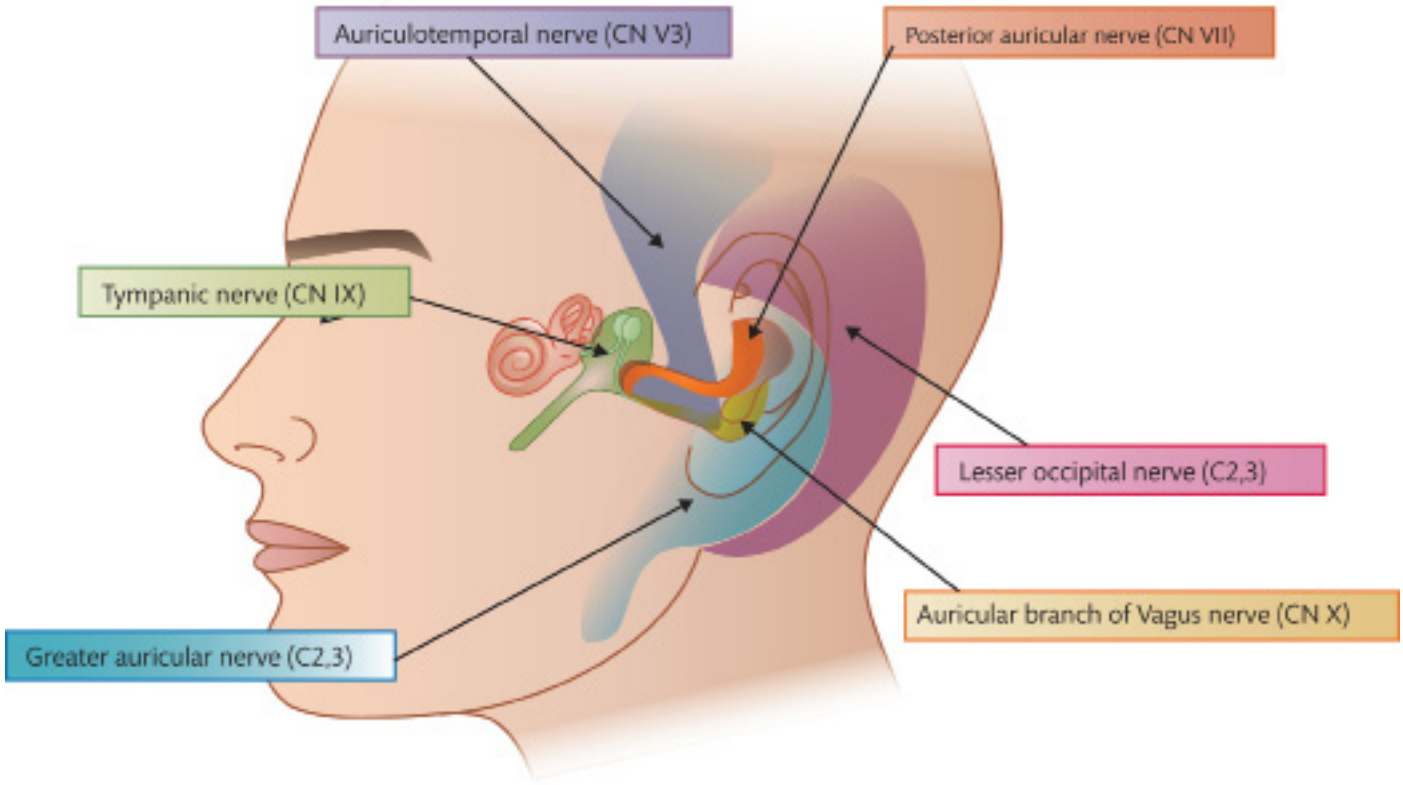
Table 8.1 A summary of the common causes of otalgia, otorrhoea, and aural fullness

	Otorrhoea	No otorrhoea
Painful	Acute otitis media with TM perforation OE Necrotizing OE Chronic otitis media Ear canal/eardrum polyps and granulations tissue (usually secondary to underlying pathology) External auditory canal foreign body Ear canal malignancy	Pinna perichondritis or cellulitis Chondrodermatitis nodularis helicis Trauma Early or mild OE Furunculosis AOM Bullous myringitis Mastoiditis Eustachian tube dysfunction HZO/Ramsey Hunt syndrome Malignancy (primary or producing referred pain) TMJ dysfunction Neuralgia Osteoradionecrosis of the temporal bone
Painless	Trauma (usually bloody otorrhoea) KO Canal wall cholesteatoma Chronic OE (often itchy) Infected TM perforation Ventilation tube (grommet)-associated otorrhoea Chronic otitis media/ cholesteatoma Granular myringitis CSF leak via a TM perforation	**Aural fullness (not reported as pain) may be caused by:** External auditory canal wax impaction External auditory canal foreign body Eustachian tube dysfunctions Otitis media with effusion TMJ dysfunction Neuralgia Ménière's disease Vestibular migraine Superior semicircular canal dehiscence syndrome

AOM, acute otitis media; CSF, cerebrospinal fluid; HZO, herpes zoster oticus; KO, keratosis obturans; OE, otitis externa; TM, tympanic membrane; TMJ, temporomandibular joint.

Assessment

In cases of otalgia or otorrhoea, diagnosis is dependent on a thorough history and examination supported by relevant investigations. Otorrhoea, wax, and debris should be carefully and thoroughly cleaned from the ear canal to allow inspection of the ear and permit topical therapy to penetrate. For most cases of otorrhoea, topical



Nerve	Origin	Sensory supply	Sources of referred otalgia
Auriculotemporal nerve	Mandibular division of trigeminal nerve (CN V_3)	Lateral surface of TM, EAC, tragus and pre-auricular area, skin of temporal scalp	Temporomandibular joint, dental pathology, herpes zoster, trigeminal neuralgia
Posterior auricular nerve	Facial nerve (CN VII, nervus intermedius part)	Lateral surface of TM, posterior wall of EAC, conchal bowl	Nose, nasopharynx, soft palate, herpes zoster/Ramsey Hunt syndrome, cerebellopontine angle lesions
Tympanic nerve (Jacobsen's)	Glossopharyngeal nerve (CN IX)	Middle ear and medial surface of the TM	Oropharynx (tonsils and tongue base)
Auricular branch of Vagus nerve (Arnold's or Aldermans)	Vagus nerve (CN X)	Lateral surface of TM, floor of EAC, conchal bowl	Pharynx and larynx
Lesser occipital nerve	Cervical plexus (C2, 3)	Superior part of pinna, postero-lateral scalp	Cervical spine pathology
Greater auricular nerve	Cervical plexus (C2, 3)	Majority of the pinna, skin over parotid and mastoid, angle of mandible, upper lateral neck	Cervical spine pathology

TM = tympanic membrane; EAC = external ear canal

Figure 8.1 Sensory nerve supply to the ear and possible causes of referred pain.

medication should be provided in order to treat underlying infection and inflammation. In cases of severe ear canal oedema and inflammation it may not be possible to adequately examine the medial ear canal and ear drum. Initial therapy with topical drops should be commenced with subsequent follow-up to complete the examination.

History

Otalgia and otorrhoea should never be viewed in isolation. The five cardinal otological symptoms of otalgia, otorrhoea, hearing loss, tinnitus, and dizziness should all be fully explored in the otological history (see Chapters 6, 7, 10 and 11). Facial nerve palsy is a significant concern when related to ear disease and should be investigated and managed as an emergency (see Chapter 10).

In cases of otalgia, the mnemonic 'SOCRATES' can be a helpful aide-memoire to explore salient features of the pain; Site, Onset, Character, Radiation, Associated features, Timing, Exacerbating/relieving factors, and Severity. Otalgia caused by primary otological pathology is usually felt deeply within the ear and is often unilateral

in nature. Sharp, shooting, or fleeting pains may indicate neuralgia. Pain radiating away from the ear may be caused by spreading pathology (such as cellulitis) or referred otalgia/neuralgia. Pain felt below the ear, behind the angle of the mandible, may be related to Eustachian tube dysfunction or referred pain from the sinuses, teeth, pharynx, larynx, or musculoskeletal issues. Pain in front of the ear may be caused by TMJ dysfunction. Insidious-onset, short-duration (hours to days) unilateral pain is most commonly caused by an infective pathology. Chronic pain raises suspicious for chronic OE, chronic otitis media, necrotizing OE, or underlying malignancy. Red flag head and neck symptoms, as well as a history of smoking and high alcohol consumption, should heighten a suspicion of malignancy (see Chapter 46).

Otorrhoea is an objective symptom and sign. The nature of the otorrhoea may give some non-specific hints about underlying pathology. Mucopurulent discharge is associated with acute and chronic infections. An offensive odour to the discharge may be an indication of underlying cholesteatoma. Serous otorrhoea may be produced by inflammatory conditions such as chronic OE, myringitis, and chronic otitis media. Bloody otorrhoea can arise from trauma, acute TM perforation, chronic inflammatory tissue, or tumours. Profuse watery otorrhoea should raise suspicion of an underlying cerebrospinal fluid (CSF) leak and TM perforation.

A past history of ear disease, including childhood infections and operations, may help identify likely pathologies. Medications or conditions that cause immunosuppression (such as diabetes) should always raise concern about atypical or deep-seated infection.

Clinical examination of otalgia and otorrhoea

Examination of the ear is undertaken in a systematic fashion starting with the pinna, mastoid, and surrounding region before progressing to otoscopic, microscopic, or oto-endoscopic evaluation of the ear canal and ear drum. Endoscopic examination of the nasal cavity and nasopharynx should be considered if sinonasal or Eustachian tube pathology is suspected. Examination of the head and neck region, including flexible pharyngolaryngoscopy, is mandatory if there is any possibility of underlying malignancy.

Many acute ear infections are associated with a mild conductive hearing loss. This can be confirmed with tuning forks and clinical tests of hearing. Formal audiometry and tympanometry should be performed in cases where inner ear involvement is suspected and those with non-resolving ear symptoms (see Chapter 5). Dizziness and acute sensorineural hearing loss should be managed appropriately (see Chapters 7 and 11).

Investigation for otalgia and otorrhoea

In cases of otalgia or otorrhoea, a thorough history and examination are usually sufficient to make a clinical diagnosis. Additional investigations are rarely helpful in the initial management of common infective conditions.

Swabbing of ear discharge for microbiological culture and sensitivities is not required in simple infections, as they tend to respond to first-line therapy irrespective of the causative organism. In refractory or recurrent infections, swabs may help identify resistant and atypical organisms thus allowing further tailoring of antimicrobial treatment. Microbiological analysis should be undertaken in cases of possible necrotizing OE. Biopsy of persistent ear canal polyps and granulation tissue should be performed to rule out malignancy.

Fine-cut computed tomography (CT) imaging of the temporal bone provides excellent anatomical definition of the bony structures of the ear and mastoid. This should be considered in cases of suspected temporal bone fracture, CSF otorrhoea, or necrotizing OE. Ossicle, scutum, and mastoid erosion associated with cholesteatoma may be demonstrated.

Magnetic resonance imaging (MRI) of the skull base and neck should be considered in cases of suspected referred otalgia and malignancy. MRI can also supplement CT in the diagnosis of necrotizing OE. Diffusion-weighted MRI scanning is increasingly being used for postoperative surveillance of cholesteatoma and may also be used as an adjunct to diagnosis at initial presentation (see Chapter 19).

Pathology of the external ear

Otitis externa

OE refers to any inflammation of the ear canal; it can be classified as acute or chronic. Acute OE tends to be infective in nature. Chronic OE may be caused by non-resolving acute infection or more dermatological conditions including contact allergy. Differential diagnosis includes foreign body impaction, necrotizing OE, canal wall cholesteatoma, keratosis obturans (KO), or malignancy.

Acute otitis externa

Acute OE often begins with a sensation of irritation, itchiness, fullness, and mild hearing loss. Within a few hours or days this progresses to more overt otalgia and otorrhoea. Infection may spread to cause localized cellulitis of the pinna, periauricular region, and face. On examination the pinna and ear canal may be tender to retraction and speculum insertion. The skin of the ear canal is oedematous and the lumen may be occluded by debris and discharge (**Figure 8.2**).

Risk factors for acute OE include water exposure, wax impaction, narrow ear canals, use of cotton buds and instruments within the ear canal, foreign bodies, trauma, hearing aid use, background chronic OE, immunocompromise, and radiotherapy. It is more common in humid, tropical climates.

Bacterial infection is the principal cause of acute OE in 98% of cases (figures from North America), with *Pseudomonas aeruginosa* and *Staphylococcus aureus* the most common pathogens. Swabbing of the ear canal for microbiological culture is not required in simple cases. Culture should be considered in refractory, recurrent, or atypical cases, especially in immunocompromised patients or if necrotizing OE is suspected. Mixed infections or infections in which no pathogenic organism can be isolated are not uncommon.

Treatment of acute OE involves aural toilet, topical medication, analgesia, and avoidance of contributing factors. In cases of severe canal oedema and occlusion, the use of a temporary expanding otowick should be considered. This should be combined with the use of aqueous ear drops; oil-based drops do not expand otowicks. Otowicks can be removed after 2–7 days and replaced if required.

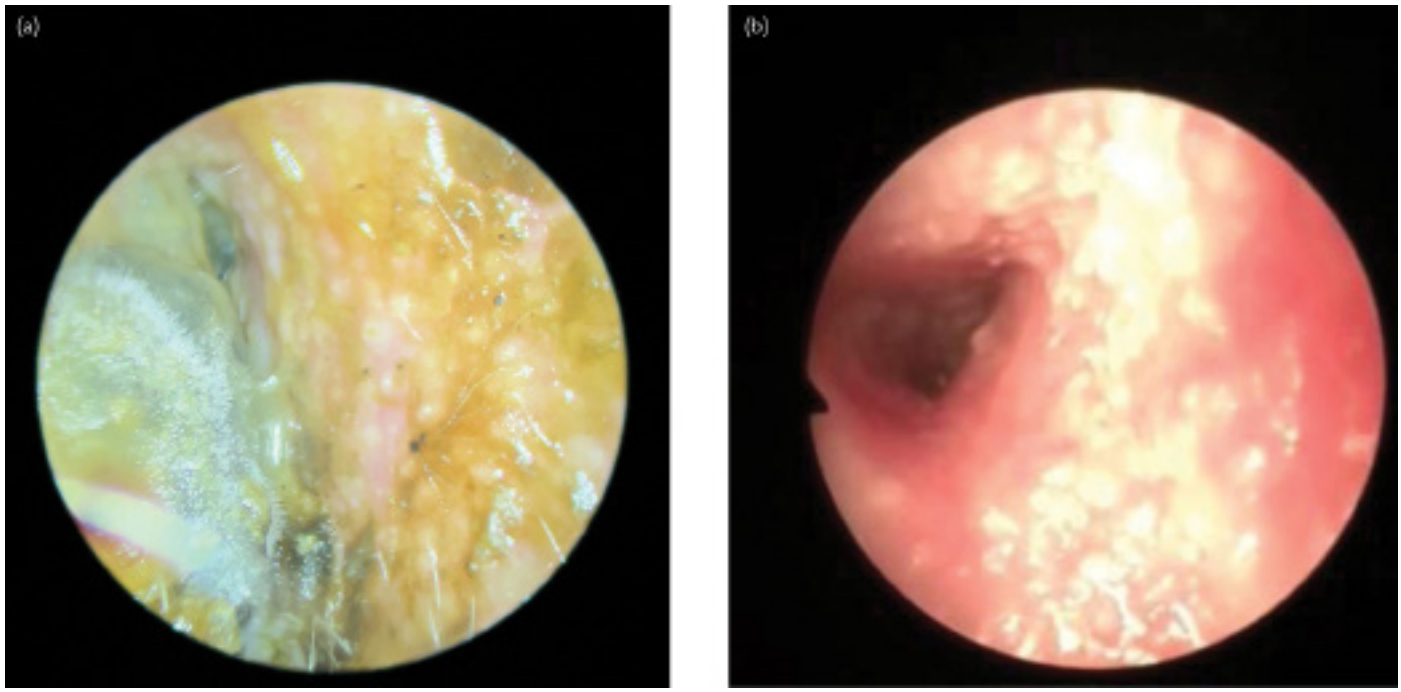


Figure 8.2 Otoendoscopic images of acute OE; (a) Mixed bacterial and fungal debris occluding the external ear canal in acute otitis externa; (b) External ear canal oedema and erythema in otitis externa, narrowing lumen.

Mild cases of OE may respond to topical antiseptic or acidifying agents (such as acetic acid, boric acid, aluminium acetate, phenol, or alcohol). Steroid drops may also be beneficial. Combination antibiotic and steroid drops are the most effective treatment for OE. They are usually prescribed for 7–10 days. The choice of drop may be guided by local prescribing policies, specific microbiological results, or drop availability. There is controversy regarding the use of potentially ototoxic aminoglycoside-containing drops, especially in the presence of a TM defect (see 'Controversies'). Reviews of the background evidence and expert opinion concludes that the need to treat acute infection outweighs the small ototoxic risk, although patients should be warned of potential side effects and advised to stop if dizziness or sensorineural hearing loss is experienced. The addition of systemic antibiotic is indicated for infections spreading beyond the ear canal, such as furunculosis, cellulitis, perichondritis and necrotizing OE (see below).

Hearing aids may cause skin irritation or moisture-trapping within the medial ear canal. In cases when this is thought to contribute to refractory or recurrent OE, alternatives such as bone conduction hearing aids can be considered (see Chapter 16).

Recurrent infections can predispose the formation of circumferential scar tissue and canal stenosis. This may eventually 'burn out' with formation of a false fundus and medial canal fibrosis. Surgical canalplasty can be considered for cases of recurrent OE associated with narrow ear canals, progressive canal stenosis, or bony exostoses.

Otomycosis

Otomycosis (also known as fungal OE) may occur primarily or as a secondary infection following treatment with topical antibiotics. It is more common in tropical climates. Fungal colonization is common in cases of chronic OE, inflamed mastoid cavities, and in association with hearing aid use.

Presenting symptoms and signs are similar to acute OE. *Candida* or *Aspergillus* species are the most frequent fungal pathogens isolated. Fungal hyphae may be evident on otoscopy and can indicate the causative organism: white hyphae or discharge in *Candida albicans*, black hyphae in *Aspergillus niger*, and yellow hyphae in *Aspergillus flavus* (**Figure 8.3**).

Treatment is similar to acute OE, with thorough microsuction of mycotic debris, topical drops, analgesia, and avoidance of precipitating factors. Specific topical antifungal preparations such as clotrimazole, nystatin, or clioquinol may be used. Fungal OE also responds to antiseptic or acidifying drops such acetic acid and aluminium acetate. More prolonged treatment is often required in order to eradicate the fungal disease (2–3 weeks).

Furunculosis

Furunculosis is a localized form of acute OE caused by infection of a hair follicle in the lateral, cartilaginous part of the ear canal. Infection may produce a localized abscess and spread to surrounding tissue as cellulitis.

Otalgia is the dominant symptom. Scant discharge may be produced by a ruptured abscess or granulation tissue. Furunculous can be hard to distinguish from acute OE. Manipulation of the pinna and insertion of an otoscope speculum into the ear canal are exquisitely tender, often disproportionately to the examination findings. Point tenderness at the tip of the speculum and identification of a localized, oedematous, and inflamed area may be seen. A discharging abscess sinus or granulation tissue are highly suggestive for furunculosis. The medial ear canal and TM are relatively normal.

The commonest causative organism is *Staphylococcus aureus*. Treatment is with antibiotics and analgesia. Mild cases may be treated with oral anti-staphylococcal agents including flucloxacillin,

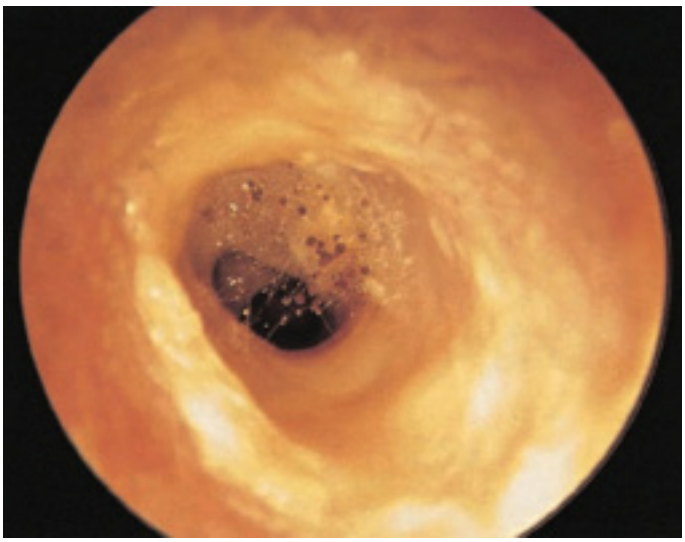


Figure 8.3 Otoendoscopic image of fungal OE (otomycosis) showing fungal hyphae of *Aspergillus niger*.

Reproduced from Carney S. Otitis externa and otomycosis. In: Watkinson JC, Clarke RW, eds. *Scott-Brown's Otorhinolaryngology and Head and Neck Surgery*, Vol. 2. 8th ed. CRC Press; 2018, with permission from CRC Press.

ciprofloxacin, clindamycin, macrolides, or cephalosporins. More severe spreading cellulitis may require intravenous antibiotics. If present, a localized abscess can be drained via aspiration or incision. Topical antibiotics may be given for associated acute OE or when the abscesses have discharged.

Chronic otitis externa

Chronic OE is characterized by long-term inflammation of the ear canal. There is no widely accepted time frame in which acute OE becomes chronic OE. As a guide, inflammation lasting more than 3 months or four attacks of acute OE within a year should be considered chronic OE.

Chronic OE tends to be less painful in nature than acute OE. Presenting symptoms include itchiness, irritation, discharge (often serous or watery), repeated blockage, and conductive hearing loss.

Chronic OE maybe the result of non-resolving acute OE and otomycosis, especially when resistant or atypical microbes are present. Infection may be perpetuated by ongoing water exposure, instrumentation of the ear canal, hearing aid use, and immunosuppression. Microbiological analysis of the ear canal discharge with cultures and sensitivities may help guide antimicrobial treatment. In cases of recurrent acute OE, a thorough review of potential precipitating factors should be undertaken as well as a screening for underlying chronic pathology. Differential diagnoses such as necrotizing OE, canal wall cholesteatoma, KO, and malignancy should be considered. Biopsy of ear canal polys and granulation tissue should be undertaken to rule out malignancy (see Chapter 18).

Chronic OE may be due to an underlying dermatological condition and is associated with more widespread psoriasis and atopic dermatitis (eczema). Acute-on-chronic infective exacerbations are common. Allergic contact dermatitis related to eardrop ingredients should be considered and investigated with patch testing if necessary.

The mainstay of treatment for chronic OE involves avoidance of irritants, water, hearing aids, and use of cotton buds. Intermittent use of steroid, acidifying, or antiseptic drops should be considered. Waterproof ear plugs should be used when showering, bathing, washing hair, or swimming in order to keep the ear canal dry and reduce exposure to potential irritants. Care should be taken to avoid ear plugs being pushed deep into the ear canal. A cotton wool ball, soaked in petroleum jelly and placed in the conchal bowl, provides a cheap, effective, and disposable option. Custom-moulded silicone ear plugs are effective, especially when prolonged use is required. A swim-cap or headband may be required to hold earplugs in place during water sports.

Necrotizing otitis externa

Necrotizing OE (also called malignant OE or benign necrotizing OE) principally affects diabetic, immunocompromised, or elderly patients. Infection spreads from the skin of the ear canal to underlying bone, resulting in a spreading osteitis of the bony ear canal, temporal bone, and skull base. The diagnosis of necrotizing OE should be considered in susceptible patients with non-resolving infection, intractable pain, facial nerve palsy, or ear canal granulations/polyps (see Chapter 24).

Perichondritis and pinna cellulitis

Perichondritis and pinna cellulitis are inflammatory conditions of the pinna that are usually infective in nature. Both present with otalgia, erythema, and oedema of the pinna. Perichondritis specifically refers to inflammation of the connective tissue layer surrounding cartilage; the ear lobe is thus spared. In contrast, cellulitis spreads within subcutaneous connective tissue and may affect all areas of the pinna. Inflammation may spread to the periauricular area and face.

Perichondritis and pinna cellulitis may occur in isolation or secondary to OE or furunculosis. Primary perichondritis and pinna cellulitis may be caused by pinna trauma, bites, piercings, and broken skin from dermatological conditions.

Treatment is with oral or parenteral antibiotics. Staphylococcal and streptococcal species are the most common causative organisms. Associated acute OE should be treated as described previously.

In the presence of OE, the choice of systemic antibiotics should be modified to include cover for *Pseudomonas aeruginosa*. Infected piercings should be removed and local abscesses drained surgically.

Relapsing perichondritis is an autoimmune condition affecting cartilage at multiple anatomical sites including the pinna, nose, ribs, respiratory tract, eyes, and joints. It may be associated with systemic autoimmune diseases such as rheumatoid arthritis, vasculitides, systemic lupus erythematosus, scleroderma, and Sjögren's syndrome. Referral to rheumatology colleagues should be considered, so that a detailed multisystem history, examination, and blood test for autoimmune conditions can be undertaken. Treatment involves immunosuppressive medications. Severe or recurrent perichondritis may lead to cartilage hyperplasia or replacement of necrotic cartilage with thick, fibrous scar tissue, a condition known as cauliflower ear (**Figure 8.4**).

Keratosis obturans

The ear canal is lined by stratified squamous epithelium. This migrates laterally from the ear drum by approximately 1 mm every 1–2 weeks. KO is thought to be a failure of this migratory system leading to build-up of keratinous debris in the medial ear canal. This presents as recurrent blockage, hearing loss, and OE. Over time, keratin build-up and infection may cause remodelling of cortical bone with a resultant dilated medial ear canal. Focal ulceration of the canal skin may result in bony sequestra and is thought to be one potential mechanism of canal wall cholesteatoma formation (see Chapter 19). KO is usually unilateral, but rarely can affect both ears. Secondary KO may occur after trauma or surgery involving the ear canal.

Diagnosis is confirmed by the appearance of a dilated medial ear canal and the need for repeated microsuction of keratinous debris. This can be investigated by fine-cut CT of the temporal bone, especially if invasive canal wall cholesteatoma or necrotizing OE are suspected. Infective exacerbations are treated in a similar manner to OE. Persistent granulation tissue should be biopsied to rule out malignancy.

KO may be managed conservatively with regular aural toilet to remove keratinous debris and reduce the frequency of infections and hearing loss. Bony or soft tissue meatoplasty may be considered for cases when the ear canal becomes irregular, hard to clean, or stenotic. Blind sac closure of the ear canal may be necessary for severe unremitting cases.

Figure 8.4 Relapsing perichondritis leading to cauliflower ear.

Ear canal malignancy

Malignancy involving the ear canal is rare. Cancers may be primary (usually squamous cell carcinoma), due to local spread (from scalp skin or parotid) or metastatic in nature. Presentation is typically with pain, discharge, bleeding, or facial nerve palsy. Differential diagnosis includes necrotizing OE. See Chapter 18 for more details.

Pathology of the tympanic membrane and middle ear

Acute otitis media

AOM is one of the commonest infectious conditions seen in children. Over 80% of children will have experienced at least one episode of AOM by their third birthday. For the diagnosis and management of paediatric AOM, including recurrent AOM, see Chapter 84.

AOM in adults is less common than in children. It may occur in combination with an upper respiratory tract infection or as a separate entity. It is often associated with chronic eustachian tube dysfunction or otitis media with effusion (see Chapter 9). The aetiology of AOM involves Eustachian tube obstruction leading to dysregulation of middle ear ventilation, inflammation of the middle ear mucosa, and otitis media with effusion. Viral and bacterial infection of the effusion leads to AOM. The microbiology of adult AOM is broadly similar to paediatric AOM. Common pathogens include *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Moraxella catarrhalis*.

The clinical presentation is of pain, hearing loss, and fever. The TM appears hypervascular, inflamed, and bulging. Acute rupture of the ear drum produces otorrhoea, often with a sudden improvement of otalgia. Treatment guidelines for adult AOM are often extrapolated from paediatric studies. Initial management of AOM in adults involves analgesia, especially in mild cases. For more persistent cases oral antibiotics are considered.

Complications of AOM include mastoiditis, facial palsy, suppurative labyrinthitis, petrous apicitis, extratemporal abscess, sigmoid sinus thrombosis, and intracranial infection. Complications of AOM should be treated urgently given the potential morbidity and mortality. Treatment consists of intravenous antibiotics and consideration should be given to surgical drainage of the infection with myringotomy, ventilation tube, and cortical mastoidectomy (in cases involving mastoiditis). For more details see Chapter 84.

Recurrent acute otitis media

Recurrent AOM is a common presentation in paediatric ear, nose, and throat clinics. This typically occurs between the ages of 6 and 24 months and is usually self-resolving. The frequency and severity of infections may be improved with a low-dose prophylactic oral antibiotic or grommet insertion. For more details see Chapter 84.

Recurrent AOM in adults is less common. It is usually associated with chronic Eustachian tube dysfunction, otitis media with effusion, or immunosuppression. Patients with recurrent acute otalgia should ideally be reviewed by an otolaryngologist during acute attacks in order to confirm the diagnosis of AOM. Recurrent acute otalgia in the absence of discharge can often be mislabelled as AOM. In addition to a thorough history and otoscopic examination, the

nose and postnasal space should be examined with nasendoscopy to screen for sinonasal pathology.

Audiometry and tympanometry in between infections is helpful in order to identify otitis media with effusion or Eustachian tube dysfunction. Specific, validated Eustachian tube dysfunction symptom questionnaires may help support the diagnosis (see Chapter 9).

MRI imaging may demonstrate anatomical causes of Eustachian tube obstruction in the postnasal space or skull base. A review of the patient's immune competency should be undertaken especially in the presence of additional non-otological infections.

Most research regarding the management of recurrent AOM has been undertaken in children. Many of the management strategies for adult AOM are extrapolated from the paediatric population. Management options are dependent on the frequency and severity of AOM, symptoms present in between attacks (such as chronic hearing loss or aural fullness/pressure), and the general health of the patient. Contributing factors such as chronic rhinosinusitis, immunosuppression, Eustachian tube dysfunction, adenoidal hypertrophy, smoking, and masses obstructing the Eustachian tube should all be addressed. In cases of recurrent AOM, treatment of acute infections with oral antibiotics (such as amoxicillin, co-amoxiclav, or clarithromycin) should be considered in order to ensure complete resolution of each infection. For frequent and confirmed recurrent AOM, low-dose prophylactic antibiotics may be considered. For cases associated with otitis media with effusion, the insertion of ventilation tubes (grommets) or T-tubes may reduce the frequency and severity of AOM. This needs to be balanced with the risk of ventilation tube-associated otorrhoea and resultant TM chronic perforation, which is likely to be higher in this subset compared to the paediatric population (where Eustachian tube immaturity often improves before the ventilation tube extrudes). Eustachian tube ballooning offers the theoretical possibility of improving underlying Eustachian tube function and recurrent AOM. However, research evidence is currently lacking in this specific cohort.

Chronic otitis media

Chronic otitis media is a broad term that describes long-term changes to the shape and function of the TM. This includes retraction pockets, perforation, and cholesteatoma.

Chronic otitis media typically presents with recurrent otorrhoea and hearing loss. Otalgia is variable. This may result in delayed presentation and diagnosis. The diagnosis and management of retraction pockets, perforations, and cholesteatoma is described elsewhere (see Chapters 19 and 22).

Myringitis

Myringitis is characterized by inflammation of the lateral layers of the TM. The aetiology of myringitis is unclear.

Acute bullous myringitis is thought to be an infective pathology, resulting in the formation of fluid-filled vesicles or bullae between the middle fibrous layer and outer squamous layer of the TM. This may be concurrent with AOM and otitis media with effusion, or more rarely as a solitary entity with a normal middle ear. In keeping with AOM, it is more common in children. Acute bullous myringitis presents with severe otalgia and fever. Hearing loss and dizziness may be present, which tend to recover after resolution of the infection. Treatment consists of oral antibiotics and analgesia. Steroids may be given if there is evidence of inner ear involvement with either sensorineural hearing loss or vertigo.

Figure 8.5 Otoendoscopic image of granular myringitis affecting the posterosuperior aspect of the right TM following tympanoplasty. Spot bleeding is caused by contact during microsuction.

Granular myringitis is a more chronic condition caused by instability of the lateral, squamous layer of the TM. The underlying middle ear is usually normal. Granular myringitis may follow surgery, trauma, or infection and often affects areas of the TM that are perforated, atelectatic, sclerotic, or avascular. A chronic inflammatory reaction results in the formation of surface granulation tissue. This may be focal or diffuse in nature and can extend to the adjacent medial ear canal (**Figure 8.5**). The clinical presentation is one of watery/serous otorrhoea which may be painless in nature. Mucopurulent otorrhoea suggests an additional infective element. Florid granulation tissue or polyps produce bloody otorrhoea. Investigation with CT or diffusion-weighted MRI may help rule out underlying middle ear pathology or hidden cholesteatoma.

Treatment of granular myringitis involves regular aural care, avoidance of water ingress, and topical therapy. Combination antimicrobial and steroid drops are used to treat any infective element. This should be guided by microbiological culture and sensitivities. This may be followed by an extended course of steroid drops as an attempt to settle down long-term granulation tissue and encourage re-epithelization (typically 6–8 weeks). A number of studies have described successful treatment of granular myringitis with topical antiseptics including aluminium acetate, acetic acid, phenol, hydrogen peroxide, and alcohol. Persistent granulation tissue can be cauterized with topical silver nitrate or debrided using cold steel or KTP laser techniques. Surgical tympanoplasty to excise and graft the myringitic ear drum segments may be required. Case series have indicated a high success rate of over 95%, but this needs to be balanced against surgical risks during an informed consent process.

Cerebrospinal fluid otorrhoea

CSF otorrhoea results from a defect in dura and bone of the temporal skull base. CSF flows into the mastoid air cells and middle ear with subsequent discharge thorough a TM defect. In the presence of an intact ear drum, CSF may flow though the Eustachian tube resulting in CSF rhinorrhoea. Postoperatively, CSF can leak from surgical wounds.

CSF otorrhoea may occur via congenital defects or be the result of surgery, temporal bone fractures, erosive temporal bone pathology, and intracranial hypertension. A painless, watery otorrhoea is noted exacerbated by rises in intracranial pressure associated with coughing, straining, bending forwards, or supine position. Diagnosis is confirmed by analysis of the discharging fluid for the presence of asialotransferrin/beta-2-transferrin. CT and MRI scanning of the temporal bone may localize the defect and identify associated pathology. Prolonged leak increases the risk of meningitis and requires conservative or operative management depending on the nature of the leak.

For further detail on the diagnosis and management of CSF leaks, see Chapter 45.

Otalgia

Otalgia may originate from primary ear disease, adjacent structures, or via referred neuralgia. Many of the otorrhoea-producing conditions described above also produce otalgia. In mild or early stages of these ear conditions, otalgia may be the only complaint. Otorrhoea may be noted on examination of the ear but not reported by the patient. A focused history and thorough examination of the ear should be undertaken to identify primary ear pathology. A number of conditions may produce otalgia without otorrhoea or apparent abnormalities on ear examination.

Herpes zoster oticus

Herpes zoster oticus (HZO) is an infection of the ear caused by the varicella zoster virus. When associated with facial palsy, the condition is also known as Ramsey Hunt syndrome (see Chapter 10). HZO presents with acute and often severe otalgia. The diagnosis can be difficult to make in the early stages, until a few hours or days later when a vesicular rash is noted around the ear, face, mouth, or tongue. The severity of the rash can range from a few vesicles to a widespread mucocutaneous crop. Distribution of the rash is in the dermatomes of nerves that supply sensation to the ear and may include the tongue and soft palate. Serological testing for varicella zoster virus may show evidence of previous infection but is rarely helpful in diagnosing acute cases (a significant rise in convalescent antibody titre is supportive of recent acute infection).

HZO is caused by reactivation of the varicella zoster virus that lies dormant in sensory nerve root ganglia including the geniculate ganglion. In addition to facial nerve involvement (Ramsey Hunt syndrome), HZO can be associated with vertigo, sensorineural and conductive hearing loss, dysgeusia, and dry eyes or mouth.

Treatment of HZO involves analgesia and antiviral mediation such as aciclovir, valaciclovir, or famciclovir. Early treatment with antivirals is associated with more rapid resolution of pain and rash; this has also been shown to reduce the risk of persistent post-herpetic neuralgia. Facial palsy or sensorineural hearing loss should prompt additional treatment with oral steroids (see Chapters 10 and 11).

Otic barotrauma

Otic barotrauma is produced by maladaptation to changes in environmental pressure. This can be a sudden pressure change (decompressive events, skydiving, explosions, Valsalva manoeuvre, blunt 'slap' injury to the ear, or high-diving) or gradual pressure change (such as air flights, scuba diving, and hyperbaric therapy).

When occurring gradually, otic barotrauma starts with a sensation of blockage or pressure in the ear associated with a desire to equalize pressure. If middle ear pressure fails to equalize via normal Eustachian tube function, symptoms progress to otalgia. A mild conductive hearing loss is caused by reduced compliance of the ear drum, oedema of middle ear mucosa, formation of a middle ear effusion, or eventual haemotympanum. Perforation of the ear drum causes sudden acute pain and otorrhoea, often with resolution of discomfort and pressure. Vertigo and sensorineural hearing loss are indicative of barotraumatic perilymph fistula. More severe conductive hearing losses are observed in cases of ossicular damage. The symptoms and signs related to middle ear barotrauma can be classified according to a modified Teed grading (Table 8.2).

Otic barotrauma usually requires symptomatic treatment: analgesia, oral or topical nasal decongestants, and avoidance of further unnecessary baro-challenges. Severe acute symptoms may be managed with myringotomy. Perforations of the TM are managed conservatively with observation or topical drops. Surgical myringoplasty is indicated for symptomatic non-healing perforations.

For management of chronic Eustachian tube obstruction, see Chapter 9.

Referred otalgia

Pain localized to the ear but produced by a distant pathology is known as referred otalgia. The ear has a complex innervation from multiple cranial and cervical nerves (Figure 8.1). In cases where otalgia is the presenting complaint, and in the absence of other otological symptoms or signs, a careful evaluation of possible sources of referred otalgia should be undertaken. Referred pain may have a characteristic neuralgic quality often described as sharp, stabbing, burning, or transient in nature. However, referred pain can also mimic primary otalgia.

Table 8.2 Modified Teed grading for barotrauma

Barotrauma grade	Sign and symptoms
Grade 0	Symptoms with no ontological signs of trauma
Grade 1	Diffuse redness and retraction of the TM
Grade 2	Grade 1 plus slight haemorrhage within the TM
Grade 3	Grade 1 plus gross haemorrhage within the TM
Grade 4	Dark and slightly bulging TM due to free blood in the middle ear (a fluid level may also be present)
Grade 5	Free haemorrhage into the middle ear, TM perforation with blood visible in the external auditory canal

Malignancy

Malignancy of the upper aerodigestive tract is a relatively rare cause of referred otalgia but should always be considered given the significance of the diagnosis and adverse outcome associated with delayed diagnosis. The commonest site of malignancy causing otalgia is the tongue base and oropharynx, via irritation of the glossopharyngeal nerve. This nerve also supplies the middle ear via the tympanic branch (Jacobson's nerve). More distal tumours affecting the larynx can stimulate the vagus nerve which shares innovation with the ear canal and lateral surface of the TM via Arnold's nerve. Sinonasal, oral, and postnasal space tumours may stimulate the trigeminal nerve that co-innervates the external ear via the auriculotemporal nerve.

A focused history should be taken to screen for aerodigestive tract symptoms including sore throat, dysphagia, odynophagia, dysphonia, neck masses, and weight loss. Risk factors for malignancy such as smoking and alcohol consumption, previous malignancy, and radiotherapy should be documented. A complete head and neck examination is performed including neck palpation, oral cavity examination, and flexible nasal endoscopy of the upper aerodigestive tract (see Chapter 46).

MRI imaging of the skull base and neck should be considered in cases of unexplained otalgia (see Chapter 18).

Temporomandibular joint dysfunction

The TMJ sits directly in front of the canal. Degenerative or inflammatory disease within the joint as well as the muscles of mastication can produce referred otalgia. There are no pathognomonic features of TMJ dysfunction; tenderness on palpation of the joint, tenderness of the muscles of mastication, crepitus within the joint, and trismus are suggestive features. TMJ dysfunction may be caused by, or result in, bruxism. A history of teeth grinding at night with resultant dental wear pattern on examination (flattening of the occlusal surfaces) may be evident. Given the lack of a standard definition, it is estimated that between 5% and 70% of the population may suffer from TMJ dysfunction at some point.

Management of TMJ dysfunction initially involves reassurance, analgesia, soft diet, and rest. Relaxation therapies may help with muscular tension. Nocturnal occlusal splints provide occlusal stability and may reduce nocturnal bruxism. Referral to maxillofacial specialist may be indicated for severe and non-resolving cases, especially if associated with mechanical dysfunction of the TMJ.

Dental disease

Inflammation, infection, and degeneration of the teeth or gums may be poorly localized and felt as otalgia. History and examination features consistent with poor dentition should prompt a dental review.

Neuralgia

Neuralgic otalgia may be idiopathic or be secondary to infection, surgery, trauma, and HZO. The distribution, character, and timing of the pain may be variable. In some cases, otalgia may have a typical neuralgic quality with sharp or stabbing pains. Pain is usually localized deep within the ear and may radiate to the adjacent scalp, neck, or face. Examination of the ear is normal. Neuralgia may be associated with numbness, paraesthesia, or hypersensitivity of the skin to touch.

Treatment consists of reassurance and analgesia. Specific neuralgic medication may be trialled, such as carbamazepine, amitriptyline, pregabalin, and gabapentin. Trigeminal neuralgia may be investigated with MRI scans to rule out neurovascular conflict or compressive lesions along the course of the nerve. Guided injection, radiofrequency ablation, stereotactic radiosurgery, or microvascular decompression of the trigeminal nerve roots can be considered in severe and refractory cases. Tympanic neurectomy may be considered for persistent glossopharyngeal otic neuralgia.

Controversies

The use of eardrops in the presence of an open middle ear

Aminoglycoside eardrops have been used to treat otological infections for many years, with side effects being rare. However, aminoglycosides are known to be ototoxic. This effect is deliberately harnessed in the treatment of vestibular conditions such as Ménière's disease (see Chapter 14). The risk of unintended ototoxicity when treating ear infections may be higher in the presence of a TM defect such as a ventilation tube or TM perforation. In clinical practice it may be difficult to confirm integrity of the TM, especially if there is profuse otorrhoea and ear canal oedema.

Many aminoglycoside eardrop manufacturers specifically advise against the use of eardrops in the presence of an open middle ear, presumably as a medicolegal consideration. This must be balanced against the pressing clinical need to treat the infection, which in itself may damage the inner ear.

A number of national specialist bodies have reviewed the concerns and evidence regarding the use of aminoglycoside eardrops in the presence of a TM defect. Most have concluded the need to treat infection (including minimizing the risk of the infection itself causing inner ear damage) outweighs the small risk of drop-related ototoxicity. Patients should be warned to stop the drops if dizziness or hearing loss occurs. A pretreatment hearing test should ideally be undertaken, but in clinical practice (especially primary care) this may not be feasible.

Quinolone-containing eardrops are now routinely available. These do not appear to have an ototoxic effect and have become more widespread in their use. There is concern about increasing rates of quinolone resistance in pseudomonal ear infections. This can be especially problematic when dealing with severe pseudomonal infection such as necrotizing OE. Some studies suggest that TM healing may be reduced following quinolone drop application.

Tips and pearls

- In cases of otorrhoea, meticulous microsuction is key to establishing diagnosis and treating infection.
- In cases of otorrhoea, topical therapy should be considered a first-line treatment. Oral or parenteral antibiotics are usually considered for refractory, deep-seated, or spreading infection.
- Generic advice to keep the ear canal dry and avoid instrumentation should be undertaken in all cases of otorrhoea.

- In cases where it is not possible to fully examine the ear drum (due to debris, discomfort, oedema of the ear canal, or patient non-compliance with aural cleaning), initial treatment with antibiotic and steroid drops should be commenced. Subsequent follow-up to complete the microsuction and reassess the ear should be arranged to rule out underlying ear canal and TM pathology.

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SECTION 2

Otology and audiology

5. **Clinical and audiological assessment of hearing in adults** *27*
   Gemma Crundwell
6. **Tinnitus and disorders of sound tolerance** *37*
   Don McFerran
7. **Vertigo** *45*
   Peter Rea and Joel A. Goebel
8. **Otalgia and otorrhoea** *59*
   Philip J. Clamp
9. **Aural fullness** *69*
   Holger H. Sudhoff
10. **The facial nerve** *72*
    Richard M. Irving
11. **Sensorineural hearing loss** *77*
    Ted A. Meyer
12. **Conductive (or mixed) hearing loss with an intact tympanic membrane** *81*
    Paul Merkus and Thadé Goderie
13. **The external ear** *92*
    Pablo M. Reyes Burneo and Miguel Vaca González
14. **Ménière's disease and vestibular migraine** *105*
    Louisa Murdin
15. **Benign paroxysmal positional vertigo** *112*
    Jeffrey D. Sharon and Danielle Gillard
16. **Audiovestibular implants** *119*
    Neil Donnelly and Patrick Axon
17. **Noise-induced hearing loss** *125*
    Peter Webber
18. **Lateral skull base disease** *131*
    J. Walter Kutz Jr and Daniel E. Killeen
19. **Cholesteatoma** *141*
    Neil Bailie and Colin Leonard
20. **Otosclerosis and stapes surgery** *148*
    Christopher Aldren
21. **Ossiculoplasty** *154*
    Christopher Aldren
22. **Tympanoplasty** *160*
    Matthew P.A. Clark
23. **Temporal bone trauma** *166*
    Nicola Quaranta and Vito Pontillo
24. **Necrotizing otitis externa** *176*
    Emma Stapleton and Glen Watson

5

Clinical and audiological assessment of hearing in adults

*Gemma Crundwell**

Anatomy and physiology of the ear and auditory pathways

The ear is the organ of hearing and balance. It is usually described as having three parts, the outer, middle, and inner ear (Figure 5.1).

The outer ear comprises the pinna and external auditory meatus. The pinna (auricle) is the fleshy outer part of the ear, composed of skin-covered elastic cartilage. It is an irregular-shaped concave ovoid with ridges and folds that are used as anatomical landmarks. While commonly considered a rather unimportant part of the auditory system, the pinna does in fact significantly modify the incoming signal, predominantly in the high frequencies above 4 kHz. These pinna cues are particularly important when listening monaurally, providing localization cues to aid identification of sound elevation and front–back distinction (Figure 5.2).

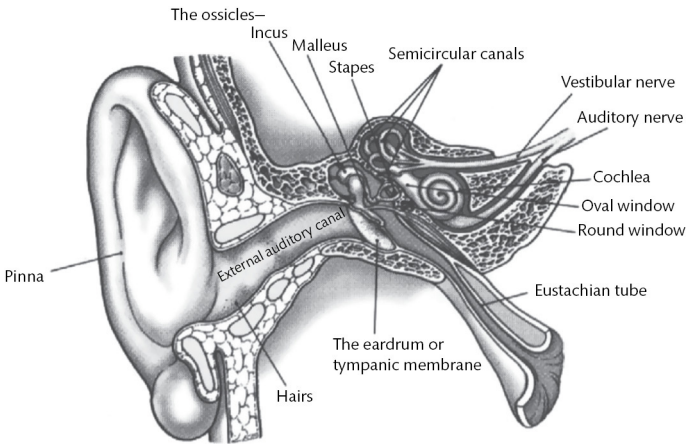

Figure 5.1 Anatomy of the ear.

Reproduced from Corbridge R, Steventon N. *Oxford Handbook of ENT and Head and Neck Surgery*. 3rd ed. Oxford University Press; 2019, with permission from Oxford University Press.

The concha of the pinna forms the entrance to the external auditory meatus (ear canal) leading to the tympanic membrane. The ear canal is divided into two parts: the outer one-third of the ear canal is cartilaginous and the inner two-thirds are bony. It is lined with skin containing cerumen (wax) and sebaceous glands which lubricate the canal and help keep it free of debris and foreign objects. It has a S-shape, running first anterosuperiorly, then posterosuperiorly and finally anteroinferiorly. In order to visualize the tympanic membrane the ear canal must be straightened by pulling the pinna up and backwards. The auricular branch of the vagus nerve, sometimes referred to as Arnold's nerve, innervates the posteroinferior section of the tympanic membrane, posterior section of skin lining the ear canal, and a small patch of skin behind the pinna. Stimulation of this nerve such as through an aural impression can trigger a cough reflex in some individuals.

The tympanic membrane (ear drum) forms the anterior–inferior and part of the posterior wall of the external auditory meatus, sitting at an angle of approximately 55°. The tympanic membrane is a thin, concave elliptical membrane formed of four layers. The outermost layer is continuous with the skin of the ear canal and the inner layer is continuous with the mucous membrane of the middle ear. The superior portion of the tympanic membrane, the pars flaccida, only contains these two layers, whereas the inferior pars tensa contains two additional fibrous layers, rich in collagen. Attached to the tympanic membrane is the manubrium of the malleus, the tip of which corresponds to the umbo. Sound waves travel down the ear canal and cause the tympanic membrane to vibrate. These vibrations are transmitted through the manubrium of the malleus into the middle ear (Figure 5.3).

The middle ear (tympanic cavity) is an air-filled space comprising the ossicles, Eustachian tube, and stapedius and tensor tympani muscles. The lateral wall of the middle ear is formed by the tympanic

* Gemma Crundwell and the editors of the *Oxford Textbook of Otolaryngology* would like to dedicate this chapter to Professor David Baguley. David sadly passed away during the production of this textbook. David was instrumental in overseeing the development of this book chapter. Our thoughts and best wishes remain with David's family.

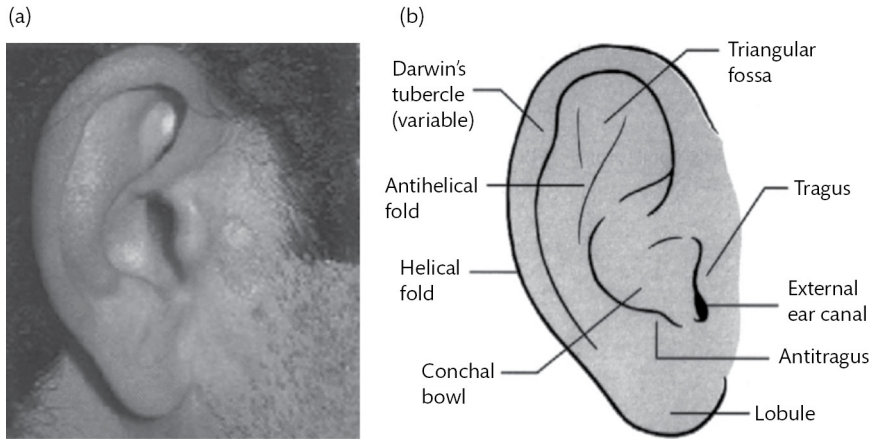

Figure 5.2 Pinna.

Reproduced from Corbridge R, Steventon N. *Oxford Handbook of ENT and Head and Neck Surgery*. 3rd ed. Oxford University Press; 2019, with permission from Oxford University Press.

membrane. Opposite this on the medial wall is the promontory of the basal turn of the cochlea and the oval window. The roof of the middle ear is formed by the tegmen tympani, separating the middle ear from the middle cranial fossa, and the inferior wall separates the middle ear cavity from the jugular bulb. The anterior wall contains the opening to the Eustachian tube and canal for the tensor tympani as it joins the tensor veli palatini. The Eustachian tube links the middle ear with the nasopharynx, equalizing the air pressure on both sides of the tympanic membrane and providing drainage. Sound is transmitted most efficiently when the pressure is equal on both sides of the tympanic membrane. The Eustachian tube is normally closed but is opened by actions including swallowing and yawning; there is a convention that the Eustachian tube is pictorially depicted in the open state. There are three ossicles in each ear: malleus, incus, and stapes (colloquially known as the hammer, anvil, and stirrup) which are collectively referred to as the ossicular chain. These are the smallest bones in the human body. The ossicles are suspended in the middle ear via a series of ligaments, tendons of the middle ear muscles, and attachments to the tympanic membrane and oval window. The middle ear contains two muscles: the tensor tympani and stapedius muscles. The tensor tympani joins onto the top of the manubrium of the malleus, pulling the malleus anteromedially when contracted. The stapedius muscle inserts on the posterior aspect of the neck of the stapes, pulling the stapes posteriorly when contracted. The stapedius muscle in particular contracts when exposed to loud sounds—known as an acoustic reflex, reducing transmission of sound through the middle ear, particularly low frequency. This reflex is commonly thought to prevent damage to the delicate structures of the cochlea, but it is too slow to provide protection against impulse sounds such as gunshots. The two other functions suggested for the reflex is to reduce the audibility of self-generated sounds and to minimize the upward spread of masking by attenuating lower-frequency sounds. The major function of the middle ear is to ensure efficient transfer of sound from the air to the fluid of the cochlea. The middle ear acts as an impedance-matching transformer without which the vibration of sound in air would not efficiently penetrate the denser fluid-containing structures of the cochlea. This is accomplished mainly through the difference in effective areas of the tympanic membrane and oval window (ratio 20:1), and to a smaller extent the lever action of the ossicles.

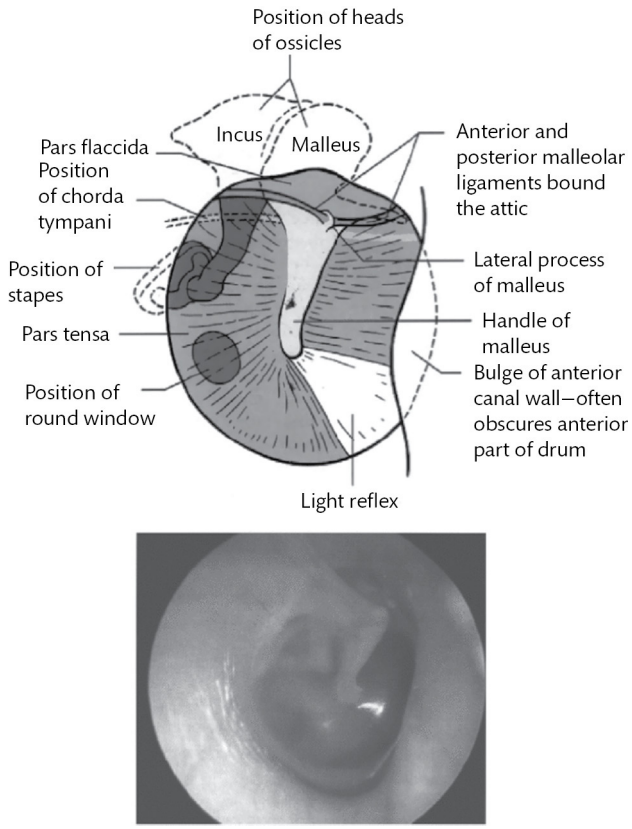

Figure 5.3 Tympanic membrane.

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The inner ear includes the sensory organs of hearing and balance. The inner ear sits within the petrous portion of the temporal bone, in a complex cavity known as the osseous labyrinth. Within this bony labyrinth sits the membranous labyrinth. The inner ear is divided into three sections: the cochlea, vestibule, and semicircular canals.

The cochlea is a fluid-filled, spiral-shaped structure, which could sit upon a UK £1 coin. It is divided along its length by Reissner's membrane and the basilar membrane into three chambers: the scala

vestibuli and scala tympani which are filled with perilymph (rich in potassium ions), and the scala media which is filled with endolymph (rich in sodium ions, and a product of perilymph). At the apical end is the helicotrema, where there is a small opening in the basilar membrane which allows fluid to pass between the scala vestibuli and scala tympani.

Movement of the stapes in the oval window at the basal end of the cochlea results in a flow of fluid travelling along the length of the cochlea through the scala vestibuli, around the helicotrema, along the scala tympani, and out as a corresponding movement in the round window. The fluid movement displaces the basilar membrane which vibrates according to the amplitude and frequency of the incoming sound. The response of the basilar membrane is strongly affected by its mechanical properties which vary significantly from the base to apex. At the base it is relatively narrow and stiff whereas the apex is wider and more flexible. As a result, the peak displacement of the basilar membrane for high-frequency sounds is near the oval window and low-frequency sounds have a maximum displacement near the apex. Attached to the basilar membrane are the hair cells which form part of the organ of Corti and sit within the scala media. These hair cells are divided into two groups: outer hair cells which are arranged in three rows, and inner hair cells arranged in a single row. There are approximately 25,000 outer hairs cells and about 3500 inner hair cells. Above the hair cells sits the tectorial membrane. Movement of the basilar membrane creates a shearing motion with the tectorial membrane which results in a bending of the stereocilia at the top of the hair cells. This in turn creates a mode of excitation which leads to the generation of action potentials in the neurons of the auditory nerve. The hair cells are innervated by bipolar spiral ganglion neurons, whose axons form the auditory nerve (Figure 5.4).

The auditory nerve (cranial nerve (CN) VIII) leaves the inner ear through the internal auditory meatus and enters the brain at the angle of the pons and cerebellum, terminating in the medulla of the brainstem at the cochlear nuclei. From here it passes through a series of structures including the trapezoid body, superior olivary complex, lateral lemniscus, inferior colliculus, medial geniculate body, and finally into the primary auditory cortex. Afferent auditory nerve fibres carry information from the peripheral system to the brainstem, type 1 fibres innervating the inner hair cells and type 2 innervating the outer hair cells. Recent research has indicated that these fibres may be particularly vulnerable to noise injury, which may be an insidious cause of hearing loss. Efferent fibres bring information from higher neural centres in the olivocochlear bundle to the cochlea hair cells.

The vestibular system is central to sensing head position in relation to the environment, producing motor reflexes to support the maintenance of equilibrium and facilitating visual gaze stability. It comprises the utricle, saccule, and semicircular canals (Figure 5.5).

Each ear has three semicircular canals that sit at right angles to each other: horizontal, superior, and posterior semicircular canals. The horizontal canals lie at 30° to the horizontal plane. The anterior canal runs anterolaterally over the roof of the utricle. The posterior canal lies anteroposteriorly behind the utricle. Ampullae in the semicircular canals contain the cupula and sensory receptors that detect rotational acceleration. Vestibular hair cells of the semicircular canals project into and are coupled with the otolithic membrane of the cupula of the semicircular canals. The cupula has the same specific

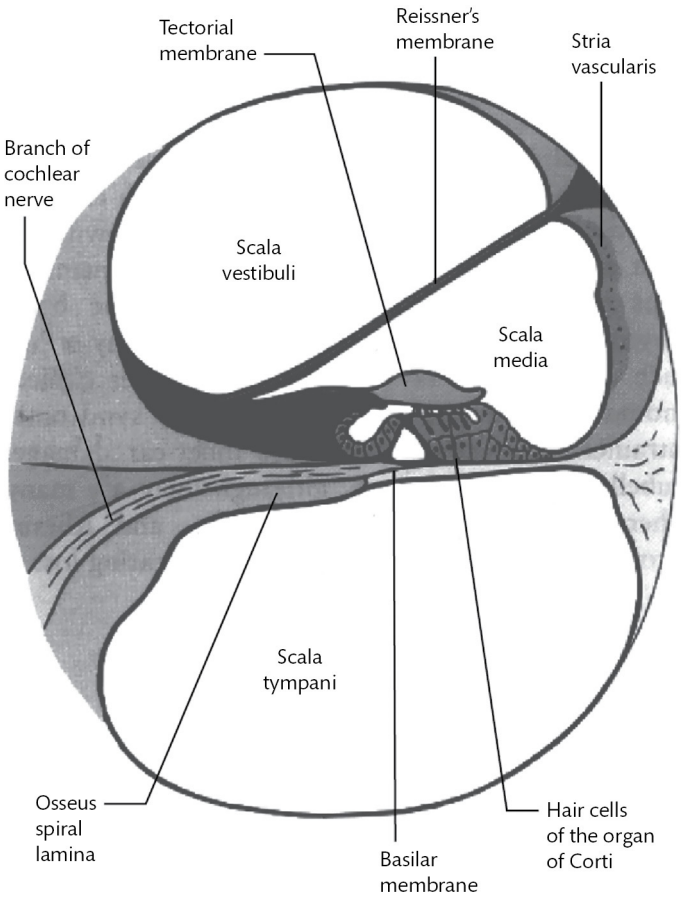

Figure 5.4 Cochlear cross-section.

Reproduced from Corbridge R, Steventon N. *Oxford Handbook of ENT and Head and Neck Surgery*. 3rd ed. Oxford University Press; 2019, with permission from Oxford University Press.

gravity as the endolymph surrounding it. Angular head acceleration causes displacement of the otolith membranes and cupulae, which stimulate the vestibular hair cells (Figure 5.6b).

The vestibule is in the central area of the vestibular system and contains the utricle and saccule, which detect linear accelerations, gravity

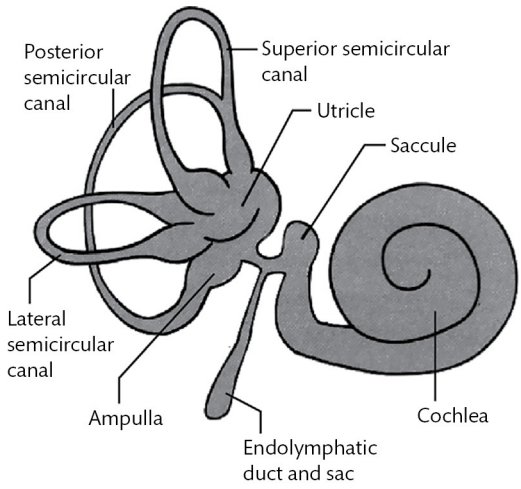

Figure 5.5 Vestibular system.

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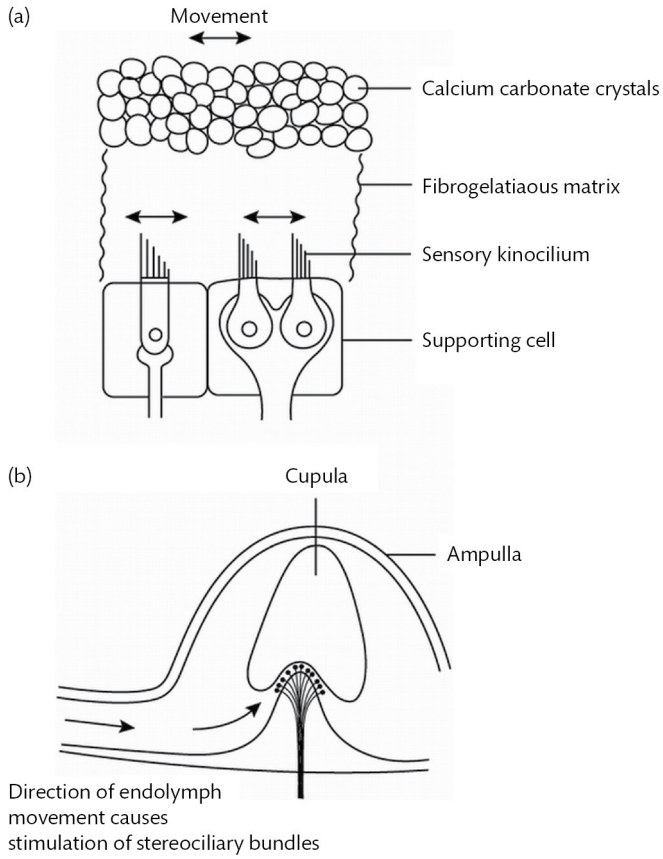

Figure 5.6 Sensory hair cells of the vestibular system: (a) otolithic membrane; (b) cupula and ampulla of semi-circular canals.

Reproduced from Warner G, Burgess A, Patel S, Martinez-Devesa R, Corbridge R. *Otolaryngology and Head and Neck Surgery*. Oxford University Press; 2009, with permission from Oxford University Press.

effects, and head tilts, in the horizontal and vertical plane respectively. The otolithic membrane in the macular of the saccule and utricle is composed of otoconia and a gelatin layer. Gravitational forces mean the greater density of the otolithic membranes overlying the hair cells results in a downward force on the neuroepithelium. During a static head tilt the weight of the membrane creates a shearing force on the hair cells result in neural stimulation (Figure 5.6a).

Orientation of the head causes bilateral vestibular organ stimulation. The central nervous system compares the resulting excitatory or inhibitory polarization from both ears to assess the plane of movement. Balance maintenance requires a complex interaction between the vestibular system and the visual and proprioceptive receptors. Using adaptive feedback loops and reflexes, the central nervous system is able to maintain gaze stability and coordinate motion in relation to the environment.

Auditory testing

Pure tone audiometry

Pure tone audiometry is presently the gold standard of assessing hearing threshold levels. Threshold is defined as the lowest-intensity sound which a patient can reliably identify the presence of 50% of the time. Pure tone audiometry is a subjective behavioural measurement used with adults and older children (typically aged 4 years and above) who can cooperate with the test procedure. Where an individual is not able to cooperate with testing (e.g. adults with intellectual disabilities and younger children), modifications to testing methods may be required (see Chapter 83). Using standardized techniques, individuals are presented with a series of frequency-specific tones, conventionally pure tones between 250 and 8000 Hz. They are asked to make a standard response, typically a button press, when they hear the sound. Automated audiometry exists where a computer program determines the presentation stimulus; however, manual audiometry where the audiologist controls the presentation of the stimulus is the most common technique. Air conduction testing measures everyday hearing ability, testing the entire auditory pathway from outer ear to inner ear. Air conduction thresholds are marked on an audiogram with O for the right side and X for the left side (Figure 5.7).

There are three main types of transducers that can be used for air conduction audiometry: supra-aural, circumaural and insert earphones. Supra-aural and circumaural earphones can be cumbersome particularly when masking bone conduction thresholds and may cause ear canal collapse. Insert earphones use a disposable foam tip to direct sound into the ear canal, preventing the ear canal from collapsing, and also have less transcranial transmission, reducing the need for masking; however, they cannot be used when there is wax or infection present. Unlike sound field audiometry, the use of these transducers in pure tone audiometry provides ear-specific results. Bone conduction audiometry sends vibrations via a bone oscillator directly to the inner ear, bypassing the outer and middle ear to directly stimulate the cochlea. Bone conduction thresholds are marked on an audiogram with a triangle. Bone conduction testing uses a reduced frequency range because of the limits of bone vibrators and vibrotactile thresholds. Without masking it is not possible to determine which ear is responding to the bone conduction testing. Masking is used to prevent cross-hearing which can occur when the difference in thresholds between the two ears is greater than the transcranial transmission loss, that is, when the sound delivered to the test ear also stimulates the contralateral non-test ear if sufficiently loud. To accurately assess thresholds where there is cross-hearing, a narrowband masking noise is used to temporarily elevate the threshold of the non-test ear. The interaural attenuation varies with the type of transducer, which requires different levels of masking to be applied, following three principal rules of masking. Masking is needed at any frequency where one of the following rules is met:

- Rule 1: where the difference between left and right not-masked air conduction thresholds are 40 dB or more using supra-aural or circumaural earphones, or 55 dB with insert earphones.
- Rule 2: where the not-masked bone conduction is better than the air conduction threshold in either ear by 10 dB or more.
- Rule 3: where rule 1 has not been applied but where bone conduction thresholds are better than the air conduction thresholds in the contralateral ear by 40 dB or more using supra-aural or circumaural earphones, or 55 dB with insert earphones.

Masked air conduction results are recorded with a shaded O/X and masked bone conduction with a square bracket [/]. The results are plotted on an audiogram which enables the assessment of type, degree, and configuration of hearing loss for diagnostic and rehabilitative management.

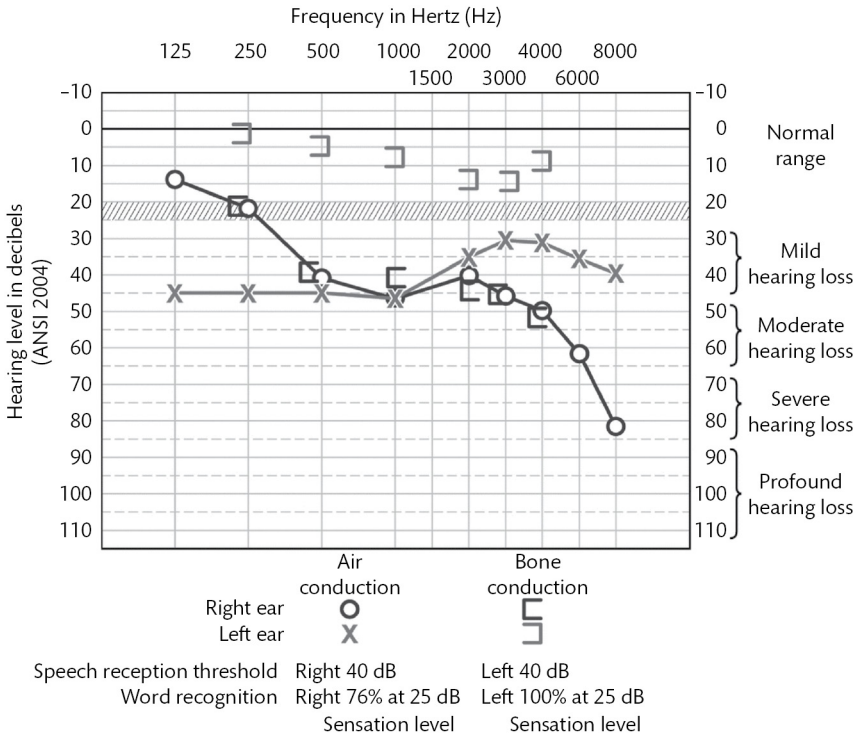

Figure 5.7 Masked audiogram.

Reproduced from Bauch CD, Olsen WO. Audiogram, acoustic reflexes, and evoked otoacoustic emissions. In: Rubin DI, Daube JR, eds. *Clinical Neurophysiology*. 4th ed. Oxford University Press; 2016:851–859, with permission from Oxford University Press.

The type of hearing loss can be divided into conductive, mixed, and sensorineural based on the relative position of the air conduction and bone conduction thresholds in each ear. A sensorineural hearing loss has an equal degree of loss for air conduction and bone conduction thresholds. A conductive loss is characterized when there is a difference of at least 15 dB between air conduction and bone conduction thresholds (air–bone gap) at the same frequency. An air–bone gap of 10 dB or less is not considered a conductive component, due to test–retest variability. A mixed hearing loss occurs when there is both a sensorineural and conductive component, where there is an air–bone gap and the bone conduction values fall outside the normal range. There are a number of classification systems used to define the degree of hearing loss. These typically use descriptors such as normal, mild, moderate, severe, and profound hearing loss based on the air conduction thresholds. Average thresholds may be reported; for example, 'four-frequency average' uses the mean air conduction value at 0.5, 1, 2, and 4 kHz. While average can be useful when comparing large numbers of audiograms, it fails to adequately account for the configuration of the audiogram and may not under represent the disability associated with the hearing loss. The configuration of the audiogram describes the shape of the audiogram which often provides insight into the aetiology. Configuration descriptors include terms such as flat, gradual sloping, steeply sloping, notch, and cookie bite (Figure 5.8).

There are limitations with pure tone audiometry because it relies on behavioural responses and does not require complex cognitive processing. As such, pure tone audiometry is not able to distinguish between cochlear and retrocochlear hearing loss, to identify cochlear dead regions where outer hair cells are intact but inner hair cells are dysfunctional, and central auditory processing disorders.

Tympanometry and middle ear reflexes

Tympanometry is an objective test of middle ear function and mobility of the tympanic membrane. Sound waves set in motion a vibration within the tympanic membrane and ossicular chain. The phase and amplitude of this vibration is determined by the acoustic admittance of the system. Tympanometry measures how admittance changes in response to a pressure changes. A probe tip is placed in the ear canal to make an airtight seal. During pressure changes a probe tone is presented, typically 226 Hz, and the tympanometer measures the functional change in ear canal pressure in response to this tone as a result of tympanic membrane movement. In infants a 1000 Hz tone may be used because of the difference in middle ear mass. A tympanogram provides data on ear canal volume, compliance, and pressure at which peak compliance is recorded. Typical values for ear canal volume are between 0.6 and 2.5 cm^3 for adults and 0.4 and 1.0 cm^3 for children. Where the tip of the probe is incorrectly positioned and occluded by the wall of the ear canal, an unexpectedly small ear canal volume would be seen. An open perforation and patent ventilation tubes (i.e. grommets) will give an abnormally large ear canal volume.

Compliance is normally in the range 0.3–1.6 cm^3 in adults. A lower limit of 0.2 cm^3 is acceptable in children under 6 years, but over 6 months. Units of mmho or mL may be used interchangeably when a 226 Hz probe tone is used.

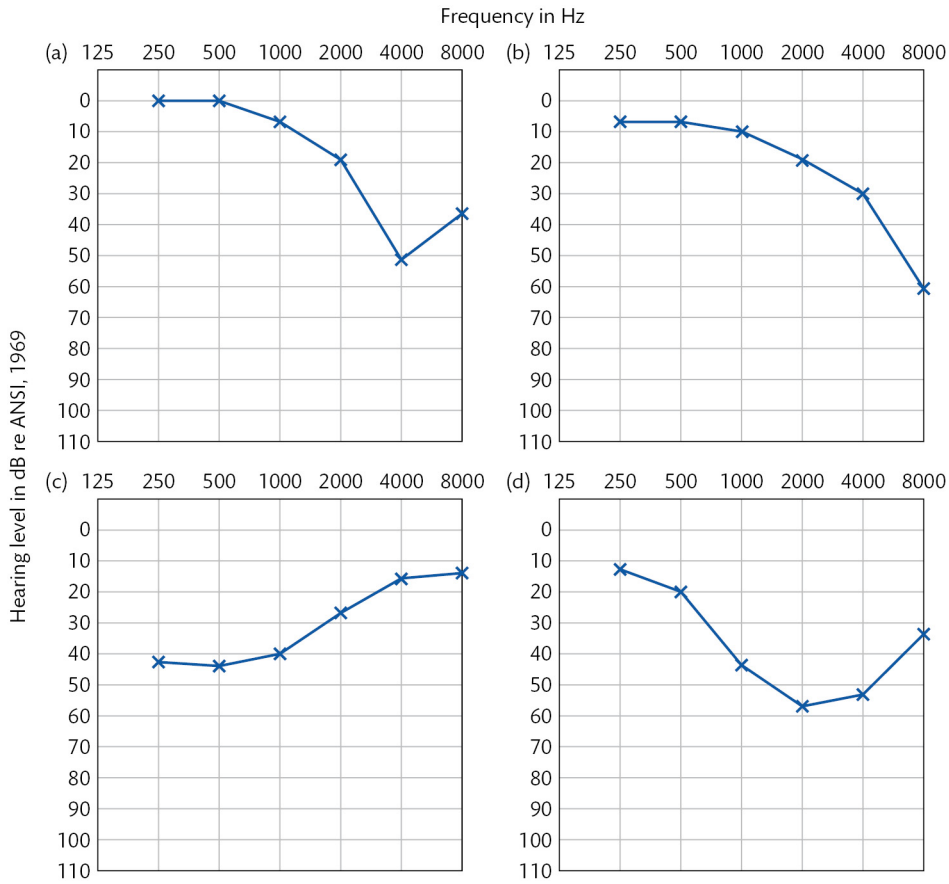

Figure 5.8 Examples of different audiogram configurations: (a) Sloping, notched; (b) high-frequency sloping; (c) reverse sloping; (d) cookie bite.
Reproduced from Baloh RW, Honrubia V, Kerber KA. Clinical evaluation of hearing. In: *Baloh and Honrubia's Clinical Neurophysiology of the Vestibular System*. 4th ed. Oxford University Press; 2011:218–230, with permission from Oxford University Press.

Peak pressure is used to estimate middle ear pressure. Normal middle ear pressure has a mean value of 0, though pressures from −50 to + 50 daPa is considered normal in adults. In children, values between −100 and + 50 daPa have little clinical significance. Tympanometric traces typically produce a single sharp peak; however, double peaks may be seen where there is scarring on the tympanic membrane.

Results are commonly categorized using classifications such as the Jerger classification system, which can be used as a diagnostic guide. It should be noted that some organizations no longer recommend the use of classification systems such as Jerger as it can sometimes lead to confusion or misreport; particularly when the original trace and values are not reported. Jerger classification divides tympanograms according to middle ear pressure and shape:

- Type A: normal middle ear pressure and compliance.
- Type As: normal middle ear pressure, low compliance.
- Type Ad: normal middle ear pressure, high compliance.
- Type B: no measurable middle ear pressure or compliance.
- Type C: significant negative middle ear pressure in the presence of normal compliance.

Long time-base tympanometry can be used as a diagnostic tool for middle ear conditions such as patulous Eustachian tube, or middle ear myoclonus (muscle spasm) by measuring acoustic immittance over an extended period of time and reviewing real-time immittance changes. In patulous Eustachian tube characteristic patterns of tympanic membrane movement with breathing may be observed, that cease with breath-holding (**Figure 5.9**). In middle ear myoclonus deflections of the TM are seen, and these are usually arrhythmic.

A tympanometer can also be used in acoustic reflex testing (**Figure 5.10**). The acoustic reflex is an involuntary muscle contraction of the stapedius muscle in response to loud sounds which stiffens the ossicular chain, decreasing middle ear admittance. Acoustic reflex thresholds measure the lowest sound pressure level at which sound triggers the acoustic reflex (**Figure 5.10**). Reflexes occur bilaterally regardless of which ear is exposed to sound so a change in immittance can be measured at either the ipsilateral or contralateral pathway. The ipsilateral reflex pathway involves stimuli travelling through the outer, middle, and inner ear, along the CN VIIIth nerve to the cochlear nucleus. From here the signal travels to the superior olivary complex and facial nerve (CN VII) nuclei, down the CN VII, and results in a contraction of the stapedius muscle. On the contralateral pathway after the cochlear nucleus, the signal travels to the contralateral superior olivary complex, CN VII nucleus, causing contraction of the contralateral stapedius muscle. Comparison of the results through the ipsilateral and contralateral pathways can contribute to the differential diagnosis. People with normal hearing and normal middle ear function

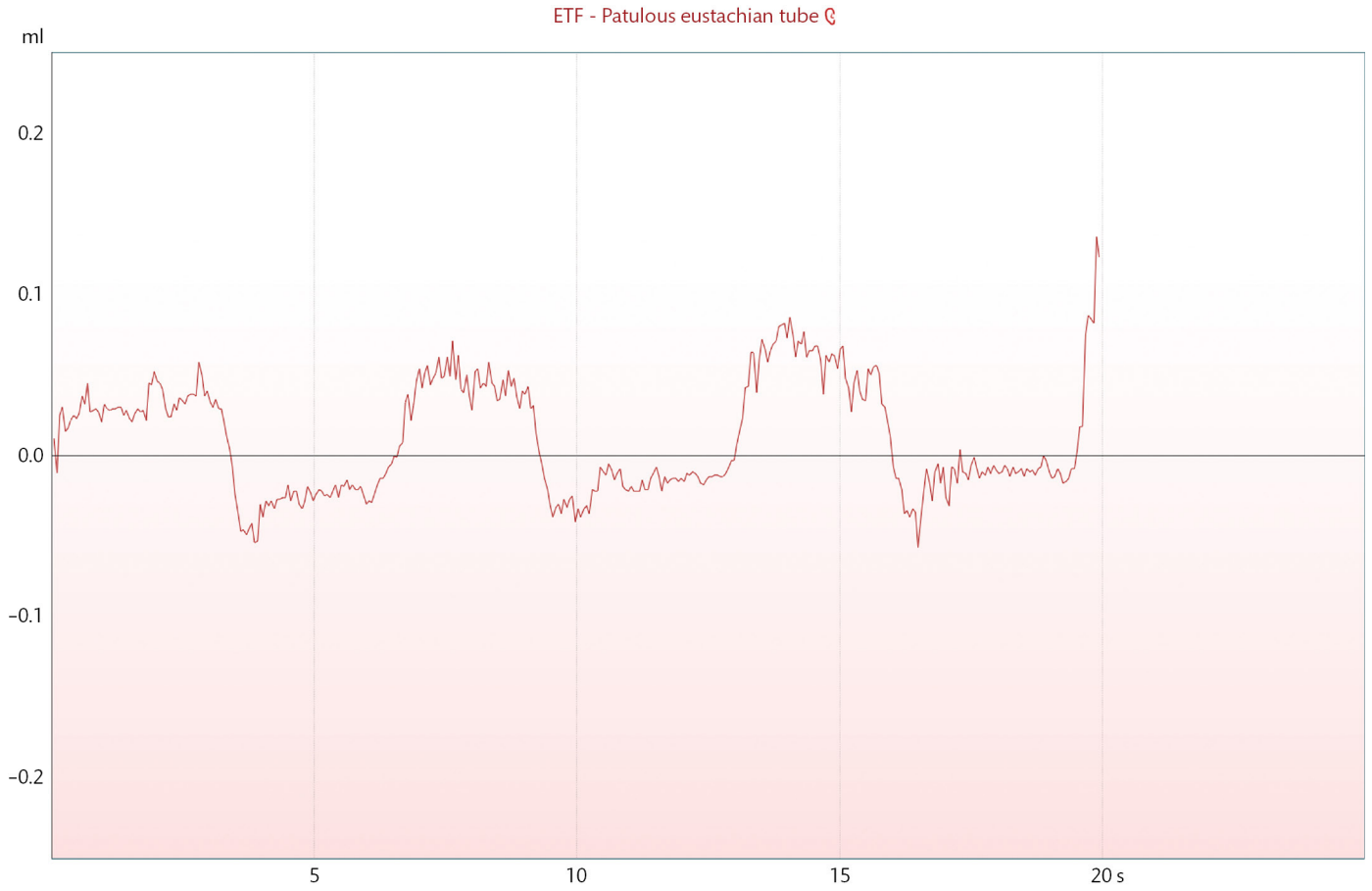

Figure 5.9 Long time-base tympanometry performed on author Gemma Crundwell with patulous Eustachian tube.

generally have ipsilateral and contralateral reflexes present at all frequencies; however, about 5% of the adult population have absent acoustic reflexes. Acoustic reflexes will be absent in ears with a middle ear disorder because they typically prevent the probe from measuring changes in compliance when the stapedius muscle contracts. Patients with cochlear hearing loss may have reflexes elicited at lower stimuli levels due to loudness recruitment. Patients with retrocochlear pathology may have elevated or absent reflexes. The

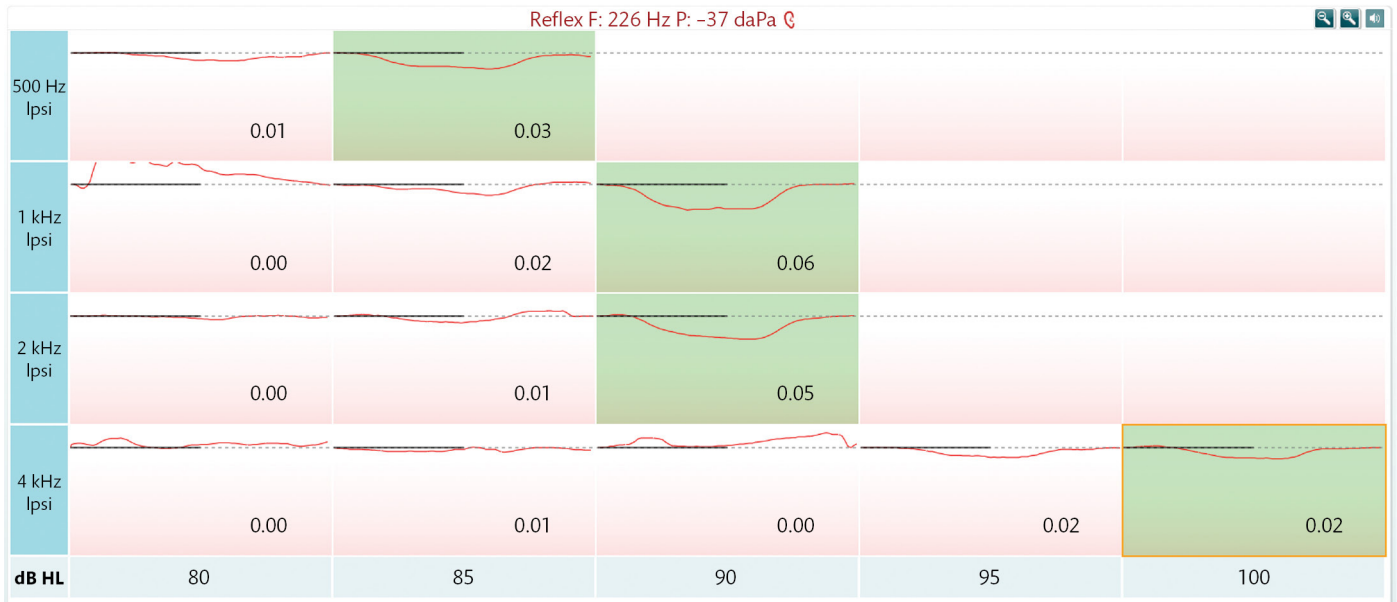

Figure 5.10 Ipsilateral acoustic reflex.

stapedius muscle is innervated by the facial nerve, therefore the stapedius muscle is likely affected in the presence of CN VII paralysis. Reflex decay testing is an additional tool that can be used to identify retrocochlear pathology. In normal ears the reflex should stay contracted for the full 10 seconds and abnormal reflex decay is indicative of nerve abnormalities.

Otoacoustic emissions

Otoacoustic emissions (OAEs) are an objective assessment providing information about the integrity and function of the outer hair cells. OAEs are commonly used as part of newborn hearing screening programmes because it is a quick non-invasive test. When the outer hair cells are stimulated they change shape to amplify and propagate sounds along the basilar membrane. This amplification function of the outer hair cells can be measured with a microphone in the external auditory canal. OAEs fall into two categories, spontaneous and evoked. Spontaneous OAEs occur naturally without external stimulation. Evoked OAE use a transient signal such as broadband clicks or tone burst (transient evoked OAEs) or a pair of pure tones referred to as f1 and f2 (distortion product OAEs). A healthy outer and middle ear are required to accurately record an OAE because of the small amplitude signal (Figure 5.11).

Auditory evoked potentials

Auditory evoked potentials are electrical signals generated by the auditory nerve and central nervous system in response to an acoustic stimulus. They are used to objectively assess hearing using electrodes on the skin, scalp, or tympanic membrane. Auditory evoked potentials are divided by their response latencies: short latencies—electrocochleography, auditory brainstem responses (ABRs), cochlear microphonics; middle latency—P1 and N1 responses; and long latency—cortical event-related evoked potentials.

Electrocochleography is a technique for recording electrical potentials within the cochlea, evoked by sound stimulation. Traditionally a needle electrode was placed on the promontory, passing through the tympanic membrane, but more recently placement of an electrode upon the tympanic membrane or in the external ear canal has become more popular and is less invasive. Click or tone pip stimuli are used to evoke cochlear action potentials, which can be used as an objective measure of threshold, and can also be analysed to diagnose Ménière's disease if the morphology of the action potential is altered in a characteristic fashion.

The most widely used evoked response is the ABR which measures the integrity of the auditory system up to the lower brainstem. An electrode montage on the head (high forehead, mastoids, and low forehead ground) measures the neural response to sound. Peaks in the response correspond to activity within particular

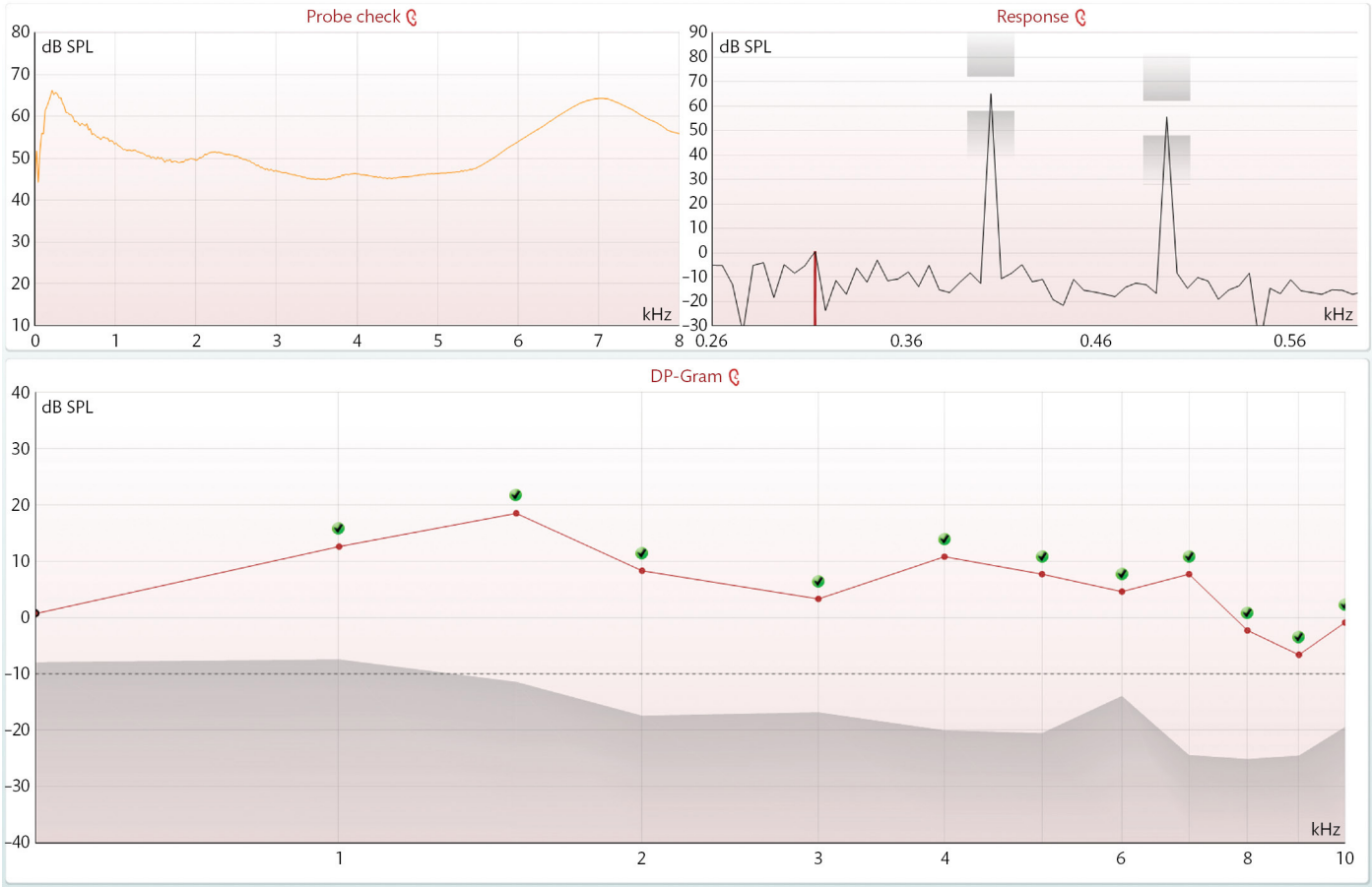

Figure 5.11 Distortion product OAE.

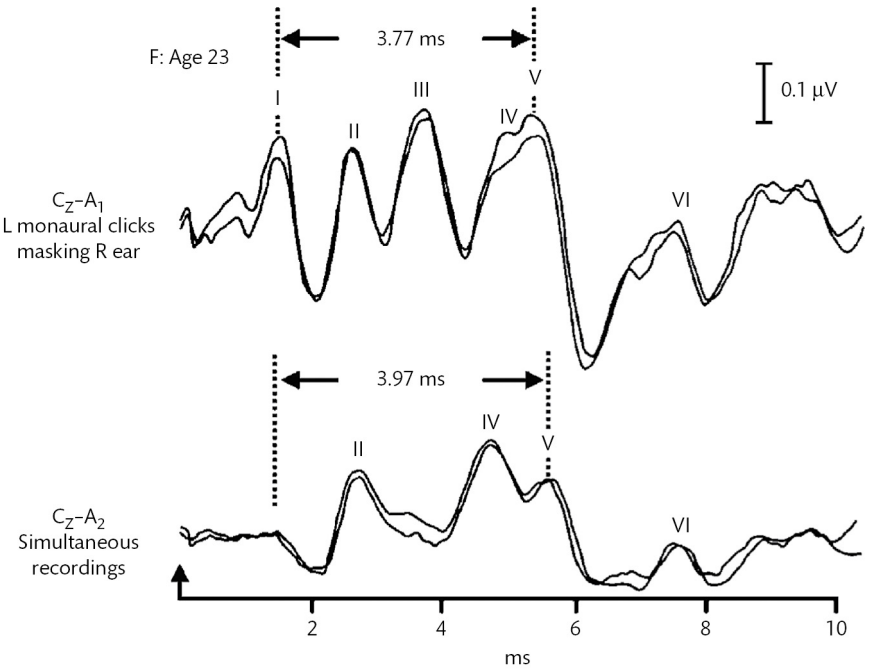

Figure 5.12 Auditory brainstem response.

Reproduced from Baloh RW, Honrubia V, Kerber KA. Clinical evaluation of hearing. In: *Baloh and Honrubia's Clinical Neurophysiology of the Vestibular System*. 4th ed. Oxford University Press; 2011:218–230, with permission from Oxford University Press.

areas of the auditory pathway which can be used for neurological evaluation of the integrity of the auditory system (Figure 5.12). ABR can also be used for threshold estimation by identifying the lowest intensity at which a response remains present. As an objective assessment of hearing, ABR is used as part of newborn hearing screening programmes and in patients who are unable to complete behavioural testing.

The cochlear microphonic (CM) is a pre-neural electrical potential arising from the cochlea, which mimics the stimulus waveform; for example, a 500 Hz pure tone will appear as a 500 Hz CM. The presence of a CM suggests outer hair cell function. CM can be used to help diagnose patients with auditory neuropathy spectrum disorder. People with auditory neuropathy spectrum disorder typically have abnormal or absent ABRs and present OAEs. CM present at the beginning of the click but no synchronous neural activity on the ABR.

Auditory steady-state response is an electrophysiological response to rapid auditory stimuli which can be used to create a statistically valid audiogram. Auditory steady-state response detects spectral peaks across the frequency domain and uses statistical measures to determine if and when a threshold is present.

Cortical auditory evoked potentials use the P1–N1–P2 response complex as an objective predictor of hearing thresholds. They can be elicited by the onset of a transient stimuli such as a click or by a change in a stimulus such as a gap in a continuous sound, change in level, or change in frequency of a continuous sound. The response can be used to provide an indication of the maturity of the auditory pathway. Unlike an ABR a cortical auditory evoked potential may not be reliably present in drowsy or sleeping patients. As a result, cortical auditory evoked potential testing is performed in awake and alert patients.

Functional hearing loss

Functional hearing loss (also called non-organic hearing loss or pseudohypacusis) is characterized by hearing loss without a detectable corresponding structural pathology in the auditory system. Typically, this is observed as a discrepancy between pure tone thresholds and speech discrimination ability. It is important to note that the term functional hearing loss does not mean there is an absence of a problem nor that there is an intentional falsification or exaggeration of a problem; it is therefore not synonymous with malingering or feigning. Functional hearing loss can present in both adults and children for a variety of psychological and psychosocial factors. Positive support and onward referral to psychology are indicated rather than the traditional approach of confrontation.

The use of objective measures such as OAEs and ABR combined with clinical judgement can be used to identify functional hearing loss. The management of functional hearing loss is challenging, and may require vigilance to the possibility of an emotional or trauma-related aspect to the patient's behaviour, which would fall outside the remit of an ear, nose, and throat doctor or audiologist, and onward referral is essential.

Hearing rehabilitation and hearing aids

Once a hearing loss has been identified an appropriate (re)habilitative strategy can be implemented.

Hearing therapy explores the psychological and emotional effects of audiological problems and provides help and advice on practical solutions.

Communication tactics are strategies that can be implemented to ameliorate social difficulties associated with hearing. Communication tactics can be used by individuals with hearing impairment as well as their communication partners. They can be used in conjunction with other rehabilitative strategies, and are particularly helpful in challenging listening situations. Examples include reducing background noise where possible, facing the person, and ensuring good lighting to be able to see visual cues, gestures, and lipreading.

Lipreading can be a useful additional skill for those with hearing loss. Learning to recognize lip patterns for different speech sounds and combining these with people's expressions and gestures can assist in everyday conversations, particularly when in noisy situations.

Assistive listening devices are pieces of equipment other than hearing aids that can be used to help people with hearing impairment in specific situations. Even with hearing aids, the limitations associated with amplification mean some listening situations remain challenging. Assistive listening devices may be designed to work on their own or work in conjunction with hearing aids. These include amplified telephones, flashing doorbells, vibrating alarm clocks, and headsets for the television.

Hearing aids are the most common intervention for hearing loss. All hearing aids share the same basic components. Sound is detected by the microphone and converted into an electrical signal. In contemporary hearing aids this is passed through a digital signal processor before being delivered by the receiver and ear coupling system into the external auditory meatus. Custom moulded hearing aids are made from impressions of the patient's ear which can be taken physically or by three-dimensional scanning.

There are a variety of factors that influence the style of hearing aid including audiological, economical, functional, and cosmetic. Completely-in-canal aids are the smallest most cosmetically appealing hearing aids with all their components contained within the ear canal. In-the-canal instruments fit within the concha and cartilaginous portion of the ear canal. In-the-ear hearing aids include the full concha. The utilization of the full area of the concha to house the device allows the use of complex electronics and reduces internal feedback compared to completely-in-canal and in-the-canal aids. Behind-the-ear hearing aids are designed to fit behind the pinna, with a sound tube directing sound into the ear canal. Traditional behind-the-ear hearing aids are used with an ear mould, which is custom fitted to the individual ear. More recently, open-fit hearing aids have been used, where a fine tube delivers sound into the ear. This allows amplification with improved cosmetics and without the occluding effects of an ear mould; however, they are not suitable for all patients. Devices where the receiver is separate from the amplifier are known as receiver-in-the-canal aids. Body-worn hearing aids have a separate unit which is connected to the hearing aid via a cord. Body-worn devices may be used where significant gain is required without feedback, by separating the microphone of the aid and the receiver or where there are significant dexterity issues that would prevent the use of smaller ear-level devices.

Contralateral routing of sound/contralateral routing of offside signals hearing aids have been specifically developed to manage the challenges posed by single-sided hearing loss. Sound is picked up on the unaidable ear and routed to the better hearing ear. Where there is also a hearing loss in the better ear, a *bi-contralateral routing of sound* hearing aid also provides amplification to overcome the hearing loss in the better ear.

Combination devices combine a hearing aid with a sound generator for patients with tinnitus, hyperacusis, and misophonia. In addition to amplifying sound these provide additional therapy sounds with the aim of habituating tinnitus.

Where conventional air conduction devices are not suitable, there are a variety of implanted hearing devices including auditory brainstem implants, cochlear implants, bone conduction devices, and middle ear implants (see Chapter 16).

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6

Tinnitus and disorders of sound tolerance

Don McFerran

Introduction

There is no completely satisfactory definition of tinnitus—'the conscious perception of an auditory sensation in the absence of a corresponding external source' is widely used but even this is not perfect at as it would include the auditory hallucinations of some psychological illnesses. Most tinnitus takes the form of simple sounds such as ringing, whistling, or buzzing. However, some people report more complex sounds including musical sounds or distant voices—in such cases, there is no meaning conveyed by the voices, in contrast to psychological auditory hallucinations. Some people with tinnitus report rhythmical sounds which may be synchronous with the arterial pulse, the respiratory cycle, or may have its own individual rhythm. Heart synchronous tinnitus is referred to as pulsatile tinnitus and may have a vascular origin (Box 6.1).

Rarely, people describe hearing their own breathing which may be due to a patulous Eustachian tube. Rhythmical tinnitus that is asynchronous with the heart or breathing is often described as a fluttering or ticking sensation and may have a muscular origin either from myoclonus of the intratympanic muscles, stapedius, and tensor tympani, or myoclonus of the palatal muscles. Rhythmical tinnitus can be a stand-alone entity, or it can coexist with non-rhythmical tinnitus. For most tinnitus there is no detectable sound to an external observer and such tinnitus is classified as subjective. In a few cases a sound can be detected, particularly in some forms of pulsatile or myoclonic tinnitus, and the tinnitus in these cases is therefore classified as objective. It is important to state, however, that most pulsatile tinnitus does not have a detectable sound even when highly sophisticated listening equipment is used and therefore most pulsatile tinnitus is subjective. There have been a few reported cases where people are aware of their own spontaneous otoacoustic emissions though this is a very rare cause of tinnitus. Some people report tinnitus that is modulated by touch or movement, particularly of the neck or jaw joints. Such tinnitus is called somatosensory tinnitus.

Defining disorders of sound tolerance is a complicated and somewhat contentious field. The word hyperacusis is frequently employed as a blanket term for all forms of sound intolerance—in its alternative usage, hyperacusis can be defined as dislike of all sounds above a certain intensity; misophonia is a word that conveys dislike or hatred of particular sounds irrespective of their intensity; phonophobia is fear of certain sounds irrespective of intensity; and recruitment is abnormal loudness growth and is generally associated with significant sensorineural hearing loss. However, the word misophonia has also been used to define a specific group of people who respond to sounds typically generated by other humans' mouths such as lip-smacking, chewing, or whistling. These people report feeling anger towards the person generating the sounds and react with avoidant behaviour. Misokinesia is a similar response to repetitive visual stimuli created by other people, such as leg-swinging or repetitively clicking the top of a pen. An international working group proposed an alternative classification of impaired sound tolerance: loudness hyperacusis, annoyance hyperacusis, fear hyperacusis, and pain hyperacusis. Although this suggestion has some merit, it has yet to be widely adopted.

Box 6.1 Some of the causes of heart synchronous pulsatile tinnitus

Arterial	Anatomical variants/abnormalities Dissections Aneurysms Stenoses Arteriovenous fistula Arteriovenous malformations
Venous	Anatomical variants/abnormalities Stenosis Intracranial hypertension
Microvascular	Vascular tumours
Bone pathology	Paget's disease Otosclerosis
Perceptual	Conductive hearing loss
General circulation	Increased cardiac output (anaemia, hypoxia, thyrotoxicosis, pregnancy)
Other	Superior semicircular canal dehiscence

Epidemiology

There are now tinnitus prevalence studies from many countries and these have produced wildly varying estimates of tinnitus prevalence,

ranging from 5.1% of the study population to 42.7%. Much of this variation can be explained by methodological differences between the studies and the consensus view is that the point prevalence probably lies between 10% and 15% of the population. A large UK study suggested that while 10.1% of the adult population have some degree of tinnitus, the number who find it intrusive is much less with approximately 1 in 30 reporting tinnitus which is at least moderately annoying and 1 in 200 reporting it at a level that severely affects the ability to lead a normal life. Most studies have shown that there is no significant sex difference in tinnitus prevalence though there have been reported differences in the way that men and women perceive their tinnitus. The prevalence of tinnitus increases with age up to the seventh decade of life. Some studies show the prevalence continuing to increase thereafter but many other studies show the prevalence plateauing or even starting to lessen with increasing age. Although children rarely volunteer that they perceive tinnitus, when they are asked in age-appropriate language, paediatric tinnitus prevalence seems not dissimilar to the adult prevalence. One study suggested that there is probably at least one child with significant tinnitus in every UK school classroom. Less is known about the incidence of tinnitus and there is a paucity of longitudinal studies. The available longitudinal data suggest that tinnitus generally become less intrusive with time for most people. Genetic studies have shown a small degree of heritability overall though ongoing work suggests this may be higher for certain subtypes of tinnitus.

The prevalence of disorders of sound tolerance remains to be fully elucidated though there is a strong link with tinnitus: the majority of people whose primary complaint is hyperacusis report some degree of tinnitus; conversely, among those whose primary complaint is tinnitus, approximately half report some impairment of sound tolerance.

Associations

The condition with the biggest link to tinnitus is hearing loss and this is an interesting relationship. All forms of hearing loss are risk factors for tinnitus—including conductive losses and sensorineural loss due to noise exposure, age, or ototoxic agents. However, the degree of hearing loss correlates poorly with tinnitus severity. It is possible to have a severe or profound hearing loss without accompanying tinnitus. Conversely, approximately one in ten people with tinnitus have normal hearing as measured by pure tone audiometry although investigation with more sophisticated hearing tests such as threshold equalizing in noise testing suggests that most if not all tinnitus sufferers have some degree of impairment. It is even possible for tinnitus to occur after division of the auditory nerve: a functioning auditory periphery is not a prerequisite for tinnitus. Some otological conditions including otosclerosis, Ménière's disease, and vestibular schwannoma feature both hearing loss and tinnitus as part of their symptom complex. These are sometimes referred to as examples of syndromic tinnitus and we understand more about both the underlying pathophysiology and the outcome of treatments in such cases. As discussed previously, tinnitus is frequently comorbid with disorders of sound tolerance. Tinnitus is often associated with various psychological conditions, particularly anxiety, depression, and post-traumatic stress disorder. Other tinnitus comorbidities include migraine, head injury, and temporomandibular joint dysfunction.

Most of the comorbidities for tinnitus also apply to disorders of sound tolerance, particularly anxiety, post-traumatic stress disorder, depression, and migraine. Conditions that affect facial nerve function and thereby impair the protective stapedial reflex are associated with increased risk of hyperacusis. Williams–Beuren syndrome is a congenital condition caused by deletion of a portion of chromosome 7 and impaired sound tolerance is one of its defining characteristics.

Pathophysiology

Many pathological mechanisms have been suggested for the causation of tinnitus and as our knowledge improves it seems increasingly evident that tinnitus is not a single entity but many different processes that end up causing similar symptoms. This heterogeneity impairs both management and research of the condition. Some of the processes have been suggested are shown in **Box 6.2**. Our understanding of the pathophysiology of disorders of sound tolerance is still rudimentary.

A practical, patient-focused model of tinnitus pathophysiology

Although it is academically interesting to study the underlying mechanisms of tinnitus, this has not yet translated into a useful management strategy and we still have no way of determining what process is occurring in any individual patient. Furthermore, the main problem is not the patient's awareness of the sound percept but rather the reaction and distress that this triggers. This process involves not only conventional auditory pathways but also many non-auditory brain regions. Rather than trying to explore tinnitus at a cellular level, it is more fruitful to examine how the auditory system functions and links to other systems using a flowchart model. Although this is somewhat simplistic, it helps to explain some of the paradoxes of tinnitus and also provides a useful framework for explaining tinnitus to patients in a clinical setting.

Incoming sound is turned into a stream of digital electrical information by the ear which is essentially an analogue-to-digital convertor. This information is conveyed to sub-awareness regions of the brain where it undergoes a process of pattern recognition and filtering. Information that is deemed important is passed onwards to higher centres of the brain at the auditory cortex where the incoming signal is consciously perceived as a sound. Unimportant sound information is ignored. But the lower brain also has an option to involve other non-auditory parts of the brain, particularly the limbic system (**Figure 6.1**), a phenomenon well demonstrated in

Box 6.2 Some of the proposed mechanisms by which tinnitus neuronal activity is generated

- Reorganization of the brain's tonotopic map following deafferentation.
- Increased spontaneous neuronal firing within the auditory brainstem and midbrain.
- Increased neuronal synchrony.
- Failure of inhibitory pathways.
- Maladaptive auditory–somatosensory plasticity.
- Errors of predictive coding.

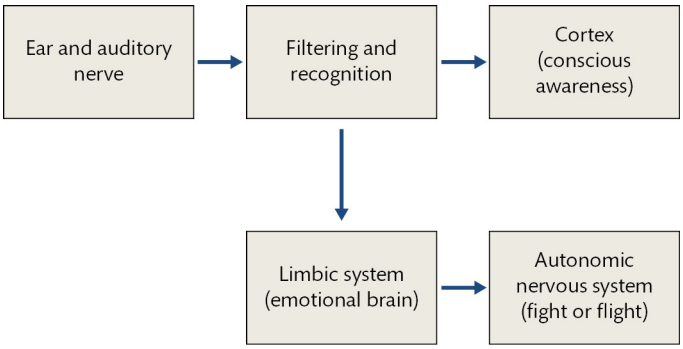

Figure 6.1 The functional elements of the auditory system.
Credit: reprinted with permission from McKenna, Baguley and McFerran, (2021) Living with Tinnitus and Hyperacusis, Sheldon Press.

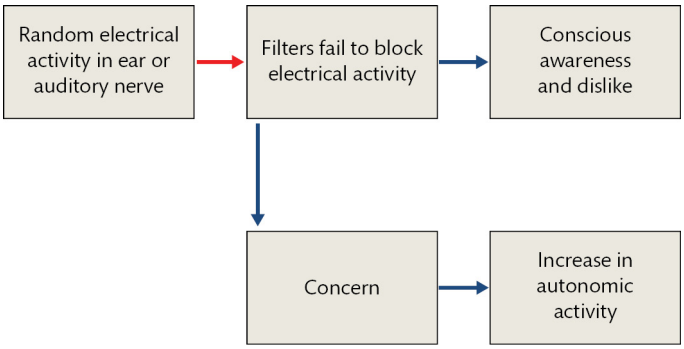

Figure 6.3 Random electrical activity in the auditory system passing through the normal filtering circuitry and being misidentified as an unpleasant sound.
Credit: reprinted with permission from McKenna, Baguley and McFerran, (2021) Living with Tinnitus and Hyperacusis, Sheldon Press.

functional magnetic resonance imaging (MRI) studies of the auditory system. It is easy to explain these processes to a tinnitus patient. We are all aware that auditory attention varies: we have all experienced being in a group of people all chatting, yet if someone mentions our name we are instantly aware and our attention is drawn to that speaker. Conversely, we may be so engrossed in doing a task that we are effectively unaware of sound around us. Hearing also has an important warning function: if we hear a sudden unexpected sound we startle, eliciting a fight or flight response. This is part of our hard-wired survival mechanism: the lower brain centres are constantly on the alert for unexpected sounds, viewing them as potential threats and alerting both the auditory cortex and limbic pathways.

One of the seminal tinnitus research projects was performed by Heller and Bergman in 1953—they put 80 people with normal hearing and no tinnitus in a soundproof chamber and asked them to listen carefully. Within 10 minutes 75 subjects reported hearing a variety of sounds including humming, buzzing, and ringing. This finding has been used to suggest that we all have random electrical activity in our auditory pathways. This is normally ignored (Figure 6.2) but if the circumstances are right, it can be perceived as sound.

If, however, the subcortical centres fail to block this activity it can be perceived as a sound in higher brain centres and can be mislabelled as a threat, eliciting a limbic response (Figure 6.3).

Once this neuronal activity has been interpreted as an unpleasant, potentially threatening sound there is an opportunity to set up positive feedback loops between various parts of the brain which enhance the sensation and exacerbate the psychological response (Figure 6.4).

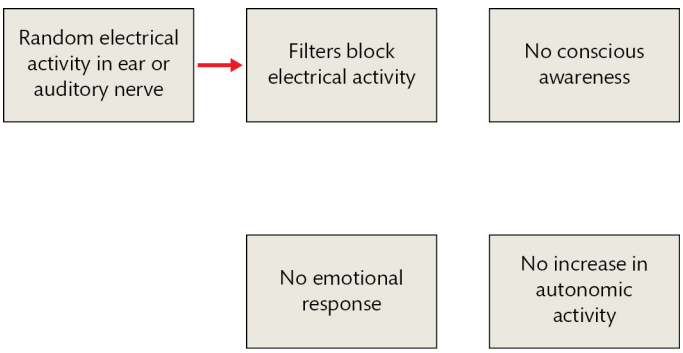

Figure 6.2 Random electrical activity in the auditory system being blocked at a subcortical level and being ignored.
Credit: reprinted with permission from McKenna, Baguley and McFerran, (2021) Living with Tinnitus and Hyperacusis, Sheldon Press.

The switch from normal to abnormal state can be triggered in several ways: hearing loss causes the brain to strain to listen, in effect opening the auditory filters and letting the tinnitus signal through; stressful events can enhance limbic system activity and because the brain pathways are reciprocal this can cause the filters to malfunction. Of course, this model is a simplification of a highly complex system, but it can be used to explain some of the seeming paradoxes of tinnitus. The model can explain why hearing loss can trigger the symptom but can also explain why hearing loss is not an essential prerequisite. It can explain why stressful life events can cause the problem. It can explain why anxiety, depression, and insomnia are common. It can explain why the symptom becomes self-perpetuating after the initial generation. It can even explain why a functioning ear is not necessary for tinnitus.

The model can also point the way towards methods of ameliorating the symptom—using methods such as correcting hearing loss, dealing with anxiety and depression, and stress reduction.

Assessment

History and examination

Taking a clear otoneurological history is paramount. The nature of the tinnitus should be assessed, particularly whether the percept is continuous or rhythmical. Key questions for exploring the impact of the patient's tinnitus are whether it is affecting sleep and/

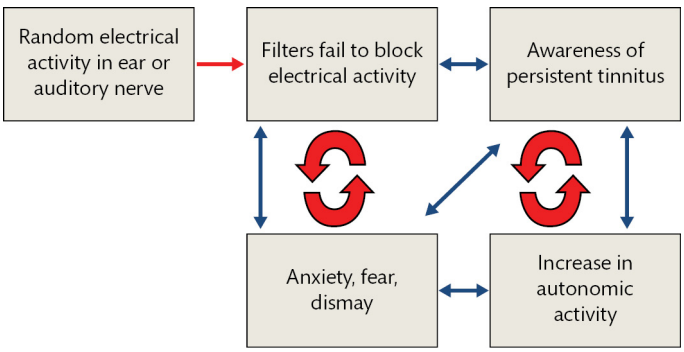

Figure 6.4 The vicious circles of tinnitus.
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or concentration. A standard otological examination should be performed though this is often unremarkable. Examination should include the temporomandibular joints and neck movements. For heart synchronous tinnitus, careful auscultation of the head and neck should be performed. Cervicocranial bruits are generally quiet and quite localized, so this step should not be hurried. If myoclonus is suspected, examination of the pharyngeal muscles, looking for involuntary muscular contraction, should be conducted. This can be performed either trans-orally or by using fibreoptic nasendoscopy.

Investigations

Audiometry

Pure tone audiometry should be performed to assess any associated hearing loss, to determine if further investigations are needed, and, when necessary, as a precursor to hearing aid fitting. Many people with tinnitus complain of the full or blocked sensation in the ears and tympanometry is useful to exclude Eustachian tube dysfunction. If middle ear myoclonus or patulous Eustachian tube is suspected, long time-base tympanometry is helpful, showing a sawtooth rapid variation of middle ear impedance in the former and a larger but slower variation in the latter, synchronous with the patient's breathing. Audiometry can be used to measure the pitch, tone, and minimal masking level of tinnitus. These measurements, however, add little to the management plan, give inconsistent results, and are time-consuming to perform. While they may have value in hearing research, there seems little point in performing these tests in the routine clinical setting. Similarly, otoacoustic emission measurements add little to the management of most tinnitus patients. Acoustic reflex testing and uncomfortable loudness level/loudness discomfort level testing both use high levels of sound that patients with tinnitus and, more particularly, hyperacusis find unpleasant. These tests should therefore be avoided unless there is a compelling clinical indication.

Imaging

For non-pulsatile tinnitus, further investigation is recommended if the tinnitus is unilateral or asymmetric, associated with an asymmetric pure tone audiogram, or if there are abnormal neurological symptoms or signs. The UK National Institute for Health and Care Excellence (NICE) published a tinnitus guideline in March 2020 which recommended that imaging should be considered if there is an asymmetry on pure tone audiometry of 15 dB or more at any two adjacent test frequencies, using test frequencies of 0.5, 1, 2, 4, and 8 kHz. The recommended imaging modality is MRI and this should be possible for the majority of tinnitus patients. Where there is a genuine contraindication, contrast-enhanced computed tomography (CT) can be used. The investigation of pulsatile tinnitus is more complex and although various algorithms have been suggested there is still some disagreement on this topic. Investigating pulsatile tinnitus may require multiple modalities including MRI, magnetic resonance angiography, CT, CT angiography, Doppler ultrasound, conventional angiography, and lumbar puncture with cerebrospinal fluid pressure measurements.

Blood tests

For most patients with tinnitus, blood tests do not supply useful information and are unnecessary. For patients with pulsatile tinnitus secondary to a hyperdynamic circulation, haemoglobin, thyroid function tests, and/or pregnancy testing may be needed. Some tinnitus arises after a sudden sensorineural hearing loss in which case the appropriate investigative algorithms should be followed (see Chapter 11). Some weak evidence suggests that tinnitus can be associated with vitamin B deficiency and, importantly, that correcting the deficiency can help the tinnitus. Patients who have low dietary intake of vitamin B or malabsorption syndromes should therefore have their vitamin B levels checked.

Questionnaires and other tools

A large problem with both the clinical management and research of tinnitus is that there are no satisfactory biomarkers or outcome measures for tinnitus. Many tinnitus clinics therefore use healthcare questionnaires to assess tinnitus impact and monitor treatment progress. There are many such instruments available for tinnitus, all with pros and cons. The most commonly used in the UK are the Tinnitus Handicap Inventory (THI), the Tinnitus Functional Index (TFI), and the Tinnitus Questionnaire (TQ). There are also questionnaires which can be used to assess comorbid conditions including insomnia, anxiety, and depression. The Insomnia Severity Index is useful for assessing insomnia. The TQ was developed by psychologists and is useful for assessing both tinnitus in general and the psychological impact. Questionnaires for disorders of sound tolerance have been developed but are less widely used than tinnitus questionnaires. The most commonly used hyperacusis instrument in the UK is the Khalfa Hyperacusis Questionnaire. In a research setting, visual analogue scales are often used to assess a range of tinnitus parameters.

Treatments

Syndromic tinnitus

For cases of syndromic tinnitus where the symptom is associated with a particular otological condition, management will follow the pathway specific to that condition. The management of otosclerosis (see Chapter 20) may involve watchful waiting, fluoride treatment, use of hearing aids, or stapes surgery. The tinnitus outcome following stapedectomy has been well documented and tinnitus generally improves, especially when the hearing outcome is good. However, surgery carries a small risk of triggering tinnitus or worsening pre-existing tinnitus and this should be carefully discussed preoperatively. Management of vestibular schwannoma (see Chapter 18) has also been explored with respect to tinnitus outcome. There is conflicting evidence about what happens to tinnitus when vestibular schwannoma is managed by watchful waiting and there is no firm evidence on how best to treat this growing population of patients. The consensus following vestibular schwannoma surgery is that pre-existing tinnitus is more likely to remain than lessen or disappear. Studies on the use of either fractionated radiotherapy or radiosurgery generally suggest outcomes similar to surgical outcomes though this is not universal and some studies have given much more optimistic results regarding tinnitus. The management of Ménière's disease is complex (see Chapter 14) and mostly focuses on control of vertigo and hearing. Where tinnitus outcomes have been explored, results are either inconclusive or show no effect.

Rhythmical tinnitus

Heart synchronous rhythmical (pulsatile) tinnitus

When investigation of heart synchronous pulsatile tinnitus reveals a specific pathology, such as carotid artery stenosis, an arteriovenous fistula, vascular skull base tumour, raised intracranial pressure, or cranial venous outflow obstruction, the appropriate management protocol should be followed. However, investigation often fails to reveal a treatable cause in which case standard tinnitus management methods can be used.

Heart asynchronous rhythmical tinnitus

If investigation of pulse asynchronous rhythmical tinnitus reveals specific pathology there may be relevant treatment options. Intratympanic myoclonus patients can be offered division of the stapedius and/or tensor tympani tendons. However, this does not always help the symptoms and is irreversible. Palatal myoclonus can be treated by botulinum toxin injection into the affected muscles. Treatment of palatal myoclonus with systemic drugs has been explored using benzodiazepines or antiepileptic drugs but results are disappointing. Patulous Eustachian tube is rare and has been the subject of many attempted treatments. Those with best evidence include procedures that increase the mass of the tympanic membrane either by applying materials to the surface of the eardrum or by performing cartilage tympanoplasty. Alternatively, various procedures have been described to try and partially block the Eustachian tube. Grommet insertion has historically been deemed inadvisable as it can worsen symptoms. The available evidence, however, suggests that it is beneficial for about half of sufferers.

Persistent spontaneous tinnitus

Most tinnitus is neither syndromic nor rhythmical and in such cases there are no current curative treatment options. There are, however, many helpful management strategies underpinned by audiological and psychological principles.

Counselling and education

It is vital that negative counselling is avoided. Patient surveys have shown that many tinnitus patients are told that nothing can be done for their symptom and they will just have to learn to live with it. They understandably report that they find such statements not only unhelpful but positively distressing. A more optimistic approach should be used, explaining that there are many options to try and ameliorate the tinnitus impact. Many people find that once they understand tinnitus it helps them to cope more effectively and therefore the first step in managing tinnitus is generally an education module to explain the condition. Counselling is used to identify patients' specific problems and discuss ways to resolve these.

Sleep, stress management, and suicide

Approximately half of people with tinnitus report some degree of sleep impairment and many people report their tinnitus escalates if they find themselves in a stressful situation. Referral to an insomnia or stress management service may be required. In the unlikely event that the patient discusses suicidal ideation, immediate referral to mental health services should be undertaken.

Address underlying hearing loss

A pivotal step in the management of tinnitus is generally to try and improve any hearing loss, even if the loss is mild and not at a level where communication is significantly impaired. There are several reasons behind this recommendation: firstly, it is thought that people with hearing loss strain to listen and this process heightens their tinnitus. Correcting the hearing loss is hypothesized to counteract this tendency. Use of external sound to partially mask or distract attention from the tinnitus is often used in tinnitus clinics and this is easier if hearing is optimized. Finally, on a pragmatic level, tinnitus management requires good communication between therapist and patient and this is easier once hearing loss has been corrected. This step may require the use of hearing aids, assistive listening devices, or, if the hearing loss is severe to profound, cochlear implantation. There is good evidence that cochlear implantation helps the majority of this subgroup of tinnitus sufferers, though there is a small risk of exacerbating any existing tinnitus or even triggering tinnitus when none was present preoperatively.

Sound therapy

Sound can be used to either try and mask the tinnitus percept or used at a lower level to blend with it, reducing its salience. There are many ways of achieving this, with the guiding principles that the sound used should be neither aversive nor overly interesting. For a patient whose tinnitus is mostly a problem at night in their sound-insulated bedroom, simply opening a window to admit normal environmental sounds may help. There are also electronic sound generators that can be used at night-time, producing low levels of repetitive, pleasant, non-threatening sounds, typically natural sounds such as surf, wind rustling trees, or running water. There are many apps available for mobile phones which perform the same function or CDs or MP3 downloads. Such electronically generated sound can be delivered via the device's loudspeaker, through a special speaker placed underneath the pillow, through a pillow with built-in speakers, or via a Bluetooth headband. Alternatively, some people prefer sounds from devices such as fish tank oxygenators, electric fans, ticking clocks, or indoor water features. Small ear-level devices are available for when people are ambulant. These resemble small hearing aids, are known as wearable sound generators, maskers, white noise generators, or tinnitus noisers, and supply low-level broadband sound.

Sound therapy is often utilized in the management of hyperacusis and in this situation it is used either at a constant low level to recalibrate the auditory system or at a varying level, just under the point where discomfort would occur, to desensitize the auditory system.

Combination devices

Ear-level devices are available that combine the functions of a hearing aid and a sound generator. The sound generator section may produce simple broadband sounds, more complex sounds, or may allow wireless streaming to the device so that any sound that the patient prefers may be used. Generally, the device is used on the hearing aid setting when in areas with significant ambient sound and the sound generator is reserved for use in quiet environments. Some devices switch automatically between the hearing and sound programmes depending on external sound levels whereas others require the patient to manually change programmes.

Drug treatments

There are no drugs that are currently recommended for the management of persistent spontaneous tinnitus. Drugs may have a role in the treatment of comorbid conditions such as anxiety and depression and in certain types of syndromic tinnitus, such as the use of fluoride in the management of otosclerosis. The recent NICE guideline on tinnitus has made a specific recommendation that betahistine should not be given to people with tinnitus with the exception of tinnitus secondary to Ménière's disease.

Combination therapies

There have been attempts at combining individual elements of tinnitus and hyperacusis management in structured protocols. These include tinnitus retraining therapy, progressive tinnitus management, and tinnitus activities treatment. Currently there is little robust evidence to say whether these are better than using an ad hoc approach.

Physiotherapy

Somatosensory tinnitus may benefit from physiotherapy to neck and jaw muscles. Nocturnal dental splints have also been proposed.

Dietary manipulation

There is no evidence that altering the diet affects tinnitus outcomes. In particular, there is no evidence that excluding caffeine helps.

Complementary and alternative medicine

There is no evidence that complementary and alternative medicine treatments prevent or cure tinnitus effects but many concentrate on relaxation, stress reduction, and promoting general well-being.

Psychological treatments

Of all the available management options for tinnitus, the modality with the largest supportive evidence base is psychological management. In particular, there is good evidence to recommend cognitive behaviour therapy (CBT) though there is also supportive evidence for newer options including mindfulness-based cognitive therapy, mindfulness-based stress reduction, and acceptance and commitment therapy. Such modalities seem effective in reducing tinnitus distress but are ineffective when the primary outcome measure of tinnitus loudness is considered. A stepped approach is recommended, starting with digital CBT delivered via computers, smartphones, or tablets; if this is ineffective, escalate to a group psychological intervention using CBT, mindfulness-based cognitive therapy, or acceptance and commitment therapy; if this is ineffective, offer individual CBT. In the UK access to such psychological management protocols is currently limited due to a dearth of appropriately trained clinicians.

Tips and pearls

- Be reassuring and optimistic. Do not use the phrase, 'There is nothing that can be done. You'll just have to live with it'.
- Offer information or direct the patient to a source of information such as the Tinnitus UK (http://www.tinnitus.org.uk).
- Offer to refer the patient onwards to tinnitus rehabilitation services in the audiology or psychology departments.
- If the tinnitus is associated with any degree of hearing loss, consider the use of hearing aids. Patients often find this difficult to understand as their complaint is tinnitus rather than deafness. Explaining that the hearing aid is being used to treat tinnitus rather than for communication is often helpful.
- Overuse of hearing protection devices tends to exacerbate hyperacusis. Explain to patients that they should only use ear defenders when they are in genuinely noisy environments.

Guidelines

NICE guideline [NG155], 'Tinnitus: assessment and management', was published in 2020 and its recommendations are summarized in **Box 6.3**. Similar guidelines have been produced in other regions including the US and Europe.

Controversies and areas of interest for the future

Non-psychologists delivering CBT

Because of the dearth of audiologically cognizant psychologists in the UK, it has been proposed that CBT should be delivered by audiologists using internet-based CBT. Initial studies have shown promising results and methods of incorporating this in the healthcare framework are being explored.

Tinnitus heterogeneity

Research suggests that tinnitus is far from a single entity and until it is possible to subtype tinnitus accurately it seems unlikely that curative options can be developed; different subtypes of tinnitus almost certainly need different treatments. To this effect, the development of a large tinnitus biobank has been suggested.

Acoustic neuromodulation

One of the suggested mechanisms for tinnitus is that it is due to abnormal synchronization of neural activity in the central auditory system (**Box 6.1**). Acoustic and dual-modality electroacoustic neuromodulation have been proposed as ways of disrupting this abnormal synchrony. Although preliminary work has been undertaken, large-scale independent research is needed.

Transcranial magnetic stimulation

Repetitive transcranial magnetic stimulation is a non-invasive way of stimulating superficial layers of the brain. It has a role in certain types of treatment-resistant depression and its use has been explored in tinnitus. However, tinnitus trials have tended to be small scale and used varying methodologies. Confusing and sometimes contradictory results have been obtained. Large-scale multicentre trials are needed.

Biomarkers

A major obstacle to tinnitus research is that there are no satisfactory biomarkers or objective outcome measures. Until these are developed, tinnitus research relies on unsatisfactory subjective measures.

Box 6.3 Summary of NICE guideline, 'Tinnitus: assessment and management'

Reassure		
Give information		
Refer for assessment and investigation: see full online document for details		
Assessment		
Overall impact	Tinnitus Functional Index Questionnaire (TFI)	
Quality of life	Discuss with patient, family, carers	
Sleep	Insomnia Severity Index (ISI)	
Psychological impact	Tinnitus Questionnaire (TQ) or mini-TQ	
	Assessment by a mental health professional	
Investigate		
Audiometry	Offer audiogram	
	Offer tympanometry	
	Do not routinely offer acoustic reflex testing, otoacoustic emission testing, tinnitus pitch, and loudness testing.	
Imaging	Non-pulsatile tinnitus plus associated neurological, otological, or head and neck signs and symptoms	MRI internal auditory meati (IAM). Contrast-enhanced CT IAM if contraindications to MRI
	Non-pulsatile tinnitus, if unilateral or asymmetric	MRI IAM. Contrast-enhanced CT IAM if contraindications to MRI
	Non-pulsatile tinnitus and no other symptoms or signs and symmetrical	Imaging not required
	Synchronous pulsatile tinnitus with no other symptoms and audiometry normal	MRI and magnetic resonance angiography head, neck, temporal bones, and IAM. Contrast-enhanced CT if contraindications to MRI
	Synchronous pulsatile tinnitus with osseous or middle ear pathology suspected	Contrast-enhanced CT of temporal bones +/– MRI
	Non-synchronous pulsatile tinnitus such as palatal myoclonus	MRI head. Contrast-enhanced CT if contraindications to MRI
Management		
Amplification devices	Offer if hearing loss affecting ability to communicate	
	Consider if there is a hearing loss but no communication difficulty	
	Do not offer if no hearing loss	
Sound therapy	Unable to reach a recommendation	
Psychological therapies	For people with tinnitus and psychological distress, use a stepped approach	Step 1: digital tinnitus-related CBT provided by psychologists
		Step 2: group-based tinnitus-related psychological interventions including mindfulness-based cognitive therapy (delivered by appropriately trained and supervised practitioners), acceptance and commitment therapy, or CBT (delivered by psychologists)
		Step 3: individual tinnitus-related CBT (delivered by psychologists)
Betahistine	Do not offer betahistine to treat tinnitus	
Combining therapies	Unable to reach a recommendation	
Neuromodulation	Unable to reach a recommendation	

Source: https://www.nice.org.uk/guidance/ng155

Conclusion

Although a cure for most types of tinnitus remains elusive, good, well-structured management plans are available. Despite this, patients report receiving negative counselling from many healthcare professionals. Research is more active than ever before but is hampered by the heterogeneity of the condition, unclear pathophysiology, and a lack of biomarkers and objective outcome measures. Understanding of disorders of sound tolerance is still at an early stage.

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7

Vertigo

Peter Rea and Joel A. Goebel

Scope of the problem

Dizziness is a common symptom affecting approximately 30% of the population before the age of 65, while for patients over 75 years of age, dizziness and/or imbalance are the most common reasons for visiting their family doctor. Falls caused by dizziness and poor balance in the elderly are associated with significant morbidity and mortality and constitute a major healthcare burden. In these cases, early detection and intervention can result in significant improvement in the quality of life as well as a decrease in disability and reduction in healthcare costs. It is important for those who diagnose and treat dizziness to recognize the complexity and impact of the problem, develop a systematic method for gathering accurate historical information, perform a reproducible neurotological examination, and institute management that addresses all major contributing factors.

Overview of balance and gaze stabilization

The ability to maintain upright posture and stable vision during head movement is a complex combination of sensory input, central sensory integration, and appropriate motor output (**Figure 7.1**). The vestibular labyrinth senses angular acceleration of the head (semicircular canals (SCCs)) and linear acceleration and gravity (utricle and saccule) and conveys this information to the eyes via the vestibulo-ocular reflex (VOR) and the vestibulospinal reflex for gaze and posture stabilization. However, visual, somatosensory, and auditory cues are also received by the brain to aid in balance and gaze stability and these inputs need to be properly integrated in the brainstem, cerebellum, and higher cortical centres to generate the appropriate motor response for posture control and gaze stability. Patients with dizziness and imbalance may suffer from any combination of insufficient sensory input, poor central integration, and suboptimal motor control.

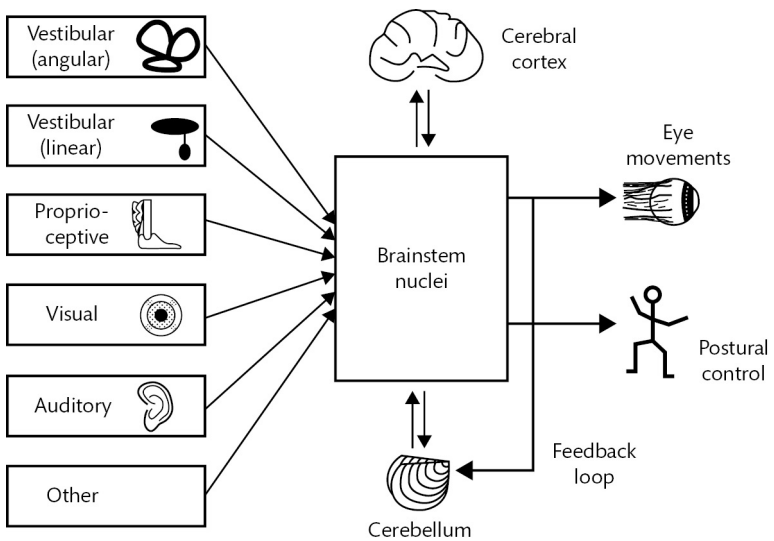

Figure 7.1 Integrated nature of balance and ocular motor control.
Reproduced from Hughes GB, Pensak ML, eds. *Clinical Otology*. 2nd ed. Thieme; 1997, with permission from Thieme Medical Publishers.

Anatomy and physiology of balance

The inner ear consists of two fluid-filled spaces housed within the otic capsule situated in the temporal bone on both sides of the skull in a mirror image. The perilymphatic spaces are in continuity with the cerebrospinal fluid via the cochlear aqueduct, whereas the endolymphatic space is self-contained and contains the sensory organs. The cochlea and vestibular labyrinth are in continuity via the ductus reuniens.

The sensory cell throughout the membranous labyrinth is the hair cell with specialized cilia at its apex (large kinocilium and smaller stereocilia) that modulate its neural firing rate from a baseline of 90–100 spikes/sec depending on the direction of ciliary deflection. In the vestibular labyrinth, these cells are further differentiated into type I and type II hair cells to encode high-frequency/impulsive and low-frequency/sustained head movement, respectively. In the SCCs, both types of hair cells populate the cristae ampullaris and are stimulated during angular head movement by deflection of a fan-like structure called the cupula. Since the otic capsules are situated in both temporal bones as a mirror image, the polarity of the hair cells is such that during head movement, the hair cells on one side are stimulated and the opposite side inhibited to create a differential firing rate that the brain interprets as head movement. It is important to note that hair cell stimulation (stereocilia towards kinocilium) produces far more neural activity than hair cell inhibition (kinocilium towards stereocilia). In the utricle and saccule, hair cells are embedded in the maculae and covered by a dense otolithic membrane that stimulates the hair cell during linear acceleration and changes in alignment to gravity. Taken together, the vestibular labyrinth senses both angular and linear head movement and conveys this information to the brainstem via two distinct pathways. The VOR conveys SCC input through the VIIIth cranial nerve, vestibular nuclei, and oculomotor nuclei for gaze stabilization. The vestibulospinal reflex relays otolith stimulation to the neck, torso, and extremity musculature to aid in posture control during linear acceleration and changes in orientation to gravity. There is also input from the otolith organs to the ocular muscles to aid in gaze stabilization during linear head translations (linear VOR) and head tilt (ocular tilt reaction). An example of the consequences of losing this stabilization is oscillopsia, when the patient reports the world 'bobbing up and down' as they walk.

The history

General observations

Watch your patient as they enter the examination room. Consider if they required assistance. Record their gait speed, stride length, and arm swing (e.g. in Parkinson's disease, a stooped, shuffling gait with loss of arm swing is observed). Did the patient touch or lean on objects to steady themselves? Listen to their gait in the corridor for stomping (the magnetic gait of normal-pressure hydrocephalus), or slapping (from foot drop).

History taking

The history is critical. In 75% of cases of chronic or recurrent dizziness and imbalance, the history alone yields an accurate differential diagnosis. This is because most causes of dizziness present a typical pattern of symptoms with stereotypic symptoms (e.g. dizziness on rolling over in bed is usually benign paroxysmal positional vertigo (BPPV) and vertigo with photophobia is usually migraine); peripheral, central, and systemic disorders typically have quite different symptoms; and most patients are not experiencing an acute sensation of dizziness at the time of examination. Hence, the development of a systematic approach to history taking allows the examiner to build a provisional differential diagnosis prior to performing the physical examination and further testing.

Symptom quality: vertigo, disequilibrium, and light-headedness

Types of dizziness can be roughly divided into three sensations: (1) vertigo, (2) disequilibrium, and (3) light-headedness. Distinguishing these alone is a powerful pointer to diagnosis as demonstrated in the balance disorder spectrum infographic (**Figure 7.2**).

Vertigo can be defined as a false sense of motion within the patient's environment and is usually described as a 'spinning, whirling, tumbling, or even rhythmic rocking' feeling. In most instances, patients will sense that the environment is in motion around them (objective vertigo) and may describe the illusion that objects within their visual world are actually moving in one direction before their eyes. In a minority of cases, the patients feel that they are in motion relative to a stationary world (subjective vertigo).

In contrast to vertigo, patients who have disequilibrium describe difficulty manoeuvring within their physical environment often without experiencing an illusion of motion. Patients with disequilibrium may use synonyms such as 'imbalanced, clumsy, uncoordinated, or fear of falling' while standing or walking. A variety of central nervous system, peripheral neuropathic, and musculoskeletal disorders can cause disequilibrium, although slowly progressive, bilateral vestibular loss without significant asymmetry may present with this complaint.

Patients with light-headedness describe a wide range of sensations such as 'wooziness, giddiness, feeling faint, or as if one is about to pass out'. Causes include migraine, persistent postural-perceptual dizziness, cardiovascular, metabolic, drug-induced, endocrine, or primary psychogenic disorders. Although light-headedness is less commonly the primary complaint of patients with damage of the peripheral vestibular pathways, it frequently intermixes with vertigo and/or disequilibrium.

Duration of dizziness

A critical question is whether the dizziness is continuous or episodic and, if so, do the episodes last seconds, minutes, hours, days, or even longer? When patients complain of vertigo, this time course distinction is exceedingly valuable since the most common peripheral vestibular disease processes produce stereotypical attacks of consistent duration. Patients with BPPV usually complain of discrete attacks that last less than 1 minute. In contrast, patients with Ménière's disease usually describe a 'spinning feeling' that can last 15 minutes to many hours. Finally, patients who develop vertigo secondary to vestibular neuritis describe attack(s) of continuous spinning lasting up to 24 hours. In patients with transient ischaemic attacks involving the vertebrobasilar circulation and brainstem, attacks generally last

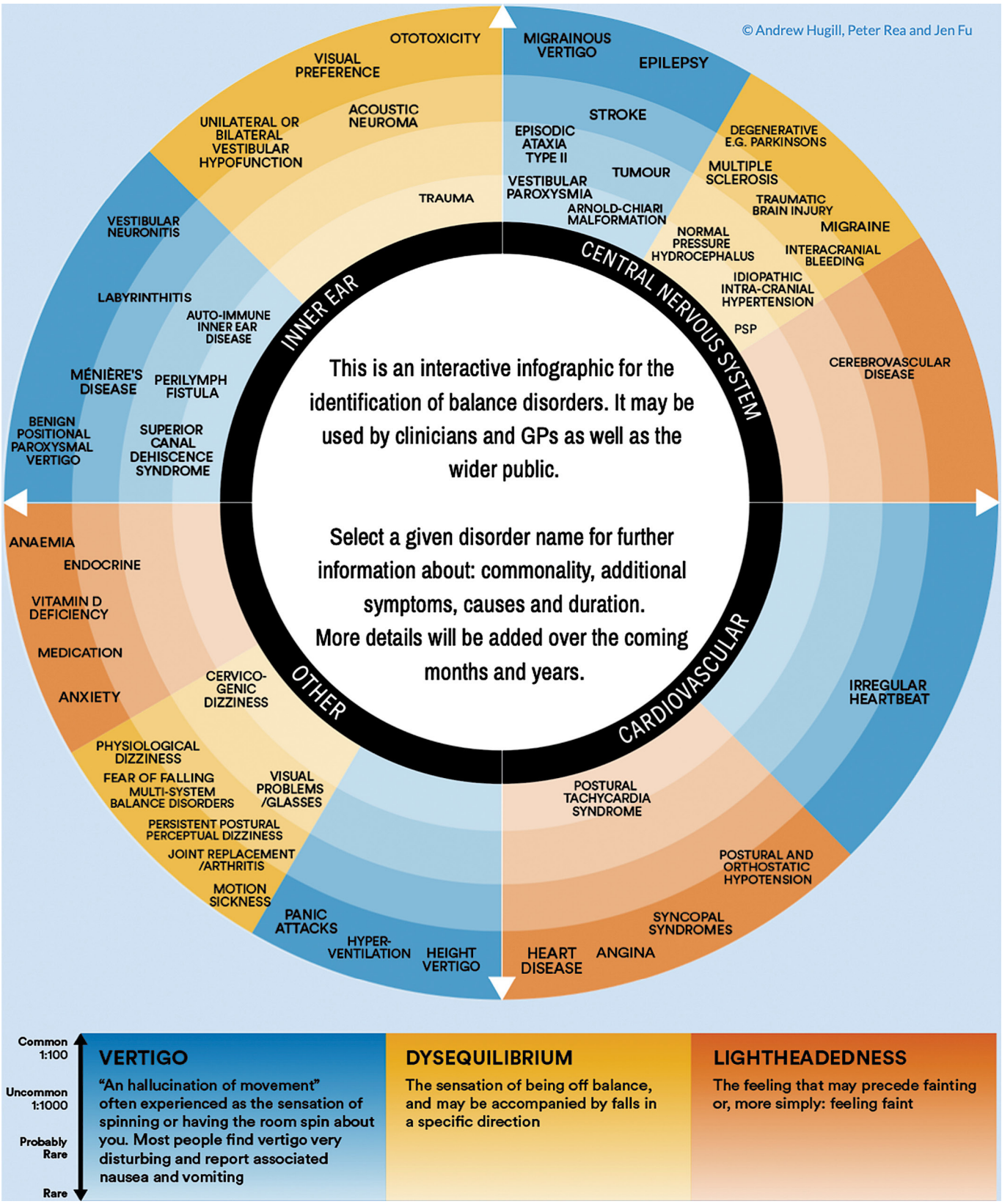

Figure 7.2 The balance disorder spectrum.

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15 minutes, whereas brainstem infarcts and cerebellar haemorrhages exhibit acute, severe vertigo lasting hours and impact the inability to stand.

The time course of attacks in patients with disequilibrium is somewhat less stereotypical of their disease process. Frequently, patients report that symptoms persist as long as they are upright. Similarly, patients with light-headedness may have a variable symptom picture, with sensations lasting anywhere from seconds to days. Finally, patients with vestibular migraine may complain variably of all three sensations, and the duration may fluctuate from seconds to days or even longer. Vestibular migraine often coexists with other vestibular disorders, adding to the diagnostic challenge.

Potential triggering events

Consider position change (dizziness on rolling over in bed or looking up is usually BPPV), diet (caffeine is one of many migraine triggers, for example), lifestyle (ask about sleep apnoea and insomnia), or pressure change (straining causing dizziness may be associated with a third-window disorder).

In cases of bilateral and uncompensated unilateral vestibular dysfunction, patients have difficulty walking in the dark or on uneven surfaces and avoid fast head movements due to bobbing or blurring of their visual field (oscillopsia). Motion sensitivity is also characteristic of persistent postural-perceptual dizziness and migraine. Patients with orthostasis become more symptomatic with changes in position against gravity such as getting up from a chair and obtain relief by lying flat.

Identify associated events that occurred near the time the dizziness began, such as trauma, recent or chronic infection(s), systemic diseases, or medication changes. Post-traumatic vertigo may be due to a perilymphatic fistula of the oval and/or round windows, delayed endolymphatic hydrops, BPPV, migraine, or middle ear surgery. Furthermore, trauma may open a dehiscence of the superior SCC if the bone covering the canal was thin prior to the injury.

Determining a history of recent infections is important. For example, vestibular neuritis is thought to be caused by viral infection, and an upper respiratory viral prodrome may occur prior to vertigo onset. Other infections that can cause vertigo include herpes zoster oticus (Ramsay Hunt syndrome), suppurative otitis media, human immunodeficiency virus, syphilis, Lyme disease, and tuberculosis.

Associated symptoms

Hearing loss, tinnitus, aural fullness, and vertigo strongly suggest labyrinthine involvement, but can also be seen in migraine. Fluctuations in these symptoms may suggest Ménière's disease. One important exception is acute vertigo with hearing loss that may indicate a brainstem stroke (anterior inferior cerebellar artery (AICA)). On the other hand, vertigo without hearing loss may be generated by the labyrinth (e.g. BPPV), VIIIth cranial nerve (e.g. vestibular neuritis), brainstem, and/or cerebral cortex (e.g. migraine, seizure). Patients with superior semicircular canal dehiscence (SSCD) can present with conductive hyperacusis or autophony. Conductive hyperacusis includes symptoms such as hearing one's own eye movements, one's own heartbeat, and/or the impact of one's feet during walking or running in the affected ear. Patients with vestibular migraine frequently complain of photophobia, phonophobia, or a heightened sense of smell. Finally, loss of consciousness implies a significant perfusion failure of the cerebrovascular circulation and is not usually associated with primary labyrinthine or VIIIth cranial nerve lesions.

Medication side effects

Dizziness is a common side effect of many medications, and, in some instances, the interaction of multiple medicines is the cause of a patient's symptoms. Be particularly alert to polypharmacy in the elderly. Medications causing dizziness, light-headedness, and/or auditory symptoms are generally divided into three categories: those that (1) are ototoxic (e.g. aminoglycosides, and chemotherapeutic agents), (2) affect systemic blood flow (especially antihypertensives), and (3) act on the central nervous system (prochlorperazine taken over a long period is frequently seen to cause Parkinsonian symptoms in patients seen in the balance clinic, and any antidepressants and anxiolytics should be reviewed).

Outpatient examination of the dizzy patient

The most effective clinical examination of the dizzy patient is a structured, stand-alone series of tests after the standard otolaryngological head and neck examination has been completed. Common disorders such as serous middle ear effusion, nasal airway obstruction, and sinusitis can cause dizziness and must be excluded. In the following section, each test is described and the interpretation discussed.

Equipment

Three tuning forks are used: 128, 256, and 512 Hz. The 128 Hz fork is used to assess vibrotactile sensation in the lower extremities (for peripheral neuropathy). The 256 Hz is used for the Rinne and Weber test, diplacusis and hyperacusis in Ménière's disease, and the malleolar test when patients with a third-window pathology (e.g. SSCD) can abnormally perceive the sound in the affected ear while the fork is placed on the ankle. The 512 Hz fork is the standard auditory stimulus for Weber and Rinne testing. Frenzel glasses allow for ocular examination without visual fixation, which can enhance peripheral vestibular nystagmus. The pneumatic otoscope (including a Siegel speculum) is used to introduce positive and negative pressures into the external auditory canal to elicit eye movement associated with SSCD or perilymph fistula. Finally, the 3-inch foam pad is used to alter sensory input to the proprioceptive system during quiet stance with eyes closed to emphasize the use of vestibular cues for balance (the clinical test of sensory interaction and balance) (**Figure 7.3**).

The neurotological examination

See **Box 7.1** for a summary list of the neurotological examination.

Eye movements form a critical part of the assessment as no clinical test can directly measure peripheral vestibular function. The most common type of nystagmus is jerk nystagmus, which consists of a slow phase and a fast phase. Vestibular control of eye movements results in the slow phase, while the fast phase is caused by the rapid resetting of the eyes in the orbit. Jerk nystagmus is named by the direction of the fast phase (e.g. left-beating horizontal).

Under normal conditions, there is no spontaneous nystagmus with visual fixation and only minimal spontaneous nystagmus if any under Frenzel lenses. Vestibular nystagmus is usually direction fixed; that is, regardless of the direction of the patient's gaze, the

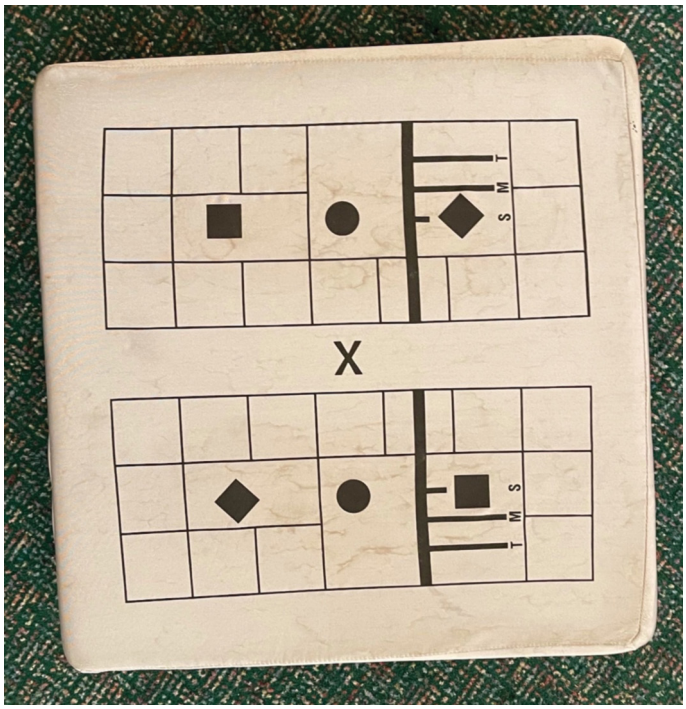

Figure 7.3 The clinical test of sensory interaction and balance.

Box 7.1 The neurotological examination

Gait and posture
- Gait.
- Tandem gait.
- Romberg.
- Unterberger.
- Clinical test of sensory interaction and balance.

Eye movements
- Gaze-evoked nystagmus.
- Smooth pursuit.
- Saccades.

Limb coordination tests
- Finger–nose–finger.
- Heal shin.
- Thumb to fingers.

Sensory
- Pin-prick.
- Joint position.
- Reflexes.

Miscellaneous
- Head impulse test (HIT).
- Test of skew.
- Post-headshake nystagmus.
- Dynamic visual acuity.
- Position testing (including Hallpike's).
- Malleolar testing.
- Pressure- and sound-induced nystagmus.
- Lying and standing pulse and blood pressure.
- Plantar responses.

direction of the nystagmus does not change. Unilateral, vestibular hypofunction causes an imbalance in the tonic activity of the VOR and tends to cause eye movement in the plane of the damaged SCC. This input asymmetry usually results in a spontaneous, direction-fixed, horizontal-rotary jerk nystagmus with fast-phase movement towards the healthy ear (away from the damaged side). Removing visual fixation (e.g. placing Frenzel lenses on a patient) or gazing in the direction of the fast phase enhances the nystagmus, while gazing in the direction of the slow phase suppresses the nystagmus. This effect is called Alexander law. Alexander classified nystagmus as first-, second-, and third-degree nystagmus. First-degree nystagmus is the least intense and is only observed with gaze towards the fast phase. Second-degree nystagmus is more intense and is observed with the eyes in primary gaze position or when gazing towards the fast phase. Third-degree nystagmus is the most intense and is present when the eyes are in primary gaze position, gazing towards the fast phase or gazing in the direction of the slow phase. The nystagmus intensity declines as the central system compensates.

In contrast, central lesions of the brainstem and cerebellum may cause direction-changing horizontal, vertical, torsional, or pendular nystagmus that may appear diminished under Frenzel lenses. Medications and alcohol may also induce a variety of nystagmus patterns.

Gaze-evoked nystagmus (GEN)

GEN is a type of nystagmus that results when the eyes assume an eccentric position in the orbit. Transient GEN is normal at extreme gaze (>30° from midline) and is called end-point nystagmus. However, any rhythmic eye movement with attempts to hold the eyes in an eccentric position within 0–30° of midline is abnormal. The patient follows the examiner's finger in the horizontal and vertical planes.

GEN may be observed in peripheral vestibular dysfunction or in lesions of the brainstem or vestibulocerebellum. Direction-fixed GEN that increases while gazing in the direction of the fast phase (Alexander law) suggests unilateral peripheral vestibular dysfunction. Direction-changing GEN with fast-phase movement in the direction of gaze and/or the presence of rebound nystagmus in primary gaze indicates impairment of the brainstem or midline cerebellum (vestibulocerebellum).

Smooth pursuit

Smooth pursuit is the visual tracking of moving objects caused by either target movement and/or movement of the viewer. Smooth pursuit requires intact central optic tracts through the brainstem, visual cortices in the occipital lobes, and the flocculonodular lobes of the cerebellum. The cerebellum ensures accurate pursuit by minimizing slippage of the image off the fovea. Next, the pursuit signal is compared and integrated with vestibular input in the brainstem, and a unified signal is transmitted to the oculomotor nuclei. Finally, the appropriate ocular muscle response is generated. It is clear, therefore, that abnormal ocular pursuit can arise from a variety of lesions within this complex pathway.

To assess smooth pursuit, the examiner positions his/her index finger directly in front of the patient and moves the target smoothly 20–30° per second, first in the horizontal plane and then in the vertical plane. The testing area is restricted to the central 60° of the visual field (30° to the left, right, up, and down from neutral position) to avoid provoking physiological end-gaze nystagmus.

Smooth pursuit is generally intact in patients with peripheral vestibular disease. Occasionally, patients with acute unilateral vestibular dysfunction and spontaneous nystagmus may exhibit impaired tracking when they move their eyes in the direction of the fast phase of their spontaneous nystagmus. Patients with poor visual acuity or inattention to the task demonstrate large irregular fast eye movements (saccades) to catch up to the target. In central lesions, a more global failure of pursuit is evident. Catch-up saccades refixate the eye on the visual target in order to compensate for the pursuit deficit (saccadic pursuit).

Most abnormalities affect smooth pursuit symmetrically. A common cause is medication, such as anticonvulsants and sedatives, as well as alcohol. Smooth pursuit performance progressively deteriorates as a patient's age increases or visual acuity declines. Furthermore, smooth pursuit is impaired by neurological conditions such as Parkinson's disease, Alzheimer's disease, supranuclear degeneration, and cerebellar degeneration.

Saccades

Saccadic eye movements involve the frontal and parietal eye fields, paramedian pontine reticular formation, medial longitudinal fasciculus, superior colliculus, and oculomotor nuclei III, IV, and VI.

Saccadic eye movements are examined by having the patient alternatively fixate on two stationary targets without moving their head. The patient is asked to look back and forth between the examiner's nose (in neutral position) and fingertip (held 15° off midline) (repeat in the right and left horizontal plane as well as up and down in the vertical plane). Observe for the following eye movement characteristics: accuracy of target acquisition, conjugate movement, velocity (especially slowing), and latency of onset (initiation). Abnormalities in saccadic eye movements may be difficult to detect in the clinic examination. Videonystagmography (VNG) may be necessary in order to identify subtle abnormalities.

Patients with peripheral vestibular pathology have normal saccadic eye movement; oculomotor abnormalities indicate ocular or central nervous system pathology. The midline cerebellum and fastigial nuclei control saccadic accuracy, whereas velocity, latency, and conjugate deviation are controlled by the brainstem and frontal eye fields. Abnormalities in saccadic initiation may be seen in Parkinson's disease. Small-amplitude saccades are characteristic of myasthenia gravis or an abnormality in the orbit. Cortical and brainstem diseases exhibit slow saccades. Progressive supranuclear palsy is characterized by slow vertical saccades initially as well as slow and hypometric (undershoot) horizontal saccades. Olivopontocerebellar atrophy, also known as spinocerebellar ataxia, is characterized by slow saccades, especially in the horizontal direction. Inaccurate saccades (dysmetria) are associated with cerebellar vermis and fastigial nuclei lesions. Pathology of the medial longitudinal fasciculus produces internuclear ophthalmoplegia, which is frequently associated with multiple sclerosis if bilateral. Internuclear ophthalmoplegia is characterized by dysconjugate eye movements with slowing of the adducting eye and overshoots and/or nystagmus of the abducting eye. Internuclear ophthalmoplegia is often associated with a monocular nystagmus in the adducting eye contralateral to the lesion.

Head impulse test (HIT)

The HIT is a bedside technique used to detect unilateral or bilateral vestibular hypofunction. To understand this test (and many of the following tests), one must understand Ewald's three laws. Ewald's first law states that the trajectory of nystagmus generated by an SCC is in the same anatomic plane of that particular SCC. Ewald's second law is based on experiments conducted on the horizontal SCC and states that ampullopetal endolymph flow (towards the ampulla) causes a greater vestibular response than ampullofugal endolymph flow (away from the ampulla). In other words, excitatory input is a stronger vestibular stimulus than inhibitory input. Ewald's third law is based on experiments conducted on the vertical (superior and posterior) SCCs. These experiments found that ampullofugal endolymph flow produces a greater vestibular response than does ampullopetal endolymph flow. Therefore, ampullofugal endolymph flow is excitatory in the two vertical SCCs.

The HIT is a method of detecting asymmetries in vestibular gain from the SCCs. In patients with profound or absent SCC function, the VOR cannot produce signals to the ocular muscles for eye stabilization during high-frequency, passive angular head rotations. Consequently, the brain must generate a refixation saccade to acquire a visual target. Saccade refixation occurs in response to passive angular head movements towards the involved ear in patients with unilateral vestibular loss. Covert saccades are small corrective saccades that occur during the actual head movement towards their affected side. These small covert saccades are difficult for the examiner to detect with their naked eye. Therefore, eye movement recordings are required to detect the abnormality (using the video head impulse test (vHIT)). This HIT can be performed in all three canal pair orientations and has become a significant addition to the neurotological examination in the office.

The patient is instructed to fixate on the examiner's nose while the head is turned 20–30° from centre and impulsively and unpredictably moved back to midline. The examiner observes for any indication of ocular globe slippage off the target. The velocity of the head movement must exceed 200° per second, and the acceleration must exceed 2000° per square second in order to exceed smooth pursuit capacity and fully inhibit the contribution of the contralateral ear for stabilization. To avoid neck injury, it is advisable to start at slower speeds and gradually increase to the desired velocity. The HIT is best performed in the plane of paired canals. If the horizontal SCCs are being tested, the head is tilted forward 30°. Hence, the movement is in the horizontal (yaw) plane, causing ampullopetal stimulation of the canal on the same side as the movement and ampullofugal inhibition of the opposite canal. As noted, it is desirable to move the head from an eccentric position back to midline rather than from midline to the side in order to minimize patient resistance and potential neck strain.

In healthy subjects, visual fixation is maintained during rapid head impulses, and no corrective saccades result. In cases of unilateral SCC dysfunction, head movement in the plane of the affected canal towards the lesioned side is accompanied by a repeatable eye slippage and corrective saccade in the opposite direction as the head movement. A positive HIT in the presence of acute vertigo and spontaneous nystagmus is suggestive of a peripheral rather than central disorder.

Skew deviation and ocular torsion reaction test

Skew deviation is a vertical misalignment of the eyes, which results from asymmetric activity along the peripheral or central pathways mediating the otolith–ocular reflex. Skew deviation often presents as

vertical or torsional diplopia. First, the examiner must rule out that the ocular misalignment is due to extraocular muscle palsies.

The alternate cover test is used to detect skew deviation. The examiner asks the patient to fixate on a visual target and then covers one eye. The examiner then uncovers that eye and covers the contralateral eye, observing for a vertical refixation eye movement indicating a vertical mal-alignment.

In the normal condition, the eyes do not move or deviate spontaneously in the vertical plane. However, if there is skew deviation, vertical corrective saccades will occur during pursuit.

Post-headshake nystagmus test

During head shaking, the vestibular labyrinths send neural input to the brainstem. As soon as head shaking ceases, this stored input is discharged over a short period of time (10–20 seconds). Unilateral damage to the peripheral vestibular organs leads to asymmetric input and discharge resulting in nystagmus. Clinical head-shaking nystagmus is useful in detecting asymmetric peripheral vestibular lesions or asymmetric central lesions of the velocity storage mechanism.

Dynamic visual acuity

Maintenance of gaze stability during head movement is a complex interaction between the VOR, foveal smooth pursuit, optokinetic stimulation of the peripheral retina, and the cervico-ocular reflex.

With best-corrected vision, visual acuity is measured at rest using the Snellen eye chart. Visual acuity is again measured during passive horizontal head oscillations at 2 Hz. The change in visual acuity is determined. An acuity decrease of more than two lines on the Snellen eye chart is abnormal. Excessive retinal slippage during head movement is a sign of vestibular dysfunction.

Position testing

Nystagmus and vertigo can be induced by certain head postures (positional) or head movement in a specific trajectory (positioning). In other words, positional nystagmus is a nystagmus induced by a static head position in space. Positioning nystagmus is induced by the actual movement of the head from one position in space to another. The goal of positional and positioning testing is to aid in vestibular lesion localization.

Positional testing

Positional testing involves observing the eyes for nystagmus in various static head positions with and without visual fixation. Observe the eyes for at least 30 seconds in each of the following head positions: upright looking straight ahead, supine, right ear down, and left ear down. If positional nystagmus is present, it will occur approximately 30 seconds after the new head position is assumed, and the effects of positioning head movement (positioning nystagmus) have waned. The examiner should note the nystagmus direction, effect with and without visual fixation, and if it is persistent and sustained. Removing visual fixation (Frenzel lenses or infrared video goggles) worsens peripheral positional nystagmus, but central positional nystagmus improves or stays the same. Technically, true BPPV is a positioning nystagmus and occurs in the plane of the canal that is stimulated, whereas central positional nystagmus may beat in the same direction regardless of head position.

Positional nystagmus can be found in both central and peripheral pathologies. Central positional nystagmus is usually asymptomatic, persistent, and may be dysconjugate. Peripheral positional nystagmus is associated with severe vertigo, transient, and usually direction fixed. A central lesion is the most likely aetiology when positional nystagmus is purely vertical (up or down) or purely torsional, and visual target fixation fails to suppress nystagmus.

Positioning testing

Positioning testing is the evaluation of nystagmus in response to head movement. Positioning testing can also help distinguish central from peripheral aetiologies, determine which ear is damaged, and localize the involved SCC. To perform positioning testing, the eyes are observed as the head is moved into various positions: upright, supine, right ear down, and left ear down. Preventing visual fixation (Frenzel lenses or infrared video goggles) may enhance peripheral positioning nystagmus and may diminish central positioning nystagmus.

BPPV is a positioning nystagmus. The most common positioning manoeuvre is the Dix–Hallpike test, which is used to identify posterior canal BPPV (**Figure 7.4**). While the patient is in a sitting position, the patient's head is turned 45° towards the side to be tested. The patient's head is rapidly moved in the plane of the posterior SCC to a head-hanging, slightly extended position (120° from the upright position). This position is held for 30 seconds. While keeping the head turned 45°, the patient is brought back to an upright position and held for 30 seconds while observing the eyes for nystagmus. This manoeuvre is then repeated on the contralateral side. Alternatively, the head may be rolled from 45° one side straight over to 45° the other, keeping the neck slightly extended. The examiner may need to perform a modified positioning manoeuvre in order to induce

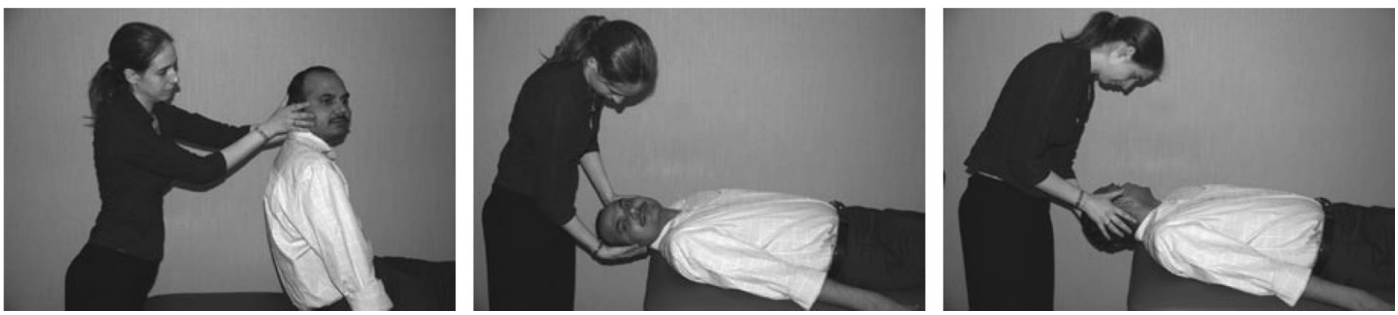

Figure 7.4 The Dix–Hallpike test.
Reproduced from Rea P. Disorders of balance. In: Graham J, Baguley D, eds. *Ballantyne's Deafness*. 7th ed. Wiley; 2009, with permission from John Wiley and Sons.

peripheral positioning nystagmus caused by horizontal SCC BPPV. In this manoeuvre, the patient is placed into the supine position, followed by turning the patient's head and body 90° to the right side, back to supine, and then turn 90° to the left side.

For BPPV of the posterior SCC, a positive response is elicited when the head is turned towards the affected side, and the ear is positioned towards the ground. The resulting nystagmus is upbeating, geotropic torsional (upper pole of eye towards the ground). The nystagmus has a brief latency (5–20 seconds), less than 30 seconds' duration, fatigability with repeated positioning, and, in some instances, reversal nystagmus upon returning to an upright position. The reversal nystagmus is a downbeating, ageotropic-torsional (upper pole of eye away from the ground) nystagmus. Vertigo without observed nystagmus may still provide a clue to the offending side and involved SCC. A positive Dix–Hallpike manoeuvre is diagnostic for posterior canal BPPV. In contrast, central positioning nystagmus is usually immediate (no latency), persists longer than 1 minute, does not produce a reversal nystagmus, does not fatigue, is direction changing, and is not usually accompanied by vertigo.

BPPV can involve the horizontal SCC and may be spontaneous or result from a repositioning manoeuvre of the posterior SCC. Depending on the location of the debris within the horizontal SCC, the pattern of nystagmus is either horizontal geotropic (towards the ground) or horizontal ageotropic (away from the ground). The direction alone does not guide to the side of the horizontal BPPV.

Superior SCC BPPV is rare. Otolithic debris does not usually enter the SCC due to its anatomic position. In superior SCC BPPV, when the affected ear is away from the ground, a downbeating, ageotropic-torsional nystagmus results. However, debris in the downmost posterior SCC moving towards the cupula can produce an identical nystagmus pattern to that of the uppermost superior SCC. Therefore, special care must be taken to avoid confusing the two types.

For further details on BPPV see Chapter 15.

Limb coordination tests

Examining limb coordination provides the examiner with important clues needed to diagnose and exclude central lesions of the cerebellum and brainstem. Peripheral vestibular lesions will not cause limb coordination abnormalities.

For the finger–nose–finger test, the patient should alternate touching the examiner's finger and his/her own nose. As the patient touches their nose, the examiner quickly moves their finger to a new horizontal position. This test assesses the patient's ability to accurately judge the position of a target. To assess fine finger movements, the examiner asks the patient to touch their own thumb to each of their four fingers on the same hand in rapid sequence. Slowing (bradykinesia) is associated with Parkinson's disease. To test lower limb coordination, the examiner can use the heel-to-shin test. The patient is asked to elevate their right leg straight, while lying on the couch. The patient then smoothly runs the heel of the right foot along the shin of the left leg. The test is repeated with the left leg elevated. During each of the above tests, the examiner observes for dysmetria or dysrhythmia.

The presence of limb dysmetria or dysdiadochokinesia is a useful indicator of cerebellar disease, which may or may not accompany midline or vestibulocerebellar oculomotor dysfunction. Peripheral vestibular dysfunction does not cause limb coordination abnormalities.

Sensory testing

Test peripheral sensation with a Neurotip on the great toes—diabetic neuropathy is the most commonly identified disorder. Assesses reflexes—the ankle jerks are most sensitive for peripheral neuropathy. Assess proprioception with up/down movement of the great toes with the patient's eyes closed. Finally, place a 128 Hz tuning fork on the malleolus of the ankle and compare the vibratory sensation to that at the wrist.

Gait and posture tests

For the tandem gait test, the patient steps heel to toe in a tandem manner, five paces, across the room. With the eyes open, this test assesses cerebellar function, but is difficult for the elderly.

For the Romberg test, the patient is asked to stand with feet together and arms at the side, first with eyes open and then with eyes closed. The examiner observes the relative amount of sway, both with and without vision, and notes if the patient falls consistently to one side. The Romberg stance primarily tests somatosensory and proprioceptive functions and not the integrity of vestibular inputs.

'Vestibular gait' does not define any specific manner and/or type of movement. Indeed, a broad array of characteristics can qualify a gait as abnormal. Furthermore, a number of causes can be the source(s) of the abnormal gait. If a patient suffers an acute unilateral loss of otolithic function, the patient will tend to veer towards the side of the lesion. Patients often rapidly compensate. Nevertheless, a variety of central brainstem and musculoskeletal lesions also produce lateral deviation during ambulation. Difficulties with gait initiation and turns as well as decreased arm swing can be seen in extrapyramidal disease. Gait ataxia implies cerebellar dysfunction and is distinctly different from the gait deviation associated with uncompensated peripheral vestibular disease. A shuffling gait should prompt the examiner to explore the possibility of Parkinson's disease and/or normal-pressure hydrocephalus. Finally, exaggerated hip sway, rhythmic deviations, and an excessive reliance on touching the wall during ambulation may constitute signs of a functional gait disorder.

The stepping test was first described in 1938 by Unterberger and later modified by Fukuda in 1959. The ability to close one's eyes and step in place without turning depends on normal vestibulospinal and proprioceptive function. Most normal subjects deviate less than 30° in rotation to one side during the step test, whereas some patients with uncompensated unilateral vestibular weakness deviate more than 30° towards the weaker side. It is important to note that patients with chronic vestibular dysfunction that have compensated for their vestibular loss may perform normally on the Unterberger (Fukuda) stepping test.

The 'malleolar sign'

Patients with SSCD may have hyperacusis to bone-conducted sounds and can present with symptoms of hearing their own pulse and/or the movements of their eyes. The bone-conducted hyperacusis can be explained by the 'third mobile window' effect. To assess this unique suprathreshold bone conduction, the examiner places a 256 Hz tuning fork on the patient's malleolus (ankle). A patient with SSCD may hear the tone in the affected ear.

Sound-evoked or pressure-evoked eye movements

Consistent eye deviations or nystagmus induced by sound or pressure changes in the middle ear are abnormal. The most commonly involved sites are the oval window (perilymph fistula, excessive footplate movement), round window (perilymph fistula, enlarged cochlear aqueduct), lateral SCC (perilymph fistula), and superior SCC (dehiscence).

Hennebert's sign is vertigo and/or nystagmus evoked by pressure-induced movement of the tympanic membrane and may occur in patients with perilymph fistula, otic syphilis, otic capsule erosion from disease, or SCC dehiscence. Pressure changes in the middle ear can be induced by tragal compression, pneumatic otoscopy (through a Siegel speculum), or Valsalva manoeuvres either through a closed glottis straining, or open glottis with nose pinched. Eye movements are typically conjugate.

Tullio phenomenon

This is the occurrence of vertigo and nystagmus with exposure to sound. The examiner introduces pure tones over a range of 250–4000 Hz at intensities of 100–110 dB. The characteristic eye findings are similar to those resulting from increased middle ear pressure.

Hyperventilation-induced nystagmus

Hyperventilation may induce dizziness and light-headedness in patients with psychiatric disorders (e.g. anxiety or panic disorder), but does not result in nystagmus. Hyperventilation does induce nystagmus in demyelination of the vestibular nerve (e.g. acoustic schwannoma or microvascular nerve compression) or central demyelination conditions (e.g. multiple sclerosis). Frenzel lenses will need to be in place, and the patient is asked to hyperventilate for 90 seconds. Hyperventilation-induced nystagmus is an excitatory nystagmus with the slow phase towards the healthy ear.

Investigations

While the history is often the key to vestibular diagnosis, laboratory testing can play an important part of the diagnostic process. It can help distinguish central from peripheral disorders, confirm a clinical diagnostic hypothesis, help quantify deficits, and offers greater sensitivity than clinical testing. However, each test answers a specific question. Calorics, vHIT, and the rotatory chair assess labyrinthine function during rotatory acceleration, posturography assesses postural stability, and vestibular evoked myogenic potentials (VEMPs) address otolith organ function, while electrocochleography (ECochG) assesses Ménière's disease. Simply requesting 'balance tests' is unlikely to be helpful.

Audiometry

Most dizzy patients will require an audiogram, the exception being classical BPPV where no laboratory investigations are required. Many causes of dizziness have classical associations with audiometric configurations, such as the low-frequency sensorineural hearing loss of Ménière's disease and the artificially low bone conduction thresholds of SSCD.

Electronystagmography and videonystagmography

Most vestibular test batteries begin by assessing the integrity of the three main ocular tracking mechanisms which are the saccadic, smooth pursuit, and the optokinetic systems. In order to record eye movements, many laboratories utilize VNG, which is a system whereby the patient wears goggles with an inbuilt infrared camera (so that it can operate in the dark) that can track eye movements. Alternatively, electronystagmography can still be used if VNG is not available or appropriate. Electronystagmography records from electrodes placed near the eye. The saccades and smooth pursuit recorded are those described in the earlier clinical section. But the data can be quantified and eye movements with and without optic fixation (i.e. recording in light and dark) are more easily obtained (**Figure 7.5**).

Caloric testing

Caloric testing is often considered the 'gold standard' test for unilateral and post-traumatic vestibular injuries.

Although only the lateral SCC is being assessed, it is sometimes taken as a proxy for overall peripheral vestibular function. It has the advantages of being a largely objective test, and the weakness, if present, persists in the long term which makes it especially useful for assessing chronic imbalance.

However, only very low-frequency motion is being assessed (that equates to very slow head turn), and a significant reduction in function is required for a significant difference to be seen between the ears. A 20–25% inter-aural difference is usually considered significant, but this will vary with local protocols.

The patient is positioned on a couch with their head at 30° to the horizontal bringing the lateral canals into the vertical plane of gravity. A thermal stimulus is then introduced via warm and cool water or air. This temperature gradient may induce convection currents in the endolymph resulting in deflection of the cupula, or simply change the excitability of the labyrinth. Given that only one ear is irrigated at a time, an asymmetry in neural firing between the left and right sides is created and as a result nystagmus is generated. Cold irrigation causes nystagmus to the opposite ear, warm to the same (COWS: cold

Figure 7.5 Videonystagmography.

Reproduced from Rea P, Sandhu J. Tests for balance. In: Musheer Hussain S, ed. *Logan Turner's Diseases of the Nose, Throat and Ear, Head and Neck Surgery*. 11th ed. CRC Press, 2015.

opposite warm same). This is then recorded using either VNG or electronystagmography and a canal paresis (CP) and directional preponderance (DP) calculated using the following equations:

$$\mathrm{CP}(\%) = \left[\frac{(R_{Warm} + R_{Cool}) - (L_{Warm} + L_{Cool})}{(R_{Warm} + R_{Cool} + L_{Warm} + L_{Cool})}\right] \times 100$$

$$\mathrm{DP}(\%) = \left[\frac{(R_{Warm} + L_{Cool}) - (L_{Warm} + R_{Cool})}{(R_{Warm} + R_{Cool} + L_{Warm} + L_{Cool})}\right] \times 100$$

The CP indicates reduction in function of one side compared to the other while DP indicates the propensity of the eyes to deviate in one direction over another and indicates asymmetric neural activity in the vestibular system as a whole. DP, however, is relatively nonspecific and does not specify the side or location of a vestibular lesion, and can occasionally be abnormal in a normal subject.

The caloric test can result in vertigo and nausea and as a result is not always well tolerated by patients, especially migraineurs.

Rotatory chair testing

During this test the patient is seated in a chair without optic fixation. The chair can rotate in either direction stimulating both lateral SCCs and the resulting nystagmus is recorded using either electronystagmography or VNG.

This test is usually only available in specialist balance centres and provides a less provocative alternative to the caloric test and hence can be more acceptable and well tolerated by children. It is also superior to the caloric test in cases of bilateral vestibular hypofunction. However, the localizing value of the test is limited as both horizontal canals are stimulated simultaneously and the equipment is expensive.

Video head impulse test

vHIT is the laboratory iteration of the clinical HIT test. It allows accurate recording of both overt and covert saccades. Saccade refixation occurs in response to rapid passive angular head movements towards the involved ear in patients with unilateral vestibular loss. Covert saccades are small corrective saccades that occur during the actual head movement towards their affected side. These small covert saccades are difficult for the examiner to detect with the naked eye. It is very helpful in identifying unilateral vestibular weakness and can be used in cases where this is not possible with calorics (e.g. in a wet ear, or mastoid cavity). And unlike calorics which assess very slow angular acceleration, the vHIT assesses more physiological and rapid rotations. Using a range of tests helps build a picture of vestibular responses through the rotation frequency range.

Computerized dynamic posturography

Dynamic posturography allows postural sway to be quantified in a number of conditions where visual and somatosensory cues are either absent or altered in some way. The test is sensitive to a number of neurological disorders and is not specific for vestibular deficits although patients with uncompensated vestibular deficits or bilateral hypofunction have difficulty maintaining their balance when visual and somatosensory cues are altered. The tool is very useful in establishing treatment and monitoring progress during rehabilitation. It has the added advantage that the patients can observe their own progress, which can be beneficial. It is able to characterize 'visual preference', effectively the situation where the visual system assists the injured vestibular apparatus, and so allow targeted vestibular physiotherapy. Other tests within the protocols may suggest the presence of peripheral neuropathies, musculoskeletal disorders, and malingering (Figure 7.6).

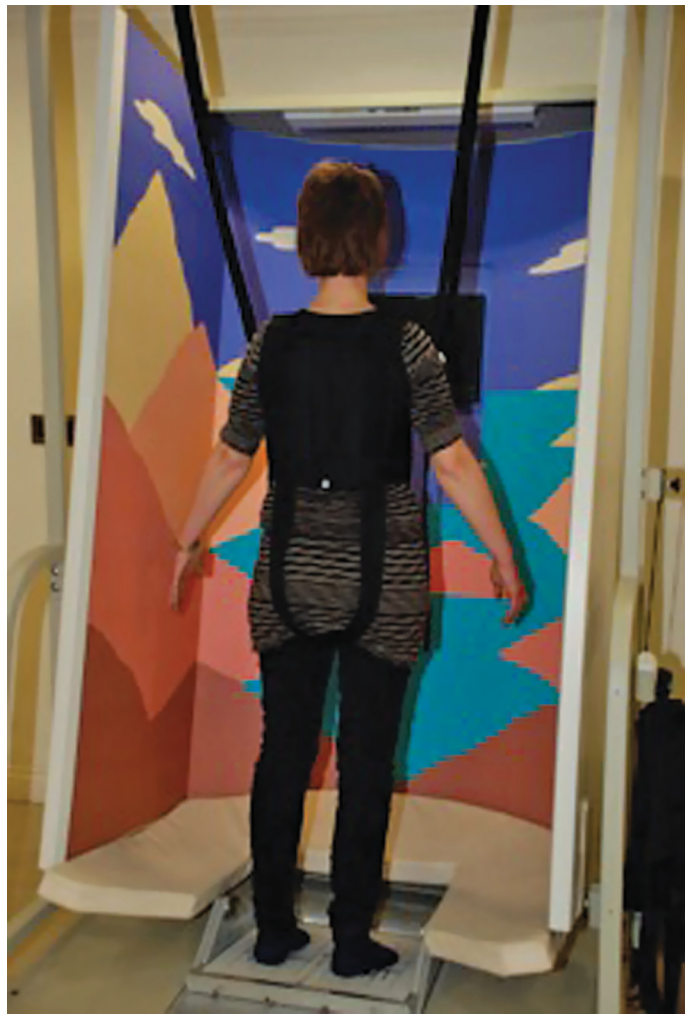

Figure 7.6 Posturography.
Reproduced from Rea P, Sandhu J. Tests for balance. From Musheer Hussain S, ed. *Logan Turner's Diseases of the Nose, Throat and Ear, Head and Neck Surgery*. 11th ed. CRC Press, 2015.

Vestibular evoked myogenic potentials

A VEMP is a myogenic response that is generated by the otolith organs as a reaction to a number of stimuli including air- and bone-conducted sound. When recorded from the tonically contracted ipsilateral sternocleidomastoid muscle they are known as the cervical VEMP (cVEMP) and when the recording electrodes are placed over the contralateral inferior oblique muscle they are referred to as the ocular VEMP (oVEMP). Both the cVEMP and oVEMP are now an established part of the vestibular test battery and allow independent assessment of the otolith organs. The cVEMP is generated by the saccule and travels via the inferior vestibular nerve. There is a large intersubject variation of the absolute magnitudes, therefore side-to-side differences are more useful. The origin of the oVEMP remains controversial but is most likely the utricle and the signal travels via the superior division of the vestibular nerve. An alternative to using

the response amplitude is to determine the threshold of response onset. As well as demonstrating reduced function in the utricle or saccule, it has become especially useful in assessing for SSCD syndrome where the third window leads to higher amplitudes and lower thresholds of VEMPs.

Electrocochleography

ECochG measures the auditory evoked response from the cochlea and provides a direct assessment of inner ear function. The auditory evoked response is electrical activity generated by the auditory system in response to an acoustic stimulus such as a click or a tone burst. The ECochG provides a method of recording this electrical activity from the cochlea and VIIIth cranial nerve by placing an electrode either extratympanically in the outer ear canal so that it is close to the tympanic membrane or alternatively it can be inserted transtympanically and rested on the cochlear promontory. The transtympanic method provides a better signal quality but is more invasive compared to the extratympanic approach.

The ECochG is a short latency response and is generated within a few milliseconds of the acoustic stimulus. It comprises three components: the cochlear microphonic, the summating potential (SP), and the compound action potential (AP). The cochlear microphonic reflects the displacement of the basilar membrane and hair cells in response to the acoustic stimulus. The SP is a current response from the inner hair cells while the AP is representative of usually more distal auditory fibres and is usually larger than the SP. ECochG results are normally reported as an SP/AP ratio.

While there are a number of specialist uses of the ECochG to monitor auditory function, it is most commonly requested for the assessment of Ménière's disease. While the exact mechanism remains elusive, it is thought that the increased pressure of endolymphatic hydrops impacts the biomechanics of the basilar membrane and hair cell structures resulting in changes in the ECochG waveform. An SP/AP ratio in excess of 0.5 is usually considered to be abnormal with the sensitivity to Ménière's disease being reported as 60–70%.

In order to generate an ECochG there must be hearing present and from a practical perspective the response is difficult to elicit beyond a sensorineural hearing loss of 40 dBHL. It is used less often than in the past (Figure 7.7).

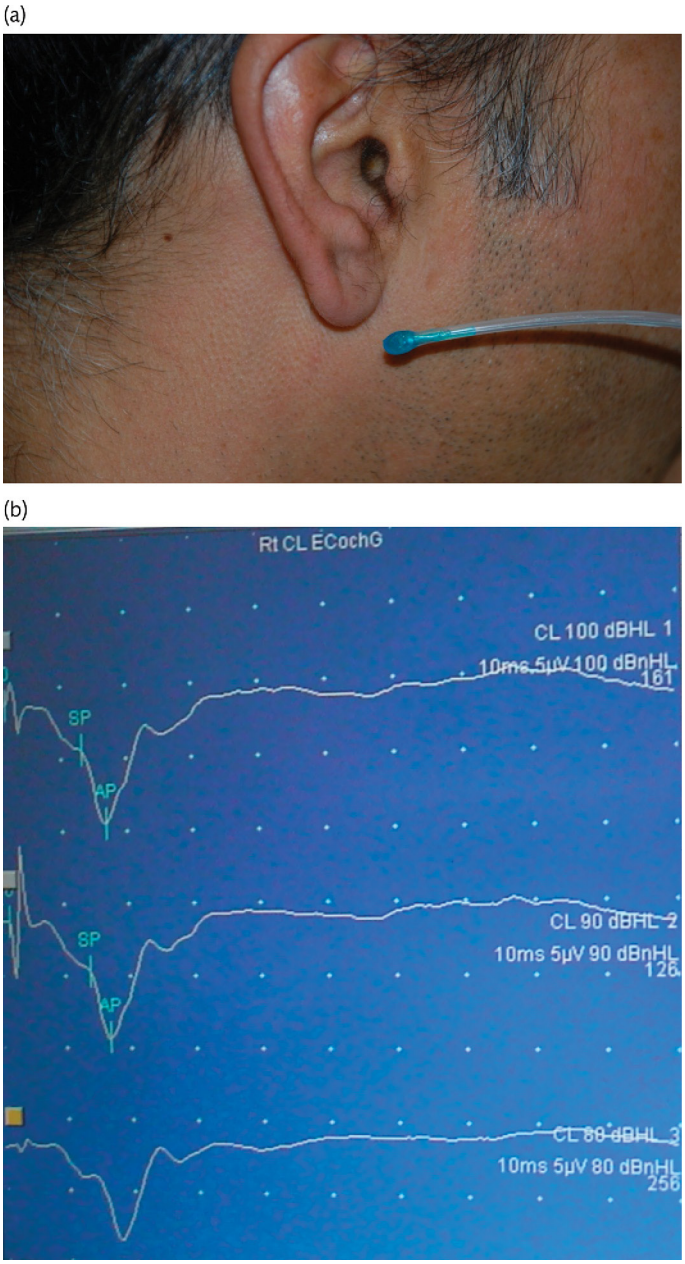

Figure 7.7 (a) ECochG probe. (b) ECochG trace.

The acute vertigo attack

In cases where patients present emergently with an abrupt onset of vertigo, the initial consideration is whether the cause is a peripheral or central vestibular event. While the majority of acute vertigo cases are caused by peripheral disorders (e.g. vestibular neuritis, Ménière's disease, BPPV, or labyrinthitis), central vascular and structural causes are diagnosed in approximately 10% of cases and should be promptly identified and treated. In general, patients with peripheral disease are able to stand unassisted for a short period of time, have unidirectional nystagmus according to Ewald's laws, no skew deviation, a positive head impulse sign, and no central oculomotor deficits. In contrast, patients suffering a brainstem/cerebellar infarct (e.g. posterior inferior cerebellar artery or AICA stroke) cannot stand unassisted and display some combination of atypical nystagmus, skew deviation, pursuit or saccadic abnormalities, and absence of a head impulse sign. Of particular interest is the patient with acute vertigo and sudden hearing loss who may represent vascular insufficiency of the labyrinth alone (peripheral) or brainstem and labyrinth (AICA infarct). The 'HINTS' acronym (Head Impulse, Nystagmus, Test of Skew) and 'HINTS Plus' (associated sudden hearing loss) has been described as a sensitive and specific method to differentiate peripheral (HINTS negative) from central (HINTS positive) causes of acute vertigo.

In managing acute vertigo, remember vertigo is a symptom not a diagnosis. Establishing the diagnosis, or at least the body system involved, is key to successful management. Patients who have become dehydrated or may have a central or medical cause may need rehydration and hospital admission as an emergency. Certain peripheral vestibular causes may respond to treatment (e.g. an Epley manoeuvre for BPPV). Antiemetics or vestibular sedatives such as cyclizine, prochlorperazine, or a benzodiazepine may be provided.

Differential diagnosis of the dizzy patient

Ménière's disease (Chapter 14), vestibular migraine (Chapter 14), and BPPV (Chapter 15) are described in detail elsewhere in this textbook.

Medical causes of dizziness

Psychological and neuropsychological

Anxiety and depression may cause dizziness and likewise complicate a vestibular disorder. Anxiety-related dizziness is characterized by situational provocation, intense autonomic activation, catastrophic thinking, and avoidance behaviour. More than 50% of patients with vestibular migraine have comorbid psychiatric disorders.

Persistent postural-perceptual dizziness is a relatively new diagnostic formulation encompassing the older terms of visual vertigo, chronic subjective vertigo, and phobic positional dizziness. It tends to occur in those with a history of vestibular insult and is often with a history of migraine and anxiety. Patients may report non-spinning vertigo, unsteadiness, light-headedness, dissociation, visual hypersensitivity, and are worse when in motion or surrounded by motion. Symptoms need to be present most days for more than 3 months. Testing may be normal. It appears to be a central processing disorder, with amplification of visual stimuli, but much work is ongoing. Treatment may include vestibular physiotherapy, cognitive behavioural therapy, and selective serotonin reuptake inhibitor antidepressants.

Autoimmune

Autoimmune inner ear disease

Autoimmune inner ear disease represents the archetypal systemic disease-causing audiovestibular dysfunction. Although classically associated with rapidly progressive bilateral sensorineural hearing loss, and often associated with tinnitus, about 50% of patients have vestibular symptoms. The dizziness may mimic Ménière's disease with episodic vertigo, tinnitus, and hearing loss. It responds very well to oral and intra-tympanic steroids.

A number of classical autoimmune conditions are associated with autoimmune inner ear disease, including granulomatosis with polyangiitis (formerly known as Wegener's granulomatosis), polyarteritis nodosa, and systemic lupus erythematosus. Systemic lupus erythematosus is particularly interesting as its diagnosis is sometimes delayed (and hence may be seen in the balance clinic) and because of the high reported incidence of associated vertigo—9% in one recent large series.

Cogan syndrome describes a rare systemic disorder presenting with sensorineural hearing loss, vertigo, and non-syphilitic interstitial keratitis. It typically affects patients in their twenties or thirties.

Susac syndrome is another rare systemic disorder that has audiovestibular features and needs rapid diagnosis. It is believed to result from an immune-mediated endotheliopathy. The classical triad is encephalopathy, branch retinal vein occlusion, and sensorineural hearing loss. What is relevant to those seeing balance patients is that the hearing loss is usually low frequency (as with Ménière's disease).

Cardiac

Presyncopal causes of dizziness are related to effects on the cardiovascular system, and are described as the sensation of light-headedness. The main causes of presyncope and syncope can be divided into three categories: neurally mediated, orthostatic, and cardiac.

Neurally mediated syndromes are associated with vasodilation of both arterial and venous beds with a relative bradycardia secondary to an inappropriate reflex arc. Conditions include vasovagal attacks and sinus node dysfunction, and also include those mediated by increased intrathoracic pressure, such as coughing, sneezing, or straining, or arising from oesophageal, nasopharyngeal, or gastrointestinal stimulation (such as swallow syncope or defaecation syncope).

Orthostatic hypotension is defined as a decrease of at least 20 mmHg in systolic and at least 10 mmHg in diastolic pressure within 10 minutes of active standing, and results from a failure of the baroreceptors to promote an adequate vasomotor response upon standing. This leads to venous pooling and subsequent cerebral hypoperfusion, giving rise to symptoms of dizziness and palpitations, and is often concomitant in patients with heart failure. The cause may be idiopathic, or may be attributed to primary or secondary autonomic control disorders. Primary causes include pure autonomic failure and multiple system atrophy, and secondary causes include diabetic and alcoholic neuropathies, Addison's disease, paraneoplastic syndrome, and prolonged periods of physical inactivity.

In contrast to the generalized autonomic conditions described above which affect both sympathetic and parasympathetic activity, postural orthostatic tachycardia syndrome affects sympathetic function only. It is defined as an increase in the heart rate of 30 beats per minute or more within 10 minutes of active standing in the absence of orthostatic hypotension, and the standing heart rate is often found to be in excess of 120 beats per minute. Patients experience a number of symptoms related to orthostatic intolerance, including dizziness, palpitations, and headache, which are typically relieved by lying down.

Cardiac syncopal syndromes arise from a fall in effective cardiac output, either from an electrical or a mechanical cause. Electrical causes comprise arrhythmias, including ventricular tachycardias, supraventricular tachycardias, and bradycardias, and mechanical obstruction can arise from conditions such as aortic stenosis, hypertrophic obstructive cardiomyopathy, or massive pulmonary emboli.

Metabolic

A number of metabolic conditions should be borne in mind during assessment of the dizzy patient, including hyper- and hypoglycaemia, thyroid disorders, and adrenal insufficiency.

Pregnancy

Pregnancy may cause symptoms of dizziness from as early as 8 weeks into the first trimester. This is a common complaint, affecting up to 75% of pregnant women.

Central neurological

Epidemiological studies suggest that around one-quarter of patients presenting with dizziness have an underlying central pathology. This includes cerebrovascular disorders, vestibular migraine (see Chapter 14), demyelinating conditions, neurodegenerative disorders, and rarely tumours of the posterior fossa. Idiopathic intracranial hypertension is common in young obese woman and is associated with pulsatile tinnitus.

Peripheral neurological

Peripheral neuropathies are commonly found in patients with multisensory dizziness, especially the elderly. Testing for them is a

While diabetes is the commonest cause seen in the balance clinic, other disorders to consider include chemotherapeutic agents, infections including Lyme disease and HIV, inherited disorders such as Charcot–Marie–Tooth disease, trauma, and vitamin deficiency. Recommended routine tests include immunoglobulins, autoantibody screen, serum protein electrophoresis, B complex vitamins, folate, homocysteine, urea and electrolytes, glycated haemoglobin, C-reactive protein, thyroid function tests, and liver function tests.

Genetic

Enquire about a family history of dizziness. Familial conditions range from the very common (e.g. migraine), uncommon (e.g. Ménière's disease, and essential tremor), through to the rare (e.g. episodic ataxia type 2).

Medication

Dizziness as an adverse reaction to medication is a commonly reported finding, with over 1000 medications listing vertigo as a side effect and 2000 listing dizziness. In addition, once polypharmacy and drug interactions have been considered, the potential effects are almost infinite. With an ageing population, and the prevalence of multiple comorbidities, polypharmacy is on the rise, leading to further diagnostic difficulties. The most commonly encountered medications causing dizziness are antidepressants, anticonvulsants, antihypertensives, and sedatives.

Orthopaedic

While cervicogenic dizziness is an entity still under debate in the literature, there is a growing body of evidence to support the importance of cervical proprioceptive input in balance and postural control. Major joint replacements are common contributors to imbalance on the elderly.

Infections

Infection should also always be considered in the differential diagnosis of dizziness. Infections of the central nervous system can broadly be divided into those arising from bacterial (neurosyphilis, meningitis, Lyme disease), viral (HIV, meningitis), fungal (meningitis), and protozoal (malaria, toxoplasmosis) sources. For example, both Lyme disease and neurosyphilis may demonstrate endolymphatic hydrops and thus mimic a clinical picture of Ménière's disease, and in addition, patients with neurosyphilis may also exhibit significant loss of type 2 vestibular hair cells.

Respiratory

Both respiratory failure and hyperventilation can cause dizziness, the latter commonly being seen in anxiety states. Obstructive sleep disorder is associated with fatigue and vestibular responses and should certainly be sought.

Vitamin deficiencies

Peripheral neuropathies may be associated with deficiencies of vitamins B_1, B_3 (niacin), B_6, B_{12}, and E. Routine testing for B complex vitamins and vitamins D and E in peripheral neuropathy is recommended. There is accumulating evidence for a link between low bone mineral density, vitamin D deficiency, and BPPV, although the relationships appear complex.

Paraneoplastic syndrome

As symptoms may develop long before the cancer is diagnosed, paraneoplastic syndromes may first present to the balance clinic. It is most commonly associated with small cell lung tumours, lymphoma, and cancers of the ovary, breast, and uterus. Typical presentations may include rapidly progressive imbalance and downbeat nystagmus sometimes termed a vestibulocerebellar syndrome.

Poisoning

Carbon monoxide poisoning may be acute or chronic, and may present not only with headache and dizziness, but also hearing loss, tinnitus, and peripheral neuropathy.

Toxin-induced cerebellar degeneration causing ataxia is most commonly associated with chronic alcohol abuse but heavy metals (lead and arsenic), drugs (such as phenytoin and amiodarone), and chemotherapeutic agents (including 5-fluorouracil) need to be screened for.

Peripheral vestibular causes of dizziness beyond Ménière's disease, BPPV, and migraine

When considering diagnoses, it is helpful to classify presentations into chronic dizziness and imbalance versus acute episodic vertigo (see also Chapters 14 and 15).

Chronic imbalance

Acute vestibular loss resulting from acute vestibular neuritis, labyrinthitis, or head injury, for example, and chronic loss resulting from ageing, leads to a fixed vestibular deficit, with asymmetric vestibular signally to the vestibular nuclei. This weakness is termed vestibulopathy, or vestibular hypofunction. If central adaptation does not occur this leads to a conflict between vestibular stimuli and other inputs to the balance system such as visual and proprioceptive. This conflict is central to the experience patients report with vestibulopathy. A similar conflict arises when we feel sea sick on a boat as visual and vestibular inputs conflict as we look at the floor of the boat (stable visual inputs) while it rocks (active vestibular inputs).

Such asymmetry is treated with vestibular physiotherapy, while vestibular suppressants (e.g. prochlorperazine) are discouraged.

Vestibulopathy may combine with any of the other disorders discussed previously, and in the elderly may be part of a multisensory or multisystem balance disorder.

Avoiding vestibular stimulation (which triggers dizziness) may then lead to deconditioning and a downward spiral of restricted mobility and mood.

Recurrent acute vertigo

BPPV, Ménière's disease, and vestibular migraine account for the great majority of patients with recurrent acute vertigo. There are many other less common peripheral vestibular disorders which lead to episodic dizziness.

Superior semicircular canal dehiscence syndrome

SSCD results from a 'third window' in the labyrinth caused by thinning and absence of the bone between the superior SCC and the middle cranial fossa. Patients present with pressure-induced vertigo (lifting, straining, sneezing), autophony, Tullio's phenomenon, aural fullness, and fluctuating hearing loss. They may describe hearing their own eyes move or their heartbeat. In clinic they should have a positive malleolar test, they may have artificially low bone conduction thresholds that look like a conductive hearing loss, and nystagmus may be seen on a Valsalva manoeuvre. The key tests are VEMPs and CT scanning. Treatment may be conserva-

middle cranial fossa approach for those severely affected is usually successful.

Perilymph fistula

Perilymph fistula, like SSCD, can mimic many other balance disorders and the symptoms are varied. A communication between the middle and inner ears through the round or oval windows is the pathology. Symptoms may include fluctuating or fixed hearing loss, tinnitus, unsteadiness on movement, and pressure-induced dizziness. Most important is an appropriate history such as stapes surgery, a head injury, or barotrauma (e.g. ear pain on scuba diving or air travel). There is no definitive test but nystagmus on VNG testing while changing ear canal pressure with tympanometry (the fistula test) is strongly supportive. Surgical plugging of the round or oval window can be curative but proceed with caution as there is inevitable diagnostic uncertainty.

Vestibular paroxysmia

Vestibular paroxysmia may present with multiple short spinning or non-spinning dizzy spells per day lasting less than a minute. Tinnitus may also occur. It is a vascular compression syndrome usually arising from compression of the VIII nerve by a loop of AICA. It may be triggered by hyperventilation or in certain head positions. The vascular loop can be seen on magnetic resonance angiography imaging while caloric testing a deficit is often seen. Carbamazepine can be an effective treatment.

Episodic ataxia type 2

Episodic ataxia type 2 results from defects of a calcium channel gene (*CACNA1A*) located on chromosome 19. It has been linked to four conditions, all of which may be seen in the balance clinic: episodic ataxia type 2, familial hemiplegic migraine, spinocerebellar ataxia, and sporadic hemiplegic migraine. Episodic ataxia type 2 is on the differential diagnostic list of patients with unexplained episodic vertigo. Other conditions include vestibular migraine, Ménière's disease, and vestibular paroxysmia among others. Vertigo attacks may last minutes to days, and may be associated with neurological symptoms, tinnitus, and headache. About 50% have migrainous headaches, and triggers do overlap with migraine (stress, exertion, caffeine). Many will have a family history. The diagnosis may be confirmed with expensive testing by requesting *CACNA1A* genetics. The key to diagnosis clinically is to seek cerebellar signs, which are likely to develop between attacks as the disease progresses. Treatment is available with acetazolamide.

Tips and pearls

- The history is key.
- Differentiate between chronic dizziness, and recurrent acute vertigo.
- For a single acute attack, differentiate between central versus peripheral attacks using the HINTS test.
- For recurrent episodes, identify the nature of the attack including time line, triggers, and associated symptoms.
- Recognize symptoms may evolve over time.
- Recognize several pathologies may occur at the same time (e.g. in Ménière's disease, 40% have migraine, 20% BPPV, and all are likely to have an emotional reaction and vestibulopathy).
- Do not forget to consider non-vestibular causes.

Conclusion

Although the dizzy patient can seem challenging, by understanding the patterns of presentation, and recognizing the range of possible diagnoses, most patients can be successfully diagnosed and treated. It is a very rewarding subspecialty which continues to evolve at pace.

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9

Aural fullness

Holger H. Sudhoff

Introduction

Aural fullness is a ubiquitous symptom among patients, frequently related to Eustachian tube dysfunction (ETD). However, the underlying pathology may involve any part of the peripheral hearing organ, from the external auditory canal to the inner ear. Aural fullness is often reported in hydropic ear diseases and idiopathic sudden sensorineural hearing loss.[1] Additionally, aural fullness can be related to temporomandibular joint disorders.[2]

The Eustachian tube (ET) is part of a system including the nose, palate, rhinopharynx, and middle ear spaces.[1] The middle ear comprises the tympanic cavity, which includes the bony ET and the mastoid cells system. The tympanic cavity and mastoid cells are interconnected and allow for gas exchanges and pressure. The ET is a complex organ consisting of a dynamic canal with its mucosa, cartilage, surrounding soft tissue, peritubal muscles, superior bony support, and the sphenoid sulcus. Clinical experience as well as numerous patient studies and animal models show that the ET plays an important role in middle ear pathology.[3–5] While knowledge of the mechanical properties of the ET has improved significantly in the last years, uncertainties still remain due to its complex anatomy, multiple functions, and the impact of intrinsic and external factors.[6,7] Intermittent transitory tubal dilation is probably the major mechanism for equilibration of middle ear cleft pressure with the ambient atmosphere.[8] Barometric and chemical receptors within the middle ear cleft are assumed to provide autonomic nervous system feedback that impact the frequency of involuntary tubal opening.[9,10] It is important to distinguish ETD from other causes of aural fullness such as patulous ET, temporomandibular joint disorders, superior semicircular canal dehiscence syndrome, Ménière's disease, and increased intracranial pressure.

Eustachian tube dysfunction

ETD can lead to clinical symptoms such as aural fullness, impaired pressure equilibration, altered middle ear aeration, hearing loss, and autophony. Because the most common cause of obstructive dysfunction is mucosal inflammation within the cartilaginous ET, patients should be questioned about inflammatory processes such as allergic rhinitis, chronic rhinosinusitis, laryngopharyngeal reflux, and smoke exposure.[11,12] Paediatric ETD may be caused by adenoidal hypertrophy and mucosal swelling due to acute or chronic upper respiratory tract infections. Cleft palate, granulomatous disease, cystic fibrosis, Samter's triad, and Kartagener's syndrome are predisposing factors. Dilatory and baro-challenged ETD may be a contributing factor to vertigo.[13,14] ETD is estimated to be present in about 1% of the general population.[15] Almost 40% of all children up to the age of 10 develop temporary ETD.[16,17] Studies assessing paediatric and adult patient cohorts demonstrate that ETD is detectable in up to 70% of patients undergoing middle ear surgery.[18] ET function is believed to be important for the successful outcome of middle ear surgery.[19,20] Patients with a patulous ET usually present with positional autophony and hearing both voice and breathing sounds and aural fullness.[21] The precise distinction between obstructive and patulous types of ETD is pivotal for appropriate medical or surgical treatment.

Patulous Eustachian tube

A patulous ET (tuba aperta) may lead to an enormous reduction in quality of life. A patulous ET can cause symptoms such as autophony, breath-synchronous tinnitus, pressure sensation in the ear, and hearing loss. In combination with so-called sniffing, it can trigger the development of cholesteatoma. Due to the diffuse symptoms the correct diagnosis of this disease can be challenging. A patulous ET can be best diagnosed through a well-structured examination including patient history, physical examination with thorough observation of movements of the tympanic membrane, and tympanometry with reflex decay. Patients with patulous ETs may benefit from augmentation or reconstructive procedures.[22,23]

Eustachian tube dysfunction assessment

A huge number of methods have been employed to assess ET function. More than 40 methods can be found in the literature.[24] None has proved to give detailed insights into all aspects of ET physiology and pathology. Clinical tests such as otoscopy, endoscopy, Politzer test, and Valsalva and Toynbee manoeuvres may

Table 9.1 The Eustachian tube score (ranging from 0 to 10 points)—facilitates quantification and inter-individual comparison of Eustachian tube function

Symptom/finding	2 points	1 point	0 points
Clicking sound when swallowing	Always	Sometimes	Never
Positive Valsalva manoeuvre	Always	Sometimes	Never
TMM 30 mbar	R <1	R ≥1	No R
TMM 40 mbar	R <1	R ≥1	No R
TMM 50 mbar	R <1	R ≥1	No R

R, tubal opening latency ratio (time between the pressure transfer from the nasopharynx to the middle ear); TMM, tubomanometry.

give initial guidance. Manometric testing includes tympanometry, reflex decay tympanometry, nine-step inflation–deflation test, modified inflation–deflation test, forced response test, tubomanometry, and the seven-item ETD questionnaire (ETDQ-7)[25] (Tables 9.1 and 9.2).

Balloon dilation Eustachian tuboplasty

Balloon dilation Eustachian tuboplasty is a minimally interventional method to treat chronic obstructive ETD. Preoperative tubomanometry is initially used to assess the level of ETD. Balloon dilatation of the ET is usually performed with the patient under general anaesthesia.[26] A balloon catheter is introduced into the ET via the nose, under transnasal endoscopic vision. Once the balloon is correctly positioned in the cartilaginous portion of the ET, it is filled with saline up to a pressure of about 10 bars. Pressure is maintained for approximately 2 minutes. The balloon is then emptied and removed. The catheter insertion device offers three differently angled extensions matching the individual anatomical situation: 30°, 45°, and 70°. Balloon dilation Eustachian tuboplasty is easy to perform and it is usually a bloodless and straightforward procedure. We recommend performing the procedure under general anaesthesia with endoscopic visualization of the nasopharynx.[27]

Table 9.2 The ETDQ-7—a tool for the evaluation of chronic obstructive Eustachian tube dysfunction

Over the past 1 month, how much has each of the following been a problem for you?	No problem		Moderate problem			Severe problem	
1. Pressure in the ears?	1	2	3	4	5	6	7
2. Pain in the ears?	1	2	3	4	5	6	7
3. A feeling that your ears are clogged or 'under water'?	1	2	3	4	5	6	7
4. Ear symptoms when you have a cold or sinusitis?	1	2	3	4	5	6	7
5. Crackling or popping sounds in the ears?	1	2	3	4	5	6	7
6. Ringing in the ears?	1	2	3	4	5	6	7
7. A feeling that your hearing is muffled?	1	2	3	4	5	6	7

Key evidence

- Sound transmission requires a stable air compartment within the middle ear spaces maintained by a sufficient ET function. If affected, aural fullness may be one of the clinical symptoms.
- Aural fullness requires a thorough clinical examination investigating patulous ET, temporomandibular joint disorders, superior semicircular canal dehiscence syndrome, Ménière's disease, and increased intracranial pressure.
- Balloon dilation Eustachian tuboplasty is a potential treatment option in dilatory and baro-challenged ETD to treat aural fullness.

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10

The facial nerve

Richard M. Irving

Introduction

The facial nerve is the commonest cranial nerve to suffer from a pathological deficit with resultant cosmetic, functional, and psychological repercussions. The nerve's vulnerability to pathology such as trauma, inflammation, and neoplasia in part stems from its long and complex course. It commences intracranially, then proceeds in a non-expandable bony canal, finally ending up in the parotid gland and superficial tissues of the face. There are many causes of facial palsy most of which, especially idiopathic, have an excellent prognosis. It is, however, important to recognize atypical cases that require further investigation and to understand the principles of management including the options for rehabilitation to provide the patient with facial palsy with the best possible outcome.

Applied anatomy

The facial nerve is a mixed nerve although it is the loss of the motor component that results in the greater disability. It has approximately 10,000 fibres of which two-thirds are motor and one-third sensory. The nerve comprises upper and lower motor neurons with the lower having intracranial, intratemporal, and extracranial segments.

Upper motor neurons originate from the precentral gyrus, project to the internal capsule, then lower brainstem, and synapse in the pontine facial nerve nucleus. These corticobulbar fibres mostly cross to the contralateral side with selective bilateral contribution to the upper facial muscles. This bilateral cortical representation results in forehead sparing in supranuclear lesions. In contrast, lower motor neuron lesions will result in ipsilateral paralysis of the both upper and lower face.

The lower motor neurons have their cell bodies in the pontine facial motor nucleus. Fibres then loop around the abducent nerve nucleus before exiting the ventral surface of the pons anteroinferior to the VIIIth nerve and nervus intermedius. Lesions at the level of the pons can result in a combined VIth and VIIth nerve weakness. The nerve then crosses the cerebellopontine angle to enter the fallopian canal via the internal auditory meatus.

The intratemporal course of the nerve measures approximately 3 cm and can be divided into the labyrinthine, tympanic, and mastoid segments. The labyrinthine segment courses anterolaterally from the internal auditory meatus, posterior and superior to the cochlea and anterolateral to the ampulla of the horizontal and superior semicircular canals, to the geniculate ganglion. This is the narrowest segment of the facial nerve canal, 0.6 mm at the meatal foramen, and is more susceptible to compression lacking epineurium and anastomosing arterial cascades.

The tympanic or horizontal segment is dehiscent in up to 50% of cases and passes from the geniculate ganglion to the second genu. This segment lies medial to the cochleariform process and tensor tympani muscle, passes above the oval window and stapes, and below the lateral semicircular canal.

From the second genu the nerve continues downwards as the mastoid or vertical segment where it lies anterior and lateral to the ampullated end of the posterior semicircular canal and medial to the tympanic annulus. This segment of the nerve gives rise to the nerve to stapedius, chorda tympani, and the sensory auricular branch.

The facial nerve exits the stylomastoid foramen and enters the parotid gland. Within the gland the nerve divides into the temporofacial and cervicofacial branches going on to form five major branches: temporal, zygomatic, buccal, marginal mandibular, and cervical.

The sensory component of the nerve originates from the superior salivatory nucleus (secretomotor) and has fibres that pass to the nucleus of the tractus solitarius (somatic sensory and special sensory) in the brainstem. The fibres travel with the nervus intermedius to the geniculate ganglion which contains the cell bodies of the somatic sensory component of the nerve and gives rise to a branch, the greater superficial petrosal nerve. The greater superficial petrosal nerve contains preganglionic parasympathetic fibres from the nervus intermedius that then join the deep petrosal, synapse in the pterygopalatine ganglion, and innervate the lacrimal gland and mucus glands of the nasal and oral cavities. The chorda tympani caries parasympathetic preganglionic fibres to the sublingual and submandibular glands and the anterior two-thirds of the tongue.

Physiology and special investigations

Special investigations can be used to assist in determining the site of the lesion, its severity, underlying cause, prognosis, and to provide a guide to rehabilitation.

Table 10.1 House–Brackmann grading

Grade	Gross	Resting tone	Forehead	Eye closure	Mouth	% function
I	Normal function					100%
II	Slight weakness	Normal	Moderate- good function	Complete closure with minimal effort	Slight asymmetry	80%
III	Obvious weakness	Normal	Slight–moderate function	Complete closure with full effort	Slight weakness	60%
IV	Disfiguring	Normal	None	Incomplete closure	Asymmetric	40%
V	Barely perceptible motion	Asymmetric	None	Incomplete closure	Slight movement	20%
VI	Total paralysis					0%

Site of lesion is best determined by imaging using a combination of computed tomography (CT) and magnetic resonance imaging (MRI). Topodiagnostic physiological tests include taste, salivary flow tests, Shirmer test, and stapedial reflex. These rarely provide useful additional information and show limited clinical correlation.

Electrophysiological tests aim to determine the severity of the lesion, potential for recovery, and can guide ongoing management, in particular the value of surgical exploration and facial reanimation. Currently, the two most common objective tests used are electroneuronography (ENoG) and electromyography (EMG).

ENoG relies on a functional contralateral nerve and is mainly of value in acute-onset complete facial nerve paralysis. The nerve is stimulated at the stylomastoid foramen and the muscle response detected with a surface or needle electrode at the nasolabial fold. A poor prognosis correlates with greater than 95% reduction in amplitude on the affected side within 3–21 days. It takes a minimum of 3 days for Wallerian degeneration to occur to the level of the stylomastoid foramen and after 3 weeks, due to concurrent degeneration and regeneration, ENoG is of less value.

EMG studies are most useful in determining prognosis and guiding rehabilitation. EMG measures voluntary motor unit potentials, using needle electrodes placed in the facial muscles. Fibrillation potentials suggest nerve degeneration and confirm the presence of viable muscle fibres. They appear 2–3 weeks following injury. Polyphasic potentials indicate early signs of reinnervation, which can precede clinical signs of recovery. Electrical silence is a poor prognostic indicator and implies the facial musculature is non-viable.

Several facial grading scales exist in an attempt to objectively quantify facial motor function. These are used in measuring recovery, potential deterioration, and comparison of facial nerve outcomes across therapeutic modalities. The House–Brackmann score (Table 10.1) is widely used clinically to measure the degree of facial motor weakness.

Pathology

By far the most common cause of facial paralysis is idiopathic or Bell's palsy, which accounts for over 70% of cases. An alternate cause must be considered if the weakness is of slow onset, recurs on the same side, and is associated with twitch, spasm, or additional neurological deficit. Other atypical features are hearing loss, skin lesions, or failure to recover within a month. The other common causes are trauma, infection, and neoplasia (Table 10.2), and of these the important cause to exclude is neoplasia which is best achieved by a combination of CT and MRI scanning.

Cases of bilateral facial palsy should alert the clinician to underlying systemic or neurological pathology, such as Lyme disease or Guillain–Barré syndrome, and in recurrent cases Melkersson–Rosenthal syndrome.

Bell's palsy

Bell's or idiopathic facial palsy is a diagnosis of exclusion and has an incidence of 30 cases per 100,000 per year. It can occur in any age group but has a peak incidence at 40 years of age. There is an 7% risk of recurrence. At onset 70% are complete and 30% partial.

Presentation is with sudden onset (24–48 hours) of unilateral weakness often accompanied by taste disturbance and hyperacusis. A viral prodrome is rare but preceding postauricular pain is quite typical. Examination focuses on excluding any alternate underlying cause and includes assessment of facial nerve function, cranial nerves, otoscopy, and palpation of the parotid.

Table 10.2 The more common causes of facial palsy

Idiopathic	Bell's palsy
Infection	Otitis media Cholesteatoma Skull base osteomyelitis Lyme disease Varicella zoster virus
Trauma	Temporal bone fracture Iatrogenic
Tumour	Schwannoma Meningioma Haemangioma Paraganglioma Temporal bone malignancy Parotid malignancy
Neurological	Lacunar/brainstem infarct Guillain–Barré syndrome
Congenital	Mobius syndrome CHARGE syndrome
Other	Sarcoidosis

There is a good evidence base to support the use of steroids especially if taken within the first 24 hours, and a typical treatment for an adult would be 60 mg a day for 5 days. While there is evidence to support a viral aetiology there is as yet no strong evidence supporting the use of antivirals. If eye closure is incomplete, regular topical lubricants throughout the day with thicker viscosity lubricant at night should be prescribed with taping of the eye shut at night. Referral to ophthalmology and for facial therapy should be considered in those with delayed recovery.

The role of surgical decompression in the management of acute idiopathic facial paralysis remains controversial; however, surgical decompression has shown promising results in recurrent idiopathic facial palsy.

All patients get some recovery with 85% showing significant recovery within 1 month of onset. Any ultimate outcome worse than House–Brackmann grade III should raise the suspicion of an underlying cause. Poor prognostic factors include complete facial palsy at onset, age over 60 years, severe pain, no recovery by 1 month, and associated conditions (e.g. hypertension and diabetes).

Ramsey Hunt syndrome

Ramsey Hunt syndrome is caused by the varicella zoster virus and is characterized by vesicles in the external ear, soft palate, or tongue. It may be misdiagnosed as Bell's palsy as the classic vesicles may not appear, be quite subtle, or may be delayed. Ramsey Hunt syndrome is frequently a polyneuritis with sensorineural hearing loss, vertigo, and other cranial nerve involvement common. This is primarily a clinical diagnosis but can be confirmed by polymerase chain reaction. If suspected, prompt treatment with steroids and antivirals is recommended. The prognosis is very poor with only 20% achieving complete recovery of facial motor function and all should be considered for facial therapy and ophthalmology review.

Acute and chronic infection

Facial paralysis secondary to acute otitis media is typically seen in young children. The prognosis is excellent and most resolve with conservative management in the form of systemic antibiotics. A myringotomy with or without ventilation tube should be considered when spontaneous perforation of the tympanic membrane is not present. A mastoidectomy may be indicated for suppurative complications, or lack of clinical improvement.

Middle ear cholesteatoma is associated with facial paralysis in under 1% of cases; however, when the disease is in the petrous apex this increases to up to 50%. Prompt surgical intervention is warranted in the form of either a mastoidectomy or petrosectomy to ensure the best outcome; however, recovery can occur even if treatment is delayed for several months in partial facial paralysis secondary to cholesteatoma.

Skull base osteomyelitis is a condition that occurs in elderly immunocompromised patients and is characterized by severe pain, aural discharge, and progressive cranial neuropathies including facial paralysis. Diagnosis is based on imaging comprising CT and MRI scanning and treatment is with long-term antibiotics.

Lyme disease

Lyme disease is a multisystemic illness caused by the spirochaete *Borrelia burgdorferi* and transmitted by the *Ixodes* tick. It can present as either unilateral or bilateral facial nerve paralysis. Erythema migrans is evident in the early stages of the disease which can be associated with joint pain, fever, and later symptoms of fatigue and neck stiffness. The diagnosis is confirmed by serology and treatment is with a course of oral antibiotics, either doxycycline or amoxicillin.

Trauma

Blunt, penetrating, and iatrogenic injuries can result in facial nerve paralysis.

Facial nerve injury is more common in transverse or otic capsule involving temporal bone fractures. Penetrating injury, especially gunshot related, results in more extensive injury with a higher incidence of facial nerve paralysis compared to closed head injury.

In middle ear surgery, the tympanic segment of the facial nerve is the most common site of iatrogenic injury, due to the high prevalence of facial canal dehiscence or exposure secondary to cholesteatoma. The nerve can also be damaged in the cerebellopontine angle during acoustic neuroma surgery or when operating on the parotid gland.

Many patients with temporal bone fractures have multiple injuries and management of any facial nerve injury may need to wait until the patient is stabilized. Imaging with high-resolution CT is essential to evaluate the temporal bone and the intracranial contents as soon as the patient's condition allows.

Delayed onset or incomplete paresis almost always fully recovers, the majority within the first 3 months. Treatment involves high-dose corticosteroids and eye care.

The decision to explore a facial nerve following trauma is complex. Factors favouring exploration and indicating a severe injury to the nerve are a penetrating injury, immediate onset of paralysis, CT evidence of nerve canal disruption or bony spicule, loss of inner ear function, persistent cerebrospinal fluid leak, and 90% or greater degeneration on ENoG. Iatrogenic injury should be repaired at the time of surgery if noticed or re-explored within days and the nerve repaired.

Benign and malignant tumours

Tumour involvement of the facial nerve can either be from intrinsic lesions or from spread from adjacent tissues. The most common intrinsic tumours are schwannomas (**Figure 10.1**) and haemangiomas. Secondary spread can occur from benign lesions such as paragangliomas or from perineural spread of malignant tumours, the most common being squamous cell carcinoma or tumours arising in the parotid gland.

Symptoms vary depending on the site of the lesion. Tumours involving the vertical or horizontal segment of the nerve may present with conductive hearing loss and a mass noted on otoscopy. Due to inner ear erosion, those at the labyrinthine segment may cause sensorineural hearing loss, tinnitus, and vertigo.

Suspicion is aroused by a history of facial spasm or twitching, a progressive weakness coming on slowly, typically over a number of weeks, and recurrent or slowly resolving weakness. Imaging is critical to the diagnosis as lesions may be very small at presentation and are easily missed. Both high-resolution CT scanning of the temporal bone and T1-weighted gadolinium-enhanced MRI scanning of the brain and skull base are recommended to provide a detailed view of the nerve.

Management of malignant tumours is very much along oncological principles. For benign tumours, however, the decision-making can be more complex and needs to be individualized. However, there

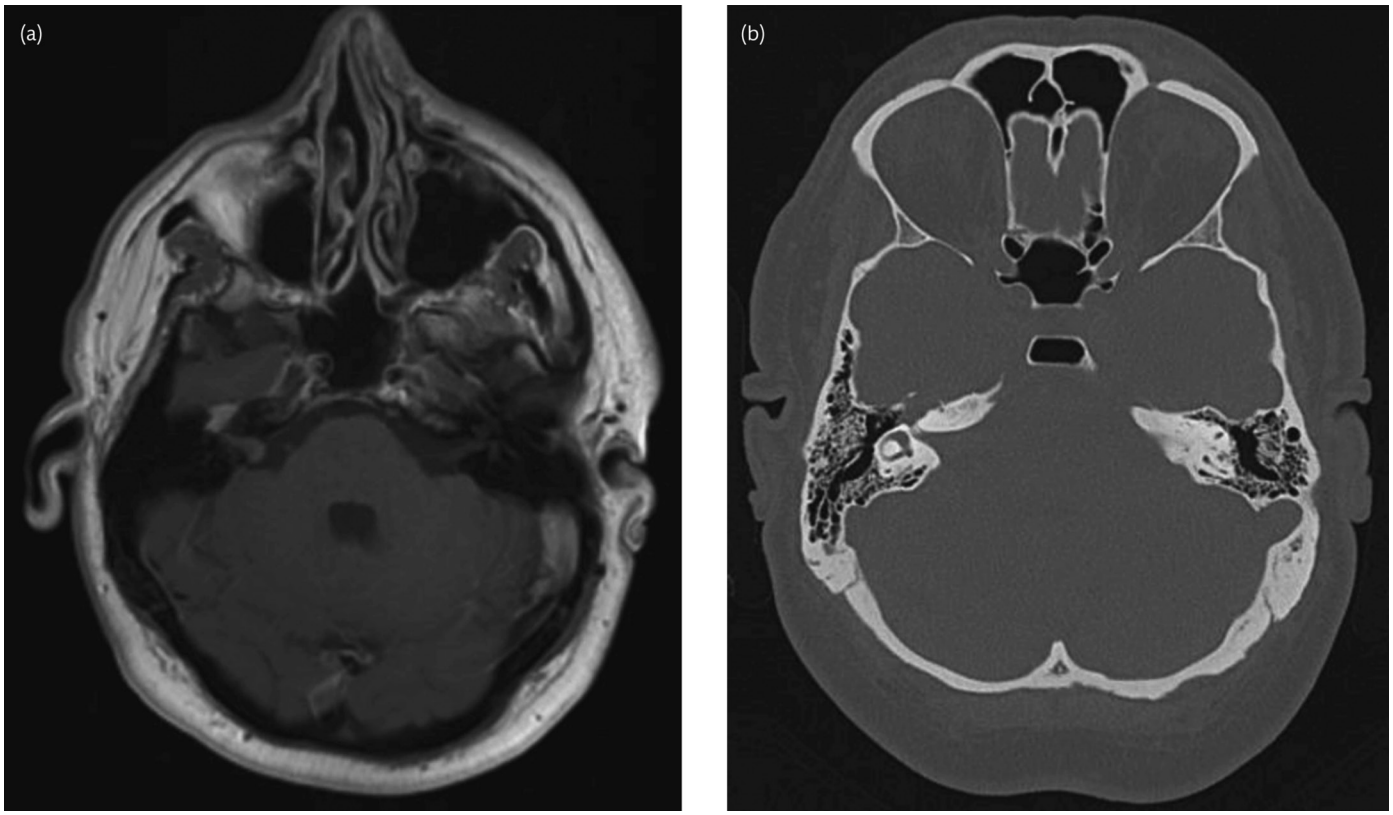

Figure 10.1 (a) T1-weighted gadolinium-enhanced MRI demonstrating a 'dumbbell' facial nerve schwannoma involving the internal auditory canal, labyrinthine segment, and geniculate ganglion. (b) High-resolution CT scan of the same tumour showing bony erosion around the geniculate ganglion.

are some basic principles that apply. Intrinsic tumour treatment is directed at optimizing and maintaining best facial motor function. Patients who are able to close their eyes and are stable are typically managed conservatively with observation, serial imaging, and annual hearing tests and EMG. Patients with House–Brackmann grade III showing clinical deterioration and those with grade IV–VI are considered for surgery. Those with good function but who suffer recurrent weakness may benefit from decompression surgery. The surgical approach considers the size and position of the tumour as well as residual hearing. A transmastoid and/or middle cranial fossa approach is recommended for patients with good hearing, and a translabyrinthine approach for those with poor hearing.

Facial therapy and reanimation

Despite optimal treatment many patients are left with some persistent facial weakness. There is not just the visible deformity to consider, these patients also suffer with functional problems relating to lagophthalmos, oral continence, and speech. This has an impact on their psychology and quality of life. There are a number of treatment modalities aimed at improving cosmetic appearance and function.

Facial therapy commences with a full assessment. The flexibility of the facial musculature is recorded, and photographic evidence of facial expression is collected to monitor progress. Initial therapy is focused on massage, which ameliorates mobility and circulation. Patients are also taught eyelid and brow stretching techniques. Patients are encouraged to tape their eyes on a regular basis. They are provided with individualized therapy dependent on the specific muscle groups identified as inactive. In those with a poor response to therapy, especially if the palsy is complicated by spasm or hyperkinesis, regular Botox injections may be beneficial.

Reanimation surgery

The main factor in deciding the surgical approach to facial nerve reconstruction is the status of the facial musculature. Successful muscle unit survival is unlikely beyond 18–24 months after nerve injury. Techniques utilizing the native musculature typically give better results so early referral for reanimation is critical.

Facial musculature viable

Primary nerve repair with accurate end-to-end anastomosis is recommended in facial nerve transection following oncological resection, temporal bone fracture, or iatrogenic injury. Where the nerve defect can not be overcome by rerouting, interposition nerve grafts, great auricular or sural, can be used. Where the proximal nerve stump is not available, nerve transfer using a branch of the masseteric or hypoglossal nerve can provide good outcomes.

Facial musculature non-viable

Where the onset of the facial palsy is more than 24 months old, with no response on EMG or in congenital cases, muscle units are either non-viable or non-existent. Facial reanimation can be achieved with muscle transposition (temporalis or masseter) or free muscle transfer (gracilis). These techniques provide animation to the lower

third of the face and may need to be combined with oculoplastic surgery.

Static procedures

Static techniques aim to achieve resting symmetry but do not achieve spontaneous dynamic movement. They can be used either alone, or in conjunction with more complex nerve and muscle techniques. Static slings such as facia lata, can be used to suspend the nasolabial fold or the perioral muscles and can achieve good resting symmetry. Facelift and brow lift procedures have a place in selective cases.

Gold and platinum weights to the upper lid can be used in lagophthalmos to help protect the cornea and medal and lateral canthoplasty to correct lower lid ectropion.

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11

Sensorineural hearing loss

Ted A. Meyer

Introduction

We begin with a straightforward, albeit simplistic, classification of hearing loss into the two major types, conductive and sensorineural. Conductive hearing loss results from a decrease in the transmission of sound through the external and/or middle ear. More recent evidence supports conductive hearing loss also arising from abnormalities in the inner ear anatomy (Box 11.1) and mechanics such as those seen with superior semicircular canal dehiscence. Sensorineural hearing loss (SNHL), on the other hand, results from an absence or loss of function of the inner ear and its connections with the remainder of the auditory system. Some patients present with both conductive hearing loss and SNHL. Additionally, some patients present with central or functional hearing losses.

Aetiology of sensorineural hearing loss

Aetiologies of SNHL are varied. A short list includes congenital anomalies, developmental disorders, autoimmune insults, inflammation, medications, exposures to toxins, metabolic disorders, infection, and trauma, including noise. In some patients, congenital or genetic anomalies play important roles in the lack of appropriate development of the ear and expression or progression of SNHL. Numerous genetic disorders, some with well-understood single point mutations, others with more complicated inheritance patterns, result in SNHL through dominant, recessive, X-linked, and mitochondrial genetic transmissions, with variable penetrance depending on the specific mutation, gene, or genes involved (Table 11.1). With some genetic disorders, hearing loss is an isolated finding, while in others, a variety of other developmental or progressive medical problems arise. For example, vision loss, renal dysfunction, and cardiac arrhythmias are commonly associated with SNHL, and the astute clinician should be aware of these and other issues in addition to hearing loss. An exhaustive list of genetic disorders and associated health-related issues in patients with SNHL is not within the scope of this chapter.

As we age, damage to the inner ear, and particularly the hair cells, occurs from a wide variety of causes. In many cases the damage corresponds to the inability of the inner ear to overcome oxidative metabolic stressors. In humans, once damage to the inner ear structures reaches a critical threshold, recovery, to a great extent, does not occur. What might have been a short-lived SNHL becomes permanent. For example, SNHL from noise exposure often improves with time. The loss is generally termed a temporary threshold shift. The hair cells recover from the noise exposure insult, and with the reparative process, hearing improves. If, however, after a significant noise exposure, the hair cells or other inner ear structures do not recover, the hearing loss persists. This is generally termed a permanent threshold shift. If we extrapolate this presentation to other disorders affecting the inner ear, the eventual inability of the body to repair damage to the hair cells and other supportive structures describes a simplified view of progressive SNHL.

Box 11.1 Inner ear developmental abnormalities span a range from absence of inner ear structures to minor anomalies in the size or shape of portions of the labyrinth

- Michel aplasia—absence of labyrinth.
- Common cavity.
- Mondini abnormality—less than 2.5 cochlear turns.
- Enlarged vestibular aqueduct.

Table 11.1 Common genetic mutations associated with sensorineural hearing loss

Gene, condition	Chromosomal location
GJB2—connexin 26 deafness	Chromosome 13
CLRN1—Usher syndrome type 3	Chromosome 11
PAX3—Waardenburg syndrome	Chromosome 2
KCNQ1—Jervell and Lange-Nielsen syndrome	Chromosome 11
PDS—Pendred syndrome	Chromosome 7
EYA1—branchio-oto-renal syndrome	Chromosome 8
COL1A1—osteogenesis imperfecta	Chromosome 17
COL1A5—Alport syndrome	Chromosome X

Presentation of sensorineural hearing loss

The majority of newborns receive hearing screening evaluations prior to leaving the hospital in many countries. For those children who fail the initial screen, timely follow-up is recommended with an audiologist for further testing including an auditory brainstem response test and an otolaryngologist for further evaluation. It is recommended that children with hearing loss identified in the newborn period receive appropriate amplification shortly after identification. Routine audiological testing and otolaryngology follow-up are critical to identify changes in hearing, provide appropriate amplification, manage related medical issues, and provide the parents of the child with information about different communication options. Referral to a speech–language pathologist is recommended for young children with hearing loss. For children with significant SNHL, cochlear implantation can be considered.

SNHL can begin and progress at any time and at any rate. For some, the progress of SNHL is slow with predictable loss over decades. For others, the progress is variable, and for a few, SNHL occurs rapidly, even suddenly, with no recovery despite appropriate medical treatment. Beyond the newborn period, it is imperative that children are monitored closely by their parents and paediatricians so that hearing loss is detected early, and medical and audiological interventions are implemented in a timely manner to benefit the child. For example, a child who is not developing speech and language at an appropriate rate could have an underlying hearing loss. A child who struggles hearing in noise or struggles with determining the direction of sound could have a unilateral hearing loss. For a child with hearing loss who generally benefits from hearing aids, a bout of otitis media with effusion could impair hearing significantly if the effusion remains in the middle ear for an extended period of time. The conductive hearing loss from the fluid added to an underlying SNHL might not be overcome with routine amplification. An evaluation with an otolaryngologist is warranted, and a myringotomy and pressure-equalization tube might be warranted as this straightforward procedure is expected to provide significant hearing benefit to the child. For a child with fluctuating SNHL, an underlying autoimmune disorder should be considered, and evaluation and management by a rheumatologist might help the child maintain or even regain useful hearing. Thankfully, many children with SNHL maintain reasonably stable hearing for many years, and they benefit substantially from consistent hearing aid use.

As we age, the percentage of the population with SNHL, as well as the amount of SNHL, increases. Many patients with mild SNHL do not realize that they have hearing loss, and they do not struggle significantly with communication in routine settings. As SNHL progresses, patients generally find background noise more challenging, the clarity of speech decreases, localization becomes more difficult, and, overall, communication takes more effort than in the past. The patient with SNHL relies more on lip-reading, asks for repetition, avoids challenging communication situations, and might even take longer to respond as processing of speech becomes more challenging. In the clinic, patients commonly report frustration with family and friends mumbling, and family and friends commonly report frustration with having to frequently repeat themselves. In addition, denial of hearing loss, social isolation, cost, and reluctance to wear hearing aids are challenges faced by both patients and clinicians.

SNHL is typically symmetric as both ears generally receive similar levels or amounts of damaging exposures that cause harm. Exposure to toxic medication, for example, unless the medication is applied directly to an ear, should cause a similar level of hearing loss in both ears. Inflammation, or other autoimmune processes, ought to affect both ears relatively equally. On the other hand, asymmetric SNHL is expected from other exposures or disorders. For example, acoustic trauma, if not experienced symmetrically, generally causes more hearing loss to the ear closer to the sound than the ear that is better shielded. As the majority of the population is right-handed, SNHL from noise exposure from shooting long guns is expected to be greater in the left ear. Ménière's disease, for reasons not well understood, generally affects one ear more than the other. Sudden SNHL also typically affects one ear. Although rare, both are seen regularly in otolaryngology practices. In these examples, the loss of hearing in one or both ears, and the associated otological symptoms such as aural fullness and tinnitus, can be quite bothersome to patients. Tinnitus is typically described as a ringing sound, but the loudness, pitch, and other auditory percepts described by patients are varied. Tinnitus can occur without hearing loss or a significant difficulty in understanding speech or other communication problems. A patient who presents with tinnitus, however, has a high likelihood of at least some degree of SNHL and should undergo audiometric evaluation.

Audiometric testing

For young children, audiometric evaluation includes otoacoustic emissions, which measure the function of the inner ear, and auditory brainstem responses, which measure the function of the lower portion of the auditory system. As children develop, they are better able to respond to auditory stimuli through looking or head turning. Further development with age leads to the child being able to undertake more advanced behavioural testing methodology such as play audiometry and then routine audiometry including testing with speech.

For older children and adults presenting with signs and symptoms of SNHL, routine behavioural audiometric studies are obtained. Both air- and bone-conduction thresholds are necessary to determine the type and amount of hearing loss. For patients with SNHL, audiograms demonstrate little difference between air- and bone-conduction thresholds. Normal hearing thresholds fall in the range from approximately −10 dB sound pressure level (SPL) to 20–25 dB SPL. Thresholds corresponding to mild hearing loss fall between 20–25 dB SPL and 40 dB SPL. Moderate hearing loss covers the range of 40–60 dB SPL, severe loss in the range of 60–90 dB SPL, and profound loss greater than 90 dB SPL. Ears are generally tested in isolation under earphones or with insert ear plugs.

The shape of the audiogram can give clues as to the aetiology of the hearing loss (**Figure 11.1**). For example, the hearing loss associated with ageing, or presbycusis, typically increases with frequency. Patients generally have relatively good hearing in the lower frequencies with progressive loss across the higher-frequency spectrum. As another example, patients with Ménière's disease often present with a fluctuating low-frequency SNHL, with return to better hearing in between vertiginous spells. Over time, however, the hearing loss

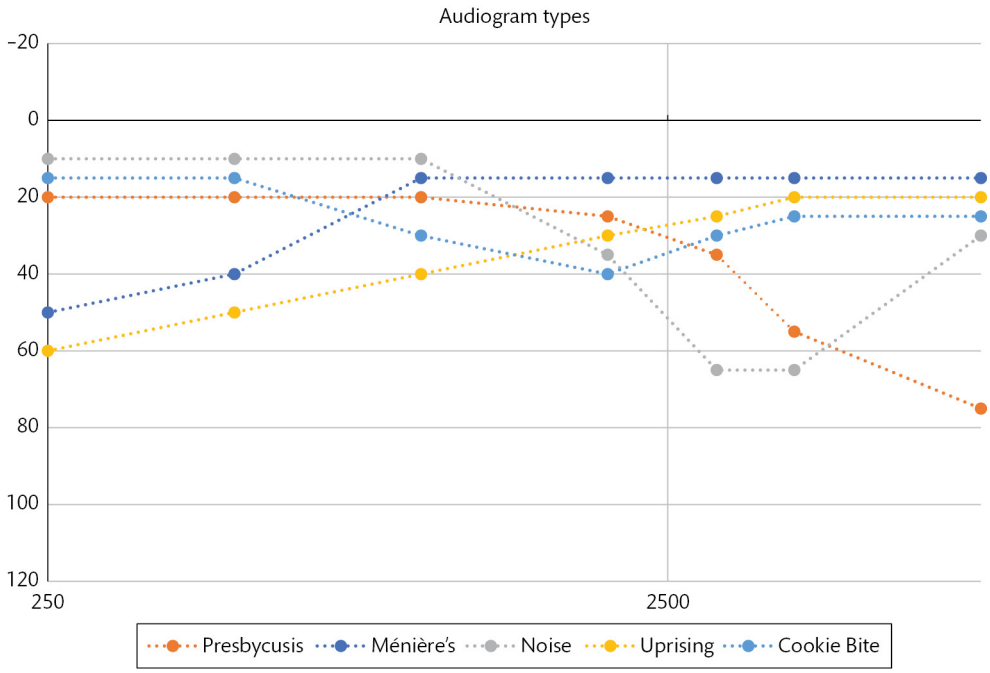

Figure 11.1 Audiogram examples.

for many patients with Ménière's disease becomes a relatively flat SNHL across all frequencies in the 60 dB SPL range. SNHL from noise exposure typically presents as a noise notch, or worse hearing, in the 3000–4000 Hz range with better hearing at lower and higher frequencies. Hearing loss with a significant Mondini malformation, a cochlea with less than the normal 2.5 turns, often presents as an up-sloping SNHL.

Speech testing should also be obtained along with pure tone audiometry. A speech reception threshold is obtained as a comparison to pure tone thresholds. A speech reception threshold uses two-syllable words with equal emphasis on both syllables (spondees), for example, the word 'RAILROAD'. Monosyllabic word testing is also performed, typically at a level 40 dB above the patient's pure tone threshold average or speech reception threshold. Differences between ears in thresholds or word recognition scores should lead to further evaluation, including testing for malingering, tuning fork testing, auditory brainstem response, or radiological testing (computed tomography or magnetic resonance imaging) to search for a cause and perhaps better explain the hearing loss of difference between the two ears.

Clarity or fidelity of hearing

The onset, the progression, the amount of hearing loss, the shape of the audiogram, the speech reception threshold, and monosyllable word testing does not tell the entire story of a patient's hearing loss or his or her experience with the hearing loss. For example, presbycusis, or hearing loss with ageing, often causes a greater loss of clarity, or word recognition, than would be expected based on numerical calculations from the audiogram. In addition, testing in the clinic is generally performed without hearing aids and without background noise. The world is noisy, and many patients remark that they do pretty well in quiet situations, but they struggle where there is background noise. Although hearing aids help many patients, some remark that their hearing aids amplify sound, but they do not make speech any easier to understand. Struggles in noise, and lack of improvement, or perhaps even a decrease in clarity with amplification for some, and cost, are major impediments to fitting more patients with SNHL with hearing aids.

Progression of sensorineural hearing loss

While SNHL for the average patient progresses as expected with age, some patients have relatively little hearing loss over time, and others progress rapidly. In general, SNHL progresses about 0.5 dB per year with greater losses in the higher frequencies. The rate of hearing loss also changes with time for some patients for many different reasons. Hearing loss in both ears generally progresses at about the same rate unless noise, trauma, or a pathological process affects one ear more than the other. In general, men have more rapid progression of hearing loss throughout adulthood than women, with greater differences in the higher frequencies. Much current research focuses on other health-related issues and their relations to hearing loss. Of great interest is the rate of cognitive decline and hearing loss with ageing, including a concentrated effort to reduce cognitive decline with proper amplification.

Prevention of sensorineural hearing loss

In general, once SNHL begins, it does not recover or improve, but exceptions occur. It is recommended to reduce or avoid exposures to substances or situations that are typically ototoxic or damaging to the inner to prevent sensorineural loss. For example, by avoiding loud noise, wearing hearing protection, avoiding trauma, and avoiding exposures to ototoxic medications, one can reduce risks of SNHL. As healthcare providers we discuss reducing risks of obesity,

hypertension, or cancer through weight management, salt intake, or smoking cessation or reducing time in the sun, for example. We should also discuss reducing risks of hearing loss with our patients. We should also consider medications or dosages of medications that limit ototoxicity. For example, if a patient is on a loop diuretic, the addition of an aminoglycoside might be reconsidered for another class of antibiotic as long as clinically appropriate in an attempt to reduce the potential additive effects of the medications in combination.

Treatment and management of sensorineural hearing loss

For patients with sudden SNHL, the general recommendation is early treatment with steroids and evaluation for retrocochlear pathology with a magnetic resonance imaging scan. Different regimens of oral steroids, intratympanic injections, and combined therapies have been recommended for many years with various success rates reported in an extensive literature. Many other therapeutic regimens have been developed, again with varying success and often not given in well-controlled studies. For patients with mild, sudden SNHL, recovery rates are higher than for those with significant losses.

The mainstay of treatment of SNHL is amplification with properly fitted and programmed hearing aids. Many related challenges can be difficult to overcome. For example, when hearing aids are not covered by health insurance, an additional financial burden is placed on the patient in the management of SNHL. For many patients, a pair of hearing aids is not financially possible, and many patients do not receive hearing aids because they do not have the financial resources. The amount of hearing loss and the shape of the audiogram differ for each individual as does the size and shape of the ear and canal. Properly fitting hearing aids, both to match the patient's audiogram and external ears, are important for performance and satisfaction with the hearing aids. Some hearing losses are relatively easy to fit with hearing aids, while other hearing losses are challenging. Patients with recruitment, for example, often find hearing aids difficult to tolerate. Patients with tinnitus often obtain a reduction in tinnitus with hearing aids, but some patients find that amplification makes their tinnitus more bothersome. Patients with challenging ear canals also often struggle with hearing aids.

Many other technologies are available to help those with hearing loss. For example, Bluetooth or FM or similar data transfer platforms incorporated into microphones, phones, televisions, and hearing aids allow for direct communication between devices. Devices that alert through light or direct vibration make alarms more generally available to those with hearing loss. Accurate and nearly real-time speech detection/translation software has improved greatly in the recent past, and many people with SNHL are able to take advantage of this technology on their smartphones or other devices. Lastly, video streaming has become a significant part of much of our work and social lives during the COVID-19 pandemic, and our improved familiarity with this technology has improved access for many patients with SNHL, even in a time where communication can be even more challenging.

For patients with significant hearing loss, if hearing aids provide only limited benefit, cochlear implants should be recommended (see Chapter 16). Typically, the patient has bilateral moderate/severe to profound SNHL and poor aided word understanding. Over the past several decades, the criteria for implantation have relaxed with more patients eligible to receive cochlear implants today than in years past. In addition, some patients with unilateral SNHL have the option to consider a cochlear implant to help with localization, hearing in background noise, and to reduce bothersome tinnitus.

As we learn more about the workings of the inner ear, including the possibility of hair cell regeneration, many novel therapeutics to protect or recover hearing are being and will be developed to provide current and future patients with SNHL options for better hearing. The future for patients with SNHL is bright.

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12

Conductive (or mixed) hearing loss with an intact tympanic membrane

Paul Merkus and Thadé Goderie

Introduction: conductive hearing loss is more than a perforation, cholesteatoma, or otosclerosis

Conductive hearing loss (CHL) seems a straightforward condition but can be challenging due to the vast differential diagnoses (Table 12.1).

This is especially the case when the three most prevalent diagnoses of CHL are ruled out, that is, tympanic membrane (TM) perforation, cholesteatoma, and otosclerosis. We will go into these conditions briefly as they are discussed in other chapters.

A *TM perforation* is the most easily diagnosed condition. On otoscopy a perforated ear drum can be seen. On pure tone audiometry the air–bone gap (ABG) is linearly associated with the size of the perforation with a maximum ABG of 25–30 dB in case of a total perforation at 4 kHz.[1] If the perforation is subtotal or less and ABG exceeds 25 dB an additional cause should be considered, such as tympanosclerosis. TM perforation is discussed in Chapter 22.

Acquired cholesteatoma is often discovered via a history of chronic suppurative otitis media and otomicroscopy showing a retracted TM with a pocket full of keratin accumulation (see Chapter 19). A high-resolution computed tomography (CT) scan will strengthen the clinical diagnosis of cholesteatoma, especially when local opacification and bone erosion are seen (Figure 12.1). Residual cholesteatoma is more difficult to diagnose in the outpatient clinic, but can be ruled out with diffusion-weighted magnetic resonance imaging (MRI).

When the TM is intact, diagnosing the cause of the CHL is not as straightforward. In case of a history of chronic suppurative otitis media, tympanosclerosis is likely, but identifying alternative diagnoses during the initial assessment can be challenging. *Otosclerosis* is the most prevalent cause of CHL with an intact TM in absence of effusion. The classic presentation is an adult-onset, progressive CHL and the audiogram shows a CHL with a 'Carhart notch' at 2 kHz. A CT scan often shows an otospongiotic area anterior of the oval window, as shown in Figure 12.2, with a sensitivity of 93%.[2] Otosclerosis is discussed in Chapter 20.

If medical history, otomicroscopy, audiometry, or CT or MRI scanning is not according to the characteristics as described above, an alternative diagnosis should be considered as more knowledge is needed to unravel the cause and the best solution. This expertise will be covered in this chapter, with a focus on CHL with an intact ear drum.

Assessment (history)

The first step in seeing a patient with a hearing loss is to establish if a patient has a conductive, a sensorineural, or a mixed (i.e. conductive and sensorineural) type hearing loss. Preferably your patient will have an audiogram at the start of the evaluation (see Chapter 5). It will help you in making a differential diagnosis and to direct your history taking as the aetiology is very different for different types of hearing loss.

Meticulous history taking provides important clues for the aetiology of a CHL. It should include *age of onset* and *duration* of hearing loss. This can distinguish between congenital and acquired CHLs. *Laterality* should be asked for and also the *evolution* of hearing loss (i.e. abrupt, gradual, or fluctuating). In case of an abrupt onset, the *triggering factor* should be noted. When a patient has a *history of ear surgery*, obtain, if possible, the original report of the procedure to assure the indication for and the details of the procedure. The *family history* is important. Some forms of CHL can be hereditary and have a delayed onset such as osteogenesis imperfecta, otosclerosis, or Paget's disease, or can be caused by hereditary tumours such as paraganglioma involving the temporal bone. Multiple syndromes are known to be associated with conductive or mixed hearing loss. Therefore, in patients with a known syndrome, it is important to ascertain whether hearing loss is part of that syndrome and, if so, to determine the precise cause of the hearing loss. Further, ask for *associated symptoms*, such as vestibular symptoms, pain, tinnitus (pulsatile/constant), aural fullness, facial nerve function, actions that change intensity of symptoms, antecedent illness, and otorrhoea. When discussing vestibular symptoms specifically ask for sound-induced unsteadiness, vertigo, or oscillopsia, known

Table 12.1 Different diagnosis of conductive hearing loss

Diagnosis	Specific history	Tympanic membrane and middle ear	Audiogram	Tympanogram	CT scan	Other
		Aerated				
Otosclerosis	Slowly progressive hearing loss, 50% have affected family members	Intact TM, aerated ME	CHL, Carhart notch 2000 Hz	Type A or As	Demineralization anterior of vestibule	Starts unilateral, becomes bilateral in 70% of cases
Otosclerosis (retrofenestral type)	Slowly progressive hearing loss	Intact TM, aerated ME	Mixed hearing loss, less prominent Carhart notch at 2000 Hz	Type A or As	Demineralization of the otic capsule	
Tympanosclerosis	History of ear infections	Intact TM, possibly myringosclerosis, aerated ME	CHL	Type A or As or B	Calcifications around the ossicular chain	
Ossicular chain disconnection	Trauma or infection	Intact TM, aerated ME, TM can be adherent to middle structures and ossicles	CHL	Type A, Ad, B or C	Most often disconnection of incudostapedial joint/ incus long process erosion	
Semicircular canal dehiscence	Hearing and balance symptoms, such as Tullio's phenomenon, pulsatile tinnitus, autophony, and hearing other bodily sounds	Intact TM, aerated ME	CHL (low-frequency)	Type A	Dehiscence of the bone overlying a semicircular canal, most commonly the superior canal	Other diagnostic tool is the cervical vestibular evoked myogenic potential
Enlarged vestibular aqueduct	Progressive (mixed) hearing loss often onset in childhood	Intact TM, aerated ME	(Mixed) hearing loss	Type A	Enlarged vestibular aqueduct best visible in axial CT	Can be unilateral, bilateral, and as part of syndrome
Persistent stapedial artery	Pulsatile tinnitus, hearing loss	Intact TM, aerated ME	CHL	Type A or As	Aplastic or hypoplastic foramen spinosum, linear soft tissue crossing the promontory	
Meningoencephalocele	Trauma or infection	Intact TM, aerated ME	CHL	Type A or As	Tegmen defect with protrusion of the meningoencephalocele through the tegmen onto the ossicular chain	
		Fluid filled				
Otitis media with effusion	Hearing loss and aural fullness history of ear infections, often bilateral, or (infrequently) pain	Intact TM, fluid-filled ME	CHL	Type As, B	Intact bony structures, no signs of destruction	
Cerebrospinal fluid leak (with or without encephalocele)	CHL, trauma, idiopathic, intracranial hypertension	Intact TM, fluid-filled ME	CHL	Type As, B	Thinned or interrupted tegmen bone and opacified mastoid	MRI showing herniated brain in case of an encephalocele
		Mass filled examples, not a complete overview)				
Congenital cholesteatoma	Hearing loss, earache, aural fullness	Intact TM, white mass often anterior superiorly	Only CHL when touching ossicular chain	Type A, As, B	Local destruction is possible, but not always present	Diffusion-weighted MRI is needed

Table 12.1 Continued

Diagnosis	Specific history	Tympanic membrane and middle ear	Audiogram	Tympanogram	CT scan	Other
Gout tophus	CHL	Yellow-whitish calcareous lobular mass located in the mesotympanum anterior to the malleus	CHL	Type A or As or B	Heterogeneous hyperdense round mass	
Tympanic/ jugulotympanic paraganglioma	Pulsatile tinnitus	Intact TM, red pulsating mass, inferiorly	CHL when touching ossicular chain, sensorineural hearing loss in case of involvement inner ear	Type A, As, B	Opacification of caudal middle ear or extension and destruction towards and around jugular bulb	MRI shows a typical salt-and-pepper appearance
Facial nerve tumour (schwannoma, haemangioma)	Facial nerve paresis and aural fullness	Intact TM, white mass variable, often superiorly	Only CHL when touching ossicular chain	Type B	Opacification adjacent to the facial nerve canal often from widened ganglion geniculi	
Middle ear adenoma and carcinoid	CHL, and aural fullness	Located around ossicular chain, colour is variable	CHL	Type A, As, B	Local destruction is possible	MRI with contrast and biopsy is needed
Middle ear malignant neoplasm	Hearing loss, pain, cranial nerve deficit	Dependent on extension, local destruction	CHL or deaf ear	Type B	Invasive growth	MRI with contrast and biopsy is needed
Aberrant carotid artery	Pulsatile tinnitus	Intact TM, red structure and pulsating anteroinferiorly	CHL	Type B	Vessel crossing the inferior cochlear promontory from posterior to anterior	
High jugular bulb	Pulsatile tinnitus	Intact TM, blue structure, posterior inferiorly	Only CHL when touching ossicular chain	Type B	Bluish mass arising from the hypotympanum in the posterior mesotympanum	

CHL, conductive hearing loss; CT, computed tomography; ME, middle ear; MRI, magnetic resonance imaging; TM, tympanic membrane.

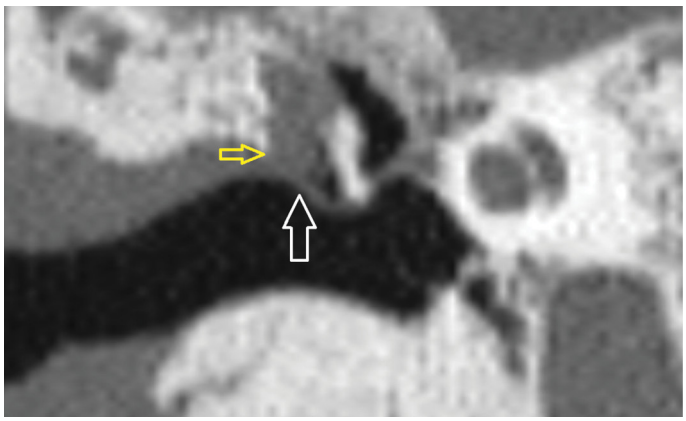

Figure 12.1 CT scan (coronal) of a cholesteatoma. The open smaller arrow shows the bony scutum erosion of medial ear canal wall enlarging Prussak's space. The bigger white arrow points at the mass in this space which is filled with keratin debris, often visible with otomicroscopy.

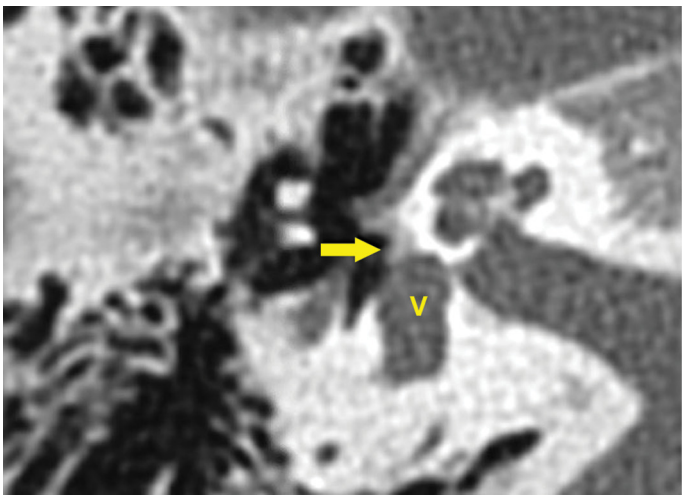

Figure 12.2 CT scan (axial) of the right temporal bone. The yellow arrow is pointing at the area anterior of the vestibule (v) which is isodense ('grey') and normally is hyperdense ('white'). This is the otospongiotic area in otosclerosis, starting anterior of the oval window ('ante fenestram').

as Tullio's phenomenon. Tullio's phenomenon is caused (but not exclusively) by a third window of the inner ear such as a dehiscence of the superior semicircular canal.[3] When a temporal bone tumour is suspected (mass behind intact ear drum), cranial nerve involvement should be ruled out.

Investigations

All patients suspected to have a hearing loss should undergo formal audiological testing, including *pure tone audiometry* with bone and air conduction and speech audiometry while masking the contralateral ear when indicated (see Chapter 5). A Weber test should confirm the side of the CHL. In case of a discrepancy between pure tone audiometry and speech audiometry or between pure tone audiometry and Weber testing, an experienced audiologist should repeat audiometry. It still happens that sensorineural hearing losses are mistaken for CHLs.

Perform *tympanometry* in CHL (see Chapter 5). A TM can be hypermobile (type Ad), suggestive of luxation of the middle ear ossicles, or stiffened (type As), suggestive of tympanosclerosis, or flat (type B), suggestive of otitis media or a TM perforation. Stapedial reflexes are used to test the mobility of the ossicular chain and more specifically of the stapes and can confirm otosclerosis or tympanosclerosis.

Additionally, *vestibular testing* can be performed (see Chapter XX). A fistula test can be done by applying tragal pressure. This can elicit a saccade (Hennebert's sign) and is associated with a third window of the inner ear, congenital syphilis, and Ménière's disease. Cervical vestibular evoked myogenic potentials (cVEMP) can be tested and assess the vestibulocollic reflex and tests specifically the saccule. Lowered threshold for eliciting a cVEMP and larger amplitudes indicate a superior canal dehiscence syndrome.[4]

Physical examination starts with the external ear. Size and shape should be assessed. Preauricular pits are sometimes associated with external auditory canal and middle ear anomalies, whereas microtia is very often associated with such anomalies. Visual inspection of the external auditory canal, TM, and middle ear (through the TM) should be performed with an otomicroscope rather than a handheld otoscope, because the former provides superior illumination, greater magnification, and allows instrumentation in the ear canal. Increased vascularization on the medial inferior side of the external auditory canal is sometimes seen in jugular paraganglioma. Inspect the TM meticulously on retractions, with or without cholesteatoma. A thickened TM (with myringosclerosis) indicates a history of middle ear infections and a CHL can be related to tympanosclerosis. The TM can also be thickened after previous ear surgery and sometimes blunting of the anterior tympanomeatal angle can be seen. In case of a transparent TM, most mesotympanic structures can be recognized. Sometimes the TM is atrophic (and can have a wrinkled appearance), which can lead to an atelectatic middle ear, incus erosion, or even a pars tensa cholesteatoma. Erosion of the long process and lenticular process of the incus and of the stapes suprastructure can usually be identified on microscopic examination.

Middle ear masses can be seen through the TM and should be assessed on colour and presence of pulsations. Colour of the middle ear mass could indicate the sort of tumour or anomaly in the middle ear (bluish (jugular bulb), reddish (paraganglioma, carotid artery, middle ear adenoma), pink (middle ear adenoma—colour can be variable), pale (congenital cholesteatoma, mostly anterior superior quadrant of the mesotympanum)) (**Table 12.1**). No biopsies of middle ear masses should be taken prior to imaging to avoid inadvertent damage to the facial nerve, or bleeding due to vascularized tumours, high jugular bulb, or an aberrant carotid artery. Always assess facial nerve function given its trajectory through the temporal bone. Examine other cranial nerves in case of temporal bone tumours and specifically the glossopharyngeal nerve, the vagal nerve, and the accessory nerve given their trajectory through the jugular foramen. Also, the trigeminal nerve (Meckel's cave) and abducens nerve (Dorello's canal) deserve special attention given their close proximity to the temporal bone.

Imaging

The middle ear ossicles and the contours of the middle ear are best seen on a high-resolution or cone beam CT scan with slice thickness of 0.5 mm or less. A CT scan will show bony structures as sharply delineated white structures, soft tissue as grey, and air as black. Examples of well-seen structures are the contours of each ossicle, the oval and round window niche, the cochleariform process, the temporal bone tegmen, and the external auditory canal and its medial sharp ending just lateral of the malleus (the scutum).

With high-resolution scanning the crura of the stapes, the tensor tympani tendon, the anterior ligament of the malleus, and the ear drum are also visible.

In case of a patient with an intact TM and CHL, one should rule out the following:

- *On axial CT images*: otosclerosis (otospongiotic bone anterior of the oval window niche), integrity and malformations of the ossicular chain, calcifications somewhere along the ossicles, calcifications of anterior ligament of the malleus, enlarged vestibular aqueduct, dehiscence of posterior semicircular canal, middle ear mass, or an inner ear or vascular anomaly.
- *On coronal CT images*: integrity and malformations of the ossicular chain (especially the connection of the lenticular process to the head of the stapes), calcifications of the tensor tympani tendon, the separation of the tegmen/brain and the ossicles, dehiscence of the superior semicircular canal, middle ear mass, or an inner ear or vascular anomaly.

Differential diagnoses and treatment

These have been separated by diagnosis (groups).

Cochlear (retrofenestral) otosclerosis

In patients with a CHL, an intact TM, and no signs of otitis, otosclerosis is the most prevalent cause of pure CHL but in 10% of cases it develops a mixed type of hearing loss.

Diagnosis: otosclerosis is primarily diagnosed by history taking and audiometry. However, a CT scan can confirm the diagnosis and shows a pathognomonic focus of decalcification anterior of the oval window (otospongiosis of the fissula ante fenestram) (**Figure 12.2**). Usually in cochlear otosclerosis a more pronounced pericochlear decalcification can be seen, known as retrofenestral otospongiosis, or a severe decalcified otic capsule (**Figure 12.3**).[5]

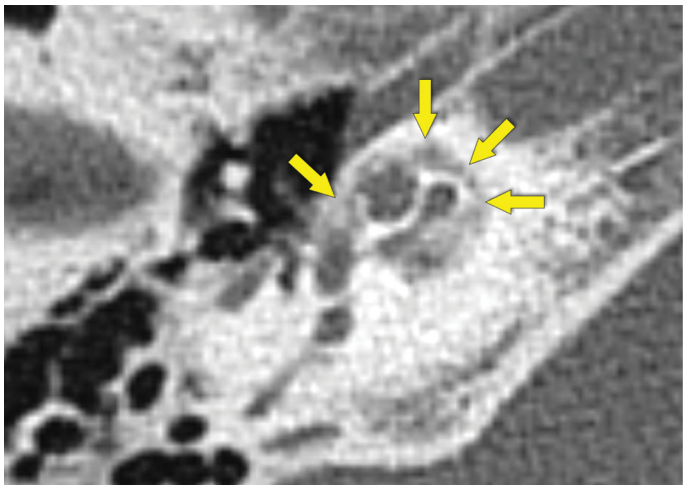

Figure 12.3 Retrofenestral otosclerosis is a radiological description of an advanced otospongiotic state of the otic capsule. In the axial scan there is a ring around the cochlea visible, a so-called halo sign, representing the otospongiotic/decalcified bone area (yellow arrows). In a severe retrofenestral state the contour of the cochlea is not visible anymore, and calcification of the scala tympani often present (not shown here).

Treatment: this type of mixed hearing loss is often treated with hearing aids and should always be referred for a CT scan. The conductive part of the moderate to severe hearing loss can be treated with a stapedotomy but because of the continuing need for hearing aids, this is often discarded. In these cases, bisphosphonates or sodium fluoride can be considered to diminish the progression of the hearing loss;[6] however, evidence supporting their efficacy is limited and long-term outcomes have yet to be established. In cases of severe to profound hearing loss, stapedotomy becomes a consideration again when hearing aids no longer provide sufficient benefit. Even—or especially—in patients eligible for a cochlear implant, a stapedotomy seems a reasonable alternative to cochlear implantation with the possibility to result in sufficient hearing with a hearing aid.[7–9] If stapedotomy fails or is unfavourable, a cochlear implant should be considered.

Tympanosclerosis

The main symptom is usually a pancochlear CHL, commencing in childhood and commonly bilateral. Tympanosclerosis is a calcification in the middle ear and TM (myringosclerosis) and a final stage of an inflammatory process (Figure 12.4). History of infection, calcification of the TM, and a CT scan with calcification around the middle ear ossicles are pathognomonic. Tympanogram is flat (B), normal with reduced mobility (As), or negative pressure (C).

Differential diagnosis: if the calcification is unilateral and presents as a solitary mass it could be an osteoma of the middle ear.[10] Another rarity resembling tympanosclerosis could be tophaceous gout of the middle ear.[11] Both rarities can be treated surgically.

Treatment: depending on the extension, most tympanosclerotic foci can be dealt with surgically. If all ossicles including the stapes are fixed, a hearing aid should be considered as an alternative to malleostapedotomy (stapes piston attached to the malleus). Malleostapedotomy is less successful compared to stapedotomy to the incus and has a higher chance of sensorineural hearing loss as a complication.

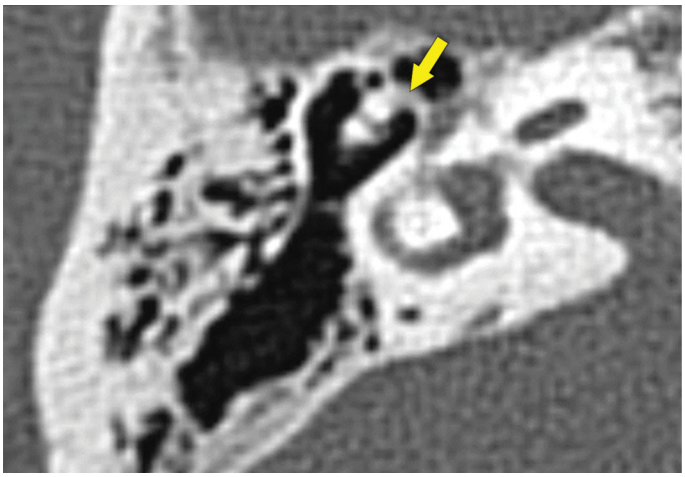

Figure 12.4 Tympanosclerosis with fixation of the malleus head.

Ossicular chain discontinuity

The history could be varied, from previous surgery and frequent infections, to trauma, but sometimes very subtle clues can lead to this diagnosis (frequent Valsalva manoeuvre needed to improve hearing and thin TM with a hypermobile tympanogram). The audiogram in case of a complete ossicular discontinuity typically shows an audiometric pattern of a large, flat ABG over all frequencies, and the tympanogram can be normal (A) to hypermobile (Ad). When encountering larger ABGs at higher frequencies compared to lower frequencies, a partial ossicular discontinuity should be considered.[12]

The most prevalent causes of ossicular chain discontinuity result from postinflammatory erosion of the lenticular process and long process of the incus (Figure 12.5) and from the traumatic disconnection of the incudostapedial joint. The medical history will differentiate between these two. High-resolution CT scanning is needed to clearly identify the long process of the incus and the connection of the lenticular process to the stapes. As the joint is only 0.5 × 0.8 mm,[13] it is often only visible on one single axial CT image and one coronal, and could be easily missed by an unexperienced radiologist or ear, nose, and throat surgeon.

Next to erosion, incus discontinuity may result from traumatic displacement or surgical removal (Figure 12.6).

A less frequent cause of ossicular chain discontinuity is a stapes that is luxated or broken (iatrogenic or traumatic, e.g. cotton tip) or a malleus handle which is broken (due to trauma or manipulation of the external ear).

Treatment: to surgically restore the connection between the TM and the stapes footplate. Chances of hearing improvement to an ABG within 20 dB are in the following sequence (from more likely to less likely): stapedotomy, incus stapes reconnection, incus interposition or a partial ossicular replacement prosthesis, and finally a total ossicular replacement prosthesis (assuming an absent stapes superstructure) (Figure 12.7).

Otitis media with effusion

Infectious and paediatric otitis media is more extensively covered in Chapters 8 and 88. In case of unilateral otitis media with effusion, a mass blocking the Eustachian tube should be ruled out by

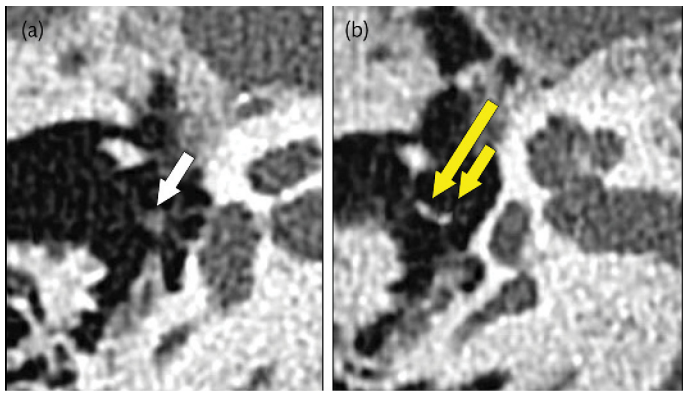

Figure 12.5 To investigate the integrity of the long process of the incus both axial and coronal images must be thoroughly examined. Often only a single image will show the connection of the incus and the stapes head, so moving between images is essential. Panel (a) shows the axial scan in which only the stapes head is visible (single white arrow). In the same image the incus long process is missing. In panel (b) (the mirrored other ear of the same patient) the two arrows show the lenticular process of the incus (long yellow arrow) and the head of the stapes (short yellow arrow) in close proximity.

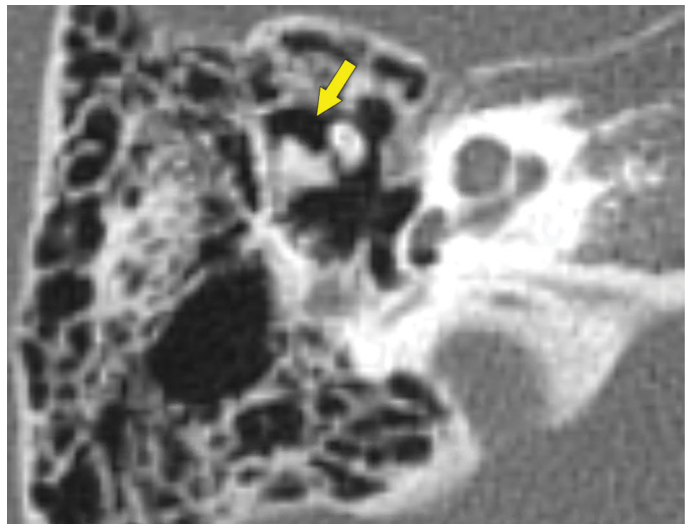

Figure 12.6 Due to trauma this incus malleus complex is dislocated. The integrity of the typical 'ice-cone' shape is easy to assess on axial images, even on a CT head with trauma protocol.

nasopharyngoscopy and/or MRI skull base, especially in patients with no prior ear pathology and accompanying complaints such as pain or cranial nerve deficits. Parapharyngeal tumours can easily be missed if no additional imaging is done. Unilateral otitis media with effusion can also be caused by cerebrospinal fluid (CSF) leaks caused by temporal bone trauma or spontaneously (discussed below).

Cerebrospinal fluid leaks and encephaloceles

Both CSF leaks and encephaloceles with CSF leak can cause an otitis media with effusion. In addition, an encephalocele can be in contact with the ossicular chain causing diminished sound transmission with CHL as result. One should be especially wary of a CSF leak if the middle ear keeps running clear fluid after putting in a tympanostomy tube. To confirm a CSF leak, middle ear fluid can be analysed for beta trace protein. CT and MRI can identify an encephalocele (Figure 12.8).

CSF leaks and encephaloceles can be caused by trauma, previous surgery, infection, cholesteatoma, and can be related to other tumours causing a bony erosion. Congenital abnormalities such as incomplete partition type 2 and a patent tympanomeningeal (Hyrtl) fissure can also cause CSF leaks.

Encephaloceles with CSF leaks can also occur spontaneously, which in 67% of cases is associated with idiopathic intracranial

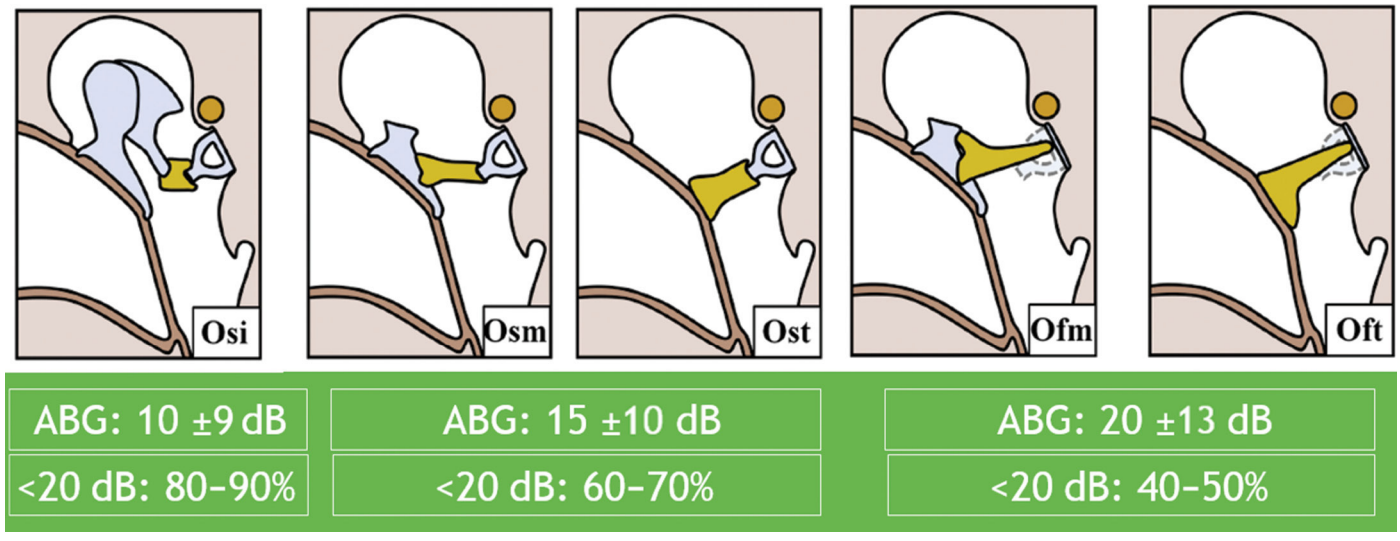

Figure 12.7 Ossiculoplasty results: this schematic overview presents the estimated results based on literature with different types of ossiculoplasties (note: the results are based on a scoping review of the literature with a rounding up of the outcomes). ABG, air-bone gap; Ofm, malleus to footplate; Oft, tympanic membrane to footplate; Osi, incus to stapes; Osm, malleus to stapes; Ost, tympanic membrane to stapes.
Drawings are of the SAMEO ATO classification, with permission of the International Otology Outcomes Group.[14]

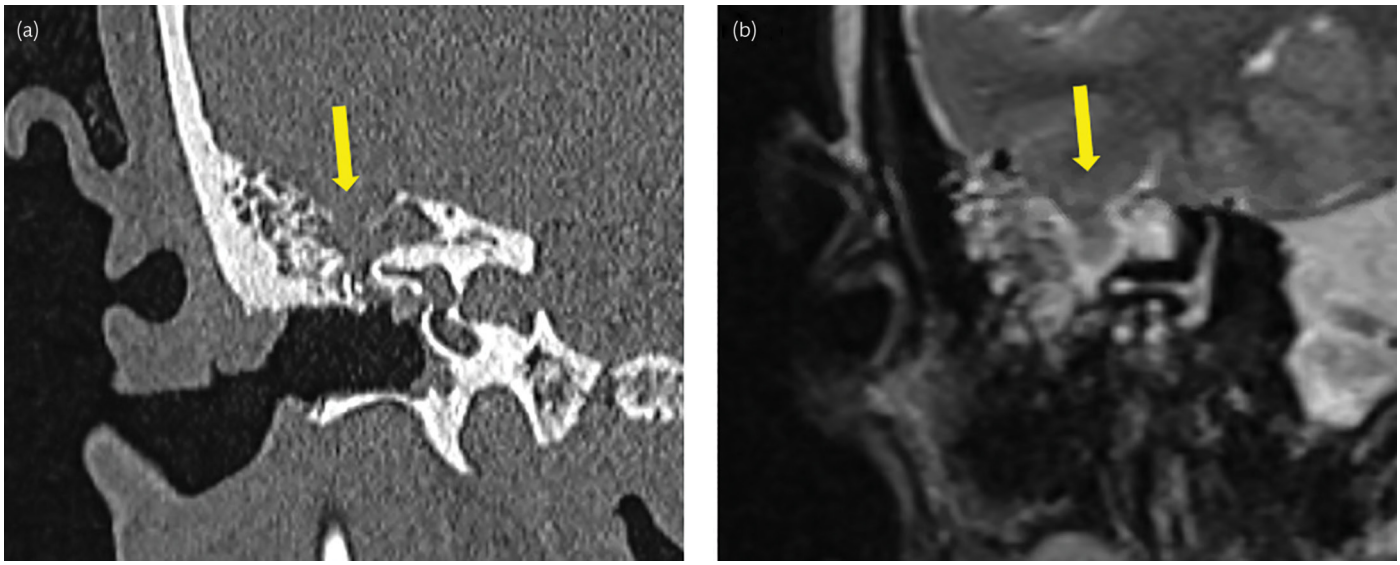

Figure 12.8 Encephalocele on CT and MRI scanning. This lady had a 10-year history of a discharging ear, after suffering a temporal bone trauma. (a) Coronal CT image of a defect in the tegmen mastoideum (yellow arrow). The tympanic membrane is partially disrupted and the remaining part is adherent to middle ear structures. (b) T2-weighted MRI of the same plane, confirming the defect in the tegmen and showing an encephalocele.

hypertension (IIH).[15] Following repair of the spontaneous CSF leak, neuro-ophthalmology should assess patients through a dilated fundoscopic examination. Lumbar puncture will be specifically administered for those displaying abnormal findings or experiencing symptoms like chronic daily headache or pulsatile tinnitus. This tailored approach is based on the understanding that papilloedema may not be present in all individuals with IIH.[16] If IIH exists, treatments to consider are weight loss, acetazolamide, or a ventriculoperitoneal shunt.[17]

CSF leaks caused by temporal bone fractures will resolve spontaneously in the first 5 days after injury, but they can persist and surgical repair is indicated if the leak persists after 10–14 days.

A meningoencephalocele without CSF leak can also cause CHL due to prolapse of the meningoencephalocele onto the ossicular chain.[18]

Sequelae of encephaloceles are CSF leak, epilepsy, and intracranial infections. Especially in meningitis cases with a previous temporal bone fracture, a CSF leak with or without encephalocele should be considered. Given the severity of the sequelae, surgical repair of the encephalocele is indicated. If the encephalocele is in contact with the middle ear ossicles, anterior to the ossicular chain, or multiple defects are present, a middle fossa approach is preferred. Defects of the tegmen mastoideum and posterior fossa are reconstructed through a transmastoid approach. Large defects are reconstructed through a middle fossa approach or a combined transmastoid–middle fossa approach. Alternatively, a subtotal petrosectomy with closure of the Eustachian tube can be performed, but this causes a maximum ABG and is hardly ever necessary.

Third-window abnormalities

Symptoms of a third-window lesion (with the stapes footplate and the round window being the first and the second window) are typically producing sound-/pressure-induced vertigo, oscillopsia or imbalance (Tullio's phenomenon), a positive fistula test (Hennebert's sign), and a low-frequency ABG. Furthermore, autophony and hearing bodily sounds can be heard such as pulsatile tinnitus, footsteps when walking, and eyes moving.[3] In these patients, pressure changes of the middle ear (Valsalva manoeuvre, coughing, or nose blowing) or intracranial pressure changes (straining at stool, straining in labour, coughing, blowing the nose, travelling in airplanes) can cause vestibular symptoms.[3] These symptoms should be actively asked about as patients often do not mention them spontaneously. A lowered cVEMP threshold ipsilateral to the CHL is suggestive of a (superior) semicircular canal dehiscence. The location of the third window in the bony structure of the inner ear can be varied.

Differential diagnoses: semicircular canal dehiscence (most often superior canal) (**Figure 12.9**), and enlarged vestibular aqueduct (**Figure 12.10**).

Treatment: there are two surgical approaches and two techniques to treat a superior semicircular canal dehiscence. Approaches: middle fossa and transmastoid. Techniques: resurfacing or plugging. The advantage of resurfacing is that the function of the canal can be preserved. It should be considered if the contralateral ear is affected as well. Resurfacing is only possible through a middle fossa approach. It seems that the middle fossa approach is the most common approach, but also plugging via a transmastoid approach has good results.[17,19]

Middle ear masses

The differential diagnosis of middle ear masses is diverse and extensive. Benign tumours of the middle ear include middle ear adenoma, cholesteatoma, mucosal adenoma, ceruminous adenoma, carcinoid, paraganglioma, pleomorphic adenoma, meningioma, schwannoma, endolymphatic sac tumour, Schneiderian-type mucosal papilloma, lipoma, epidermoid, and tophaceous gout (**Figure 12.11**).[20–22]

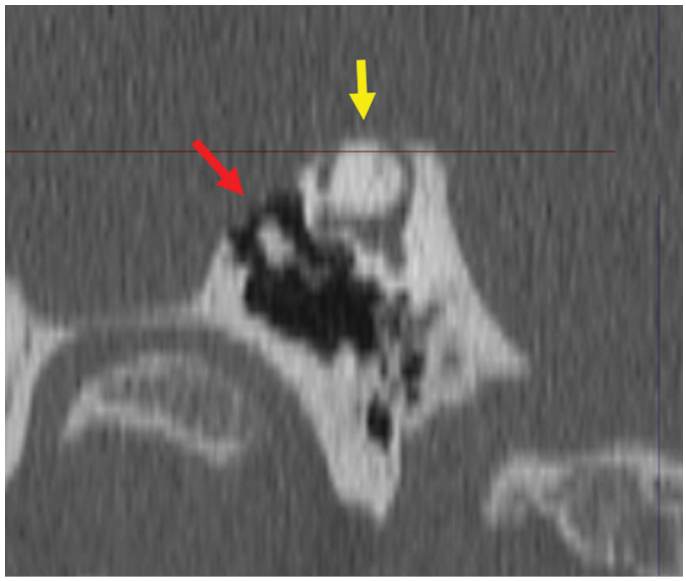

Figure 12.9 Superior semicircular canal dehiscence. This CT image is rotated in the plane of the superior semicircular canal. The yellow arrow indicates the superior semicircular canal dehiscence. The red arrow indicates a tegmen dehiscence.

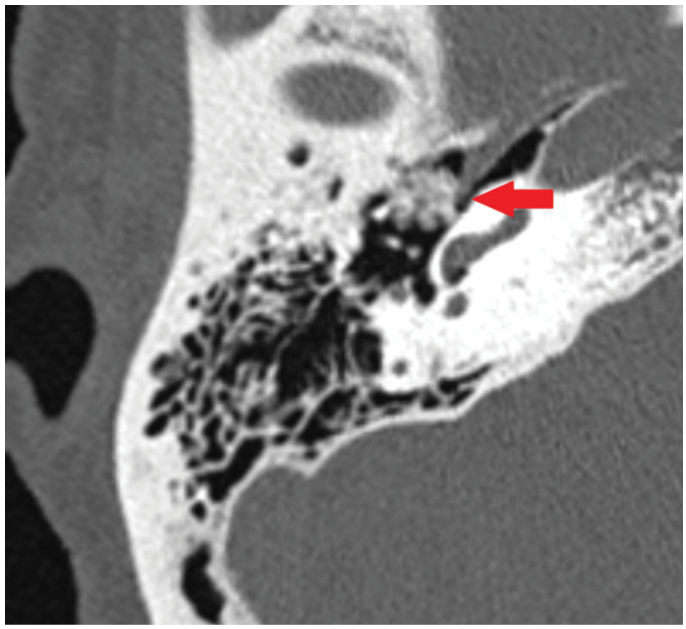

Figure 12.11 Axial CT imaging of the middle ear showing a heterogeneous hyperdense round mass of 5.5 mm in the right middle ear with a close relation to the malleus, eardrum, and tegmen.

Malignant tumours of the middle ear are rare with an estimated 0.18 cases per million people.[23] Most commonly they are squamous cell carcinomas (50%). But a variety of other malignancies have been described including rhabdomyosarcoma (especially in children) (5%) adenoid cystic carcinoma (4%), papillary adenocarcinoma (4%), and malignant melanoma (2%).[24]

Symptoms: otalgia, otorrhoea, and hearing loss are the most common presenting symptoms of malignant tumours of the middle ear. Therefore, it can be hard to distinguish malignant tumours of the middle ear from benign tumours or even chronic suppurative otitis media. In case of therapy-resistant chronic suppurative otitis media, consider taking a biopsy to rule out malignant disease. Facial nerve paresis, hoarseness, trismus, and dysphagia are signs of advanced disease. Tumours can be differentiated on several imaging and histological characteristics. It is important to differentiate between benign and malign masses to tailor treatment. Due to the extensive list it goes beyond the extent of this chapter to discuss all treatment options of the different middle ear masses.

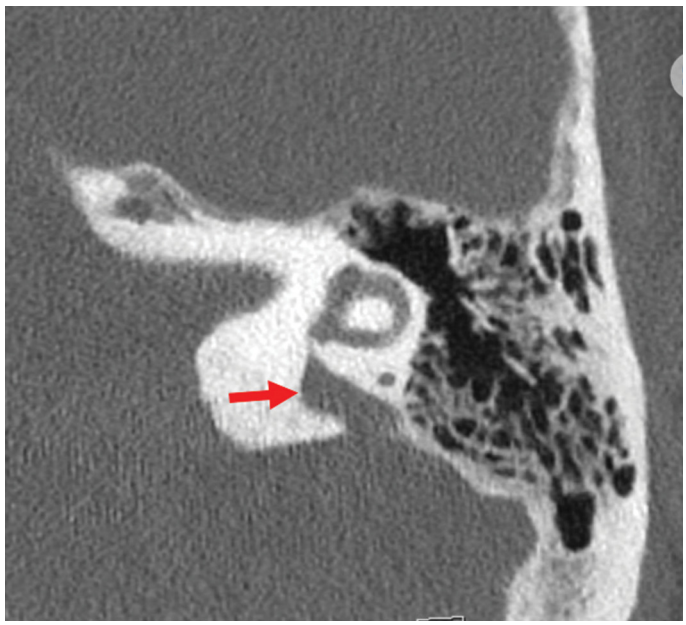

Figure 12.10 Enlarged vestibular aqueduct. Axial CT image. The red arrow indicates the enlarged vestibular aqueduct.

Vascular anomaly

A red mass behind the ear drum and pulsatile tinnitus could be caused by a tympanic paraganglioma, jugulotympanic paraganglioma, or a vascular anomaly, like aberrant internal carotid artery, persistent stapedial artery, or a high-riding jugular bulb. Middle ear masses could cause a similar presentation, and are discussed in the previous section (see 'Middle ear masses').

Symptoms: most of the time pulsatile tinnitus is the most prominent symptom and CHL could be absent. In case of an aberrant carotid artery (**Figure 12.12**) or a paraganglioma, pulsations can be seen when the mass is in contact with the TM. Recruitment of vessels can occur in case of jugulotympanic paraganglioma and these are visible at the inferior medial side of the external auditory canal. The diagnosis is made using imaging, preferably CT, but also with MRI.

Treatment: no follow-up is needed for high-riding jugular bulb or an aberrant internal carotid artery, unless complaints are increasing. For treatment of a persistent stapedial artery, suffering from both pulsatile tinnitus and hearing loss, the combination of persistent stapedial artery transection and stapes surgery seems appropriate in experienced hands.[25]

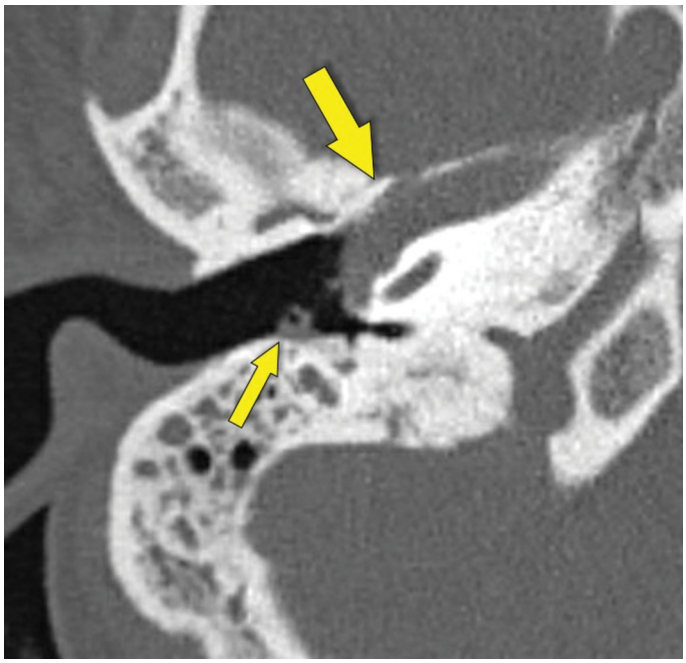

Figure 12.12 This axial CT image shows an aberrant carotid artery anterior and lateral to the promontory (large yellow arrow). It blocks the Eustachian tube. The small yellow arrow shows a ventilation tube in the posterior side of the ear drum.

Congenital ossicular chain malformations

Otitis media with effusion is the most common cause of CHL in young children. But after ruling out or treating otitis media with effusion, congenital unilateral CHL is often an isolated finding. Still, because of the association with more than 70 syndromes, a preoperative syndromic workup is needed in cases with a congenital ossicular chain malformation[26] (see Chapter 21). Some ossicular chain abnormalities are present in combination with microtia. Because this condition has many more implications, it will not be discussed further in this chapter.

Symptoms: congenital ossicular chain malformations can cause a mild CHL to a maximum ABG (i.e. 50–60 dB), but can be missed on newborn hearing screening and may not be diagnosed until the child can sit for behavioural testing.

Diagnostics: cases with an aplasia or dysplasia of the ossicular chain (such as dysplasia of the long process of the incus, or the stapes' superstructure) and aplasia of the oval window can be easily recognized on CT. But one has to be aware that congenital stapes fixation ('stapes ankylosis') can be present without visible abnormalities on CT scanning.[27,28]

Treatment[29]: patients with CHL due to isolated ossicular chain malformations are usually good candidates for surgical rehabilitation of hearing. An exception is congenital aplasia (absence) of the oval window, in which no good long-term hearing outcomes are seen. Other surgical reconstructions of a malformed ossicular chain have fairly good results and surgical management and outcomes of congenital stapes fixation are similar to that of otosclerosis. Hearing aids or a bone conduction device can be helpful when surgery is not possible or indicated (yet).

Temporal bone fractures

Temporal bone fractures can be subdivided into longitudinal (from cortex towards apex), axial, and oblique fractures, or, clinically more relevant, as otic capsule-sparing or otic capsule-involving fractures.[30] Conductive or mixed hearing losses mostly occur in longitudinal and oblique fractures. In the acute phase mostly a haemotympanum is seen, explaining part of the hearing loss. A CT head with trauma protocol should be performed to rule out intracranial bleedings in the acute phase. Also, a fracture through the inner ear can be seen, which is associated with a deaf ear. Often this CT head with trauma protocol has thicker slices than the specific CT for mastoid and middle ear, impeding optimal interpretation of the ossicular chain. For accurate assessment of hearing it is best to wait for 6 weeks, primarily for the blood in the middle ear to resolve and repeat audiometry with tympanometry. A CHL with a hypermobile TM is suggestive of luxation of the middle ear ossicles. If the CT head with trauma protocol was without evident abnormalities, an additional thin-slice CT scan should be performed to rule out ossicular chain disruptions. If no abnormalities are seen, middle ear inspection could be discussed to check ossicular chain mobility. Alternatively, a patient could start with hearing aids or a bone conduction device. See also Chapter 23.

Bone diseases

Otosclerosis and osteogenesis imperfecta are bone diseases that can cause conductive, mixed, or sensorineural hearing losses. Otosclerosis and stapedotomy are covered more extensively in Chapter 20. Osteogenesis imperfecta is a hereditable disorder affecting collagen type 1. Osteogenesis imperfecta can cause deformities of the middle ear ossicles resulting in fractures of ossicles or fixation of the stapes footplate. Though stapedotomy can be successful (i.e. ABG <10 dB), outcomes are generally less favourable than those seen in stapedotomy for otosclerosis.

Congenital cholesteatoma

Congenital cholesteatoma is a very rare appearance of a well-known entity. It may be undiagnosed for years because of the intact ear drum and can grow undetected until symptoms appear, such as hearing loss, otalgia, or even meningitis or a brain abscess. Congenital cholesteatomas located in the middle ear will present themselves in childhood, but when located in the mastoid it could take decades for the diagnosis to be revealed (**Figure 12.13**).[31,32] In case of (a child with) hearing loss with a white mass behind an intact TM, imaging is needed before proceeding with any intervention.

Differential diagnoses: meningo(encephalo)cele, schwannoma, myringosclerosis, meningioma, or other middle ear tumours.

Treatment: cholesteatomas can only be treated surgically. The location, the extension, and hearing status will direct the surgical approach and technique.

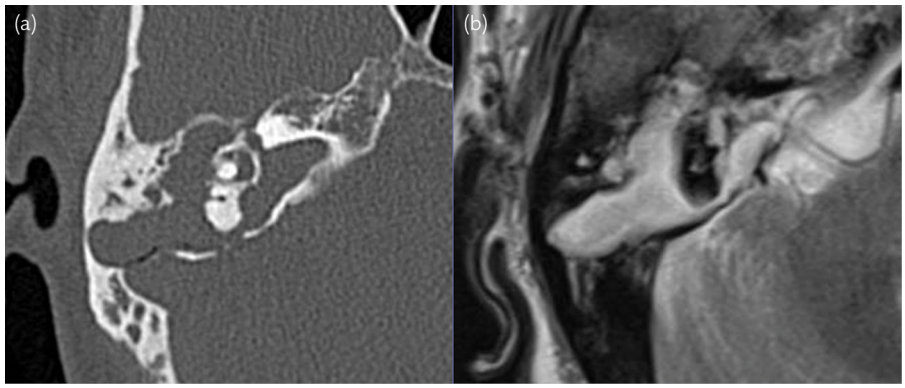

Figure 12.13 Massive cholesteatoma shown on axial CT scan (a) and MRI T2-weighted scan (b). In this case, a congenital cholesteatoma—without preceding otalgia, otitis, or other otological complaints—was only recognised in adulthood as the underlying cause of recurrent episodes of meningitis. Extensive destruction of the mastoid and labyrinth is visible, with involvement of the lateral semicircular canal, and extension into the supralabyrinthine and infralabyrinthine spaces, as well as the petrous apex.

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13

The external ear

Pablo M. Reyes Burneo and Miguel Vaca González

Introduction

The external ear can suffer from a wide spectrum of diseases which have great importance in daily life as a result of the functional and aesthetical effects that they may produce. This chapter seeks to give the reader a comprehensive view of the characteristics and management of these diseases.

Infectious diseases of the external ear as well as tumours are treated in other chapters of this book (see Chapters 8 and 24).

Foreign bodies of the external auditory canal

Foreign bodies of the external auditory canal (EAC) are a common occurrence in both otorhinolaryngology practice and in emergency services; they can represent a challenge for both the patient and the physician. In children, they are usually small objects, placed there in the child's eagerness to explore their body or just as part of usual play (pieces of rubber, beans, beads); in contrast, adults tend to lose objects used to scratch the ear canal (paper clips, toilet paper, nails) and also parts and pieces of hearing aids, especially the adapters to the ear canal (**Figure 13.1**). Moreover, insects can also sneak into the ear canal, producing great discomfort.

As for symptoms, adults usually immediately notice that an object that was being used around the EAC is missing a part of it. In contrast, a foreign body could be asymptomatic and found in a regular examination in children and institutionalized patients; therefore, the possibility of a foreign body of the EAC should always be kept in mind in cases of pain, otorrhoea, and cellulitis in this last group. Hypoacusis, ear fullness, and otitis media-like symptoms could also be present.

All foreign bodies in the EAC must be removed, but they should not be considered under a general strategy since the timing and the choice of technique for removal vary depending on the type of foreign body, ear canal, and patient's condition. Immediate removal must be considered for (1) insects, which can be extremely uncomfortable and also bear the risk of laying eggs and growing larvae in the EAC; (2) batteries, which can cause great tissue destruction in short periods of time when in contact with moisture; and (3) vegetable matter that can grow with humidity and lead to a problematic removal afterwards (**Figure 13.2**). The list of other types of objects can be infinite and the removal time varied.

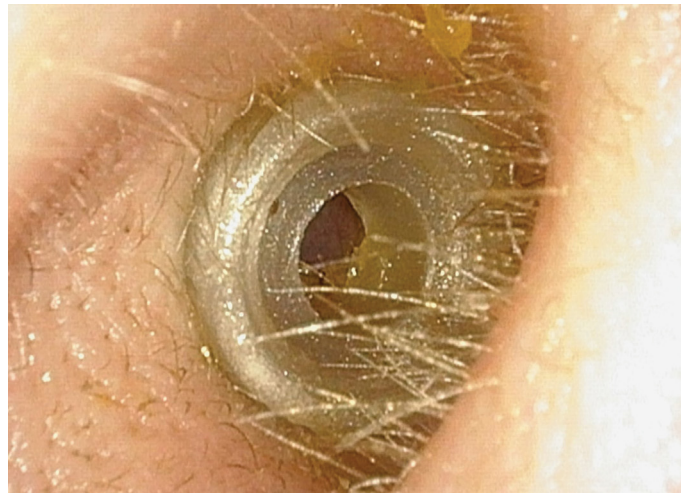

Figure 13.1 Right ear, foreign body of the EAC; the patient was an adult who lost the adapter after taking out his hearing aid.

Removal of a foreign body of the EAC by the otolaryngologist can usually be planed, allowing the gathering of elements such as proper light source, ear forceps, speculums, hooks, endoscopes or microscope, type of anaesthesia, and strategy for removal.

Insects are of special interest due to the painful and unpleasant sensation that they may produce. Before trying to extract a live insect, first it needs to be drowned and immobilized without manipulation; any attempts to extract a live mobile insect may cause it to move towards the tympanic membrane with risk of perforation and lacerations on the skin produced by the insect's exoskeleton. Several liquids have been suggested for this purpose: oil and liquid local anaesthetics such as lidocaine have been proposed, all of them with similar results; other liquids such as alcohol and acetone have been also proposed but they need the important condition of an intact tympanic membrane—if the tympanic membrane cannot be seen, they should not be used.

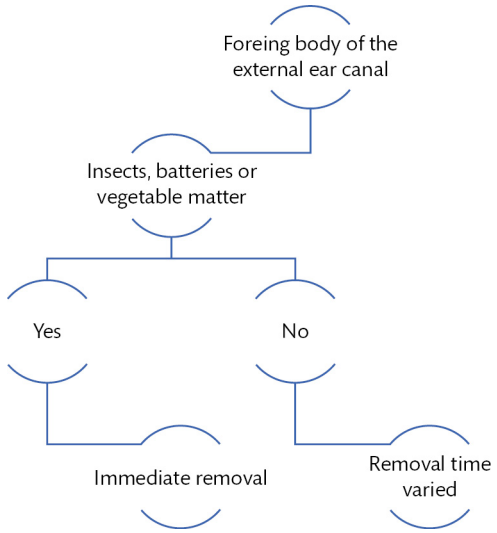

Figure 13.2 Timing for removal of foreign bodies of the EAC.

There are several ways to classify any other object in the ear canal, for practical purposes they can be grouped in two types: (1) soft, graspable, and irregular objects such as paper or cotton; and (2) hard, regular, non-graspable objects such as beads, batteries, or seeds. This classification is useful for planning the proper material and technique for extraction. The first group can usually be easily grasped with an ear forceps or with an aspirator; for the second group, the use of forceps is no recommended due to the risk of moving the object towards the tympanic membrane—an angled hook, a curette, or a suction probe should be tried instead. Ear irrigation/syringing has also been proposed, although it is usually not effective and can carry other complications such as skin maceration; therefore its use as a general strategy is not recommended in the otolaryngology consult where other methods can be used. EAC foreign body extraction can usually be performed in the outpatient setting, although in patients with prior attempts of extraction, anxiety, pain, or irritability, especially in children, it may be better performed in the operation room under sedation or general anaesthesia. Ear drops with steroids alone or combined with antibiotics are not mandatory after extraction, but they should be considered depending on the condition of the EAC (Figure 13.3).

Pearls and pitfalls

- Insects, batteries, and vegetable matter should be extracted immediately.
- Before trying to extract a live insect, first it needs to be drowned and immobilized without manipulation—liquid local anaesthetic or oil can be useful for this purpose.
- Avoid using forceps for round, regular, non-graspable objects since it can move the foreign body towards the tympanic membrane.

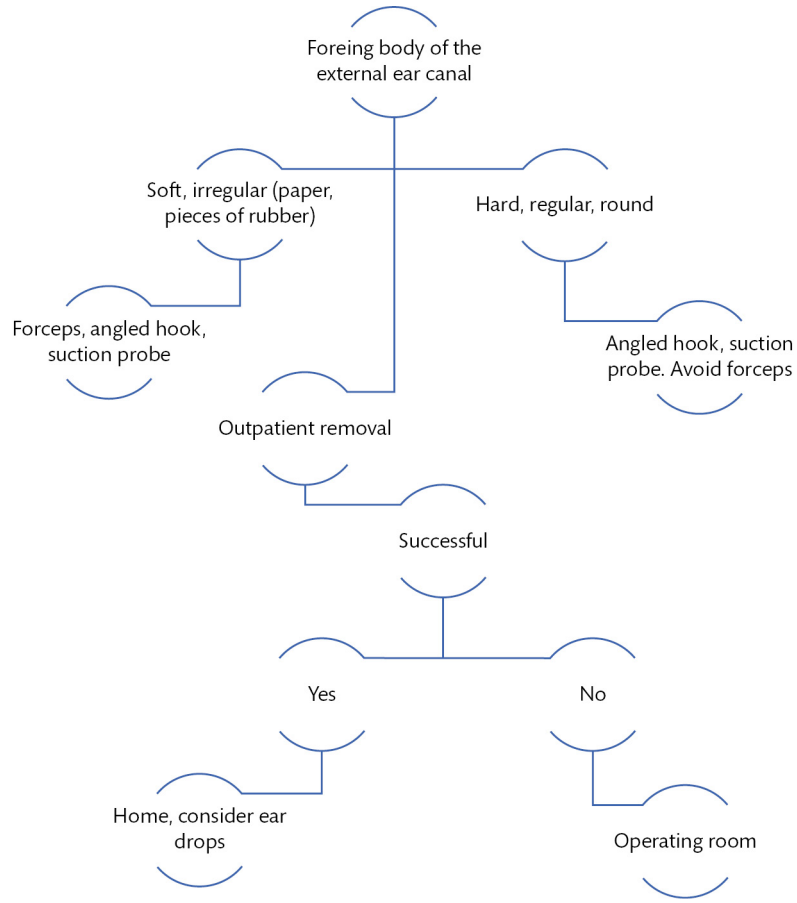

Figure 13.3 Management of foreign bodies of the EAC.

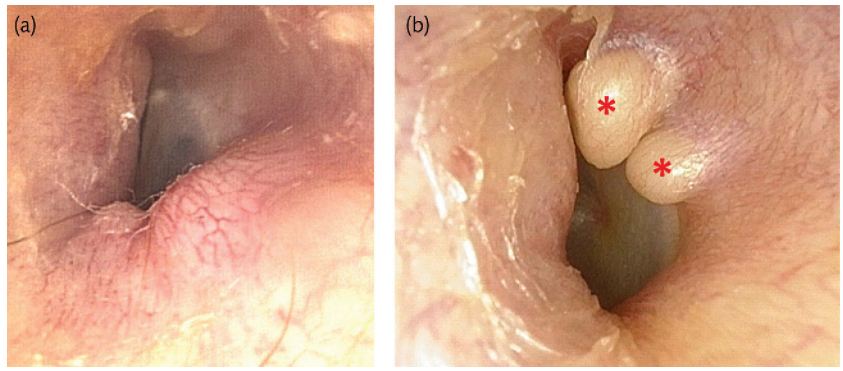

Figure 13.4 (a) Exostoses. (b) Exostoses coexisting with osteoma (red stars).

Exostoses and osteomas of the external auditory canal

Exostoses and osteomas are defined as benign osseous proliferative growths of the bony portion of the EAC. Although exostoses and osteomas differ in their clinical presentation, there is controversy regarding if they should be considered as different histopathological entities.

Exostoses are sessile, broad-based lesions which typically present bilaterally and are located medially in the bony portion of the EAC. It is believed that they have a reactive origin against external aggressions—typically, exposure to cold water. On the other hand, osteomas are pedunculated, isolated lesions, usually arising along the tympanomastoid and tympanosquamous sutures (Figure 13.4).

Treatment

Most patients with exostoses and osteomas remain asymptomatic; nevertheless, both types of lesion can become large enough to induce symptoms such as wax retention, recurrent otitis externa, conductive hearing loss, skin debris retention with risk of EAC cholesteatoma, and tinnitus secondary to the blockage caused by the bony obstruction. Asymptomatic patients do not require any particular treatment, besides the knowledge that they have this bony lesion in their EAC. Treatment for symptomatic patients ranges from in-clinic aspiration and cleaning of the EAC with intermittent use of ear drops to definite treatment which requires surgical removal of the exostosis or osteoma.

As for surgical treatment, chiselling and/or drilling have been proposed as the two main techniques to remove these lesions by a transcanal, endaural, or retroauricular approach. Despite the predominance of the modern drilling techniques, chiselling can still be useful in cases of small pedunculated lesions which can be safely removed by this approach under local anaesthesia and usually without requiring a retroauricular approach.

Drilling techniques are better performed by a retroauricular incision which allows proper exposure of the bony ear canal, as well as meticulous repositioning of the flaps made in meatal skin.

Surgery involves the following steps:

1. A retroauricular incision is made where the mucoperiosteal flap is detached from the bone at the level of the EAC.
2. The posterior meatal skin is partially detached from the meatus.
3. The posterior meatal skin is incised, revealing the bony portion of the EAC with the exostosis and/or osteoma medial to the incision performed.
4. Circular incisions are made lateral to the bony structures that need to be removed, radial incisions are made to achieve proper management of the skin (Figure 13.5a).
5. The meatal skin that covers the lesion is pushed towards the tympanic membrane with the help of a cottonoid and blunt dissection (Figure 13.5b).
6. Drilling of the bony overgrowth is performed, usually starting by the posterior wall of the EAC (Figure 13.5c).
7. After the approached lesion has been drilled, the medial meatal skin that used to cover the bony growth can be put back in place (Figure 13.5d); the same procedure is applied to the rest of the walls if needed. A piece of aluminium can be of tremendous help in protecting both the meatal skin and the tympanic membrane.

Pearls and pitfalls

- A piece of aluminium can be helpful in protecting the meatal skin and the tympanic membrane when performing drilling manoeuvres; it can be obtained from the aluminium wrap of the sutures that are going to be used in closure, and can be tailored to adapt to the EAC (Figure 13.6).

Cerumen impaction

Cerumen or ear wax is a natural substance which is a combination of the secretion of the ceruminous glands in the cartilaginous portion of the EAC mixed with epithelial cells gathered by desquamation. It has the function and potential of cleaning, lubricating, and protecting the EAC (Figure 13.7). Cerumen is expelled from the EAC spontaneously via a self-cleaning mechanism that is assisted by jaw movement.

Cerumen is a harmless substance but if self-cleaning mechanisms fail, it can become impacted, causing blockage of the EAC which can lead to a variety of conditions ranging from asymptomatic to hearing loss, odour, itching, fullness, and pain. It can also prevent a proper assessment of the EAC and tympanic membrane or interfere with audiovestibular testing and evaluation (i.e. tympanometry, audiometry, and caloric tests). Cerumen is also more likely to

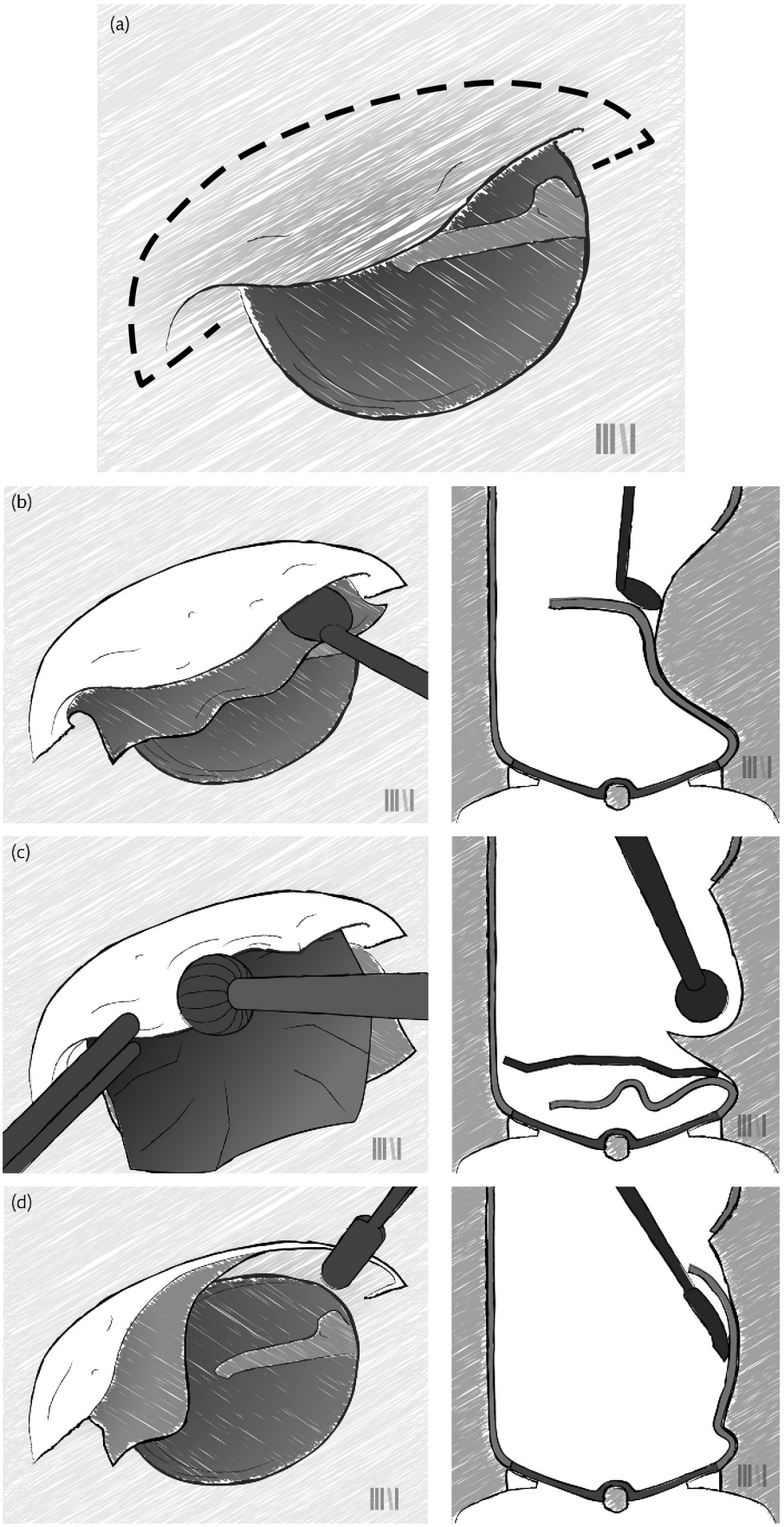

Figure 13.5 (a) Circular incisions are made lateral to the bony structures that need to be removed, radial incisions are made to achieve proper management of the skin. (b) The meatal skin that covers the exostoses is pushed towards the tympanic membrane with the help of a cottonoid and blunt

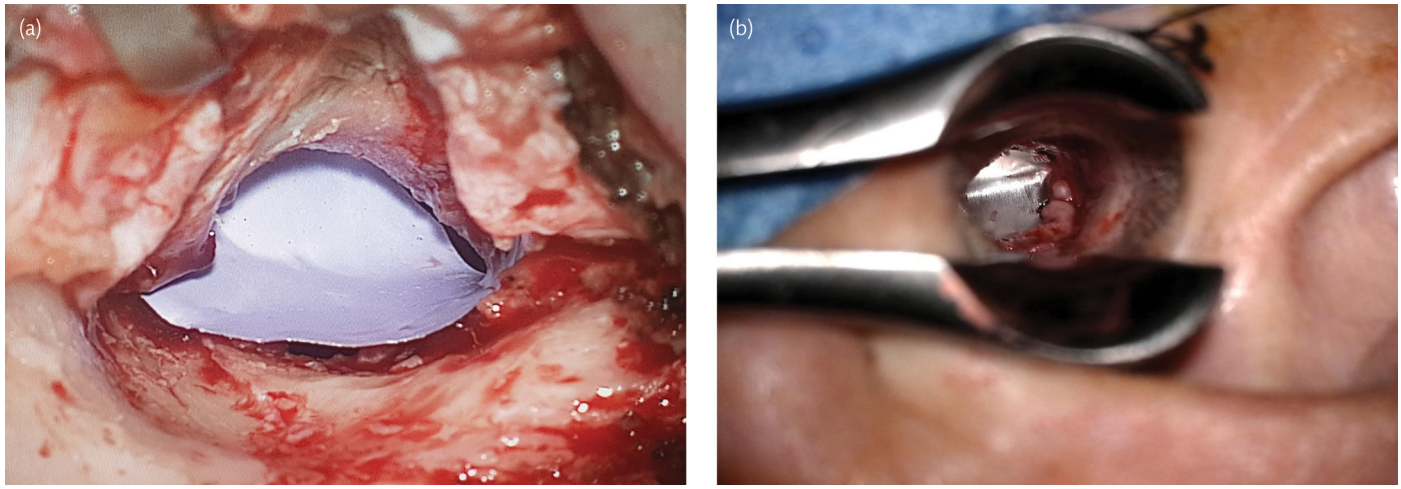

Figure 13.6 A piece of aluminium protecting the meatal skin and the tympanic membrane. (a) Retroauricular approach. (b) Right transcanal approach.

become impacted when self-cleansing mechanisms are prevented by the use of hearing aids, ear plugs, or cotton aids to clean the EAC.

Cerumen accumulation and related symptoms is one of the most common reasons for seeking medical attention either in primary care or from an otolaryngologist.

Diagnosis is made by correlation of the symptoms described by the patient with direct visualization of the EAC with an otoscope, which will show a brown or dark substance blocking the ear canal.

Treatment

A fundamental part of treatment is assessing patients' habits, with great attention paid to the use of cotton aids or buds and explaining its relationship to the ear wax impaction.

Several treatment options have been proposed, such as the following:

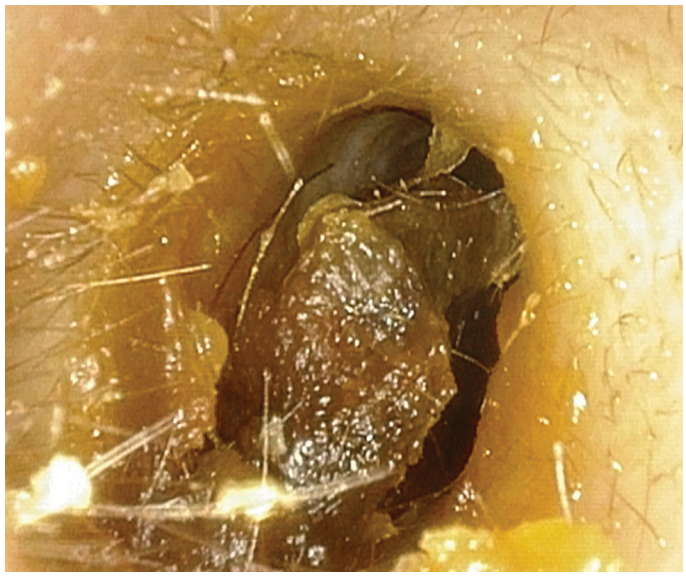

Figure 13.7 Right ear; ear wax partially occluding the EAC. The patient was referred with a scratchy noise over his right ear, symptoms were relieved after removal.

- *Cerumenolytic ear drops*: various types of oil- and water-based cerumenolytics are commercially available. None of them has proven to have a better effect on cerumen dissolution; they usually require at least 5 days of three-times-a-day treatment to become effective.
- *Ear irrigation*: although ear irrigation can be performed without preparation, some providers prefer prior preparation with any of the available cerumenolytic agents for 5–7 days before the procedure; instillation just prior to irrigation for 15–30 minutes has also been described.
  - As for technique, the patient has to sit upright, and a good-capacity syringe (at least 20 mL), or ear irrigation pump, should be filled with warm water (cold water can produce dizziness). Draw up the syringe to the ear canal no further than the bone/cartilage junction. A collecting device such as a basin should be tightly held to the skin below the ear. Direct the syringe superior and posteriorly to allow separation of cerumen from the tympanic membrane. Care must be taken not to direct the water flow towards to the tympanic membrane as this can lead to complications such as perforation of the tympanic membrane.
  - Ear irrigation is considered a safe procedure, nonetheless it may lead to complications such as vertigo, otitis externa, tympanic membrane perforation, and hearing loss. Ear irrigation must not be performed in patients with tympanic membrane perforation and should also be avoided in the only hearing ear of patients with single-sided deafness due to the risk of hearing loss. Because of its potential risks, irrigation is now less used in the outpatient setting, and has been replaced by other techniques such as aspiration or mechanical extraction.
- *Manual removal with instrumentation*: this can be performed under direct visualization with a head light, otoscope, or microscope by using a curette, hook, or suction probe.

After treatment, clinicians should assess the resolution of impaction by direct visualization; if persisting impaction is detected, a different technique or referral should be considered. In addition, resolution of symptoms must be addressed; if partial or full symptoms persist, differential diagnoses should be considered.

Pearls and pitfalls

- Under proper visualization, manual removal with instrumentation can usually solve cerumen impaction in the outpatient setting and could resolve impaction on the first visit.
- Preparation with cerumenolytics 5–7 days before attempting extraction can facilitate both aspiration and irrigation techniques.
- If using irrigation, direct the syringe superior and posteriorly to allow separation of cerumen from the tympanic membrane.
- Ear irrigation must not be performed in patients with tympanic membrane perforation and should also be avoided in the only hearing ear of patients with single-sided deafness due to the risk of hearing loss.

Malformations of the external ear

There is a great variety of congenital malformations that affect the elements of the external ear. From a practical point of view, these can be divided into malformations of the auricle and congenital aural atresia. Both are frequently associated, and a certain correlation between the external appearance of the auricle and the size of the ear canal is usually observed.

Congenital malformations of the external ear are relatively frequent in ear, nose, and throat practice. The incidence of minor malformations of the pinna is described as 2–3/10,000 newborns, and severe malformations/atresia as 1/20,000 newborns. In around 85–90% of cases the malformation is unilateral, and the right ear is more frequently affected than the left ear.

Basic science: embryology

External ear malformations can occur as an isolated disease, or frequently appear in association with other middle or inner ear malformations, even though their embryological origins are different. This can be partially explained by the fact that the development of all the elements of the auditory system overlap during embryogenesis, and so any disorder could potentially affect any of them.

The primitive auricle originates from the fusion of six mesenchymal hillocks that originate from the first and second branchial arches, known as the hillocks of His. This fusion takes place at the end of the second month of gestation, and any failure during the development of these hillocks or their fusion can result in a malformation of the pinna.

The external auditory meatus originates from the first branchial groove. At the eighth week of gestation, a proliferation of ectodermal cells at the dorsal part of the groove starts to expand medially forming the meatal plate. Around the 21st week of gestation this plate starts to resorb and hollow, thus forming the external meatus, ear canal, and the superficial layer of the tympanic membrane. Any failure at the resorption will result in different degrees of EAC atresia.

Assessment and classification

Malformations of the auricle are easily addressed by simple inspection. The area around the pinna should also be palpated to discover any cartilaginous remnants under the skin, and to estimate the size of the mastoid. The term microtia is used to describe malformations of the auricle, as they usually mean a reduction on its size. According to Weerda, there are three grades of microtia that consider the severity of the malformation and the characteristics of the auricular remnant. Grade I is a virtually normal auricle, with all its features present, but reduced in size. Grade II is a rudimentary auricle, but some elements can still be recognized. In grade III microtia, there are not any recognizable elements, but only some protuberances of tissue with occasional remnants of cartilage under the skin. Because of its resemblance, grade III microtia is also known as 'peanut ear'. In the most severe cases the term anotia is often used to describe the complete absence of the ear (Figure 13.8).

If there is associated atresia of the ear canal, the size of the EAC is evaluated with otoscopy. It can be classified accordingly in total or partial atresia. In the latter, the tympanic membrane status and the malleus handle appearance should be assessed if

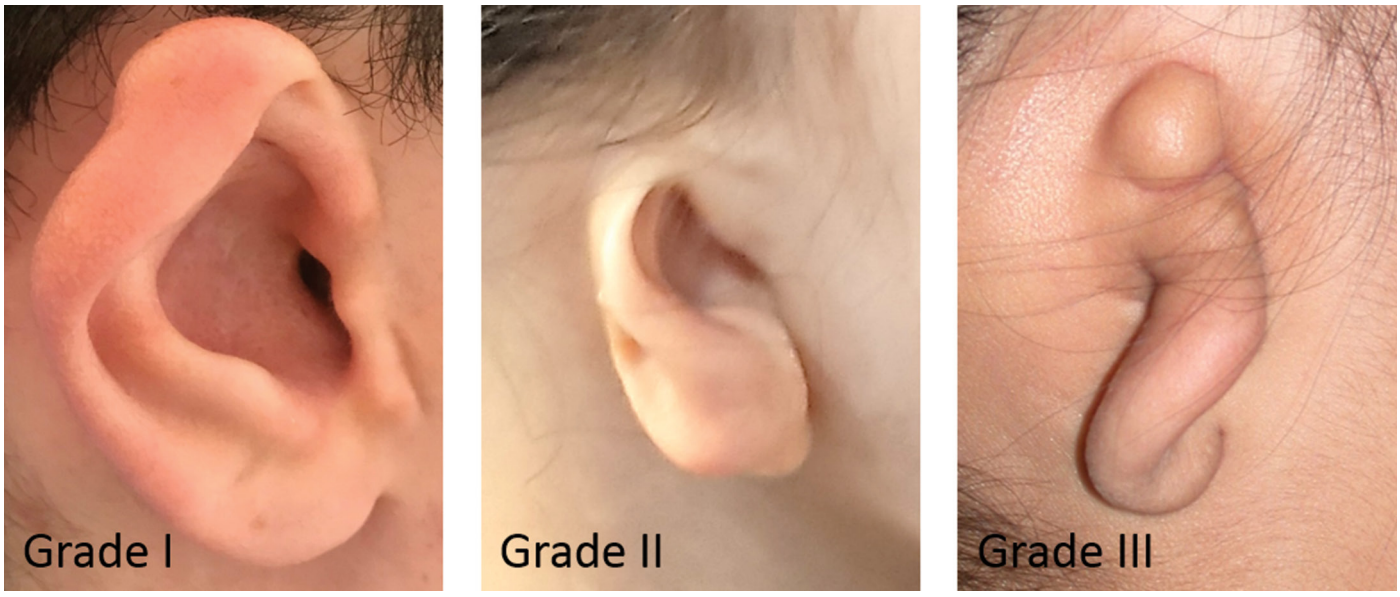

Figure 13.8 Classification of microtia.

Table 13.1 Jahrsdoerfer grading system for the selection of patients with congenital aural atresia

Parameter	Points
Stapes present	2
Oval window open	1
Middle ear space	1
Facial nerve	1
Malleus–incus complex	1
Mastoid pneumatization	1
Incus–stapes connection	1
Round window	1
Appearance of external ear	1
Total score	10

possible, because they may be abnormal in the context of an associated middle ear malformation. It is also important to rule out a potential cholesteatoma as a consequence of the atretic ear canal. In severe cases or total atresia, otoscopy does not yield such information, and these considerations have to be assessed by radiological examinations.

A computed tomography (CT) scan is paramount in the evaluation of external ear atresia, especially when surgery is considered. Nevertheless, this examination can be postponed until the age of 5 years or later if no other symptoms are present. With a practical perspective to address surgical candidacy, Jahrsdoerfer et al. developed a grading system based on preoperative CT scanning (Table 13.1). Along with the EAC, middle ear and inner ear structures should be carefully evaluated in both axial and coronal planes. It is also important to assess the facial nerve position and its relationship with other structures.

Apart from these morphological aspects, the key point to be addressed is the impact of the malformation on hearing levels. In this context, the inner ear is rarely affected, and so the external ear malformation causes conductive hearing loss to different degrees according to its severity. An isolated malformation of the auricle is not likely to affect air conduction significantly. On the other hand, atresia usually means a moderate to severe conductive hearing loss which will definitely have a clinical impact.

In small children, audiological diagnosis can represent a challenge, and auditory brainstem response tests or behavioural audiometry have to be conducted according to the age of the patient. In unilateral cases, hearing thresholds of the contralateral ear have to be investigated as early as possible to rule out sensorineural hearing loss that may affect normal development of language. If the malformation is bilateral, bone conduction auditory brainstem response can be performed in newborns to establish hearing levels, and subsequently prepare a treatment plan that guarantees an adequate development of language.

Close monitoring of the hearing levels and language acquisition throughout the first years of life should be carried in these patients until surgical correction is considered, usually by the age of 5–6 years.

Differential diagnosis/associated conditions

As external ear malformations are usually evident from the very early moments of life, the diagnosis is quite clear. However, mild cases with a normal pinna and incomplete atresia may be difficult to distinguish from acquired stenosis of the canal due to exostosis. Therefore, when approaching a child with an exostosis, careful examination and a CT scan are important to rule out a potential malformation with all its concerns.

In around 10% of the cases, external ear malformations are part of more complex syndromes such as Treacher Collins, Crouzon, Goldenhar, or CHARGE association. Each of them has its own clinical profile and specific considerations, and so a brief general anamnesis and physical examination are recommended to rule out other congenital abnormalities in the context of these syndromes.

Treatment

The objectives of the treatment of external ear malformations are to get a cosmetically acceptable pinna and, more importantly, to re-establish normal hearing when hearing loss is associated.

In unilateral malformations, the normal contralateral side guarantees language development and therapeutical decisions can be postponed until the child reaches the appropriate age. Until that moment, bone conduction devices with soft or hard bands can provide temporary stimulation of the malformed side, resulting in binaural hearing with its associated benefits. In bilateral malformations, language acquisition is of the greatest concern. Early stimulation with bone conduction hearing aids with soft or hard bands will ensure language development until the child is old enough to undergo a definitive treatment.

Treatment of microtia

Auricular reconstruction for severe microtia and anotia is usually recommended at 6–8 years of age. This helps to diminish the risks associated with the cartilage graft harvesting, and in unilateral cases allows the normal ear to grow to nearly its adult size and act as a template. Also, the child can express their concerns related to the deformity, and take part in the decision-making. The management options and the balance between risks and benefits should be discussed with both the patient and their parents.

The status of the auricular remnant and its cartilage has to be carefully assessed to establish a surgical plan. For mild cases of microtia (grade I), no additional tissue is required to perform the otoplasty, and the deformity can be approached by reshaping the existing cartilage frame or skin. Severe cases of microtia (grades II and III) require autologous cartilage grafts, usually from the rib, or synthetic materials.

Autologous grafts are well tolerated and more resistant to infection, but lead to worse cosmetic results than synthetic materials. However, synthetic materials (porous polyethylene or silicone) have an intrinsic risk of developing a foreign body reaction and extrusion over time. In any case, these reconstruction procedures are complex and involve multiple stages. In the first procedure, the auricular framework, whether autologous or synthetic, is implanted under the skin. In subsequent staged interventions, the retroauricular sulcus and the rest of the elements of the pinna are created accordingly to

Figure 13.9 Multistaged auricular reconstruction. In the first stage, a large piece of cartilage is harvested from the rib (a), and the auricular framework is carved to resemble the normal landmarks of the pinna (b). This framework is placed under the skin (c). In a second stage, retroauricular sulcus and tragus are created using the auricular remnant (d).
Pictures courtesy of Dr. P. Benito.

configure a normal-looking auricle (Figure 13.9). Complications of these procedures, apart from those related to the donor site, include skin necrosis and extrusion of the graft, or malpositioning of the reconstructed pinna, which may require additional surgical interventions.

Auricular prostheses remain as an option for those severe cases that are not good candidates or reject surgical reconstruction. Modern prostheses can be retained with an adhesive or with an osseointegrated implant, and can be modelled to resemble the contralateral ear offering an excellent aesthetical appearance with minimum risks (Figure 13.10).

Figure 13.10 Auricular prosthesis.

Treatment of congenital atresia

Surgery for external ear atresia should be considered for children older than 5 years. This allows for complete development and pneumatization of the mastoid process. In cases with associated auricular malformations, auricular reconstruction with autologous cartilage should be performed first to benefit from an unaltered vascular supply. If an artificial porous polyethylene framework is chosen, then atresia surgery can be performed in first place.

In this context, the surgical goals are to get a dry and stable EAC with an improvement in air conduction hearing levels. In general, surgical success and risk of complications are related to the degree of malformation of the middle ear elements and the mastoid pneumatization. Careful preoperative CT scan evaluation and use of the Jahrsdoerfer grading scale (Table 13.1) help in candidacy

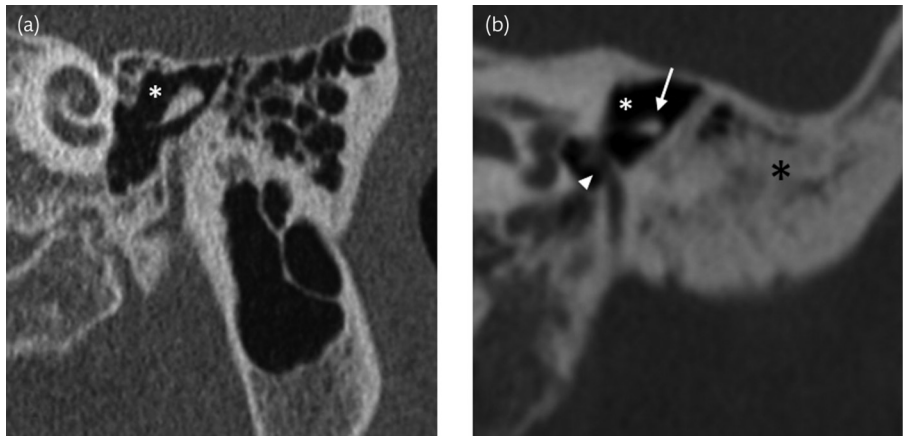

Figure 13.11 Coronal CT scan images of a good surgical candidate (a), versus a poor surgical candidate (b). Though both have good middle ear space (white asterisk), notice the absence of malleus–incus complex (arrow), lack of mastoid pneumatization (black asterisk), and the facial nerve over the oval fossa (arrowhead) in (b). Considering the rest of the items, the total Jahrsdoerfer score in case (a) was 8, versus 4 in case (b).

selection. Patients with a score over 7 are usually considered good surgical candidates. This pictures a patient with adequate mastoid development, and normal relationship between facial nerve and oval window/stapes. On the other hand, patients with scores below 5 are poor surgical candidates, and the risks clearly discourage the reconstructive option (Figure 13.11). Decisions in patients with scores between 5 and 7 should be individualized, considering the alternatives and the results and experience of the surgeon. In bilateral cases, the worst ear should be operated first.

When the atresia is partial and the ear canal is patent, then a bony canaloplasty and conchal meatoplasty can be performed. In these cases, the ossicular chain and the tympanic membrane might be affected by the malformation, and so should be reconstructed during the same operation, or in a staged procedure. Care has to be taken to preserve as much ear canal skin as possible; however, skin grafts are usually needed to cover all the exposed bone and prevent restenosis.

The operation to reconstruct the EAC in complete atresia is performed under general anaesthesia and facial nerve monitoring. The position of the external meatus is determined by palpation of the mastoid tip and the temporomandibular joint, next to the auricle. A C-shaped postauricular incision is made with the aim of preserving the vascular supply to the pinna. Subcutaneous tissue and periosteum are elevated anteriorly to expose the mastoid and the glenoid fossa. Sometimes a depression in the remnant of the tympanic bone can be identified, which serves as a superficial landmark for the drilling. If no superficial landmarks are identified, then the drilling should start in the area limited superiorly by the middle cranial fossa, and anteriorly by the temporomandibular joint (Figure 13.12). After the identification of both structures, which will eventually serve as superior and anterior canal walls, dissection proceeds medially through the tympanic plate up to epitympanum. The diameter of the new canal should be around 12–15 mm, and extensive opening of mastoid cells is to be avoided as far as possible.

As surgical landmarks in this context are scarce and the facial nerve may follow an aberrant course, utmost care should be taken in the creation of the new canal, especially at the inferior and posterior areas. Also, when approaching the theoretical position of the ossicular chain, low-speed drilling or curetting are advised to avoid trauma to the ossicles. Finally, the bony surface of the newly created ear canal has to be smoothed with diamond burs, and any mastoid cells opened to this surface must be plugged with soft tissue to prevent iatrogenic cholesteatoma.

Considering the ossicular chain, it may be normal or frequently associated with any degree of malformation. If that is the case, ossicular reconstruction can be performed under the same principles as for chronic otitis media. The malleus–incus complex may be fixed to the atretic plate by fibrous tissue or an osseous bar, which can be eliminated to liberate it. Reconstruction to a mobile stapes or to the footplate can be performed with a partial or total ossicular replacement prosthesis, or with cartilage or bone autografts.

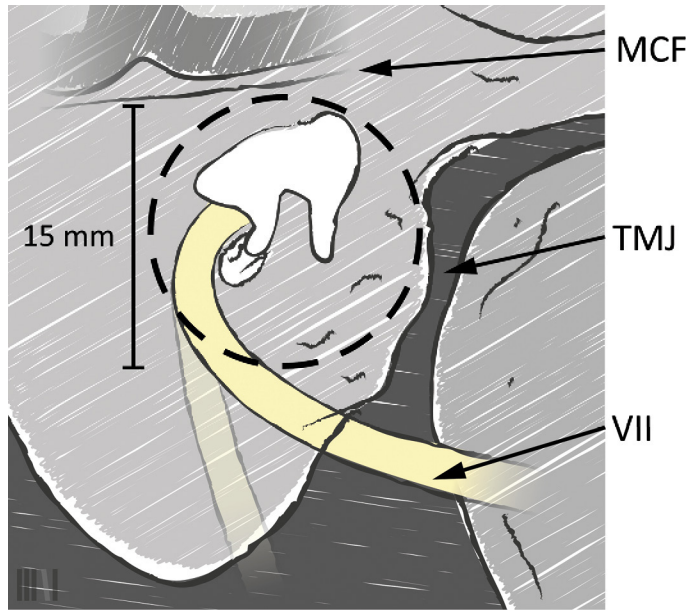

Figure 13.12 References for the drilling of the EAC. It is limited superiorly by the middle cranial fossa (MCF) and anteriorly by the temporomandibular joint (TMJ). Note the facial nerve trajectory (VII), in contrast to its usual disposition.

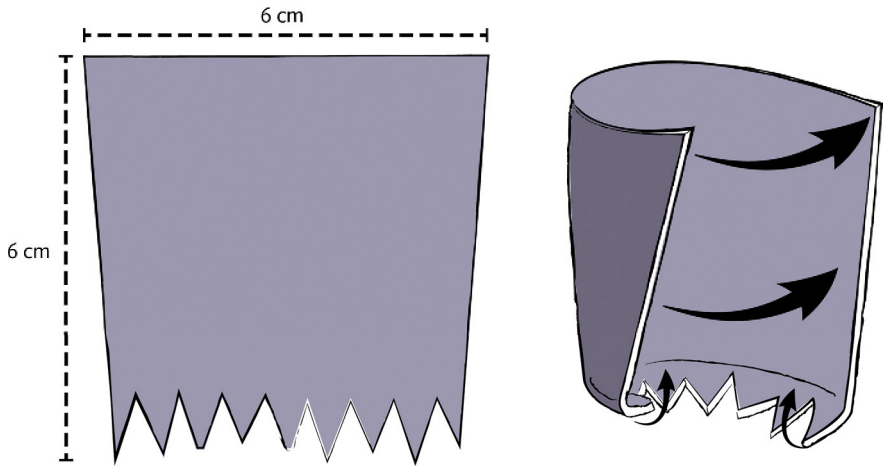

Figure 13.13 Template for the split-thickness skin graft. It is meant to cover completely the bony canal, and its trimmed edges are folded upwards to rest over the fascial graft.

Tympanic membrane reconstruction starts with the drilling of a bony rim in the medial end of the ear canal, where previously harvested temporalis muscle fascia will rest. This prevents blunting of the membrane angles. The dried fascial graft is then placed over the ossicular chain with its edges supported by the bony rim. To prevent lateralization of the membrane, two cuts are made in the anterior edge of the fascial graft to create a small strip that will be placed medial to the bony rim in the protympanum or hipotympanum.

The bony surface of the ear canal has to be completely covered with skin. A 0.2–0.3 mm thick, 6 × 6 cm wide split-thickness skin graft is harvested with a dermatome (**Figure 13.13**). Typical donor sites include hypogastrium or the medial surface of the upper part of the arm. To adapt the skin graft to the ear canal it should have a slight trapezoidal shape, as the external meatus is usually wider than its medial part. Also, the medial edge is cut in a zigzag fashion, which is meant to be folded over the fascial graft. To trim the skin graft accurately, the length of the ear canal and its circumference can be measured using suture threads or a sterile paper as a template.

The new external meatus is created in the pinna, in a position that overlies the lateral part of the canal. The skin and soft tissues are excised, taking care not to expose the cartilage or the material of the previous auricular reconstruction unless it is strictly necessary. A 15 mm wide opening is performed to compensate the effects of postoperative retraction.

The skin graft is carefully positioned on the canal and over the fascia. It is important to verify that all the bone is covered, and that no skin is folded on itself. To hold the graft in place, the canal is covered with Silastic strips, and the tympanic membrane with a Silastic disc. This helps graft integration and prevents blunting and lateralization of the membrane. The lateral edge of the graft is finally sutured to the meatal area with 5-0 absorbable sutures, and the canal is packed with a Merocel wick (**Figure 13.14**).

The packing and the Silastic strips will be removed after 2 weeks. Then, the patient should be seen every 2 weeks for cleaning and debridement of any granulation tissue that may appear. Antibiotic ear drops with steroids are advised in this period. At 2 months, the healing process is usually complete and the first audiogram can be performed.

Complications of atresia surgery are rare if there has been adequate candidate selection. Facial nerve injury is the most fearsome complication that can be prevented with careful dissection and monitoring. High-speed drilling in the vicinity of the ossicular chain can result in sensorineural hearing loss, which may appear in up to 7% of cases. Complications related to the patency of the ear canal are the most frequent. Lateralization and blunting of the tympanic membrane can occur in up to 15% of cases, even at 1 year after surgery. Meatal or canal restenosis by soft tissue is related to uncovered bone or granulation tissue. In the early postoperative period these can be managed by aggressive debridement and topical steroids. In

Figure 13.14 Final disposition of the grafts. The fascial graft (green) is placed over the ossicular chain and the bony rim. The skin graft (purple) is placed over the fascia and the bony walls. Laterally, it is sutured to the pinna.

the late postoperative period, it may require local steroids and dilatation to avoid progression and the need for surgical revision. Bony restenosis may also happen in patients with new bone growth. This is typical of puberty and can compromise the patency of the canal or hearing, often requiring a revision procedure.

The results of surgery for canal atresia usually depend on the severity of the malformation. Air conduction levels below 30 dB are achieved in approximately two-thirds of patients. In those cases, with suboptimal audiological results but good ear canals, conventional hearing aids are advised. When surgery does not achieve any of its goals, neither audiological nor to get a patent ear canal, auditory implants remain as an alternative.

Middle ear implants constitute a therapeutic option for selected cases of aural atresia. They provide excellent audiological outcomes and enable optimal sound localization. However, their implantation depends on favorable anatomical conditions, which often restricts their applicability in this context.

Bone conduction devices play a key role in the current management of atresia. Their rationale is to provide direct stimulation to the inner ear through the skull, therefore bypassing the external and middle ear. Patients with external ear malformations usually keep a good cochlear reserve, so the results with these devices are optimal with minimum risks. Their indication is obvious in those bilateral cases which are not candidates for atresia surgery (Jahrsdoerfer score ≤5). In unilateral cases, or candidates for reconstructive procedures, they remain as an option that should be discussed with the patient and the family. For more details, see Chapter 16.

Currently, there are several bone conduction devices, each one with its own range of audiological indications, recommendations, and surgical techniques. As a general rule, they require an intervention to place an implant on the bone, which can be performed under local or general anaesthesia. Though simple, this operation can interfere with local vascularization. Therefore, if surgery of the pinna is considered, the device should be placed in an alternative location using a technique that preserves local vascularization, or postponed until the reconstructive procedure has been completed. In any case, the minimum age for the implantation is 5 years old.

Pearls and pitfalls

- External ear malformations represent a challenging clinical scenario. Both functional and aesthetical concerns must be addressed by a multidisciplinary team that includes audiologists, otologists, and reconstructive surgeons.
- Audiological rehabilitation from the early days of life with bone conduction hearing aids is paramount in bilateral cases. Reconstructive procedures offer reasonably good results with adequate candidate selection. However, they often involve multistaged procedures with associated risks. Bone conduction implants offer a good alternative for hearing rehabilitation with excellent results.
- An individualized treatment plan that deals with all these considerations has to be created for each case. It should include both the family's and the child's expectations. To provide recommendations and help in the decision-making, the International Microtia and Atresia Workgroup published in 2019 a guideline entitled 'International Consensus Recommendations on Microtia, Aural Atresia and Functional Ear Reconstruction'.

Acquired atresia of the external auditory canal

Acquired non-osseous atresia of the external ear is an uncommon condition in which the ear canal is obliterated by fibrous tissue. This can be a consequence of trauma, burns, dermatological conditions, or chronic infections. All these causes lead to chronic myringitis and granulomatous inflammation of the EAC, which eventually results in fibrous tissue formation and a progressive stenosis of the canal. It also can occur as a complication of ear surgery. In this context, it is usually related to inadequate surgical technique with extensive removal of the ear canal skin.

The atresia develops over the course of several months. In the first inflammatory phase, discharge and hearing loss are the main symptoms. Once the atresia has completely developed, moderate conductive hearing loss is the main concern. CT scanning is important for the evaluation of these patients as an external ear cholesteatoma may develop medial to the fibrous plug.

The treatment for atresia depends on the clinical phase. In the early stages, control of the underlying infection and inflammatory process are sought to prevent scar tissue formation. Antibiotic drops with steroids and local debridement are useful at this time. If a dermatological condition underlies the atresia formation, then its specific treatment may help to prevent it. When the atresia is complete, surgical treatment remains the only curative option.

The objective of acquired atresia surgery is to provide a patent ear canal, thus improving air conduction thresholds. The results of the operation are worse in cases of active inflammation or infection; therefore, it should be delayed until the appropriate moment.

Surgery is performed through a retroauricular approach. The ear canal skin and the fibrous plug are gently elevated and separated from the posterior bony wall, and the tympanic membrane fibrous layer is identified and preserved. Then, the fibrotic plug and the pathological skin are excised and the bony canal is enlarged with burrs. A split-thickness skin graft is required to cover all the exposed bone. Packing of the ear canal and postoperative management are similar to those described for congenital atresia surgery.

The main concern in acquired atresia surgery is the 10–20% risk of restenosis. This is minimized with the use of split-thickness grafts and meticulous care after surgery. Chronic ear discharge is another complication that can be prevented with adequate covering of the bone with skin, and postoperative debridement of any granulomatous tissue.

Hearing usually improves if the canal remains patent. If the patient is not satisfied with their hearing results, or mild restenosis occurs, conventional hearing aids can be an option. Bone conduction hearing devices are an alternative for severe restenosis, or for cases where reconstructive surgery is not recommended.

Keratosis obturans and external ear cholesteatoma

Keratosis obturans is a rare entity in which lateral migration of the external ear epithelium is disrupted. A plug of keratin builds up due to the accumulation of normal desquamation of the superficial layer

Table 13.2 Naim's classification of histopathological findings in external auditory canal cholesteatoma

Stage I	Hyperplasia of the epithelium with increased apoptotic rate
Stage II	Localized inflammation and periostitis
Stage III	Bone erosion, aseptic osteonecrosis
Stage IV	Destruction of the adjacent structures (mastoid, temporomandibular joint, skull base, facial nerve)

of the ear skin, which can form a true cast of the ear canal and reach the tympanic membrane. It is usually bilateral, and has been associated with other conditions such as bronchiectasis or sinusitis.

Conductive hearing loss due to the obstruction of the canal is frequently the only symptom in keratosis obturans; moreover, otorrhoea is rare, in contrast to ear canal cholesteatoma. In the long term, constant pressure of the plug on the skin and subjacent bone walls can result in a circumferential widening of the canal that can be observed in temporal bone CT scanning. Conservative management with emollients and periodical removal of keratotic plugs is an appropriate treatment for most patients. However, canaloplasty may be required for intractable cases.

External ear cholesteatoma is characterized by a local accumulation of keratinized epithelium that leads to focal bone erosion and osteitis. This entity can be idiopathic, or more frequently appear as a consequence of congenital atresia, temporal bone trauma, or ear surgery, or as a complication of any inflammatory condition of the external ear. Normal migration and desquamation of the epithelium are blocked by the predisposing condition, which eventually causes the keratin cyst to grow and erode the underlying bone (Table 13.2).

Most of the patients with external ear cholesteatomas have otorrhoea and/or conductive hearing loss as their main symptoms. Dizziness, sensorineural hearing loss, or even facial paralysis may occur with the progression of the disease.

The initial otoscopic appearance of external ear cholesteatoma is a unilateral non-obstructive keratin pearl in the floor of the canal. This contrasts with the generally obstructive and bilateral keratosis obturans. With progression of the disease, the pearl grows and exteriorizes, revealing the matrix over the eroded tympanic bone. Local infection and granulation tissue are frequent at this stage. Therefore, necrotizing external otitis or carcinoma must be ruled out. Biopsy of the margins of the erosion is recommended, and the relatively rare (20–30%) and mild otalgia in external ear cholesteatoma helps to distinguish it from malignant external otitis.

High-resolution CT scanning is important to assess the extension of the bony erosion and the involvement of adjacent structures. Idiopathic external ear cholesteatomas tend to be located in the floor of the canal, whereas secondary cholesteatomas tend to appear in atypical locations or even may be multifocal (Figure 13.15).

In early-stage cholesteatomas with good control of the limits, conservative management with periodical aural toileting and debridement is a feasible option. Nevertheless, surgery is the only definitive treatment that can prevent progression, and so it is advised for symptomatic patients, erosive cholesteatomas, and complicated cases. The technique of choice for most cases is bone canaloplasty with removal of the devitalized skin and bone up to healthy tissue. Smoothing of the surface with diamond burrs, obliteration of the defect, and the use of skin grafts to completely cover the exposed bone are advised to prevent failures. Mastoidectomy may be required for complicated cases with the involvement of the mastoid or other structures.

Pearls and pitfalls

- Conductive hearing loss due to obstruction of the ear canal is frequently the only symptom in keratosis obturans; moreover, otorrhoea is rare, in contrast to ear canal cholesteatoma where otorrhoea is quite usual.

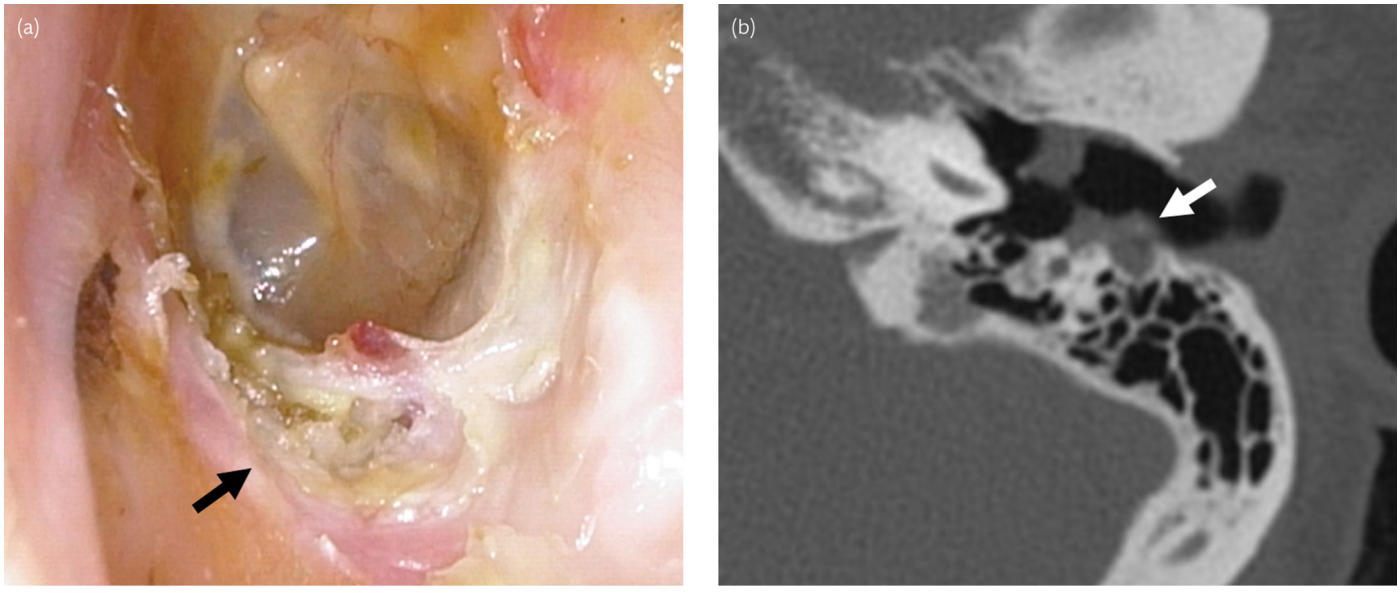

Figure 13.15 External ear cholesteatoma: (a) otoscopy and (b) axial CT scan. Note the focally eroded tympanic bone with cholesteatoma matrix and keratin.

- The initial otoscopic appearance of external ear cholesteatoma is a unilateral non-obstructive keratin pearl in the floor of the canal. This contrasts with the generally obstructive and bilateral keratosis obturans.

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14

Ménière's disease and vestibular migraine

Louisa Murdin

Introduction

Ménière's disease (MD) and vestibular migraine (VM) together form a significant proportion of the caseload of episodic vertigo likely to be encountered in the ear, nose, and throat (ENT) clinic. VM is one of the commonest causes of episodic non-positional vertigo likely to be encountered in emergency medicine and in community settings, and thus also forms a high proportion of referrals to ENT clinics from those sources.

Diagnosis

Both MD and VM are defined by a set of characteristic clinical features rather than by any objective diagnostic objective test. The cardinal feature in both conditions is the proneness to episodic vestibular symptoms, usually vertigo.

Widely used and internationally agreed diagnostic criteria are given in Boxes 14.1 and 14.2.[1,2]

Box 14.1 Diagnostic criteria for Ménière's disease (Bárány Society)

Definite Ménière's disease

- **A.** Two or more spontaneous episodes of vertigo each lasting 20 minutes to 12 hours.
- **B.** Audiometrically documented low- to medium-frequency sensorineural hearing loss in one ear, defining the affected ear on at least one occasion before, during, or after one of the episodes of vertigo.
- **C.** Fluctuating aural symptoms (hearing, tinnitus, or fullness) in the affected ear.
- **D.** Not better accounted for by another vestibular diagnosis.

Probable Ménière's disease

- **A.** Two or more episodes of vertigo or dizziness, each lasting 20 minutes to 24 hours.
- **B.** Fluctuating aural symptoms (hearing, tinnitus, or fullness) in the affected ear
- **C.** Not better accounted for by another vestibular diagnosis.

Box 14.2 Diagnostic criteria for vestibular migraine (Bárány Society)

- **A.** At least five episodes fulfilling criteria C and D.
- **B.** A current or past history of migraine.
- **C.** Vestibular symptoms of moderate or severe intensity, lasting between 5 minutes and 72 hours.
- **D.** At least half of episodes are associated with at least one of the following:
  1. Headache with at least two of the following four characteristics:
     - Unilateral location.
     - Pulsating quality.
     - Moderate or severe intensity.
     - Aggravation by routine physical activity.
  2. Photophobia and phonophobia.
  3. Visual aura.
- **E.** Not better accounted for by another International Classification of Headache Disorders, third edition, diagnosis or by another vestibular disorder.

For MD, the condition can be diagnosed with two levels of certainty, 'definite' and 'probable', with the 'probable' category removing the requirement in criterion B for the documented hearing loss on pure tone audiometry.

From these criteria, it is apparent that there are clinical features in common to both disorders, namely the presence of episodes of vestibular symptoms lasting between 20 minutes and 24 hours. This can make the differential diagnosis between the two conditions difficult, especially in the earlier phases of the condition. There is also a statistical association between the two conditions, so that there is a group of patients who have both MD and VM, further complicating matters.

Pearls for differentiating Ménière's disease and vestibular migraine

- Fluctuating hearing loss is occasionally reported in relation to VM attacks but is not a prominent feature. Strongly lateralized and prominent auditory symptoms point to MD.
- If there is documented hearing loss which meets MD criteria (especially low-frequency unilateral fluctuating loss), MD should be preferentially diagnosed.

- The sensory sensitivity symptoms (phonophobia, photophobia, and also osmophobia and allodynia) point more strongly towards VM.

Pathophysiology

The pathophysiology of MD is incompletely understood, but it is usually thought to relate to endolymphatic hydrops. Endolymphatic hydrops causes characteristic changes in inner ear fluid volumes visible on postmortem examination. In theory, increases in endolymph fluid volumes in endolymphatic hydrops are associated with episodes of symptoms. However, endolymphatic hydrops and MD do not have a perfect one-to-one correspondence, as endolymphatic hydrops is reported in the temporal bones of patients with other otological conditions and also those with no symptoms.

Migraine is a complex disorder and, like MD, its pathophysiology is also incompletely understood. It is known to have a familial predilection; however, most of the common forms of migraine are not single-gene disorders. There are some rare single-gene conditions, such as familial hemiplegic migraine, where migraine is a characteristic feature. Some of these single-gene migraine-associated conditions have a specific relationship with vestibular symptoms. Episodic ataxia type 2, for example, is linked to mutations in the *CACNA1A* gene and predisposes to episodes of ataxia with vestibular symptoms. One hypothesis to explain the heterogeneity of the condition is that there are genetic defects in different groups of ion channels. These defects could be responsible both for inner ear-related symptoms and also for migraine headache by activation of the so-called trigeminovascular reflex, the neurovascular pathomechanism thought to underlie headache. The inner ear receives innervation from the trigeminal nerve. Also, there are direct projections between the vestibular nuclei and the aminergic brainstem nuclei activated in the trigeminovascular reflex. As well as genes, migraine phenotypes are also related to other aetiological pathomechanisms, such as structural lesions and head injuries.

Differential diagnosis

The differential diagnosis of episodic vertigo, with or without headache or auditory symptoms, is very broad.

At first presentation of acute vertigo, a neurological assessment is required to identify a possible vascular cause in the posterior circulation. Adults with a sudden-onset acute vestibular syndrome in whom benign paroxysmal positional vertigo or postural hypotension do not account for the presentation, should be referred urgently in line with local stroke pathways.[3] Symptoms and signs of posterior circulation disease including limb weakness, cranial nerve signs, hiccups, confusion, long tract signs, dysarthria, Horner syndrome, hearing loss, reduced consciousness, and gait or limb ataxia also indicate the need for a neurological opinion. Vascular disease as a cause of recurrent vertigo is more likely in older adults and those with vascular risk factors such as hypertension and diabetes, and less likely where there is a very long history of episodes. Chiari malformations can also present with episodic vertigo and headache similar to the VM phenotype.

Occasionally VM can mimic benign paroxysmal positional vertigo (see Chapter 15) and vice versa, therefore positional testing should always be carried out in patients with episodic vertigo. This can be particularly challenging when benign paroxysmal positional vertigo episodes act as triggers for migraine headache in susceptible individuals (secondary VM).

Episodic ataxia type 2 has some features in common with VM. It usually presents before the age of 20 years and there are frequently interictal eye movement abnormalities. Genetic testing is available.

Other causes of episodic vertigo include superior semicircular canal dehiscence, where additional features of third-window symptoms are characteristic. In vestibular paroxysmia the episodes are stereotyped—very short and often sensitive to treatment with carbamazepine.

Autonomic system disorders such as postural orthostatic tachycardia syndrome, cardiovascular conditions such as arrhythmias, and psychiatric disorders such as anxiety and hyperventilation syndrome can also be associated with episodic vertigo.

MD can be mimicked by a number of conditions including autoimmune inner ear disease, temporal bone fracture, otosyphilis, end-stage otosclerosis, endolymphatic sac tumours, and acoustic neuromas.

Natural history and epidemiology

VM is around ten times as common as MD with an estimated lifetime prevalence of 1%.[4]

Migraine may date back to childhood, teens, or young adulthood, or present later in life. It is less common for it to present for the first time in the elderly, when a secondary cause needs to be suspected. Episodes of vertigo in an adult that date back to childhood without features of cardiac or epileptic cause are highly likely to be migrainous (see Chapter 85). VM, like other forms of migraine, may be a persistent problem over many years. In a prospective study in a specialist centre following patients over 11 years, 87% still had vertigo. More than half found the episodes were less frequent and less severe, but in around a third the frequency and severity increased. Therefore VM must be regarded as being a chronic condition in the majority of cases.

MD likewise has a variable outcome. In some cases, there may be complete recovery and remission. In others, symptoms are more persistent and progressive. In more extreme cases, the hearing loss becomes permanent and can progress to become severe or profound, usually over around 5–10 years. A number of cases of MD can become bilateral and cochlear implantation (see Chapter 16) may become necessary. Estimates of the proportion of cases of MD that become bilateral vary, but are of the order of up to 50% over 20 years. The potential for MD to become bilateral is an important consideration in some types of treatment. **Figure 14.1** shows the characteristic changes in a patient with unilateral MD.

The frequency of vertigo attacks in MD typically diminishes within 5–10 years, but the attacks can be replaced by permanent vestibular dysfunction analogous to the cochlear hearing loss. The vestibular dysfunction can therefore be unilateral (in the majority) or bilateral. Affected individuals can experience more persistent imbalance and head motion intolerance accordingly.

Drop attacks are a relatively uncommon feature of MD where patients can suddenly, without warning, fall to the ground but without

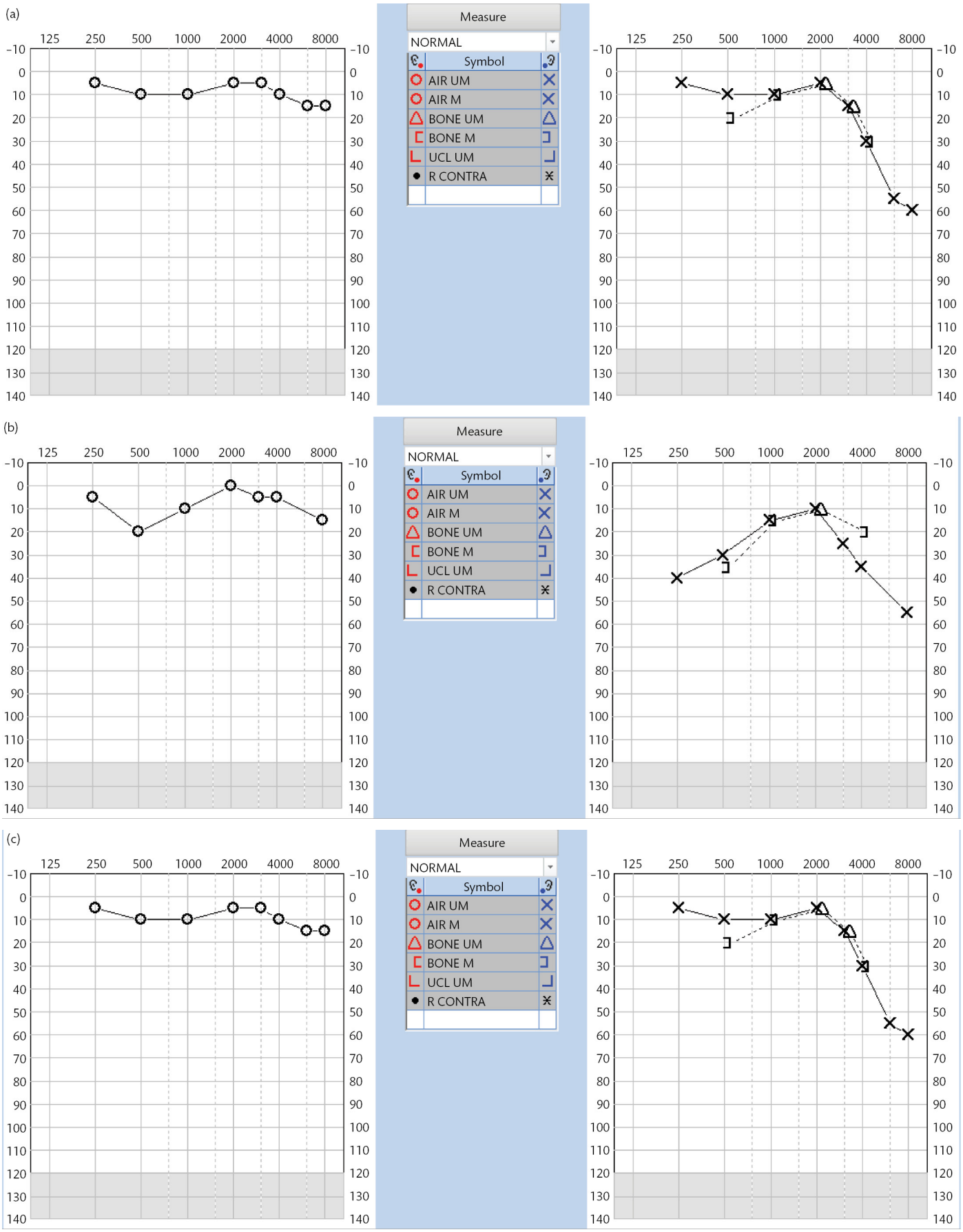

Figure 14.1 The typical audiometric changes of MD over time. This patient has a long-standing unilateral high-frequency loss at baseline (a) of unknown cause. Initially there is some minor low-
frequency hearing deteriorates (d) and eventually there is a flat sensorineural hearing loss (e).

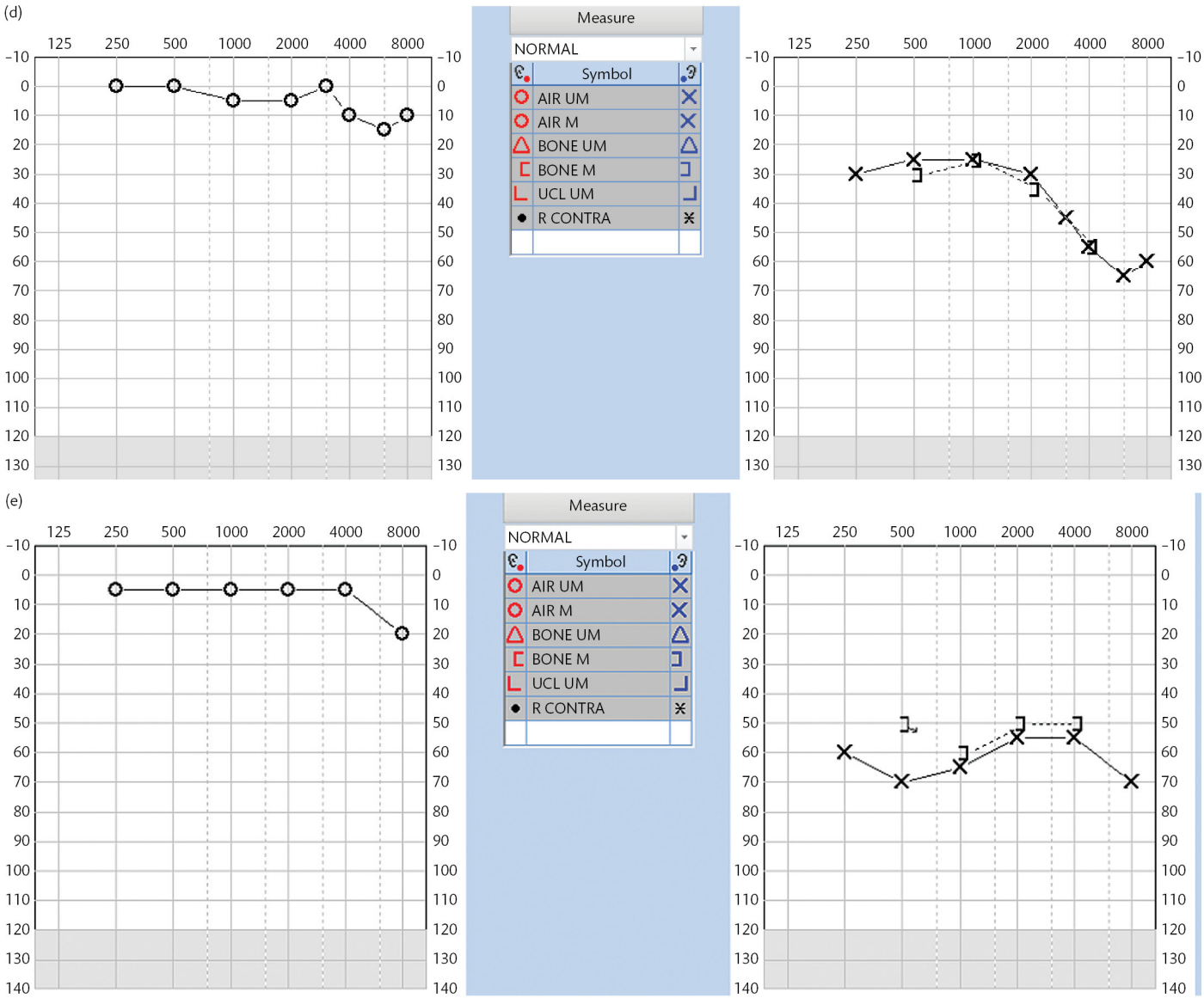

Figure 14.1 Continued

any disturbance of awareness. Drop attacks may occur early or late in the course of the disease, and remission is often spontaneous.

Persistent vestibular symptoms can occur at the more extreme end of the clinical severity spectrum in either VM or MD where attacks are very frequent so recovery from one attack is not complete before the next begin; or in MD where the condition has progressed to the stage where permanent vestibular dysfunction is present, or where either condition has been complicated by persistent perceptual postural dizziness.[5]

History

The diagnosis for MD and VM is primarily made on the basis of the history. The history should aim to gather enough information to determine the likely diagnosis, but also the level of disability and distress the patient is experiencing as this is critical in deciding the management strategy. Self-treatment and prescribed treatment history must also therefore be sought. It is also wise to determine the patient's own health beliefs about the cause of the condition and required action to address it, in order to better develop a shared understanding with the patient for a collaborative management strategy (Box 14.3).

Examination

When seen in between episodes, the examination of patients with VM is usually normal. During an episode, eye movement abnormalities may be seen such as peripheral or central types of nystagmus, or positional nystagmus can also be observed. These recover at follow-up when the attack has resolved.

Box 14.3 History taking for episodic vertigo

The presenting episodes

- What type of dizziness? Vertigo, head motion intolerance?
- Duration of episodes? Typical episode, longest, shortest?
- Associated symptoms? Headache, photophobia, phonophobia, hearing loss, tinnitus? What is the timeline for each of these symptoms separately?
- What is the current frequency and severity of episodes, and how is this condition affecting work, family responsibilities, and social life?

Between the episodes

- Is there persistent imbalance, head/visual motion intolerance, hearing loss, tinnitus?
- Is there secondary psychological disorder such as anxiety or depression?

Migraine history

- Is there any lifetime history of migraine? Remember that headache and migraine can be normalized, so patients with infrequent or milder forms of migraine may well not have a formal diagnosis.
- Is there a family history? Note, however, that migraine is a very common condition so a family history is not very specific.
- Are there any features which suggest a secondary cause (Box 14.4)?

Treatment history

- Is there any use of non-prescribed treatments? If there is migraine headache, then ask particularly about use of analgesics including those available over the counter. Analgesic-related headache is common.

Likewise, MD patients seen in between attacks will frequently have a normal physical examination. As the condition progresses, there may be signs of a persistent unilateral sensorineural hearing loss, which can become bilateral. Similarly, there can eventually be signs of a persistent unilateral or bilateral vestibular hypofunction. During attacks, irritative nystagmus may be seen (beating towards the affected ear), whereas between attacks nystagmus is more likely to be paretic (beating away from the affected ear).

A key aspect of physical examination in both cases is to ensure that alternative diagnoses are not missed, therefore as well as a careful ENT assessment, neurological examination including fundoscopy and cardiovascular examination may be necessary.

Investigations

The diagnosis of VM is made on clinical grounds and there are no confirmatory investigations available. Where the diagnosis is completely clear, it may be possible to avoid any additional investigation. However, investigation may well be required to exclude secondary or alternative causes for symptoms and should be particularly considered at the first presentation, where symptom pattern is atypical or where response to treatment is poor, as well as where red flags are present (Box 14.4).[6]

For MD, a pure tone audiogram is required to make a definite diagnosis. Unilateral sensorineural hearing loss will frequently require investigation with magnetic resonance imaging to exclude secondary causes. If there is evidence of autoimmune inner ear disease, appropriate blood work may be considered. Vestibular function tests

Box 14.4 Red flag features of headache (National Institute for Health and Care Excellence 2022 guidelines)

- Worsening headache with fever.
- Sudden-onset headache reaching maximum intensity within 5 minutes.
- New-onset neurological deficit.
- New-onset cognitive dysfunction.
- Change in personality.
- Impaired level of consciousness.
- Recent (typically within the past 3 months) head trauma.
- Headache triggered by cough, Valsalva manoeuvre, or sneeze.
- Headache triggered by exercise.
- Headache that changes with posture.
- Symptoms suggestive of giant cell arteritis.
- Symptoms and signs of acute narrow angle glaucoma.
- Substantial change in the characteristics of the headache.

are not necessary in all cases but can be helpful in some selected cases, for example, if ablative treatment is being planned.

Treatments

In both MD and VM, as neither condition is of itself directly life-threatening, the aim of treatment is to minimize the number and severity of attacks and to improve quality of life. There are no treatments that are known to affect outcome in the long term; this includes the hearing loss that is a complication of MD. As both conditions are likely to be chronic, the aim should be for clinicians to facilitate patients in developing empowered self-management.

Vestibular migraine

Conservative

Patients may be able to identify triggers for attacks, and should be encouraged to consider this. However, it is not always possible to identify clear triggers, and further it is not always possible to modify triggers even when they are clear. Dietary triggers are apparent in some cases; however, there is little evidence to support a role for exclusion diets which can be very burdensome for patients. Alcohol and caffeine are recognized to have an effect in many cases and should be moderated or excluded. Stress management advice may be helpful.

Acute attack

Vestibular symptoms in the acute attack can be relieved with vestibular suppressants such as prochlorperazine (oral 10 mg or buccal 3 mg). The patient needs to be advised to restrict use to short-term relief only. Headache can be managed using simple analgesics or, if this is ineffective or the headaches are severe, then a triptan such as sumatriptan 100 mg, naratriptan 2.5 mg, or almotriptan 12.5 mg can be offered.[7] Triptans can be combined with a long-lasting NSAID such as naproxen. If oral medications are offered then adding an antiemetic (prochlorperazine 10 mg, domperidone10 mg, or metoclopramide 10 mg) to an acute treatment improves efficacy even if nausea and vomiting are not prominent features.

Preventive

There are a large number of different prophylactic preparations available for migraine, which vary hugely in terms of pharmacological properties, contraindications, evidence for efficacy, and tolerability. Beta blockers are the most popular choice as a first-line agent worldwide, and propranolol 10–20 mg twice daily is a common starting regimen.[7] Common side effects include fatigue, hypotension, and bradycardia. Tricyclics such as amitriptyline or nortriptyline starting at 10–25 mg/day are also considered first-line options in many published guidelines, especially if there is comorbid sleep disturbance related to depression or anxiety. Common side effects include sedation and weight gain. Candesartan has a small evidence base in favour of efficacy but is better tolerated than many other options, although blood pressure and renal function need to be monitored. Specialist clinics may also explore other types of treatment such as greater occipital nerve injections, Botox, or neuromodulators. Calcitonin gene-related peptide monoclonal antibodies such as erenumab are becoming more widely available and at the time of wirting there is preliminary evidence of effectiveness for VM. There are some small studies that support the use of the nutraceuticals riboflavin, coenzyme Q10, or magnesium. Although these have a lower evidence base of efficacy, some patients will prefer this option over prescribed medication for personal reasons and because they are usually very well tolerated.

It is worth noting that the evidence in favour of these medications derives from studies on migraine headache rather than VM. To date, there have been no completed prospective randomized controlled trials on prophylactics for VM.[8]

Ménière's disease

Conservative

As for VM, some patients may be able to identify triggers for attacks, and this can be supported. Low-salt diets have long been advocated by experts; however, a Cochrane review has no shown no high-quality evidence either for or against this intervention.[9] Where symptoms of hearing loss or tinnitus are persistent between attacks, patients should be referred to audiology for consideration of hearing aids or other rehabilitative devices as well as appropriate hearing therapy support. Similarly, where head motion intolerance, postural instability, or visually triggered symptoms persist between episodes for prolonged periods, vestibular rehabilitation should be offered.

Acute attack

The symptoms of vertigo, nausea, and vomiting can be addressed with antiemetics such as prochlorperazine, cyclizine, and cinnarizine. These must be reserved for acute attacks only and should not be given regularly over long periods.

Preventive

There are no medications with a strong evidence base to support usage as prevention in MD. Betahistine has been a popular choice in the UK for a long time. There is no good-quality evidence for its use, but it is very well tolerated with a good safety profile and therefore remains in usage.[10] Diuretics have been advocated by some experts but a Cochrane review showed there is no good evidence for their use and there are significant potential unwanted effects.[11]

Interventional and surgical

Intratympanic injection of steroid injections is increasingly offered as a treatment to patients whose attacks are frequent and interfering with quality of life.[12] There are at least two randomized trials and several systematic reviews indicating efficacy in the treatment of vertigo with a preponderance of benefit over harm. Intratympanic gentamicin therapy is also offered when symptom control is inadequate with other more conservative treatments. Steroids or gentamicin may be injected directly through the tympanic membrane, via a middle ear ventilation tube with or without a catheter or via a surgically inserted microcatheter. With the use of gentamicin there is a greater concern about the possibility of worsening both vestibular and cochlear function, although trials show that rates of hearing loss with gentamicin are no different than for use of steroids. Caution must be applied to those with bilateral MD and it must be remembered that the potential for bilateral MD exists in those with unilateral disease. The vestibular status of the non-treated ear should be assessed before chemical ablative treatment with gentamicin. If treatment with chemical agents fails, and hearing is not serviceable, patients may be referred for consideration of labyrinthectomy or vestibular nerve section, although this should rarely be necessary. Endolymphatic sac decompression and ventilation tube insertion are also sometimes done but are of uncertain benefit.[13]

Patient experience

The feeling of loss of control and unpredictability are common themes in patient perspective reports of living with a liability to episodes of vertigo. There is the potential for wide ranging impacts on work, leisure, social life, and family responsibility. Driving may be impacted with an associated loss of independence as well as the implications for work and quality of life. Another common report is the sense of living with an invisible disability. During attacks, sufferers can be mistaken as being intoxicated with alcohol, leading to unsympathetic or even hostile treatment from those around them. A full biopsychosocial approach which validates these experiences is necessary. Some patients may find benefit in contact with peer support groups.

Controversies/areas of interest for the future

Both conditions would benefit from the existence of a reliable objective biomarker. There is interest in the potential ability for magnetic resonance imaging to show the presence of endolymphatic hydrops (Figure 14.2), and this is a rapidly evolving area.[14] Such a diagnostic tool could potentially help differentiate MD from other forms of episodic vertigo such as VM in the early stages, or be used to detect early changes that predict bilaterality. There is a need likewise to establish a clinically useful biomarker for VM. For both VM and MD, research needs to identify subtypes that predict outcomes such as bilaterality (in MD),[15] prognosis, or treatment response (for both VM and MD). There are many treatments (drugs and others) being researched for use in migraine at the present time, and it remains to

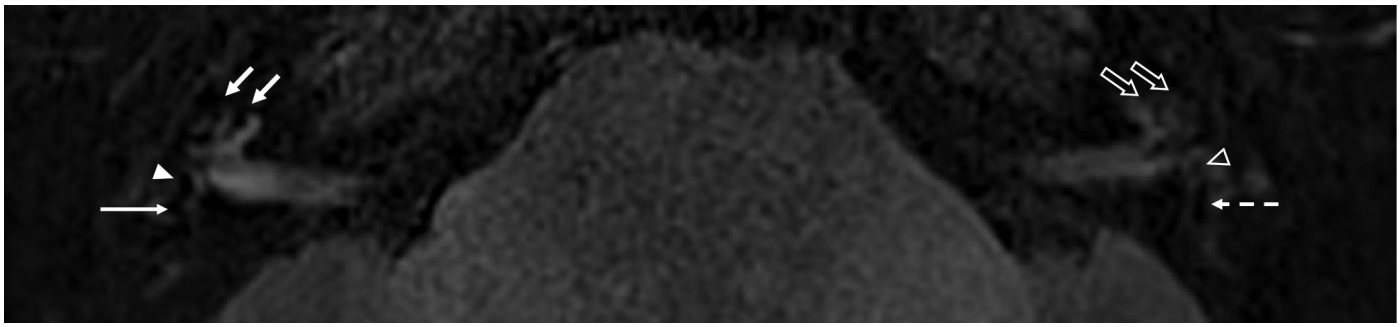

Figure 14.2 Axial image from a three-dimensional T2 FLAIR sequence through the temporal bones 4 hours following gadolinium administration in a patient with clinical features of right-sided Ménière's disease. On the right, there is distension of the non-enhancing cochlear duct (short solid arrows), effacing the scala vestibuli, in keeping with cochlear hydrops. This compares to normal left side (short outlined arrows). The right vestibular endolymphatic structures, including the saccule (solid arrowhead) and utricle (long arrow) are also distended relative to the normal contralateral saccule and utricle (outlined arrowhead and dashed arrow respectively); these appearances are compatible with right-sided vestibular hydrops.
Acknowledgement: Dr Philip Touska.

be seen the extent to which any of these may be more or less specifically useful for VM rather than other subtypes of migraine.

Relevant guidelines (national and international)

There are a number of different UK national guidelines on the management of migraine from the headache perspective, including from the National Institute for Health and Care Excellence,[6] the British Association for the Study of Headache [7] and the Scottish Intercollegiate Guideline Network.[16]

The American Academy of Otorhinolaryngology–Head and Neck Surgery has also developed a clinical practice guideline on MD.[17]

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15

Benign paroxysmal positional vertigo

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Introduction

Benign paroxysmal positional vertigo (BPPV) is a common and treatable vestibular disease, causing vertigo triggered by positional changes, and characteristic nystagmus. Between 17% and 40% of patients with peripheral vestibular disease are diagnosed with BPPV.[1,2] There is a common misperception that BPPV is the sole cause of vertigo, or that all vertigo should be treated with the Epley manoeuvre. BPPV is one of many causes of vertigo and, as with any medical condition, precise diagnosis is required for effective treatment. The incidence is reported to be between 10.7 and 140 cases per 100,000 people each year.[3,4] This increases with age, with an overall lifetime prevalence of 2.4%.[5] BPPV is more common in women, with a ratio of about 2:1.[6] In addition to discomfort caused by vertigo, there is also increased risk of falls and resulting injury, which can cause severe morbidity and/or mortality in the elderly. Overall, older patients who report 'dizziness' as a symptom have a 12-fold increased risk of falls.[7] In geriatric patients, about three-quarters of those with a diagnosis of BPPV had fallen in the 3 months prior to their referral for geriatric evaluation.[8] Underdiagnosis is a problem as well—9% of patients undergoing routine geriatric evaluation had undiagnosed BPPV in one study.[8]

Basic science: anatomy and physiology

BPPV was first described in 1921 by Bárány, who believed the syndrome to be due to otolith disease.[9] In 1952, Dix and Hallpike described a provocative test for BPPV, which is still used today.[10] Schuknecht proposed the cupulolithiasis theory of BPPV in 1969.[11] He theorized that dislodged otoconia from the utricle and saccule otoliths become fixed to the cupula of the posterior semicircular canal. When motion occurs relative to gravity, the fixed otoconia deflect the cupula, causing characteristic symptoms. This theory predominated until 1979 when Hall, Ruby, and McClure published the canalolithiasis theory,[12] in which dislodged otoconia from the saccule and utricle otoliths make their way into the posterior semicircular canal through the membranous labyrinth. Once in the posterior canal, otoconial movement due to positional changes mimics natural head movements, tricking the brain into thinking that intense spinning is occurring, which in turn prompts compensatory eye movements (nystagmus) and perceptual illusions (vertigo). In support of this theory studies have shown that ocular vestibular evoked myogenic potential[13] and subjective visual vertical[14] are impaired in patients with BPPV, which supports the idea that dysfunction is caused by the loss of otoconia in the utricle. Additionally, specimens collected from the posterior canal during posterior semicircular canal occlusion found intact and degenerating otoconia.[15] The current working theory for posterior canal BPPV is the canalolithiasis model, and the hugely successful canalith repositioning manoeuvres, like the Epley manoeuvre, are predicated on that theory. However, the cupulolithiasis model may explain certain rare variants, like ageotropic horizontal canal BPPV.

Although the model explaining the symptoms behind BPPV has been understood for many years, the causative mechanism behind the dislodged otoconia remains unclear. The majority of cases are idiopathic. However, BPPV has been shown to be associated with post-traumatic head injuries, vestibular neuritis, labyrinthitis, migraine, and Ménière's disease. The posterior canal is the most gravity-dependent portion of the inner ear, which makes it the most likely place for dislodged otoconia to sit. That anatomy explains why 95% of diagnosed BPPV is due to posterior canal pathology.[16] Horizontal canal BPPV has also been described far less commonly, making up between 5% and 15% of diagnosed BPPV cases.[16,17]

Investigations

Patients with BPPV commonly report episodic vertigo. These episodes are usually provoked with head motion, including rolling over in bed, tilting the head upward, or bending forward. The vertigo is brief, generally lasting several seconds and rarely longer than 1 minute. However, nausea, dizziness, and imbalance may persist once the vertigo itself resolves.[5] Additionally, descriptions of vertigo and dizziness in patients are often inconsistent and unreliable, so the astute clinician should always consider BPPV in the differential diagnosis of vertigo and dizziness.[18]

The gold standard for diagnosis for posterior canal BPPV is the Dix–Hallpike manoeuvre.[19] Posterior BPPV has a characteristic nystagmus elicited by this provocative test. The fast component of the

nystagmus is torsional and upbeating, as would be expected from posterior canal activation. This is because the canaliths float away from the posterior canal ampulla during the manoeuvre, resulting in ampullofugal deflection, which is excitatory for the posterior canal. The first of Ewald's laws, which govern vestibular physiology, states the evoked nystagmus will be in the same plane as the canal being activated. That means that BPPV nystagmus will change appearance as eyes are brought in and out of the plane of the posterior canal. For example, with the right posterior canal, eyes are coplanar with the canal when looking to the left, so nystagmus will be purely upbeating. Conversely, when looking to the right, the eyes will be at a 90° angle to the canal, so nystagmus will be rotatory, with the top pole of the eye rotating to the right. In the neutral gaze position, we see a mix, due to the eyes being 45° off the plane of the posterior canal, resulting in the typical upbeating and torsional nystagmus. Since for either ear the direction of the torsional component is always towards the ground in the Dix–Hallpike position, it is also referred to as 'geotropic' (**Figure 15.1**).

Additionally, there should be a short latency period between positioning and the onset of the nystagmus and symptomatic vertigo. The exact time frame of the latency period is not fully agreed upon. In the initial paper,[10,20] the latency period was described as falling between 2 and 5 seconds, and this is consistent with the author's clinical experience. The nystagmus also generally resolves within 60 seconds of the onset. It is commonly described as being a crescendo–decrescendo pattern of nystagmus; after the latency period the nystagmus begins, increases in intensity, reaches a peak and then decreases in intensity until it resolves. The nystagmus also commonly reverses in direction once the patient sits up at the end of the manoeuvre, presumably due to otoconia falling back towards the ampulla. The nystagmus is fatigable, diminishing with repeated trials of the Dix–Hallpike manoeuvre. Cases with long-duration nystagmus or without fatigue may be due to otoconia near or attached to the cupula.

To complete the Dix–Hallpike manoeuvre the patient begins seated and the provider rotates the patient's head 45° to the affected side, aligning the posterior canal with the sagittal plane. The practitioner then moves the patient from the seated to the supine position with the affected ear down and the neck extended 20°. Nystagmus and symptoms are then characterized. The patient is then returned to the upright seated position. If the Dix–Hallpike manoeuvre is

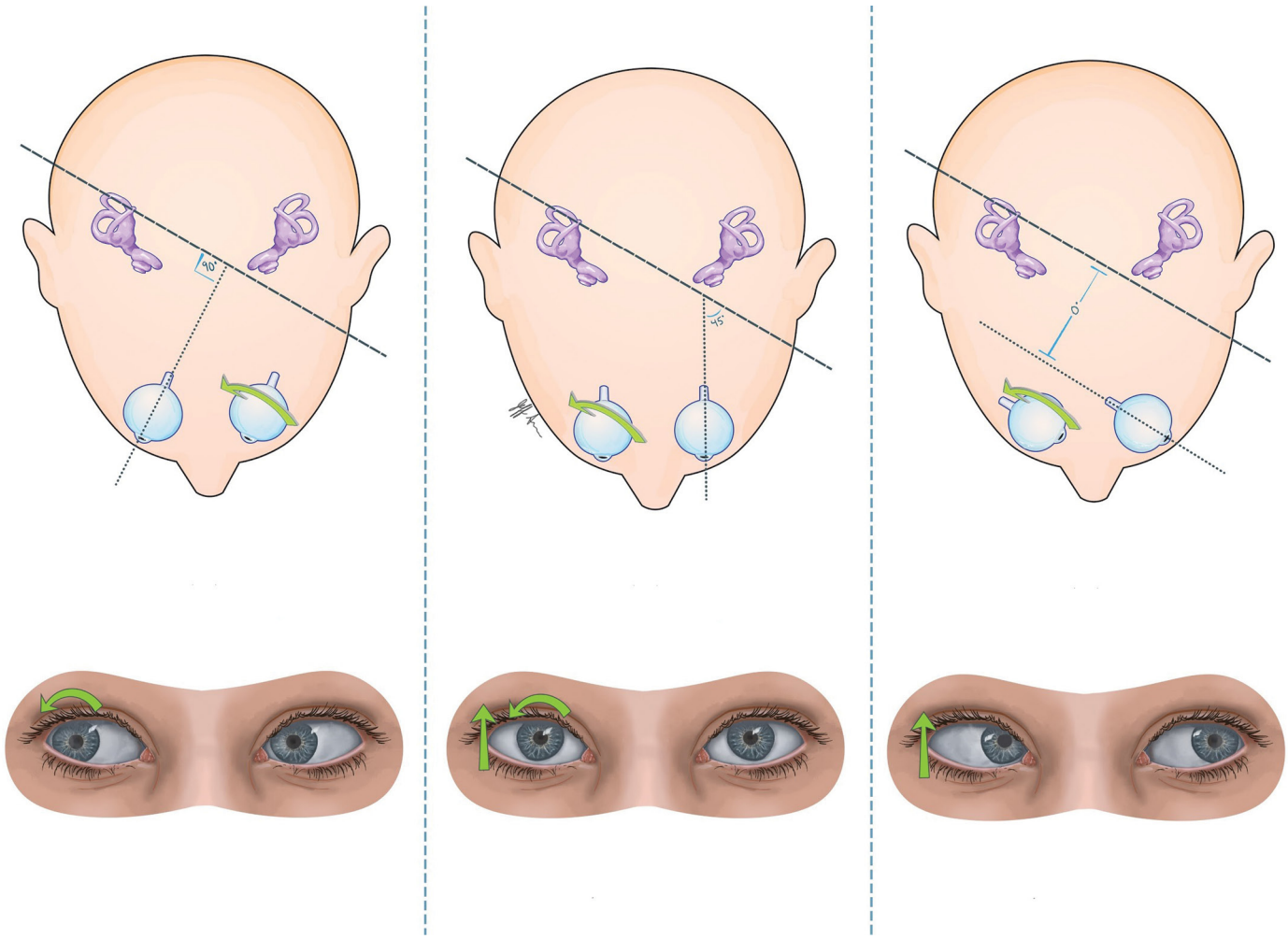

Figure 15.1 Characteristic nystagmus in posterior canal BPPV. This shows the characteristic nystagmus seen with right-sided posterior canal BPPV as the eyes are brought in and out of the plane of the posterior canal. In the first panel the eyes are looking to the right, at 90° to the canal, so the nystagmus is rotatory. In the second panel the eyes are in a neutral gaze position 45° off the plane of the canal so the nystagmus is mixed upbeating and torsional. In the third panel the eyes are looking to the left, coplanar with the canal causing a purely upbeating nystagmus.

positive, rather than returning to the sitting position, the patient can be directly transitioned to the repositioning manoeuvre. Although the Dix–Hallpike manoeuvre is the gold standard, up to one-quarter of patients with BPPV may have false-negative testing.[21] The sensitivity may be increased by using infrared or Fresnel goggles, or by 'loading' the manoeuvre with a downward head tilt for 30 seconds prior.[22] The current clinical practice guidelines state that a provider should diagnose a patient with posterior canal BPPV if the patient has subjective vertigo and torsional upbeating nystagmus provoked by the Dix–Hallpike manoeuvre.[20] Although this manoeuvre provokes vertigo and may cause symptoms of nausea, it is generally well tolerated. Special consideration should be taken in patients with decreased cervical range of motion as neck extension is required to perform the manoeuvre, unless performed on a stretcher capable of Trendelenburg positioning. If no nystagmus is observed, then the Dix–Hallpike manoeuvre is repeated on the other side. To be able to check both ears, the authors usually check the suspected non-affected side first.

In cases where the clinical picture is consistent with BPPV, but the Dix–Hallpike manoeuvre shows either horizontal nystagmus or no nystagmus, providers should consider horizontal canal BPPV.[20] To perform the supine roll test[23,24] the patient begins lying supine with the head on a 20° incline, so that the horizontal canal is vertical. The head is quickly turned 90° to one side. Any observed nystagmus is characterized, and then the head is moved back to neutral and then quickly turned 90° to the other side and again observed for nystagmus. Patients with horizontal canal BPPV have horizontal nystagmus with the head turned in either direction. The nystagmus is either geotropic (fast phase towards the ground) or apogeotropic (fast phase away from the ground). Compared to the posterior canals, it is harder to lateralize horizontal canal BPPV, because the two horizontal canals are coplanar. Noting which side provokes stronger nystagmus and symptoms can help determine the affected ear, as excitation provokes a better response than inhibition.

The geotropic variant of horizontal canal nystagmus is more common. In this type, when the patient's head is rolled to the affected side, the patient will have an intense nystagmus towards the affected ear. When the head is rolled to the unaffected side, the patient will have a less intense nystagmus that is still horizontal and geotropic. This is believed to be due to canalolithiasis, otoconia present within the horizontal canal. The apogeotropic type is less common. When the head is turned towards the affected ear the patient has nystagmus that beats towards the unaffected ear. When the head changes positioning to the unaffected ear downwards the nystagmus will continue to beat away from the ground, now towards the affected ear. This type of horizontal canal BPPV is believed to be due to otoconia that are adherent to the cupula of the canal.[24,25] Additional methods have been described to aid providers with lateralization including the bow and lean test.[26]

Superior canal BPPV is rarely seen and poorly understood. It is believed to represent about 2% of all BPPV cases.[26,27] The characteristic nystagmus is described as downbeating, torsional nystagmus in which the top poles of the eyes beat towards the affected ear.[28,29] Similar to posterior canal BPPV, the nature of the nystagmus should change as eyes are brought in and out of the plane of the canal. For cases with unusual nystagmus, the authors will frequently record the nystagmus (with the patient's written permission) with either infrared goggles, or a high-speed camera as available on many of today's smartphones, so that an 'instant replay' is possible. Purely downbeat nystagmus is usually due to central pathology, and frequently increases in intensity with Dix–Hallpike manoeuvre positioning.

Differential diagnoses and associated conditions

BPPV can be diagnosed with clinical history and positive provocative manoeuvres alone. If patients meet clinical criteria for BPPV, routine radiographic imaging or vestibular testing is not recommended.[20] However, those who present with atypical nystagmus, signs or symptoms that raise concerns for an additional underlying vestibular pathology, failed response to repositioning manoeuvres, or frequent BPPV recurrence may require additional workup. In these patients, important causes to consider include stroke, vestibular neuritis, vestibular migraine, or Ménière's disease. As BPPV can be associated with additional underlying vestibular pathology, it is important to consider additional diagnoses if symptoms persist after appropriate treatment.[20] An often-overlooked feature of vestibular migraine is that it can cause positional vertigo, which can be associated with a low-intensity nystagmus.[30] **Table 15.1** reviews the differential diagnosis of positional vertigo.

Medical treatments and regimens

Although sometimes self-limiting, canalith repositioning procedures are recommended for treatment of BPPV. These manoeuvres are thought to work by moving the otoconia out of the posterior canal back into the vestibule. The Epley manoeuvre is a safe and effective repositioning procedure for the treatment of posterior canal BPPV, first described in 1992 (**Figure 15.2**).[31] It begins with the patient seated upright with their head turned 45° towards the affected ear. The patient is then rapidly placed into the supine position with their head extended 20°, like the Dix–Hallpike manoeuvre. Once the diagnosis is confirmed, the patient is held in this position for 30–60 seconds. Their head is then turned 90° towards the unaffected side and held for 30–60 seconds. Next, the head is turned another 90° while the body is brought into the lateral decubitus position and held there for 30–60 seconds. The patient is finally brought back up to the seated position, with the neck flexed down. This procedure has been shown to have significant symptom resolution and conversion to a negative Dix–Hallpike manoeuvre compared to sham procedures or no procedure. Resolution has been reported to be as high as 80% after one session, and 92% with four sessions.[32] However, the optimal number of treatments and sessions remains unclear. Although the success rates are higher with specialists than general practitioners, it can even have an efficacy of up to 95% with self-administration.[33] No serious adverse effects usually seen with this procedure. Some don't tolerate the procedure due to cervical spine issues and emesis during the treatment. Up to 5% of posterior canal BPPV is inadvertently converted to horizontal canal BPPV during the Epley repositioning procedure.[34,35] Recurrences of BPPV are common, and up to 36% of patients have symptomatic recurrence within the first 48 months after treatment with the Epley manoeuvre.[36]

Another repositioning procedure is the Semont manoeuvre, which was first described in 1988.[37] In this procedure the patient

Table 15.1 Differential diagnosis of positional vertigo

Diagnosis	Onset and course	Nystagmus	Associated symptoms
BPPV	Discrete episodes of vertigo that last generally <1 minute	Fast component of nystagmus is torsional and upbeating during the Dix–Hallpike manoeuvre, has a brief latency period, and is fatigable (posterior canal) Horizontal nystagmus seen in supine roll (horizontal canal)	Nausea is common May report imbalance or lightheadedness between discrete episodes
Vestibular migraine	Classically vertigo lasts minutes to hours but can also have brief attacks lasting minutes or seconds	Variable: low velocity, can be horizontal, vertical, or torsional	Headache may occur with vertigo or independently, photophobia/phonophobia can occur
Light cupula	Discrete episodes of vertigo that generally last >1 minute	Horizontal geotropic nystagmus that is non-fatigable	Can be seen with alcohol consumption
Superior semicircular canal dehiscence	Vertigo often triggered by sound or pressure (Tullio phenomenon)	Can be induced by vibration, downbeat torsional nystagmus	Conductive hyperacusis, autophony, pulsatile tinnitus, hearing loss, phonophobia, dizziness with coughing/sneezing/straining
Head trauma/post-concussive syndrome	Onset acutely after head impact or can be delayed, frequently causes BPPV	Similar to BPPV, but more likely to be bilateral or multi-canal—however, post-concussive syndrome can cause positional dizziness without BPPV as well	History of trauma More resistant to treatment
Chiari malformation and other craniocervical anomalies	Frequently have Valsalva-induced vertigo	Persistent downbeating nystagmus	Weakness, headache, ataxia, lower cranial neuropathy
Vertebrobasilar insufficiency	Transient ischaemic attacks last <24 hours Stroke results in permanent disability	Central pattern nystagmus including upbeat, downbeat, or rotary nystagmus, rarely can mimic peripheral pattern nystagmus	Diplopia, visual field defects, headache, hearing loss, paresthesias, weakness
Multiple sclerosis	Variable	Pendular nystagmus, downbeating or upbeating nystagmus, gaze-evoked nystagmus	Internuclear ophthalmoplegia, loss of vestibulo-ocular reflex inhibition, optic neuritis, transverse myelitis, other focal neurological signs
Cerebellar degenerations	Nystagmus is often more significant than vertigo	Persistent downbeating nystagmus that may be stronger with positioning, no latency period, non-fatigable	Focal neurological signs, including cerebellar, limb or truncal ataxia, abnormalities of smooth pursuit or saccades, or central patterns of nystagmus including gaze evoked, rebound, and downbeating

begins seated with their head turned away from the affected ear. The provider then quickly lies the patient down towards the affected ear with the head turned away. The patient remains in this position for about 30 seconds, or until nystagmus resolves. The patient is then quickly moved through the seated position to lying on the opposite side with the head facing down. They remain in this position for 30 seconds. The patient is then slowly returned to seated. It has been shown to be equally effective as the Epley repositioning procedure in resolving posterior BPPV.[36] However, it is less effective than the Epley manoeuvre with self-administration.[36] Reported adverse events are similar to those reported with the Epley manoeuvre. Currently neither the Epley or Semont procedure requires postural restrictions after completion as they have not been shown to prevent recurrence.[20] Follow-up 1 month after repositioning procedures is recommended to reevaluate symptoms. Videos detailing repositioning techniques can be found online (https://ohns.ucsf.edu/balance-falls/BPPV).

There are other options, including vestibular rehabilitation, that have been proposed as treatments for posterior canal BPPV. Exercises like Cawthorne–Cooksey and Brandt–Daroff have been shown to improve BPPV. However, the repositioning procedures are significantly more effective.[35] The current role of vestibular rehabilitation is as an adjuvant treatment which can be offered for those who have persistent disability following repositioning procedures or if an individual is not a candidate for repositioning due to cervical spine issues.

There is no role for the routine treatment of BPPV with medical therapy.[20] Medications used in the management of vestibular symptoms including benzodiazepines and antihistamines can interfere with central compensation in peripheral vestibular disorders and prolong the duration of subjective symptoms. Additionally, these medications have significant side effects, which are often more pronounced in the elderly population. An antiemetic can be given prophylactically prior to the repositioning manoeuvres if needed.

Several repositioning procedures for horizontal canal BPPV have also been described; however, less data are available. It is important for the provider to identify which side is affected prior to repositioning. The geotropic form of horizontal canal BPPV is more responsive to these manoeuvres than the ageotropic form. The most well-described manoeuvres include the Lempert 360° roll manoeuvre (also known as the Barbeque or log roll)[27,38] and the Gufoni manoeuvre.[39] The log roll starts with the patient lying with the affected ear down. The head (and body) are then rotated away from the affected ear in 90° increments, stopping at each position for 30

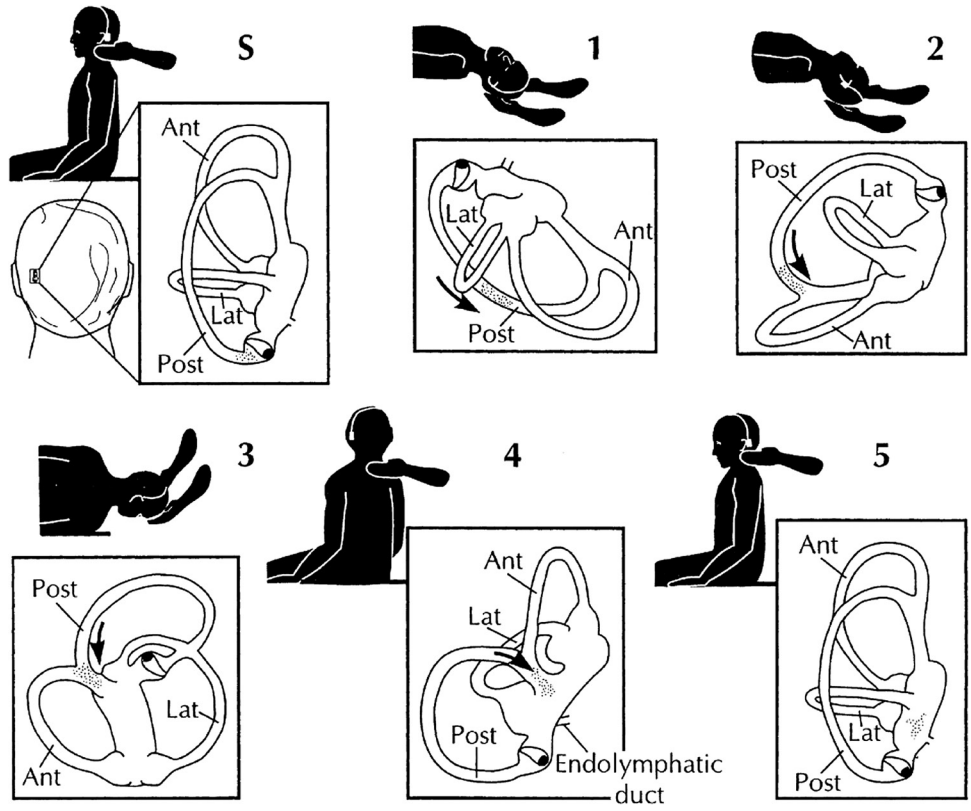

Figure 15.2 Epley manoeuvre. Diagram of the steps of the Epley manoeuvre showing patient positioning and movement of otoconia from the posterior semicircular canal towards the utricle.

Reproduced from Corbridge R, Steventon N. The inner ear. In: *Oxford Handbook of ENT and Head and Neck Surgery*. 3rd ed. Oxford University Press; 2019, https://doi.org/10.1093/med/9780198725312.003.0007, with permission from Oxford University Press.

seconds. A full 360° spin is completed, and then the patient is returned to seated position.

The Gufoni manoeuvre is completed differently if the patient has geotropic or apogeotropic nystagmus. For geotropic nystagmus the patient is seated and then moved to a lateral decubitus position on the unaffected side for 30 seconds. The head is then turned downward 45–60° and held for 1–2 minutes. The head is then turned towards the unaffected shoulder and the patient is returned to sitting with the head turned. For apogeotropic nystagmus the manoeuvre begins with moving from the seated position to side lying on the affected side. After 30 seconds, the head can either be turned up quickly (theoretically treating debris on the canal side of the cupula), and held for a minute, or turned down quickly, and held for a minute (theoretically treating debris on the utricular side of the cupula). The head is then turned to the unaffected shoulder, and the patient is brought back into sitting position. Other methods include forced prolonged positioning, where a patient lies for 12 hours on the affected (geotropic) or unaffected (apogeotropic) side. Overall, the log roll and Gufoni procedures have been determined to be moderately effective for geotropic horizontal BPPV and there is insufficient evidence for their efficacy in apogeotropic horizontal BPPV.[39]

Evidence is limited for treatment of anterior canal BPPV. However, several repositioning manoeuvres have been described. In the Yacovino or deep head hanging manoeuvre, the patient begins in the sitting position.[40] From there the patient is laid supine with the head extended 30° below the horizontal for 30 seconds. Next, the neck is taken from extension to flexion, and held with the chin against the chest for 30 seconds. Finally, the patient is brought back to sitting position. One advantage of this approach is that it theoretically treats both anterior canals, since the head is not turned. However, given the rarity of anterior canal BPPV, there are no large randomized trials evaluating the efficacy of treatment.

Surgical treatments

There is a limited role for surgical treatments in BPPV. In patients with recurrent intractable BPPV, occlusion of the posterior semicircular canal leads to resolution of symptoms.[41] In patients with frequent BPPV recurrence that is successfully treated with the Epley manoeuvre, it is reasonable to offer posterior semicircular canal occlusion as a definitive way to stop recurrence. Historically, singular neurectomy was used to treat intractable BPPV,[42] but the procedure has fallen out of favour. It is important to confirm the diagnosis of BPPV before offering surgical therapy. If patients are not responding to the Epley manoeuvre, it is important to consider alternative diagnoses.

Tips and pearls

- Examine the suspected non-affected ear first, otherwise it can be difficult to examine after treatment.

- The Dix–Hallpike position is very stimulating, so prepare the patient ahead of time that if you see BPPV, you will go straight into treatment, rather than repeating it later in the visit.
- Be sure to ask about neck issues before positioning, and check neck range of motion.
- Treat BPPV yourself. It can be tempting in a busy clinic to provide the patient with home instructions, but treatment time is very well spent.
- Based on evidence, initial treatment should be with the Epley or Semont manoeuvre.
- Look carefully at the nystagmus pattern to confirm diagnosis—there are other causes of positional vertigo.
- For uncomplicated cases, do not request imaging or audiovestibular testing.
- Check the patient again after treatment with the Dix–Hallpike manoeuvre, and repeat treatment if necessary.
- During an Epley manoeuvre, look for nystagmus during each position change. If the nystagmus continues in the same direction as original, that means that otoconia are moving further and further away from the posterior canal ampulla, which is good.
- Be familiar with the log roll manoeuvre for horizontal canal BPPV—some posterior canal cases will convert during treatment, and it is best to be prepared for that.

Conclusion

BPPV represents a common and treatable cause of vertigo. There is increased incidence in the elderly with significant implications due to the increased risk of falls. The pathophysiology is due to dislodged otoconia that falsely activate the semicircular canals. It is a clinical diagnosis that can be made based on characteristic patterns of nystagmus. Posterior canal BPPV is the most common form and the characteristic upbeating torsional nystagmus can be elicited with the Dix–Hallpike manoeuvre. It responds well to treatment with either the Epley manoeuvre or the Semont manoeuvre. Other forms of BPPV can present in the horizontal or superior canal, but are far less common.

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16

Audiovestibular implants

Neil Donnelly and Patrick Axon

Introduction

Rehabilitation of hearing loss is determined by both the nature and the severity of the loss. The past decades have seen a wide range of implantable hearing rehabilitation options develop. While air conduction hearing aids (ACHAs) are the standard of care and provide excellent amplification for the majority of patients, there are occasions where they are not suitable. In this situation, there are a range of implantable auditory device: these include bone conduction hearing devices (BCHDs), active middle ear implants (AMEIs), cochlear implants (CIs), and auditory brainstem implants (ABIs). These implantable devices rehabilitate hearing by interacting with different components of the auditory pathway.

The hearing implant team

It is now accepted that effective hearing implantation of a population requires a multidisciplinary team with knowledge of and access to a full range of hearing implants. There are many factors to consider when contemplating hearing implantation and a team incorporating a wide range of members covering different aspects of paediatric and adult care is essential. Teams will include audiologists, speech and language therapists, teachers of the deaf, psychologists, nurse specialists and otolaryngologists.

Patient selection

Patients are holistically assessed for implant suitability and then the results discussed by the full team taking into account all available information. This joint decision process ensures that patients are well informed about all options available to them as well as the associated risks and benefits of each. Assessment includes:

- medical history and physical examination
- pure tone audiometry to quantify the nature and severity of the hearing loss
- functional audiometry to assess speech discrimination in quiet and noise—this is usually undertaken using the patient's ACHA
- objective audiometry such as brainstem auditory responses and cortical evoked responses
- vestibular function tests
- imaging.

Relationships between the implant centre and referring regional departments are aimed at helping to identify as many potential implantees as possible so it is expected that some will fall outside implant criteria and be better served by optimizing hearing aid function. Some patients will decline surgery because they were unaware of the implications of surgery and the commitment required for post-surgery hearing rehabilitation. Others might benefit from middle ear surgery.

Guidelines

All hearing implants in the UK are governed by NHS specialist commissioning policies.

The current NHS England policy for BCHDs and AMEIs was published in July 2016 and makes these devices available to patients for whom:

- ACHAs are not suitable or do not provide adequate benefit, *and*
- have a level of hearing loss that falls within the manufacturer's guidelines.

Cochlear implantation is commissioned according to the National Institute for Health and Care Excellence criteria and was last revised in 2018:

- In both adults and children, the audiometric requirement is hearing thresholds that are worse than or equal to 80 dB hearing level at two or more frequencies (500, 1000, 2000, 3000, and 4000 Hz) in the better-hearing ear.
- The functional hearing requirement between adults and children differ:
  - Adults need an Arthur Boothroyd word list score of less than 50%, where presented at 70 dB sound pressure level in best ACHA conditions.
  - Children require speech, language, and listening skills non-appropriate for age after a 3-month ACHA trial.

ABIs are commissioned differently, depending on whether they are for individuals with neurofibromatosis type 2 (NF2) or whether they are for congenitally deaf children with cochlear malformations or cochlear nerve agenesis.

Bone conduction hearing devices

BCHDs are hearing implants that rely on the acoustic properties of bone to transmit sound with minimal attenuation directly to the cochlea, bypassing the outer and middle ear. Surgical BCHDs consist of an implanted component and an external sound processor. They are categorized into either direct-drive devices or skin-drive devices.

Direct-drive devices vibrate the skull via direct contact with the bone: they can be further subdivided into percutaneous or transcutaneous, dependent upon whether the skin is intact or not.

- Direct-drive percutaneous devices include the Cochlear Baha Connect System and the Oticon Ponto System. These rely on a titanium implant that is sited within the bone of the skull and forms a secure interface with the bone via a process known as osseointegration. A percutaneous abutment attaches to the titanium screw. The direct-drive is delivered by the sound processor which connects to the abutment.
- Direct-drive transcutaneous devices include the MED-EL Bonebridge, the Cochlear Osia and the Oticon Sentio. These consist of an internal receiver stimulator package that is placed subcutaneously and anchored to the skull using different techniques depending on the implant. The external sound processor connects to the receiver stimulator magnetically through the intact skin, providing power and processed sound. The direct drive is delivered via the active bone conduction implant rather than the processor.

Skin-drive devices are not directly in contact with the bone and instead transmit vibrations from the sound processor through the skin to an implanted magnet or directly to the bone. Devices include the Cochlear BAHA Attract system and the MED-EL Adhear. Such devices require normal bone conduction thresholds and are being superseded by direct-drive implants.

Implanted BCHDs are commissioned for use in adults and children (as per manufacturers' CE markings) who have:

1a. Unilateral or bilateral conductive or mixed hearing loss (**Box 16.1**) within the manufacturers fitting criteria, *and*

Stable bone conduction thresholds (<15 dB deterioration in greater than two frequencies in a 2-year period).

Or:

1b. Unilateral sensorineural hearing impairment (including single-sided deafness) where the better ear has bone conduction hearing thresholds within the manufacturer's fitting criteria.

And:

2. Trialled an ACHA or wireless contralateral routing of signal (CROS)/bi- contralateral routing of signal (BiCROS) hearing aid for a minimum of 4 weeks, or who are anatomically or physiologically unable to undertake a trial of an ACHA.

And:

3. Trialled a BCHD on a headband for a minimum of 14 days and show benefit in speech tests.

Box 16.1 Common causes of conductive hearing loss

- Aural atresia.
- Abnormality or fixation of ossicles.
- Fluid in middle ear.
- Eustachian tube dysfunction.
- Chronic suppurative otitis media with cholesteatoma.
- Tympanic membrane perforation.
- Iatrogenic following mastoid surgery.
- Trauma.
- Third-window effect.

Active middle ear implants

Middle ear implants are divided into active and passive implants. Passive implants are ossiculoplasty prostheses that attempt to restore middle ear mechanics by coupling the tympanic membrane to remnants of the ossicular chain or cochlea. AMEIs are devices placed in the middle ear that transduce sound into mechanical energy to drive the ossicular chain or cochlear windows. This is achieved by coupling to the ossicles or cochlear windows in what has become known as vibroplasty. Vibroplasty is defined as the treatment for hearing loss via vibratory stimulation in the middle ear.

The most widely used AMEI worldwide is the MED-EL Vibrant Soundbridge. The Vibrant Soundbridge comprises an external part, the sound processor, and an implanted part, the vibrating ossicular prosthesis. The sound processor is worn on the head and contains the microphones, the digital signal processing unit, and a zinc–air battery as the power source. The implantable aspect of the Vibrant Soundbridge consists of a receiver coil, a conductor link, and a FMT. The receiver coil is placed beneath the scalp. The conductor link connects the receiver to the FMT, a small electromagnet, that in turn is attached to the ossicles or round window membrane via couplers (**Figure 16.1**). The choice of coupler is dependent upon the nature of the deafness and available anatomy but a rule of thumb is that the preference, in decreasing order, is short process of incus then long process of incus followed by stapes and finally round window.

AMEIs are commissioned for use in adults and children over 5 years of age (or as per the manufacturer's CE markings) who have:

- unilateral or bilateral conductive, mixed, or sensorineural hearing loss (**Box 16.2**) within the manufacturer's fitting criteria, *and*
- middle ear anatomy suitable to accommodate an AMEI as determined by radiological and audiometrical testing (**Figure 16.2**), *and*
- stable bone conduction thresholds (<15 dB deterioration in greater than two frequencies in a 2-year period), *and*
- trialled an ACHA or wireless CROS/BiCROS hearing aid for a minimum of 4 weeks, or who are anatomically or physiologically unable to undertake a trial of an ACHA.

The direct drive of the AMEI has the following advantages over the indirect drive of the conventional ACHA:

- Elimination of occlusion effect.
- Reduced feedback.
- Improved high-frequency sound fidelity.
- More natural sound quality.

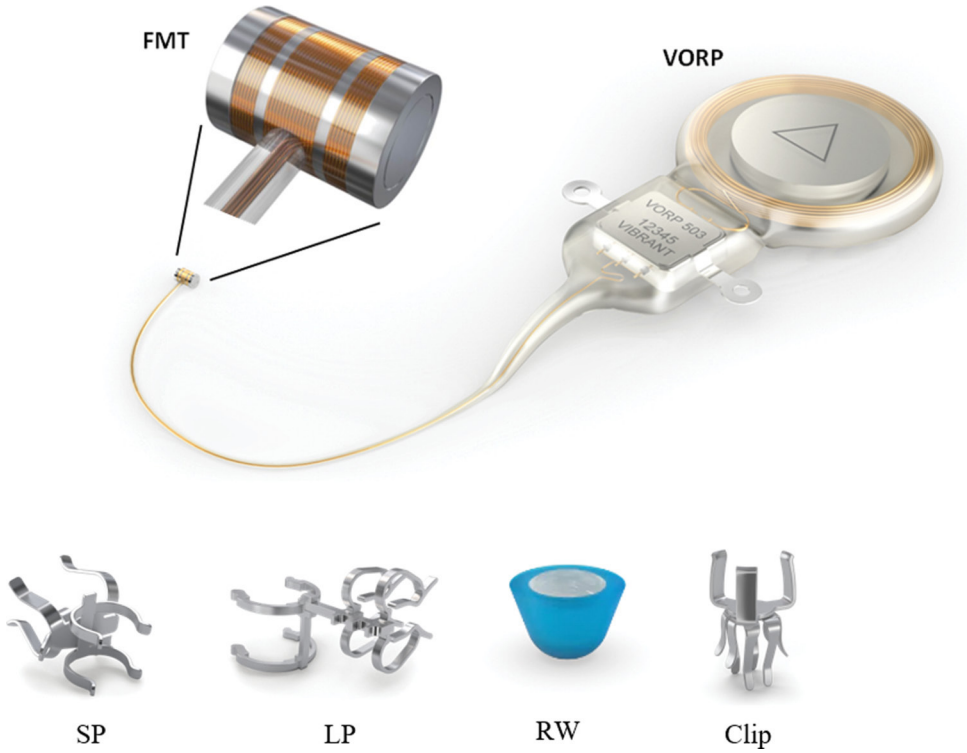

Figure 16.1 Vibrant Soundbridge (MED EL) AMEI and couplers.

Clip, clip coupler to stapes head; FMT, floating mass transducer; LP, long process of the incus coupler; RW, round window coupler; SP, short process of the incus coupler; VORP, vibrant ossicular prosthesis.

Images courtesy of MED-EL UK Ltd.

Box 16.2 Common causes of sensorineural hearing loss

- Genetic.
- Infection.
- Age.
- Noise.
- Trauma.
- Drugs.
- Tumour.
- Autoimmune.
- Iatrogenic.
- Ménière's disease.
- Neuropathy.
- Idiopathic.

Cochlear implants

CIs unlike ACHAs, BCHDs, and AMEIs rehabilitate hearing by electrically stimulating the cochlear nerve, rather than amplifying and an acoustic stimulus.

A variety of CI manufacturers have devices on the market: these have, over time, converged on an accepted implant design bar a few important differences highlighted as innovative by those manufacturers. As with BCHD and AMEI, CI systems consist of an external speech processor and an internal implant requiring surgical placement.

Implant design and surgical technique

The internal implant consists of a slim receiver stimulator package that is placed beneath the scalp. This is connected to a multichannel intracochlear electrode that is typically routed to the cochlea via a cortical mastoidectomy and posterior tympanotomy. The intracochlear electrode has differing numbers of electrical contacts dependent upon the manufacturer. Likewise, the position of the electrode in the cochlea—lateral wall, mid-scala, or perimodiolar—varies among manufacturers and reflects the ongoing debate as to which placement is superior. One important advance has been the development of the atraumatic electrode array. Different implant manufacturers have approached the problem of cochlear trauma and, as a result, reduced the risk of audiovestibular loss in different ways. What is accepted is that a thinner electrode array, incorporating variable stiffness along its length, reduces the risk of basilar membrane rupture.

Surgical technique has evolved over the past 20 years, with the aim of preserving cochlear anatomy and residual audiovestibular function. The term 'soft surgery' describes an approach that minimizes cochlear trauma. It has been helped considerably by implant electrode design but there are other crucial steps to maximize outcome. It is recognized that the best CI outcomes are achieved by a scala tympani placement of the electrode rather than a scala vestibuli insertion or where scala translocation arises as a result of electrode rupture of the basilar membrane. The decision whether to approach the scala tympani via the round window, a separate cochleostomy, or a combination of both is a surgeon's personal choice and can, on

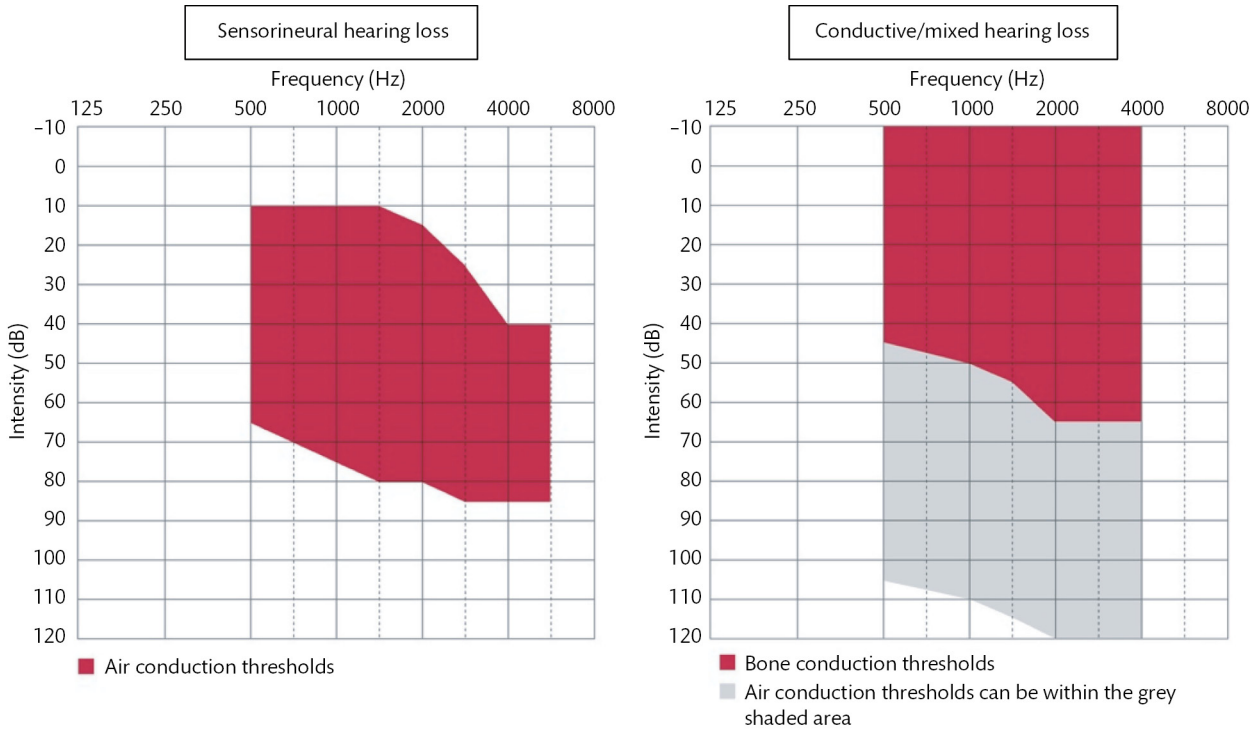

Figure 16.2 MED-EL audiometric indications for the Vibrant Soundbridge AMEI.
Image courtesy of MED-EL UK Ltd.

occasion, be dictated by electrode design. The use of intraoperative dexamethasone, both systemically and topically at the round window, is widely advocated. Other factors include the speed and trajectory of electrode insertion. There is now considerable evidence to support ultra-slow insertion over minutes, so reducing the risk of basilar membrane rupture and translocation of the electrode into the scala vestibuli.

The surgical complication rate is acceptably low and major complications are rare. Immediate postoperative vertigo, tinnitus, and taste disturbance are the most common as well as delayed implant infection, pain around the implant site, and implant failure, which do occasionally occur.

Speech processing strategies

The external speech processor contains the microphones, the digital signal processing unit, and the power source. It encodes incoming speech sounds and ensures that stimulation is delivered via the implant at an appropriate and comfortable level to each stimulating electrode. Historically, the objective of CIs has mainly been to improve speech intelligibility which is determined by spectral and temporal characteristics of the acoustic signal. The spectral information is delivered by the multichannel electrode array, coarsely representing the auditory system's natural tonotopic organization from low to high frequency in spatial progression along the length of the cochlea. Temporal speech information is commonly classified into three categories: (1) the speech envelope, defined as the fluctuations in overall amplitude at rates between 2 and 20 Hz; (2) the periodicity from around 50 to 500 Hz, usually due to the fundamental frequency (F0); and (3) temporal fine structure. Temporal fine structure can be defined as the variations in wave shape within single periods of periodic sounds, or over short time intervals of aperiodic ones. It has dominant fluctuation rates from around 500 Hz to 10 kHz.

Speech processing strategies have moved from simple feature extraction strategies, focusing on estimates of F0 and formants F1 and F2 of speech signals to continuous interleaved sampling. Continuous interleaved sampling runs a spectral analysis of the pre-processed digital input sound signal performed by a bank of band-pass filters or a fast Fourier transform. The filters have partially overlapping frequency responses and bandwidths that generally become broader with increasing frequency. Each filter is assigned to (at least) one intracochlear electrode following the frequency-place tonotopic organization of the cochlea.

CI development has undoubtedly benefited from the advances in hearing aid technology. Most CI manufacturers have a close relationship with one of the major hearing aid companies and advances in microphone technology, speech processing strategies to help in complex listening environments, miniaturization, as well as Bluetooth connectivity are the direct result of those relationships.

Postoperative CI programming requires the audiologist to establish both the lowest level of electrical stimulation that produces an auditory sensation (threshold or T-level), and the highest comfortable level (C-level) for each of the stimulating electrodes. These values, often expressed in terms of stimulation-level units that relate to the current and pulse-width delivered, are stored as a 'map' in the speech processor. Post-lingually deafened individuals are usually able to describe the sounds they are perceiving and to report loudness growth, pitch changes, and so on. In pre-lingually deaf individuals, the programming is inevitably slower and more time-consuming. In the very young child, a conditioned response to sound may not be established and the audiologist must therefore rely on observation of

involuntary responses to stimulation, such as stilling, blinking, head turns, and so on. Modified visual reinforcement audiometry can sometimes be used to obtain estimates of threshold levels. Accurate estimation of the highest comfortable levels is frequently difficult in young children. All current implant designs incorporate a form of neural response telemetry that provide both evidence of functioning electrodes and estimated C-levels. More complex algorithms can now accurately predict residual auditory hearing post-implantation which helps which helps optimize performance by 'tailoring' electrical stimulation to match the neural conditions in the cochlea.

Auditory brainstem implants

ABIs are considered for patients who have profound hearing loss and where cochlear implantation is not possible. The most common recipients are children or adults who have bilateral vestibular schwannoma as a result of the autosomal dominant disease NF2. On occasion, these patients will undergo tumour resection which can result in sacrifice of the auditory nerve. Other rarer indications include:

- adults who have post-meningitis cochlear ossification (labyrinthitis ossificans)
- labyrinthitis
- fractures
- otosclerosis
- children with bilateral cochlear aplasia or bilateral auditory nerve aplasia or dysplasia.

The ABI has similar components to the CI, the main difference being that the electrode contacts are arranged on a flat paddle. The retrosigmoid or translabyrinthine approaches are typically used for access to the brainstem and the electrode paddle is placed in the lateral recess of the fourth ventricle adjacent to the cochlear nucleus. Paddle position can be optimized based on implant-evoked ABR responses.

Hearing outcomes from ABIs are highly variable and are certainly inferior to CIs. In addition, it takes many years for patients to reach peak performance. In most cases, they allow awareness of sounds and an aid to lip reading, although a minority can achieve open-set speech recognition. Outcomes in adults and children without tumour-related hearing loss are generally superior to those who have NF2 and this might be because of tumour-related brainstem distortion making paddle placement more difficult and also difficult hearing rehabilitation as a result of other NF2-related morbidity.

Outcomes

The child

A shorter duration of deafness is associated with better outcomes regardless of the nature and severity of the hearing loss. Early rehabilitation of any hearing loss is key. In the paediatric population, audiology and implant teams accept that the best long-term outcomes in children are likely to be achieved if rehabilitation occurs at a very young age: the goal being to limit the period of auditory deprivation. In cochlear implantation, it is now the aim of most implant centres to perform bilateral simultaneous implantation between the age of 9 and 12 months. This drive to early implantation has been very much aided by the introduction of the UK Newborn Hearing Screening Programme in 2006.

The goal for children with aural atresia is for early stimulation of the atretic ear with a BCHD on a headband from 6 months of age. Definitive BCHD or AMEI implantation is then undertaken as early as possible, typically at 4–5 years of age dependent upon the device choice.

The adult

There is increasing published evidence that hearing loss is associated with higher rates of dementia and the incidence of dementia increases proportionately to the severity of the loss. There is also evidence that appropriate hearing rehabilitation can decrease the speed of progress of dementia and in some cases result in some improvement in cognition.

Cochlear implantation is incredibly beneficial to most recipients leading to the majority acquiring good open-set speech recognition. There is, however, a small but significant subset of implant users that only gain marginal benefit, which may amount to awareness of sounds, enhanced lip reading, and reduced awareness of tinnitus, with no open-set speech recognition. Predicting outcome is notoriously difficult and much energy must therefore be spent on a few struggling implant recipients, drawing out the best possible function by optimizing the speech processor map and encouraging them to persevere with auditory rehabilitation exercises. Speech and hearing therapists are an indispensable part of the implant team in this regard.

Magnetic resonance imaging compatibility

Magnetic resonance imaging (MRI) is an important and rapidly expanding area in diagnostic imaging. It is regarded as the standard of care for soft tissue imaging, in particular that of the brain and spinal cord. It is likely that a majority of individuals will require an MRI scan at some stage during their life and as such it is an important consideration when discussing implantable hearing devices.

The principal concerns regarding the use of MRI in the presence of hearing implants are that:

- powerful magnetic fields may demagnetize or damage the electrical components in the implants
- radiofrequency energy may result in heating that can cause tissue damage
- the artefact caused by the implant may impair the diagnostic usefulness of the scan.

Implants can be divided into MRI conditional, posing no hazard when placed in a specified MRI environment and accepted protocols are used, and MRI unsafe, posing a hazard in all MRI environments.

Significant manufacturer developments have occurred in this field so that we are now able to offer an MRI-conditional version of all devices. The percutaneous direct-drive and skin-drive BCHDs are conditional in MRI scanners up to 3 tesla. The Bonebridge is MRI conditional to 1.5 tesla, as is the latest version of the Vibrant Soundbridge. New magnet technology has also led to the development of 3-tesla MRI-conditional CIs and ABIs. It must be remembered that older CI and ABI devices are only 1.5-tesla MRI

conditional and require specific bandaging protocols to prevent magnet displacement and significant patient discomfort or harm.

Conclusion

Developments in hearing aids and implantable hearing devices over the past 30 years have facilitated a more tailored and nuanced approach to hearing rehabilitation depending on patient need and choice.

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17

Noise-induced hearing loss

Peter Webber

Introduction

Noise is the term generally used to describe the range of sounds from nuisance to harmful. Sound is a mechanical wave, caused when energy is expressed which causes particles to move and collide with each other. Noise is unwanted sound; as pointed out by Dobie,[1] a Bach fugue, played loud enough and long enough, can be as hazardous to the ear as the noise from a chainsaw. This particular point has relevance to a recent legal case in England, discussed later.

Inner ear injury from noise has been well described in human postmortem studies and from experimental animal studies. The most vulnerable structures in the cochlea are the outer hair cells; initially, the hair cell stereocilia lose their stiffness and hence the ability to vibrate, resulting in a temporary threshold shift. With continuing noise exposure, the hair cells die, and a permanent threshold shift occurs.[2] With continuing damage through noise, inner hair cell and auditory nerve fibre loss occurs as well. This is believed to be due to metabolic issues, rather than direct trauma, at least for occupational noise-induced hearing loss (NIHL).

History

Pliny the Elder (29–79 AD), a Roman naturalist and historian writing during the early first century, noted that noise was a workplace risk for fishermen of the upper Nile. A high rate of deafness among these workers was attributed to the constant noise produced by the rapids and waterfalls that the fishermen worked and lived among.[3] NIHL was seen in the Middle Ages among miners and bell ringers, who were known to become deaf in their later years as a result of their work.[4]

During the Victorian era, the affliction of NIHL was known as blacksmiths' or boilermakers' deafness.[5] During this period the Industrial Revolution introduced mechanization and mechanically powered equipment to factories throughout the developed world, making high levels of noise exposure a daily reality for much of the working class. Acute acoustic trauma was also recognized in the nineteenth century, with Toynbee[6] describing a patient with hearing difficulties after being exposed to a single pistol shot. However, the now well-known feature of noise damage producing a high-frequency sensory hearing loss was not appreciated until the development of the electric audiometer.[7]

While originally seen as an unavoidable consequence of employment or even a natural form of protection from further noise,[8] NIHL is now viewed as both undesirable and, to a considerable extent, preventable. However, despite the knowledge of the primary cause-and-effect relationship of occupational deafness for over two millennia, NIHL still exists and is very much a modern problem.

Definitions

Occupational NIHL is a slowly developing hearing loss, usually occurring over many years with continuing noise exposure; it is usually bilateral, and not usually profound. As the hearing threshold increases because of noise exposure, the rate of loss decreases. Following cessation of exposure, or adequate protection from noise, there is no significant further progression of the hearing loss due to the previous noise exposure. The earliest damage to the inner ear affects 3, 4, and 6 kHz, with the greatest loss generally occurring at 4 kHz.[9]

Acute acoustic trauma is a sudden and immediate change in hearing as a result of a single exposure to a loud noise, usually impulse or blast wave noise.[10] Unlike with occupational NIHL, there is no intervening temporary threshold shift. Although the frequencies affected can be similar to occupational NIHL, a variety of other audiogram shapes can be produced, of which the commonest are down-sloping or flat audiograms.

Acoustic shock refers to the combination of exposure to a brief, sudden, unexpected, high-frequency, high-intensity sound emitted (the stimulus) and the subsequent symptoms (the response) which can develop (Health Services Australia Group definition—there are several other definitions of this condition). Hearing loss does not have to be present. A wide-frequency spectrum can be causative, and the sounds are not usually those of occupational NIHL or acute acoustic trauma.

The law in England and Wales regarding occupational noise

In 1963, the Government published 'Noise and the Worker', which highlighted the dangers of excessive noise, and what measures should be taken to protect employees from excessive noise exposure. Subsequently, the judgement in Thompson v Smith Ship Repairs (1984) held that any employers from 1963 onwards who had exposed their employees unprotected to noise in excess of 90 dB were in breach of their duty of care (NB: there are exceptions to the '1963' rule, e.g. British Railways (1955); Shipyards (1957); Armed Forces (1987); the entertainment industry (2008)).

1972 Code of Practice

This represented guidance and not statutory regulation but did provide the framework for the duty of employers under common law, to ensure that employees were not exposed unprotected to continuous noise levels in excess of 90 dB.

The Noise at Work Regulations 1989

This replaced the 1972 Code of Practice, and stated that from 1 January 1990 the following applied:

- First action level: ear protection to be provided on a voluntary basis/or when requested by an employee, where the daily noise exposure is 85 dB.
- Second action level: ear protection compulsory where the daily noise exposure reaches and/or exceeds 90 dB.

The Control of Noise at Work Regulations 2005

These are the current regulations. In brief, these regulations from 6 April 2006 reduced the first action level from 85 dB to 80 dB, and the second action level from 90 dB to 85 dB. There is also a requirement for the employer to calculate weekly noise levels, where there is day-to-day variability in levels of noise. Employers also have a duty to carry out risk assessments and noise surveys, reduce noise levels to the lowest practicable levels, instigate defined hearing protection zones, carry out health surveillance, and provide appropriate training for employees.

Diagnosis

It is easy to make a diagnosis of occupational NIHL where there is a history of unprotected exposure to high levels of noise over a long period of time, and an audiogram demonstrating a notch in both ears at 4 kHz. However, diagnostic competitors, and the effects of age-associated hearing loss obscuring the features of noise damage, are just two factors which can complicate the diagnosis. Personal susceptibility to noise damage and the hearing thresholds prior to noise exposure are also complicating factors. Additionally, as often occurs in the medicolegal field, audiometry performed many years after the period of noise exposure in question has ceased, together with the matter of audiometric variability on repeated testing, will further complicate the attempt at diagnosis (**Figure 17.1**).

Because of the above issues, particularly in the medicolegal context, for the need to be able to identify a readily measurable amount of NIHL on the audiogram, the guidelines published in 2000 by Coles, Lutman, and Buffin[11]—the CLB guidelines—have become the cornerstone of medicolegal practice in the UK in the diagnosis of NIHL. The principles of diagnosis, however, are common to other methods. (e.g. the American Medical Association guides).

The CLB guidelines generally applied to what can be termed industrial noise exposure, that is, predominantly steady-state noise exposure and not to acute acoustic trauma, or to sounds predominantly tonal, or low/very high frequency in spectrum.

In brief, the guidelines comprise three requirements:

R1 A high-frequency sensorineural hearing loss on the audiogram: where the measured hearing threshold level (HTL) at 3, 4, or 6 kHz, is at least 10 dB greater than the HTL at 1 or 2 kHz.

R2 An adequacy of noise exposure sufficient to be likely to give rise to NIHL. This is considered to be equivalent to a daily (8 hours) of noise exposure of not less than 85 dB(A) for a sufficient number of years to lead to a cumulative exposure of not less than 100 dB NIL—the Noise Immission Level. This defines R2(a). R2(b) recognizes that substantial NIHL can be caused in certain susceptible individuals exposed to less than 100 dB NIL. In such cases the cumulative noise exposure is reduced to 90 dB(A) NIL. The estimation of the NIL should be undertaken by an expert in acoustic engineering, but it is often the case that the claimant's recollection of the noise exposure can be used instead, without such an expert report.

R3 Evidence for NIHL is considered to be present if there is a downward notch or bulge in the range 3–6 kHz large enough to be identified, that is, 10 dB for R3(a), and 20 dB for R3(b), both contingent on R2(a) and R2(b) being met respectively.
A notch can be simply identified from the audiogram by way of the HTL at 3 and/or 4 and/or 6 kHz being at least 10 dB (R3(a)), or 20 dB (R3(b)), greater than at 1 or 2 kHz and at 6 or 8 kHz.
By contrast, a bulge (**Figure 17.2**) is defined as the HTL being at least 10 or 20 dB greater at the above higher frequencies relative to the comparison values for age-related hearing loss. Although this can simply be identified by plotting the expected age-related hearing loss on the audiogram against the measured HTL at the higher frequencies, the CLB guidelines refine this further by individualizing the hearing losses for each ear.

This calculation relies on the use of 'anchor points' at 1 and 8 kHz; both these frequencies are less susceptible to the effects of steady state noise. A closer approximation to the actual percentile of age-related hearing loss on which the individual sits, can thus be obtained. There is usually a difference between the statistical figures for the age-related hearing loss and the measured HTL at the anchor points. From this difference is derived the so-called misfit values at the frequencies between 1 and 8 kHz, which are then applied, producing an adjusted age-related hearing loss at the measured frequencies. These adjusted values are then compared with the measured HTLs at the higher frequencies to determine the presence of a bulge. There are a number of additional factors which may need to be taken into account when using the CLB guidelines, including so-called modifying factors, which include such issues as the clinical picture (to include any diagnostic competitors for NIHL, as well as factors which could give rise to a hearing loss additional to ageing and NIHL), age and type of noise exposure, the application of Robinson's probability tests, and complicated cases including audiometric asymmetry and the presence of conductive hearing losses. Detailed discussion of these matters lies outside the scope of this chapter.

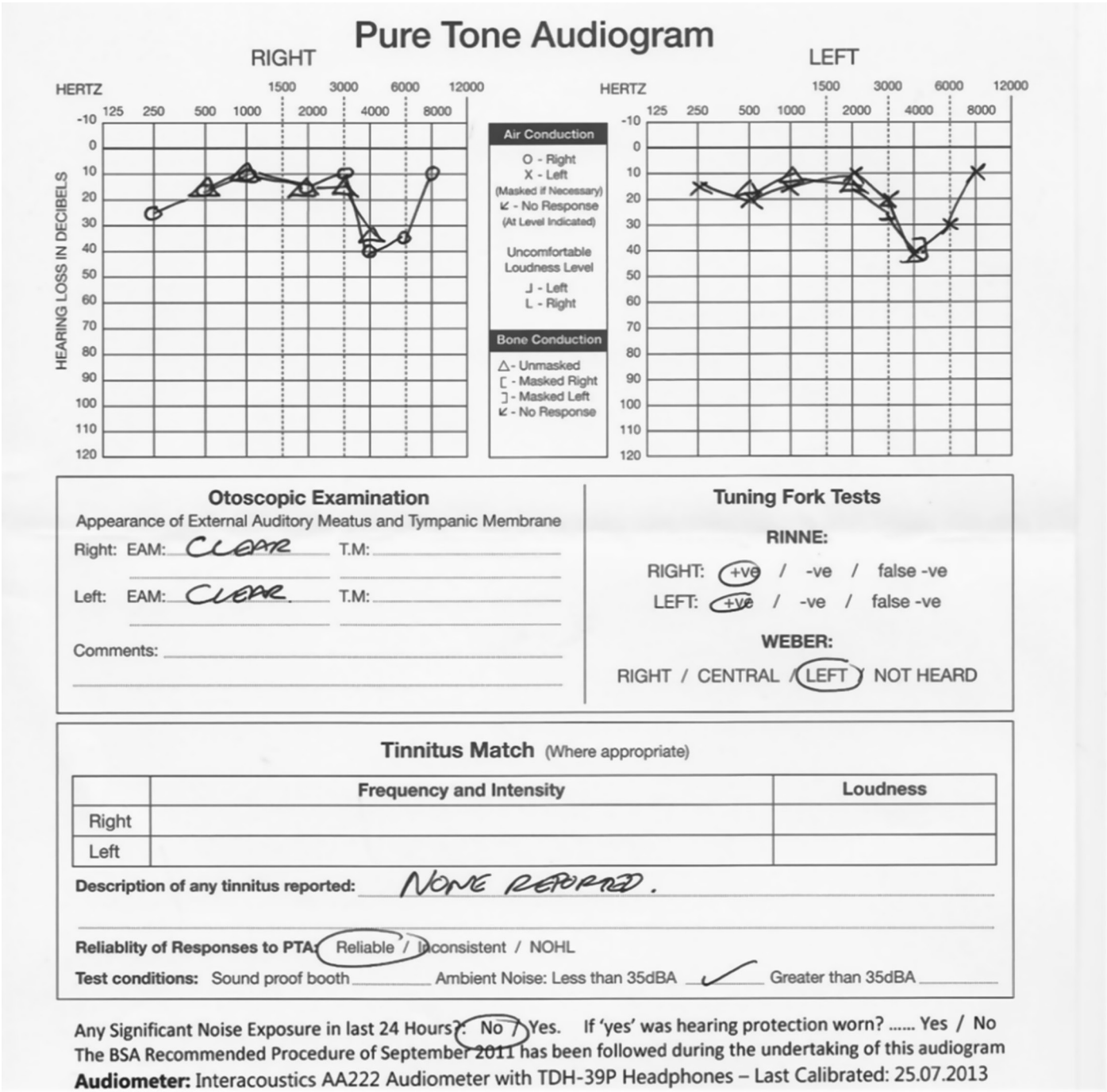

Figure 17.1 Medicolegal audiogram exhibiting notches bilaterally at 4 and 6 kHz.

In recent years, there has been interest in the study by Kujawa and Liberman[12] who postulated that noise-induced damage to the ear has consequences that are considerably more widespread than revealed by conventional threshold testing (i.e. pure tone audiometry). This damage is stated to be due to damage and loss of presynaptic hair cell nerves in the inner ear and postsynaptic cochlear nerve terminals despite hearing thresholds returning to normal after temporary impairment. Kujawa and Liberman suggested that noise-induced cochlear synaptopathy in humans could explain the hearing difficulties experienced by individuals in noisy environments who have normal hearing on audiometry.[12] This could therefore be extrapolated to state that even where no material loss due to NIHL can be demonstrated on audiometry, cochlear synaptopathy could be responsible for the perceived impairment.

However, the above hypotheses were based on animal studies, as pointed out by Beach.[13] Unlike the animal studies, humans do not come from one genetic strain, and the amount and extent of noise exposure is usually highly variable in terms of intensity, duration, and source. The research by Beach and others suggests that noise-induced cochlear synaptopathy in humans is highly variable and is likely to be only one of several factors determining the ability to perceive speech in the presence of noise.

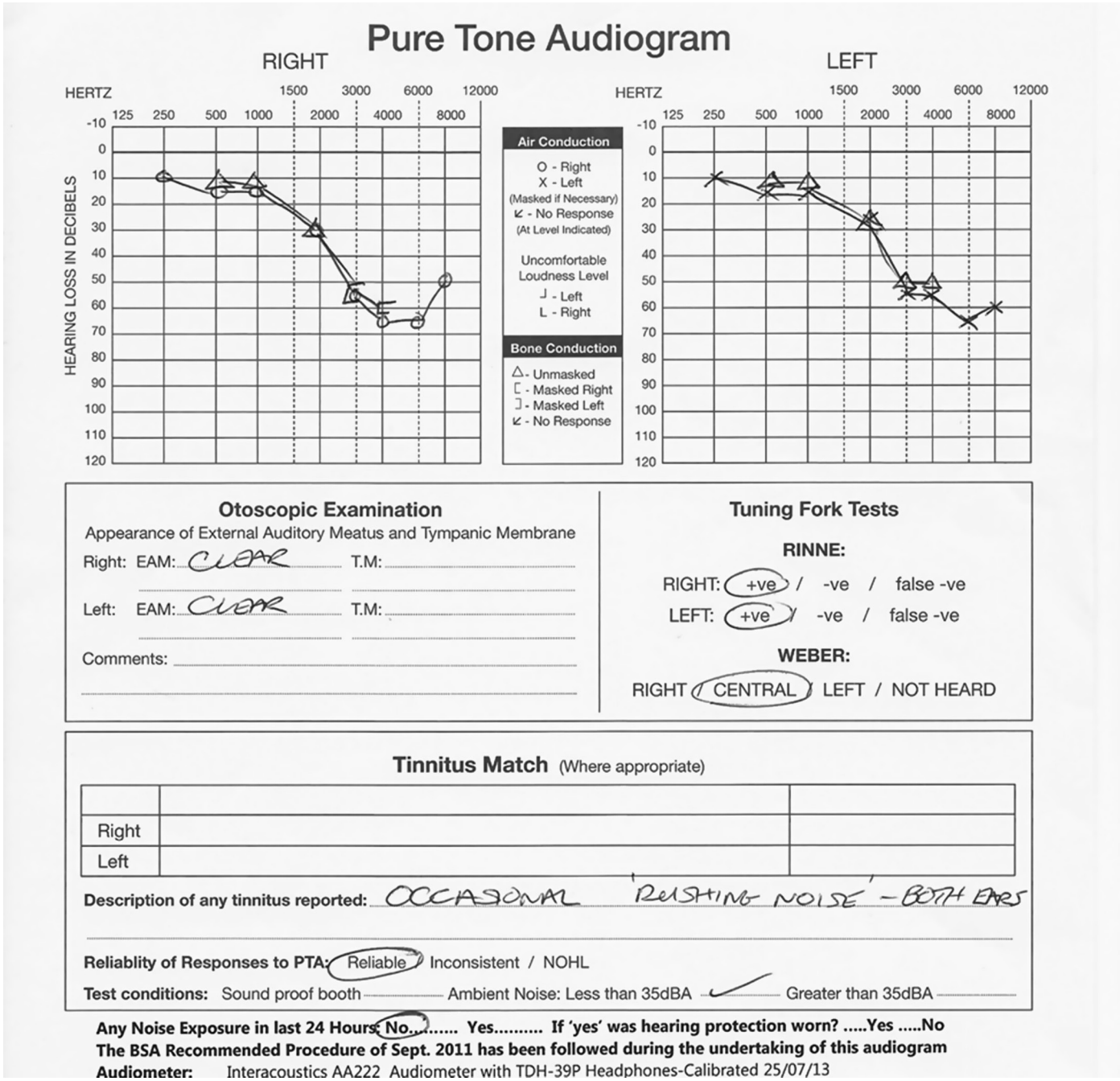

Figure 17.2 Medicolegal audiogram, 60-year-old male, demonstrating audiometric bulge (straight line = expected age-related hearing loss) Modified from ISO 7029.

Quantification of noise-induced hearing loss

As with much to do with occupational NIHL, this is primarily of medicolegal interest, in assisting the determination by the courts of the value of a hearing loss claim.

The conventional method in the UK has been the so-called DSS or DHSS method. This method has been in existence (or at least variants of it) from 1973. It was first adopted in 1973 with the then DHSS statutory compensation scheme for occupational deafness but was first set out explicitly in the Social Security (Industrial Injuries) (Prescribed Diseases) Amendment (No. 2) Regulations 1983 at Schedule 2A Part III.

This method is applied to 1–3 kHz. First, the overall binaural hearing loss is calculated for the averaged threshold value of these frequencies, weighted towards the better-hearing ear. The averaged value for the expected age-related hearing loss, conventionally based on the 50th percentile, by reference to the age-related hearing loss tables in ISO 7029, is then deducted from the overall binaural hearing loss. What remains is considered to be the NIHL.

The limitation of this method is that it assumes no other diagnostic competitors, and as such, will usually tend to overestimate the NIHL. For example, idiopathic sensorineural hearing losses can commonly affect the lower frequencies, particularly in the older

population,[14] and by applying this calculation in such a case, the NIHL will be exaggerated.

A newer method of quantification was published in 2015 by Lutman, Coles, and Buffin—the LCB guidelines.[14] This method is based on the CLB guidelines, with the addition of estimating the contribution of NIHL at the 1 and 8 kHz anchor points. To do so, a NIHL model is used to estimate the likely NIHL; this is based on the 1974 meta-analysis by Passchier-Vermeer,[15] specifically audiogram graphs showing the varying median NIHL after 10 years of exposure to four differing levels of noise. The calculations to adjust the threshold values at 1 and 8 kHz are carried out by reference to the additional hearing loss (bulge) at 4 kHz, and adjusting the anchor point thresholds at 1 and 8 kHz by multiplying the bulge value at 4 kHz (by a factor of ×0.15 and ×0.4 respectively), and applying the result to adjust the threshold values at the anchor points. The calculation then proceeds as with the CLB calculation. The resulting additional hearing losses are then averaged as with the DHSS calculations, weighted towards the better-hearing ear.

There are limitations with this method, chiefly where the individual has been exposed to very high levels of noise, and also where noise exposure has exceeded 10 years. Further, although 4 kHz is generally the most noise-sensitive frequency, this is not invariably so, and in cases where the maximal noise damage is at 3 kHz, the use of this method will probably underestimate the NIHL. Additionally, this method, predicated on additional hearing losses at 4 kHz, assumes noise damage at 1 and 8 kHz; by contrast, particularly at the lower anchor point, NIHL tables ISO 1999, for example, suggest that noise damage will only arise at 1 kHz after a daily noise exposure to at least 95 dB(A) for at least 10 years.

A less used quantification method is the median-based allocation method. This sums the expected age associated hearing loss and the expected NIHL (from ISO 1999), expressed as a median value over 0.5, 1, 2, and 3 kHz. This is then set against the measured HTL and adjusted dependent on the relative percentages of age and noise based on the relevant statistical tables.

As noted above, estimation of the impact of NIHL on hearing, at least in the medicolegal sense, has traditionally concentrated on the 1–3 kHz frequencies. However, the 2016 review by Professor Moore[16] concluded that there was strong evidence that NIHL for frequencies above 3 kHz had adverse effects on the ability to understand speech, especially when background noise was present. Professor Moore also concluded that a good predictor of the ability to understand speech in noise for people with NIHL was the average audiometric threshold at 2 and 4 kHz. This last conclusion was based on the earlier work by Smoorenburg.[17] The range of speech served by the 4 kHz frequency is restricted mainly to the 's' sounds of certain speakers, and usually is only problematic when the threshold at that frequency exceeds 30 dB.

Professor Moore et al.[18] have also recently published an alternative set of guidelines for NIHL (2022), termed the MLC guidelines after the authors. The stated rationale for publication is that existing guidelines are inadequate in dealing with impulse noise, including military exposure (see below), or to very intense tones. Guidelines for the diagnosis and quantification of military noise, termed M-NIHL had previously been published by Moore[19] and is also covered by the MLC guidelines in respect of impulse noise. Comment has been made regarding the poorer specificity of these newer guidelines, leading to a higher false-positive rate of NIHL than with, for example the CLB guidelines.[20] This had been acknowledged by Moore himself in respect of M-NIHL, leading to publication of revised guidelines, rM-NIHL.[21] Further details lie outside the scope of this chapter.

Tinnitus

Tinnitus often accompanies NIHL. Up to 1% of adults have tinnitus that affects the quality of their life. The risk of tinnitus increases with age: 1% of people under 45 experience tinnitus, this increases to 25–30% in the over 70s. Tinnitus is more prevalent in those who have been exposed to noise compared with the general population. The general prevalence rate is 10%; however, in those exposed to noise the prevalence is anything from 23.3% to 87.5%.

It is widely accepted by experts that in the presence of hearing loss, whatever is the cause of the hearing loss is also likely to be the cause of any tinnitus. Some experts suggest that tinnitus should only be attributable to noise exposure if it comes on during a period of exposure, or within 1 year of cessation of exposure. Others suggest that tinnitus can be attributable to noise exposure no matter at what stage it starts following cessation of exposure to noise. Certainly, a 1-year cut off appears to be purely arbitrary, and it seems illogical to presume that an individual is any less likely to develop tinnitus after a longer duration of time; a number of well-regarded authors have stated that noise can result in tinnitus a number of years after noise exposure has ceased.[22,23]

Acute acoustic trauma

Acute acoustic trauma and NIHL are clinically distinct entities, probably with different mechanisms. The peer-reviewed literature on acute acoustic trauma almost exclusively concerns studies carried out in the armed forces.

Pure tone audiograms after acoustic trauma may show a similar pattern to that which is seen in occupational NIHL, and hence show notches or bulges over the 3–6 kHz frequency range. However, a variety of audiogram shapes can be seen. As previously stated, the down-sloping type is probably the most common.

Mrena et al.[24] showed that acute acoustic trauma was most commonly caused in service personnel by the assault rifle, with the acoustic trauma occurring in over 87% of cases where the ear(s) were unprotected. Immediately after the acoustic trauma, over 46% were suffering from hearing loss, and over 94% from tinnitus. At extended follow-up, hearing loss, tinnitus, or both were still experienced by 45%.

By way of example, Paddan and Lower[25] noted that the peak sound pressure produced by the standard rifle used by the British Army, the SA80, is estimated to be 137 dB(A) at a distance of approximately 20 metres to the left-hand side of the rifle. The authors stated that hearing protection should be worn by anyone closer than 20 metres to a person firing. Repeat sound pressure level was estimated to be 135 dB(A) to a distance of approximately 25 metres, and therefore hearing protection was recommended at distances up to 25 metres. At 20 metres, it was noted that the sound pressure exposure level of 98 dB(A) was sufficient to cause noise damage, after 1440 rounds had been fired. It is thus clear that an individual firing this rifle is exposed to very high levels of noise indeed.

According to Dobie,[1] exposure levels of 130 dB are sufficient to risk causing permanent hearing loss, after 5 minutes' exposure.

Acoustic shock

As well as hearing loss, it is stated that many other symptoms can occur with this condition. These include tinnitus, hyperacusis, otalgia, headache, pain or a burning sensation in the head, neck, or jaw on the affected side, depression, and anxiety.

Originally described in telephone headset users working in call centres,[26] the sounds are usually brief, and typically between 56 and 110 dB at 100 Hz to 3.8 kHz. However, there is, to date, no medical consensus on what constitutes acoustic shock syndrome, although McFerran and Baguley stated that acoustic shock was a 'recently recognised clinical entity'.[27]

Some sources have considered that the condition is primarily psychological.[28,29] Parker et al.[30] in a retrospective analysis of 30 cases, reported that 90% had tinnitus, 70% previous otological pathology, 17% a history of head injury, and 63% psychological symptoms.

Physical hypotheses supporting acoustic shock syndrome have included prolonged contraction of the tensor tympani muscle, although it has been argued that there is currently no scientific evidence to support that view.[31] A recent study examining changes in middle ear function in acoustic shock sufferers suggested middle ear inflammation may be a factor particularly in terms of pain and tinnitus.[32]

The recent legal case of Goldscheider v the Royal Opera House Covent Garden Foundation (2018) EWHC 687 (QB), stands out as it was the first case to involve a musician claiming acoustic shock as a result of exposure to excessive noise from brass instruments, rather than from electronic noise delivered directly to the ear through a headset.

The claimant won his case on two counts. Firstly, the Royal Opera House was found to be in breach of The Control of Noise at Work Regulations 2005: it was held that the Act did not differentiate between noise generated as a by-product in a factory, and noise (music) generated for enjoyment, by the Royal Opera House. Secondly, the Judge was minded towards accepting a diagnosis of acoustic shock, though mainly by a process of elimination, given that the medical evidence in this case supporting the diagnosis was deemed to be rather 'opaque'.

In conclusion, the weight of published evidence has been concerned with occupational hearing loss; acute acoustic trauma has received far less attention. The newer phenomenon of acoustic shock remains, at present, rather poorly defined.

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18

Lateral skull base disease

J. Walter Kutz Jr and Daniel E. Killeen

Introduction

Lateral skull base disease encompasses a wide range of conditions that may cause significant morbidity. These conditions include neoplasms, infections, and inflammatory conditions. Because of the compact anatomy of the lateral skull base with multiple cranial nerves and proximity to the carotid artery and jugular bulb, treating diseases of this area may be challenging with significant potential for morbidity with treatment as well as from disease. This chapter will focus on lateral skull base disease with a review of the relevant anatomy and the common disease processes that affect this complex area.

Anatomy

By understanding the anatomy of the skull base, the physician can begin to understand the possible location and nature of the disease (Figure 18.1).

Temporal bone

The temporal bone is divided into four portions: squamous, petrous, tympanic, and mastoid.[1] The squamous portion of the temporal bone forms the anterior superior portion of the temporal bone and is a thin plate of bone that connects to the parietal and sphenoidal bones. It joins the tympanic portion of the external

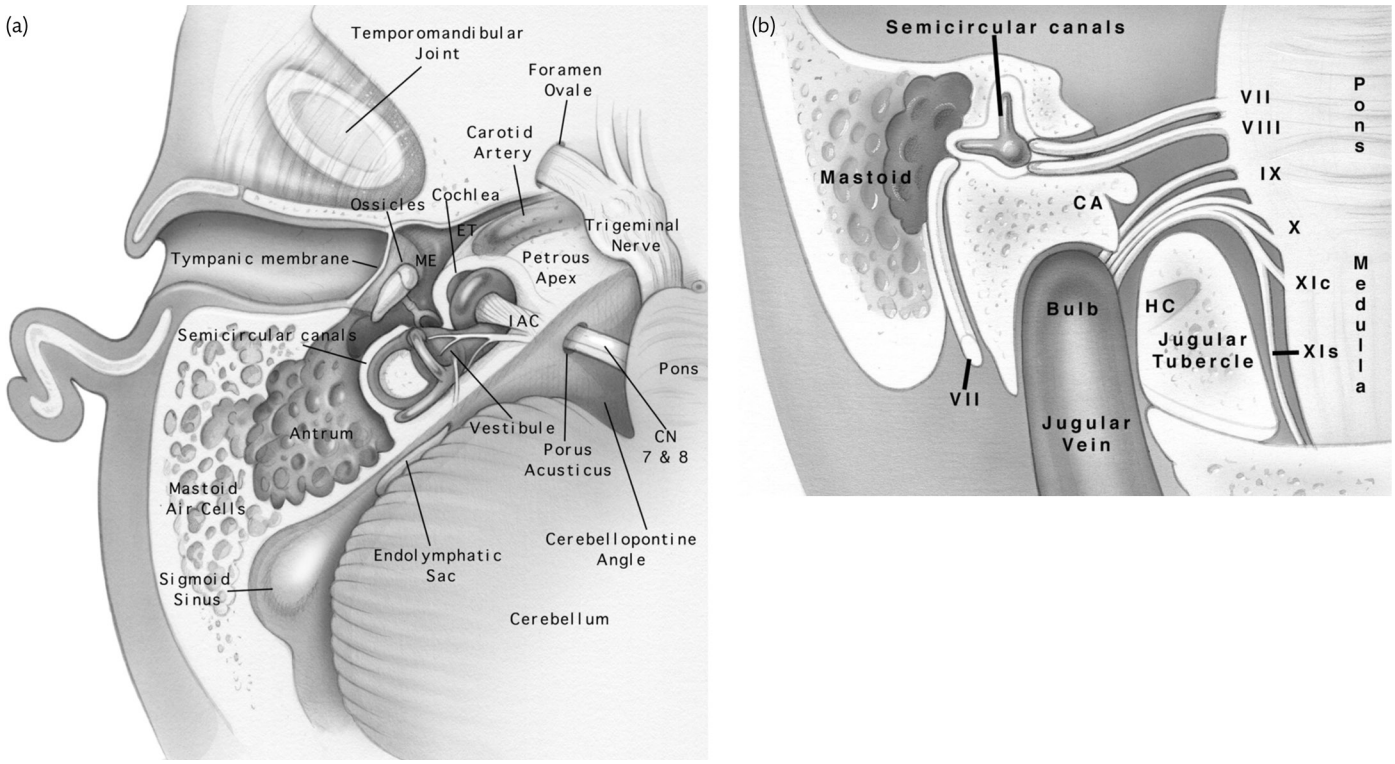

Figure 18.1 (a) Axial and (b) coronal illustrations showing the anatomy of the lateral skull base. A differential diagnosis for skull base disease can be formed by understanding the anatomy and locating the disease by the affected cranial nerves. CA, cochlear aqueduct; CN, cranial nerve; ET,

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auditory canal (EAC) at the tympanosquamous suture line. The mastoid portion of the temporal bone forms the posterior portion of the temporal bone and articulates with the occipital bone.[1] The medial border of the mastoid portion is lateral to the lateral semicircular canal.[1] The mastoid part connects to the tympanic portion at the tympanomastoid suture line of the EAC.[1] The EAC is composed of two portions, the lateral one-third of the EAC is encased in cartilage, and the medial two-thirds of the EAC is encased in bone.[1] The tympanic portion of the temporal bone encases the bony portion of the EAC and the tympanic membrane.[1] The petrous portion of the temporal bone forms the medial portion of the temporal bone, containing the inner ear, petrous carotid artery, internal auditory canal, and jugular bulb.[1] The petrous portion of the temporal bone is shaped like a pyramid with a base facing laterally, and the apex, which may either be pneumatized or filled with marrow, positioned anteriorly and medially.[1]

Cranial nerves

Understanding the anatomy and function of the cranial nerves will allow for more precise localization of disease based on impaired cranial nerve function. (Table 18.1) The most common cranial nerves affected by lateral skull base diseases are cranial nerves V–XII.

Deficits in cranial nerve V (trigeminal nerve) will cause facial numbness, pain, or paraesthesia in the distribution of the affected branches. The trigeminal nerve exits the pons and travels to the trigeminal ganglion in Meckel's cave.[2] The trigeminal nerve then divides into three branches: the ophthalmic (V_1), the maxillary (V_2), and the mandibular (V_3) branches.[2] The ophthalmic branch then courses through the lateral wall of the cavernous sinus, exiting through the superior orbital fissure, the maxillary branch then briefly courses through the lateral wall of the cavernous sinus leaving through the foramen rotundum, and the mandibular branch does not enter the cavernous sinus and leaves the skull base through the foramen ovale.[2]

Deficits of cranial nerve VI (abducens nerve) will cause diplopia with lateral gaze to the affected side.[2] The abducens nerve leaves the brainstem at the junction of the pons and the medulla medial to cranial nerve VII. It passes through Dorello's canal near the petrous apex, entering the cavernous sinus and exiting through the superior orbital fissure.[2]

Table 18.1 Presenting symptoms of lateral skull base lesions

Location	Symptoms
Internal auditory canal and cerebellopontine angle	Common: hearing loss, imbalance, tinnitus Possible: facial paresis, facial numbness Rare: diplopia, hoarseness, dysphagia
Jugular foramen	Common: hoarseness, dysphagia, dysarthria, shoulder weakness Possible: hearing loss, imbalance, tinnitus, facial paresis Rare: diplopia, facial numbness
Facial nerve	Common: facial weakness Possible: hearing loss, imbalance, tinnitus, conductive hearing loss
Petrous apex	Common: asymptomatic, headaches Possible: retro-orbital pain, facial pain or numbness, diplopia, hearing loss, imbalance, tinnitus, otorrhoea

If cranial nerve VII (facial nerve) is affected, the patient will present with facial paresis or paralysis.[2] The facial nerve has a complicated course, emerging from the junction of the pons and the medulla and passing through the internal auditory canal with cranial nerve VIII.[2] The facial nerve then transitions to the labyrinthine segment of the facial nerve and travels anteriorly and laterally to the geniculate ganglion.[2] At this point, the facial nerve gives off the greater superficial petrosal nerve and changes direction at the first genu. The facial nerve then courses posteriorly and laterally through the middle ear as the tympanic segment of the facial nerve.[2] After passing superior to the stapes and inferior to the lateral semicircular canal, the facial nerve changes course again at the second genu and courses inferiorly in the vertical segment of the facial nerve. The facial nerve then exits the temporal bone at the stylomastoid foramen.[2] The facial nerve then pierces through the parotid gland as it branches into five branches: temporal (frontal), zygomatic, buccal, marginal mandibular, and cervical.

Diseases affecting cranial nerve VIII (cochleovestibular nerve) will cause hearing loss, tinnitus, vertigo, and imbalance. Cranial nerve VIII exits the brainstem at the junction of the pons and medulla near cranial nerve VII. It then travels with cranial nerve VII in the internal auditory canal and divides into the cochlear nerve, innervating the cochlea, the superior vestibular nerve, innervating the utricle, lateral semicircular canal, and superior semicircular canal, the inferior vestibular nerve, innervating the saccule, and the singular nerve, which only receives input from the posterior semicircular canal.[2]

Deficits of cranial nerves IX and X (glossopharyngeal and vagus nerves) will cause hoarseness and dysphagia, with the disease being most likely in the jugular foramen. Deficits involving cranial nerve XI (the spinal accessory nerve) lead to ipsilateral shoulder weakness and difficulty turning the head to the contralateral side. Cranial nerves IX–XI exit the skull base through the jugular foramen.[2] Cranial nerve XII (hypoglossal nerve) dysfunction causes deviation of the tongue towards the side of the lesion with possible atrophy of the tongue and occurs with diseases affecting the hypoglossal canal.[2]

Diagnostic evaluation of skull base disease

Computed tomography (CT) and magnetic resonance imaging (MRI) are the most common imaging modalities for skull base disease and are often complementary. CT is excellent at detailing the bony anatomy of the temporal bone and petrous apex. CT is superior to MRI to evaluate lesions such as glomus tumours, encephaloceles, intertemporal facial nerve tumours, cholesterol granuloma, and cholesteatoma. MRI is better at evaluating lesions of the cerebellopontine angle, petrous apex, and brainstem and has the advantage of using different imaging types to differentiate the lesion.

Vestibular schwannoma

Vestibular schwannomas are benign neoplasms that originate from the vestibulocochlear nerve with an incidence in the US of 1.09 per 100,000 people[3] and 19 per 1 million people per year based on an extensive study from Denmark.[4] Patients with vestibular schwannomas present with hearing loss (95%), dizziness (65%), and tinnitus (63%),[5]

although, with the more frequent use of MRI, incidental diagnosis is common. Large tumours may present with trigeminal symptoms of facial numbness (7–9%), hydrocephalus, or ataxia (14%).[5] Facial paresis is rare (5.2%), even with the largest tumors.[5] MRI is the diagnostic modality of choice when evaluating a vestibular schwannoma. MRI characteristics of a vestibular schwannoma include the tumour centred around and often involve the internal auditory canal, hypointense on pre-contrast T1-weighted imaging, hyperintense on T2-weighted imaging, and brightly enhanced with gadolinium administration.[6] Treatment options include observation with serial MRIs, radiosurgery, or microsurgery.

Because many vestibular schwannomas are slow-growing or appear not to progress, an initial period of observation is becoming more common. Stangerup et al. have shown in a series of 552 patients with vestibular schwannoma managed with observation and 4-year follow-up that 17% of intracanalicular tumours and 29% of extrameatal tumours will progress.[7] Even if tumours enlarge, the typical growth rate is 1–2 mm per year, so a period of watchful waiting does not affect future treatment. One exception to increased morbidity with observation is hearing, since patients may lose hearing during the waiting period. Because of this, some patients will choose hearing preservation surgery as the initial treatment. In a cohort of 466 patients observing a vestibular schwannoma, Hunter et al. found that 66% and 44% of patients maintained serviceable hearing over 5 years and 10 years, respectively.[8] When observing a tumour, the first MRI is scheduled 6 months after the initial MRI to ensure a rapidly growing tumour or different diagnosis is not present. If the tumour is stable, further tumour growth is monitored with annual MRIs.

Radiosurgery has become more commonly used throughout the years. Most centers use the one-time treatment of radiosurgery compared to multiple treatment sessions with radiotherapy. The International Stereotactic Radiosurgery Society guidelines recommend a radiosurgery dose of 11–14 Gy.[9] The overall control rate is around 90–99% over 5 years[9]; however, a 2017 study from the Mayo Clinic showed tumours that enlarged greater than 2.5 mm in greatest diameter per year before radiation had a control rate of 69%.[10] Side effects and complications are rare and include facial paresis (1%),[11] facial pain or numbness (3.1%),[11] hydrocephalus (2–3%),[12] and malignant transformation (0.3%).[13] In tumours that require microsurgical resection after failed radiosurgery, facial nerve outcomes may be worse than unirradiated tumors.[14]

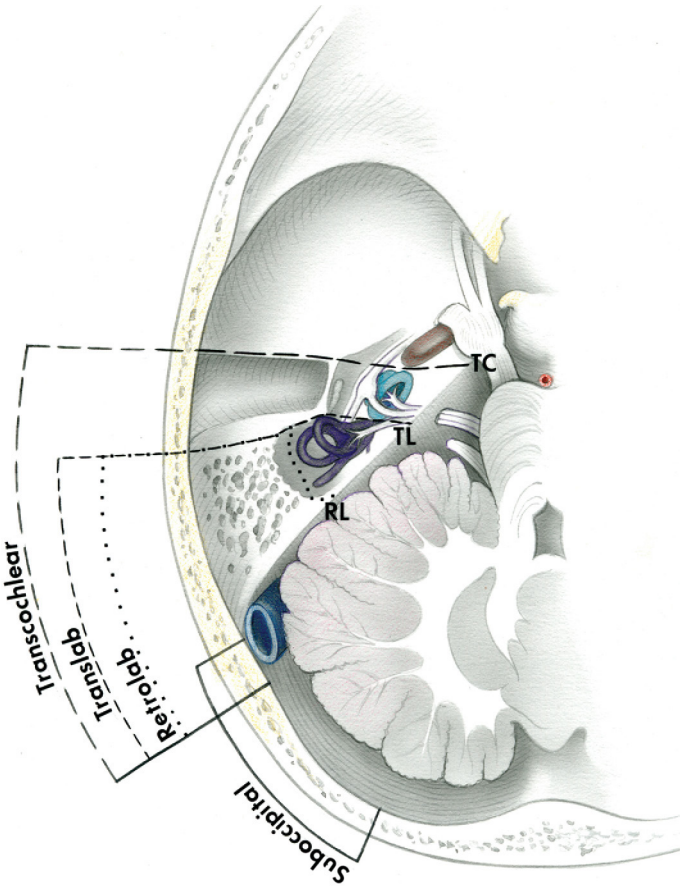

Figure 18.2 The translabyrinthine and suboccipital (retrosigmoid) approaches are the most common approaches to remove neoplasms from the lateral skull base. The translabyrinthine approach sacrifices residual hearing and vestibular function since the labyrinth is entered. The retrosigmoid approach has the advantage of potential hearing preservation, but the lateral internal auditory canal and remaining temporal bone is less accessible. The retrolabyrinthine approach is similar to the translabyrinthine approach, but the labyrinth is spared, allowing for hearing preservation at the cost of less exposure. The transcochlear approach allows the widest access to the lateral skull base but may require transposing the facial nerve, which will likely result in chronic facial paresis.

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Microsurgery

There are three common approaches to the surgical management of vestibular schwannomas. The retrosigmoid and middle fossa approaches have the advantage of potential hearing preservation since the labyrinth is not entered during the procedure. The translabyrinthine approach removes the labyrinth, which results in a profound sensorineural hearing loss (**Figure 18.2**).

Translabyrinthine approach

The translabyrinthine approach has the advantage of a direct view of the cerebellopontine angle without retraction of the temporal lobe or cerebellum. Also, the facial nerve is identified in the fundus of the internal auditory canal before tumour dissection begins. At many skull base centres, the translabyrinthine approach is the most common approach for the removal of a vestibular schwannoma, and even the largest vestibular schwannoma can be removed with a translabyrinthine approach. The translabyrinthine is used to remove other tumours in the cerebellopontine angle or temporal bone in patients with non-serviceable hearing. Concerning facial nerve outcomes, 84% of patients with tumours less than 3.5 cm in diameter had facial function of (House–Brackmann) HB 1–2 function after 1 year, compared to 53% of patients with tumours greater than 3.5 cm in a retrospective study of 392 patients who underwent translabyrinthine resection of a vestibular schwannoma.[15] In another retrospective review of 1244 patients who underwent translabyrinthine resection of a vestibular schwannoma, 92.3% of patients with intracanalicular tumours had facial function of HB 1–2, 82% of patients with tumours less than 25 mm in diameter had HB 1–2 facial function, and 55.6% of patients with tumours greater than 25 mm had HB 1–2 facial function.[16]

Variations of the translabyrinthine approach

The transotic and transcochlear approaches allow improved access when there is an anterior extension of the tumour with the cochlea blocking access. Both procedures involve over closing the EAC, removing the posterior bony EAC, and opening the promontory to allow for more anterior exposure. The difference between these two approaches is the need to transpose the facial nerve, with the transcochlear approach requiring transposition to create an unobstructed view of the brainstem.[17] Transposing the nerve poses a significant risk for permanent facial nerve dysfunction. In a retrospective series of 20 patients who underwent the transcochlear approach, only 30% of patients had HB 1–2 facial function at last follow-up.[17] The transotic approach involves working both anterior and posterior to the facial nerve with the facial nerve remaining in the fallopian canal.[18] The facial nerve outcomes with the transotic approach in a retrospective study of 36 patients with vestibular schwannoma are 94% HB 1–2 (19% HB 1, 75% HB 2).[18]

Middle fossa approach

The middle fossa approach is used to remove vestibular schwannomas in cases of serviceable hearing. A temporal craniotomy is performed, the dura is lifted off the middle fossa floor in a posterior to anterior direction to prevent injury to the facial nerve, and a retractor is placed under the petrous ridge. The arcuate eminence and greater superficial petrosal nerve identify the location of the internal auditory canal, with the position of the internal auditory canal bisecting this angle. The internal auditory canal is skeletonized with the superior semicircular canal being the posterior limit of dissection and the cochlea and trigeminal nerve being the anterior limit of bone removal. The fundus and labyrinthine segment of the facial nerve are identified. Intraoperative auditory brainstem response or near-field compound action potential is used to monitor the status of the cochlear nerve during tumour dissection. The maximum size of a vestibular schwannoma to consider a middle fossa approach is 1.5 cm with more cerebellopontine angle involvement making tumour dissection and hearing preservation more difficult. In a retrospective study of 38 patients with vestibular schwannoma and serviceable hearing who underwent middle fossa craniotomy, 63.2% of all patients, and 73.3% of patients with tumours smaller than 10 mm, maintained class A or B hearing, respectively.[19] With regard to facial nerve outcomes, 76.1% of patients had HB 1 facial function, 13% of patients had HB 2 facial function, and 10.9% of patients had HB 3 facial function.[19] Disadvantages include temporal lobe retraction that could result in temporal lobe injury or cognitive issues.

Retrosigmoid approach

The retrosigmoid approach has the advantage of potential hearing preservation and provides the surgeon with a panoramic view of the cerebellopontine angle. To facilitate tumour removal in the internal auditory canal, the bone around the internal auditory canal usually needs to be removed. The labyrinth prevents full exposure of the internal auditory canal in most cases, which makes tumour removal in the lateral internal auditory canal (i.e. fundus) difficult. The retrosigmoid craniotomy is used in patients with serviceable hearing, tumours less than 20 mm, and tumours not extending greater than two-thirds into the internal auditory canal. In a recent systematic review of 2034 vestibular schwannomas removed with the retrosigmoid craniotomy, 57%, 37%, and 12% of patients maintained class A or B hearing for intracanalicular, tumours less than 20 mm, and tumours greater than 20 mm, respectively.[20] When comparing the retrosigmoid craniotomy and middle fossa craniotomy in a retrospective study of 138 vestibular schwannomas, both retrosigmoid craniotomy and middle cranial fossa craniotomy had similar rates of patients with HB 1 or 2 facial function—88% for the middle fossa group and 90% for the retrosigmoid group.[21] However, the middle cranial fossa group had significantly lower rates of long-term HB 1, 72%, compared to the retrosigmoid group, 88%.[21] Despite these findings, guidelines by the Congress of Neurological Surgeons found insufficient evidence to determine the superiority of any of the three most common approaches —middle fossa craniotomy, retrosigmoid craniotomy, and translabyrinthine—in terms of facial nerve outcomes.[22]

Less common tumours of the cerebellopontine angle and internal auditory canal

Meningioma

Meningiomas are benign tumours that arise from arachnoid cap cells in the dura and comprise 10% of tumours involving the cerebellopontine angle.[23] Although meningiomas present less frequently with audiovestibular symptoms compared to vestibular schwannomas, hearing loss is still the most common presenting symptom, affecting 75% of patients, followed by vestibular symptoms in 59% of patients, and trigeminal symptoms with facial numbness or facial pain in 28% of patients with meningioma.[24]

Imaging characteristics can often differentiate these neoplasms (**Figure 18.3**). Meningiomas tend to be eccentric to the internal auditory canal, have dural tails, more homogeneous enhancement, and appear sessile.[23] As with vestibular schwannomas, meningiomas are managed with observation, radiosurgery, or microsurgical resection.[23] Radiosurgery controls 93% and 77% of meningiomas over 5 and 10 years, respectively.[25] Meningiomas may be removed surgically through many of the same approaches used for vestibular schwannoma, namely translabyrinthine craniotomy, retrosigmoid craniotomy, or middle fossa craniotomy approaches. However, surgical resection of meningiomas has better hearing preservation outcomes compared to vestibular schwannoma, and for this reason, hearing preservation approaches may be favoured.

One additional approach that is more applicable to meningioma is a transpetrosal approach, which is a combination of the retrolabyrinthine and the middle fossa craniotomy approaches. In this approach, a standard mastoidectomy is performed, followed by skeletonizing the bony labyrinth.[26] The posterior cranial fossa dura, middle cranial fossa dura, and sigmoid sinus are decompressed.[26] A middle fossa craniotomy approach is then performed with skeletonization of the internal auditory canal and opening of the petrous apex.[26] The superior petrosal sinus is then ligated and the tentorium is divided, which allows for a combined cavity and improved visualization of the tumor.[26] This approach allows for possible hearing preservation. This procedure has the additional risk of cranial nerve IV injury when dividing the tentorium.[26] The transpetrosal approach is most useful for meningiomas that have a

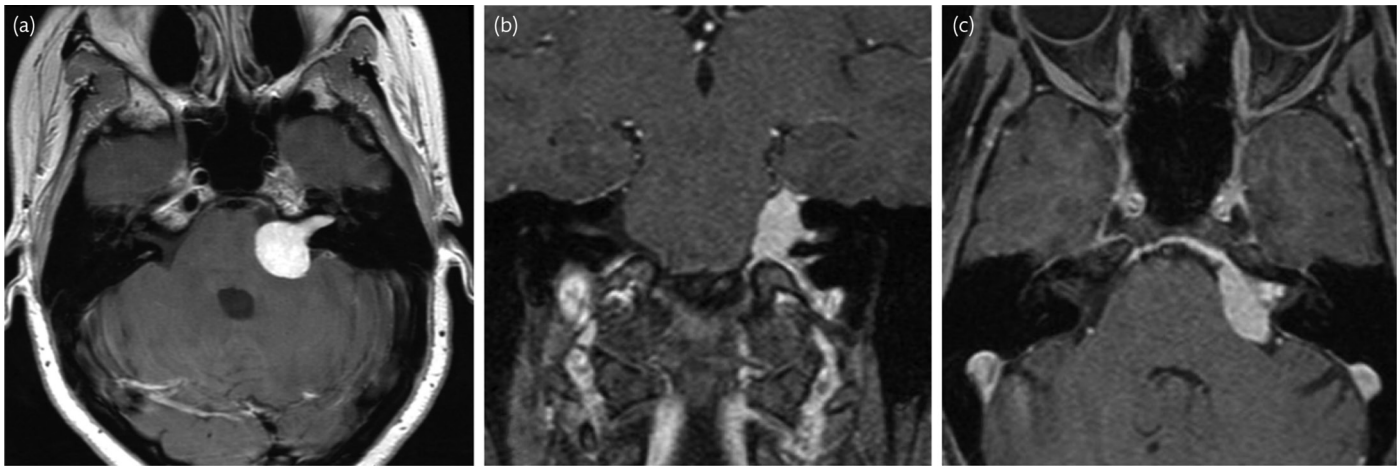

Figure 18.3 MRI comparison of vestibular schwannoma and meningioma. (a) A left vestibular schwannoma characterized by being centred on the internal auditory canal, homogeneous and bright enhancement, and a rounded cerebellopontine angle component. (b) and (c) A meningioma characterized by less enhancement, eccentric to the internal auditory canal, presence of dural tails, and more sessile appearance in the cerebellopontine angle.

significant anterior and superior extension. In a series of 34 meningiomas treated surgically, the majority of which underwent retrosigmoid craniotomy, 91.2% of tumours were controlled with surgery over a mean follow-up of 16.6 months.[27] None of these postsurgical patients had new hearing loss, and 5.9% had a new facial weakness.[27] The most common new neurological deficit was cranial nerve X (11.8%), which occurred when the tumour involved the jugular foramen.[27]

Lipoma

Lipomas of the internal auditory canal and cerebellopontine angle comprise 0.1% of tumours in the location.[28,29] In a study of 15 patients with a lipoma, 40% of the sample presented with hearing loss, 33% presented with tinnitus, 13% presented with dizziness, and 6.7% presented with facial weakness.[29] Of note, only one patient (6.7%) showed tumour progression.[29] Because these tumours rarely progress, observation is recommended as the management strategy of choice. Distinguishing between lipoma and vestibular schwannoma is best accomplished with a T1-weighted MRI without contrast (Figure 18.4). A lipoma will be hyperintense, but a vestibular schwannoma will be isointense and enhance only with the addition of gadolinium.[29] A T1-weighted MRI with fat suppression is also helpful to differentiate a lipoma from a vestibular schwannoma. Therefore, it is imperative to always look at the T1-weighted MRI without contrast to rule out a lipoma.

Epidermoid

Similar to a cholesteatoma originating in the middle ear, epidermoids are benign tumours comprised of squamous epithelium found in the cerebellopontine angle, petrous apex, and posterior fossa. Epidermoids comprise 1% of cerebellopontine angle tumors.[23] Symptoms of epidermoids include hearing loss (66%),

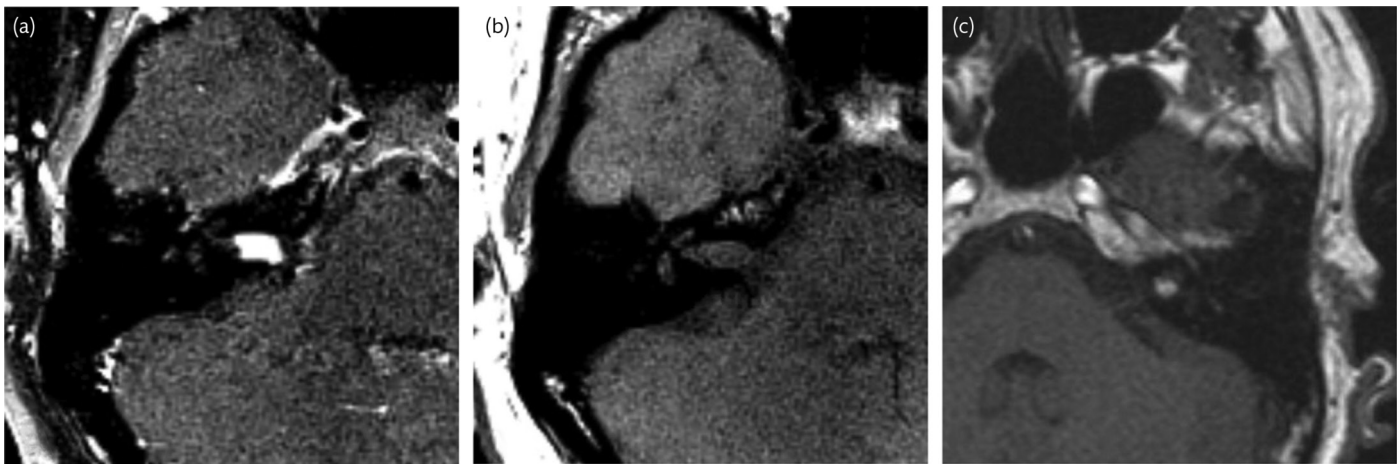

Figure 18.4 MRI comparison of an internal auditory canal lipoma and vestibular schwannoma. (a) T1-weighted MRI with gadolinium contrast consistent with a right vestibular schwannoma involving the internal auditory canal. (b) The same tumour without contrast. (c) A hyperintense lesion in the left internal auditory canal in a T1-weighted MRI without gadolinium contrast. Notice the lack of contrast in the nasal and sinus mucosa. This is consistent with a lipoma of the internal auditory canal.

facial weakness (32%), and trigeminal symptoms (42%).[30] Unlike other cerebellopontine angle neoplasms, the keratin debris from epidermoids may cause recurrent episodes of aseptic meningitis.[23] MRI demonstrates a lesion that is isointense on T1-weighted MRI that does not enhance with contrast. Epidermoids are hyperintense on T2-weighted MRI. Since arachnoid cysts also have similar MRI characteristics, a diffusion-weighted MRI differentiates these lesions.[23] An epidermoid will show restricted diffusion (appears bright on diffusion-weighted MRI), whereas an arachnoid cyst will not show restricted diffusion.[23] In patients with cranial neuropathies, hydrocephalus, or proven growth on MRI, surgical resection should be considered.[31] In 47 patients with cerebellopontine angle epidermoid managed with surgery (the majority with the retrosigmoid approach), 8% experienced recurrence of the tumour, 93% of patients with preoperative hearing maintained serviceable hearing, and 3% had mild (HB 2) facial weakness.[31]

Arachnoid cysts

Arachnoid cysts are malformations of the arachnoid, leading to cystic spaces filled with cerebrospinal fluid.[23] Arachnoid cysts are only symptomatic in 15% of cases, which can present with cerebellar compression, hearing loss, balance disturbance, or suboccipital headache.[23] On MRI, these lesions appear identical in signal to cerebrospinal fluid and have rounded and smooth edges.[23] In a study of 213 arachnoid cysts, only 2.3% of these cysts enlarged in size over a median follow-up period of 3.3 years.[32]

Tumours of the jugular foramen

Tumours of the jugular foramen affect the lower cranial nerves and present with dysphagia, hoarseness, and tongue weakness as opposed to tumours involving the cerebellopontine angle, which may present with hearing loss, dizziness, or pulsation tinnitus. MRI and CT are complementary in the evaluation of lesions in the jugular foramen.

Jugular foramen paraganglioma

Paragangliomas, previously described as glomus tumours, are the most common tumours of the jugular foramen.[33] Paragangliomas are slow-growing, highly vascular tumours that arise from the paraganglion cells of the adventitia of the jugular bulb.[33] These tumours are most often benign, though they can rarely be considered malignant and spread to other organ systems.[33] A paraganglioma is deemed to be malignant only if metastasis is present since benign and malignant paragangliomas are identical on histopathological examination. The most common presenting symptom is pulsatile tinnitus, but hearing loss, tinnitus, dizziness, and lower cranial nerve dysfunction can also be present.[33] Additionally, patients may present with sweating, palpitations, flushing, hypertension, syncope, and headaches due to catecholamine secretion from the tumor.[33] However, less than 4% of paragangliomas of the head and neck secrete catecholamines.[33] Blood and urine catecholamines are sent as part of the initial workup to rule out a secreting paraganglioma. If a secreting tumour is identified, surgical removal is often preferred. On physical examination, these tumours can extend into the middle ear and appear as a violaceous retrotympanic mass on otoscopy.[33] Because a glomus tumour is vascular, the tumour has a characteristic 'salt-and-pepper' appearance on T1-weighted MRI due to flow voids and enhances avidly with gadolinium contrast on MRI.[33] CT shows a lesion with bone destruction and erosion of the jugulocarotid spine.[33] Thirty per cent of head and neck paragangliomas may be attributable to germline mutations in the succinate dehydrogenase (*SDH*) genes.[33-34] Patients with *SDH* mutations, specifically *SDHB*, *SDHC*, and *SDHD* mutations, are at risk for multiple paragangliomas, and particularly with *SDHB* mutations, have a higher risk of malignancy, defined by the presence of metastasis.[34] For this reason, genetic screening should be considered in cases of jugular foramen paraganglioma. Because paragangliomas are slow-growing, even large tumours may not cause apparent lower cranial nerve deficits. The lack of symptoms, even in large tumours, has resulted in the more common practice of observation or radiation therapy. In a multicentre study of 132 patients with jugular foramen paraganglioma treated with Gamma Knife radiosurgery, 93% of tumours were controlled, with an estimated 5-year control rate of 88%.[35] Pulsatile tinnitus improved in 49% of patients.[35] Fifteen per cent of patients developed a new cranial nerve deficit after radiation, and 11% of patients with pre-radiation cranial neuropathies experienced an improvement in their cranial nerve deficit.[15] Surgery is generally reserved for growing tumours that are causing lower cranial neuropathies. It is essential to know that even in patients with apparent lower cranial nerve involvement (vocal cord paralysis, dysphagia, etc.), removing the tumour will usually exacerbate these symptoms because residual preoperative function is almost always present.

Traditional microsurgical resection with the infratemporal fossa approach involves a mastoidectomy, over closing the ear canal, removing the posterior ear canal wall to access the middle ear, removing the mastoid tip, and exposing the cranial nerves, jugular vein, and carotid artery in the neck.[36] The facial nerve is skeletonized and transposed to provide better access to the infratemporal fossa approach.[36] The sigmoid sinus is packed, and the jugular bulb is resected along with the tumor.[36] As with the transcochlear approach, transposition of the facial nerve results in a significant risk of permanent facial weakness. For this reason, our practice is to typically skeletonize the facial nerve, but preserve the normal course of the facial nerve, and resect the tumour working around the facial nerve, as in the transotic approach. Resecting the jugular bulb places lower cranial nerves at significant risk of injury. In a retrospective series of 202 patients with jugular foramen paraganglioma treated with microsurgery, 90% had gross total resection with tumour recurrence in 6% of patients.[36] New cranial neuropathy occurred in 60% of patients, with 40%, 24%, 26%, and 21% developing dysfunction in cranial nerves IX, X, XI, and XII, respectively.[36] Given the high rate of cranial neuropathy and morbidity secondary to gross tumour resection, in recent years, subtotal resection with subsequent radiation for tumour growth has become a more favoured approach. In a study of 12 patients who underwent planned subtotal resection, none developed new permanent postoperative lower cranial nerve deficits.[37] Four of these patients (33.3%) had postoperative tumour growth, though in patients with residual tumours less than 20% of the original tumour volume, no tumour progression occurred.[37]

Schwannoma

Schwannomas of the jugular foramen comprise less than 1% of all temporal bone lesions.[38] These neoplasms can present with similar

symptoms to jugular foramen paragangliomas, which include lower cranial nerve dysfunction or audiovestibular symptoms.[38] Lower cranial nerves schwannomas of the jugular foramen are isointense on T1, hyperintense on T2 with a homogeneous enhancement pattern with gadolinium on MRI, and a smooth enlargement of the jugular foramen without dural tails or flow voids.[38] Because schwannomas arise from the lower cranial nerves, removal will often result in lower cranial nerves deficits. Because of this, schwannomas of the jugular foramen are treated conservatively with serial imaging. If growth is demonstrated, stereotactic radiation therapy, radiosurgery, or surgical resection is advised.

Facial nerve tumours

The most common cause of facial paralysis is idiopathic facial paralysis (Bell's plays), which is characterized by acute onset of facial paralysis and recovery in most cases. Insidious facial weakness or incomplete recovery should raise the suspicion of a more serious condition. The most common tumours of the facial nerve are facial nerve schwannoma and facial nerve haemangioma. Malignancies of the parotid or skin can affect the facial nerve. Progressive facial paresis, pain, and history of malignancy are hallmarks of malignancy. As a rule, facial paralysis or paresis that is progressive or does not recover should raise suspicion of a neoplasm and not idiopathic facial paralysis.

Facial nerve schwannoma

Facial nerve schwannomas most commonly occur near the geniculate ganglion, but may occur anywhere along the course of the facial nerve.[39] Progressive facial paralysis is common: 41–82% of patients present with facial weakness,[39] although many patients never develop significant paralysis. In a study of 79 facial schwannoma patients, 40.5%, 22.8%, 16.5%, 6.3%, 10.1%, and 3.8% of patients had HB 1, HB 2, HB 3, HB 4, HB 5, and HB 6 at presentation, respectively.[40] Removing a facial nerve schwannoma requires sacrificing and grafting the facial nerve, and the best outcome after a facial nerve graft is HB grade 3 function. Therefore, surgical removal of a facial nerve schwannoma should be reserved for tumours that cause facial weakness worse than HB grade 3.[40] Choice of surgical approach will depend on the location of the tumour and hearing status. In patients with serviceable hearing, a transmastoid approach can be used for tumour distal to the geniculate ganglion and a middle fossa craniotomy approach for tumours that are proximal or involve the geniculate ganglion. In cases where the hearing is no longer serviceable, a translabyrinthine approach is an option. Radiosurgery may be used to treat enlarging facial nerve schwannomas that are not causing significant facial paresis. In a study of facial schwannomas treated with radiosurgery, 90% of tumours were controlled over 5 years, with 90% preserving their pre-radiation facial nerve function.[41] If an enlarging tumour is in the fallopian canal, decompression of the tumour followed by radiosurgery is advisable to prevent the facial nerve and tumour from swelling in the tight space of the fallopian canal.

Facial nerve haemangioma

Facial nerve haemangiomas, which are less consistent with haemangiomas on histopathology and should be termed venous malformations,[42] comprise 0.7% of temporal bone lesions.[43] These lesions are slow-growing, most commonly affect the geniculate region of the facial nerve, and cause more facial weakness than expected by their size.[44] In a study of 18 patients with facial nerve haemangiomas, 94% presented with facial weakness.[45] Imaging characteristics on MRI of a haemangioma show hyperintensity on T2 with areas of hypointensity and heterogeneous enhancement with areas of less enhancement on T1 with gadolinium.[44] On CT scanning, haemangiomas have irregular borders and intratumoral bony spicules.[44] Unlike with resection of schwannomas, removal of the haemangioma while preserving the facial nerve is possible and preferable, therefore earlier surgical resection may be considered. The primary surgical approach is middle fossa craniotomy, though the translabyrinthine approach is another option if hearing is poor. In a series of 15 patients with facial nerve haemangioma removed surgically, 11 (73%) tumours were removed with preservation of the underlying nerve.[45] All 11 patients in which the facial nerve was preserved had at least HB 3 facial function, and 72% recovered to HB 1 or HB 2 facial function.[45]

Petrous apex lesions

Petrous apex lesions are often discovered incidentally.[46] Retroorbital pain and generalized temporoparietal headaches may be symptoms caused by petrous apex lesions secondary to irritation of the trigeminal nerve.[46] Diplopia may also occur secondary to involvement of cranial nerve VI as it passes through Dorello's canal.[46] Facial weakness may occur secondary to compression of the facial nerve in the cerebellopontine angle or internal auditory canal.[46] Hearing loss, tinnitus, and imbalance may similarly occur if lesions from the petrous apex compress the VIIIth nerve in the cerebellopontine angle or internal auditory canal.[46] Rarely, malignant processes may cause dysphagia, hoarseness, or shoulder weakness.[46] MRI is excellent at differentiating lesions of the petrous apex (Table 18.2).

Cholesterol granuloma

Cholesterol granulomas have an incidence of 0.6 cases per 1 million population,[46] and cholesterol granulomas of the petrous apex are ten times more common than petrous apex cholesteatomas.[47] Cholesterol granulomas form secondary to blood entry into a mucosa-lined space.[46] Over time, anaerobic breakdown of blood leads to the formation of cholesterol crystals, which in turn incite a foreign body giant cell reaction, leading to cyst formation.[46] On MRI, cholesterol granulomas are hyperintense on both T1 pre-contrast and T2-weighted images.[46] On CT scanning, cholesterol granulomas demonstrate smooth erosion of the petrous apex.[46] Cholesterol granulomas are most commonly managed with observation and serial imaging examination.[46] Rapid growth with the onset of new symptoms may prompt surgical management, either through excision with a middle fossa craniotomy or marsupialization.[46] Marsupialization may be performed through a transcanal infracochlear approach or transmastoid infralabyrinthine approach.[46] The benefit of marsupialization is avoiding the need for craniotomy, though there is increased risk of recurrence if scarring blocks the newly created ventilation tracts.[46]

Table 18.2 Magnetic resonance imaging characteristics

Tumour	T1-weighted	T2-weighted	Enhances with contrast	Location	Description
Vestibular schwannoma	Hypointense	Iso- to hyperintense	Yes	IAC, CPA	Centred over IAC in CPA with IAC extension, may be cystic or solid
Meningioma	Hypointense	Hyperintense	Yes	IAC, CPA, petrous apex, jugular foramen	Eccentrically positioned relative to IAC, dural tails, sessile appearance
Lipoma	Hyperintense	Hyperintense	No	IAC, CPA	Use pre-contrast T1 to differentiate from vestibular schwannoma, disappears with fat saturation
Epidermoid	Isointense	Hyperintense	No	IAC, CPA, petrous apex	Significant restricted diffusion, use diffusion-weighted MRI to differentiate from arachnoid cyst
Arachnoid cyst	Isointense	Hyperintense	No	IAC, CPA	No restricted diffusion, appears like cerebrospinal fluid
Paraganglioma	Isointense with flow voids	Hyperintense with flow voids	Yes	Middle ear, jugular foramen	Salt-and-pepper appearance due to flow-voids
Lower cranial nerve schwannoma	Hypointense	Hyperintense	Yes	Jugular foramen	Smooth tumours that enhance without flow-voids
Facial nerve schwannoma	Hypointense	Hyperintense	Yes	Facial nerve	Geniculate ganglion—most common site, smooth appearance on CT
Facial nerve haemangioma	Isointense	Hyperintense	Yes	Facial nerve	Heterogeneous enhancement, CT shows bony spicules and irregular borders
Cholesterol granuloma	Hyperintense	Hyperintense	No	Petrous apex	Smooth erosion of petrous apex
Chordoma and chondrosarcoma	Isointense	Hyperintense	Yes	Clivus, petrous apex	Erosion seen on CT

CPA, cerebellopontine angle; IAC, internal auditory canal.

Other lesions of the petrous apex

Cholesteatomas or epidermoids may involve the petrous apex.[46] The origin of the epithelium distinguishes an epidermoid from a cholesteatoma. Cholesteatomas originate from the middle ear, and epidermoids occur from residual epithelial rests from embryogenesis. Otherwise, these lesions are identical on both histopathology and imaging. On MRI, these lesions are hypointense on T1 imaging, hyperintense on T2, and restricted diffusion on diffusion-weighted MRI.[46] When lower cranial nerve deficits are present, there should be an elevated concern for malignancy.[46] Chordomas are lobulated masses that destroy bone and typically involve the clivus and petrous apex. Chordomas enhance with gadolinium, though less intense enhancement when compared to chondrosarcoma.[46] Chondrosarcoma may originate from the petrous apex.[46] On MRI, these lesions are hypointense on T1, hyperintense on T2, avidly enhance with contrast, and are centred on the petrous apex near the foramen lacerum.[46]

Infectious skull base disorders

Skull base osteomyelitis

Skull base osteomyelitis can cause a myriad of symptoms, which include pain, otorrhea, hearing loss, and cranial neuropathies.[48] The condition occurs secondary to an infection, which occurs most commonly due to *Pseudomonas aeruginosa*, but may also occur secondary to non-pseudomonal bacteria, such as methicillin-resistant *Staphylococcus aureus* or secondary to fungal infection.[49] The examination classically shows granulation tissue in the EAC at the bony cartilaginous junction.[48] Skull base osteomyelitis is more common in patients with diabetes mellitus or who are immunocompromised.[48] Diagnosis has traditionally used technetium-99 and gadolinium bone scans. Follow-up gallium scans are used to measure a response to treatment since technetium-99 scans will stay positive indefinitely.[48] On a recent meta-analysis, technetium-99 scans were found to have a sensitivity of 85.1%, and gallium scans had a sensitivity of 71.2%, both with reduced specificity.[50] A recent best practices paper, therefore, advocates against nuclear studies, preferring CT, which demonstrates bony destruction, or MRI with contrast, with potential for fluorodeoxyglucose-positron emission tomography/CT as an additional diagnostic imaging.[51] Erythrocyte segmentation rate and C-reactive protein are serum inflammatory markers that are markedly elevated in this condition and can be used to follow response to therapy.[52] The mainstay of treatment for skull base osteomyelitis is long-term intravenous antibiotics and reversal of immunocompromised state if possible.[48] Surgery is indicated in the presence of abscess formation or disease not responding to medical therapy; however, surgery may also be needed for biopsy to rule out a malignant process and obtain cultures to direct long-term antibiotic therapy.[48] See also Chapter 24 on necrotizing otitis externa.

Petrous apicitis

Petrous apicitis classically manifests as a triad of otorrhoea secondary to suppurative otitis media (61.4%), deep retro-orbital pain secondary to trigeminal nerve inflammation (54.5%), and abducens cranial nerve palsy, secondary to inflammation in Dorello's canal (15.9%).[53] CT may demonstrate an effusion and erosive changes in the petrous apex.[53] MRI often shows a rim enhancing lesion in the petrous apex and is important if there is concern for intracranial involvement.[53] *Pseudomonas aeruginosa*, followed by *Staphylococcus aureus*, are the most common organisms.[53] Treatment is usually with intravenous antibiotics; 77.3% of patients respond well to medical therapy alone or with the placement of a tympanostomy tube.[53] Surgical treatment is reserved for patients who are not responding to antibiotic therapy. The transcanal infracochlear or the transmastoid infralabyrinthine approaches are most commonly used to drain abscesses.

Conclusion

Lateral skull base disease presents unique management challenges due to the complex anatomy of the region and vital structures. Thus, morbidity from surgical intervention may exceed the morbidity from the disease process. Because of the complexity of this region, multidisciplinary care with input from neurotologists, neurosurgeons, and radiation oncologists is important for patients to make educated and informed care decisions to ensure the best possible management. If surgical intervention is ultimately selected, optimal operative results may benefit from the close collaboration between neurotological and neurosurgical teams.

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19

Cholesteatoma

Neil Bailie and Colin Leonard

Introduction

A patient with a cholesteatoma (active squamous chronic otitis media) classically presents with recurrent episodes of foul-smelling otorrhoea and hearing loss. Unrecognized and untreated, a cholesteatoma can progressively destroy structures within and around the temporal bone, resulting in lasting morbidity, and may ultimately prove fatal. Indeed, throughout history and until the advent of modern surgical treatment, chronic middle ear suppuration was greatly feared as a frequent cause of lasting misery and early death. In order to appreciate the nature of the challenges posed by this disease, one must first understand what constitutes a cholesteatoma, how, why, and where they arise within the middle ear cleft, and what structures may be put at risk. Delay in diagnosis and treatment is associated with greater morbidity, more extensive surgery, higher rates of recidivism, and greater requirement for revision surgery. Strategies for assessment and treatment aim to ensure timely, safe, and effective surgical care despite the diversity of symptoms, pathophysiology, and anatomy that may present in both elective and emergency settings.

Definition, classification, and pathophysiology of cholesteatoma

A cholesteatoma is an epithelioid (skin) cyst within the middle ear cleft. It consists of a matrix of keratinizing squamous epithelium, a perimatrix of a variable thickness of connective tissue, and contains keratin debris. It may be classified as being congenital or acquired. The pathophysiological mechanisms underpinning both congenital and acquired cholesteatoma are incompletely understood and remain a matter of debate and controversy.

A congenital cholesteatoma is believed to arise due to failure of normal involution of an embryonic rest of epithelial tissue medial to the tympanic membrane. It is assumed to have been present since birth but is most commonly diagnosed in infancy or early childhood as a white mass medial to the anterosuperior quadrant of an intact tympanic membrane in a patient with no history of perforation or previous ear surgery, and often little or no history of otitis media (Figure 19.1).

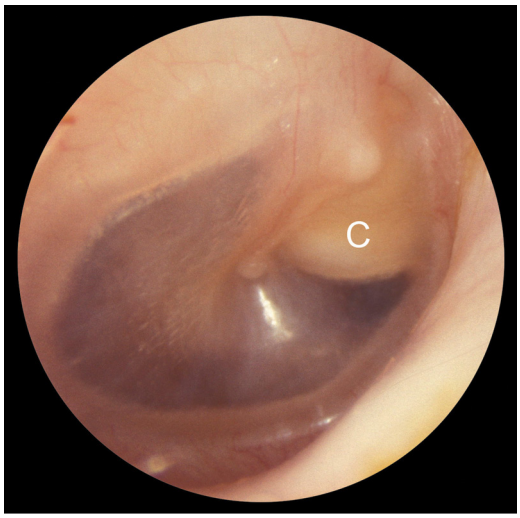

Figure 19.1 Congenital cholesteatoma with intact tympanic membrane, seen as a small pearl of cholesteatoma (C) in the anterosuperior quadrant.

Acquired cholesteatoma is much more common than congenital cholesteatoma and arises as a result of ingress of squamous epithelium into the middle ear cleft, with subsequent retention of keratin debris and secondary mucosal inflammation. A number of different mechanisms have been proposed to account for the development of acquired cholesteatoma, with no single mechanism gaining widespread acceptance. Acquired cholesteatoma may be subclassified into retraction pocket cholesteatoma and non-retraction pocket cholesteatoma. Retraction pocket cholesteatoma is thought to arise secondary to chronic Eustachian tube dysfunction: negative middle ear pressure acting on naturally weak or previously weakened areas of tympanic membrane results in a retraction that interferes with normal epithelial migration, with progressive accumulation of keratin debris helping to force the retraction deeper into the middle ear cleft. In non-retraction cholesteatoma, there is migration of squamous epithelium through a perforation of the tympanic membrane that may have arisen secondary to previous acute suppurative otitis media, trauma, or iatrogenic intervention.

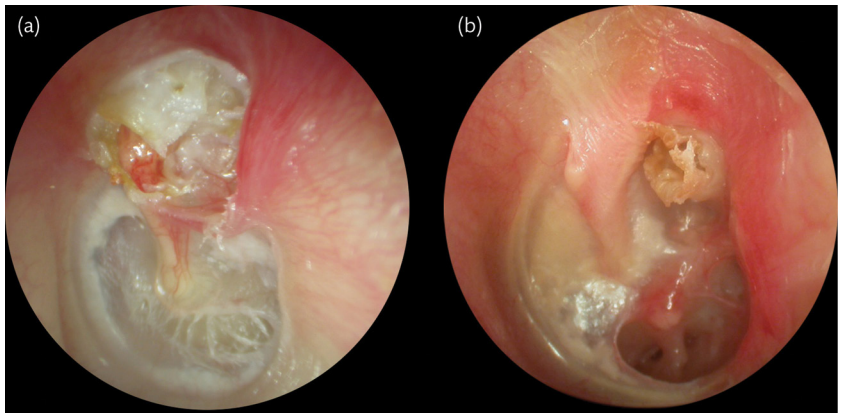

Figure 19.2 Acquired cholesteatoma. (a) Attic cholesteatoma. (b) Pars tensa retraction with cholesteatoma visible in the posterosuperior quadrant.

Other proposed theories include basal cell hyperplasia and squamous metaplasia. The basal cell hyperplasia theory proposes that proliferation of the basal layer of the pars flaccida in response to inflammation results in invagination through of squamous epithelium through the basement membrane with subsequent production of a keratin cyst. The metaplasia theory proposes that chronic middle ear inflammation gives rise to squamous epithelial metaplasia of the mucosa of the middle ear. It is entirely possible that a number of separate mechanisms can give rise to acquired cholesteatoma and indeed, more than one mechanism can exist in a single patient.

Once established, there is a tendency for gradual enlargement of the cholesteatoma within the middle ear cleft, with destruction of bone and frequently with secondary infection that can be difficult to eradicate. A range of infecting organisms may be involved, including aerobic and anaerobic bacteria and fungal species, with increasing evidence of biofilm formation. The most commonly encountered infections are *Pseudomonas aeruginosa*, *Staphylococcus aureus*, *Proteus mirabilis*, *Klebsiella pneumoniae*, and *Escherichia coli*. The mechanism by which cholesteatoma causes bone resorption is also incompletely understood but includes direct pressure, chemical osteolysis, and activation of osteoclasts in response to inflammatory mediators and biofilms (Figure 19.2).

Middle ear and mastoid anatomy

See Figures 19.3–19.7.

Clinical assessment and investigation

Any patient presenting to the outpatient clinic with recurrent episodes of foul-smelling otorrhoea or hearing impairment should be suspected of having an underlying cholesteatoma, despite what previous diagnoses may have been proffered. As such, it is advisable to treat each patient as if you are the first doctor assessing them and begin each consultation with a detailed history, considering both current otological symptoms (Table 19.1) as well as any issues which arose in childhood that may be suggestive of or carry increased risk of middle ear dysfunction (Table 19.1).

Clear visualization of the entire tympanic membrane is essential and photographic documentation is desirable. Microscopic or endoscopic examination and aural toilet will be required in cases where discharge or debris is present. However, it is advisable in all cases as it provides a clearer view of the tympanic membrane and allows for both photographic documentation and an assessment of the extent of surgery that may be achievable via a microscopic or endoscopic approach. In ears where there is little or no discharge present, cholesteatoma may be visible only as a as a small crust or defect in the attic region that may easily be overlooked by the unwary. In more inflamed ears, granulation tissue or polypoidal middle ear mucosa may obscure the view of the tympanic membrane and limit assessment. In this instance, a microbiology swab and antimicrobial

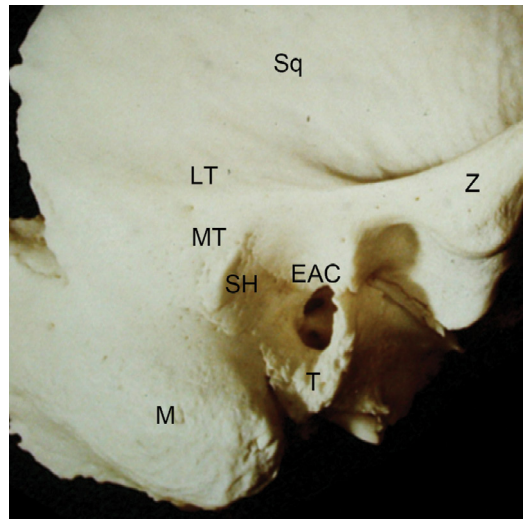

Figure 19.3 Lateral view of the temporal bone showing the squamous bone (Sq), mastoid bone (M), tympanic bone (T), and the zygomatic process (Z). The arrangement of the external auditory canal (EAC), spine of Henle (SH), linea temporalis (LT) and MacEwen's triangle (MT) is clearly seen. The spine of Henle overlies the mastoid antrum, the linea temporalis is a rough estimate for the level of the tegmen tympani. When drilling a cortical mastoidectomy, the triangle formed by the EAC, linea temporalis, and the line that bisects them is known as MacEwen's triangle and represents a safe position with which to enter the mastoid air cells.
Courtesy of Dr John Rutka.

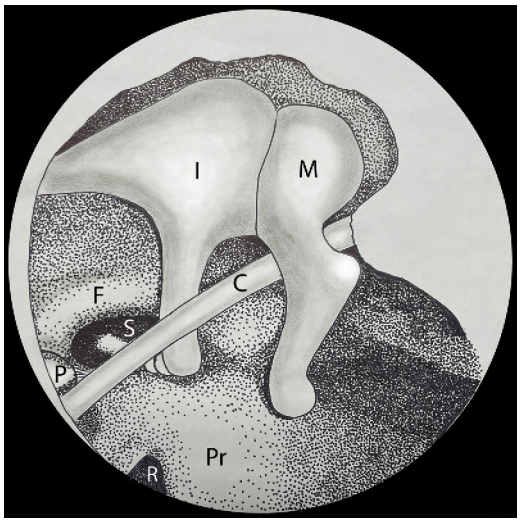

Figure 19.4 The attic and mesotympanum visualized following removal of the tympanic membrane and scutum showing the malleus (M), incus (I), stapes (S), chorda tympani (C), horizontal segment of the facial nerve (F), pyramid (P), round window niche (R), and promontory (Pr).

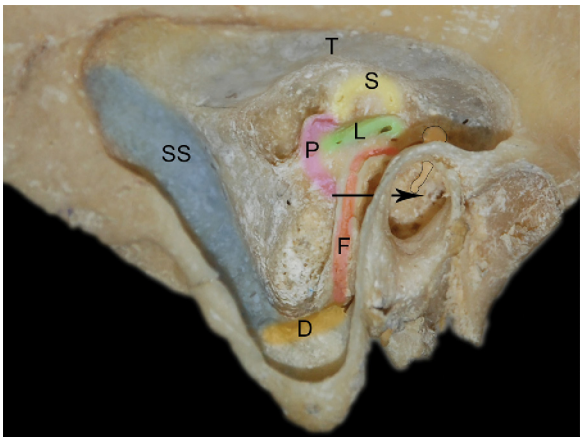

Figure 19.6 Combined approach tympanoplasty or CWU mastoidectomy. The canal wall is intact with the tegmen tympani (T), superior semicircular canal (S), lateral semicircular canal (L), posterior semicircular canal (P), facial nerve (F), sigmoid sinus (SS), and digastric ridge (D) visible. An arrow passes through the posterior tympanotomy from mastoid cavity to middle ear. If proceeding to modified radical mastoidectomy, the superior and posterior parts of the bony ear canal would be removed to allow smooth communication between the mastoid, attic, and ear canal.

therapy in line with local antimicrobial policy should be initiated. However, until assessment has demonstrated otherwise, these patients should be assumed to have a cholesteatoma and kept under close surveillance to facilitate visualization of the tympanic membrane and cholesteatoma. Once free from inflammation it is advisable to complete the clinical examination with a rigid 0° endoscope. The wider field of view may allow visualization of the full extent of a retraction pocket obscured on microscopic view and allows for more satisfactory clinical photography.

All patients should undergo pure tone audiogram, with a minimum of air conduction and bone conduction being recorded at 500 Hz, 1 kHz, 2 kHz, 3 kHz, and 4 kHz in both ears. Where the tympanic membrane is intact, tympanometry may be recorded as an indicator of Eustachian tube function.

Computed tomography (CT) should form an integral part of preoperative planning in the majority of cases. While CT cannot differentiate between cholesteatoma and inflamed mucosa, it provides excellent resolution of bony anatomical landmarks, including extent and aeration of the mastoid. CT will also provide indication of damage caused by cholesteatoma such as erosion of the lateral semicircular canal or deeper entry into the labyrinth, dehiscence of the facial nerve, or erosion of the tegmen tympani, all of which would have significant bearing on the preoperative consent process. While such a finding should occasion extra care when approaching these structures during surgery, it would nonetheless be foolhardy to assume that a complication is not present because it was not found on

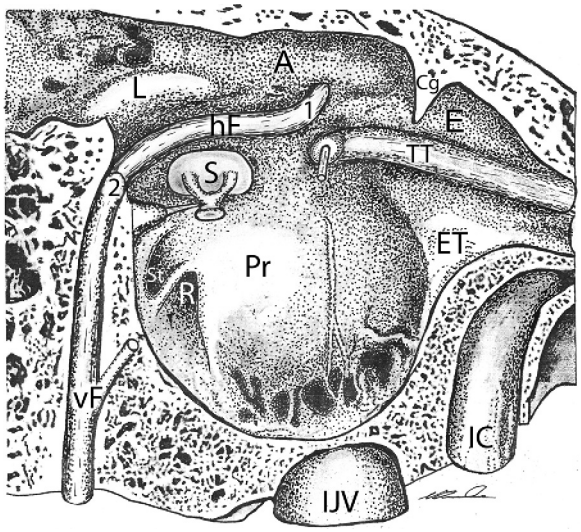

Figure 19.5 View of the medial wall of the middle ear and attic after removal of the malleus and incus showing the stapes (S), first genu of the facial nerve (1), horizontal segment of the facial nerve (hF), second genu of the facial nerve (2), vertical segment of the facial nerve (VF), lateral semicircular canal (L), promontory (Pr), round window niche (R), sinus tympani (ST), attic (A), cog (Cg), anterior epitympanic space (E), tensor tympani (TT), Eustachian tube (ET), internal jugular vein (IJV), and internal carotid artery (IC).

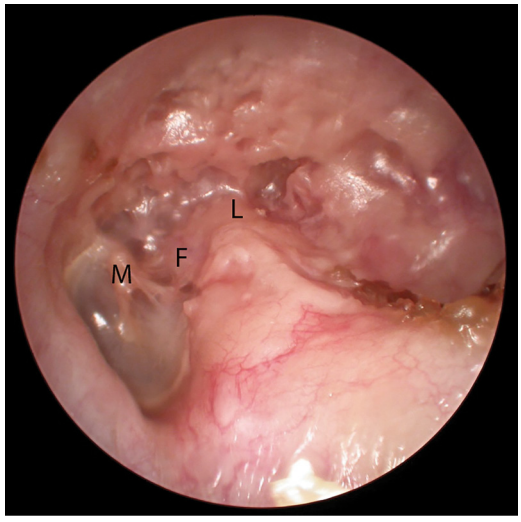

Figure 19.7 Modified radical mastoid cavity, with the malleus handle (M), lateral semicircular canal (L), and facial nerve (F) marked.

Table 19.1 Symptoms associated with cholesteatoma

Symptoms associated with cholesteatoma	Risk factors for developing cholesteatoma
Recurrent otorrhoea	Cleft palate
Recurrent odour	Syndromes (such as trisomy 21) that impact upon mid-facial development and Eustachian tube function
Permanent or intermittent hearing loss	Recurrent ventilation tubes in childhood
Bleeding	
Pain	

CT, particularly where there has been significant delay between the time of the CT and surgery.

Magnetic resonance imaging (MRI) is less commonly performed preoperatively as it does not provide detail of the bony anatomy and the diagnosis and the extent of disease can frequently be made from clinical examination and CT scanning. However, where there is doubt about the diagnosis, a diffusion-weighted MRI can be of help in differentiating cholesteatoma from other tissue. In addition, where there is involvement of intracranial structures, an MRI is advisable to quantify the extent of disease and facilitate planning for a joint procedure with neurosurgical colleagues.

Management

Surgical removal of cholesteatoma is the only effective treatment. The principles of surgery are often summed up by the adage that the aim of any surgery for cholesteatoma is to provide, in order of importance, a 'safe, dry, hearing ear'. This provides a technical standard for surgeons but also the primary aim reminds us of our duties as a doctor. An ear should not necessarily be considered 'safe' because it is free from cholesteatoma and associated risks. Rather, in the modern era, a 'safe' ear is one where the surgeon and patient have made informed choices about the potential risks and benefits of surgery. The surgeon must be aware of the likely more symptom-orientated aims and hopes of the patient, who may primarily be hoping to achieve an improvement in hearing, or to be able to return to activities limited by the cholesteatoma. Accordingly, a 'safe' ear is one treated utilizing techniques that facilitate complete removal of disease while minimizing the intraoperative risk to the core functions of the ear as well as other structures that pass through the temporal bone and mastoid.

Where the patient declines surgery, or is deemed to be unfit for prolonged anaesthetic, careful removal of as much squamous debris from the retraction pocket and prompt topical treatment of granulation tissue or infection is required for symptom control. While these patients will often suffer from hearing impairment, the use of the conventional hearing aids may result in increased frequency of otorrhoea, in which case a bone conducting hearing aid should be considered.

Surgical options

Soft tissue approach

The middle ear and mastoid have traditionally been approached either through a transcanal (no incisions outside the external auditory canal), endaural incision(s) (where incisions extend from the external auditory canal between the tragus and root of the helix towards the temporalis muscle) or via a postauricular or Wilde approach (where an incision is made behind the ear and the pinna elevated forward). While the transcanal approach limits morbidity to the patient, it also limits the extent of disease that may be treated and thus its use is typically for limited middle ear and attic disease. The endaural and postauricular approaches may both be utilized to treat more extensive disease, with the choice of approach being primarily guided by the experience and preference of the surgeon. The key principle to both is that the soft tissue approach should allow for clear visualization of the full extent of disease and the operative field required for removal. In both approaches, it is now commonplace for patients to be discharged from hospital on the day of surgery, although those with endaural surgery do so without the placement of a pressure head bandage.

Total endoscopic ear surgery, using an endoscope via a transcanal approach, is becoming increasingly popular for treating cholesteatoma limited to the middle ear and attic. The 120° viewing angle of the endoscope allows for better visualization of the cholesteatoma and structures within the middle ear and attic than is achievable with a microscopic transcanal approach. An increasing array of efficient endoscopic surgical instruments, combined with the benefits to the patient of seemingly incisionless surgery and potential for reduced recovery times, make this an appealing and interesting area of development. However, the technique is more technically demanding of the surgeon and, to date, it is unclear as to what impact total endoscopic ear surgery has on recidivism or hearing outcomes in cholesteatoma.

Canal wall up versus canal wall down

Where cholesteatoma extends beyond the limits of transcanal or total endoscopic ear surgery removal, then removal of some portion of the mastoid bone is necessary in order to achieve complete removal of disease. While a wide variety of techniques have been described, they may be broadly categorized as to whether the posterior bony wall of the ear canal is preserved, referred to as canal wall up (CWU) or closed cavity procedures, or removed, the so-called canal wall down (CWD) or open cavity procedures. CWD procedures represent a more traditional approach, joining the exenterated mastoid and attic air cells to the external auditory meatus to create a much wider meatus, known as a mastoid cavity. CWU surgery was developed and popularized in the 1960s and 1970s in an attempt to avoid the creation of a mastoid cavity and facilitate better hearing results. Both approaches have well-recognized pros and cons and, while advocates of both techniques continue to argue vigorously, no single technique has been adopted as a gold standard.

In CWU surgery, also called a combined approach tympanoplasty, the cholesteatoma is removed via a combination of a transcanal and a transmastoid approach, leaving the posterior bony wall of the external meatus intact. Following an initial cortical mastoidectomy, further exposure of the cholesteatoma is achieved via a posterior and superior tympanotomy (Figure 19.6). Depending on the integrity of the ossicular chain and configuration of the cholesteatoma, it is usual to remove the incus and head of the malleus. The small buttress of bone cupping the anterior aspect of the malleus head known as the cog is then removed to expose the anterior epitympanic recess. The removal of cholesteatoma is then completed via the posterior

exposure and ear canal. While offering the advantage of avoidance of a mastoid cavity, CWU surgery is associated with significantly higher rates of recidivism than CWD surgery. In light of this, CWU surgery is often planned as a staged procedure, with a first stage aimed at removal of disease and a subsequent stage(s) at intervals of 3–6 months with a plan for ossicular reconstruction if the ear is free of disease. However, arguments exist for performance of the surgery and ossicular reconstruction as a single stage with subsequent surveillance for recidivism with diffusion-weighed MRI scanning.

CWD surgery proceeds in a similar fashion to CWU surgery, with an initial cortical mastoidectomy, often with earlier removal of the posterior bony canal wall—a so-called posterior-to-anterior, or back-to-front, approach. CWD surgery may also be performed using an anterior-to-posterior (front-to-back) approach. In this instance, the scutum and adjacent mastoid air cells are removed from anterior to posterior to expose the entirety of the cholesteatoma. While technically more challenging, the advantages of this technique are that the ossicles, lateral semicircular canal, and facial nerve are identified early in the procedure, with subsequent surgery proceeding away from rather than towards these structures, and that the removal of normal mastoid bone is kept to a minimum. Thus, the extent of surgery is determined not by the need for surgical access but is tailored to the extent of the cholesteatoma. Furthermore, where the cholesteatoma is limited, a small cavity may be left open and will often require no aural toilet but yet will remain clearly visible on microscopy for postoperative surveillance. CWD surgery is associated with a lower rate of recidivism than CWU surgery, but commits the patient to the disadvantages of an open cavity (**Figure 19.7**), not least the need for lifelong regular aural toilet. Approximately 40% of CWD patients will suffer repeated or persistent otorrhoea and the cavity is often intolerant of water ingress. In order to facilitate cleaning, a meatoplasty is often required and may result in altered cosmesis of the external ear.

Where an anterior-to-posterior CWD results in a large canal wall defect, rather than create a cavity with the associated quality-of-life implications and requirement for long-term attendance at the outpatient department, the cavity may be obliterated. In addition to avoidance of a mastoid cavity, obliteration is associated with a lower rate of recidivism than an open mastoid cavity. The reason for this reduction in recidivism is unclear, perhaps relating to reduced mastoid gas exchange or, more pragmatically, providing a physical barrier to regrowth of cholesteatoma. Cartilage from either the tragus or concha may be used to reconstruct the canal wall, and bone dust, muscular flaps, or synthetic materials such as bioactive glass used to fill the mastoid defect. Given that a small attic cavity will be the first location of retraction and recurrence and frequently requires no intervention, it may be left unobliterated to facilitate surveillance.

The evidence regarding hearing outcomes between CWU and CWD surgery is equivocal and randomized trials are both difficult to conceive due to the individualized nature of care required and ethically dubious due to the greater risk of recidivism as a result of CWU surgery. When a CT scan has been performed, the likely extent of surgery can be discussed with the patient preoperatively and an appropriate approach chosen. Irrelevant of CWU or CWD technique, patients should be consented for the same risks including the presence of a mastoid cavity (Table 19.2).

Table 19.2 Risks of tympanomastoid surgery

	Wilde/postaural	Endaural	Endoscopic
Approach	Pain	Pain	Pain but significantly reduced
	Bleeding	Bleeding	Small risk of thermal damage from endoscope
	Infection	Infection	Infection
	Scar	Scar (potentially but not necessarily in a cosmetically sensitive area)	
	Altered wound healing (keloid/hypertrophic scar)	Altered wound healing (keloid/hypertrophic scar)	
	Head bandage for 24 hours		
Resection	Incomplete resection of disease/residual/need for second-look surgery	Incomplete resection of disease/residual/need for second-look surgery	Incomplete resection of disease/residual/need for second-look surgery
	Damage to facial nerve (CN VII)	Damage to facial nerve (CN VII)	Damage to facial nerve (CN VII)
	Damage to LSCC	Damage to LSCC	Damage to LSCC
	Damage or removal of ossicles and resultant conductive hearing loss	Damage or removal of ossicles and resultant conductive hearing loss	Damage or removal of ossicles and resultant conductive hearing loss
	Damage to sensorineural hearing/audiologically dead ear	Damage to sensorineural hearing/audiologically dead ear	Damage to sensorineural hearing/audiologically dead ear
	Cerebrospinal fluid leak	Cerebrospinal fluid leak	Cerebrospinal fluid leak
Reconstruction	Failure of graft to take and tympanic perforation	Failure of graft to take and tympanic perforation	Failure of graft to take and tympanic perforation
	Failure of ossiculoplasty to improve hearing	Failure of ossiculoplasty to improve hearing	Failure of ossiculoplasty to improve hearing
	Failure of obliteration	Failure of obliteration	Failure of obliteration

CN, cranial nerve; LSCC, lateral semicircular canal.

Table 19.3 Options available to surgeons depending upon the status of the ossicles

Ossicular status following resection	Ossiculoplasty principle	Method
Incus eroded	Incus to stapes	Cartilage/bone cement/ prosthesis
	Malleus to stapes	Incus transposition/prosthesis
	Tympanic membrane to stapes	Myringostapediopexy/ cartilage reinforced Myringostapediopexy/partial ossicular reconstruction prosthesis
Incus and stapes superstructure absent	Malleus to footplate	Incus transposition/prosthesis
	Tympanic membrane to footplate	Total ossicular reconstruction prosthesis/oval window baffle/ malleovestibulopexy
Incus and stapes absent	Malleus to vestibule	Malleovestibulopexy
	Tympanic membrane to vestibule	

At the time of surgery, one must also consider the implications for the patient of impaired hearing. There are many options as to when, how, and with what to reconstruct the connection between the tympanic membrane and the oval window. Table 19.3 shows the options available to surgeons depending upon the status of the ossicles. A variety of factors including ossicular integrity, experience and preference of the surgeon, and the cost and availability of prostheses affect the choice of the reconstruction performed. A detailed discussion on ossicular reconstruction is outside the limits of this chapter (see Chapter 21). Indeed, the individualized nature of cholesteatoma treatment means that it remains unclear as to which reconstruction technique will result in the best hearing outcomes, with some advocating that the best predictor of postoperative hearing outcome remains the preoperative hearing. That is to say, those who are worse off to begin with will remain worse off postoperatively. However, irrespective of the technique chosen, it is important that the surgeon bases their estimates of the likelihood of successful hearing outcome on their own outcomes when counselling the patient, rather than those published by others.

Postoperative surveillance

Cholesteatomas possess an obstinate tendency to recur and regrow despite surgical intervention. Such recidivism may be either, 'residual', where the cholesteatoma is incompletely removed or 'recurrent', where despite successful initial treatment the tympanic membrane re-retracts and the cholesteatoma reforms. Combined, the tendencies to residual and recurrent disease present a considerable challenge to modern healthcare services and patients continue to require long-term follow-up.

The risks of initially occult residual disease mean that there is ongoing debate surrounding the optimal course of action for postoperative surveillance. In CWU surgery, a planned second stage is a common means of surveillance and, irrespective of technique, some surgeons will perform a second look in all cases, while others decide on a patient-by-patient basis. This consideration must weigh up not only the initial surgical findings but also the wishes, beliefs, and medical history of the patient. Inevitably, each surgeon's view and methodology will be coloured by their training, experience, and indeed the availability of operating theatre space to facilitate timely treatments. Whatever decision is made with the patient, outpatient department follow-up with aural toilet and serial endoscopic imaging of the tympanic membrane is prudent.

Where a patient is being followed up in the outpatient clinic and a suspicion of residual disease arises, or the patient strongly wishes to avoid planned second-stage surgery unless absolutely necessary, a diffusion-weighted imaging (DWI) MRI may be requested. DWI MRI is advantageous over CT in that it avoids further exposure to ionizing radiation and can distinguish cholesteatoma from other tissues. DWI MRI is not without limitations. While the sensitivity (90%) and specificity (95%) for detecting cholesteatoma are both high, they are not 100%. DWI MRI is commonly performed using a 3 mm slice thickness and so disease may not be detected reliably where it is less than 3 mm in diameter. Thus, all images must be correlated with clinical concern and, where available, review of serial endoscopic imaging of the ear. Where clinical suspicion is low, repeat MRI will screen for false negatives, but where clinical suspicion is high, a negative DWI MRI is not cause to avoid a second-look surgery.

Avoidance and management of recurrent disease

Recurrent cholesteatoma can provide an even greater surgical challenge than the initial disease and is probably best managed by experienced otologists. We have already discussed that obliteration reduces recurrence but by what means remains unclear. When assessing the middle ear, it is important not only to assess the ossicles and for the presence of cholesteatoma, but also to consider the ventilation pathways of the middle ear. Where adhesions impair the ventilation to the Eustachian tube they should be taken down in order that postoperatively the entire tympanum may ventilate. This ventilation is with the aim of normalizing middle ear pressure and preventing retraction. Despite these measures, many ears will retract, and where areas are of particular concern they may safely be reconstructed with cartilage. As a more robust graft material than temporalis fascia it is thought that the cartilage resists the negative pressures of the middle ear more effectively, although inevitably in some cases a retraction may occur around the cartilage. While the cartilage undoubtedly increases the mass of the tympanic membrane there is little evidence to suggest a deleterious impact on the patient's hearing. Where the ossicular chain has been damaged, it can be argued that the cartilage graft is of benefit to hearing as it may be placed onto the stapes and form a cartilage myringostapediopexy. The use of ventilator tubes is also an intuitive option to reduce recurrence but there is little evidence supporting the practice and detractors highlight that a ventilator may ventilate

the mesotympanum but that this space is likely separated from the epitympanum by adhesion.

Tips and pearls

- Cholesteatoma should be considered in all patients presenting with recurrent foul smelling otorrhoea.
- Where cholesteatoma is suspected it should not be ruled out without adequate aural toilet and examination with a microscopic or endoscopic image capture.
- A CT scan should be considered in all cases of clinical cholesteatoma prior to embarking on surgical intervention—forewarned is forearmed.
- In cases where doubt exists about the presence of cholesteatoma, a DWI MRI scan can be of benefit in making the diagnosis.
- Careful aural toilet and topical antimicrobial therapy can be helpful in controlling otorrhoea and otalgia prior to surgical treatment.
- Complete surgical removal is the only definitive treatment of cholesteatoma.
- While there is still significant controversy regarding optimal surgical treatment, surgeons should audit their own outcomes to ensure they are not falling significantly below reasonable standards.

Controversies

Pretty much everything other than you have got to get the cholesteatoma out!

Conclusion

In treating patients with cholesteatoma, the modern otologist is in many ways standing on the shoulders of giants. Historically, cholesteatoma would progressively destroy structures within and around the temporal bone, resulting in lasting morbidity, and ultimately could prove fatal. In contrast, the giant leaps of microsurgery and disease clearance mean that modern otologists may focus on more than achieving a 'safe, dry, hearing ear'. Despite the myriad of differing symptoms, pathophysiology, and anatomy that may present, it is our ambition to offer patients not a technical standard but a patient-centred outcome based on symptom management and disease clearance. The full breadth of surgical approaches and reconstructive options may be used to achieve this aim.

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20

Otosclerosis and stapes surgery

Christopher Aldren

Introduction

The main focus of this chapter will be on otosclerosis and its management; however, the management of otogenesis imperfecta and congenital stapes abnormalities will also be considered in more detail.

Basic science

Definition

Otosclerosis is a localized disorder of bone metabolism of the otic capsule that is characterized by disordered resorption and deposition of bone. It is thought to result from increased pathological bone remodelling and the basic lesion consists of areas of bone resorption by osteoclasts and new bone formation by osteoblasts, accompanied by vascular proliferation.

Many patients with otosclerosis are asymptomatic and clinical otosclerosis with associated hearing loss only occurs when the lesions involve the stapes footplate or, much more rarely, the round window, causing a conductive hearing loss. Sensory hearing loss is thought to occur when lesions involve the cochlear endosteum.

The commonest site of otosclerosis is the cochlear wall just anterior to the stapes footplate.

Aetiology

The aetiology of otosclerosis remains obscure. There is a clear familial history of hearing loss in approximately half of patients.

The measles virus has been suggested as a possible cofactor in the development of otosclerosis but the available evidence remains conflicting and a number of recent studies have cast doubt on this link.[1]

Genetics

Otosclerosis occurs in familial and non-familial forms. The familial form accounts for 25–50% of cases and has an autosomal dominant inheritance pattern with incomplete penetrance. While a number of gene associations have been noted in affected families the genetic cause of otosclerosis remains largely unidentified.[2]

Incidence

Histological otosclerosis in random series of temporal bones harvested in Belgium showed histological evidence of otosclerosis in 3.4%. The British National Study of Hearing showed the clinical prevalence is around 2% in adults. This increased with age to 3% in the 60- to 80-year-old age group. Only 10% of those patients had surgery which explains the lower incidence reported in surgical series. Interestingly, this study showed similar prevalence in men and women but that the women had more severe hearing loss which may account for the finding, commonly reported in clinical series, that otosclerosis is commoner in women by approximately 2:1.[3]

Natural history

Although no perfect studies are available, the average air–bone gap typically deteriorates about 1 dB per year. This is in addition to a sensory hearing deterioration of another 1 dB per year. Clearly in many cases the deterioration is significantly quicker.

Assessment

Patients are typically young adults presenting with a history of progressive hearing loss. Most patients develop bilateral hearing loss over time but the loss may progress at different rates in the two ears and so initially may appear to be unilateral. Half of patients have a first-degree family member with otosclerosis or hearing loss. Two out of three have associated tinnitus.

The tympanic membrane is usually normal in appearance. In some patients it is possible to see a reddened area over the promontory through the tympanic membrane. This is due to dilated arteries on the medial wall of the middle ear due to increased vascularity of the otosclerotic foci. It is known as a flamingo flush or Schwartze's sign.

The patients have a conductive hearing loss usually with a negative Rinne test.

Investigations

Audiometry

Pure tone audiometry will confirm the conductive hearing loss which is usually worse at lower frequencies. The bone conduction is

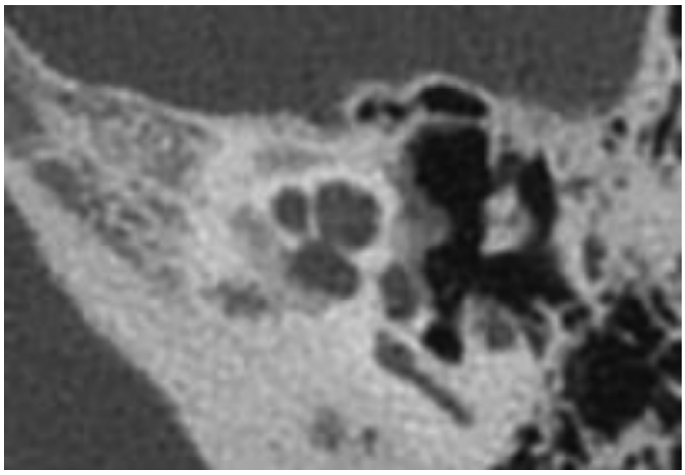

Figure 20.1 Severe otosclerosis around cochlear and obliterating the stapes footplate, left ear.

This is known as Carhart's notch (see Chapter 12). If the cochlear is involved, patients may have a mixed hearing loss. Pure sensory hearing loss from cochlear otosclerosis with no involvement of the ossicular chain is rare. Tympanometry may show reduced compliance due to fixation of the ossicular chain. Stapedial reflexes are normally reduced or absent.

Imaging

While stapes surgery has been performed very successfully before the times of computed tomography (CT) scanning, many surgeons now like to have a preoperative CT scan. A high-resolution or cone beam CT scan can frequently confirm the diagnosis with the appearance of a lucency in the area anterior to the oval window (Figure 20.1). It can also be useful to show other possible differential diagnoses such as superior semicircular canal dehiscence or fixation of the malleus (Figure 20.2). CT scanning can forewarn of possible difficult surgical situations such as an obliterative footplate or dehiscence of the facial nerve and is therefore very useful for more junior surgeons who may wish to refer these potentially tricky cases elsewhere (Figure 20.3).

Differential diagnoses

Any cause of conductive hearing loss with a normal tympanic membrane can be a differential diagnosis. These include the following:

- Congenital fixation: hearing loss would be expected from birth.
- Tympanosclerotic fixation: there may be a history of previous ear infections and tympanosclerosis of the tympanic membrane may be visible on otoscopy.
- Stapes fixation owing to osteogenesis imperfecta (OI): patients with this hereditary connective tissue disorder usually are aware of their OI by the time they develop hearing loss due to multiple bone fractures, but may not necessarily associate it. Up to 90% of OI patients will develop hearing loss which typically comes on at an earlier age (second and third decades) than seen in otosclerosis patients. Hearing loss can be conductive, mixed, or sensory neural. Patients frequently have a family history of OI. They may have blue sclera, underdeveloped teeth, and short stature.

Treatments

Hearing aids

As most patients with otosclerosis have a predominantly conductive hearing loss, conventional hearing aids offer an excellent management option and should always be discussed with the patient. In patients with bilateral hearing loss opting for surgery, a hearing aid should be offered for the non-operated ear long term or until they opt for second-side surgery.

Bone-anchored hearing aids

Bone-anchored hearing aids are a useful method of hearing rehabilitation in patients who are unsuitable for conventional aids and who do not wish to have surgery.

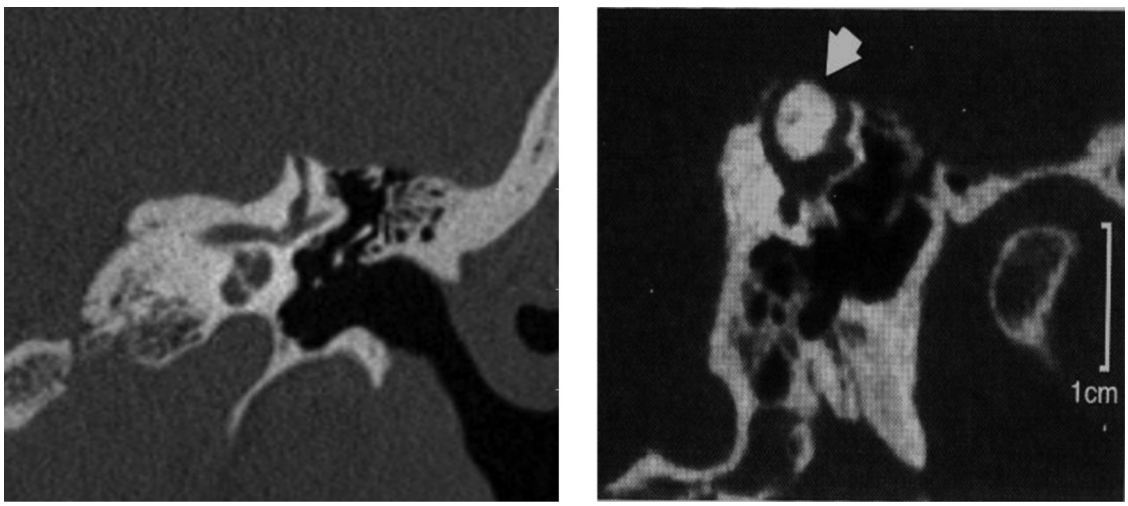

Figure 20.2 Superior semicircular canal dehiscence, left ear.

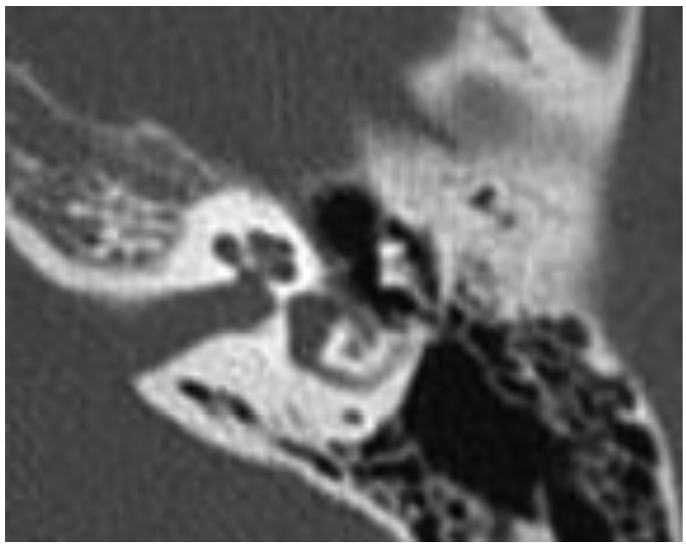

Figure 20.3 Dehiscent facial nerve over oval window, left ear.

Far-advanced otosclerosis

Far-advanced otosclerosis is defined as average hearing thresholds of over 85 dB with unmeasurable bone conduction. This can occur in patients with conductive hearing loss from otosclerosis combined with sensory hearing loss from ageing, cochlear otosclerosis, or any other cause of sensory loss. These patients are treated with surgery and a hearing aid, direct acoustic cochlear stimulation, or by cochlear implantation. Most clinicians offer surgery and hearing aids in the first instance with cochlear implantation reserved for those who fail the first treatment option.

Cochlear implantation

This is offered for patients with far-advanced otosclerosis who are unsuitable for or who have failed conventional surgery and provision of a hearing aid. The surgery can be more tricky than normal due to occasional obliteration of the round window with otosclerosis. Facial nerve stimulation by the implant causing facial twitching is commoner in otosclerosis patients possibly due to demineralization of the otic capsule.

Direct acoustic cochlear stimulation

This device is a powered stapes prosthesis. A stapedotomy is performed but the prosthesis is powered with an active middle ear implant. This in effect combines the benefits of the stapedotomy with additional gain from a hearing aid. It is useful for far-advanced cases who are deemed too good for a cochlear implant.

Medical management

While oral fluoride has been proposed to prevent further deterioration in otosclerosis the evidence for its use is debated and currently it is not used in the UK.

The use of bisphosphonates to prevent deterioration is in its infancy and studies show conflicting evidence on benefit. Future studies may show efficacy but they are currently not used widely.

Surgical management

Consent for surgery

Patients with unilateral or mild bilateral loss may elect for no treatment; however, as hearing aids give an excellent outcome for otosclerosis patients they should be offered to all patients. Those contemplating surgery should be aware of the risks and success rates. As risks are operator dependent, ideally surgeons should quote their own figures. Total sensory hearing loss is reported to be in the region of 0.4–2%. Closure of the air–bone gap to within 10 dB is achievable in 80% up to the high 90s%. Some distortion of hearing and discomfort with louds sounds may be reported in the first few weeks after surgery but typically settles quickly. In patients with preoperative tinnitus it typically will improve or resolve completely in 75% of cases but can become worse in 5%. In those with no preoperative tinnitus it can develop in up to 10% of cases but usually is not troublesome.[4] Alteration in taste due to damage to the chorda tympani occurs in up to 30% but in most cases lasts only a few months.[5] Vertigo has become much less common with the move to small fenestra stapedotomy. When it occurs, it is usually in the immediate postoperative period and typically settles quickly. Benign positional vertigo presumably from the trauma of surgery is reported in the literature following surgery but can be treated with particle repositioning. Facial nerve injury is very rare; however, direct trauma from laser or drill is possible. A delayed facial paresis can occur as with any case of middle ear surgery. While the aetiology of such delayed palsies is debated, they are usually treated with oral steroids and typically recover completely within 6 weeks.

Surgery

Stapedectomy was first described by John Shea in 1956. This operation involves removing the fixed stapes and placing a piston from the incus through the stapes footplate into the vestibule. The basic technique Shea described remains in place; however, refinements have taken place over the last 64 years.[6] The main modification of the technique has been to perform a small stapedotomy rather than removing the whole stapes footplate. This has reduced the risk of total sensory hearing loss and probably improved high-frequency hearing results.

The surgery can be performed per aurally through a speculum or using an endaural incision. More rarely a postauricular incision is used. The author prefers a per aural approach using a speculum held with a Yasargil speculum holder. This gives an excellent view, is quick, and relatively bloodless. The holder and speculum also provide an excellent support to stabilize the surgeon's hands.

The stapedotomy surgery consists of a number of steps:

The ear canal is injected. The author only uses adrenaline 1:100,000. If a local anaesthetic is used, on occasions some will track under the canal flap into the middle ear and can cause a temporary facial palsy. While this will settle it does create anxiety in patient and surgeon.

If a vein graft is to be used to seal the inner ear it is now harvested.

A posterior tympanomeatal flap is raised and the annulus raised.

The stapes now has to be exposed which usually requires removal of some posterosuperior canal bone to expose the stapes back to the pyramid and the incus long process up to the level of the facial nerve superiorly. Adequate exposure makes the subsequent surgery significantly easier. The bone removal is performed with a bone curette or a drill. Some surgeons use a hammer and chisel which can remove bone quickly but need to be used with care. Whichever technique is used care must take care to preserve the chorda tympani which is present in this area. Evidence suggests it is better to stretch and preserve the nerve rather than cut it.[5]

The incudostapedial joint is next divided with a joint knife. The joint lies more medially than at first obvious. Once the joint is divided the mobility of the malleus, incus, and stapes is checked. This confirms the diagnosis.

The stapes superstructure is now removed. There are a variety of methods of removing the stapes superstructure. The stapes crura can be drilled, cut with scissors, or fractured with a needle. The author prefers to use the KTP or diode laser as this reduces the risk of stapes dislocation when removing the superstructure. Many surgeons use a laser when performing the stapedotomy but there is no good evidence to favour laser over microdrill or hand trephine.

Ugo Fisch describes a technique called reverse stapedotomy where the stapedotomy is performed before dividing the incudostapedial joint. The piston is then placed onto the relatively fixed and stable incus. Only when the piston is in place is the incudostapedial joint divided and the stapes superstructure removed. This makes the procedure of placing the piston a little easier; however, it is only possible when there is a good view of the stapes footplate and enough room to perform the stapedotomy with the stapes superstructure in place. There are also concerns that separating the stapes superstructure when there is a piston in place may expose the inner ear structures to trauma.

Once the stapedotomy has been fashioned a vein graft is placed if used. The piston is then placed and crimped (**Figure 20.4**). There are many piston designs, but the literature shows little to choose between them. Some surgeons prefer self-crimping designs and thermalloy smart pistons are increasingly popular. These pistons are placed onto the incus, heat is then applied and the piston self-crimps onto the incus. Advocates say the surgery is more straightforward and results are more consistent.

A larger piston diameter should theoretically give better hearing but the differences are small. Most surgeons use a piston with a diameter between 0.4 and 0.6 mm.[7]

A soft tissue seal is often placed to prevent a leak of perilymph through the stapedotomy and around the piston. Some favour a vein or perichondrial graft placed over the stapedotomy before the piston is placed to seal around it. In these techniques allowance for the graft thickness must be made when drilling the stapedotomy. Typically, a 0.7 or 0.8 mm stapedotomy would be created for a 0.4 mm piston. If no graft is placed under the piston a smaller hole is created. In this case, typically 0.5 mm for a 0.4 mm piston or 0.7 mm for a 0.6 mm piston. In this circumstance, the fit is tighter, and surgeons may then choose to place either no graft or a blood or fat graft over the stapes footplate around the piston. There is no convincing evidence of superior results with either technique although the vein graft technique is technically more challenging. French air force pilots are only allowed to fly after stapes surgery if they have had a vein graft placed under the piston.

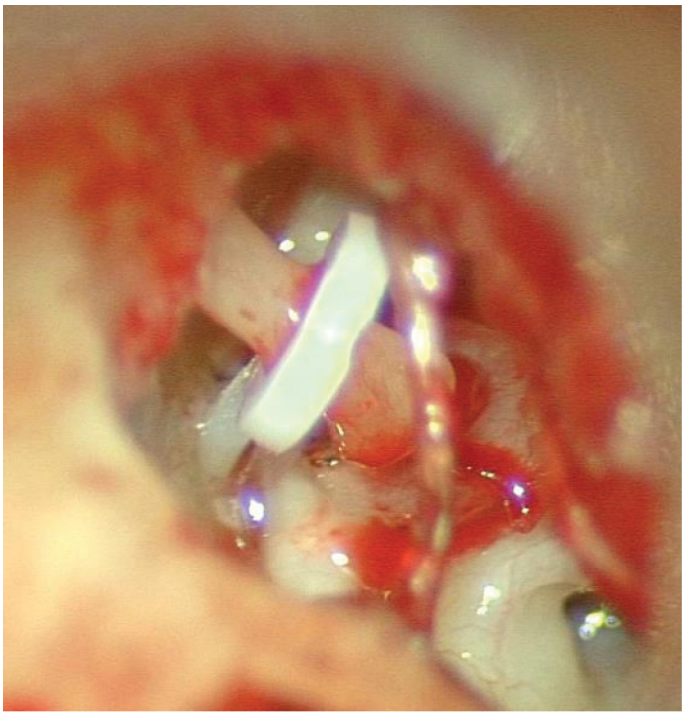

Figure 20.4 Causse Teflon piston in place on incus, right ear.

Once in place the piston should be checked to make sure it is in the stapedotomy by gently pushing the shaft forwards. If in place there is little movement but if the piston is too short it will move forwards out of the stapedotomy. It is also possible to move the malleus and look for movement of fluid placed over the round window.

The tympanomeatal flap is then replaced and the ear packed.

Many surgeons perform the surgery as a day case with the patient kept for 2–3 hours postoperatively. The hearing should be checked by the bedside before the patient is allowed home. Also check for nystagmus and facial nerve function.

Surgery for osteogenesis imperfecta

The surgery is very similar to that of otosclerosis but the chance of a footplate fracture and sensory hearing loss are thought to be higher.

Surgery for congenital stapes fixation

CT scanning for congenital fixation is valuable as these patients can have other ear abnormalities. It is important to look for a widened vestibular aqueduct which may lead to a perilymph gusher if a stapedotomy is performed. It is also important to establish the course of the facial nerve which may be abnormal and on occasions can cross the stapes footplate.

Revision stapes surgery

This is usually performed when there is a significant postoperative conductive hearing loss which maybe immediate or come on at a later stage. The commonest reasons are erosion of the long process of the incus, displaced prosthesis, too short prosthesis, dislocated incus, or incorrect diagnosis (**Figure 20.5**). This surgery should only be performed by experienced stapes surgeons. CT scanning is useful preoperatively (**Figure 20.6**). A laser is very useful in revision surgery to divide soft tissue at the footplate.

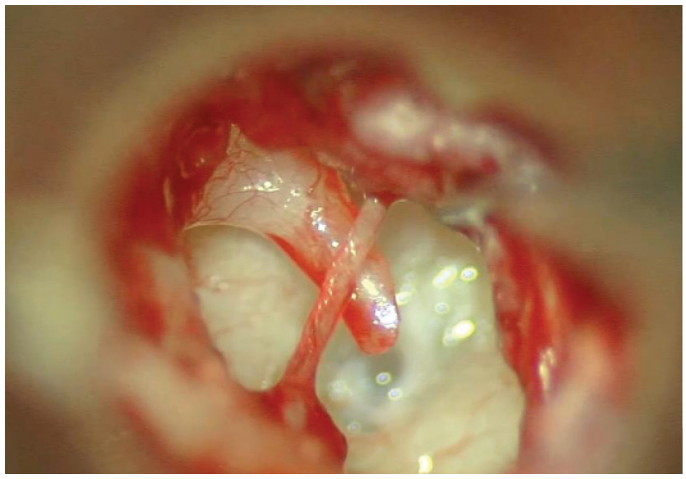

Figure 20.5 Eroded incus, right ear.

In cases of eroded incus, it may be possible to secure a self-crimping piston tightly to the shortened incus. Cement can be used to secure the new piston to the eroded incus. If the incus is very short then a malleostapediopexy can be performed where a longer piston can be placed from the malleus to the stapedotomy.

Managing complications

Peroperative complications

Damage to the chorda tympani

If the chorda is damaged there is debate whether it is better to fully divide it rather than leave it stretched. The author favours retaining the nerve whenever possible.

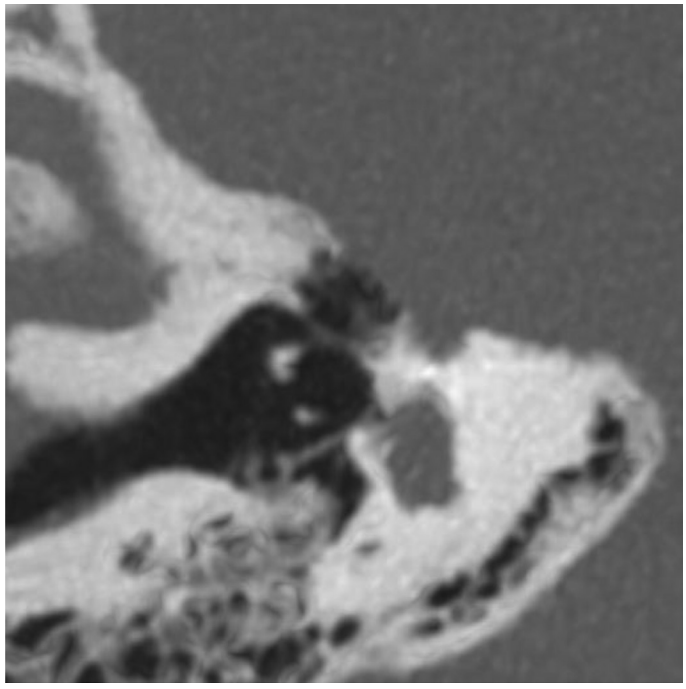

Figure 20.6 CT scan of revision case with piston seen off incus lying free in middle ear, right ear.

Dislocation of the incus

The incus can be dislocated from the malleus when the posterior bony rim is curetted or during division of the incudostapedial joint. If this occurs there a number of options. Some surgeons would endeavour to relocate the incus and abandon the procedure planning to return in 6 months when hopefully the malleoincudal joint would have reformed. The author would favour attaching the stapes piston directly to the malleus to perform a malleostapediopexy.

Floating footplate

If the stapes becomes mobile during division on the incudostapedial joint or of the crura then most surgeons would abandon the procedure. The mobile footplate may allow good hearing if the stapes superstructure and the incudostapedial joint are both still in place. If the stapes has been removed and only the footplate remains then it is possible to place a piston from the incus onto the now mobile footplate. It is likely that the stapes footplate will refix over time and the operation can be completed 6 months later. Some surgeons would perform a laser stapedotomy through the mobile footplate and place the piston.

If a fragment of the footplate falls into the vestibule it is best to leave it but usually possible to place a piston as usual.

Perilymph gusher

Rarely when the footplate is penetrated perilymph can well up into the middle ear. The footplate can be packed with vein, fascia, fat, or Gelfoam. If a vein graft is used it is sometimes possible to place the graft and piston to plug the stapedotomy.

Postoperative complications

Vertigo

Vertigo is rare after surgery and when it does occur is usually mild, short-lived, and can be managed with reassurance. Occasionally, however, there can be severe true rotatory vertigo occurring suddenly within the first few days postoperatively. It may be accompanied by significant sensory hearing loss. While this clearly represents an insult to the inner ear the exact cause is still unknown. Suggested causes are intralabyrinthine infection, bleeding, or perilymph leak. There is little evidence that interventions influence outcome in these cases, but most surgeons would give high-dose oral steroids and antibiotics. There is possibly also a role for intratympanic steroids. The vertigo should be managed with vestibular sedatives and will usually settle fully but the sensory hearing loss may not.

Some patients can develop benign positional vertigo after surgery. This can be treated with particle repositioning manoeuvres.

Dead ear

Sudden onset of severe or profound sensory hearing loss postoperatively is managed as above and is frequently accompanied by severe vertigo.

Tinnitus

In most patient tinnitus improves with successful surgery but on occasions tinnitus can develop postoperatively. It should be managed with reassurance that it will improve over time but if it fails to do so then should be managed as tinnitus from other causes (see Chapter 6).

Controversies in stapes surgery

Age of patient

There is no upper age limit for stapes surgery and as described above in patients with severe mixed hearing loss, stapedotomy may allow patients to use a hearing aid.

While true otosclerosis can occur in children it is very rare and congenital stapes fixation should be considered as an alternative diagnosis. Most surgeons would defer surgery until the child can understand the issues of consent and the risks of surgery. In the interim, hearing aids should be offered. Most surgeons would perform a CT scan before operating on a child for stapes fixation.

Flying after stapes surgery

While there are no absolute rules most surgeons recommend not flying after stapes surgery for at least 2 weeks.

Scuba diving

Most surgeons recommend at least a delay to returning to diving with 50% of surgeons recommending a life-long ban on scuba diving.

Unilateral otosclerosis

When considering surgery for patients with unilateral otosclerosis, it is important to consider the chance of achieving symmetry of hearing after surgery. This means looking carefully at the bone conduction thresholds of the proposed ear. Patients need careful counselling on the likely outcomes of surgery.

Second-side surgery

Most stapes surgeons will happily operate on the second ear provided the first-side surgery was successful and the appropriate audiometric criteria for the second ear are met. It is prudent to wait at least 6 months before operating on the second ear.

Air–bone gap

Traditionally a minimum average air–bone gap of 20 dB is felt to be a sensible threshold to justify surgery although in some circumstances experienced surgeons may reduce this required level.

Guidelines

There are currently no guidelines for the management of otosclerosis.

Conclusion

The aetiology of otosclerosis remains obscure. The hearing loss of otosclerosis is well treated with hearing aids which must always be offered to patients. The operation of stapedotomy remains an excellent treatment but does have inherent risks. Surgeons performing this surgery should be dedicated otologists and should keep records of their results so they can properly consent their patients.

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21

Ossiculoplasty

Christopher Aldren

Introduction

This chapter focuses on surgical reconstruction of the ossicular chain to restore hearing in cases of conductive hearing loss.

Basic science

Definition

Ossiculoplasty is the reconstruction of the ossicular chain and is performed to restore hearing in cases of conductive hearing loss. It is often performed together with reconstruction of the tympanic membrane, when the term tympanoplasty may be used.

Aetiology

Ossicular chain abnormalities leading to conductive hearing loss can be congenital or acquired. They can be due to discontinuity or fixation.

Congenital

Congenital causes of conductive hearing loss can be due to isolated ossicular problems but may be associated with abnormalities of the external auditory canal, pinna, and inner ear.[1]

Fixation of the stapes footplate is the commonest abnormality. There can also be fixation of the ossicles in the epitympanum. Stapes superstructure abnormalities are also seen. These abnormalities can be associated with abnormal course of the facial nerve. Preoperative computed tomography (CT) scanning is essential for these challenging cases. Surgery is usually delayed until the child is able to provide informed consent.

Acquired

Acquired defects of the ossicular chain usually occur due to chronic otitis media with or without cholesteatoma. The commonest abnormality is erosion of the incudostapedial joint and the long process of the incus. The stapes superstructure, body of incus, and malleus head can also be eroded, usually in the presence of cholesteatoma, but erosion of the malleus handle is less common.[2]

The other major cause of acquired ossicular discontinuity is secondary to trauma where the incus and stapes are most at risk of damage.

Assessment

History

Patients should be asked about magnitude and duration of hearing loss and associated disability. Associated symptoms such as tinnitus and vertigo should be sought. Discharge should raise the possibility of current active middle ear disease.

Otoscopy should always be performed preferably with a microscope and/or a Hopkins rod otoscope. Look for abnormalities of the tympanic membrane and the attic area. Check for an associated cholesteatoma. Look at the incudostapedial joint which is frequently eroded.

Try to assess the middle ear ventilation as this has a major impact on the chances of getting a good audiological result from surgery.

Investigations

Audiometry

A pure tone audiogram should be performed. An air–bone gap of greater than 30 dB would be a good indication for surgery although smaller gaps can be accepted if surgery also aims to reduce infection or prevent development of or remove a cholesteatoma.

Tympanometry may help in the assessment of middle ear ventilation.

Imaging

Cone beam or high-resolution CT scanning is helpful in assessing the ossicular status and middle ear ventilation prior to surgery. Scanning may identify pathology in the nose and paranasal sinuses. Anatomical variations which may make surgery technically challenging can also be identified, such as a dehiscent facial nerve overhanging the stapes footplate or ossicular fixation (**Figures 21.1 and 21.2**).

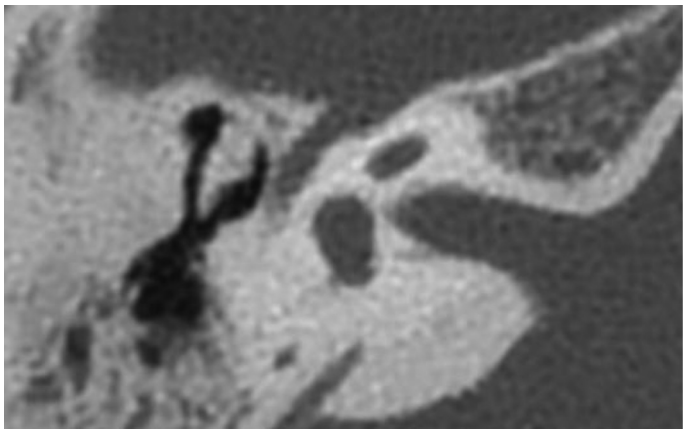

Figure 21.1 CT scan fixed malleus, right ear.

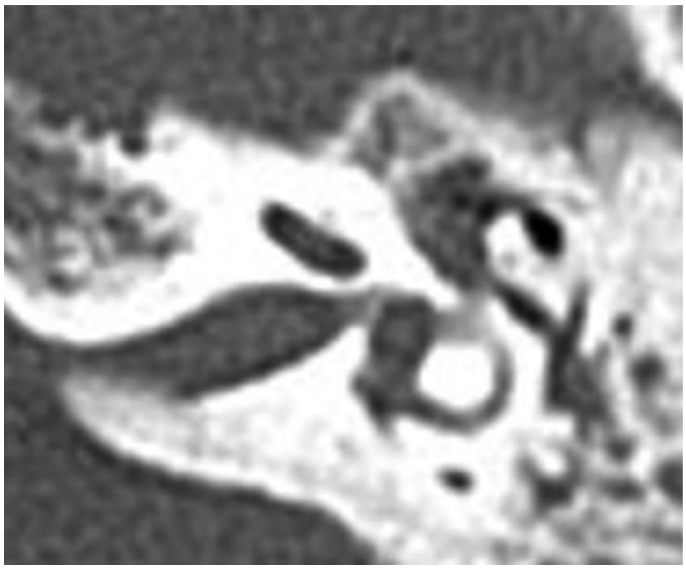

Figure 21.2 CT scan fixed incus, left ear.

Ossicular status

Several scoring systems for assessing the status of the ossicles are available. The Austin classification (Table 21.1) is perhaps the most commonly used.

Scoring of middle ear status

Results from ossiculoplasty are dependent on many factors. In particular:

- State of middle ear ventilation.
- Status of the ossicular chain.
- Presence of cholesteatoma.
- Presence of infection.
- Stability of the prosthesis.
- Ability and experience of operating surgeon.

A number of surgeons have tried to score the above factors to allow prediction of likely success. Scoring systems such as Jack Kartush's Middle Ear Risk Index (MERI) (Table 21.2) and Bruce Black's SPITE (Surgical, Prosthetic, Infection, Tissue, Eustachian) have been used.[3] These scoring systems help to predict the likely outcome from surgery. While none is perfect in predicting success, they reflect the belief of surgeons that the preoperative status of the middle ear has a major impact on the likelihood of a successful hearing outcome.

Table 21.1 Austin classification

Group	Ossicular status	
A	Malleus and stapes present	M+S+
B	Malleus present, stapes absent	M+S−
C	Malleus absent, stapes present	M−S+
D	Malleus and stapes both absent	M−S−

Table 21.2 Kartush Middle Ear Risk Index (MERI)

Risk factor	Score
Otorrhoea (Belluci)	
I Dry	0
II Occasionally wet	1
III Persistently wet	2
IV Wet, cleft palate	3
Perforation	
None	0
Present	1
Cholesteatoma	
None	0
Present	2
Ossicular status (Austin)	
M+I+S+	0
M+S+	1
M+S−	2
M−S+	3
M−S−	4
Ossicular head fixation	2
Stapes fixation	3
Middle ear granulations or effusion	
No	0
Yes	2
Previous surgery	
No	0
Staged	1
Revision	2
Smoker	
No	0
Yes	2

Differential diagnoses

These patients typically have a conductive hearing loss. The differential diagnosis therefore includes middle ear effusion, otosclerosis, and superior semicircular canal dehiscence.

Associated conditions which may cause an ossicular discontinuity or fixation are chronic suppurative otitis media, cholesteatoma, and tympanosclerosis.

Treatments

Patients have three treatment options:

1. No treatment and accept the hearing loss which may be the best option if the hearing loss is mild and particularly if the other ear has normal hearing.
2. Unilateral or bilateral hearing aids.
3. Surgical intervention.

The presence of associated conditions mentioned above may help determine the treatment option recommended.

Hearing aids are frequently a good option for these patients although in the presence of associated pathologies causing otorrhoea they may not be appropriate.

Bone-anchored hearing aids may be a good option for patients where a conventional aid is not appropriate and if surgical conditions do not look favourable.

Powered middle ear implants. The Vibrant Soundbridge can be an option either attaching directly to the stapes or to the incus. Other active powered middle ear implants have been withdrawn in the UK.

Cochlear implantation is used for patients who have a severe or profound hearing loss due to a mixed conductive and sensory hearing loss making them eligible on audiological grounds.

Medical management is for the underlying associated conditions such as drops for chronic otitis media or externa and steroid nasal sprays for rhinosinusitis. Middle ear ventilation may be helped with the use of an Otovent balloon. The role of balloon dilatation of the Eustachian tube in improving middle ear ventilation prior to surgery is still under investigation.

Surgical management

Consent for surgery

Ideally surgeons should have a database of their own surgical results to allow them to consent patients realistically. Generally, hearing results for ossicular reconstruction are better when an intact mobile stapes is present. Good results are hard to achieve in a poorly ventilated middle ear and are generally worse in the presence of cholesteatoma.

Hearing improvement to an air–bone gap of less than 20 dB is variably quoted as between 60% and 90% when a stapes is present and between 30% and 80% when it is absent. The large variation in results reported in the literature may reflect the choice of patients in the studies but emphasizes the need for surgeons to keep their own database to allow accurate informed consent.

Patients should be warned of the risk of partial and total hearing loss, alteration in taste, tinnitus, vertigo, and facial weakness. The risks of these complications vary according to the underlying pathologies causing the ossicular chain disruption.

Surgical approach

The surgical approach may be dictated by associated pathologies such as cholesteatoma in the mastoid or a perforated tympanic membrane. Where possible, the author prefers a per aural approach using a speculum and a Yasargil speculum holder. This speeds up the surgery and healing, reduces bleeding, and the speculum holder gives a stable platform on which to rest the hands and reduce tremor.

Endoscopic surgery

The use of otoendoscopes in ossiculoplasty is growing in popularity. Hearing results appear similar to those achieved using the microscope although with a trend towards shorter operating times.[4] Visualization of the reconstruction may be better. Associated pathologies such as extensive cholesteatoma may determine whether a purely endoscopic approach is possible. The author mainly uses the microscope but frequently uses the endoscopes to check the position of the reconstructed ossicular chain.

Choice of prostheses

To be effective and long-lasting, prostheses need to be stable and biocompatible. Over the years many materials have been used. Most of the current prostheses are made of titanium or hydroxyapatite. Both materials are stable in the middle ear; however, titanium may incite a reaction of the tympanic membrane and so most surgeons will place cartilage between the tympanic membrane and the prosthesis head.[5] Hydroxyapatite is usually placed without intervening cartilage.[6]

Stapes present

Reconstruction of incudostapedial joint

When the incudostapedial joint is partially eroded, or if the defect between the eroded incus and the stapes head is small, then it is possible to try to recreate the incus long process. This is most commonly accomplished with cement from the incus to the stapes head (Figure 21.3). Cements are either hydroxyapatite or glass ionomers. Fascia,

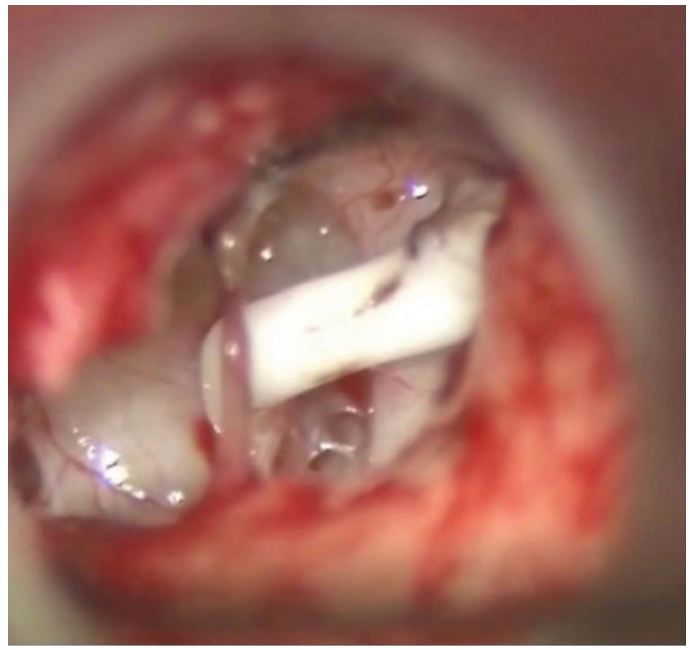

Figure 21.3

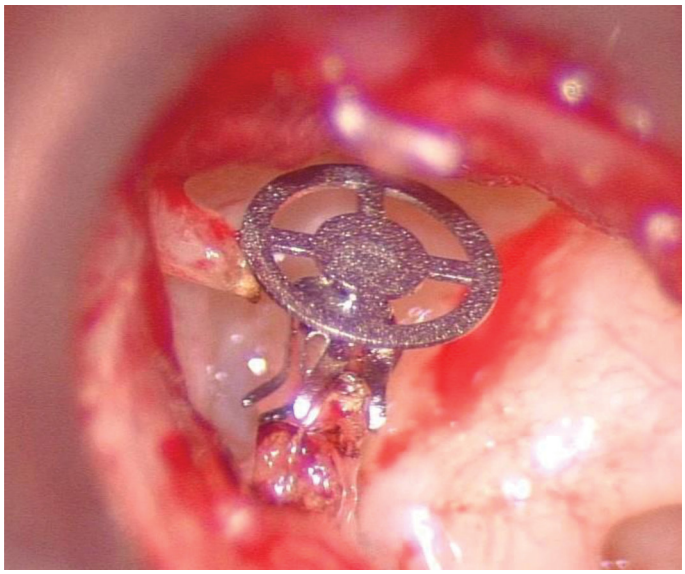

Figure 21.4 Kurz partial clip prosthesis on stapes head, right ear.

cartilage, bone grafts, and titanium prostheses can all also be used to achieve a reconstruction of the joint.[7]

Myringostapediopexy

When the stapes is present and the incus is absent, it is possible on occasion to place the tympanic membrane directly onto the stapes head. This is called a myringostapediopexy. It is most frequently used when a modified radical mastoidectomy has been performed as the posterior canal wall can be lowered to the level of the facial ridge which reduces the depth of the middle ear and allows the reconstructed tympanic membrane to sit on the stapes head. If the middle ear is well ventilated, good hearing results can be obtained.

Partial ossicular reconstruction

In most cases where the posterior canal wall is in the normal position the tympanic membrane lies too far lateral to allow a myringostapediopexy to be performed. In such cases most surgeons place a prosthesis or refashioned ossicle between the stapes head and the malleus handle or tympanic membrane. The prostheses are known as partial ossicular replacement prostheses or PORPs. Stability is critical here and the author has found, in his hands, the KURZ partial clip prosthesis (**Figure 21.4**) to be easy to place and to give superior audiological results due to its excellent stability (**Table 21.3**).

Other surgeons use a total ossicular replacement prosthesis (TORP) from the malleus handle or tympanic membrane to the stapes footplate even when the stapes superstructure is present. Robert Vincent describes a technique of connecting a TORP to the stapes superstructure with a rubber band from the shaft of the prosthesis to the stapes head which is secured underneath the divided stapedius tendon. This extra point of stabilization is said to improve hearing results.[8]

Table 21.3 Results using KURZ partial clip prosthesis

	Postoperative air–bone gap <20dB	Preoperative air–bone gap dB	Postoperative air–bone gap dB
With cholesteatoma	84%	23.2	12.9
Without cholesteatoma	95%	28.3	9.4

Stapes absent

Where the stapes is absent, the ossicular reconstruction must bridge the gap between the vibrating tympanic membrane and the mobile stapes footplate. In this situation a TORP or fashioned autologous columellar is used. The difficulty in this situation lies in not only fashioning a reconstruction of the correct length but also in maintaining stability. Laterally this can be improved by placing a prosthesis with a groove under the malleus handle, but this only works when the malleus handle is present and overlying the stapes footplate. If the malleus is anteriorly placed or very medial it cannot be used in its original position. In these situations, most surgeons place the prosthesis directly under the tympanic membrane with or without an intervening cartilage graft. Some prostheses have a spike in the head which locates into the overlying cartilage in an attempt to improve lateral stability.

Medially, the prosthesis sits on the footplate. A number of footplate shoes have been devised to try to improve the stability at the footplate (**Figures 21.5 and 21.6**).[9]

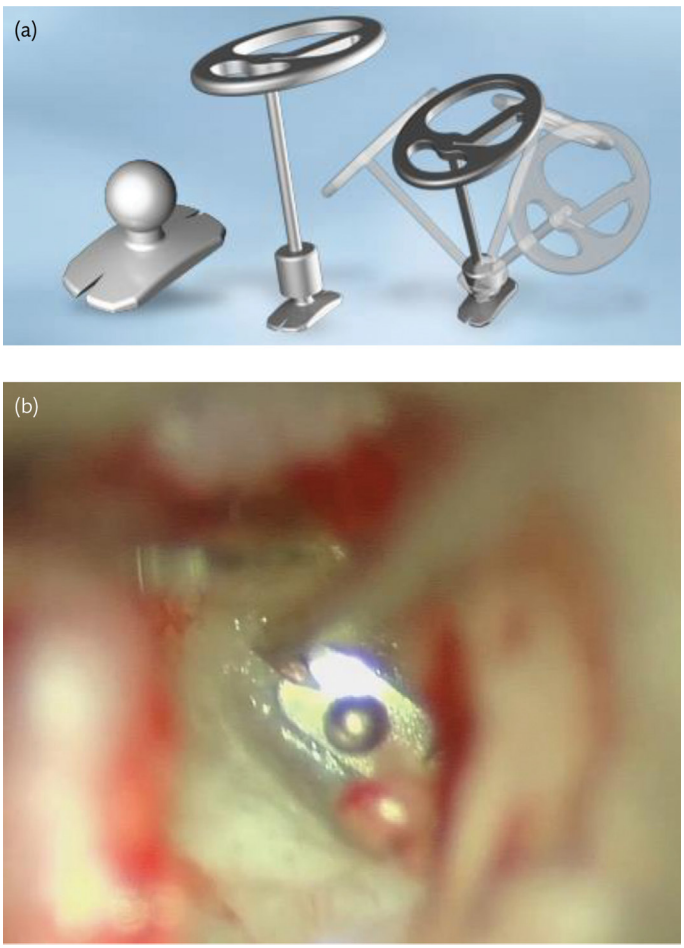

Figure 21.5 (a) Kurz Omega connector. (b) Omega connector on stapes footplate, left ear.

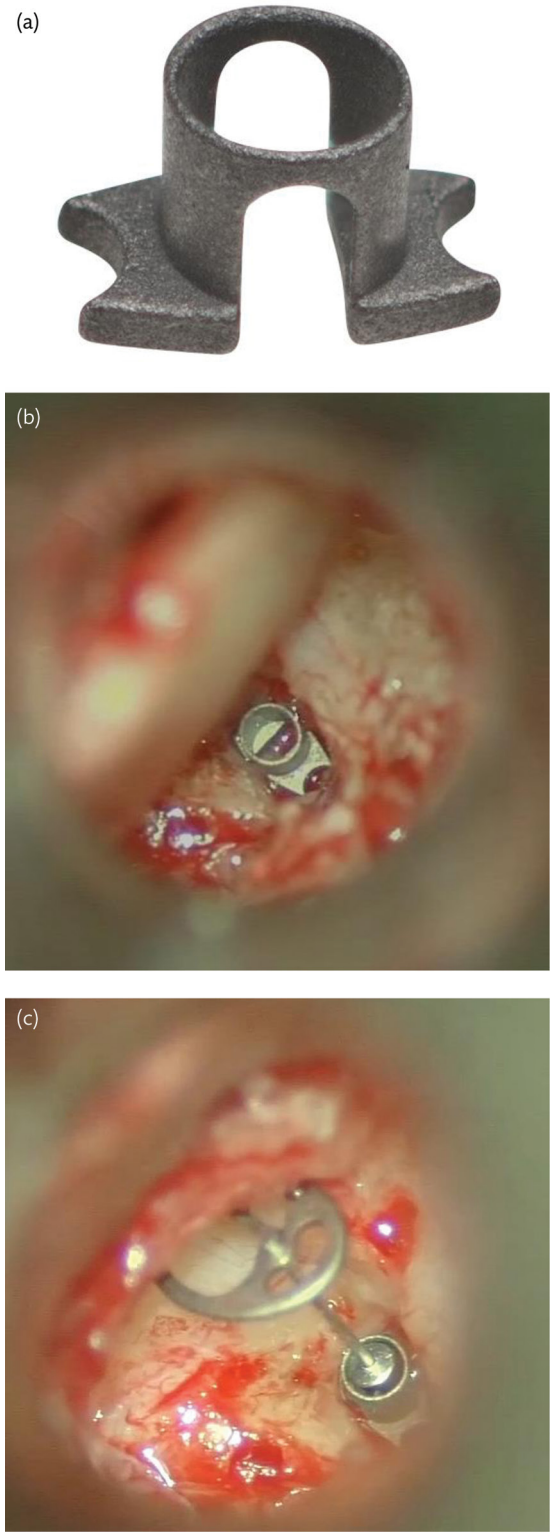

Figure 21.6 (a) Dornhoffer shoe. (b) Dornhoffer shoe on stapes footplate, left ear. (c) Kurz Variac TORP in Dornhoffer shoe, left ear.

Relocation of an anterior malleus

Some surgeons faced with an anterior malleus handle choose to relocate it to a more posterior position overlying the stapes footplate. This is achieved by separating the malleus from the tympanic membrane, then removing the incus remnant, dividing the tensor tympani tendon, and pulling the malleus posteriorly. The anterior malleolar ligament will suddenly stretch, and the malleus is then free to be placed posteriorly. It is only held by the superior malleolar ligament.[10]

Figure 21.7 Malleus replacement prosthesis.

Absent malleus

When the malleus handle is absent, surgeons usually place the prosthesis directly under the tympanic membrane with or without intervening cartilage. A malleus replacement prosthesis has been designed by Robert Vincent.[11] This titanium prosthesis is located into two holes drilled through the scutum. The neomalleus handle can then be placed directly over the stapes footplate to allow lateral stability. This prosthesis works particularly well when used with a prosthesis with a central groove such as the Vincent ALTO from Grace Medical (Figure 21.7).

Revision of ossicular surgery

If the underlying conductive hearing loss does not improve with surgery, or if it recurs at a later date, then revision surgery may be justified. A rigorous reassessment of the case with a repeat CT scan and audiology should be performed. The scan may show why the case has been unsuccessful. Again, the state of middle ear ventilation is important in determining the chances of success.

At revision surgery the chorda tympani is frequently attached to the tympanic membrane by scarring and the chance of damage is increased. Patients must be properly informed of the benefits and risks of surgery.

Postoperative complications

Vertigo is rare after surgery and when it does occur it is usually mild, short-lived, and can be managed with reassurance. Occasionally, however, there can be severe true rotatory vertigo occurring suddenly within the first few days postoperatively. This may be due to a crack in the stapes footplate leading to intralabyrinthine infection, bleeding, or perilymph leak. There is little evidence that interventions influence outcome in these cases but most surgeons would give high-dose oral steroids and antibiotics. Re-exploration with removal of the prosthesis maybe justified if the vertigo is prolonged.

Alteration in taste is common especially if the prosthesis is touching the chorda tympani. Figures of up to 50% are mentioned in the short term but only around 4% have permanent disturbance.

Patients who get a good hearing result are much more tolerant of taste disturbance than those who get a poor hearing result.

Sensory hearing loss is rare but total sensory hearing loss can occur. Again, most surgeons would give high-dose oral steroids and antibiotics but with little evidence that they affect the outcome.

Facial weakness is fortunately rare. If it occurs it is usually delayed and temporary. Oral steroids for a week are frequently given.

Prosthesis extrusion. While modern prostheses are biocompatible, extrusion will still occur typically if there is tympanic membrane retraction. Long-term extrusion rates of 1–5% are quoted.[12]

Long-term results

While good long-term results are achievable this is heavily influenced by the state of the middle ear.[13] In patients with ongoing poor middle ear ventilation, prosthesis are prone to displacement and/or extrusion over time.[14]

Guidelines

There are currently no guidelines for the performance of ossiculoplasty.

Areas for future development

There is a shortage of high-quality randomized controlled trials comparing different prostheses.

Custom-made three-dimensional printed prostheses based on preoperative CT scanning are being developed.

Robotic reconstruction of the ossicular chain is likely to develop.

Conclusion

Ossiculoplasty can be rewarding and successful; however, results are variable and dependent on the middle ear conditions encountered. It is important for surgeons to maintain their own databases so that patients can be accurately advised about the potential for success.

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22

Tympanoplasty

Matthew P.A. Clark

Introduction

Perforated tympanic membranes (TMs) are a common otological presentation. They are often repaired surgically; myringoplasty is the term for surgical repair of a perforated TM. Tympanoplasty is a less clear term that is often used synonymously with myringoplasty, yet can also include procedures for removal of widespread cholesteatoma and/or ossiculoplasty. There are several classification systems described in the literature for tympanoplasty that are popular exam topics but perhaps less practical to decision-making in the clinic. By definition, tympanoplasty is differentiated from myringoplasty in that it involves addressing some form of middle ear pathology, although this could be a minor as dividing a middle ear adhesion at the time of TM repair. Some would argue that the main difference lies in the procedure coding and subsequent financial remuneration! For the purpose of this chapter, tympanoplasty will refer to the various techniques that are available to repair the perforated TM. Other chapters in the book address ossiculoplasty (see Chapter 21) and surgery for cholesteatoma (see Chapter 19).

Anatomy

The TM is oval, approximately 8 × 10 mm in diameter, and separates the squamous epithelial-lined external ear canal from the mucosal-lined middle ear. It is largely comprised of the pars tensa, a three-layered membrane (outer epithelial, middle fibrous, and inner mucosal), thickened at its circumference to form the annulus. In the attic region the pars flaccida is a weaker structure as a result of a less organized fibrous layer.

Perforations of the TM, or incisions used to access the middle ear space via the TM, may have their location recorded according to the face of a clock (the manubrium of the malleus forms the clock hand from the centre of the TM to the 12 o'clock position).

In its normal state the TM has a concavity towards the middle ear space. It also slopes medially from posterior to anterior aspects. While its blood supply comes in radially, a leash of vessels feed the TM from the 12 o'clock position along the malleus manubrium.

Assessment/investigations

Standard assessment of a perforated TM will involve otoscopy. Many perforations will have presented with a history of recurrent discharge necessitating removal of wax or debris to obtain a clear, unobstructed view. An operating microscope is often employed.

Tympanometry will further aid diagnosis. In the case of a perforation, a flat, type B, tympanogram is expected. This can be differentiated from other causes of a type B result (as with a middle ear effusion) by considering the recorded ear canal volume, which in the case of a perforation will be expected to be greater than normal.

Pure tone audiometry should be performed. This may demonstrate a conductive hearing loss, but not always. Where the ossicular chain is intact it is unusual for the conductive loss to exceed 35 dB regardless of the size or site of the perforation. It is important to record the hearing level before ear surgery in order to be able to make a comparison after surgery.

Perhaps the most important aspect of the assessment is to consider the contralateral ear. This should strongly influence the management of the affected ear as only in exceptional circumstances would one operate upon the better/only hearing ear. Comorbidity and the patient's wishes are also taken into account.

The age of the patient should be considered. Generally, repair of the TM is avoided in young patients who are still prone to otitis media with effusion or recurrent acute otitis media. There is no defined lower age limit, but research from several paediatric ear, nose, and throat centres suggest that repair can be considered from about 8 years of age. One is trying to consider that the child has outgrown the conditions prevalent in a younger age group that might adversely affect the repair, and also has reached an age mature enough to undergo the procedure and required postoperative care. This is a judgement call to be made by the operating surgeon. There is no specific upper age limit.

Emergency treatment

Acute TM perforations may arise secondary to middle ear infection, through foreign objects entering the ear canal, from slaps to the ear, or

from more serious head injuries, skull base fractures, and barotrauma. The majority of these will heal without surgical intervention (up to 95% within a 6-week period). As such, most are initially managed in primary care before being considered for referral to specialist care.

Once identified, the mainstay of initial treatment is to exclude water from the ear canal during an approximate 6-week period and to treat any identified infection (oral and/or topical antibiotics as required). Microsuction or aural toilet to remove excessive blood or debris from the ear canal may be considered. After a 6-week period the TM can be assessed as above and the treatment options below can now be considered.

Surgery

Indications

In established, or chronic, TM perforations, the main aim for repair of the perforated TM is to 'waterproof' the middle ear space. A perforated TM tends not to tolerate water exposure, which can frequently lead to infection symptomatic by way of discharge. This may affect the ability to undertake water sports or recreational activities but may also be an issue with simple bathing. Washing of the ears is part of the practice of Wudu for some Muslims.

Commonly in the more elderly population a perforation may become infected when a hearing aid is worn in the same ear. This is presumably due to the reduced aeration of the ear canal when an aid is in place. Repair may allow for better tolerance of hearing aid usage and its associated benefits and can be justified for this indication.

Surgical repair of TM perforations where the primary aim is for hearing improvement becomes more contentious. It is not possible before an operation to know to what degree the hearing will improve by repairing a perforation. While many conductive losses will be improved, surgery for repair also carries the risk of causing inadvertent sensorineural hearing loss.

Consent

In consenting for myringoplasty, the following risks and complications should be discussed:

- Graft failure/chance of successful grafting—success rates of 90–95% should be expected for simple TM perforations.
- Hearing loss.
- Imbalance/dizziness.
- Tinnitus.
- Taste disturbance.
- Infection.
- Scar (for post-aural/endaural approaches).
- Some surgeons advocate mentioning facial nerve palsy, but it is the author's opinion that this is not necessary for simple TM perforation repair.

Surgical options

There are many surgical techniques for TM perforation repair. Undoubtedly there is no one correct technique and each has its own merits and disadvantages. Technical approaches are often influenced by the size and position of the perforation such that one technique is unlikely to be best for all. The shape and diameter of the ear canal will also influence this decision and at times canalplasty (widening of the external ear canal by removal of bone) may accompany tympanoplasty in order to gain an adequate surgical view of the TM. The techniques described below may also form part of a larger ear operation.

There are three surgical approaches:

1. *Transcanal*: techniques performed down the ear canal involving incisions within the ear canal. When used with a microscope this requires a reasonably wide ear canal and provides a limited view of the TM (often limiting access to posterior perforations only). This is the standard approach for cases using the endoscope, in which a clear view of the entire TM is to be expected.
2. *Endaural*: an incision made in the superior, lateral aspect of the external auditory canal between the tragus and crus helix, to allow wider access via the ear canal (and so more anteriorly placed perforations can be better visualized). Temporalis fascia can be harvested from the same incision.
3. *Postauricular*: an incision made behind the pinna to access the posterior aspect of the ear canal. This is usually positioned a few millimetres posterior to the postauricular sulcus or skin crease. The skin of the posterior ear canal is elevated until the space of the ear canal is entered via ear canal incisions (often made prior to this stage at the start of the procedure when the TM is formally inspected). Temporalis fascia can be harvested from the same incision. Allows visualization of the entire TM in most cases and so can be helpful for approaching anterior perforations.

There are also two technical approaches, those using an operating microscope (for options 1–3 above) and those using an endoscopic approach (for transcanal approaches).[1]

- The operating microscope provides a binocular view down the ear canal. As the optics are positioned outside of the ear, the field of view is narrow. Both hands are free to hold micro-instruments (usually a suction device in the non-dominant hand and micro-middle ear instrument in the dominant hand). Microscopes often have a side-viewing arm for use by a trainee or assistant, and may have a camera attachment for display on a screen, yet each of these two views effectively only gets a one-eyed view that is therefore inferior to that seen by the operating surgeon.
- Endoscopic ear surgery places the optics within the ear canal and so provides a much wider field of view. One hand needs to hold the endoscope leaving only one to handle all other instruments, and the endoscope itself takes up limited space within the canal. A camera attachment to the endoscope displays the surgical view identically to the surgeon and to all others present. As endoscopic ear surgery is performed via a transcanal approach there is no external incision, which can contribute towards improved postoperative comfort, no need for head bandaging after the operation, and potentially an easier discharge from hospital the same day as the procedure (Figure 22.1).

Positioning

The patient is positioned supine, with a degree of head-up tilt (reverse-Trendelenberg position). The head is turned away from the operated side and stabilized using a head ring.

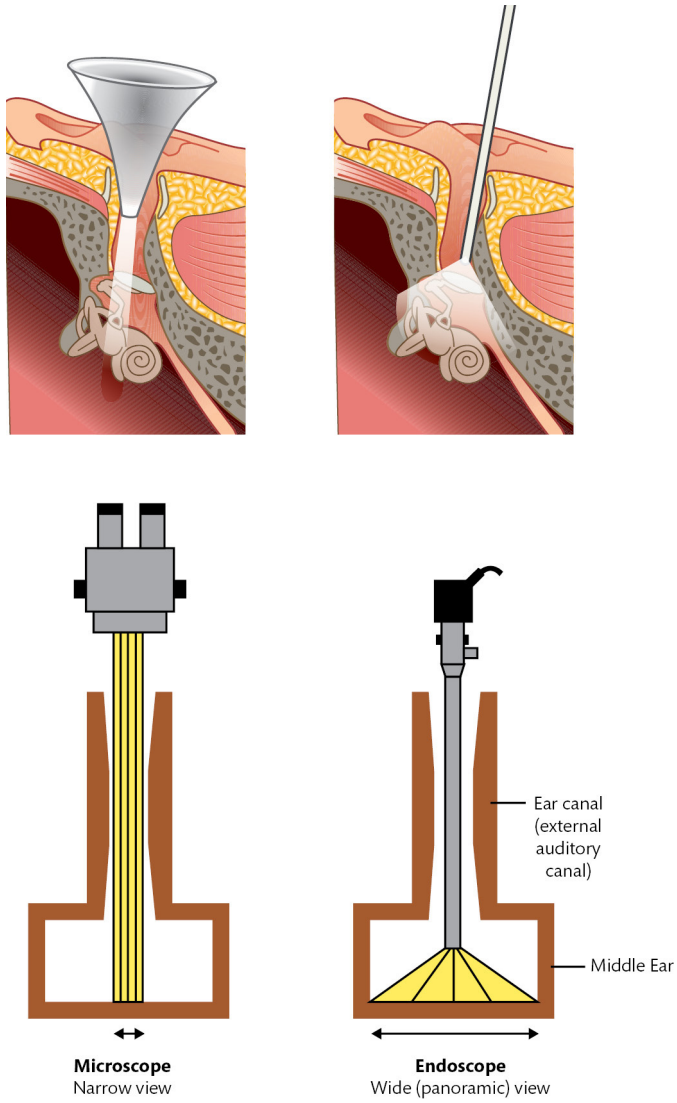

Figure 22.1 Field of view; microscopic versus endoscopic techniques.

Preparation

- Some hair shaving may be required in the post-aural region. In endoscopic ear surgery, external auditory canal hair removal is important to prevent repetitive smearing of the scope lens.
- It is not possible to fully clean or disinfect an ear canal, but preparation with aqueous povidone–iodine, instilled into the ear canal and then washed out with saline, is commonly performed.
- A 'bloodless' field is preferable and this is helped with:
  - hypotensive anaesthesia
  - the use of topical adrenaline, applied with local anaesthetic administration and used topically on cotton wool balls (or similar)
  - reverse-Trendelenberg positioning
  - preoperative consideration of medication that may affect bleeding times.
- Usually a combination of local anaesthetic with adrenaline is used, for example, 2% lidocaine with 1:80,000 adrenaline applied with a dental syringe and 25 G needle. Ear canal infiltration is applied at the cartilage/bony junction subcutaneously, with care to avoid 'blebbing'. Blanching of the skin can be observed upon infiltration. Further infiltration should be used at the site of any post-aural, endaural, or graft harvest incisions that are made.
- Cotton wool balls soaked in topical adrenaline (1:1000) can be useful when raising the tympanomeatal flap, to minimize bleeding but also to help protect the delicate skin flap from being traumatized by the suction or micro-instruments.

Graft choice

- Temporalis fascia: suitable for all size grafts including large ones required for total TM repair; easier to handle when dry.
- Perichondrium: easily accessed from the tragus.
- Fat: suitable for small grafts, available from the ear lobule.
- Composite: cartilage with or without perichondrium grafts, available from tragus, anti-tragus, and concha, for example; offer more structural support.
- Xenograft: e.g. Biodesign (collagen material derived from porcine intestine); costly but avoids the need for autologous harvest.

General considerations

- Before any form of graft is applied, the edges of a perforation should be 'freshened' with a gently curved needle and cupped forceps. In those perforations that do not heal, the inner and outer TM layers fuse at the perforation margin. The aim of freshening the edges is to remove this seal and to stimulate fresh wound healing and therefore skin migration and graft integration. The rim may be perforated multiple times (like the edge of a stamp) and removed with micro-forceps.
- Where the umbo or handle of malleus forms part of the border of the perforation, it can be more difficult for a graft to correctly seat. Dissection of the remaining TM from the malleus may facilitate graft placement although care is required to prevent transmission of movement through the ossicular chain. Disarticulation of the incudostapedial joint prior to such manipulation may reduce the risk of causing sensorineural hearing loss.
- Where a patch of tympanosclerosis forms part of the border of the perforation it may affect graft 'take'. Sometimes such chalky patches are best excised to give the perforation bleeding edges circumferentially, potentially at the expense of increasing the size of the perforation.
- Packing/dressings: there are a large range of materials available to dress the ear canal. One aim of the dressing material is to lie against the grafted material to support its positioning during the healing/integration stage. Some support may be provided in this way by surface tension. Within the ear canal where the posterior canal wall skin has been raised (most significant for postauricular or endaural approaches), the dressing also aims to prevent haematoma formation that could disrupt the postoperative result. Iodoform-containing dressings (e.g. bismuth iodoform paraffin paste) may lead to allergic hypersensitivity reactions.
- Postoperative instructions should consider:
  - water exclusion (usually for ~8 weeks)
  - avoidance of using a hearing aid/earphones
  - when an individual is OK to fly (see 'Controversies')
  - use of topical antibiotic drops.

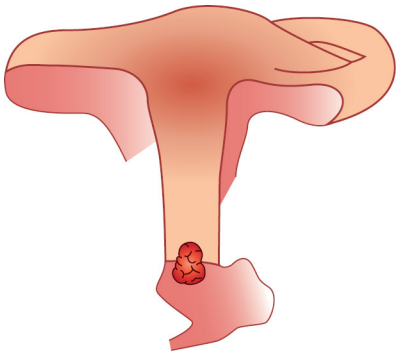

Figure 22.2 Fat-plug myringoplasty *in situ*.

Procedures that do not raise a tympanomeatal flap

Some perforations may be repaired with approaches that avoid raising a tympanomeatal flap, with either a microscope or endoscope. These techniques will have reduced risks of hearing loss and taste disturbance and will generally be more amenable to being performed under local anaesthesia. However, they are most suitable for smaller perforations and there may be a reduced graft success rate despite the lower risks of the procedure.

Fat plug

- Fat can be harvested from the ear lobule.
- Suitable for perforations up to 1–2 mm in diameter.
- The harvested fat is oversized compared to the perforation, such that when in place it forms a dumbbell shape with the narrow waist section passing through the perforation (Figure 22.2).

Push-though

- Similar to a fat-plug myringoplasty but using fascia/perichondrium/xenograft.
- May be supported in place by dissolvable dressing applied through perforation to the middle ear promontory (e.g. Spongostan or Gelfoam, absorbable gelatin sponges).

Butterfly graft

- Uses composite cartilage/perichondrium graft.
- Suitable for perforations approximately 2–4 mm in diameter.
- A disc of cartilage is taken and its margin scored, to fit (with overlap) into the perforation rather akin to a grommet (Figure 22.3).

Overlay (lateral) grafting

- Requires removal/retraction of the medial ear canal skin and TM remnant (and so has the risk of epithelial pearls if this is performed incompletely).
- Usually necessitates drilling of the deep bony canal for satisfactory graft placement.
- Graft is placed medial to the malleus to minimize risk of graft lateralization and blunting.
- This technique may be used for large perforations.

Procedures that do raise a tympanomeatal flap

In raising a tympanomeatal flap, the middle ear space can be accessed. Ear canal incision siting will vary, but typically the posterior aspect of the ear canal skin is incised and elevated to allow the back half of the TM to be folded forward (like the page of a book with the handle of the malleus acting as the book's spine). Radial incisions run from approximately the 12 and 6 o'clock positions from deep to superficial. These are joined with a circumferential incision set a few millimetres back from the annulus. The further back from the annulus this is placed, the greater the amount of dissection required to elevate the flap. If the incision is placed too close to the annulus, the flap may

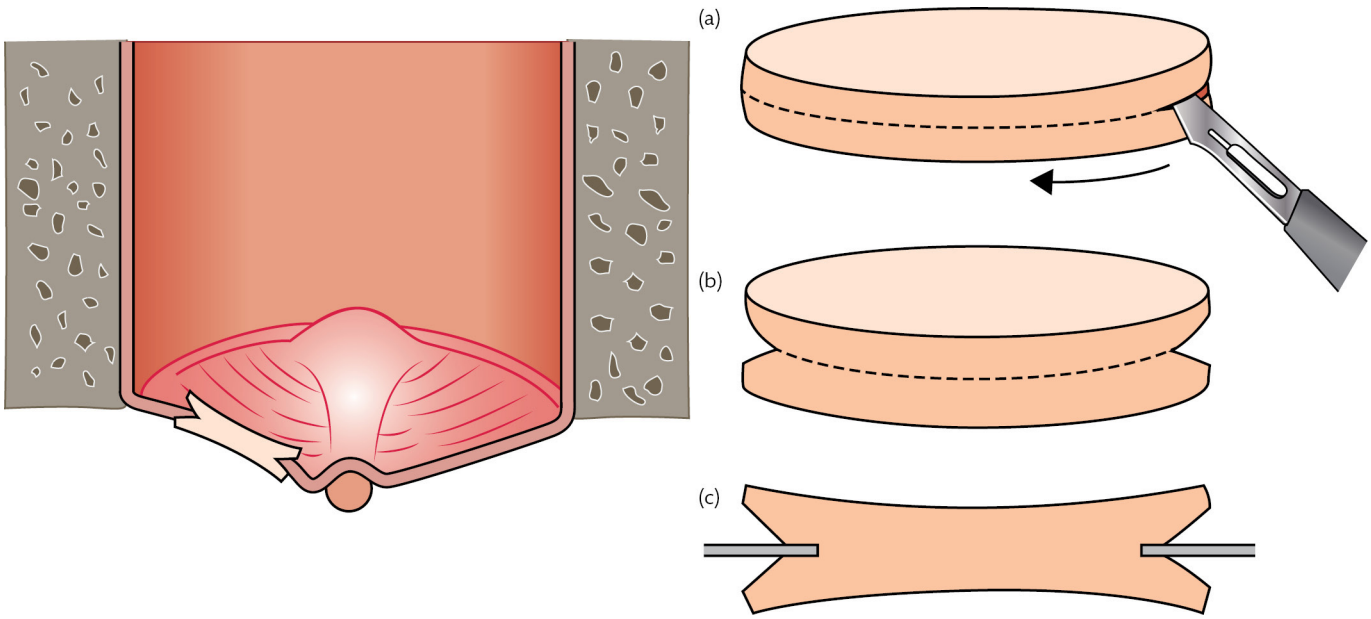

Figure 22.3 Cartilage butterfly graft.

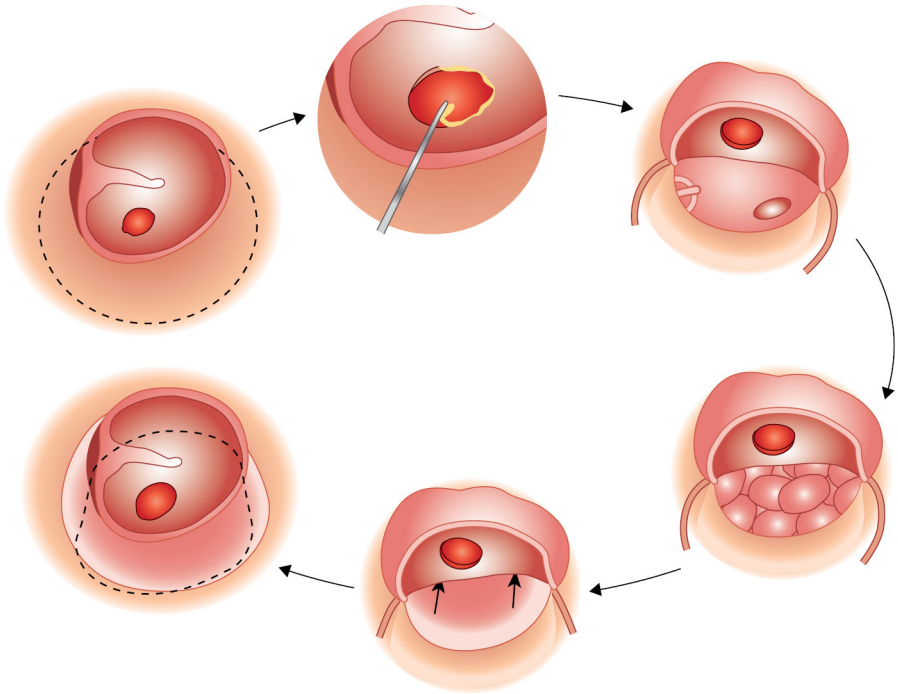

Figure 22.4 Temporalis fascia graft underlay.

shrink as it dries under the operating lights and fail to lie back onto the posterior canal wall (also an issue if there has been any posterior canal bone removal next to the annulus). Thus a happy medium between the two is sought, usually about 3–4 mm lateral to the annulus. While the exact positioning of incisions can vary depending upon the site of the perforation, wide elevation of the anterior annulus is avoided to prevent blunting of the tympanomeatal angle (which may affect the vibratory properties of the TM).

Underlay

Probably the most commonly performed repair of a TM perforation is using the temporalis fascia underlay technique. This may be achieved both with the microscope or endoscope via transcanal, endaural, or post-aural approaches (Figure 22.4).

- The TM flap is elevated and the middle ear space accessed.
- Graft is positioned underneath the perforation and is held in place largely by surface tension.
- Absorbable sponge (e.g. Spongostan or similar) may be used to support the graft on both its medial and lateral aspects.
- In large subtotal or total perforations there is a lack of remaining TM from which the graft may take support. In such cases the graft may be fashioned in a variety of ways to help hold it in place and prevent migration of the graft in the healing phase (and therefore graft failure) (Figure 22.5):

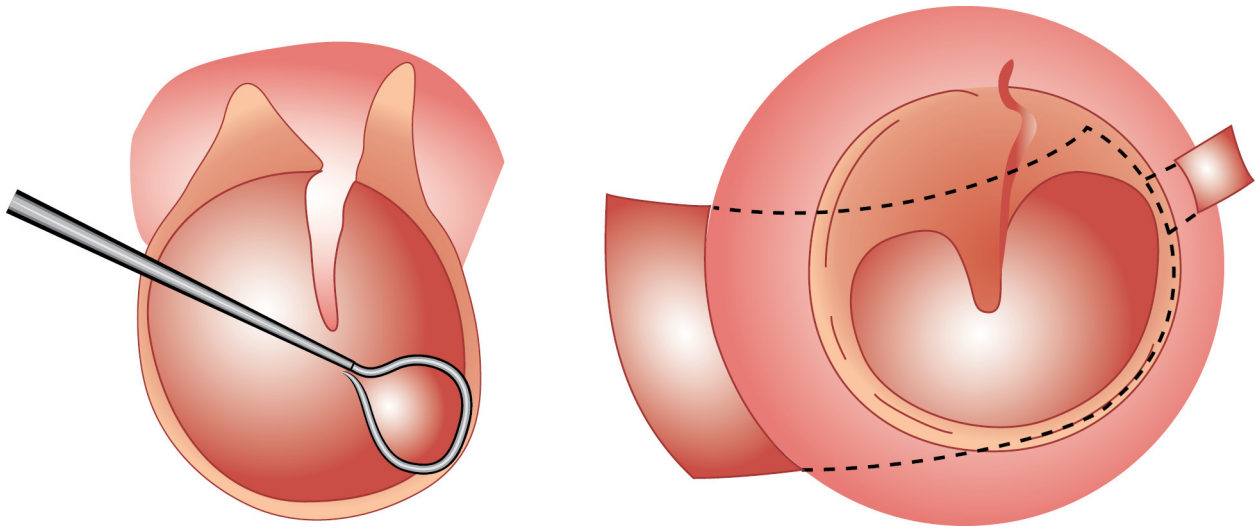

Figure 22.5 'Bunny ears' and anterior tab grafts.

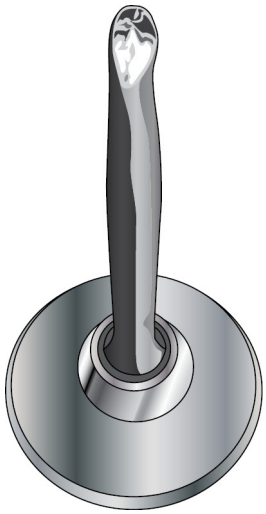

Figure 22.6 Formaldehyde-fixed fasciaform graft mould.

- 'Kerr flap': an anteriorly based flap extension that tunnels under the anterior annulus to help hold the graft in place at this location.
- 'Bunny ears': superiorly positioned flaps that wrap around the neck of the malleus to help support the graft.

Formaldehyde-fixed fascia

Temporalis fascia may be treated with formaldehyde. This process leads to cross-linking of the collagen fibres to increase the tensile strength and memory such that the graft retains its shape after rehydration or exposure to blood. The fascial graft is draped over a metal mould, resulting in a graft the same shape as a TM with 'memory' once treated (Figure 22.6). The technique lends itself to repair of total or subtotal perforations.[2]

Cartilage reinforced

The use of cartilage (with or without attached perichondrium) as a TM graft offers advantages of providing greater structural support. This may be particularly helpful in an ear prone to TM retraction, for example.

Many different modifications or shaping of the cartilage have been described, including palisading or the 'Mercedes-Benz' logo design, for example (Figure 22.7). Where the cartilage is taken with attached perichondrium, provided the latter is left intact the cartilage can be incised to create 'islands'. This will allow for greater TM mobility and potentially better hearing results, or for a more natural TM shape to be created.

Controversies

- The use of various growth factors has been the subject of several research papers relating to potential beneficial effects on TM repair (for both acute and chronic situations). The benefit is currently unclear and the technique is recommended only in research centres.
- Surgery upon an ear with active discharge remains a point of contention. Where possible, it is sensible to treat an actively infected ear prior to surgery, but as most TM perforation repairs are indicated by recurrent discharge, they are often inflamed or infected at the time of repair.
- Flying after ear surgery: there is no clear consensus regarding when an individual can fly after tympanoplasty. The concern is that the changes in middle ear pressure may cause a graft to move in position before being integrated. However, in the immediate period after surgery the middle ear space is likely to be full of blood/fluid/absorbable dressing such that there may be little or no pressure chamber requiring equalization. Studies on patients flying home the day after surgery have failed to show an increased risk of failure.[3] Despite this, many surgeons will take a cautious approach and advise 4–6 weeks of avoidance if possible.
- Drops: the use of topical antibiotic drops in the perforated ear raises frequent discussion. Some antibiotic drops are aminoglycoside- or polymyxin-based, raising a concern of potential ototoxicity and sensorineural hearing loss. However, in the presence of active infection, for a limited duration (up to 2 weeks) these drops can reasonably be used. Ciprofloxacin drops have become commonplace and do not have this risk.

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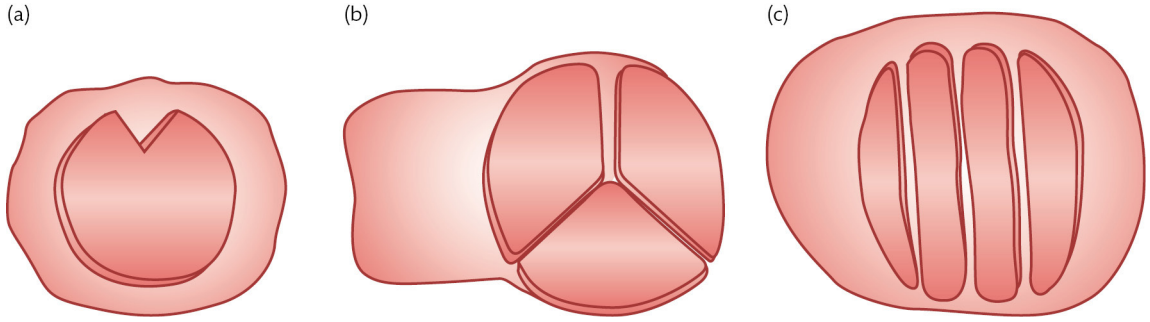

Figure 22.7 Cartilage grafts.

23

Temporal bone trauma

Nicola Quaranta and Vito Pontillo

Introduction

Thirty to seventy per cent of all head trauma cases involves the temporal bone, but only 14–22% of them produce a veritable temporal bone fracture.

Blunt head injuries (motor vehicle accidents 31–47%, falls 17–33%, aggressions 10–16%) are the main causes of trauma, while gunshot wounds are rare (3–6%).

Temporal bone fractures can occur at any age, with a peak of incidence during the second, third and fourth decades of life, and affect men more frequently (3:1).

Anatomy

Temporal bones are paired structures that contribute to the base and lateral walls of the skull. Each temporal bone is composed of five portions: squamous, tympanic, mastoid, styloid, and petrous. The petrous bone is a pyramid-shaped component of the middle and posterior skull base that encloses or has direct contact with many crucial sensorial, vascular, and nervous structures (**Figure 23.1**). Hence the clinical relevance of temporal bone trauma.

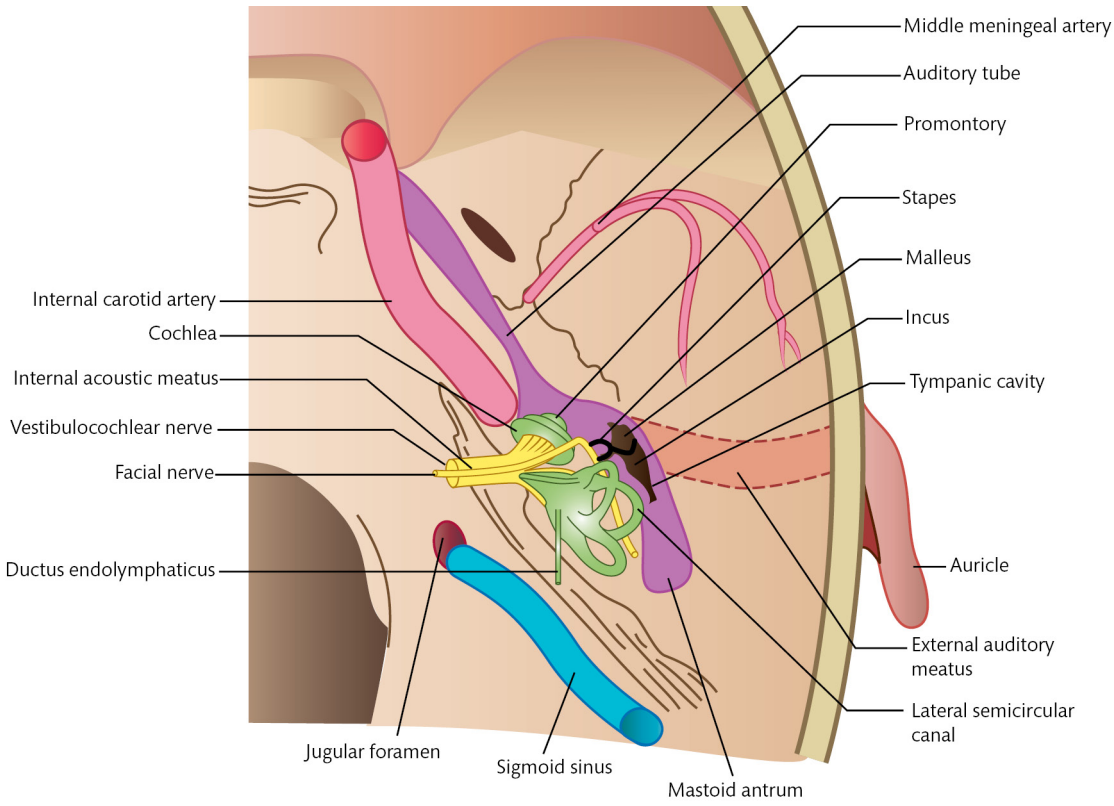

Figure 23.1 Illustration of a superior view of temporal bone with neural and vascular elements.

Table 23.1 Classification of temporal bone fractures

	Longitudinal	Transverse
Frequency	70–90%	10–30%
Collision	Horizontal temporo-parietal blow	Posterior occipital/vertex blow
Fractural line	Vertical squama → EAC → tympanic cavity → turns anteriorly at the geniculate ganglion → parallel to long axis → foramen spinosum/lacerum/Eustachian tube	Foramen magnum → foramen spinosum across IAC/cochlea (medial subtype) or posterior labyrinth (lateral subtype)
Tympanic membrane	Perforation, bleeding	Intact, haemotympanum
Hearing loss	Conductive	Sensorineural
Facial paralysis	Rare (7–10%) and mostly delayed Geniculate ganglion (premeatal or suprameatal subtypes) and 3rd segment (retromeatal subtype)	Frequent (50%) and mostly immediate 1st (medial subtype) and 2nd segment (lateral subtype)
	Possible	Frequent
CSF leak	Tegmen	Posteromedial surface of petrous bone (lateral subtype) or bottom of IAC (medial subtype)
	Otoliquorrhea	Unmanifested (tympanic membrane intact) or rhinoliquorrhea

Physiopathology and classification

The temporal bones are wedged in the thick bone of the skull base and therefore require a great force to break them, estimated at 1875 pounds (850 kilograms). Nevertheless, a number of post-traumatic lesions are still possible even in the absence of a temporal bone fracture and are not to be ignored.

Temporal bone fractures

When the force of impact is strong enough to break the bone, the fracture takes the path of least resistance, including skull base foramina, pneumatized cells, and cranial sutures. This implies that in most cases the fracture line follows some preformed anatomic pathways. On the basis of the relationship between these pathways and the axis of the petrous pyramid, temporal bone fractures are traditionally classified as reported in Table 23.1 and Figures 23.2 and 23.3.

Longitudinal fractures are typically parallelly oriented to the long axis of the petrous bone and, according to the entry point on the squamous bone, are divided into premeatal, suprameatal, and retromeatal fractures (Figure 23.2(I)).

Transverse fractures are perpendicularly oriented to the long axis of the petrous ridge and normally require a higher energy of impact. For this reason, intracranial and vascular complications are more frequent.

The fracture line may cross the internal auditory canal (IAC) and cochlea (medial subtype) or traverse the posterior labyrinth, mostly at the level of the posterior semicircular canal or vestibular aqueduct (lateral subtype) (Figure 23.2(II)).

Although this classification is quite simple to understand, it has been demonstrated not to be able to categorize the frequent mixed patterns and to precisely predict the functional sequelae and complications. Hence, the rationale of a new and simpler classification that divides temporal bone fractures based on the presence of absence of otic capsule involvement:

- *Otic capsule-disrupting fractures* (2.5–5.8%) always result in sensorineural hearing loss (HL), quite often in a facial nerve paralysis (30–50%), and present a greater risk of cerebrospinal fluid (CSF) fistula (twofold to fourfold) and of intracranial complications.
- *Otic capsule-sparing fractures* are often associated with conductive HL, while facial nerve paralysis (6%) and CSF leak are infrequent.

Associated lesions

External ear injuries classically result from direct impact on the tympanic bone or from an indirect posteriorly directed collision on the

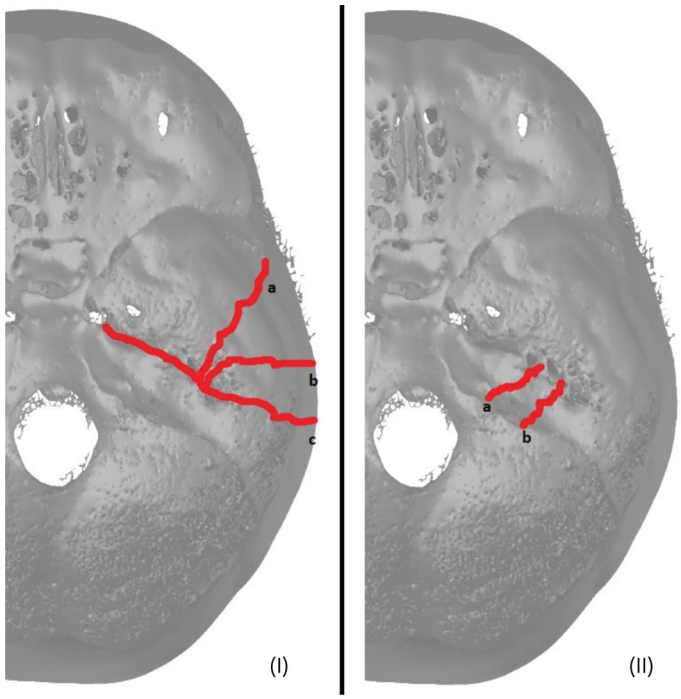

Figure 23.2 Superior view of a three-dimensional model with a graphical representation of temporal bone fractures based on the main orientation of the fracture line. (I) Longitudinal fracture with (a) premeatal, (b) suprameatal, and (c) retromeatal entry points. (II) Transverse fracture, (a) medial and (b) lateral subtypes.

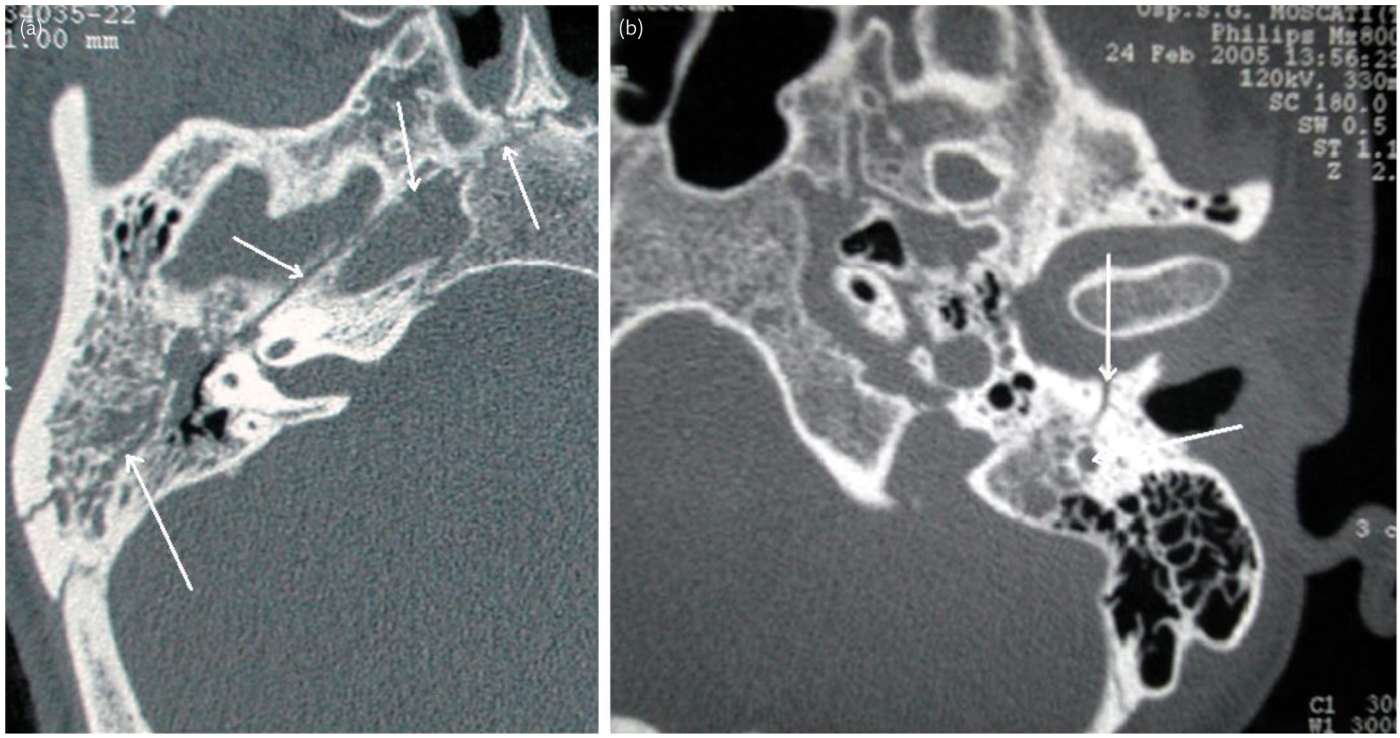

Figure 23.3 Axial computed tomography scan images of (a) left longitudinal temporal bone fracture passing laterally and ahead of facial nerve, otic capsule, and carotid artery and reaching the foramen lacerum and the sphenoid sinus; (b) right transverse temporal bone fracture involving the mastoid portion of the facial nerve.

mandibular condyle. They are commonly associated with an anterior wall displacement, with subsequent external auditory canal (EAC) stenosis and risk of cholesteatoma formation.

Temporal bone trauma, with or without fracture, can be associated with *ossicular chain lesions*. This is the product of a relative movement between stapes and malleus, which are integral with the inner and external ear, respectively. It may result in an ossicular luxation, more often of the incudostapedial (82%) and incudomalleolar (52%) joints, or in an ossicular fracture, particularly of the long process of the incus or the stapes footstep/crus. It results in significant conductive HL, of at least 30 dB.

Inner ear lesions occur in almost 90% of all transverse fractures, as a result of several mechanisms:

- Immediate vestibular deafferentation with subsequent acute major symptomatology.
- Fracture of the bony capsule, with consequent perilymphatic fistula and possible evolution to the so-called ex-vacuo endolymphatic hydrops.
- Perilymphatic fistulas often affect oval and round windows and can occur immediately or belatedly after the traumatic event, because of the incomplete healing process in the endochondral layer of the otic capsule.
- Rupture of the membranous labyrinth leads to homeostasis and electrophysiological changes, with subsequent toxic insult to inner ear neural cells.
- Evolution to labyrinthine fibrosis and labyrinthitis ossificans.
- Rupture of the vestibular aqueduct, with subsequent cicatricial stenosis and endolymphatic hydrops.
- Immune-mediated mechanisms are thought to be responsible for contralateral delayed endolymphatic hydrops, which determines fluctuating HL with episodic vertigo after a prolonged period of time from the trauma.
- 'Labyrinthine concussion' is determined by intralabyrinthine bleeding and microcirculatory disorders causing neural cell toxicity. Sudden deceleration can cause the rupture of the utricular macula with subsequent massive release of otoliths and benign paroxysmal positional vertigo.
- In case of motor vehicle accidents, air-bag explosion or noise caused by the collision can result in inner ear barotrauma.

Almost 25% of all temporal bone fractures (50% of transverse fractures and 10% of longitudinal ones) are associated with *facial nerve paralysis*. It can be complete or partial, and its onset can be immediate (27%) or delayed by 1–16 days (73%) and caused by several mechanisms: bone fragments may cut, compress, or stretch the nerve, or intra-/extra-nervous haematoma or oedema may press on it. All the segments can be involved, regarding the type of fracture (Table 23.1).

A trauma of major amplitude could eventually *damage other peripheral nerves*, such as the V, VI, VIII, IX, X, and XI cranial nerves.

Post-traumatic *internal carotid artery lesions* can determine profuse ear bleeding, epistaxis, or cerebrovascular accidents. The internal carotid artery is quite exposed at the level of the foramen lacerum, where relevant torsional or transverse forces may cause both its laceration or dissection, and potentially progress to thrombosis, aneurysms, or carotid–cavernous fistulas.

Laceration of the middle meningeal artery can cause epidural haematoma.

Sigmoid sinus and *jugular bulb* can develop post-traumatic *thrombosis*.

Fast and violent translation and torsion movements of the skull content can *damage the brain* by both direct and backlash mechanisms. Epidural, subdural, or intracerebral haematomas and cerebral oedema may occur too. Cases of delayed evolution to neurodegenerative encephalopathy have been also described.

When the fracture line involves the skull base, its cutting bony edges may dislocate and lacerate the dura, thus creating a '*osteomeningeal breach*' (17% of temporal bone fractures). The breach is classically located at the tegmen in longitudinal fractures and at the bottom of the IAC or at the posteromedial surface of the petrous bone in transverse fractures.

While tegmen breaches usually tend to the spontaneous closure, posteromedial and IAC breaches do not have the same tendency, because of the presence of the subarachnoid cistern. Moreover, in translabyrinthine breaches, the reparative inability of the endochondral bone of the otic capsule leads to an incomplete and fragile healing barrier. This may determine a delayed onset or recurrence of the leak, as a result of an effort (coughing or sneezing). For the same reasons, translabyrinthine fractures carry a higher risk of delayed *meningitis*.

Bone breaches can predispose to the development of *meningoceles* or *meningoencephaloceles* that are classically discovered several years after the trauma, during the assessment of a conductive HL, otogenic meningitis, or delayed CSF leak.

Clinical aspects

From a practical point of view, it is possible to describe three phases in most temporal bone traumas:

- A *primary* or *acute stage*, in which the haemodynamic and neurological status are of primary importance and neurosurgical interventions may be necessary. It is usually in an emergency or intensive care unit that the ear, nose, and throat (ENT) specialist is called to evaluate a comatose patient.
- A *secondary* or *subacute stage*, that starts at coma arousal, when all haemodynamic and neurovascular issues have been stabilized and a second ENT evaluation is requested to assess eventual pertinent complications.
- A *tertiary stage* or *stage of sequelae*, that normally follows months or years after the trauma, in which the otologist is called to assess some conditions derived from the 'chronicization' or re-exacerbation of some old issues.

Assessment

History collection should focus on the dynamic of injury and immediate or delayed complaints. A fundamental detail is the immediate or delayed onset of eventual facial paralysis, since this can dramatically affect the prognosis and the treatment strategy. In acute and subacute phases, history collection may be complicated by eventual coma or amnesia.

Eventual ear/head pain, otorrhoea, otorrhagia, unilateral or bilateral HL, tinnitus, fullness, and vertigo/dizziness should be registered. In case of CSF fistula, patients may report continuous ear or nose fluid drainage, salty taste, and dull, permanent, and bilateral headaches. Episodes of otogenic meningitis have been reported even years after the traumatic event and should be properly reported in the third phase. Orbital pain, progressive visual loss, chemosis, and pulsatile proptosis can be expressions of a carotid–cavernous fistula.

A complete neurotological *examination* should include the following assessments:

- Physical examination is crucial, especially in the early phases, to detect direct or indirect signs of temporal trauma: scalp wounds, skull compound fractures, mastoid ecchymosis (Battle's sign), ear bleeding. If a fracture of the anterior cranial fossa is also present, bilateral periorbital ecchymosis ('raccoon eyes' or 'panda eyes' sign) and epistaxis may be evident.
- Ear examination should be as aseptic as possible and limited to simple otoscopy at the acute stage: blood and cerumen may obstruct the vision, but they should never be debrided by irrigation. Proper otomicroscopy should be postponed after stabilization and transfer. EAC fractures, haemotympanum, tympanic membrane perforations, and brain herniations should be pointed out when present. Clear, watery otorrhoea or, in the presence of an intact tympanic membrane, middle ear effusion may represent a sign of CSF leak. It often goes unnoticed in early phases, masked by bleeding, but when the latter starts to wane the classical 'halo sign' should be noticed.
- Rhinoscopy can show continuous clear, watery drainage that may evoke a dural fistula.
- Facial nerve function assessment and grading should be performed as soon as possible after the trauma and repeated at any stage, possibly keeping the same grading system. The *House–Brackmann classification* (**Table 23.2**) is the most frequently used system, although the *Sunnybrook grading system* (**Table 23.3**) allows a more precise standardization of synkinesis and other sequelae in late phases. **Box 23.1** summarizes the main clinical features to look for at first assessment in the emergency room. Acute trauma patients may present with important facial oedema that may mask eventual asymmetries or instead limit facial expression and overestimate eventual dysfunctions. In a sedated or comatose patient, the only way to assess facial nerve function may be through the 'Marie–Foix manoeuvre'. In post-acute stages it is possible to ask the awake patient to make some moves (i.e. raise their eyebrows, close their eyes, smile, grimace).
- Neurotological examination is usually completed by assessing other cranial nerve function and ocular motricity. In the case of associated anterior skull base or orbit fractures, there is the possibility of optic nerve injury. In a comatose patient, it can be suspected by the presence of unilateral mydriasis with no photopupillary reflex.
- Epi-aortic trunk and orbital auscultation can be useful in the third phase to detect pathological murmurs underlying a carotid aneurysm or a carotid–venous fistula, respectively.

Table 23.2 House–Brackmann classification for facial nerve function

Grade	Description	Characteristics			Estimated function (%)
		Gross	At Rest	Motion	
I	Normal	Normal	Normal	Normal	100
II	Mild dysfunction	Slight weakness noticeable on close inspection, may have very slight synkinesis	Normal symmetry and tone	Forehead: moderate to good function; eye: complete closure with minimum effort; mouth: slight asymmetry	80
III	Moderate dysfunction	Obvious but not disfiguring difference between two sides; noticeable but not severe synkinesis, contracture, and/or hemifacial spasm	Normal symmetry and tone	Forehead: slight to moderate movement; eye: complete closure with effort; mouth: slightly weak with maximum effort	60
IV	Moderately severe dysfunction	Obvious weakness and/or disfiguring asymmetry	Normal symmetry and tone	Forehead: none; eye: incomplete closure; mouth: asymmetric with maximum effort	40
V	Severe dysfunction	Only barely perceptible motion	Asymmetry	Forehead: none; eye: incomplete closure; mouth: slight movement	20
VI	Total paralysis	No movement	Asymmetry	No movement	0

Investigations

Functional tests

Audiological assessment of the acute phase, considering the limited possibility of displacement of traumatic patients, should be limited to bedside hearing assessment (Weber and Rinne tests) and a first individuation and characterization of nystagmus, when not inhibited by coma, neurological conditions, or medical agents.

Tonal, vocal, and impedance audiometry are usually postponed to secondary and tertiary stages, which permit the characterization

Table 23.3 Sunnybrook facial grading system

Resting Symmetry

Compared to normal side

Eye (choose one only)
- normal ☐ 0
- narrow ☐ 1
- wide ☐ 1
- eyelid surgery ☐ 1

Cheek (naso-labial fold)
- normal ☐ 0
- absent ☐ 2
- less pronounced ☐ 1
- more pronounced ☐ 1

Mouth
- normal ☐ 0
- corner drooped ☐ 1
- corner pulled up/out ☐ 1

Total 0

Resting Symmetry score

Total X 5 0

Patient's Name

Diagnosis

8/16/2009
Date

Symmetry of Voluntary Movement

Degree of muscle EXCURSION compared to normal side

Standard Expressions	Unable to initiate movement	Initiates slight movement	Initiates movement with mild excursion	Movement almost complete	Movement complete	
Brow lift (FRO)	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	0
Geltle eye closure (OCS)	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	0
Open mouth Smile (SYG/RIS)	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	0
Snarl (LLA/LLS)	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	0
Lip Pucker (OOS/OOI)	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	0

Gross Asymmetry	Severe Asymmetry	Moderate Asymmetry	Mild Asymmetry	Normal Asymmetry	Total
					0

Voluntary movement score: Total X 4 = 0

Synkinesis

Rate the degree of INVOLUNTARY MUSCLE CONTRACTION associated with each expression

Standard Expressions	NONE: no synkinesis or mass movement	MILD: slight synkinesis of one or more muscles	MODERATE: obvious synkinesis of one or more muscles	SEVERE: disfiguring synkinesis/ gross mass movement of several muscles	
Brow lift (FRO)	☐ 0	☐ 1	☐ 2	☐ 3	0
Geltle eye closure (OCS)	☐ 0	☐ 1	☐ 2	☐ 3	0
Open mouth Smile (SYG/RIS)	☐ 0	☐ 1	☐ 2	☐ 3	0
Snarl (LLA/LLS)	☐ 0	☐ 1	☐ 2	☐ 3	0
Lip Pucker (OOS/OOI)	☐ 0	☐ 1	☐ 2	☐ 3	0

Synkinesis score: Total 0

Vol mov't score 0 - Resting symm score 0 - Synk score 0 = Composite Score: 0

Box 23.1 Signs to look for—tips and pearls

- *Battle's sign*: indicator of fracture of the middle cranial fossa skull base, it consists of arch-shaped bruising of the retroauricular sulcus and over the mastoid process as a result of bleeding of the posterior auricular artery. It appears at least 24 hours after the traumatic event.
- *Raccoon eyes (or panda eyes) sign*: unilateral or bilateral periorbital ecchymosis is a sign of anterior basal skull fracture. It may also be a sign of disseminated neuroblastoma or of amyloidosis, or temporarily occur after rhinoplasty.
- *Halo (or double ring) sign*: a distinctive pattern of pillow chromatography in patients affected by oto-rhino-liquorrhea. Blood tends to separate from CSF coalescing into the centre, while leaving an outer and clearer ring of CSF. Mixtures of blood with tears or rhinorrhoea produce similar results.
- *Marie–Foix manoeuvre*: may be useful to assess facial nerve function in comatose patients. By bilaterally and vigorously compressing the retromandibular notch, the clinician explores the eventual elicitation of a reflexive facial spasm and its symmetry/asymmetry.

of hearing impairment. Auditory brainstem responses are useful for assessing permanent sensorineural HLs, especially in candidates for cochlear implantation.

Vestibular assessment in trauma patients should be undertaken only after an eventual cranial or cervical spine injury has been already ruled out. Vestibular bedside examination is sufficient in most cases for the characterization of a post-traumatic vertigo. Further examinations (videonystagmography, caloric reflex test, and vestibular evoked myogenic potentials) may be required. Dix–Hallpike and Pagnini–McClure manoeuvres may reveal post-traumatic benign paroxysmal positional vertigo, while Hennebert and Tullio phenomenon tests may unmask a perilymphatic fistula.

Electrogustometry, stapedial reflexometry, and the Schirmer test are of little or no value in facial nerve mapping, in contrast to the better results obtained by the recent computed tomography (CT) techniques. Electrodiagnostic techniques such as electromyography and electroneurography have a diagnostic and prognostic significance. They should ideally be performed 2–3 days after the injury, in any case after coma arousal and within 2–3 weeks, and periodically repeated to monitor the evolution. Electromyography has a prognostic significance in the third phase: failure to detect electromyographic signs of reinnervation, spasms, or synkinesis after 1 year indicates the extreme unlikeness of spontaneous recovery in the future.

See also Chapter 10 on facial nerve paralysis.

Laboratory tests

The suspicion of CSF leak can be explored by the means of several diagnostic tests that are carried out on ear or nose fluid samples. Glucose test strips represent a rapid screening that takes advantage of the high glucose content of CSF. However, more specific tests such as beta-2-transferrin and beta-trace protein dosage should be preferred.

Imaging

At the time of the first ENT consultation, almost all patients with severe head trauma usually have already undergone a first cranial CT imaging to assess for intracranial haemorrhage or brain damage. A high-resolution computed tomography (HRCT) scan should be able at least to detect the fracture pathway as well as the indirect signs of complications, such as bone dehiscence, pneumocephalus, pneumolabyrinth, and pneumocochlea.

An additional temporal bone HRCT scan may be indicated in the second phase, in order to permit the exact definition of eventual facial nerve lesions and osteo-dural breaches.

Magnetic resonance imaging (MRI) can help in the assessment of cranial nerve injuries, CSF leak, or labyrinthine lesions.

Angiographic sequences (angiography, CT angiography, or magnetic resonance angiography) may be indicated in case of occurrence of neurological symptoms that are non-consistent with CT findings.

Magnetic resonance, CT, or radionuclide cisternography are useful in localizing the CSF leaking source (**Figure 23.4**).

When there is a significant time delay between the initial CT scan and tertiary phase consultation (>2 years), repeat imaging studies may be convenient. Both HRCT scanning and MRI are indicated to characterize permanent HL, neural lesions, or intratympanic masses and to guide the eventual surgical planning in case of tympanoplasty or cochlear implantation.

Table 23.4 summarizes CT and MRI findings in temporal bone fractures.

Treatment

Acute management

Impellent life-saving aspects such as airway security and haemodynamic and neurological stabilization should be prioritized in the primary phase. All patients with head trauma should be hospitalized in an intensive care regimen and strictly monitored for at least 48 hours, in order to promptly detect the eventual triphasic evolution of epidural haematoma.

The temporal bone is a non-weight-bearing bone and its fractures do not result in significant aesthetical structural deficits. Therefore, no reduction and fixation interventions are requested.

Indications for urgent ENT intervention are very limited in this phase:

- Brain herniation into the middle ear, EAC, or mastoid requires prompt immediate surgical correction, by both neurosurgical and neurotology teams.
- Ear bleeding should be initially controlled by EAC packing using rubber-coated sponge tampons (such as Merocel). If packing cannot control a massive haemorrhage, the possibility of internal carotid artery laceration should be considered. It is ideally approached by interventional radiologists, but a surgical ligation may be attempted in case of inaccessibility to interventional radiology units.
- Non-life-threatening but sequelae-conditioning situations to take care of in this phase could be auricular lacerations or haematomas. Lacerations should be sutured, while haematomas should be drained; pressure dressings should be sutured face-to-face in order to close the dead space and prevent blood recollection. When untreated, auricular haematomas may result in a non-aesthetical auricular chondropathy, the so-called cauliflower ear.

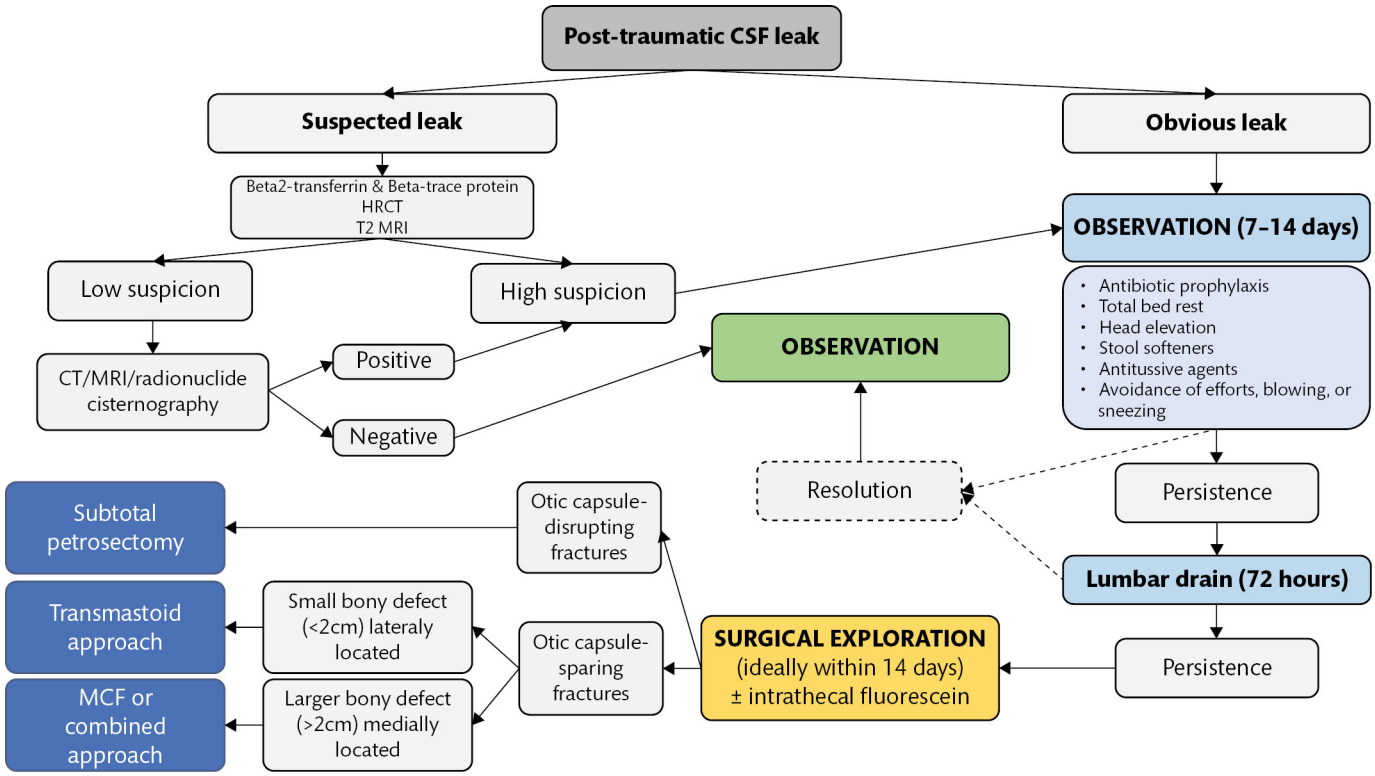

Figure 23.4 Management algorithm for post-traumatic CSF leak.

Table 23.4 Imaging findings in temporal bone trauma

Lesion	HRCT scan	MRI
Bone fractures	Bone discontinuity	
External ear lesions	Fracture/displacement/stenosis	
Ossicular chain lesions	Fractures and/or luxations	
Intralabyrinthine bleeding		Hyper-T1
Labyrinthine fibrosis	Hyperdensity	HR-T2: narrowing and/or loss of fluid signal
Labyrinthine ossificans		HR-T2: narrowing and/or loss of fluid signal
Perilymphatic fistula	Bone defect + pneumolabyrinth (oval/round windows)	HR-T2: fluid leak
Endolymphatic hydrops		Delayed FLAIR: dilated endolymphatic spaces
Osteo-dural breach	Bone defect + pneumocephalus	HR-T2: fluid leak
Meningocele/meningoencephalocele	Bone defect + soft tissue herniation	Hypo-T1, hyper-T1
Post-traumatic cholesteatoma	Bone erosion + soft tissue	Hypo-T1, hyper-T1, diffusion+
Facial nerve injury	Facial canal irregularities + bony fragments	Perineural bleeding (hyper-T1) or oedema (hyper-T1)

HR, high resolution.

Any other ENT intervention should be postponed until proper investigations have been performed and the patient's general status becomes stable. Meanwhile, medical treatment assumes primary importance at this stage.

Medical treatment

Data about the use of intravenous corticosteroids in the early phase are limited. Nevertheless, considering the low costs and limited risks of complications, we believe that a short course of steroids should be suggested, unless medically contraindicated, for both sensorineural HL and facial nerve injury.

Antibiotics, in addition to tetanus toxoid, are compulsory in case of contaminated open fractures with penetrating brain injury. In contrast, they are not routinely prescribed in closed skull base fractures, unless associated with CSF leakage.

All patients with penetrating brain injury, skull base fractures, or CSF leaks should be given pneumococcal vaccination if not previously administered: a single dose of PCV13 followed by

PPSV23 at least 8 weeks later is normally recommended. Anti-meningococcal and anti-*Haemophilus influenzae* vaccinations are not recommended.

A 7-day course of ototopical antibiotics (ofloxacin) is routinely prescribed in case of tympanic membrane perforation. Together with their antibiotic properties, ear drops may promote the cleansing of the EAC from crusting and debris, making future examination clearer.

Management of complications and sequelae

Facial nerve injury

Controversy still exists regarding the management of facial paralysis: data on surgical indications, times, and techniques and on the use of systemic steroids are limited.

The indications for facial nerve exploration and/or decompression are still debated but can be summarized in Figure 23.5. Complete and immediate paralysis require *urgent* surgical exploration in case of radiological evidence of neural compression or electroneurographical signs of important (>95%) degeneration. In all other cases a 'watchful waiting' attitude is chosen: a 2-week course of systemic steroids (1–2 mg/kg) and proper eye protection (daytime artificial tears, night-time ocular bandage, and vitamin A pomade) are normally introduced. Facial kinesitherapy may be useful too.

Modern literature agrees that facial nerve decompression should be performed as soon as possible to minimize further degeneration, ideally within 3 weeks from the trauma. However, delayed surgical exploration within 6 months has been demonstrated to be efficient in some cases.

In the event that the treatment is ineffective or delayed for more than 1 year, classical surgery cannot be successful, and recourse to a variety of facial reanimation techniques is required. Patients who recover some motor function, spontaneously or surgically, but maintain some functional sequelae, such as residual weakness or synkinesis, can benefit from injections of botulinum toxin.

Inveterate paralysis also requires eye protection or even eyelid surgery, in order to prevent unpleasant ophthalmic complications.

See also Chapter 10 on facial nerve paralysis.

CSF leak

CSF leaks caused by temporal bone trauma usually tend to resolve spontaneously within 2 weeks. Therefore, this condition should be treated conservatively for 7–14 days. This approach includes several measures aimed at maintaining a low CSF pressure: total bed rest with elevation of the head, stool softeners, antitussive agents, and avoidance of coughing, nose blowing, or sneezing. Pneumococcal immunization has already been engaged, if indicated, in the acute phase. Many authors do not routinely prescribe antibiotics in CSF leakage, for fear of masking early signs of local infection. If the leak persists for more than 7–10 days, a lumbar drain should be positioned for 72 hours and if it fails too, a surgical closure of the fistula is recommended within 14 days. Translabyrinthine and posterior fossa fistula, as already mentioned, have very little tendency to spontaneous closure, thus they can be operated even before.

Figure 23.4 summarizes the diagnostic and treatment algorithm for post-traumatic CSF leak.

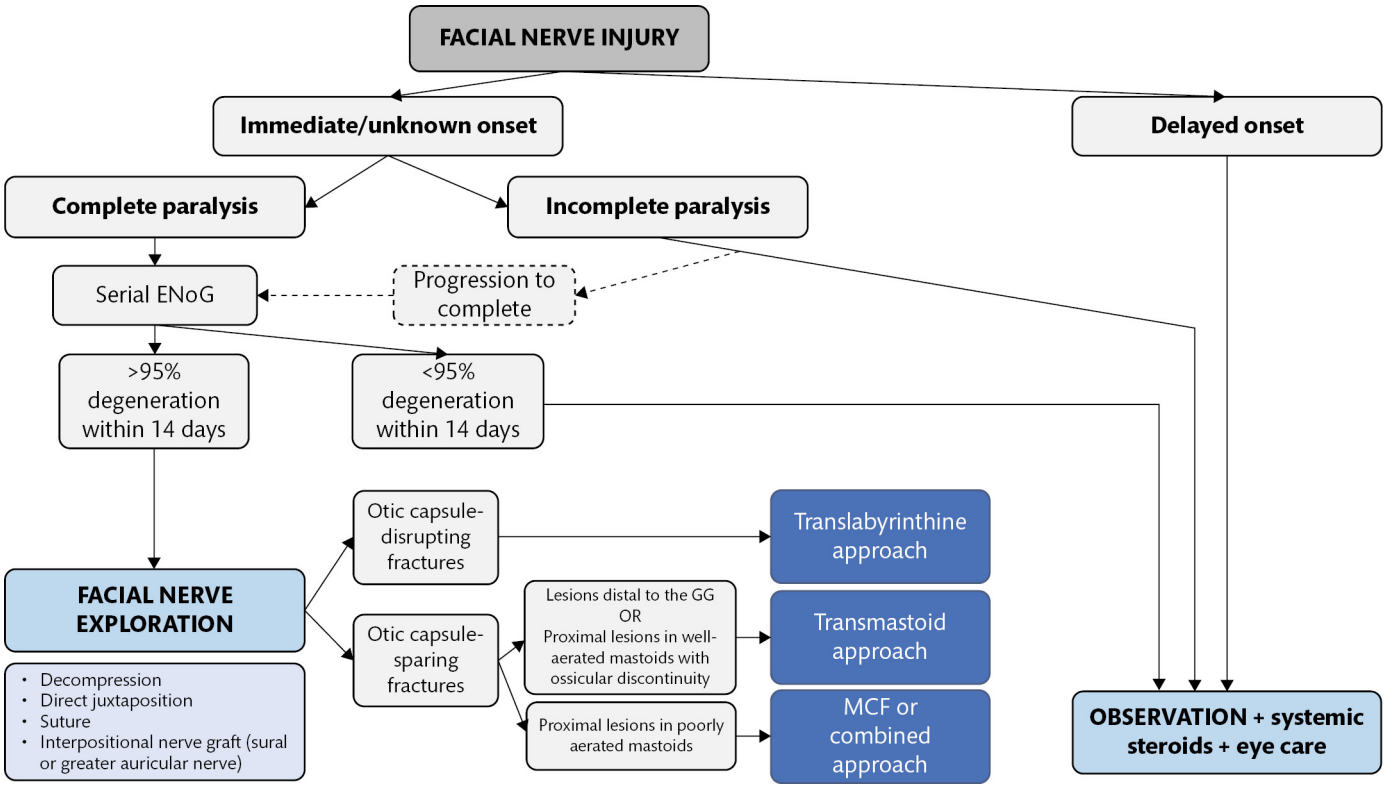

Figure 23.5 Management algorithm for post-traumatic facial nerve paralysis. ENoG, electroneurography; GG, geniculate ganglion; MCF, middle cranial fossa.

EAC stenosis

In case of EAC stenosis, early intervention in the first weeks can prevent a problem that is much more difficult to deal with once the bone callus is mature. The aim is to re-establish the patency and diameter of the canal and remove eventual fragments of invaginated skin in order to prevent cholesteatoma formation. Post-treatment ear packing by the use of modelled rubber-coated sponge tampons (Merocel type) should be removed after 3–7 days.

Conductive HL

Eighty per cent of post-traumatic conductive HLs recover spontaneously by resolution of the haemotympanum and healing of the tympanic membrane.

Surgical exploration is indicated when an air–bone gap of more than 30 dB persists for longer than 2 months after trauma. Surgery should not be recommended in the case of being the only hearing ear or severe to profound mixed HL not benefiting from hearing aids.

Eventual tympanic membrane grafting should be always complemented by an exploration of the ossicular chain mobility and eventual ossiculoplasty. Post-traumatic cholesteatoma and stenosis should be treated by habitual techniques. Post-traumatic stenosis of the Eustachian tube, if not recognized and treated, can compromise the functional outcomes of surgery.

In case of surgical failure or contraindication, bone-anchored hearing aids can be indicated in conductive or mixed HL.

Sensorineural HL and vertigo

Inner ear impairment can be empirically treated in the acute/subacute phases by systemic corticosteroid therapy (1–2 mg/kg), antinausea agents, and antivertigo agents but their utility has not been demonstrated in a traumatic scenario so far. Labyrinthine concussion may benefit from a 72-hour total bed rest. In post-traumatic perilymphatic fistulas and hydrops the initial treatment is usually medical, while patients with particularly discomforting vestibular symptomatology may benefit from surgery.

Vertigo deriving from post-traumatic peripheral vestibular deafferentation is expected to fade over time, thanks to the central compensation. However, a certain degree of disequilibrium always persists. Vestibular rehabilitation therapy can be useful in these situations.

In case of recurrent benign paroxysmal positional vertigo, repeated repositioning manoeuvres and behavioural recommendations are useful.

Sensorineural HL secondary to temporal bone trauma is typically severe to profound and permanent.

Traditional hearing aids are often inadequate but patients with profound deafness become candidates for cochlear implantation. A few issues must be considered when indicating a cochlear implant after a temporal bone fracture. First, post-traumatic ossification of the cochlea may complicate electrode insertion and request extra drilling or scala vestibuli insertion. Second, in the presence of a translabyrinthine CSF fistula, implantation is best avoided, due to the high predisposition towards developing postoperative meningitis. Third, post-traumatic implantations present a higher incidence of facial nerve stimulation, because of the generation of current leaks through the low-impedance ossification and fibrosis areas. The use of perimodiolar electrodes may reduce this risk. Finally, the sooner a cochlear implant is positioned after the traumatic event, the better results are. Preoperative HRCT and MRI are both mandatory, with the aim of verifying cochlear patency and ossification and exclude any IAC involvement or CSF fistula.

When the retrocochlear pathway is damaged too, auditory brainstem implantation may be considered.

Controversies

Indications for temporal bone HRCT

All patients with a head trauma routinely have a standard cerebral CT scan, while a more specific HRCT scan of the temporal bones is envisaged in case of clinical signs of temporal bone fracture. Fourteen to thirty-five percent of all temporal bone fractures detected by HRCT are not clinically evident. This is why some authors indicate temporal bone HRCT in all cases of head trauma.

Ear packing

The EAC is not to be packed initially in temporal bone fractures, due to the risk of iatrogenic injury. However, profuse bleeding can necessitate this procedure. Moreover, early enlargement and stenting of post-traumatic stenosis helps prevent a problem that is much more difficult to deal with once the bone callus is mature. Therefore, programming a well-timed intervention is critical for a correct risk/benefit ratio.

Immediate-onset versus unknown-onset facial paralysis

When the patient is not promptly examined at the emergency department, the facial function cannot be documented before intubation and administration of muscle relaxants. Thus, the diagnosis of facial paralysis is delayed by a few days, after coma arousal and extubation. This makes the definition of the time of onset very difficult, or even impossible. It is then crucial to differentiate 'delayed onset' from 'delayed diagnosis'. These cases should be defined as 'unknown' or 'unestablished' onset and treated equally to immediate-onset paralysis.

Antibiotic prophylaxis in cerebrospinal fluid leaks

Considering the high risk of meningitis in CSF fistulas, the role of prophylactic antibiotics must be examined. Many authors do not routinely prescribe antibiotics in CSF leakage, for fear of masking early signs of local infection. Multiple studies over the past tree decades have discussed this topic, with conflicting results. We personally believe that, knowing that the most common infecting bacteria in otogenic meningitis are *Streptococcus pneumoniae* and *Haemophilus influenzae*, an adequate antibiotic prophylaxis is a relatively inexpensive and safe strategy in the acute phase.

Efficacy of late facial nerve decompression

Modern literature agrees that facial nerve decompression should be performed as soon as possible to minimize further degeneration, ideally within 3 weeks from the trauma. However, in some circumstances immediate facial paralysis is not diagnosed or cannot be treated in the first weeks as recommended. In such cases late surgical exploration could be still indicated, ideally within 6 months. Our group reported on 13 patients who underwent facial nerve decompression 27–90 days after trauma after their temporal bone fractures, with recovery rates to a normal or subnormal facial nerve function (House–Brackmann grade I or II) at 1 year of 78%.

Guidelines

Nowadays there are no published national or international guidelines on the management of temporal bone trauma and fractures.

The Advisory Committee on Immunization Practices and the Centers for Disease Control and Prevention have released recommendations about the use of pneumococcal vaccination in the US. Similar guidelines for all Western countries suggest a single dose of PCV13 followed by PPSV23 at least 8 weeks later for adults with CSF leak who have not received either vaccine previously.

Conclusion

The management of temporal bone trauma has developed significantly in the last decades. Thanks to the improvement of intensive care, interventional radiology, and neurosurgery techniques, more and more patients bypass the critical phase; consequently, an increasing number of functional and aesthetical sequelae come to the attention of the otolaryngologist. Hence the importance of a complete and careful evaluation at each phase of treatment.

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24

Necrotizing otitis externa

Emma Stapleton and Glen Watson

Introduction

Necrotizing otitis externa (NOE) is a severe, invasive infection which has a significant impact on patients' quality of life. It starts in the external auditory canal, spreading via the fissures of Santorini at the osseocartilaginous junction, to involve the infratemporal fossa. It can cause a progressive osteomyelitis of the skull base, causing facial and lower cranial nerve palsies, venous and arterial thrombosis, intracranial abscesses, and ultimately death if untreated.

It typically presents with severe otalgia, purulent otorrhoea unresponsive to oral and topical antibiotics, and granulation tissue arising from the floor of the external auditory canal (**Figure 24.1**). Such a presentation, especially in the presence of risk factors, should alert the clinician to this diagnosis.

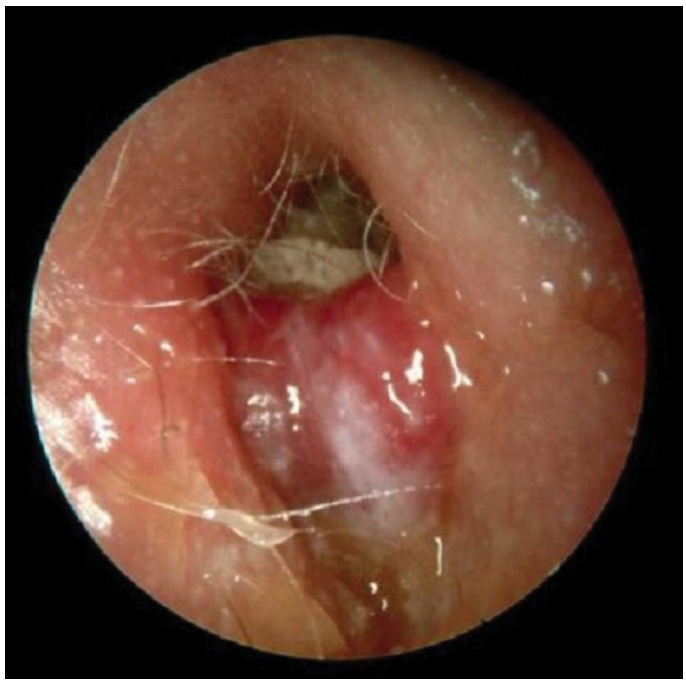

Figure 24.1 Image of granulations in the floor of the external auditory canal.

Risk factors include poorly controlled diabetes mellitus, advanced age, and other causes of immunosuppression including HIV/AIDS, steroid usage, chemotherapy, organ transplantation, leukaemia, lymphoma, and neoplasia. Local risk factors such as ear canal trauma or prolonged exposure to moisture in susceptible populations, and radiotherapy to the temporal bone, are also recognized. In published series, between 65% and 100% of patients have diabetes mellitus. The commonest causative organism is *Pseudomonas aeruginosa*; other bacteria and fungi can also be implicated. Patients with HIV and low CD4 counts are more likely to have a fungal infection. Prior to the introduction of antipseudomonal therapies, surgery was a mainstay of treatment and mortality as high as 67%, hence the now outdated term 'malignant otitis externa'.

First described in 1959, NOE has increased dramatically in incidence in the UK according to Hospital Episode Statistics data. There is currently no widely accepted treatment guideline for NOE. As its burden on health resources is significant, it is essential to identify cases early and manage them optimally.

Anatomy

The fissures of Santorini are natural openings within the anterior cartilage in the lateral aspect of the canal that may allow infectious or neoplastic processes to pass between the infratemporal fossa and the external auditory canal (**Figure 24.2**). A patent foramen of Huschke (foramen tympanicum) in the anteroinferior osseous canal also allows a similar process.

Diagnosis

Diagnostic criteria for NOE were published by Cohen and Friedman in 1987, though these are rarely used in the current literature. Interestingly, published case series incorporate a wide variety of diagnostic definitions for NOE, often including cases on the basis of a clinical diagnosis made by an experienced clinician. Indeed, a high index of clinical suspicion is required to make a diagnosis of NOE, and it is not uncommon for patients to have numerous hospital visits and failed conservative treatments before such a diagnosis is made.

The diagnosis should be made on the basis of the patient's history, risk factors, and examination, and confirmed with investigations

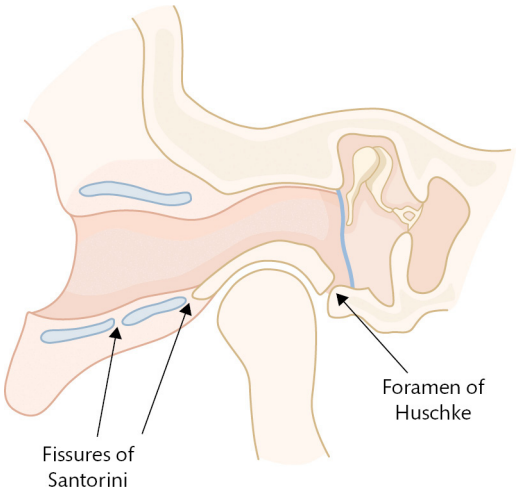

Figure 24.2 Ear canal anatomy featuring fissures of Santorini and the foramen of Huschke.

(Table 24.1). Patients may not always offer the classic presentation of 'an elderly diabetic with a painful discharging ear'. Presentations may be atypical, especially in the immunocompromised patient, and a high index of suspicion should be maintained when some, but not all, of these criteria are met.

The pain associated with NOE is typically severe, unremitting, and disproportionate to early clinical signs. Night pain and deep pain are suggestive of associated osteomyelitis. It is not uncommon for patients to complain of a hearing loss on the affected side; this is most likely to be a conductive hearing loss due to oedema, pus, and granulation tissue in the external auditory canal.

Advanced cases may present with cranial nerve palsies, most commonly a facial palsy, less frequently palsies of the glossopharyngeal, vagus, accessory, and hypoglossal nerves, and rarely palsies of the abducens or optic nerve. Very advanced cases can cross the midline and cause contralateral cranial nerve palsies, and can spread beyond the temporal bone causing abscess, meningitis, and death.

Table 24.1 Making a diagnosis of necrotizing otitis externa

Clinical history	Severe otalgia and a history of otorrhoea Non-response to oral and topical therapies
Patient risk factors	Diabetes mellitus Age over 65 years Immunosuppression (HIV/AIDS, steroid use, transplantation, chemotherapy, leukaemia, lymphoma, neoplasia) Trauma or prolonged moisture exposure causing epidermal compromise of the ear canal in susceptible populations Radiation to skin/parotid/temporal bone
Clinical examination	Oedema and/or granulation tissue in external auditory canal Facial or lower cranial nerve palsy (indicates advanced disease)
Investigations	Microbiology—swab and solid tissue analysis prior to antibiotics Histology—analysis of granulation tissue to exclude neoplasia Radiology—CT and MRI useful, discuss with local radiologist

Microbiology

The commonest causative organism in NOE is *Pseudomonas aeruginosa*, an opportunistic Gram-negative obligate aerobe which becomes pathogenic when host immunity is impaired. In such an environment it can invade local vasculature, cause focal necrosis, and form a biofilm. This means that topical therapies commonly used for pseudomonal otitis externa are ineffective, and superficial bacteriology swabs may yield no culture. Across published series there is a mean 25% incidence of antibiotic-resistant organisms in NOE. Other causative organisms are detailed in Table 24.2.

Fungal infection should be considered in the severely immunosuppressed. Fungal infection is associated with a worse prognosis than bacterial infection, though this may be due to host factors and later diagnosis. In the current world literature, fungal cultures are acquired in over 20% of primary and refractory cases.

In a patient with clinically diagnosed NOE, it is therefore essential to send both liquid (swab) and solid (granulation biopsy) samples for microbiology analysis, and to request both bacterial and fungal culture. Early discussion with colleagues in your local microbiology laboratory is essential. Prompt identification of the causative organism facilitates the commencement of appropriate therapies.

Histology and differential diagnoses

One of the main differential diagnoses of NOE is squamous cell carcinoma of the temporal bone, which can present as a rapidly progressing destructive process involving granulation tissue in the external auditory canal, and cranial nerve palsies. It rarely causes the severe, unremitting otalgia, which is characteristic of NOE; however, it is essential that a sample of granulation tissue is sent for histological examination, to confirm an inflammatory process and to exclude a neoplastic process. In the absence of tissue to biopsy in the external auditory meatus, deep surgical sampling via a cortical mastoidectomy may be required.

Inflamed or infected cholesteatoma can present with otalgia, otorrhoea, and granulation tissue which arises from the tympanic membrane or middle ear. There is often a long history of discharge and conductive hearing loss. Infected cholesteatoma can lead to osteomyelitis of the temporal bone in susceptible individuals.

Table 24.2 Causative organisms in necrotizing otitis externa

Bacterial	Pseudomonas aeruginosa (65%) Staphylococcus aureus Staphylococcus epidermidis Proteus mirabilis Klebsiella oxytoca Klebsiella pneumonia Burkholderia cenocepacia Enterobacter cloacae Streptococcus constellatus
Fungal	Aspergillus fumigatus Aspergillus niger Aspergillus flavus Candida spp. Malassezia sympodialis Scedosporium apiospermum

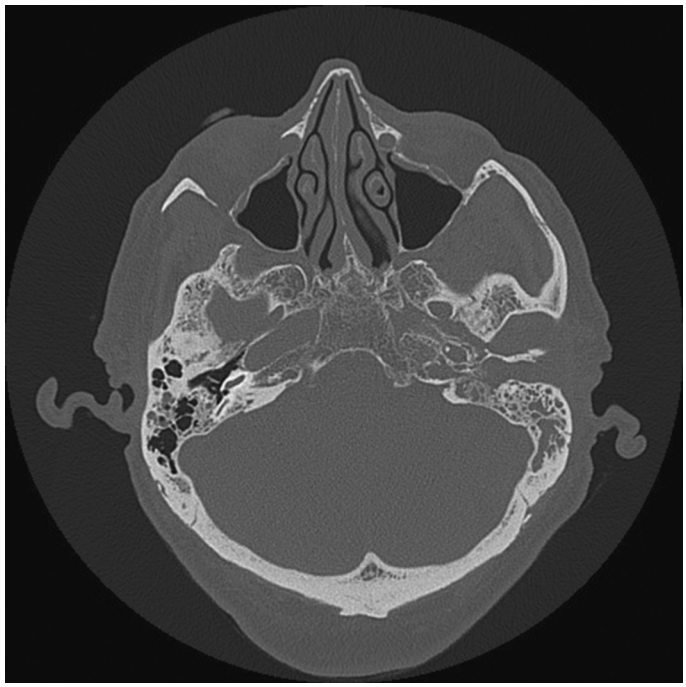

Figure 24.3 CT scan demonstrating bony erosion of the left skull base and ear canal.

Careful history taking and examination can identify cholesteatoma and guide appropriate management.

Other differential diagnoses include sarcoidosis and Paget's disease of the temporal bone. See also Chapters 18 and 19.

Radiology

Diagnostic imaging, and the use of imaging in monitoring NOE, remains heterogeneous across the world literature. Choice of imaging modality is often based on availability and local expertise, as there is little prospective evidence-based literature to support any particular modality. Across the world literature, at the time of writing, high-resolution computed tomography (CT) remains the modality of choice in over 90% of published case series, with 50% using magnetic resonance imaging (MRI) and 30% gamma- or positron-emitting nuclear tracers. It is common for more than one imaging modality to be utilized in the diagnosis and monitoring of patients with NOE.

CT is widely available in most centres, and despite delivering radiation, is a quick and inexpensive diagnostic test. Classic appearances of NOE on CT include erosion of the bone of the external auditory canal and skull base (**Figure 24.3**). More than 30% demineralization of the cortical bone is required for erosion to be apparent on CT. This means it is not an accurate tool for diagnosing early cases of NOE. Studies have identified that the lack of apparent soft tissue changes in early disease can be falsely reassuring, leading to undermanagement and early recurrence of inadequately treated NOE.

Post-contrast MRI scans with fat suppression and diffusion weighting can be very useful in detecting early soft tissue and bone marrow changes, as well as providing anatomical detail and aiding diagnosis of intracranial complications (**Figure 24.4**). Apparent diffusion coefficient can help differentiate between malignancy and inflammation. In patients for whom intravenous contrast is

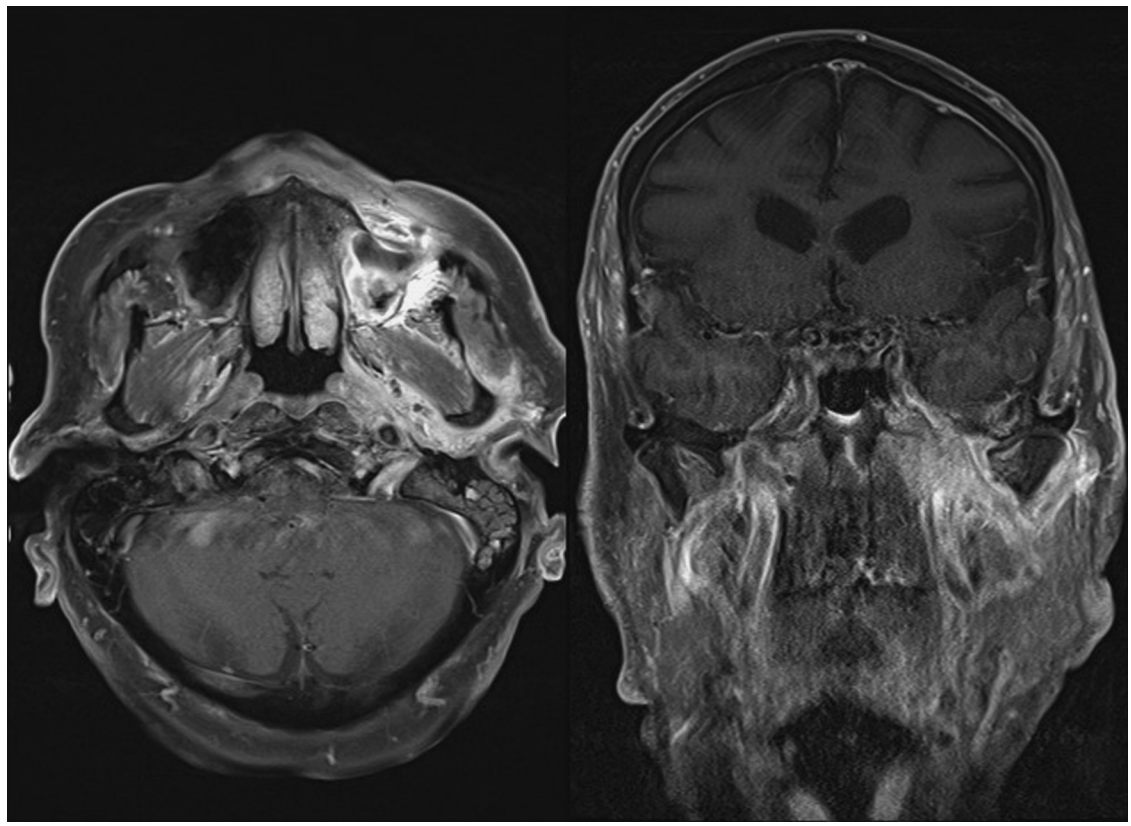

Figure 24.4 MRI scan with contrast demonstrating enhancement in NOE.

contraindicated, T1-weighted imaging can provide valuable information. It is always worth discussing cases with a radiologist in your local centre, as their experience and input can be invaluable.

Historically, gamma-emitting tracers such as technetium, indium, and gallium have been recommended as diagnostic and monitoring imaging modalities in NOE. However, they are not always easily accessible, they are expensive, have poor anatomical resolution, are time-consuming, and involve a high radiation dose. Their use has largely been surpassed by modern MRI. There is a growing body of evidence to support the use of beta-emitting fluorodeoxyglucose (FDG) combined with positron emission tomography (PET)-CT as useful for diagnosing and monitoring NOE. It is becoming more widely available, is effective in demonstrating response to treatment, and has a low radiation dose.

The use of CT and MRI to monitor response to treatment and diagnose recurrence has low specificity. Up to 100% of CT and 60% of soft tissue, and 30% of bone marrow changes on MRI, remain present for up to a year after the cessation of effective treatment. They still remain the imaging modalities of choice for monitoring due to availability. The use of FDG PET-CT for monitoring treatment response is gaining popularity.

Management

There is no widely accepted treatment algorithm or guideline for the management of NOE. The world literature demonstrates wide variation in treatment recommendations, some case series having treatment disparities between their own cases. A common theme is the immediate commencement of empiric intravenous antibiotic therapy, which can later be guided by the results of microbiological cultures. See Table 24.3.

Table 24.3 Management of patients with NOE

Conservative	Regular aural toilet Control of granulation tissue Analgesia
Multidisciplinary input	Radiology Diabetic team Elderly medicine/haematology Microbiology/infection
Systemic antibiotics	Empiric dual antipseudomonal therapy on recommendation of local microbiologist Adjustment of antibiotics according to culture, addition of antifungals as indicated Continue treatment for minimum 6 weeks, monitor patients clinically and radiologically
Cranial nerve deficits	Patients with facial palsy require meticulous eye care and ophthalmology review Patients with lower cranial nerve palsies require urgent speech and language therapist review and may require nasogastric tube or percutaneous endoscopic gastrostomy feeding
Surgery	Biopsy to obtain culture and histology Unclear role for surgery in refractory cases No evidence for facial nerve decompression
Hyperbaric oxygen	Can be considered for refractory cases

Multidisciplinary team input

Patients are frequently elderly, frail, and have comorbidities. They therefore require admission to hospital and multidisciplinary team input from specialists in otolaryngology, endocrinology (for optimal diabetic control), microbiology/infection diseases, radiology, elderly medicine, or haematology if they have a haematological malignancy. Patients with cranial nerve palsies require focused care for these. There is surprisingly little in the literature regarding dedicated multidisciplinary team care for patients with NOE; however, it remains essential to involve multidisciplinary colleagues to optimize patient care.

Conservative therapies

Regular aural toilet and control of granulations can aid pain control. The use of topical antibiotic therapies is unlikely to confer therapeutic benefit. Analgesia should be titrated according to patient requirement. It is well recognized that optimization of diabetic control leads to better clinical outcomes in NOE. Involvement of the specialist diabetic team is essential to achieve this, which can be challenging in patients with active infection.

Antimicrobial and antifungal therapy

Antimicrobial therapy should be commenced empirically, prior to the results of cultures. Advice should be taken from microbiology or infection colleagues, as there may be local policies in place. There is evidence to support dual therapy rather than monotherapy; this is likely to be due to dual therapy being effective against both sensitive and resistant strains of *Pseudomonas*. Typical empiric therapies are piperacillin and tazobactam, or ceftazidime with oral or intravenous fluoroquinolone. Fluoroquinolones are antipseudomonal, have good bone penetration, and few side effects. The addition of a cephalosporin is useful in the context of potential antibiotic resistance. It is reasonable to consider empiric antifungal therapy in patients with HIV and a low CD4 count. The antibiotic regimen can be altered, and antifungals added, depending on culture results.

There is no standardized guideline regarding the required duration of antibiotic therapy, though the consensus in the literature is that a 6-week course of intravenous therapy should be delivered, with clinical and radiological monitoring (see below) to define an improvement in the patient's clinical condition. In the UK it is usual for patients to be discharged from hospital and complete their course of intravenous therapy at home, delivered by a community team.

Surgery

Surgery was previously a mainstay of treatment for NOE. Prior to the availability of antipseudomonal antibiotics, radical debridement was the treatment of choice. Across the recent world literature, surgery for NOE continues to feature, mainly for the purpose of acquiring biopsies of granulation tissue, deep surgical sampling via cortical mastoidectomy, or release of pus from abscess cavities. Some centres advocate surgery for the removal of sequestered bone in refractory cases, and some for facial nerve decompression in cases where facial palsy is a feature, although there is no evidence that these are beneficial therapeutic interventions.

Hyperbaric oxygen

The use of hyperbaric oxygen as an adjunct to systemic antibiotic therapy is widespread in the world literature, with some centres

using it routinely for advanced or non-improving cases. A 2013 Cochrane review concluded that there was no evidence from randomized controlled trials to support this treatment. Its proposed mechanism of action is through increasing the partial pressure of oxygen, increasing the oxidative killing of microbes. It is likely that further data will emerge as more cases are treated.

Monitoring and outcomes

Monitoring of patients with NOE is essential to determine the safe cessation of treatment and to identify relapse. It is recognized that there is no single test which reliably demonstrates disease resolution. Across the literature, patient monitoring largely takes the form of pain resolution, clinical examination, inflammatory marker normalization, and serial radiological imaging.

Regular clinical examination of the patient during and after treatment is essential. Inflammatory markers are more controversial. Diabetic and immunosuppressed patients may not mount a normal response to systemic infection; however, white cell count, C-reactive protein, and erythrocyte sedimentation rate can be useful to monitor response to treatment in those who do. MRI is more useful than CT for monitoring response to treatment.

Definitive prognostic factors for NOE remain elusive, though advanced age, poor glycaemic control, and advanced disease at the time of diagnosis are associated with poorer outcomes. Overall, recurrence rates remain below 15% and mortality rate below 10%.

Key points

- NOE is a severe, invasive infection which requires prompt recognition and treatment.
- Patients are classically elderly, diabetic, or immunocompromised, and present with a painful, discharging ear refractory to oral and topical therapies.
- Atypical presentations are becoming more common, such as younger patients without pre-existing immunosuppression. A high index of clinical suspicion is required to identify these.
- Both liquid (pus) and solid (granulation tissue) samples must be sent to microbiology prior to the commencement of empiric antipseudomonal therapy. Granulation tissue should also be sent for histology to exclude malignancy.
- Radiological advice should be sought regarding imaging options in your centre.
- Multidisciplinary team input is required for optimal patient care (microbiology, radiology, diabetic team, care of the elderly).

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SECTION 3
Rhinology

25. **Clinical assessment of the nose and paranasal sinuses** *183*
Andrew C. Swift

26. **Olfactory disorders** *191*
Carl Philpott

27. **Epistaxis** *198*
Rajiv Bhalla and John S. Phillips

28. **Nasal septal perforation** *204*
Tim Woolford

29. **Rhinorrhoea** *208*
Amin R. Javer

30. **Postnasal drip** *215*
Isma Iqbal

31. **Headache and facial pain** *220*
Iain Hathorn

32. **Epiphora** *227*
Rohit D. Gohil and Ashok Rokade

33. **Nasal obstruction** *236*
Peter Andrews and Alfonso Luca Pendolino

34. **Nasal crusting** *242*
Catherine Rennie

35. **Rhinoplasty** *247*
Fazil Apaydin

36. **Acute rhinosinusitis** *267*
Isam Alobid

37. **Endoscopic sinus surgery** *274*
Claire Hopkins

38. **Chronic rhinosinusitis** *281*
Richard J. Harvey and Janet Rimmer

39. **Sinonasal tumours** *290*
Carl H. Snyderman and Eric W. Wang

40. **Surgical facial rejuvenation** *297*
Ozcan Cakmak and Mahmoud Daoud

41. **Non-surgical facial rejuvenation** *309*
Alwyn R. D'Souza and Chew Lip Ng

42. **Allergy of the nose** *314*
Richard Douglas

43. **Hereditary haemorrhagic telangiectasia** *319*
Simon Gane

44. **Endoscopic approaches to the anterior skull base** *326*
Haissan Iftikhar and Shahzada Ahmed

45. **Anterior skull base reconstruction** *333*
Caroline Smith, Omar N. Pathmanaban, and Rajiv Bhalla

25

Clinical assessment of the nose and paranasal sinuses

Andrew C. Swift

Introduction

In previous years, chapters that dealt with a protocol for the clinical assessment of the nose and sinuses would have been able to commence with a factual account of ideal techniques without the need for caveats. However, the severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) pandemic necessitated rapid change in clinical practice. While this chapter aims to provide suggestions to attain good clinical practice and excellence, guidance is included on how we minimized the risks induced by SARS-CoV-2 viral infection. These are important lessons that are mentioned should another pandemic ever recur.

The clinical history

The clinical history is of paramount importance in rhinological practice and will often reveal the likely diagnosis prior to examination. Taking a good clinical history is a skill that develops with experience. However, this process is dependent on various factors and can, on occasions, pose a significant challenge, even to experienced clinicians. The clinical challenges have been increased by patients researching their symptoms on the internet, leading to preconceived expectations about their diagnosis, management, and outcome, often associated with significant anxiety. However, certain symptoms are uncommon but require urgent attention (Table 25.1).

Specific rhinological symptoms

Nasal obstruction

Elucidate what patients are describing by using a variety of terms such as nasal stuffiness, congestion, blockage, or obstruction.

Clarify whether nasal obstruction is unilateral, bilateral, persistent, recurrent, cyclical, partial, or complete.

Rhinorrhoea/postnasal drip/nasal crusts

Patients will use a variety of terms influenced by regional dialect: runny nose, snot, cobb, bogey, discharge. Differentiate between a clear watery or mucoid discharge; intermittent or persistent discharge; discolouration (green, yellow); blood-stained; or viscous and glue-like.

Postnasal drip: this is a very common symptom and not a diagnosis that refers to a sensation of mucus running from the back of the nose to the throat. This is a normal physiological mechanism for mucus clearance but awareness is likely to occur for any disorder that increases nasal mucus flow or viscosity. Perception may also increase in the absence of recognizable sinonasal disease, as may occur with anxiety inducing focused attention on the throat.

Nasal crusts: retained mucus secretions may develop into moist or dry crusts. These are often erroneously referred to as 'crustaceans'.

Facial pressure, discomfort, and pain

Facial pain/discomfort is unlikely to be due to chronic rhinosinusitis (CRS) but patients will typically assume that they have sinusitis and remain unconvinced, even after sinusitis has been excluded.

A clear description of the pain or discomfort is essential. The history should elucidate details of individual pain episodes, an overview

Table 25.1 'Red flags' that require urgent face-to-face assessment

	Reason for concern
Unilateral symptoms	
Nasal obstruction	Sinonasal tumour
Blood-stained discharge	Sinonasal tumour
Persistent facial paraesthesia/ numbness or discomfort	Tumour in pterygomaxillary fossa
Proptosis	Mass behind eye
Watery nasal discharge	Cerebrospinal fluid rhinorrhoea
Complications of sinus disease	
Orbital swelling/displacement	Frontal sinus mucocele
Forehead swelling	Frontal sinus mucocele Mucopyocele and osteitis
Proptosis	Ethmoid sinus mucocele

of the symptom complex, the onset of symptoms, and details of individual episodes. Key information includes the frequency, duration, timing, inducing events, accompanying symptoms, and any phasic pattern of symptoms. Facial pain may initially be described as being episodic but it is important to determine if this is chronic with exacerbations.

Allergy-like symptoms

Allergy can induce a combination of nasal complaints and may be the prime cause or a contributory cause of the problem.

Key allergic-type symptoms include excessive sneezing, nasal itching, and in children, nasal rubbing (the nasal salute), and itching of eyes, throat, roof of mouth, and ears.

Patients may not perceive symptoms as being due to allergy or realize that a condition such as hay fever (seasonal rhinitis) is an allergy. Direct questioning is therefore recommended and should include seasonal variation, inducing factors, recognizable patterns of events, enquiry about pets, reaction to various foods, and specific drug allergy.

Sense of smell

Sense of smell is an integral function of the normal healthy nose that is impaired by many nasal disorders.

Anosmia versus severe impairment: an accurate description is essential—patients will often refer to an absent sense of smell when they have severe impairment or an intermittent absence of smell, as may occur in CRS.

True anosmia: this may be linked to head trauma and viral infections (particularly coronavirus disease 2019 (COVID-19)), but patients will worry most about the unlikely possibility of having a brain tumour.

Other smell disorders: an unpleasant smell, often detected by others, can arise with local infection of the nose and sinuses.

Upper respiratory tract viral infection can damage smell receptors and cause partial loss of smell (hyposmia) or distorted smell (dysosmia, parosmia) where a smell stimulus is typically perceived as an unpleasant smell such as burning wood or rotting food.

Phantosmia refers to recurrent episodic sensations of smell in the absence of an aroma/stimulus, caused by central factors including olfactory migraine.

Taste: impaired taste often accompanies loss of smell and typically recovers once smell improves. However, COVID-19 affected smell and taste separately, often causing complete loss of taste. Whilst most patients recovered, some were left with partial loss or abnormal taste (ageusia/hypogeusia/pargeusia).

NOTE

Maintain a wide vocabulary as this will help to understand the patient's clinical complaints.

Listen to the patient, allow them to speak before interjecting with closed questions. They will tell you what is wrong with them.

Be aware that patient's may have preconceived ideas about their complaint and increased anxiety from internet research.

An accurate diagnosis is dependent on a clear history. If the history is unclear, do not hesitate to re-check it or see the patient on another occasion.

Supplementary relevant medical information

This section is essential and shortcuts often eventually lead to an avoidable problem.

Previous medical history: this often contains highly relevant information, particularly with regard to previous nasal surgery or nasal trauma.

Comorbidities: comorbidities are surprisingly common and some, such as asthma, bronchiectasis, and aspirin-exacerbated respiratory disease/non-steroidal-exacerbated respiratory disease, are closely related to rhinosinusitis.

Granulomatosis with polyangiitis (GPA), cocaine abuse, and nasal sarcoidosis can all cause excessive crust formation within the nose as well as local destruction.

Diabetes, cardiovascular disease, cerebrovascular disease, rheumatoid arthritis, coagulopathies, and any other chronic conditions should be noted. Their potential impact on further management, and particularly surgery, should be considered.

Sleep: the effect of nasal obstruction on sleep and sleep deprivation is important to elucidate and may not always be forthcoming without leading questions.

Medication: enquiry into this domain is best dealt with in two sections—sinonasal medication including saline, and medication for various comorbidities.

Anticoagulants and antiplatelet medications will need specific management over the perioperative period if surgery is contemplated.

Other relevant factors: smoking, alcohol, and in some cases social drugs should be noted. Familial disorders may be relevant.

Occupation can be highly relevant, particularly exposure to dust, fumes, and air conditioning. Environmental factors can also play a role and include the home environment, country versus urban living, and effects of foreign travel.

Clinical examination

Most clinicians now use a strong narrow beam headlight, but circular mirrors on a headband with indirect light are still used in some countries.

General observation

Evaluation of the nose can begin early during the consultation, both with facial appearance but also with audible clues such as hyponasal speech, excessive sniffing, and mouth breathing.

Excessive smoking will induce an aroma on clothes, and excessive use of perfumes or hair products in women implies impairment of their sense of smell.

Initial ear, nose, and throat examination

The mouth and pharynx, ears, and neck should be included, noting the condition of dentition, mucosal surfaces and postnasal mucus discharge.

The face

Objective assessment of the face is easily overlooked when someone complains of nasal dysfunction. However, a quick review of the face will often prove beneficial when assessing nasal asymmetry. It will

also ensure areas that may be affected by underlying sinus pathology, such as the forehead, orbits, and the lacrimal systems, are included in the clinical assessment.

The external nose and face

The observer should specifically note facial symmetry and nasal alignment. Specific features such as forehead swelling and orbital signs such as displacement, proptosis, strabismus, eye movements, pupillary size and reaction, and epiphora, should be noted.

Specific features of the external nose should be considered prior to examination of the internal nose.

A detailed review of the external nose is required if the patient complains of a cosmetic deformity or a procedure that could affect nasal shape is recommended. Skin quality, thickness, and colour should be noted and the nose should be observed from all directions. Tip support, bony irregularity, and nasal tenderness should be noted. A formal photographic record should be arranged, particularly if surgery is recommended.

NOTE

It is surprisingly easy to overlook nasal deformity in patients with nasal obstruction, septal deviation, or sinonasal disease, only to regret it at a later stage.

The internal nose

The technique of examining the internal structures of the nose with a speculum is known as anterior rhinoscopy.

Prior to looking into the nose, the patient is asked to breathe normally through their nose, with the mouth closed. Each nostril should be occluded in turn by the examiner's thumb, without displacing the nasal septum. Misting can also be observed by exhalation onto a cold shiny Lac metal tongue depressor.

Anterior rhinoscopy: prior to nasal examination, the patient should be positioned so that the examiner has a good line of vision—the patient's neck should be slightly extended and the examination chair elevated. Nostril symmetry and columella dislocation is noted; the nasal tip is gently elevated with the thumb; the vestibules can then be examined and the nasal valve assessed. The valve area is triangular with the apex superiorly; the medial boundary is fixed by the cartilaginous septum and the pre-maxillary bone inferiorly; the lateral boundary is mobile and formed largely by the lower edge of the upper lateral cartilage, fibro fatty tissue, and the head of the inferior turbinate; the floor consists of skin overlying the piriform aperture.

Alar collapse during normal inspiration implies significant nasal obstruction in the nasal valve region. Lateral distraction of the cheek adjacent to the nose will open the nostril and often relieve obstruction if the valve is narrow (Cottle's test).

The septum

The nasal septum is viewed both without and with a nasal speculum, such as a Thudichum speculum. Choose a suitably sized speculum, control the speculum to avoid discomfort, and modify head position with the free hand to obtain the optimum view (Figure 25.1). Specific deformities such as septal deviation, spurs, or a perforation should be documented.

NOTE

Septal deformity may be complex and difficult to assess from the functional point of view. If in doubt, decongest the nose and re-examine.

Inferior turbinates, nasal cavities, and middle meatus

The inferior turbinates are dynamic and change the nasal cycle. However, their size and degree of congestion should be noted. See 'Nasal obstruction'.

The middle turbinates and middle meatus should be examined but may not be clearly seen in the congested nose. Excess mucus, mucopus, nasal crusts, polyps or masses should be documented.

NOTE

The degree of turbinate enlargement is subjective and re-examination after decongestion is recommended.

Large congested inferior turbinates are often mistaken for inflamed nasal polyps. Gentle palpation with a blunt probe will differentiate a polyp from a turbinate.

Deviation of the nasal septum is often accompanied by compensatory enlargement of the contralateral inferior turbinate.

Interventional procedures

Endoscopy

Nasal endoscopy can be performed with a rigid endoscope, a flexible endoscope, or a combination of the two. High-quality clear images can be viewed on a monitor and also recorded. The monitor should be placed directly behind the patient so that facial expression can be clearly observed should discomfort be experienced.

Rigid endoscopy

A standard endoscope is 4 mm in diameter, but a narrow 2.7 mm diameter endoscope is ideal for outpatient use and facilitates a wide-angle view at 0° or 30°. A narrow 30° endoscope facilitates a clear view of the middle meatus, middle turbinate, posterior septum, and nasopharynx, and has the advantage of not always requiring nasal decongestion or topical anaesthesia. The sphenoid os is not easily seen in the clinic setting.

Good endoscopic control is obtained by using a two-hand technique; the left hand supports the endoscope and camera while the right thumb remains in contact with the endoscope and the patient's face, thus avoiding inducing pain or bleeding from mucosal trauma (Figure 25.2).

Flexible endoscopy

A flexible endoscope facilitates examination of the nasopharynx and maxillary sinus should there be a patent antrostomy. The endoscope can also be passed through the nasopharyngeal isthmus into the oropharynx, hypopharynx, larynx, and oesophagus.

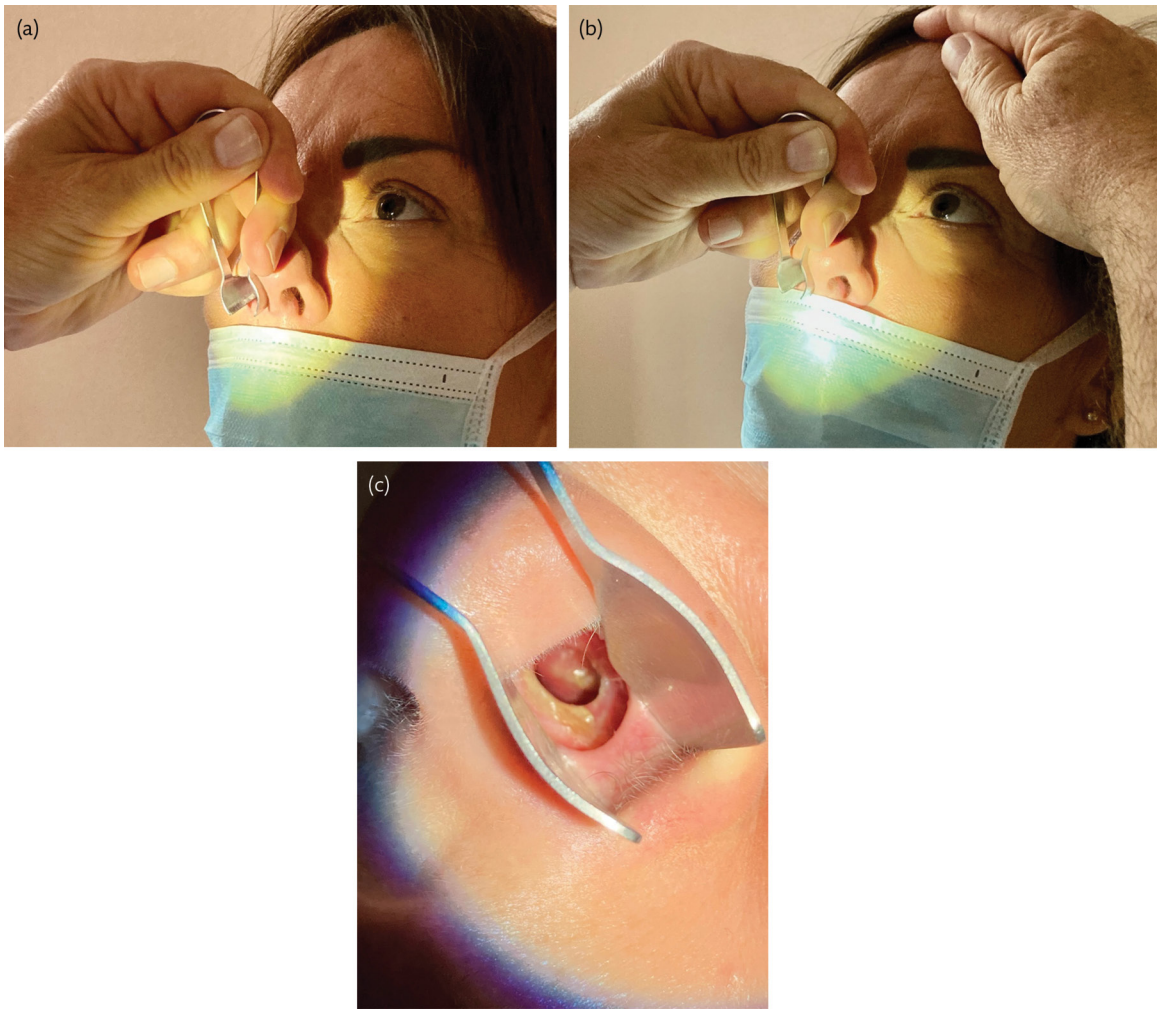

Figure 25.1 Recommended use of a Thudichum nasal speculum (face mask only required if risk of respiratory viral infection is high). (a) Tension of speculum spring controlled by fingers. Contact maintained on nasal dorsum. (b) Additional optional support by applying free hand to head. (c) View of anterior septal perforation via a Thudichum nasal speculum.

Diagnostic flexible endoscopes may be fibreoptic or chip-on-the-tip, and do not include a biopsy or insufflation channel.

Anatomical variants and pathological features

The middle turbinate may be widened by a turbinate air cell (concha bullosa), or be narrow and laterally displaced (paradoxically curved middle turbinate), narrowing the middle meatus.

The ostiomeatal complex refers to the area formed by the lateral surface of the middle turbinate, the uncinate process, and the ethmoid bulla, and includes the drainage pathways from the frontal, anterior ethmoid, and maxillary sinuses.

The true maxillary os is hidden lateral to the uncinate process and a visible os will be an accessory os.

Mucus tracks from the middle meatus are consistent with anterior ethmoid, frontal, or maxillary disease. Discharge from the superior meatus suggests posterior ethmoid disease.

In patients with an unremarkable unilateral polyp, close examination of the contralateral ethmoid may reveal subtle signs of polypoid mucosa, supporting a diagnosis of inflammatory sinus disease.

NOTE

A vomerine septal spur may be seen endoscopically, but may not always cause functional obstruction.

Remember to check endoscopes regularly. Light transmission of rigid and fibreoptic endoscopes relies on multiple flexible glass fibres which break with time, leading to poor image quality.

Nasal biopsy

Nasal biopsy is often considered in patients with newly diagnosed septal perforation, possible GPA or sarcoidosis, and in unilateral nasal polyps or masses. However, biopsy of septal perforations is rarely helpful and is not recommended unless there are unusual associated features, such as ulceration of the mucosa. In GPA and sarcoidosis, only abnormal mucosa should be sampled, and again, changes are typically non-specific.

Figure 25.2 Method of rigid nasal endoscopy utilizing free hand to steady the head and the thumb to control the endoscope. (Face mask only required if risk of respiratory viral infection is high).

NOTE

Taking a biopsy of a tumours and or unilateral polyp in the clinic may be tempting, but is best resisted and performed in theatre, where bleeding can be controlled and a representative sample can be taken.

A biopsy in theatre facilitates an excellent opportunity for detailed endoscopic assessment of the nose with maximal decongestion.

Investigations

Imaging

Plain sinus radiographs are no longer recommended.

The main imaging gold standard modality is the computed tomography (CT) scan of the sinuses. Modern-day multidetector CT scanners facilitate low-dose, rapid, multiplanar imaging with multidirectional reconstruction. Images can be set to display bony architecture or soft tissue detail, showing pathology and areas of osteoneogenesis and bony dehiscence. Details of intracranial structures are not included and need to be specifically requested in special circumstances, such as in patients with complications from orbital cellulitis.

Cone beam CT is rapidly advancing as a means of rapid, low-dose, good-quality imaging that may soon become more widely available within ear, nose, and throat clinics, facilitating a one-stop diagnostic service.

Magnetic resonance imaging may provide valuable supplementary information of sinus pathology in patients with inverted papilloma or malignant nasal tumours. This is particularly relevant where there is involvement of the orbit, intracranial structures, or facial soft tissues. Contrast may be required and will need a blood sample for renal assessment prior to imaging. Magnetic resonance imaging is often requested in patients with facial pain, and while pathology is often excluded, this is powerful reassurance for most patients.

Highly specialist positron emission tomography–CT scans are occasionally requested if tumours with metastatic disease are suspected.

NOTE

Ensure that the scan covers the whole sinus complex, including the dentition, inferior maxilla, and the superior aspect of the frontal sinuses. If the nasal tip is not included in a preoperative scan, calibration of an optical navigation unit may not be possible.

Discuss special requests with a radiologist, including the need for specific requirements and whether contrast is needed.

Always review the CT images and do not rely solely on radiology reports.

Interpret the radiological findings with the clinical presentation. Sinus opacity is often seen in the absence of clinical complaints, particularly in the base of the maxillary sinus.

Always look carefully at the dentition and bone adjacent to the teeth in patients who present with facial pain.

Objective smell tests

Objective smell tests are really useful for patients with a significant smell disorder and include the following:

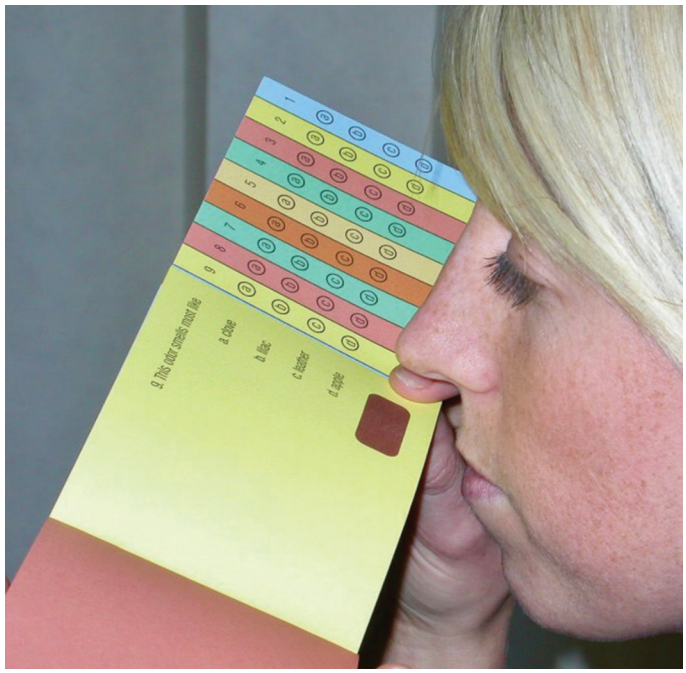

Figure 25.3 University of Pennsylvania Smell Identification Test (UPSIT) scratch and sniff smell test booklet. The smell panel is scratched with a pencil to release the odour. One of the four options must be marked for each page of the four test booklets (ten pages per booklet).

- Zurich smell diskettes—a simple smell disorder screening tool that assesses eight common odorants.
- University of Pennsylvania Smell Identification test (UPSIT)—a 40-item forced-choice super-threshold scratch and sniff test (Figure 25.3). This is now modified for use in the UK. It is easy to do but must be clearly explained and supervised. Good at identifying potential malingerers.
- Sniffin' stix—these provide both quantitative and qualitative information on the sense of smell that is particularly useful for more detailed analysis and research.

These individual tests are fully described elsewhere within this textbook (see Chapter 26).

Allergy tests

Allergy is an important disorder to identify in rhinological disorders. The clinical history is most important but diagnostic tests play a crucial role in confirming the diagnosis, to indicate the degree of atopy, and to manage expectations in refractory cases particularly where compliance with medical treatments may be an issue.

Skin prick tests

Skin prick tests have the advantage of providing both the patient and specialist with an immediate allergy result to eight to ten commonly inhaled selected allergens. Each screen is accompanied by a positive control (histamine) and a negative control (allergen carrier solution). Allergens typically include European and American house dust mite (*Dermatophagoides pteronyssinus* and *farinae*), cat and dog dander, grass pollen, *Aspergillus fumigatus*, *Alternaria alternata*, and *Cladosporium herbatum*.

A positive result will induce a wheal and a surrounding flare. The diameter of the wheal is measured and recorded. Mild reactions are open to interpretation and should be compared with the negative control. Food allergens are not normally tested because of the risk of inducing severe anaphylaxis.

Blood tests

These include total immunoglobulin E levels and specific immunoglobulin E antibody testing (radioallergosorbent tests (RASTs)) for a panel of commonly inhaled selected allergens, and food panels such as wheat and dairy products if clinically suspected. RAST tests offer a large range of potential allergens that can be tested, according to the medical history. They are both sensitive and specific, are unaffected by systemic medication including antihistamines, and provide a quantitative result for the level of allergen antibodies.

Choice of allergy test

Most specialists interested in allergy would recommend that both skin prick and blood tests should be used according to clinical need. RASTs are safe and particularly useful for patients with possible food allergies. Systemic antihistamines suppress skin prick tests and these will either need to be stopped 3–5 days prior to testing or alternatively, proceed with RAST only. RAST may be expensive though and this should be borne in mind. Skin prick tests should be avoided if the test site is affected by eczema or the patient has known dermographia.

NOTE

The patient's history is most important for a diagnosis of allergy.

Skin prick tests and RASTs are sensitive and specific, with good correlation of results. They may be used individually but are often complementary when used together.

RASTs should be used for potential food allergens.

Blood investigations

Blood tests often provide important diagnostic information in rhinological practice and the following should be considered:

- *Full blood count*: identifies anaemia and eosinophilia. The latter occurs particularly with eosinophilic granulomatosis with polyangiitis (EGPA, previously known as Churg–Strauss syndrome).
- *Renal function (urea, estimated glomerular filtration rate, serum creatinine)*: renal failure may induce smell disorders, epistaxis, nasal dryness, and crusting.
- *Total immunoglobulin E and RASTs to specific allergens.*
- *Immunoglobulins and functional antibody titres*: to detect immune deficiency disorders.
- *Thyroid-stimulating hormone and free thyroxine*: hypothyroidism is easily overlooked.
- *Vascular screen*: antinuclear cytoplasmic antibody screen, anti-myeloperoxidase and anti-proteinase 3 antibodies, angiotensin-converting enzyme, erythrocyte sedimentation rate, C-reactive protein, and renal function are essential investigations for

vasculitic disorders such as granulomatous disease (e.g. GPA, EGPA, and sarcoidosis).

Mucociliary flow

Mucus flows from the anterior nose to the nasopharynx by mucociliary transport that generally takes about 20 minutes. Should mucus stasis occur, nasal crusts will form. Mucociliary flow may be impaired by infection but is occasionally caused by primary mucociliary dyskinesis.

Mucociliary flow can be assessed by placing saccharin on the inferior turbinate and noting the time delay before it is tasted by the patient. A normal result is if the saccharin can be tasted within 12–20 minutes.

Nasal physiology

Nasal physiology investigations, if available, provide objective measurements of nasal function:

- *Peak nasal inspiratory flow*: assesses the overall inspiratory airflow during inspiration with maximum effort.
- *Anterior rhinomanometry*: assesses nasal resistance.
- *Rhinospirometry*: assesses the relative flow between the nasal airways.
- *Acoustic rhinometry*: indicates the relative deviations or obstructions within the nasal cavity. It is most useful for assessing the nasal septum.

Details of all of these techniques are described in detail in Chapter 33.

Clinical scores

Many clinical scoring systems have been devised but this section provides a brief description of the most commonly utilized scores.

Sinonasal Outcome Test: the SNOT-22 is a disease-specific quality-of-life questionnaire with 22 questions that is validated as a patient-related outcome measure for CRS. It is particularly useful for monitoring the response to medical and surgical interventions for CRS, but also alerts the clinician to the likelihood of anxiety and depression.

Visual Analogue Scale (VAS): the VAS is widely used to quantify total nasal symptoms from 0 for absent symptoms to 10 for very severe symptoms. Symptoms of CRS can be classified into mild, moderate, or severe. The total symptom VAS has been used for assessing disease control, but individual VAS scores for specific symptoms may not be equally useful.

Nasal Obstruction Symptom Evaluation (NOSE): this is another patient-related outcome measure in which five questions related to nasal obstruction are scored by the patient on a Likert scale from 0 to 4 and the total score is multiplied by 5, giving a total highest possible score of 100. It has also been referred to as the *Nasal Obstruction and Septoplasty Effectiveness Scale*.

Lund–Mackay Score (LMS): the LMS is the most common, validated means of scoring CT sinus scans for CRS, ranging from 0 to 24.

Lund–Kennedy Endoscopic Score (LKES): this score is based on endoscopic findings that include oedema, discharge, scarring, crusting, and polyps.

Adaptive change after COVID-19

The nature of the clinical activity of otolaryngologists puts them at direct risk of viral transmission from any patient who is affected.

Protocols have been developed to reduce this potential risk. Telephone consultation is now refined and accepted, facilitating investigations to be organized before a direct face-to-face consultation.

During the risk phase of the pandemic, additional precautions were taken. Patients attending for face-to-face consultations were screened prior to arrival to exclude symptoms of COVID-19, and temperature was checked on arrival at clinic. Social distancing was practised in waiting rooms. It was also compulsory for patients and hospital personnel in the UK to wear a facemask. These measures have currently been relaxed.

Nasal/nasopharyngeal endoscopy was accepted as a potential aerosol-generating procedure within the UK. Recommendations included full personal protective equipment, and endoscopy was performed in a separate procedure room with good ventilation. Patients are asked to avoid sneezing and coughing if at all possible, and to keep a mask over their mouth throughout the endoscopy.

Areas of controversy and uncertainty

Many classifications of sinonasal disorders have been described but typically lack precision and rely on subjective assessment across a spectrum of severity.

Early classification of CRS was primarily determined by the presence or absence of polyps, but current thinking is now based on the phenotype and genotype. Readers are referred to the 'European Position Paper on Rhinosinusitis and Nasal Polyps 2020' document for details (see 'Further reading').

Nasal polyp score: this initially appears to be a simple issue but the fact that there are several scoring systems and modifications implies the difficulty. A typical example is described:

- *Polyp grade 1–3*: mild polyps (1) not reaching the upper edge of the inferior turbinate, moderate polyps not extending below the inferior edge of the turbinate (2), and severe polyps (3) extending below the inferior edge of the inferior turbinate.
- *Polyp grade 0–4 (Meltzer)*: no polyps (0), polyps confined to the middle meatus (1), multiple polyps within the middle meatus (2), polyps extending beyond the middle meatus (3), and polyps completely obstructing the nasal cavity (4).

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26

Olfactory disorders

Carl Philpott

Basic science: anatomy and physiology

The neural pathways

The olfactory neuroepithelium is primarily located underneath the cribriform plate but a variable distribution may also be found on parts of the superior and middle turbinates and the nasal septum. Each olfactory cleft on either side of the nose is about 1–2 mm wide with about 200–400 mm^2 of olfactory epithelium. Proportionally, humans derive less from olfaction in the context of nasal surface area; only 15% of human nasal mucosa is designated to olfactory function compared with 40% in rats.

The cellular make-up of this epithelium includes the olfactory sensory neurons (OSNs) along with an array of supporting cells that include sustentacular, microvillar, globose basal, horizontal basal, and duct cells, as well as Bowman's glands. The OSNs have cilia containing seven transmembrane receptors and project into the mucus layer. The OSNs give rise to basal axons that form roughly 20 bundles of approximately 50 axons each, known as 'fila', which connect to the olfactory bulbs (OBs) above the skull base. Within the OBs, the OSNs synapse with the second-order neurons (mitral and tufted cells) inside the glomeruli, which is the second layer of the OBs. Negative and positive feedback is provided by means of parallel circuits in the periglomerular and external plexiform layers. The olfactory epithelium undergoes retraction with increasing age and the numbers of glomeruli decrease in conjunction with the decline in olfactory receptor cell numbers within the epithelium; they are almost non-existent by the age of 80 years.

From the OBs, second-order neurons project posteriorly as the olfactory tracts to various areas including the thalamus, the limbic system, and the orbitofrontal neocortex (secondary olfactory cortex). Odour discrimination takes place in the secondary cortex and affective responses are controlled by the limbic system. Projections of the second-order neurons to the primary olfactory cortex are direct connections with some neurons connecting in turn directly to the secondary olfactory cortex and some relaying via the thalamus between these two cortical areas. The primary olfactory cortex includes areas such as the anterior olfactory nucleus, the olfactory tubercle, and the piriform cortex. The anterior commissure may also carry decussating fibres from pyramidal cells of the anterior olfactory nucleus to contralateral elements of the primary olfactory cortex.

How 'smell' happens

Smell can be considered to occur in two phases, like hearing: conductive and sensorineural. Conduction involves the passage of odours into the nose and then olfactory transduction relies on interaction between odour molecules dissolved in the mucus layer and the transmembrane receptors of the cilia. During orthonasal olfaction, up to 15% of the incoming air stream flows towards the olfactory cleft during inhalation and turbulence provided by the turbinates mixes odours during inspiration. Retronasal olfaction is the passage of food odours from the oral cavity while eating and accounts for 75–80% of flavour perception. However, as it happens at the same time as gustatory stimuli, people find it hard to separate the two processes in their mind. Sniffing improves the processes of increasing airflow and mixing the air and is modified by the cerebellum. Odorant molecules that do not reach the olfactory receptors by diffusing through the mucus are actively transported via odorant binding proteins. Sustentacular cells may also deactivate some odorants and xenobiotic agents.

Odorant arrival at the G-protein coupled receptors stimulates firing of an action potential in the primary afferent fibre and this is in proportion to the odorant concentration. OSNs appear to be located in specific groups according to the class of odours that they are sensitive to and this arrangement enables the central nervous system to receive a spatially coded signal that conveys in part the quality of the odours. Each OSN is genetically coded to express a specific receptor gene and all OSNs with the same expression project to the same glomeruli of the OB. Cellular mechanisms allow the human nose to detect approximately 10,000 odours; in comparison 40,000 can be detected by a bloodhound. The central connections are described above, with olfactory discrimination and identification occurring centrally and thresholds modified centrally by means of a negative feedback loop in the OBs. **Figure 26.1** summarizes the key mechanisms at the receptor and bulb level.

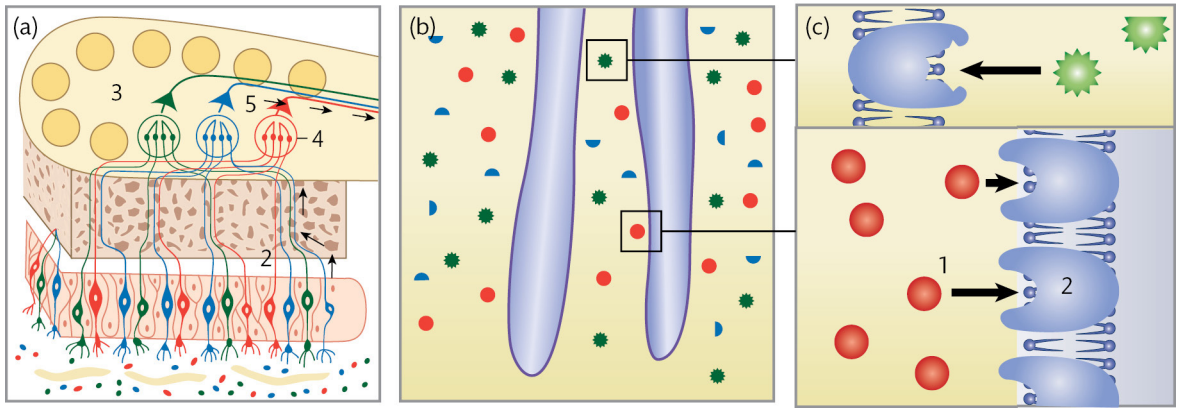

Figure 26.1 Olfactory detection. (a) (1) Scent molecules dock on to receptors; (2) bipolar nerve cells; (3) olfactory bulb with (4) glomerulus (microcentre) and (5) mitral cells. (b) (1) Scent molecules; (2) scent receptors. Each olfactory cell bears only a single type of receptor. There are 350 receptor types in humans. A receptor can recognize up to 100 structurally similar molecules in a scent category. (c) Scent molecules fit only one specific receptor. They trigger a biochemical reaction and a subsequent electrical signal, which is transmitted to the olfactory function in the brain.
Source data from Welge-Luessen A, Hummel T. *Management of Smell and Taste Disorders: A Practical Guide for Clinicians*. Thieme; 2013.

Assessment

As ever, a careful history will elucidate many key details regarding the underlying disorder (Table 26.1). Patients may complain of a number of primary symptoms including loss of smell and taste, smell distortions and imagining smells which are not present, or of a bad smell inside their nose. Key factors include the onset, timing, and duration of the olfactory disorder including the recall of any precipitating events and of any associated symptoms. Listening carefully to what the patient describes and then clarifying what they mean helps to discern the details. If they complain of loss of taste, ask them specifically if they would notice a teaspoon of sugar in a cup of tea or the salt on some crisps. Details of the past medical history and drug history are also important where iatrogenic and systemic causes may be relevant.

Examination of the nose should ideally be performed with a rigid 30° endoscope to visualize olfactory clefts. Examination of the tongue, oral cavity, and oropharynx with or without hypopharynx may also be needed. As recommended by the 2023 Position Paper on olfactory dysfunction, psychophysical testing should be considered mandatory. Patients' ability to accurately assess their own sense of olfactory performance is poor and therefore needs qualifying (Figure 26.2). The information may also help to guide prognosis and treatment options. There are a number of commercially available and validated smell test kits ranging from screening tests such as the Pocket SIT to the 40-item UPSIT and the comprehensive Sniffin' Sticks test.

Investigations

Investigations must be clearly directed by the history and examination and imaging is not needed for all cases. Choice of imaging continues to be a source of confusion in clinical practice, so a simple guide is as follows:

- Any patient with additional rhinological symptoms and/or endoscopic findings of chronic rhinosinusitis (CRS) should be managed

Table 26.1 Terminology related to olfactory disorders

Olfactory term	Definition	Related terms
Dysosmial*	Olfactory dysfunction, in general (quantitative or qualitative)	Quantitative olfactory dysfunction Qualitative olfactory dysfunction
Anosmia	Quantitatively reduced olfactory function to the extent that the sense of Functional anosmia smell is not useful in daily life	
Hyposmia	Quantitatively reduced olfactory function	Microsmia
Normosmia	Quantitatively normal olfactory function	
Hyperosmia	Quantitative increase in olfactory function, corresponding to an ceptionally high score in a psychophysical olfactory test (e.g., above the 90th percentile)	
Olfactory intolerance	Qualitative olfactory dysfunction where individuals, without odor distortions, complain of a subjectively enhanced perception of odors and are intolerant of everyday odors	
Parosmia*	Distorted perception of an odor in the presence of an odor stimulus	Euosmia
Phantosmia or olfactory hallucination	Odor perception in the absence of an odor stimulus	

* For clarity, 'parosmia' and 'dysosmia' are not regarded as synonymous.

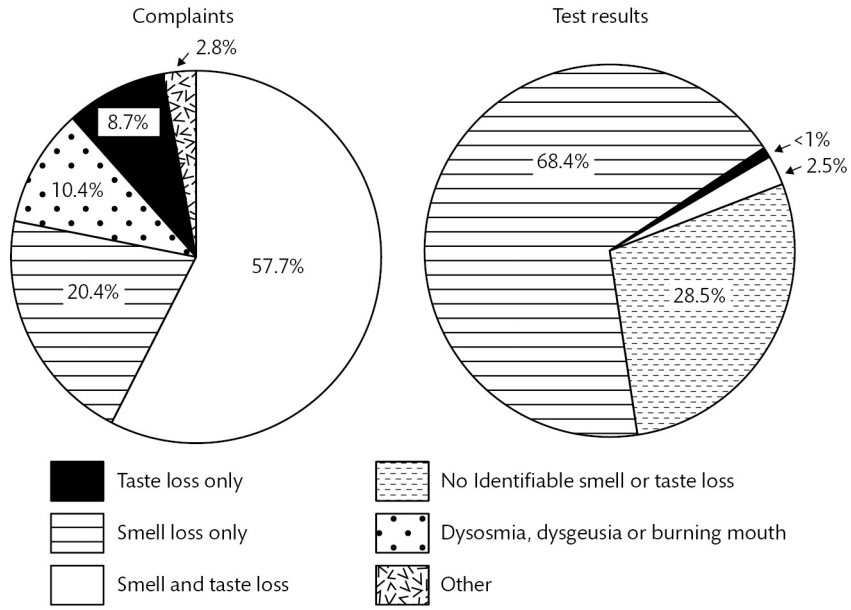

Figure 26.2 The comparison between patient complaints and performance on psychophysical testing.
Reproduced from Doty R. *The Handbook of Olfaction and Gustation*. 2015 with permission from John Wiley and Sons.

in accordance with European Position Paper on Rhinosinusitis and Nasal Polyps 2020 (EPOS2020) guidelines including a computed tomography scan where either confirmation of the diagnosis is needed or where sinus surgery is being contemplated or perhaps if complications of sinusitis are considered possible.

- Any patient with a clear history of viral aetiology in the absence of any endoscopic findings does not need any imaging unless it is for research purposes.
- All other cases should be considered for magnetic resonance imaging (MRI) scanning, ensuring the sequence includes a T2 coronal sequence via the OBs. This allows for assessment of neuronal gliosis in post-traumatic cases, assessment of OB volume, exclusion of other central pathology, and assessment of the olfactory clefts.

Occasionally, the MRI findings may prompt the need for an additional computed tomography scan to be performed.

In cases where there is no clear precipitating factor or obvious aetiology, selected blood tests should be considered in line with the patient's medical history and metabolic and trace element deficiencies can sometimes be a reversible cause for dysfunction.

Other investigations may typically be considered in a specialist clinic, or a research domain, or for medicolegal purposes. These include olfactory event related potentials, functional MRI, retronasal olfactory testing, and gustatory testing. The latter may be needed where the history indicates that true gustatory dysfunction may be present; as is seen in Figure 26.2. Although many patients will complain of taste disturbances, they are actually experiencing poor retronasal olfaction and will still be able to detect the basic taste modalities of salt, sweet, sour, bitter, and umami.

Differential diagnoses

See Table 26.2 and Figure 26.3.

Chronic rhinosinusitis

Disturbances of smell are one of the diagnostic criteria for CRS according to the EPOS2020 guidelines (see Chapter 38). CRS is a common disorder and therefore it is not surprising that it represents the biggest cause of olfactory dysfunction. Usually patients present with the array of symptoms that CRS gives rise to but occasionally the insidious nature of onset may lead to a patient to complain only of the olfactory disorder despite quite advanced inflammatory changes. Endoscopic and radiological findings are used to confirm the diagnosis. While research to determine the timing and appropriateness of long-term antibiotics and sinus surgery is ongoing, use of topical and systemic steroids along with saline nasal irrigations are supported by evidence from meta-analyses (see Cochrane database). Smell loss is likely to improve with disease control but in some patients, respiratory metaplasia can occur in the olfactory epithelium as well as the deleterious effect of bacterial toxins on neuronal

Table 26.2 Differential diagnoses and relative frequency as a percentage of all cases that present for medical advice

Conductive	Sensorineural
Chronic rhinosinusitis (50%)	Postviral olfactory dysfunction (19%)
Other sinonasal disorders (deviated septum, allergic rhinitis, adenoidal hypertrophy, olfactory cleft disease/stenosis) (15%)	Post-traumatic olfactory dysfunction (5%)
Iatrogenic (sinonasal surgery) (3%)*	Idiopathic (6%)
Toxic rhinitis (2%)	Congenital (1%)
	Iatrogenic (medications) (3%)*
	Other neurological and systemic causes

* 3% is for all iatrogenic causes.

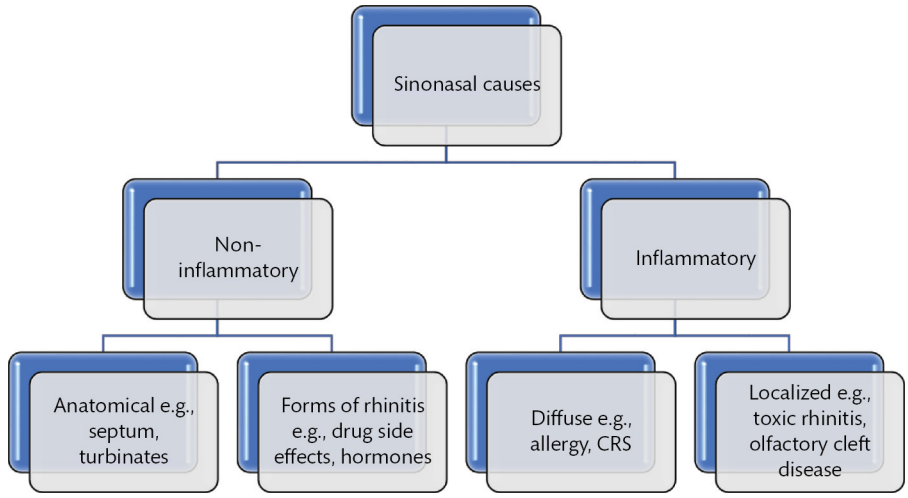

Figure 26.3 Conductive causes of olfactory dysfunction.

function. Systemic corticosteroids will normally determine the reversibility of any smell loss in these cases.

Postinfectious olfactory dysfunction

Patients who experience post-infectious olfactory dysfunction (PIOD) typically notice the unusual coryzal symptoms of an upper respiratory tract infection but as the other symptoms dissipate, their sense of smell does not return. Many report prior episodes where the loss of smell is temporary and also that this infection is more severe than previous upper respiratory tract infections. The typical demographics are in the age range 40–70 years with a female-to-male ratio of 2:1. The history should clarify as to whether any of the other rhinological symptoms have also persisted as the upper respiratory tract infection could have been the tipping point for them developing CRS. Patients in this group also typically experience parosmia, especially when hyposmic rather than anosmic and although disturbing for the patients, it is often a good prognostic sign of recovery. Since 2020, the COVID-19 variants have had a significant impact with regards to PIOD, with there being an estimated 0.5–1 million cases in the UK. Many patients who were infected early in the pandemic are still finding their way through the referral system to tertiary clinics. The characteristic feature of COVID-19 PVOD is the sudden onset of the anosmia which is sharply noted by patients. There is some potential difference in the pathophysiology of COVID PIOD compared to non-COVID cases and they may need to be considered separately in future research. In general, about one in three cases with PVOD will spontaneously recover over a 3-year period.

Post-traumatic olfactory dysfunction

Patients in this category usually have a clear timeline of a head injury preceding their loss of smell. However, in more severe cases, unconscious patients will obviously not be aware of the missing sense and even after consciousness returns, may have other distractions due to other injuries/morbidity. Many patients will make the assumption that the poor quality of hospital food is what accounts for the food seeming bland and 'tasteless'. Presentation timing can therefore vary hugely, especially as advice given after head injury in relation to smell loss is often very limited. Details of the mechanism of injury may help to determine the location of the impact within the olfactory apparatus. Where facial injury is involved, there may in addition be a conductive component due to septal deviation and possibly even secondary rhinosinusitis. In the presence of clear watery rhinorrhoea, a cerebrospinal fluid leak must also be suspected. Blows to the occiput may result in contrecoup injuries to the frontal cortical regions; acceleration/deceleration injuries may lead to shearing trauma to the olfactory fila and/or OBs. Alongside smell loss, patients may also typically experience phantosmia. The chances of spontaneous recovery may be linked to the degree of damage cause by the initial injury, but overall about 30–50% may notice some improvement over a 10-year time frame.

Iatrogenic

Cases in this category can be medical and surgical. Medications leading to olfactory dysfunction are reported to include allopurinol, amitriptyline, amphetamines, amphotericin B, azathioprine, carbimazole, cocaine, codeine, colchicine, diltiazem, gold, methotrexate, metronidazole, morphine, nifedipine, streptomycin, and thiouracil. Clinicians are advised to consult the *British National Formulary* if a patient reports smell loss that has started following the commencement of new medications. In noting that cocaine can have a deleterious effect on the olfactory epithelium, surgeons should be cautious in their method of application in preparation for endoscopic sinus surgery.

Surgery in itself can obviously have a deleterious effect on olfaction, especially if there is resection of potentially OSN-bearing areas such as the middle and superior turbinates during endoscopic sinus surgery and trans-sphenoidal hypophysectomy. This is because of the aforementioned distribution of the olfactory epithelium. Meticulous technique and avoidance of synechiae formation postoperatively will help to reduce occlusion of the olfactory clefts. Patients undergoing skull base surgery or neurosurgery should be warned of the potential for smell loss as appropriate, particularly if there is close proximity to the OBs or the olfactory clefts.

Toxic rhinitis

If a patient presents with smell loss following exposure to a hazardous chemical, they will usually experience burning/pain in the nose at the time of the exposure, followed by subsequent smell loss. Examination findings may indicate inflammatory changes within the nasal cavity and olfactory clefts. Some patients may describe sensitization following exposure to strong chemical smells with ongoing qualitative disturbances on repeated exposure. The latter does not fit with a diagnosis of toxic rhinitis and they will not have experienced any discomfort at exposure, nor should there be anything to see on examination.

Neurological disorders

Smell loss can potentially be associated with a number of neurological disorders but two key ones to consider are Parkinson's disease and Alzheimer's disease. At the point of diagnosis of Parkinson's disease, 90% of patients will be anosmic and it is thought that this may occur up to 5 years prior to the onset of the other clinical symptoms and signs. Smell loss is estimated to affect about 50% of those with Alzheimer's but furthermore, it becomes increasingly common over the age of 65 and as indicated above, especially over the age of 80. If neurological disease is suspected, the history should explore other focal neurological symptoms and signs and appropriate onward referral may well be needed. Anosmia is also now shown be an independent risk factor for reduced longevity and is associated with increased frailty.

Idiopathic

While representing a small percentage of cases in the wider community, smell loss with no attributable cause represents about 20% of cases presenting to specialist smell and taste clinics. Patients in this category present with a gradual onset of smell loss, often over several years, and in some cases may be prompted by a family member when they suddenly realize they are missing everyday smells. It is important to carefully pick through the history and exclude any potential precipitating factors or any relevant medical history. These patients typically need screening with blood tests and an MRI scan to confirm they are idiopathic. It is always possible that, in some cases, a patient may have suffered a subclinical upper respiratory tract infection and as indicated above, it is also possible that smell loss is a heralding sign of neurological disease and important to warn patients to be vigilant for other signs, especially over the age of 50.

Congenital

A history of never having been able to smell since early in childhood is likely to represent a congenital case but does require objective confirmation. An MRI scan will help to determine if there is complete aplasia of the OBs or any other congenital malformations; on occasion there may be evidence of olfactory cleft stenosis due to the position of the middle and superior turbinates. Kallman syndrome represents the combination of OB aplasia/hypoplasia in conjunction with hypogonadotropic hypogonadism. In prepubescent children there will be a need to involve a paediatrician in the management; if puberty has occurred normally, it is unlikely that Kallman syndrome exists and thus it is simply an isolated case of congenital anosmia for which, at present, there is no specific treatment.

Other differential diagnoses

As indicated in Table 26.2, there are a number of rarer causes including metabolic, neurovascular (e.g. stroke), psychiatric, and other nasal disorders. Patients with cacosmia, especially where localized rhinological symptoms exist, may have localized pathology such as a sinus mycelium (fungal ball) which will be identified on examination and/or radiological investigation.

Treatments

Treatment of olfactory disorders should be guided by the cause. In patients with a conductive disorder, these cases typically respond to medical and/or surgical treatment to modify the blockage of airflow to the olfactory clefts. Monoclonal antibodies in numerous trials have now shown beneficial effects on global CRS symptoms but especially on olfaction; unfortunately at the time of publication they remain unapproved by NICE for CRS in the UK but are in use more routinely internationally. In cases of sensorineural disorders, the treatment options are more limited, largely due to the lack of level 1 evidence to support them. Nonetheless there are several options which have shown some promise in certain cases and can be considered in discussion with patients (Figure 26.4). One simple but effective strategy that can be applied to many causes of olfactory dysfunction is smell training. This appears to have the greatest potential in patients with PIOD, but also in cases of post-traumatic olfactory dysfunction. Modified smell training, where the odours are changed every 3 months, appears to have even greater benefits for recovery of olfactory function. Further guidance to patients can be sought through resources such as SmellTaste, a charity for smell and taste disorders (www.smelltaste.org.uk). Oral steroids are probably unlikely to benefit patients with PIOD unless given soon after the exposure. However, if endoscopy reveals persistence of localized olfactory cleft oedema, using them in conjunction with topical steroids may prove beneficial. Oral steroids are also most likely to be of benefit early after onset of post-traumatic olfactory dysfunction. Other treatments are probably best reserved for the domain of a specialist clinic where experience exists in counselling patients on the potential benefits and side effects of what are usually unlicensed medications.

Tips and pearls

One of the most common complaints from patients with olfactory disorders is that clinicians fail to take the matter seriously. These patients frequently report being dismissed out-of-hand or belittled because they should feel grateful because they have not lost their sight or their hearing. Little attention is also paid to the psychological dimension of their disorder. Imagine yourself for a moment if every meal felt like chewing cardboard and passing a bakery gave you nothing at all. Sexual attraction, memories, emotion, and professional endeavours can all rely on smell. So do consider the wider impact these patients experience in losing their sense of smell.

Sometimes worse is the qualitative disorders, parosmia and phantosmia, as these can be severely disabling, particularly in the

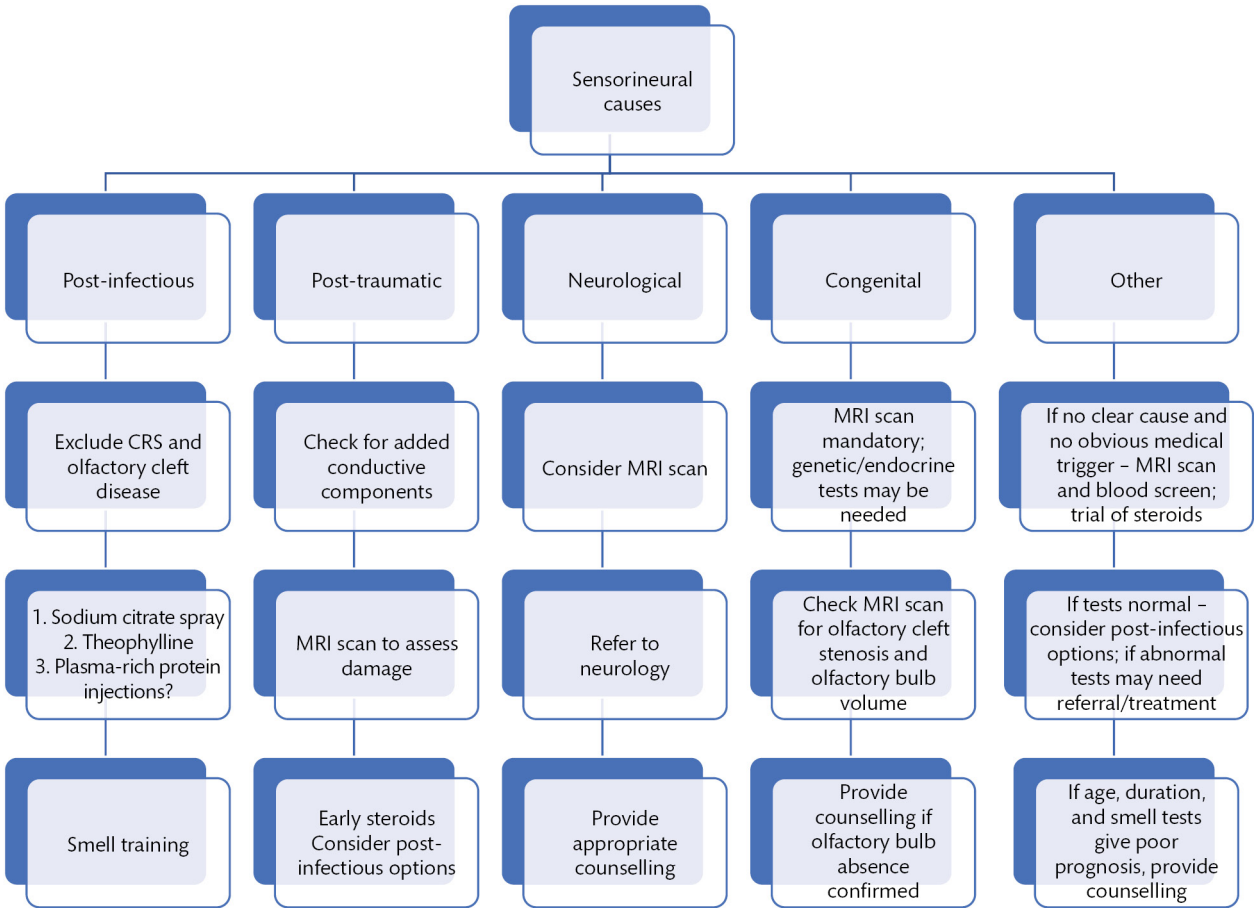

Figure 26.4 Management of sensorineural causes of olfactory disorders.

context of eating. Treatment of the primary disorder may often help alleviate them, but in select cases, antiepileptic agents such as gabapentin may prove useful in moderating these symptoms. Duration of these symptoms can be hugely variable but tend to be self-limiting eventually. Again, acknowledgement and support are key in these cases.

Controversies/areas of interest for the future

As alluded to above due to the paucity of level 1 and 2 evidence, there is a clear need for good-quality randomized controlled trials to be conducted for treatments in olfactory disorders. The lack of evidence beyond case series and pseudo-randomized trials underpins some of the apathy around managing olfactory disorders. There is also a need for greater basic science research into understanding targets for new pharmacological agents and, in some cases, diagnostics for understanding why smell loss has occurred. Despite the global recognition of anosmia being a symptom of COVID-19, it remains a challenging area for research funding. There has been recent interest in plasma rich protein injections to the olfactory cleft and whilst early evidence is promising, more suitably powered and multi-centre studies are needed to confirm this.

Guidelines

In 2023, the Position Paper on Olfactory Dysfunction was produced by the Clinical Olfactory Working Group and is freely available to download on the *Rhinology Journal* website (https://www.rhinologyjournal.com/Documents/Supplements/Supplement_33.pdf). This directs clinicians through the overall recommended assessment of olfactory dysfunction and relevant supporting evidence for existing treatments. Specific guidance around the management of CRS can be found in the EPOS2020: (https://www.rhinologyjournal.com/Documents/Supplements/supplement_29.pdf).

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27

Epistaxis

Rajiv Bhalla and John S. Phillips

Introduction

Epistaxis remains one of the most common causes for emergency access to an ear, nose, and throat service, with around 25,000 acute presentations each year in the UK.[1] Unfortunately, this is likely to be severely underrepresented due to coding anomalies and uncaptured data. While most cases of epistaxis are innocent, some may be the hallmark of significant underlying local or systemic disease and can be life-threatening.[2] Epistaxis presents according to a bimodal distribution with peaks in children and young adults, and the older adult (45–65 years). While triage and subsequent management should be relatively straightforward a wide variation in practice still exists, including clinical inactivity.[3] Although treatment must be individualized, strategies exist that can expedite management. Prompt and effective management of this condition is essential.

Anatomy of epistaxis

The vascular supply to the internal nose is from both the external and internal carotid artery systems. Bleeds are categorized as being either anterior or posterior based on an arbitrary junction at the head of the middle turbinate. About 95% of epistaxis originates from Kiesselbach's plexus in Little's area (**Figure 27.1**).

This is an area at the anterior aspect of the nasal septum where five major arteries terminate: the anterior and posterior ethmoidal arteries are branches of the ophthalmic artery and arise from the internal carotid system; the greater palatine, sphenopalatine, and superior labial arteries arise from the external carotid system. This area is easily visualized and managed with the aid of a headlight and speculum, further details are provided below. The most significant vascular supply to the internal nose is from the sphenopalatine artery. The area of the nose supplied by this artery cannot be visualized and managed with just a headlight, so an endoscope must be used. Approximately 85% of individuals are considered to be 'sphenopalatine artery dominant'; the remaining group are 'anterior ethmoidal artery dominant'. Special consideration must be given to tumour management as the vascular supply can be complex.

Acute management

Patient history, individual circumstances, and the result of a thorough nasal assessment will determine whether epistaxis can be managed easily in an appropriately equipped outpatient setting. The age of the patient, haemodynamic status, underlying anxiety, briskness of bleeding, site of bleeding (anterior or posterior), comorbidities, and clinician experience are all factors that also need to be taken into consideration. The very young may struggle to have their nose examined and treated but if a rapport can be achieved, and adequate decongestion and surface anaesthesia obtained, even this age group can do very well without the need for a general anaesthetic.

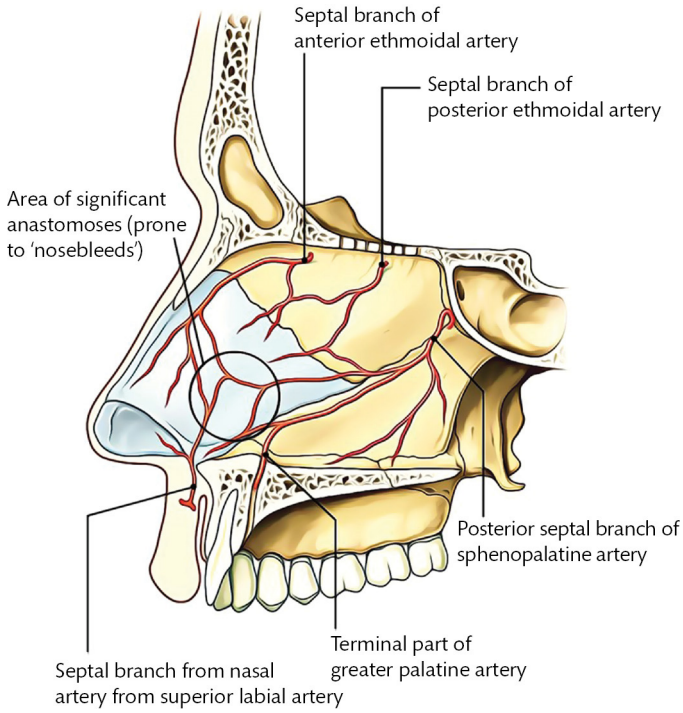

Figure 27.1 Illustration of Little's area.

Box 27.1 Causes of epistaxis

Local

- Idiopathic (85%).
- Trauma (digital, nose blowing, surgical, chronic irritation from foreign body, blunt, penetrating).
- Inflammatory (infection, allergic rhinosinusitis, atrophic rhinitis, autoimmune, nasal polyps).
- Environmental (temperature, humidity, altitude).
- Septal deviation or perforation.
- Neoplasia (benign and malignant).
- Granulomatous disease (granulomatosis with polyangiitis (previously known as Churg–Strauss syndrome), sarcoidosis).
- Environmental (nasal prongs, continuous positive airway pressure).

Systemic

- Coagulopathies.
- Thrombocytopenia.
- Platelet dysfunction.
- Hereditary haemorrhagic telangiectasia.
- Organ failure (liver, uraemia).

The history should be comprehensive and include a discussion regarding relevant associated medical history (hypertension, diabetes mellitus, and ischaemic heart disease), aetiological factors (Box 27.1), and drug history (Box 27.2). A variety of homeopathic treatments may adversely affect coagulation and must be borne in mind. It is useful to document the side and site (anterior or posterior) of bleeding, frequency, severity, and whether there have been prior episodes. One should always aim to exclude a tumour as the cause for unilateral epistaxis.

For initial assessment, begin by sitting the patient comfortably in a slightly forward-leaning position with a bowl if actively bleeding to avoid aspiration of blood. Pressure can be placed below the nasal bones in an effort to stem the flow of blood as a first aid manoeuvre. There is no evidence for the benefit of ice used over the bridge of the nose although the patient may find this soothing (and distracting). Make a rapid ABC assessment and secure intravenous access in the first instance if there is any concern that bleeding has been prolonged and severe. Take blood for a full blood count and clotting screen. You should have a good working headlight, nasal suction, nasal speculum, zero-degree fibreoptic rod, and a surface decongestant with anaesthetic to hand. Co-phenylcaine (lidocaine hydrochloride 5% weight/volume and phenylephrine hydrochloride 0.5% weight/volume topical solution) is a convenient multipurpose topical preparation that is commonly available in the outpatient setting.

Box 27.2 Drugs implicated in epistaxis

- Antiplatelet drugs (aspirin, clopidogrel).
- Anticoagulants (warfarin, heparin).
- Factor Xa inhibitors.
- Topical corticosteroid spray.
- Over-the-counter decongestants (xylometazoline, oxymetazoline).
- Cocaine.
- Natural 'blood thinners' (garlic, ginger, gingko biloba, turmeric, vitamin E).

Small cotton wool balls are a valuable adjunct. An experienced assistant is invaluable when managing epistaxis.

Step 1. Use personal protective equipment for both yourself as well as the patient (apron, gloves, eye protection, and mask) where indicated, especially if the nose is bleeding actively and briskly.

Step 2. Clear the nares and nostrils of dried blood, use suction to carefully debride any clot. While focusing on the side of the bleed, decongest both nasal passages to facilitate a thorough endoscopic examination.

Step 3. Thoroughly examine both nasal passages and the nasopharynx using anterior rhinoscopy and a zero-degree Hopkins rod.

Step 4. For vessels in Little's area, chemical cautery can be achieved using silver nitrate sticks (see below). In very young children this same manoeuvre may need to be performed under a short general anaesthetic. It is important to neutralize the silver nitrate using normal saline irrigation following cauterization to avoid skin burns as a complication of the procedure.

Step 5. For a posterior bleed from the region of the sphenopalatine notch or mucosal vascular anomalies elsewhere in the nose (e.g. a haemangioma), cauterization can be performed using bipolar diathermy (see below) under direct endoscopic visualization and suction.

Step 6. For an uncomplicated anterior bleed successfully treated by chemical cauterization the patient can be discharged immediately with a topical antiseptic/antibiotic cream to use for 14 days with advice to avoid digitally traumatizing the cauterized area for the same duration. Outpatient review 1 month later will confirm mucoperichondrial regeneration and septal integrity.

Step 7. Where the bleed was posterior and likely to have been significant, observing the patient for 6–12 hours before discharge with the same advice as Step 6 above is recommended.

Step 8. Where there is a failure to control the bleeding, the nose should be packed either anteriorly or anteriorly and posteriorly to provide adequate vascular tamponade (see below). Only the side of the bleed should be packed if at all possible, though this would assume there is a good account of which side of the nose was bleeding at presentation.

Step 9. If the nose is packed, admit the patient for observation. The provision of appropriate analgesia and an anxiolytic should be considered. Hypertension should be controlled in collaboration with the appropriate hospital medical team. Where there is ongoing bleeding through the pack in very difficult cases, keep the patient starved and prepared for emergency theatre, allowing clear fluid intake if blood ingestion has not resulted in significant nausea or vomiting; intravenous fluid hydration may be required.

Investigations

In significant cases of epistaxis, a full blood count and a 'group & save' should be performed. Transfusion should be guided by appropriate guidelines (see 'Guidelines'). Renal function should be monitored in patients who have undergone fluid resuscitation. Perform clotting

studies in patients with a suspected clotting diathesis, when the patient is taking anticoagulant medication, or if bleeding is difficult to control.[4]

Differential diagnosis

Box 27.1 lists the most commonly cited causes of epistaxis. The cause of epistaxis in the overwhelming majority of patients is idiopathic in nature. However, in certain groups of patients—such as those with hereditary haemorrhage telangiectasia, haemophilia, or platelet dysfunction—a predisposition to present with epistaxis is relatively common. More sinister causes of epistaxis should be considered in patients who present with unilateral symptoms and signs, including epistaxis, nasal obstruction, paraesthesia, or unilateral nasal polyposis with ipsilateral swelling or extension into the paranasal sinuses or orbit. The following chapters are of particular relevance as patients with these conditions commonly present with epistaxis: Chapter 39 ('Sinonasal tumours') and Chapter 43 ('Hereditary haemorrhagic telangiectasia').

Treatment

Nasal cautery

Nasal cautery is usually carried out using a silver nitrate stick (75% silver nitrate, 35% potassium nitrate BP weight/weight). When applied to the nasal mucosa a local chemical burn is produced. The burn results in the coagulation of superficial blood vessels. It has been recommended that, where available, electrocautery should be used in preference to silver nitrate cautery as it is associated with lower treatment failure and recurrence rates, a reduced requirement for nasal packing and hospital admission, and lower pain scores. This is because a deeper level of coagulation can be achieved.

Nasal packing

Nasal packing can be performed using a variety of materials and there is a wide range of commercially available preformed nasal packs. Traditionally, formal nasal packing involved the insertion of a pack composed of ribbon gauze impregnated with bismuth, iodine, and paraffin paste (BIPP). Modern options for effective anterior packing include rubber-coated sponge tampons such as Merocel or expandable nasal packs such as Rapid Rhino—which are coated in a carboxymethyl cellulose fabric to promote thrombocytic aggregation. Options for effective posterior packing include the placement of a Foley or Brighton balloon catheter. Nasal packing can lead to serious complications—see Box 27.3.

Endoscopic ligation of the sphenopalatine artery

Endoscopic ligation of the sphenopalatine surgery for refractory epistaxis is an effective manner to treat epistaxis due to its low rates of failure and complications, with cauterization being considered to be more effective than ligation.[5] As compared with alternative methods, sphenopalatine artery ligation has been found to be superior in controlling epistaxis in 98% of cases.[6] The surgical steps involved in performing a sphenopalatine ligation are illustrated in Figure 27.2, steps (a) to (d).

The nose is prepared, and the site of the sphenopalatine foramen is anticipated near the posterior attachment of the middle turbinate between the inferior turbinate and the ethmoid sinus. A broad vertical incision in made anterior to this location (a). A mucoperiosteal flap is raised until the sphenopalatine foramen is identified (b). The location of the sphenopalatine foramen is anticipated by its location posterior and slightly inferior to a consistent landmark: the crista ethmoidalis. The sphenopalatine artery may present itself either as a single trunk or as multiple branches. The trunk and/or branches are either clipped and/or diathermized with bipolar diathermy (c). The mucoperiosteal flap is replaced (d).

Considerations for informed consent

Cited potential complications to discuss when consenting a patient for sphenopalatine ligation include recurrence of bleeding, intranasal dryness with crust formation, sinusitis, impaired nasal and palatal sensitivity, adhesion formation, chronic epiphora, septal perforation, and blindness.

Tips and pearls

- A 'medically optimized' patient is more likely to have an uneventful anaesthetic. Furthermore, the surgical field would be clearer and the patient is less likely to endure any significant postoperative complications.
- Ensure that the nose is well decongested before initiating the dissection.
- Consider performing a septoplasty (conventional or endoscopic) to improve access and exposure of the sphenopalatine artery.
- Some surgeons advocate performing an uncinectomy and middle mental antrostomy to aid the anticipation of where the sphenopalatine artery will arise from its foramen.

Ligation of other big blood vessels

Ethmoid arteries

Ligation of the ethmoid arteries is indicated in cases of persistent bleeding as a consequence of nasal trauma. Surgery can be performed using either an open approach via a Lynch incision or endoscopically via a maxillary antrostomy.

Maxillary artery

Before the advent of sphenopalatine artery ligation, maxillary artery ligation represented a valid means to treat recalcitrant epistaxis. This procedure is rarely performed in modern times.

External carotid artery

Like maxillary artery ligation, ligation of the external carotid artery is rarely performed these days. Due to the relative remoteness of the external carotid artery from the nose there is a significant opportunity for rebleeding via collateral vessels. Rebleeding rates of up to 45% have been reported.

Box 27.3 Complications of nasal packing

- Aspiration of the nasal pack.
- Toxic shock syndrome.
- Hypoxia secondary to nasal obstruction.
- Acute sinusitis secondary to blockage of the ostiomeatal complex.
- Hypotension and bradycardia secondary to the nasovagal reflex.

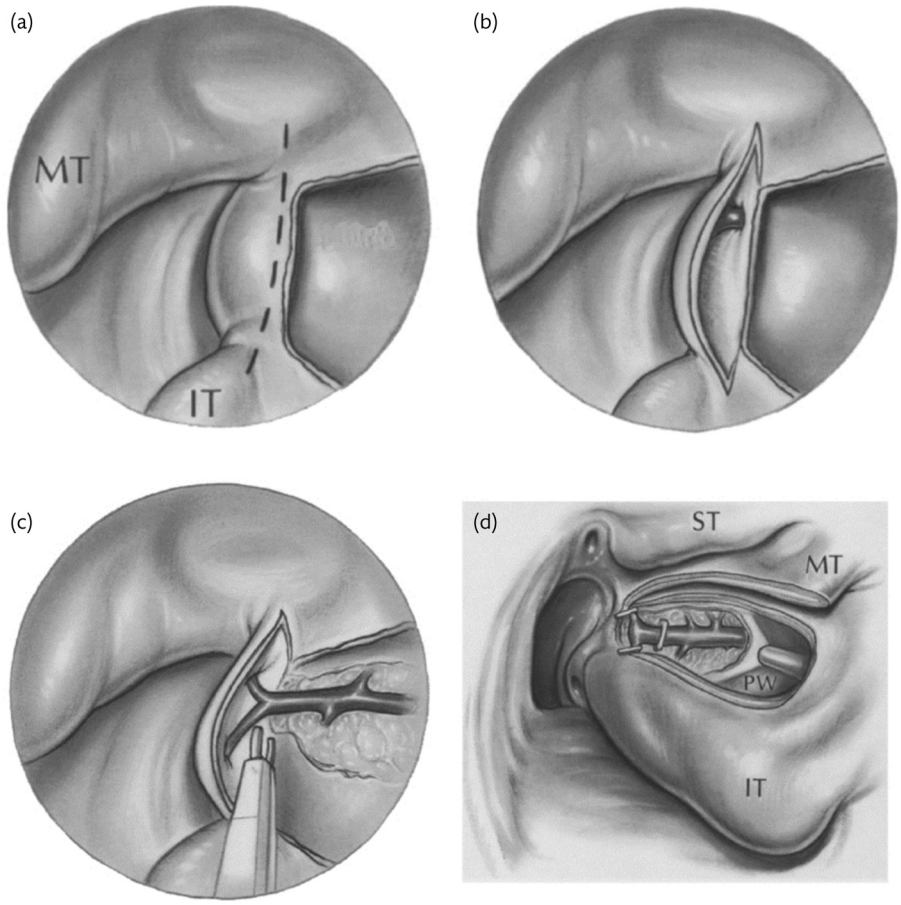

Figure 27.2 Surgical steps for the ligation of the sphenopalatine artery. IT, inferior turbinate; MT, middle turbinate; PW, posterior wall of antrum; ST, superior turbinate.

Controversies

Nasal cautery versus lubrication

There has been some evidence promoted within the literature to suggest that patients with epistaxis gain comparable relief from conservative treatment with a nasal lubricant as compared with the application of silver nitrate cautery.[7]

Ligation of the sphenopalatine artery as a first-line treatment for epistaxis

Due to the expense involved in admitting patients subsequent to nasal packing, some surgeons have advocated the provision of endoscopic ligation of the sphenopalatine artery as a first-line intervention.[8] Ligation of the sphenopalatine artery may be performed under local anaesthesia and in patients taking anticoagulant medications such as warfarin.[9]

Tranexamic acid

Tranexamic acid is an antifibrinolytic drug and its use for the treatment, and prevention, of epistaxis has been carefully considered. In scenarios outside otolaryngology, clinical trials have demonstrated that tranexamic acid has reduced the probability of requiring a blood transfusion by approximately one-third.[10] A review by Kamhieh and colleagues identified five randomized controlled trials of tranexamic acid use for epistaxis and it was determined that there was a trend towards more favourable outcomes with tranexamic acid.[11] Further studies of intravenous or topical tranexamic acid are required.

Floseal

Floseal is a biodegradable matrix haemostatic sealant. It is applied via an applicator that is introduced into the nose. There has been mounting support that Floseal may represent an effective, comfortable, and cost-effective alternative treatment of persistent epistaxis when compared to traditional packing methods.[12]

Prophylactic antibiotics

A range of opinion exists regarding the necessity for antibiotics in individuals with a packed nose. Evidence is lacking but many specialists recommend the provision of antibiotics when an anterior pack has been placed for more than 48 hours. All individuals with a posterior pack should receive antibiotics. Preferred antibiotics include amoxicillin–clavulanic acid, amoxicillin alone, or cephalosporins.[13]

Duration of nasal packing

The literature provides a wide range of values for the recommended duration a nasal pack should remain *in situ*, from 12 or 24 hours, to 3–5 days after placement.[14] Nasopharyngeal balloons should be deflated after 24 hours.

Box 27.4 Scottish Intercollegiate Guidelines Network: guidelines for the management of antithrombotic drugs (SIGN 129)

- *Serious bleeding with international normalized ratio (INR) greater than 1.1*: stop warfarin, and administer intravenous vitamin K (5–10 mg) and prothrombin complex concentrate (usually 30– 50 IU/kg, but dose-adjusted according to INR (under haematologist supervision whenever possible)). Fresh frozen plasma (at least 15 mL/kg) may be used only if prothrombin complex concentrate is unavailable.
- *Minor bleeding and supratherapeutic INR*: interrupt warfarin, reintroducing at a lower maintenance dose when situation is under control. Administer oral or intravenous vitamin K (1.0–2.5 mg).
- *No bleeding and supratherapeutic INR*: interrupt warfarin, monitor INR, restart warfarin at lower dose when INR less than 5.0. Where perceived risk of bleeding is high (e.g. INR >8) or other risk factors for bleeding are present, consider oral vitamin K administration (1.0–2.5 mg).

From the British Rhinological Society multidisciplinary consensus recommendations.

Box 27.5 British Committee for Standards in Haematology guidelines

Major haemorrhage is defined as bleeding leading to a heart rate of greater than 110 bpm and/or systolic blood pressure less than 90 mmHg.

- Hospitals must have local major haemorrhage protocols with adaptations for specific clinical areas. Such protocols should be followed when appropriate.
- Optimum target haemoglobin concentration in bleeding management is not established. However, updated European guidelines recommend a target haemoglobin level of 70–90 g/L. Patients with cardiorespiratory morbidity may require a higher target of 80–90 g/L.
- Fresh frozen plasma should be part of initial resuscitation in major haemorrhage in at least a 1:2 unit ratio with red cells until coagulation monitoring results are available. Once bleeding is under control, further fresh frozen plasma should be guided by laboratory result abnormalities, with transfusion trigger of prothrombin time and/or activated partial thromboplastin time greater than 1.5× normal level for a standard dose (e.g. 15–20 mg/kg).
- Fibrinogen supplementation should be given if fibrinogen levels decrease to less than 1.5 g/L.

From the British Rhinological Society multidisciplinary consensus recommendations.

Radiological embolization

Endovascular embolization for patients with epistaxis is considered by some to be an alternative to ligating vessels within the head and neck. Selective arterial embolization can be an attractive option for the management of severe posterior epistaxis in certain individuals with success rates between 87% and 93%.[14] However, the overall short-term and long-term rebleed rates are often higher in those patients who are ultimately selected for these types of procedure.[15] While the procedure can be performed under local anaesthesia, the risk of cerebrovascular accident of around 4% should not be underappreciated.

Guidelines

The American Academy of Otolaryngology–Head and Neck Surgery has produced guidelines for the management of epistaxis.[16] National guidelines have been proposed in the UK by ENT UK based on systematic review, national audit of practice, and multidisciplinary consensus recommendations.[17]

Two other related guidelines are worthy of consideration when managing epistaxis. The first is the Scottish Intercollegiate Guidelines Network guidelines (SIGN 129)[18] for the management of antithrombotic drugs (these guidelines have been reproduced in Box 27.4). The second is the British Committee for Standards in Haematology guidelines which among other things provide advice on managing major haemorrhage[19]—see Box 27.5.

Conclusion

The management of epistaxis has developed significantly over the past decade with respect to the materials employed to pack the nose, the available surgical techniques, and the management of coexisting medical conditions. National guidelines are emerging in a number of countries to allow a consistent and evidence-based approach to this common condition. Unfortunately, a reciprocal reduction in hospital admissions has not been observed, and this is likely to reflect an ever-increasing elderly population with more complex comorbidities necessitating the administration of newer anticoagulant formulations.

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28

Nasal septal perforation

Tim Woolford

Introduction

A nasal septal perforation is a hole through all layers of the nasal septum. The perforation can be of variable size, ranging from a tiny pinhole to a very large defect where all the septal cartilage and much of the bone is absent. The incidence of this condition is rather unclear; however, an otolaryngologist with a general practice will encounter these cases throughout their career and a specialist rhinologist will see several new cases each year.

There are many causes of a septal perforation as detailed below. However, the majority of those requiring surgical closure are a complication of septal surgery and most of these are preventable with meticulous technique. Prevention is therefore an ongoing issue for otolaryngology.

Although some septal perforations are asymptomatic, most cause symptoms which adversely affect a patient's quality of life. Conservative treatment to improve nasal hygiene will reduce, but not eliminate, these symptoms. Obturators remain an option, and recent advances have made these more acceptable to patients than the traditional silastic septal button. Many patients do not tolerate an obturator and wish to pursue the option of surgical closure.

For many years surgical closure has involved a technique using local mucosal advancement flaps covering various types of graft in the septal defect. This surgery is challenging and although smaller perforations can often be closed successfully, there is a consensus view that larger perforations greater than 2 cm are very difficult to close. In recent years, experience in endoscopic anterior skull base surgery has provided expertise in pedicled mucosal flaps based on the anterior ethmoidal artery. These sizeable flaps have been rotated to close large septal perforations previously not realistically amenable to surgical closure. It seems likely that these techniques will in time become widely adopted, with great benefit to patients.

Aetiology

There are many conditions which can cause septal perforation (**Box 28.1**), although in reality only a few of these are seen commonly in clinical practice. In many cases the cause will be clear from the outset, particularly if the patient has undergone previous septal surgery. The history taken should therefore cover any nasal surgery even if it took place a number of years previously as patients may not recall specifically having undergone septal surgery. Other nasal treatment such as cautery and nasal packing should be noted.

Patients should be asked about recreational drug use such as cocaine and this should be considered in any age group, and not limited to younger patients. Any symptoms suggestive of granulomatous or vasculitic disease should be noted, although this diagnosis is usually self-evident. Very rarely malignancy presents with a septal perforation, although these lesions generally present with localized ulceration and tissue destruction rather than a perforation.

Rarely, patients will candidly admit that they have picked their nose for years, and the perforation is self-inflicted (see Chapter 34).

Box 28.1 Aetiology of nasal septal perforations

- Septal trauma:
  - Septal surgery.
  - Digital trauma.
  - Nasal packing.
  - Septal haematoma/abscess.
- Chemical injury:
  - Cocaine.
  - Silver nitrate cauterization.
  - Decongestants/steroids.
  - Industrial chemical exposure (e.g. phosphorus, mercury).
- Infections:
  - Syphilis.
  - Tuberculosis.
  - Rhinoscleroma.
  - Actinomycosis.
- Granulomatous and vasculitic conditions:
  - Granulomatosis with polyangiitis.
  - Sarcoidosis.
  - Relapsing polychondritis.
- Malignancy:
  - Squamous cell carcinoma.
  - Adenocarcinoma.
  - Malignant melanoma.
  - Lymphoma.

Box 28.2 Symptoms caused by a nasal septal perforation

- Congestion and obstruction.
- Crusting.
- Dryness.
- Discomfort.
- Bleeding.

In many other cases the cause of the septal perforation is unclear, and the condition is considered 'idiopathic'. Iatrogenic perforations caused by surgery and the idiopathic group cover the majority of symptomatic cases in the author's experience.

Septal perforation symptoms

Septal perforations may be asymptomatic, particularly if they are small and situated posteriorly. It is not uncommon to identify an asymptomatic posterior perforation on endoscopy in a patient who has undergone a previous septoplasty.

The symptoms related to a septal perforation are many and varied (Box 28.2). Typically, anterior and larger perforations cause more symptoms, although small anterior perforations can whistle—a symptom which patients find particularly distressing. Larger perforations commonly cause a sensation of nasal obstruction and congestion due to disruption of airflow. Crusting and discomfort are common, as is bleeding when the crusts are cleared. More significant epistaxis is rare with anterior perforations, whereas posterior perforations affecting the bony-cartilaginous junction may cause challenging epistaxis with a small cohort of patients requiring multiple hospital admissions.

Significant pain, combined with septal ulceration, tissue destruction, and a septal perforation should alert the clinician of a possible diagnosis of malignancy. These cases are rare, and urgent action is required.

Investigations

Investigation is required if there is a clinical suspicion that a vasculitic or granulomatous condition such as granulomatosis with polyangiitis or sarcoidosis is implicated, and initially these should be the blood tests. These investigations are covered in detail in Chapter 34.

The indication for a septal biopsy has evolved considerably in recent years. In a condition such as granulomatosis with polyangiitis the diagnosis is usually already clear or confirmed with appropriate blood tests. A biopsy is very rarely helpful in this situation in the author's experience. If there is a clear history of septal surgery causing the perforation a biopsy is also not required. Although traditional teaching called for a biopsy for all other cases of septal perforation where the diagnosis was unclear, it is now accepted that this is unnecessary. This is certainly the case with healthy mucosa. If the perforation is considered 'idiopathic', it is highly unlikely that a biopsy will be helpful.

Any suspicion of malignancy, with ulceration and tissue destruction associated with a perforation requires an urgent diagnostic biopsy followed by appropriate multidisciplinary referral.

Conservative management

A nasal hygiene regimen using a combination of lubrication and moisturization will improve the symptoms caused by a septal perforation in the majority of patients, although this will of course depend upon compliance. The products recommended vary greatly from specialist to specialist and patients often arrive at a regimen which they find most effective after trial and error. Lubricants include petroleum jelly or water-based lubricants, with moisturization or douching provided by proprietary saline sprays or higher-volume saline rinses.

It is the author's experience that a daily regimen using a combination of water-based lubricants and a saline spray is particularly helpful, being both effective and well tolerated with good compliance. It is impossible to predict how successful these various regimens will be for a particular patient. Rarely, a troublesome perforation will become asymptomatic; unfortunately, in most patients the symptoms will be reduced but not completely eliminated even with good compliance.

Obturators

There is a range of opinion regarding the use of obturators. Traditionally, many patients with smaller perforations were advised to try a silastic septal button, and a 'trial' of a button was often a condition before proceeding to surgical perforation repair. This practice is no longer commonly adopted and many rhinologists never recommend obturators.

Although in some patients a septal button does reduce symptoms, particularly their crusting, in others the button causes irritation with increased obstruction. This results in a number of patients requesting removal. The observation that a septal button can increase the size of a perforation is another reason many rhinologists have moved away from using this intervention.

In recent years a number of more sophisticated obturators have been introduced, but with limited evidence as to their effectiveness and advantage over a silastic button. These include a thin magnetic device and fitted, soft, bespoke obturators.

Surgical treatment of septal perforations

Enlarging the perforation

Surgically enlarging a small perforation was advocated in the past to reduce the symptom of whistling, or to enable a septal button to be fitted. This is no longer considered to be appropriate because a perforation of this size can usually be successfully closed using the techniques described below.

One rare situation where enlarging the perforation may have a role is when crusting and bleeding arise from the posterior edge of a sizable perforation not amenable to closure. Usually the bleeding is from the wide bony-cartilaginous junction and can be difficult to control. The perforation is enlarged by removing cartilage and bone from its posterior edge while preserving mucosa to leave sufficiently sized flaps. These mucosal flaps can then be closed over the exposed posterior edge of the perforation, reducing bleeding and crusting.

Surgical closure of septal perforations

Numerous surgical techniques for septal perforation closure have been described over many years, confirming that there is no ideal technique. This is challenging surgery even in experienced hands.

Closure using mucosal advancement and grafts

Until recently, the majority of surgeons adopted an approach involving mobilization of mucosa surrounding the perforation. This allowed a superior and inferior flap to be advanced over the perforation, usually covering a graft in the cartilaginous defect. In order to allow a tension-free closure of mucosa, extensive mobilization is required with the superior flap providing most of the coverage (Figure 28.1).

Given that many cases will have undergone previous septal surgery, mobilization of mucosal flaps in a sub-perichondrial plane is usually difficult and time-consuming due to scarring. In these iatrogenic cases the mucosal perforation can be considerably smaller than the cartilaginous defect, with the size of the perforation and the complexity of the case increasing dramatically as the case progresses.

Surgery can be approached via an endonasal or open rhinoplastic approach. Most surgeons use an open approach for larger perforations to allow greater access, and many perform all of these cases via an open approach. An initial endoscopic elevation of the inferior flap is often helpful.

There is a range of opinion regarding the size of perforation which can be closed using these techniques, with claims of successful closure of very large perforations. There is, however, a consensus that perforations greater than 2 cm in any diameter and those situated on the floor of the septum are very difficult to close. Although partial closure may improve a patient's symptoms, particularly if the anterior aspect of the perforation is closed, this is by no means guaranteed. Symptoms can also deteriorate if the surgery is not completely successful. In view of this, subjecting a patient to a complex procedure with other associated risks does not seem reasonable unless there is a realistic prospect of success.

Endoscopic pedicled mucosal rotation flaps

In recent years there has been a significant advance in the closure of septal perforations using endoscopically raised pedicled vascular flaps based on the anterior ethmoid artery. As experience in endoscopic anterior skull-based surgery has increased, otolaryngologists specializing in this field have become familiar with these sizeable flaps designed to close skull base defects. By rotating the mucosal flap inferiorly, and often in combination with free grafts such as a collagen matrix graft, they can be used to successfully close large perforations (Figure 28.2). This technique is now being successfully employed in cases where there was little realistic prospect of closure using the mucosal advancement techniques described above.

With increasing experience and based on the reported high success rates of this technique, it seems likely that in future the majority of septal perforations will be closed using endoscopic pedicled mucosal rotation flaps rather than mucosal advancement techniques, representing a major advance in this field.

Avoidance of septal perforations

Clinical experience confirms that septal perforations often cause our patients unpleasant symptoms. It is not uncommon to see a patient who described minimal symptoms prior to their septoplasty which resulted in a septal perforation, subsequently causing a major adverse impact on their quality of life.

It is therefore of paramount importance to avoid septal perforations during septal surgery and this should be a very rare complication. Mucosal tears do of course occur during septal surgery and should be repaired if possible. Often this is technically difficult, and the key to reducing the risk of perforations is to avoid wide areas of mucosa-to-mucosa contact unsupported by cartilage. If deviated

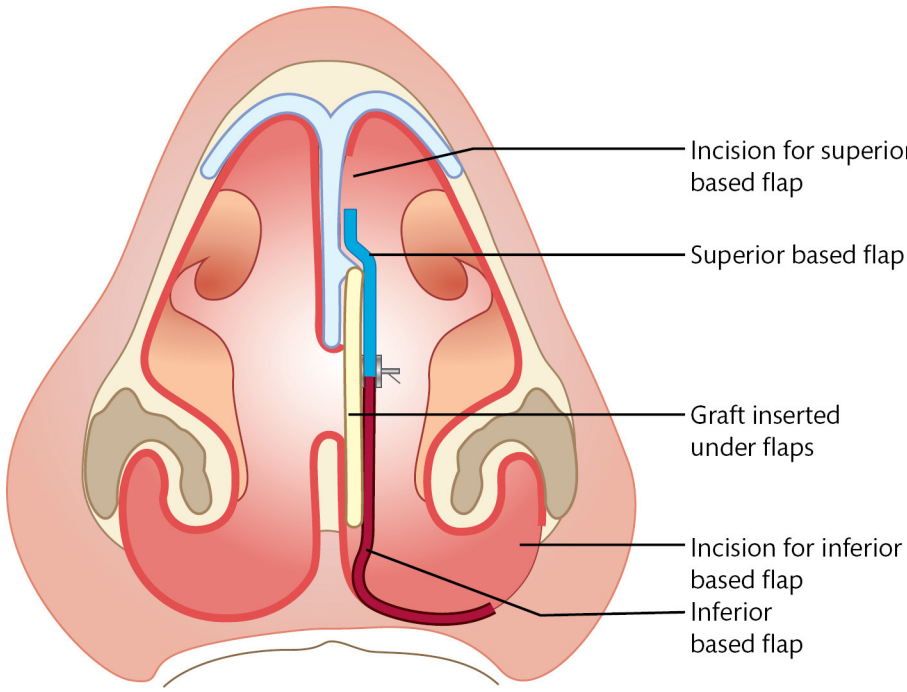

Figure 28.1 Mucosal rotation flaps and cartilage graft to repair a septal perforation.

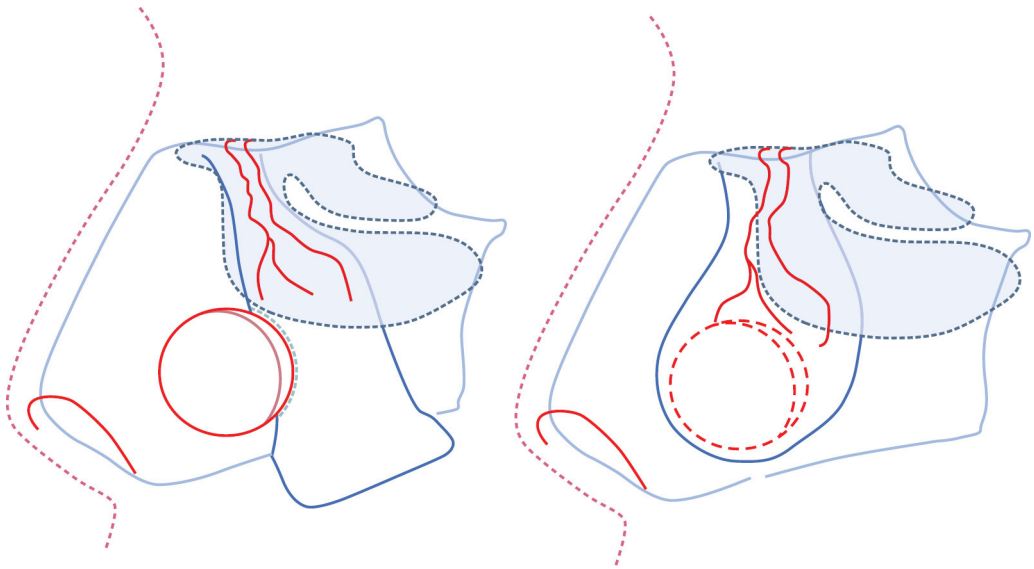

Figure 28.2 Pedicled anterior ethmoid artery flap in nasal septal perforation repair.

or fractured cartilage has to be removed during a septoplasty it should be carefully crushed and replaced to support the mucosa rather than discarded. The submucosal resection approach to the deviated septum, only preserving a dorsal and caudal strut, is now an outdated operation which should be resigned to history in the view of the author. When cartilage is harvested from the septum to use as grafts, large areas of unsupported mucosal contact should be avoided, with small pieces of bridging cartilage carefully placed to reduce the risk of a subsequent perforation.

Nasal cautery using silver nitrate is another recognized cause of a septal perforation and should not be performed concurrently bilaterally. This is particularly important for a child's delicate septum, when any cautery should be unilateral and superficial. When patients are seen with crusting of the septum and exposed cartilage due to digital trauma, they must be warned in stark terms that they are at risk of progressing to a septal perforation. Likewise, with recreational drug use, it must be impressed upon a patient that they risk developing a perforation or greater destruction of nasal tissue with continued use. With levamisole-potentiated cocaine now in widespread use, single episodes of drug use can result in a septal perforation.

In patients with granulomatous or vasculitic disease, a septal biopsy has to be deep into the submucosa to obtain a diagnosis and is rarely helpful. In cases of active granulomatosis with polyangiitis, a biopsy may result in rapid progression to a septal perforation. Diagnosis should be made using blood tests if possible. See also Chapter 33.

Conclusion

Nasal septal perforations remain a common condition, often causing distressing symptoms for patients. Many are caused by septal surgery, and techniques should be adopted in contemporary rhinological practice to minimize this complication. Although a diagnostic biopsy may be indicated in cases of septal perforation, in reality this is rarely helpful and is not needed if the mucosa is normal.

Surgery to close septal perforations using mucosal advancement techniques is challenging even in experienced hands. In recent years an advance in technique has taken place based on large pedicled mucosal rotation flaps used in endoscopic anterior skull base surgery. This technique has proved successful in closing sizeable perforations which would previously have been almost impossible to close. It is likely that with increased experience this technique will be adopted in the majority of cases.

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29

Rhinorrhoea

Amin R. Javer

Introduction

Rhinorrhoea, a medical term for runny nose or excessive thin, watery, clear mucus secretions from the nose, originates from the Greek words 'rhinos' meaning 'of the nose' and 'rhoia' meaning 'a flowing'. It is estimated that the problem affects over 1.4 billion people worldwide, including over 60 million Americans, either in a chronic form or in a recurrent seasonal manner. For the rhinologist, it is imperative to distinguish unilateral rhinorrhoea from bilateral runny nose, and to especially consider the possibility of a cerebrospinal fluid (CSF) leak. The potential differential diagnosis for bilateral rhinorrhoea can be divided into inflammatory versus non-inflammatory causes while for unilateral rhinorrhoea it can be divided into CSF leak versus non-CSF leak (Table 29.1). Inflammatory rhinorrhoea can be further divided into infectious versus non-infectious rhinorrhoea. Unilateral rhinorrhoea is generally considered more ominous with the diagnosis of exclusion being a CSF leak. Other potential diagnoses for a unilateral rhinorrhoea include a foreign body in the nose or other sinister unilateral pathology such as choanal atresia or a sinonasal tumour. The first steps in differentiating between the causes of rhinorrhoea require good history taking, an excellent endoscopic examination technique, and appropriate testing. While not life-threatening in most instances, it is important to be able to come up with the correct diagnosis and start treatment without wasting time.

Bilateral rhinorrhoea: non-infectious

Allergic rhinitis

Allergy is classically considered to arise from an immunoglobulin (Ig)-E-mediated response associated with nasal inflammation, although it is important to remember that not all people with an elevated IgE level have environmental allergies. Allergies affect 28% of the European population with women (31.6%) being significantly more affected than men (24.5%). Younger and mid-aged adults report allergies much more often than elderly people. Appropriate history taking can provide significant information for the astute physician to categorize bilateral rhinorrhoea into its differential components. The presence of an allergy with its associated symptoms can be fairly classic and easy to recognize with good history taking. The patient usually presents with complaints of sneezing, watery rhinorrhoea, nasal and ocular pruritus, and watery eyes. Also important is a history of seasonal versus perennial behaviour where the presence of geographical factors and pets that set off symptoms are all important criteria to provide an adequate diagnosis. The classification of allergic rhinitis as per the Allergic Rhinitis and its Impact on Asthma (ARIA) guidelines uses symptoms and quality-of-life parameters, and is based on duration (subdivided into 'intermittent' or 'persistent') and severity (subdivided into 'mild' or 'moderate–severe') depending on symptoms and quality of life. It is considered moderate–severe if the symptoms impair daily activities, cause problems at work or school, and create sleep difficulties (Figure 29.1).

Table 29.1 Differential diagnosis for rhinorrhoea

Bilateral rhinorrhoea			Unilateral rhinorrhoea	
Inflammatory		Non-inflammatory	No CSF leak	CSF leak
Non-infectious	Infectious	Non-allergic rhinitis (vasomotor rhinitis)	Foreign body Tumour Choanal atresia Other	Traumatic Spontaneous
Allergic rhinitis: • Seasonal • Perennial	Viral (upper respiratory tract infection)			
Non-allergic rhinitis with eosinophilia (NARES)	Bacterial (acute bacterial rhinosinusitis/ chronic rhinosinusitis)			

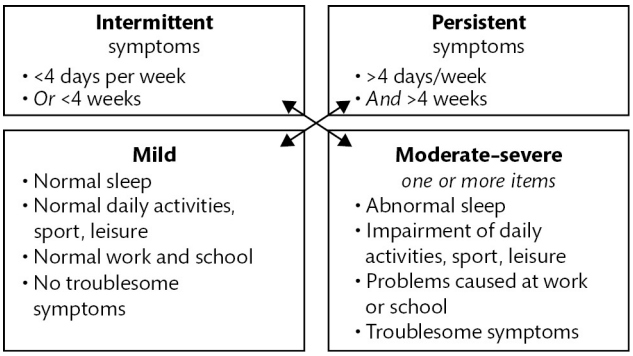

Figure 29.1 Classification of allergic rhinitis as per ARIA guidelines.

Triggers of allergic rhinitis can include aeroallergens such as mites, domestic animals, pollens, and moulds. Occupational rhinitis, latex allergy, and pollutants including indoor air pollution and tobacco smoke, as well as urban-type pollution primarily of automobile origin can often be responsible for causing rhinitis. Aspirin and other non-steroidal anti-inflammatory drugs can also induce rhinitis. Seasonal allergies to pollen and grass or tree allergens occur in the spring and summer months while rhinorrhoea secondary to dust mites is worse in the winter months. Allergy to animal hair and dander is very classically associated with exposure. Perennial symptoms that are worse in the winter months are often secondary to dust mite exposure when the allergen-containing dust is stirred up by heating systems despite the fact that the allergen levels are highest between May and October, which is the peak breeding season of house dust mites. Dust mites flourish in warm, damp environments, temperatures greater than 25°C, and humidity of 70–80%, and feed on dead skin shed from humans and animals. Their presence in the bedroom will often cause symptoms that are worse at night or in the morning. Allergy testing such as skin prick tests to local allergens is easily available and can provide a quick and ready answer for the clinician. Blood tests such as the radioallergosorbent serum test for specific IgE antibodies are more expensive and not justified in most instances, and may not give any more information than the skin prick tests. Imaging is not necessary for classic presentation of allergic rhinitis.

Treatment

Treatment strategies should include educating the patient with regard to the active allergens and avoidance of the inciting allergen, as well as using topical nasal steroids and oral or topical antihistamines. For dust mite allergies, reducing the number of dust mites in the environment, especially with appropriate washing and care of the bedding (wash in hot water twice per week), avoidance of carpets in the bedroom, having a high-efficiency particulate air filter, and not allowing pets in the bedroom can all be very helpful in reducing symptoms. Keeping the bedroom windows shut is very helpful during the seasonal allergy period. For patients with pet hair allergy, avoiding contact is an important therapeutic strategy.

When allergen avoidance by itself is not possible or ineffective, medical management includes intranasal corticosteroids, intranasal H_1 blocker, a combination of the two (Dymista or Ryaltris), and oral H_1 blockers. For patients with ocular symptoms, an intraocular H_1 blocker or intraocular chromone may be added. Treatment with specific immunotherapy, whether by the subcutaneous route or the sublingual route, may be a good treatment option for recalcitrant cases. Surgery may be used as an adjunctive intervention in a few highly selected patients and includes procedures such as coblation or reduction of the inferior turbinates and septal bodies, as well as posterior nasal nerve resection or cryotherapy which is discussed further later in this chapter. Oral corticosteroids and intramuscular or intranasal injection of glucocorticosteroids are not recommended due to the possible occurrence of severe side effects.

Non-allergic rhinitis

Non-allergic rhinitis (previously vasomotor rhinitis) patients can present in an identical manner with very similar symptoms of sneezing, itchy, congested nasal passages, itchy eyes, and cough. It is defined as rhinitis without an identifiable aetiology and without an IgE-dependent component. Allergy skin tests are negative in this group of patients. All forms are associated with some form of autonomic dysregulation with an increase in cholinergic glandular secretory activity, and can be subdivided into various different subtypes such as drug induced, gustatory, hormonal (rhinitis of pregnancy), senile, occupational, vasomotor, emotional, and non-allergic rhinitis with eosinophilia syndrome (NARES). The symptoms can be excessive at times and often exacerbated by certain odours (e.g. perfumes, cigarette smoke), alcohol, spicy foods, emotions, and environmental factors such as changes in temperature, barometric pressure, and bright lights.

Non-allergic rhinitis with eosinophilia syndrome

Eosinophilia in the nasal secretions or in the blood is expected in the allergic patient but not in the non-allergic rhinitis patient. However, a subgroup of patients has NARES that accounts for 10–14% of rhinitis patients and is defined by nasal hyper-reactivity, absence of atopy, and eosinophilia of nasal secretions 20% greater than the leucocytes. This group of patients often present with accompanying hyposmia and fail to respond to antihistamines and topical corticosteroid sprays. There is some evidence to show that NARES may be a precursor to the triad of nasal polyposis, intrinsic asthma, and intolerance to aspirin.

Treatment

Medical treatment is generally favoured as the primary mode of management for the non-allergic rhinitis patient. It is limited to topical modalities including topical intranasal corticosteroids, topical antihistamines, or topical anticholinergics. If the predominant symptom is rhinorrhoea with sneezing and obstruction, a topical intranasal corticosteroid may be tried as a first-line agent with or without a topical antihistamine. Fluticasone together with azelastine is now available as a combination topical treatment (Dymista), as is olapatadine and mometasone (Ryaltris), and may work well in this group of patients. However, if the predominant symptom is rhinorrhoea, then the topical drug of choice should be a topical anticholinergic such as ipratropium bromide (Atrovent) given in a three-times-daily dosing schedule. Oral antihistamines, oral steroids, and topical decongestants have no place in the management of non-allergic rhinitis.

While allergic and non-allergic rhinitis is conventionally perceived to be an illness primarily treated medically, surgical or procedural therapies have evolved over the past five decades. They include inferior turbinate reduction with or without coblation of the septal bodies, botulinum toxin injection, vidian neurectomy,

posterior nasal neurectomy, and, most recently, cryoablation of the posterior nasal nerve. The vidian nerve supplies most of the parasympathetic innervation to the secretory nasal mucosa via the preganglionic parasympathetic fibres of the greater petrosal nerve, which synapses at the pterygopalatine ganglion, giving rise to the postganglionic innervation of the nasal mucosa via the posterior nasal nerves. Vidian neurectomy has therefore been the primary surgical modality of choice with demonstrated efficacy for vasomotor rhinitis. Vidian neurectomy may have beneficial effects for rhinitis, but can be complicated by persistent dry eye symptoms from collateral damage of the parasympathetic innervation of the lacrimal gland partially arising from the vidian nerve. Therefore, newer techniques have favoured selective resection or ablation of the posterior nasal nerves, whose postganglionic parasympathetic fibres are anatomically distal to the lacrimal innervation branch point. This allows for obliteration of the parasympathetic supply to the mucosal membrane and provides the same benefit as vidian neurectomy without any of its complications. Endoscopic posterior nasal neurectomy has therefore gained favour more recently as the procedure of choice for recalcitrant allergic and non-allergic rhinitis when the symptoms are severe and refractory to maximal medical therapy. It is considered a safe and less invasive procedure that can provide significant relief in symptoms for patients with intractable rhinitis with fewer complications than endoscopic vidian neurectomy.

Bilateral rhinorrhoea: infectious

Viral rhinorrhoea (upper respiratory tract infection)

The common cold presents with significant bilateral rhinorrhoea that is often short-lived through the duration of the viral illness and resolves spontaneously. The pathogens most commonly responsible are rhinoviruses, coronaviruses, and the respiratory syncytial virus. An acute inflammatory response occurs and is characterized by the release of a variety of inflammatory cytokines and infiltration of the mucosa by inflammatory cells. There is agreement across most guidelines that the common cold lasts anywhere between 5 and 10 days and resolves without treatment. Topical intranasal corticosteroids and topical ipratropium bromide do produce a small but non-significant decrease in rhinorrhoea and congestion in these patients but evidence for their use is poor. Watchful waiting with adequate hydration is the recommended treatment of choice with most symptoms resolving within 10 days. No radiographic or other testing is recommended.

Bacterial rhinorrhoea

Bacterial rhinorrhoea is almost always mucopurulent where the nasal discharge is coloured, thick, and opaque. Acute bacterial rhinosinusitis (ABRS) is almost always a manifestation of self-limited, uncomplicated viral upper respiratory tract infections. If a common cold or upper respiratory tract infection does not resolve within 5–10 days and progresses from thin, clear rhinorrhoea to a coloured, thick discharge with worsening symptoms, it is highly suggestive of ABRS. In most cases ABRS will resolve with saline irrigations and intranasal topical steroid sprays, but in a small minority it will not resolve and continue beyond 12 weeks, at which point it is considered a chronic rhinosinusitis. In cases of ABRS with intractable symptoms despite saline irrigations and intranasal topical steroid therapy, a course of antibiotics may be trialled with substantial improvement expected within 48–72 hours.

For children with mucopurulent rhinorrhoea that does not resolve with medical management, underlying conditions such as adenoiditis, cystic fibrosis, ciliary dyskinesia, polyps, tumour, neoplasm, or congenital nasal anomalies (choanal atresia or stenosis) should be entertained in the differential diagnosis, and appropriate imaging and/or other appropriate studies (e.g. sweat chloride testing for cystic fibrosis) undertaken. For further descriptions see Chapters 77 and 78.

Chronic rhinosinusitis with or without polyposis can present with chronic, long-standing bilateral rhinorrhoea secondary to chronic inflammation of the sinus and nasal mucus membranes. In the majority of cases, the rhinorrhoea is mucopurulent with other associated symptoms such as postnasal drainage, facial pain or pressure, nasal obstruction, and hyposmia or anosmia. Medical management includes intranasal topical steroid sprays, nasal irrigations, and long-term oral antibiotics as well as oral corticosteroids. Failure of adequate medical management in this group of patients is high, and most symptomatic patients will require endoscopic sinus surgery for resolution of their symptoms.

Unilateral rhinorrhoea

Presentation of a chronic clear, watery discharge from one nostril is indicative of underlying pathology that necessitates further investigation and imaging. A good history followed by a meticulous endoscopic examination of the nose and nasopharyngeal region is mandatory to identify potential causes of unilateral rhinorrhoea. Unilateral rhinorrhoea can be caused by unilateral pathology such as choanal atresia or stenosis which rarely presents in adults. Other causes include unilateral tumours and an embedded foreign body either from previous sinus or dental surgery or from a traumatic event. Unilateral rhinorrhoea can also occur from unilateral cluster headaches and it is therefore imperative to obtain a headache history. A history of illicit drug use is also important as drug withdrawal can present with rhinorrhoea. It is essential for the rhinologist to be cognizant of the fact that unilateral rhinorrhoea is often the result of a CSF leak which requires immediate attention. It is further discussed below in some detail.

Imaging studies, including a computed tomography (CT) scan and often a magnetic resonance imaging scan is necessary to discern the potential aetiological cause of the leakage. In non-CSF leaks, the presence of a foreign body in the nose or sinuses or an obstructive aetiology such as a mass or tumour is often easily visualized on a CT scan. In the child with unilateral rhinorrhoea the clinician must entertain the diagnosis of choanal atresia or embedded foreign body as a possibility. Choanal atresia is rarely seen in adults, but on rare occasion, cases have presented in the adult age group. Further imaging with magnetic resonance imaging may be necessary in appropriate cases.

Cerebrospinal fluid rhinorrhoea

CSF rhinorrhoea is a condition that results from an abnormal communication between the central nervous system, specifically the subarachnoid space and the sinonasal cavity. It is a serious and potentially fatal condition that can still on occasion cause

a major challenge in its diagnosis and management. If left untreated it may lead to ascending meningitis that develops in approximately 10–25% of patients, and up to 10% of them can die as a result.

The cause of CSF rhinorrhoea can be divided into traumatic versus non-traumatic, or spontaneous. Traumatic aetiologies are much more common and account for 80% of all cases. They are usually caused by head injuries with approximately 15% occurring iatrogenically. Localized haemorrhage may seal the defect site, and in rare instances patients can present months or years post the traumatic event, after atrophy of granulation tissue or bony fragments that eventually erode the dura. It is therefore essential to take a full and detailed history for prior trauma or surgical procedures in this group of patients.

Non-traumatic CSF leaks account for approximately 10% of CSF rhinorrhoea cases and can be challenging for the clinician to diagnose correctly. Non-traumatic CSF leaks are further subdivided into normal-pressure and high-pressure CSF leaks. Examples of high-pressure leaks include intracranial space-occupying lesions, hydrocephalus, and idiopathic intracranial hypertension. The causes of normal-pressure CSF leaks may be infections, encephalocele, cholesteatoma, arachnoid granulations, and empty sella syndrome. The latter was traditionally considered an idiopathic cause, but more recently has been shown to be associated with elevated intracranial pressure. Spontaneous CSF rhinorrhoea comprises 14–46% of all cases of non-traumatic CSF rhinorrhoea and carries the highest recurrence rates ranging from 25% to 87% of all causes. This is most likely related to its aetiology, believed to be persistent or intermittent elevated intracranial pressure, and is often seen in middle-aged females with a high body mass index.

CSF rhinorrhoea diagnosis

Reliable diagnosis of CSF rhinorrhoea in cases of suspicious nasal secretion is the cornerstone and first step in proper management. This is often not easy, especially in cases of subclinical CSF leak. Diagnosis of CSF rhinorrhoea is usually made through a combination of a thorough clinical history and physical examinations followed by imaging and laboratory diagnostic procedures.

The central feature in the history includes unilateral rhinorrhoea. This is usually described as watery, clear, and salty in taste. It often occurs intermittently but can be continual, copious, or minimal. Although it may appear similar to perennial or seasonal allergic rhinoconjunctivitis, CSF rhinorrhoea does not cause lacrimation, ocular pruritus, nasal pruritus, or sneezing. CSF rhinorrhoea can be exacerbated by the Valsalva manoeuvre or by bending forward (teapot sign), and may present as postnasal drip when the patient lies supine. It does not resolve with oral antihistamines, intranasal corticosteroids, antihistamines, or anticholinergics.

The ideal initial laboratory workup includes sending the collected fluid for either beta-2 (β_2) transferrin and/or β-trace protein which is found in CSF fluid. If the fluid is not collectible in the clinic with the patient leaning forward, it is advisable to give the patient a couple of sterile containers to try and collect the fluid at home.

β_2-transferrin

β_2-transferrin is a glycoprotein (β_2-isoform of transferrin) that is almost exclusively present in CSF and serves as a reliable marker for extracranial CSF. However, it has also been demonstrated in low concentrations in the perilymph of cochlea and aqueous and vitreous humour of the eye.

The β_2-transferrin assay is a non-invasive, highly sensitive test that could help avoid conduction of further invasive and/or expensive investigations. The reported sensitivity has ranged from 84% to 100% and specificity from 71% to 100%. Various methods of β_2-transferrin detection have been described; unfortunately, these techniques are labour intensive and require 2–4 hours to perform in a specialty laboratory. The cost of performing the test is around 50 USD. With current electrophoretic techniques, the minimum required sample volume to detect β_2-transferrin is 2 μL. However, it is generally recommended to collect at least 0.5 mL of fluid to account for possible contamination with other fluids.

Clinical conditions that could also lead to false-negative β_2-transferrin assay results would be central nervous system infections caused specifically by *Streptococcus pneumoniae*. False-positive results can occur due to an abnormal presence of β_2-transferrin in the serum. This has been reported in patients with chronic liver disorders, neuropsychiatric disorders, and hereditary disorders of glycoprotein metabolism. It is therefore advisable to routinely analyse both the serum and nasal fluid of each individual patient for simultaneous comparison.

A major benefit of the test is that it is non-invasive with no harm to the patient. Refrigeration of the sample is not required provided that it is not exposed to temperatures above 25°C.

β-trace protein

Prostaglandin D_2 synthase, commonly known as β-trace protein, is the second most abundant protein in CSF after albumin and is produced mainly in the leptomeninges and choroid plexus. High concentrations of β-trace protein are also found in the perilymph, urine, amniotic fluid, and seminal plasma. It has been noted to be absent in tear fluid and nasal secretion making it an ideal marker for the presence of CSF in rhinorrhoea samples. Automated nephelometry-based assays have shortened the analysis time to 15 minutes and reduced the cost of the test to approximately 20 USD.

The minimum sample volume for a β-trace protein assay is reported to be around 5 μL. It has also been noted that β-trace protein is stable in room temperature for several days.

Markedly elevated levels of β-trace protein have been documented in the serum of renal failure patients which would eventually elevate the concentration of the protein in nasal secretions. It is important to also note that bacterial meningitis could lower β-trace protein concentration in CSF, potentially leading to false-negative assay results. The sensitivity and specificity of β-trace protein in various studies ranged between 91% and 100%, and specificity between 96% and 100%.

Imaging studies

Once the CSF leak has been confirmed, the next step should be the organization of imaging studies for evaluation of leak localization. Numerous imaging studies are available such as high-resolution CT, contrast-enhanced CT cisternography, magnetic resonance imaging, radionuclide cisternography, and contrastenhanced magnetic resonance cisternogram that can be utilized for specific clinical scenarios.

These latter studies have the potential to demonstrate visualization of active CSF leaks through dural disruption. However, several

of these modalities are more invasive due to the requirement of intrathecal injection of contrast media or radioactive tracers. High-resolution computed tomography (HRCT) and magnetic resonance cisternography are the most relied upon studies as they are non-invasive and give excellent information as to the localization of the leak site.

HRCT

The initial study of choice that is usually used for preoperative planning is HRCT scanning of the sinuses and mastoid done using an image guidance protocol due to its high sensitivity. In the setting of a positive β_2-transferrin, it has been reported that HRCT has a positive predictive value of 100% and negative predictive value of 50–70% with an overall accuracy of 87%. Another advantage of HRCT is that it provides the surgeon with the best bony detail of the sinonasal anatomy as well as an ability of using image guidance during surgery. When multiple bony defects are seen on HRCT, superimposing magnetic resonance data can significantly increase the sensitivity of identifying the leakage site.

Magnetic resonance cisternography

With advances in magnetic resonance technology, intrathecal injection is discouraged. A 1.5-tesla or higher-strength magnetic resonance imaging is commonly utilized both for confirmation and site localization. It relies on the principle of demonstrating a contiguous fluid signal between the cisternal space and the nasal cavity on heavily weighted T2 images and image reversal to highlight CSF with an ability to detect herniated soft tissue through a skull base defect. The development of heavily weighted T2 and gradient sequences such as FIESTA (GE) and CISS (Siemens) have greatly helped in identification of the exact site of CSF leak. The studies report a positive predictive value of 64–92% and a negative predictive value of 71% with an overall accuracy of 78–96% in detecting CSF leak sites.

Intrathecal fluorescein

If the leakage site has not been accurately identified despite the imaging studies, or if there is concern of more than one leakage site, the use of intrathecal fluorescein intraoperatively may be utilized to effectively identify the leakage site(s). Intravenous fluorescein is not approved for use in the intrathecal space, and therefore a separate consent with proper discussion of the potential risks and complications needs to be carried out with the patient. The risk of a seizure is extremely rare and has only been documented with much higher doses of intrathecal fluorescein than currently used. A 0.05 mL 10% intravenous fluorescein solution mixed with 10 mL of CSF obtained during the preoperative spinal tap is re-injected back into the intrathecal space over a 10-minute period just prior to starting the surgery. The fluorescein percolates throughout the CSF and is easily seen endoscopically at the leakage site. A blue light filter may be utilized to enhance the fluorescein to make it easier to visualize.

Nuclear medicine studies (radionuclide cisternography)

The use of nuclear medicine imaging to diagnose and localize CSF leakage is not common practice. It is only useful occasionally when the patient is unable to collect any fluid for analysis, yet continues to have complaints that indicate a potential but inconclusive CSF leak despite negative studies with the more superior techniques described above. One to three cotton pledgets are placed in each nasal cavity after which a radionuclide tracer such as indium-111-labelled diethylenetriaminepentaacetic acid is injected intrathecally via a lumbar puncture. The pledgets are removed after 24 hours. The presence of radioactive tracer in the pledget(s) indicates a positive study but does not help identify the exact location of the leak.

Common sites of CSF leakage

The most common site of spontaneous CSF leakage is the cribriform plate due to the fragility of the plate and intimate juxtaposition of the arachnoid to the bone where the olfactory fibres pierce the skull base. Other common sites of leakage are the lateral recess of the sphenoid sinus which accounts for 35% of all CSF rhinorrhoea, and the roof of the ethmoid (**Figure 29.2**). CSF leakage can also occur in occasional cases from the posterior wall of the frontal sinus, the planum sphenoidale, and can originate from the middle cranial fossa via the middle ear and drain into the nose through the Eustachian tube.

Treatment

Once accurately diagnosed and properly identified, CSF rhinorrhoea that has not resolved spontaneously requires a surgical approach to properly repair the leakage site. Left untreated, the risk of ascending meningitis for the spontaneous CSF leak patient is greater than 10–50% and increases to 85% over 10 years. For the traumatic group of

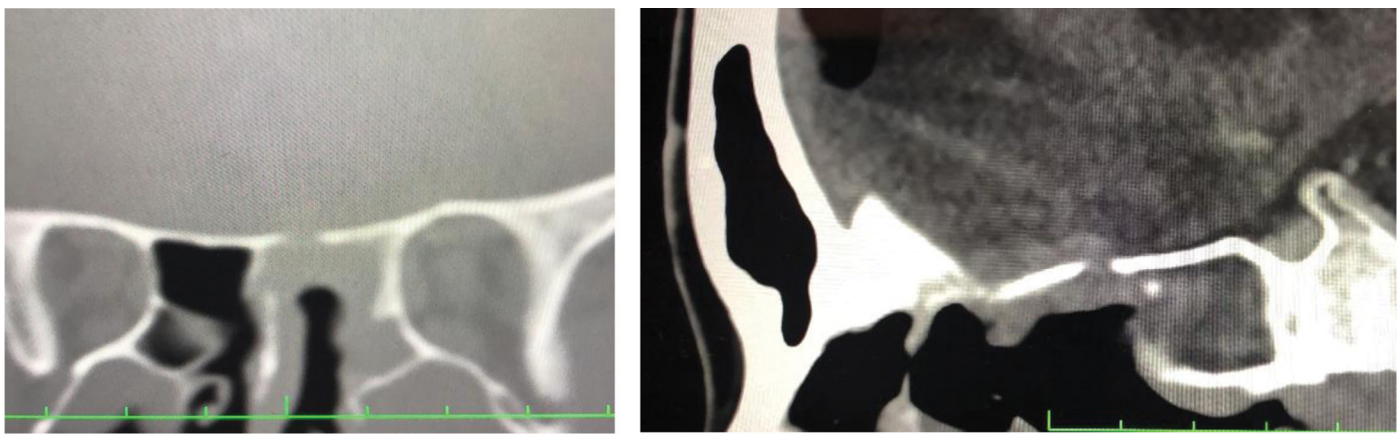

Figure 29.2 Iatrogenic defect of the posterior ethmoid with associated CSF leak.

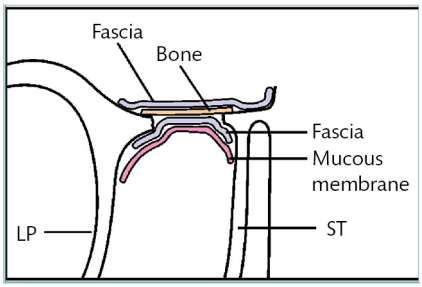

Figure 29.3 Underlay versus overlay techniques with various available autograft materials. LP, lamina papyracea; ST, superior turbinate.

patients, the rate of spontaneous repair is higher with a subsequent long-term meningitis risk of 25–30%. Surgical closure of defects at the skull base should therefore be considered treatment of choice. The literature does not support the routine use of prophylactic antibiotics or of lumbar drainage as treatment options.

Surgical treatment options historically included open intracranial approaches, but in recent decades these have largely been replaced by endonasal endoscopic approaches given their high success rate and low morbidity profile. Numerous endoscopic repair techniques including free tissue grafts, vascularized flaps, tissue sealants, and various multilayer combinations of these methods have developed as preferred by individual surgeons. Vascularized flaps have shown superiority in the setting of large dural defects or high-flow CSF leaks, otherwise, specific repair techniques are directed more by surgeon preference than by evidence-based guidelines. Adequate preparation of the leakage site is felt by most endoscopic sinus surgeons to be the most important aspect for obtaining a successful seal and repair. Removing surrounding mucosal membrane from the site of the bony defect is a *most important first step* that then allows the repair material a site to adhere to and seal the leak.

There has been no significant advantage demonstrated with the underlay technique over an overlay technique as long as adequate site preparation has been performed. Many different materials are now available for repair of CSF leak sites, but many authors still prefer using the patient's own tissue as seen in Figure 29.3. Dural sealants, fibrin glues, and related adjuncts are often used to maximize graft stability and seal the leak. It is unclear whether or not these adjuncts contribute to a successful repair.

Controversies

Allergic rhinitis

It is surprising that there are so many unanswered questions about phenotypes and endotypes of allergic rhinitis and its main multimorbidities (asthma, conjunctivitis), possible strategies for its fast and accurate diagnosis, and the best options of treating the disease and preventing its progression. Allergic rhinitis is becoming more challenging to treat, with intranasal corticosteroids providing insufficient symptom relief for many patients. There is a need for new allergic rhinitis treatment options which provide faster and more complete symptom relief than intranasal corticosteroids. A simplified approach to guide treatment decisions in the real world is needed with a simple visual analogue scale. Such a scale has been advocated by Contre les Maladies Chronique pour un Vieillissement Actif Allergic Rhinitis and its Impact on Asthma as the new language of allergic rhinitis control, and has been incorporated into the new allergic rhinitis management guideline called the allergic rhinitis integrated care pathway.

Food allergies

Some foods play a role in exacerbation of allergic rhinitis symptoms, but controversies exist regarding the role of diet on incidence or worsening of symptoms. Research demonstrates a possible relationship between certain foods and their induction of allergic reactions by modulating IgE.

Cranial cerebrospinal fluid leaks and spontaneous intracranial hypotension

Spontaneous intracranial hypotension is an important cause of headaches and an underlying spinal CSF leak can be demonstrated in most patients. Whether CSF leakage at the skull base can cause spontaneous intracranial hypotension remains a matter of controversy. Research indicates that there is no evidence for an association between spontaneous intracranial hypotension and CSF leaks at the skull base, and that a spinal source for CSF leakage should be suspected in patients with orthostatic headaches who have a documented skull base CSF leak.

Sealant use in endoscopic repair of cerebrospinal fluid leaks

The use of sealants to create a watertight repair of a CSF leak is controversial. While the majority of endoscopic sinus surgeons may use sealants, studies indicate that currently available sealants seem not to reduce the rate of CSF leakage and that site preparation and technique are likely the more important determinants of a successful outcome.

Guidelines

Several well-written clinical practice guidelines exist for rhinitis and allergic rhinitis. Many of the guidelines are endorsed by major societies. The American Academy of Otolaryngology–Head and Neck Surgery published its comprehensive guidelines on allergic rhinitis in 2015. The Canadian guidelines for the diagnosis and treatment of allergic rhinitis were published in 2007 with an overview and update of the recommendations published in 2018. The American Academy of Family Practice published its clinical practice guidelines on allergic rhinitis in April 2020 with several key recommendations. The ARIA guidelines were developed in collaboration with the World Health Organization in 2001. The original comprehensive guidelines have been updated and revised in 2008, 2010, and 2016. The 2010 revision resulted from a complete re-review of the underlying evidence and developed recommendations following the Grading of Recommendations, Assessment, Development and Evaluation (GRADE) approach: an evidence-based way of making healthcare decision acknowledging that evidence alone is insufficient, and that values and preferences, clinical circumstances, as well as clinical expertise inevitably influence decisions. The 2016 guidelines provide updated and new recommendations about the pharmacological treatment of allergic rhinitis. It specifically addresses the relative merits of using oral H_1-antihistamines, intranasal H_1-antihistamines, intranasal corticosteroids, and leukotriene receptor

antagonists either alone or in combination. Next-generation guidelines for the pharmacological treatment of allergic rhinitis were developed in 2018 by using the GRADE-based guidelines. Real-world evidence, data, and chamber studies were used to refine the algorithm for allergic rhinitis by a consensus group. An algorithm based on the visual analogue scale was devised for selection of pharmacotherapy for patients with allergic rhinitis and to step up or down treatment depending on control.

Conclusion

In order to properly diagnose the cause of rhinorrhoea, it is imperative that the clinician takes a detailed history and carries out a meticulous ear, nose, and throat, as well as an endoscopic examination of the nose and sinonasal cavities. It is important for the clinician to differentiate between a bilateral and unilateral rhinorrhoea. It is exceedingly important that a unilateral rhinorrhoea, caused by some ominous pathology, does not get misdiagnosed as an allergic rhinitis. Adequate knowledge of the various testing modalities and imaging parameters is important, especially in the case of a CSF rhinorrhoea.

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30

Postnasal drip

Isma Iqbal

Introduction

Postnasal drip (PND) is a common symptom. Its definition is complex and may be described as a sensation of mucus running down the back of the throat, discomfort in the back of the nose, or a need to clear the postnasal space or throat. It may be associated with cough and throat clearing. PND is associated with a number of sinonasal conditions. Normal mucosal function involves mucus produced by the paranasal sinuses and nasal cavity being transported to the nasopharynx and subsequently swallowed. An increase in mucus production, change in viscosity, or inflammatory composition of the mucus can lead to a sensation of PND.

Basic science

Anatomy

The postnasal space (or nasopharynx) is defined as the area posterior to the termination of the nasal septum and extends superiorly from the floor of the sphenoid sinus to the soft palate inferiorly.

Boundaries

- *Roof and posterior wall*: basisphenoid and basiocciput. Posterior wall formed by C1 and C2 vertebrae covered by prevertebral muscles and fascia. Nasopharyngeal adenoidal tissue present at the junction of the roof and the posterior wall.
- *Floor*: soft palate anteriorly. Communicates inferiorly with the oropharynx.
- *Anterior wall*: communicates with nasal cavities through choanae.
- *Lateral wall*: pharyngeal opening of Eustachian tube, torus tubarius, tubal tonsil, and fossa of Rosenmüller.

Blood supply

Branches of the external carotid artery:

- Ascending palatine branch of facial artery.
- Ascending pharyngeal artery.
- Pharyngeal branch of third part of maxillary artery.

Physiology

Nasal mucus, the nasal cycle, mucociliary clearance, and plasma extravasation of nitric oxide assist in immune defence. Nasal mucus is produced by submucosal and seromucous glands, goblet cells, transudation of blood plasma, mucosal tissue fluid, and tear fluid. An external stimulus to the nasal mucosa produces inflammatory mediators such as histamine. There is subsequent stimulation of the nasal sensory nerves which include unmyelinated (nociceptive, C fibre (slow response)) and myelinated (Aƍ (fast response)) fibres, as well as the nasociliary nerve of the trigeminal nerve, which transmit a signal to the nucleus of the spinal trigeminal tract. A projection of the vagus nerve is also transmitted to a central area called the nucleus tractus solitaris. The theory hypothesizes that environmental stimuli cause activation of nasal mucosa through inflammation which leads to signals being transmitted to the nucleus of the spinal trigeminal tract and to the nucleus tractus solitaris via the trigeminal nerve. Subsequently the vagus nerve will be activated to cause neurogenic inflammation in the lower airway (**Figure 30.1**).

Pathophysiology

The pathophysiology of PND is not clearly understood. Patients may present with a triad of chronic cough, sensation of mucus running down the back of the throat, and throat clearing. However, all three symptoms are usually not present in all patients presenting with this condition. It is not clear as to whether there is increased volume of nasal secretions or if there is an alteration in the nerve receptors causing hypersensitivity to airway inflammation, or even a neurochemical imbalance. Some studies have also suggested that the viscosity of mucus is, in fact, increased and patients complained that they were unable to expectorate secretions. The composition of mucus may also vary and different mediators within mucus may trigger differences in symptomatology and drive hypersensitivity.

The American College of Chest Physicians uses the term upper airway cough syndrome (UACS) to replace PND syndrome. However, some patients may have cough hypersensitivity which is described later in this chapter (see 'Chronic cough'). There is evidence demonstrating that a significant number of patients with PND do not have a cough.

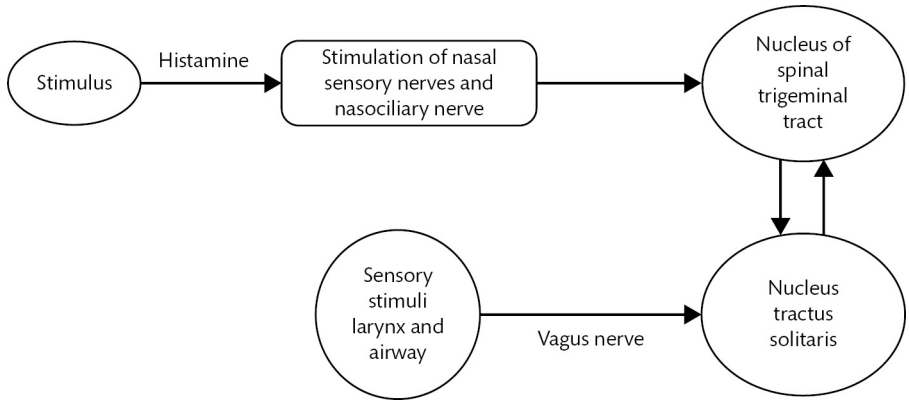

Figure 30.1 Neural pathway showing the relationship between upper and lower airway stimulation.

Assessment

History

Patients complain of mucus dripping or sitting at the back of the throat which may be constant or intermittent. The viscosity of the fluid in the postnasal space can vary between patients. The symptoms may be worse in the morning with individuals complaining of a sore throat when they wake up. Throat clearing or spitting a small amount of mucus may also be described. Patients may also refer to this as catarrh. For those patients presenting with cough, smoking history, use of an angiotensin-converting enzyme inhibitor, and smoking-related pulmonary disease should be explored.

In addition to this, identifying any dietary triggers (e.g. dairy) may be helpful. Occupational history may be relevant with patients reporting increased symptoms when working in a dry environment (e.g. flight crew or dust workers).

Examination

Examination may be entirely normal. Particular care should be taken to examine the middle and superior meati, as well as the sphenoethmoidal recess to rule out a potential sinogenic cause. Inspection of the oropharynx may demonstrate some mucus arising from the postnasal space hanging down. There may be inflammation of the mucosa on the posterior pharyngeal wall which is described as a cobblestone or nodular appearance. Flexible nasendoscopy is essential to assess the nasal cavity and rule out primary causes such as acute, chronic, or fungal rhinosinusitis, allergic and non-allergic rhinitis, or sinonasal malignancy. In the presence of these conditions, management of PND involves treating the primary cause.

Patient experience

In many cases PND can be a habitual response. PND can be a troublesome condition which many patients find embarrassing and which adversely affects their quality of life. This is due to the constant need to throat clear, sniff, or cough. Many patients fail to produce any significant expectorant which can lead to further frustration. Additional symptoms that patients may describe include halitosis, dry throat, and sleep disturbance.

Diagnosis

The mainstay of diagnosis is based on history and examination. There are no objective criteria which can make diagnosis difficult.

Causes of postnasal drip

The causes of PND are highlighted in Box 30.1.

Investigations

The diagnosis is predominantly based on history and examination. Imaging is used to exclude other causes and computed tomography scanning of the sinuses would be indicated in such cases.

Differential diagnoses

Chronic cough

This is defined as a persistent dry cough present for at least 8 weeks' duration. It can also be known as cough hypersensitivity syndrome. There are three different phenotypes for cough hypersensitivity syndrome: predominantly rhinitic and sinus-related symptoms, T-helper-2 cell dominant, and acid reflux-related. The symptoms may be exacerbated by seasonal or occupational triggers. Patients may also describe other environmental triggers such as aerosols, change

Box 30.1 Causes of postnasal drip

- Adenoidal hypertrophy.
- Infection (bacterial/viral).
- Anatomical anomalies.
- Rhinitis (allergic/non allergic).
- Mucociliary dysfunction.
- Rhinosinusitis (acute/chronic).
- Smoking and exposure to toxins.
- Cystic fibrosis.
- Laryngopharyngeal reflux.
- Gastro-oesophageal reflux.

in room temperature, or pollen. A salient feature in the history is an unpleasant sensation in the throat. There may also be associated voice changes.

The pathophysiology is related to direct or chemical triggers causing activation of the cough reflex. The exact mechanism is, however, not understood completely.

Examination findings may again reveal a cobblestone appearance of the posterior oropharyngeal wall and/or visible mucus.

Avoidance of triggers and any environmental allergens is recommended. Treatment includes a trial of antihistamine and nasal decongestant for 2 weeks. First-generation antihistamines such as chlorpheniramine seem to be more effective than second-generation ones. Non-medical measures with laryngeal hygiene therapy by a speech and language therapist has had some success through counselling, reducing the throat clearing and sensation of needing to cough.

Reflux-related cough

This may be present as gastro-oesophageal reflux or laryngopharyngeal reflux. Those with gastro-oesophageal reflux present with heartburn and cough. Laryngopharyngeal reflux manifests as silent reflux without any heartburn. In addition to this, symptoms include throat clearing, hoarseness, and a globus sensation. The Reflux Symptom Index can be used as a quantitative measure to assess severity. Diagnosis should be based on history and examination. In some cases, 24-hour pH probe monitoring and endoscopy may be required. Examination findings include laryngeal oedema and erythema, laryngeal pseudosulcus, and posterior commissure hypertrophy or pachydermia.

Lifestyle and dietary modifications as well as treatment of acid reflux are recommended in both conditions. Treatment with proton pump inhibitors or alginate-based medication is recommended. Symptoms may start to settle within 2 weeks of commencing treatment. However, a 6–8-week trial is recommended.

Rhinitis medicamentosa

Rhinitis secondary to chronic use of nasal decongestants may cause symptoms of PND. The commonest topical agents used are oxymetazoline and phenylephrine. Overuse of topical vasoconstrictors causes tolerance and tachyphylaxis with severe rebound rhinitis. In the first instance this should be treated with cessation of the offending factor. Treatment with topical nasal steroids may be helpful during this period. In some patients cessation of medication can lead to severe nasal congestion which may be refractory to treatment.

Non-allergic rhinitis

Patients may present with symptoms of nasal obstruction and thickened nasal mucus causing PND. There may also be excessive watery anterior rhinorrhoea which can be triggered by environmental temperature change, alcohol use, or environmental odours or aerosols. There may be associated sneezing, nasal congestion, and pruritus. This can be exacerbated by inhaled agents, smoking, certain foods, chemicals, and medications. Hormonal changes may also have an impact on symptoms.

Subtypes of non-allergic rhinitis:

- *Occupational*: symptoms caused by inhaled triggers. These include chemicals, solvents, and cigarette smoke. Avoidance of these triggers controls symptoms.
- *Hormonal*: this is reportedly most prominent in the later stages of pregnancy and resolves within 2–4 weeks of delivery.
- *Drug-induced*: certain medications can have physiological side effects on the nose. These include angiotensin-converting enzyme inhibitors, beta blockers, oral contraceptives, and antipsychotics.
- *Gustatory*: watery rhinorrhoea which may be associated with PND. Occurs in anticipation of eating or even with the smell of food. Responses mediated by parasympathetic stimulation of nasal secretomotor fibres on stimulation of salivary secretomotor activity.
- *Vasomotor*: a diagnosis of exclusion. The most common form of non-allergic rhinitis. Seen commonly in the elderly population. Thought to be due to an imbalance in the autonomic system resulting in parasympathetic hyperactivity. Patients complain of severe watery rhinorrhoea on exposure to triggers such as a change in weather, alcohol ingestion, or perfume.

Treatments

Treatment of PND should be aimed at identifying the underlying cause. In the absence of other sinonasal pathology, treatment is primarily medical or surgical if there is evidence of structural abnormality leading to PND. In refractory cases, counselling in the form of cognitive behavioural therapy may be required.

Medical treatments

Treatment is based on symptoms and examination findings. A review of the patient's current medication may identify drugs which may be causing a drying sensation in the postnasal space such as anxiolytics, antidepressants, and anticholinergics.

Lifestyle changes

Lifestyle changes are useful in order to improve mucus flow. Thick secretions can be particularly problematic in older patients. Measures such as increased fluid intake, reduced caffeine intake, and, where possible, avoiding diuretics may be helpful.

Medication

Some patients may respond to simple measures such as a saline nasal douche. Management of patients without associated cough is with topical intranasal treatment. In patients with severe symptoms or refractory disease, combination therapy (fluticasone nasal drops, ipratropium nasal spray, and oral azelastine) is recommended. Those with concomitant lower airway disease may benefit from a leukotriene inhibitor (montelukast) or a prolonged course of macrolide antibiotics. In severe cases unresponsive to treatment gabapentin has been reported to be effective in those with troublesome cough.

Medical treatment is summarized in Table 30.1.

Surgical treatments

In cases refractory to topical treatment, surgical management may be effective in the presence of a clear structural or mucosal abnormality which may be causing functional problems. Surgical treatments are targeted at improving any structural abnormalities such

Table 30.1 Summary of medical treatment for postnasal drip

Treatment	Dose	Side effects	Additional information
Topical intranasal corticosteroida Fluticasone propionate (1 spray = 50 mcg) Fluticasone furoate (1 spray = 27.5 mcg) Mometasone furoate (1 spray = 50 mcg) Beclomethasone dipropionate (1 spray = 50 mcg)	 1–2 sprays each nostril 1–2 times a day 1–2 sprays each nostril once daily 1–2 sprays each nostril 1–2 times a day 1–2 sprays each nostril 1–2 times a day	Epistaxis Headache Throat irritation	
Antihistaminea Chlorpheniramine (1st generation) Cetirizine (2nd generation) Azelastine hydrochloride (1 spray = 140 mcg) Dymista (fluticasone propionate + azelastine hydrochloride) (1 spray = 137 mcg/50 mcg)	4 mg every 4–6 hours (24 mg maximum daily) 10 mg daily 1 spray to each nostril 2 times a day 1 spray to each nostril 2 times a day	Impaired concentration Dizziness Dry mouth Fatigue Headache Blurred vision	1st-generation antihistamines are more effective at drying postnasal secretions. Recommended to use with topical nasal decongestant for 1–2 weeks
Leukotriene inhibitorsb Montelukast	10 mg daily	Diarrhoea Fever Gastrointestinal discomfort Headache Skin reactions	
Topical ipratropium bromidec (1 spray = 21 mcg)	2 sprays each nostril 2–3 times a day	Bladder outflow obstruction Acute angle glaucoma	
Gabapentind	300 mg once daily (300 mg 3 times a day maximum)	Respiratory depression in elderly Anxiety, arthralgia, confusion, drowsiness, nausea, dry mouth, dysarthria, diarrhoea, vertigo	Increase dosage incrementally

[a] National Institute for Health and Care Excellence guidelines on chronic cough due to UACS.
[b] European Respiratory Society guidelines on chronic cough.
[c] National Institute for Health and Care Excellence guidelines on rhinitis, evidence in literature.
[d] Evidence from case report and European Respiratory Society guidelines on chronic cough.

as turbinate hypertrophy and septal deviation. This would aid in improvement of mucociliary clearance.

Inferior turbinate reduction

Submucous diathermy or turbinoplasty is recommended to address turbinate hypertrophy. This subsequently reduces nasal secretions as well as allowing improved drainage of secretions.

The methods include:

- coblation
- mini-microdebrider
- outfracture
- submucous diathermy to inferior turbinates
- radiofrequency ablation
- laser
- conchopexy.

Septoplasty

Septoplasty may be performed via an open technique or endoscopically. Correction of any septal deformity would facilitate delivery of intranasal medications which may provide symptomatic relief as well as aiding drainage of the ostiomeatal complex.

Vidian neurectomy

This should only be considered in exceptional cases. The autonomic supply is disrupted leading to a reduction in nasal secretions. Success is limited and patients may complain of excessive dryness of the nose post procedure. Patients should be warned about possibility of dry eyes.

Figure 30.2 provides a schematic to aid management of PND.

Controversies

PND is a difficult syndrome to define quantitatively with no objective measures. In addition to this, patient perception of what constitutes PND varies significantly. This presents controversy in identifying and managing these patients.

The American College of Chest Physicians recommends the term UACS in preference to PND syndrome. This is in contrast to the UK. Due to the nature of PND there is no objective measure and hence a debate still exists as to the exact nature of this condition. As the disease mechanism is still not clear, further research is needed.

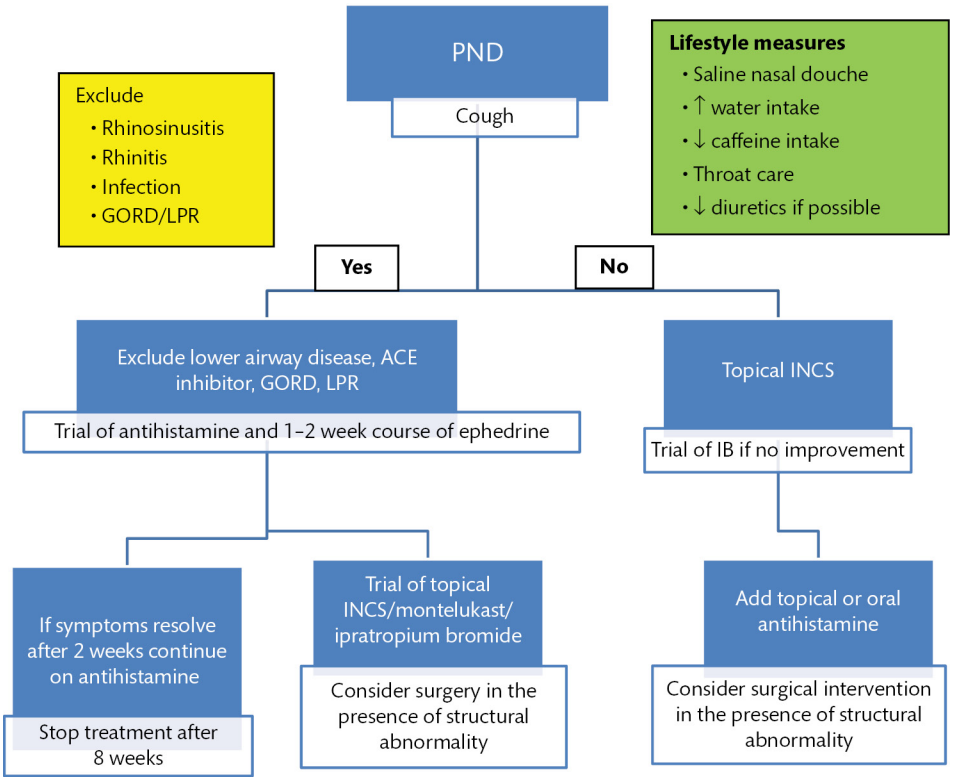

Figure 30.2 Management framework for PND. ACE, angiotensin-converting enzyme; GORD, gastro-oesophageal reflux; IB, ipratropium bromide; INCS, intranasal corticosteroid; LPR, laryngopharyngeal reflux.

Tips and pearls

- Identify and treat underlying cause.
- Avoid triggers.
- Where cough is present, consider lower airway disease.
- Consider red flag symptoms: haemoptysis, weight loss, dysphagia, unilateral nasal symptoms.
- Judicious medical treatment.
- Avoid surgery unless structural abnormality or mucosal disease is present.
- Management of patient expectations is key.

Guidelines

There is no specific guidance on management of PND without cough in the UK. However, UACS guidance is available:

- National Institute for Health and Care Excellence guidelines on cough include treatment of UACS.
- European Respiratory Society guidelines on UACS.

Related chapters

See related chapters for further information on allergy of the nose (Chapter 42), rhinorrhoea (Chapter 29), nasal obstruction (Chapter 33), chronic sinus disease (Chapter 38), acute sinus disease (Chapter 36), and endoscopic sinus surgery (Chapter 37).

Conclusion

PND is a common condition related to a number of sinonasal conditions. PND syndrome is therefore diagnosed when these conditions are excluded. The severity and characteristics of the condition vary among individuals. Due to the subjective nature of the condition no formal test is available, making diagnosis difficult. The mainstay of management is topical treatment with surgical consideration in cases with clear structural abnormalities or in those refractory to treatment.

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31

Headache and facial pain

Iain Hathorn

Introduction

Headache and facial pain are complaints frequently encountered by otolaryngologists and have a broad differential diagnosis. There is a common misconception that they are mainly caused by sinusitis. Far too often patients undergo treatment, including surgery, for sinonasal disease when a clear history would identify a far more likely non-sinonasal cause. It is important to correctly identify the underlying problem so that the patient can be managed effectively and not simply to exclude underlying sinus disease.

Assessment

A detailed history and examination are essential to identify the cause of facial pain. The history should establish the following (**Table 31.1**):

- *Quality of pain*: sharp, dull, throbbing, burning, aching, pressure.
- *Distribution*: unilateral, bilateral. Although rare, tumours should be excluded in patients with unilateral, progressive pain especially if associated with neurological deficits.
- *Frequency and periodicity of symptoms*: migraine-related pain is often associated with the premenstrual period in women. Cluster headaches occur several mornings in a row with periods of remission. It is unlikely that daily, constant facial pain is related to sinusitis and is more likely due to midface segment pain.
- *Onset*: sudden, gradual.
- *Severity*: visual analogue score 1–10, 1 being minimal pain and 10 being the worst pain imaginable. A score of 10/10 is typical for migraine or cluster headache.
- *Duration*: seconds, minutes, hours.
- *Associated symptoms*: aura, nausea, photophobia, hyperaesthesia of skin, sinonasal symptoms. Associated sinonasal symptoms may include rhinorrhoea, postnasal drip, nasal congestion, and altered sensation of smell. Facial pain, caused by the trigeminal autonomic cephalgias (see below), can be associated with autonomic symptoms, such as nasal congestion and rhinorrhoea, making the correct diagnosis challenging.
- *Exacerbating or relieving factors*:
  - There are some recognized triggers such as alcohol for cluster headaches. Tension-type headaches and midfacial segment pain do not have any classical exacerbating or relieving factors but patients do sometimes describe bending forwards can make them worse. This often leads to incorrectly diagnosing them with rhinosinusitis.
  - The impact the pain is having on the patient's daily life should be established. Finally, a drug, family, and social history should be taken and any history of dental disease, trauma, or surgery prior to onset of symptoms noted.
  - In addition to a focused history, nasendoscopy to visualize the middle meatus, and a targeted examination are essential. In the absence of mucopus or inflammation in the middle meatus, a diagnosis of rhinosinusitis is very unlikely.
  - An ear, nose, and throat examination should be completed, and targeted areas included. The oral cavity and the teeth should be examined for any lesions, decay, or abnormalities. The temporomandibular joint (TMJ) and the muscles of the head and neck should be palpated for any tenderness and signs of trigger points. The trigeminal nerve and other cranial nerves should be examined when numbness or visual symptoms are reported.

Investigation

Routine requesting of computed tomography (CT) scans should be carefully considered due to the relatively high frequency of finding incidental mucosal thickening in one or more sinuses. CT scans should be considered to investigate those patients where there is suspicion of malignancy, fungal disease, or complications of sinusitis, and also in chronic rhinosinusitis (CRS) which has failed medical treatment and surgery is planned. Dental radiographs and CT scans can be useful if there is suspicion of maxillary sinus disease secondary to periapical infection (**Figure 31.1**).

Magnetic resonance imaging (MRI) may help evaluate TMJ disorders. MRI of the brain may be useful for intracranial pathology, such as tumour or hydrocephalus, and vascular causes of headache. In trigeminal neuralgia, MRI will detect vascular compression or demyelination of the nerve root.

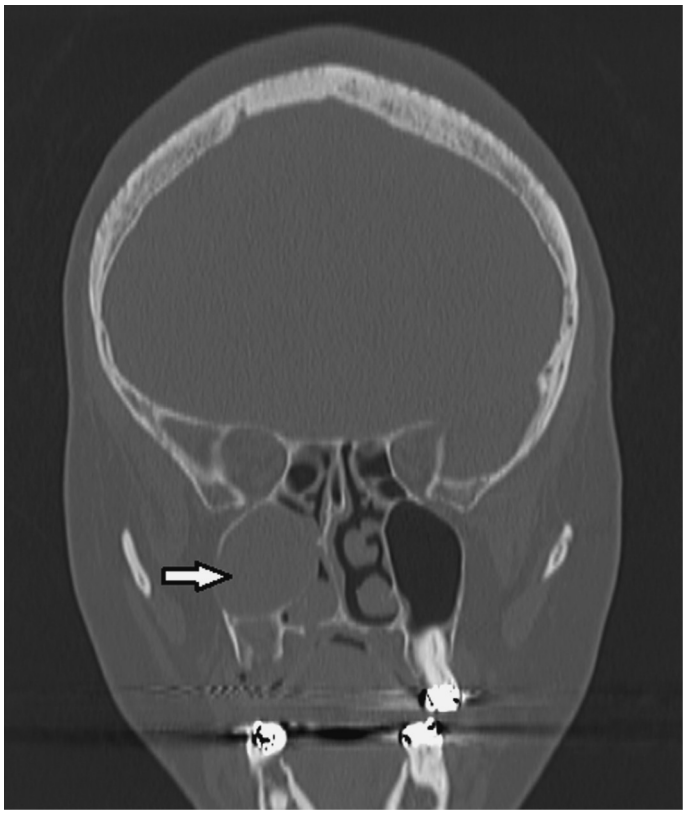

Figure 31.1 Right maxillary sinusitis secondary to a dental cause. Arrow identifies an opacified right maxillary sinus due to sinusitis following tooth removal.

Differential diagnosis

There are a wide range of possible causes of headache and facial pain, which often have overlapping features making diagnosis challenging.

The International Headache Society International Classification of Headache Disorders, third edition (ICHD-3) divides headaches into primary headache disorder, secondary headaches, and neuropathies (Figure 31.2). We will consider the clinical features and treatments of specific conditions causing headaches and facial pain encountered by otolaryngologists. These are summarized in Tables 31.1 and 31.2, respectively.

Clinical features and treatment of specific conditions

Primary headache disorders

Migraine

Migraine is common, affecting about one in seven people globally. It can be episodic or chronic and is characterized by attacks of unilateral, severe, throbbing headache. In classic migraine (25%), patients describe associated nausea, aura, and photophobia. An aura is characterized by transient focal neurological symptoms, such as zigzag lines in vision or pins and needles, which precede, or occasionally accompany, the headache. The episodes last from 4 to 72 hours and can be triggered by a range of external and internal

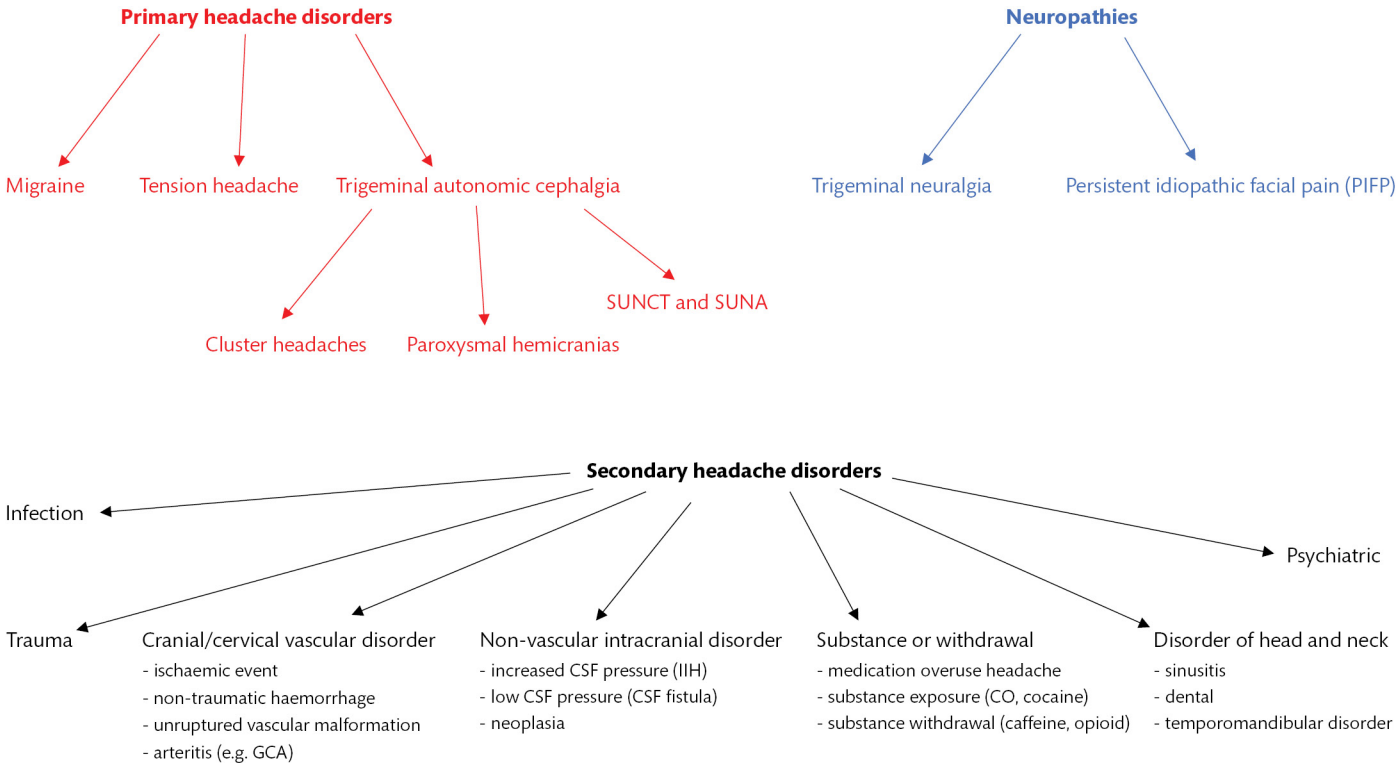

Figure 31.2 Classification of headache and facial pain—ICHD-3. CO, carbon monoxide; CSF, cerebrospinal fluid; GCA, giant cell arteritis; IIH, idiopathic intracranial hypertension; SUNA, short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms; SUNCT, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing.

Source data from the International Headache Society ICHD-3 (https://ichd-3.org/).

Table 31.1 Clinical features of specific causes of headache and facial pain

Feature	Neuropathic	Primary headache disorder						Midfacial segment pain
	Trigeminal neuralgia	Paroxysmal hemicrania	Cluster headache		Migraine		Tension-type headache		
Location	Distribution of trigeminal nerve divisions (1 or more)	Orbital, supraorbital	Around the eye		Frontotemporal		Vertex		Midface (nasal bridge, paranasal, periorbital)
Side	Unilateral	Unilateral	Unilateral		Mostly unilateral		Bilateral, symmetrical		Bilateral, symmetrical
Intensity	Severe	Severe	Very severe		Moderate/severe		Mild/moderate		Mild/moderate
Quality	Shooting, stabbing, sharp (electric shock-like)	Burning, piercing	Variable (sharp, piercing, burning, boring)		Pulsating, throbbing		Pressing/tightening (non-pulsating)		Pressing/tightening (non-pulsating)
Duration	Seconds	5–30 min	15–180 min		4–72 h		30 min to continuous		Continuous
Associated symptoms	Mild autonomic symptoms (lacrimation, redness of ipsilateral eye)	Ipsilateral nasal congestion, rhinorrhoea, lacrimation, miosis or ptosis, red eye, swollen eyelid, facial sweating, agitation	Ipsilateral nasal congestion, rhinorrhoea, lacrimation, miosis or ptosis, red eye, swollen eyelid, facial sweating, agitation		Nausea, vomiting, photophobia, phonophobia		Nil		Worsens on bending over, hyperaesthesia May have sense of nasal obstruction
Others	Can be precipitated by innocuous stimuli over affected distribution Predominantly V_2/V_3 affected	Shorter, more frequent attacks than cluster, chronic form also exists (hemicrania continua)	Mostly affects males, usually come in bouts lasting weeks to months		Aura—can occur with or without headache and are fully reversible, develop over ≥5 min, last 5–60 min				Patients often overuse analgesics without clear benefit
Frequency	Paroxysmal 1–5/day	Paroxysmal up to 5/h	1 every other day—8/day, with remission >3 months	1 every other day—8/day, with continuous remission <1 month	<15 days/month	≥15 days/month for >3 months	<15 days/month	≥15 days/month for >3 months	Paroxysmal/continuous
Diagnosis			Episodic cluster headache	Chronic cluster headache	Episodic migraine	Chronic migraine	Episodic tension-type headache	Chronic tension-type headache	

Table 31.2 Treatment of specific headache conditions

Condition	First-line treatment	Dose
Migraine	Acute—sumatriptan Prevent—propranolol	50–100 mg, maximum 300 mg/24 h 80–160 mg/24 h
Tension-type	Amitriptyline	10–25 mg, maximum 125 mg/24 h
Cluster	Oxygen Sumatriptan	7–15 L/min 6 mg subcutaneously, maximum 12 mg/24 h
Paroxysmal hemicrania	Indomethacin	Start 25 mg three times daily for 7 days, titrate to response, maximum 200 mg/24 h
SUNCT/SUNA	Lamotrigine	Up to 400 mg/24 h
Midfacial segment pain	Amitriptyline	10 mg
Trigeminal neuralgia	Carbamazepine	Start 100 mg twice daily, titrate until pain controlled, maximum 1600 mg/24 h
PIFP	Amitriptyline	25–100 mg/24 h

factors including stress, diet, premenstrual state, and barometric pressure. It is more common in women and there is often a positive family history.

The symptoms of migraine may overlap with those of other conditions including tension, cluster, or 'sinus' headaches which can cause confusion with diagnosis. Also, migraines may cause some autonomic symptoms (nasal congestion, discharge, watery eyes) during attacks, adding to the diagnostic uncertainty. In fact, facial pain may be the only presenting feature in migraine with no associated headache.

Management of migraine includes trigger avoidance and lifestyle changes. Medical treatment can be divided into abortive or prophylactic therapy. The use of abortive medications for acute migraine should be limited to a few days a week to avoid a rebound headache phenomenon developing. Medications include simple analgesia (paracetamol 1 g, ibuprofen 400 mg, or aspirin 900 mg), or a triptan. Oral sumatriptan (50–100 mg) is first choice, either alone or in combination with paracetamol or a non-steroidal anti-inflammatory drug.

Preventative treatment should be considered if:

- migraine attacks are having a significant impact on quality of life, due to frequency (more than once a week on average) or severity, despite optimal acute treatment
- acute treatments are contraindicated or ineffective
- the patient is at risk of medication overuse headaches due to frequent use of acute drugs.

The choice of preventative treatment depends on contraindications, comorbidities, and risk of adverse events. Drug treatments should start at a low dose, titrated according to efficacy and tolerability. The medications commonly used are propranolol (80–160 mg daily in divided doses), topiramate (50–100 mg daily in divided doses), or amitriptyline (25–150 mg at night). Topiramate is *contraindicated* in pregnancy and gabapentin should not be offered for migraine prophylaxis. Other non-drug therapies can be considered as an adjunct or alternative including cognitive behavioural therapy, relaxation techniques, or acupuncture. Botulinum toxin type A is an option for prophylaxis for chronic migraine where medication overuse has been addressed and patients have been appropriately treated with three or more oral prophylactic medications.

Tension-type headache

Tension-type headaches are common. They are characterized by mild/moderate pain with a symmetrical bilateral distribution. Patients describe a constant, tight, band-like sensation across the frontal and temporal regions. They tend to last for hours and they may have trigger points in the cervical muscles. Neurological examination is normal (**Box 31.1**). Stress, sleep deprivation, hunger, and eye strain are all thought to be precipitating factors in susceptible individuals.

Trigeminal autonomic cephalgias

This group of neuralgias cause recurrent, severe, lancinating pain over the ophthalmic branch of the trigeminal nerve. They tend to be associated with autonomic symptoms such as ipsilateral watery eye, rhinorrhoea, ptosis, and miosis, and so may incorrectly be attributed to a sinus aetiology.

Box 31.1 Diagnostic criteria for chronic tension-type headache (ICHD-3)

1. Headache occurring on at least 15 days/month on average for more than 3 months (≥180 days/year), fulfilling criteria 2–4.
2. Lasting hours to days, or unremitting.
3. At least two of the following four characteristics:
   - Bilateral location.
   - Pressing or tightening (non-pulsating) quality.
   - Mild or moderate intensity.
   - Not aggravated by routine physical activity such as walking or climbing stairs.
4. Both of the following:
   - No more than one of photophobia, phonophobia, or mild nausea.
   - Moderate or severe nausea or vomiting not present.
5. Not better accounted for by another ICHD-3 diagnosis.

Cluster headaches

These present with very severe, unilateral pain in the periorbital region. The headaches are characterized by recurrent attacks lasting around 30–60 minutes, associated with ipsilateral autonomic symptoms. They predominantly affect men aged 30–40 years.

Treatment can be divided into acute and prophylactic therapy. For the acute attack, oxygen (100% via facemask for 10 minutes) is used and, if started early, it can stop the headache. Subcutaneous sumatriptan (6 mg) injection can be effective and acts rapidly. Sumatriptan via nasal spray can also be used but is not as effective.

There is a role for the use of pizotifen or verapamil as prophylactic therapy for cluster headaches.

Paroxysmal hemicrania

This is characterized by recurrent episodes of severe, unilateral headaches in the same distribution as cluster headache with associated ipsilateral autonomic symptoms. Symptoms can last up to 30 minutes and patients may have many episodes in a day. If untreated, hemicrania continua can develop, when the headache is continuous but with fluctuation.

Symptoms show complete resolution on use of indomethacin (usually 150 mg/day sees response within 48 hours).

SUNCT and SUNA

Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) and short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) cause unilateral periorbital pain. Patients describe recurrent, sharp (like electric shock), severe pain, lasting for a few seconds. They can have very frequent attacks (hundreds/day) and have associated autonomic symptoms.

First-line treatment is lamotrigine, 100–400 mg/day.

Midfacial segment pain

Midfacial segment pain is not recognized as a separate primary headache syndrome in the ICHD-3 but it is an established clinical entity seen in rhinology outpatient clinics. Clinical features described include symmetrical pressure across the nasion, nasal dorsum, periorbital region, or across the cheeks. The forehead and occipital region may also be affected, and hyperaesthesia of the skin may be found.

Treatment is with low-dose nortriptyline (10–50 mg) or amitriptyline (10 mg) for 6 months. Complementary therapies to the neck and scalp may also be effective.

Neuropathies

Trigeminal neuralgia

Trigeminal neuralgia affects approximately 5/100,000 people per year. Patients experience severe, short-lasting paroxysms of lancinating pain in the distribution of one or more of the Vth cranial nerve branches. The pain occurs across both the maxillary and mandibular divisions in over a third of cases. The pain is triggered by minimal stimulation of the affected area.

MRI is required as, although the majority are idiopathic, trigeminal neuralgia can be secondary to vascular or neoplastic compression, or infiltration of the trigeminal ganglion. Depending on the cause treatment may be medical or surgical.

Medical treatment includes carbamazepine, although other antiepileptic drugs such as lamotrigine may be used.

If secondary to compression of the trigeminal nerve from pontine vessels, then surgical microvascular decompression or Gamma Knife radiation may be used. There is some evidence that microvascular decompression may result in the best long-term remission of symptoms.

Persistent idiopathic facial pain

Previously known as atypical facial pain, persistent idiopathic facial pain (PIFP) is an excruciating condition with persistent pain, usually unilateral and without autonomic symptoms or signs (**Box 31.2**). It is often described as a burning sensation or severe ache and can be compared to trigeminal neuralgia, except that the symptoms are persistent rather than intermittent. The diagnosis should be considered in patients with neuropathic facial pain whose symptoms do not fit with more common causes.

The pain is often poorly localized with widespread pain extending from the face onto other areas of the head and neck. There may be a history of other pain syndromes or depression and patients are often unable to function normally due to the pain.

There are no curative therapies for PIFP. However, some treatments have been shown to be moderately effective. The mainstay of treatment is low-dose amitriptyline, at an initial dose of 25–100 mg per day. Second-line pharmacological treatment includes gabapentin and pregabalin. There is some evidence of better outcomes with cognitive behavioural therapy as an adjunct to medication. Clinical psychology and pain specialists may also have a role to play in management. These patients often struggle to accept the diagnosis and are frequently convinced their pain is due to an underlying tumour. This can result in the involvement of multiple specialties in search of a diagnosis. They subsequently may receive invasive and unnecessary procedures with no significant improvement in their pain. Some interventional approaches have been used with some benefit including pulsed radiofrequency to the sphenopalatine ganglion.

Secondary headache disorder

Sinogenic

Facial pain is usually a mild complaint in CRS and facial pain alone is rarely caused by CRS. Facial pressure is deemed a more reliable symptom of CRS than pain per se. There also seems to be a difference in the prevalence of severity of headache and facial pain between patients with CRS with nasal polyps and those with CRS without nasal polyps. CRS without nasal polyps patients tend to have higher levels of pain and other nasal symptoms are more prevalent in CRS with nasal polyps patients. Sinogenic facial pain tends to arise from acute rhinosinusitis and is usually a deep, aching pain, which develops simultaneously with the onset of nasal symptoms, including nasal obstruction and coloured nasal discharge. Nasal endoscopy is essential and has a strong negative predictive value to exclude sinogenic pain. Facial pain should only be ascribed to CRS if there are other nasal symptoms, and concordant abnormalities on nasal endoscopy and/or imaging.

Management of acute rhinosinusitis and CRS is based on the 'European Position Paper on Rhinosinusitis and Nasal Polyps 2020' (EPOS2020) (see Chapter 36). Medical treatment for acute rhinosinusitis includes intranasal corticosteroids, decongestants (<10 days), saline spray/rinses, and antibiotics. Management of CRS is summarized in **Box 31.3**.

Significant caution should be exercised when considering endoscopic sinus surgery in those patients with facial pain due to the high long-term failure rates and subsequent identification of other causes for the pain in many cases. It is suggested that patients with facial pain without other nasal complaints or abnormalities on examination should not (primarily) be managed surgically.

Dental pain

Tooth-related facial pain is very common. Dental caries, infection, abscess, or periodontal disease can cause referred pain across the head and face. A secondary maxillary sinusitis can also result from dental infection and present with unilateral pain across the cheek, nasal congestion, coloured rhinorrhoea, and foul smell.

Dental review is required to address the underlying cause. This may involve drainage of dental abscess, removal of affected tooth, and oral antibiotics. If there is secondary maxillary sinusitis a CT scan and endoscopic sinus surgery may be indicated (**Figure 31.1**).

Temporomandibular joint

TMJ problems, including chronic myofascial pain, internal derangement, and degenerative joint disease (osteoarthritis), can cause facial pain and headache. Pain is usually a unilateral, dull ache in the preauricular area. Myofascial pain tends to be made worse by chewing or pressure on trigger points over the muscle. Internal derangement, usually due to displacement of the disc anteriorly, results in an audible or palpable click.

Treatment depends on the cause. Osteoarthritis of the TMJ should be treated with soft diet, non-steroidal anti-inflammatory drugs, and potentially surgery under the maxillofacial team. Myofascial

Box 31.2 Diagnostic criteria for persistent idiopathic facial pain (ICHD-3)

1. Facial and/or oral pain.
2. Recurring daily for more than 2 hours/day for longer than 3 months.
3. Pain has both of the following characteristics:
   - Poorly localized, and not following the distribution of a peripheral nerve.
   - Dull, aching, or nagging quality.
4. Clinical neurological examination is normal.
5. A dental cause has been excluded by appropriate investigations.
6. Not better accounted for by another ICHD-3 diagnosis.

Box 31.3 Chronic rhinosinusitis management (EPOS2020)

Appropriate medical treatment (AMT):

- Nasal steroids (sprays/drops/rinses).
- Saline rinses.
- Educate technique and compliance.
- Consider oral corticosteroids.

AMT (+/− long-term antibiotic) *or* functional endoscopic sinus surgery—CRS non-type 2, pus.

AMT (+/− oral corticosteroids) *or* functional endoscopic sinus surgery—CRS type 2, polyps.

pain can be managed with analgesia, corticosteroids, and local anaesthetic blocks. There is also a role for addressing contributory psychological factors and muscle stretching. If there is derangement of the joint, orthotic appliances may be an option. Surgery can be considered in refractory cases.

Medication overuse headaches

This is estimated to occur in 1–2% of the population worldwide and is more prevalent in women. The pain is dull, diffuse, and band-like and often starts early in the morning. The pathophysiology is not clear but can occur from regular overuse of headache medication such as ergotamine, triptans, opioids, non-steroidal anti-inflammatory drugs (including aspirin), and paracetamol. The headache usually resolves after the overused medication is stopped. There are specific diagnostic criteria (**Box 31.4**) and it is important to establish that the headache is not better accounted for by another cause.

Management involves good explanation of the diagnosis and breaking the cycle by withdrawing the overused medication. The medications should be stopped for at least a month. Patients may experience an initial worsening of their symptoms on withdrawal and headache improvement may not occur for weeks after discontinuation.

Neuropathic facial pain after surgery or trauma

Neuropathic pain can result following facial trauma or surgery, including dental procedures, endoscopic sinus surgery, or septorhinoplasty. The pain tends to be described as burning, gnawing, or stabbing in nature. There may also be altered sensation in the affected area. Treatment with gabapentin or pregabalin works well, but it needs to be taken for up to 2 months before assessing if it is effective. Local anaesthetic nerve blocks can be beneficial for localized pain. If the pain is unresponsive to medical treatment then help from clinical psychology, cognitive behavioural therapy, and the pain management team can be sought.

Controversies

Neurology referral

Many patients whose major complaint is facial pain will be seen by otolaryngologists. If the pain is not caused by CRS it is up to the treating clinician to what extent the treatment is carried out personally. A trial of initial treatment would seem reasonable based on the features ascertained from the history and examination, while in some centres a team-based approach may be available and beneficial. However, treating primary headache syndromes can be very challenging and may require multiple medications to succeed. It would therefore seem sensible to involve a specialized neurologist in those cases where first-line treatment fails.

Box 31.4 Diagnostic criteria for medication-overuse headache (ICHD-3)

1. Headache occurring on at least 15 days/month in a patient with a pre-existing headache disorder.
2. Regular overuse for more than 3 months of one or more drugs that can be taken for acute and/or symptomatic treatment of headache.
3. Not better accounted for by another ICHD-3 diagnosis.

Imaging

There is no correlation between the location of facial pain and imaging findings of anatomically related sinuses. Also, in CRS, patients reporting facial pain as their main symptom have less abnormalities on imaging than those with the other characteristic symptoms of CRS. Furthermore, it has been noted that facial pain scores in CRS are negatively correlated with CT sinus abnormalities.

Outside of an emergency setting, the risk of finding serious secondary pathology in patients with isolated headache and a normal neurological examination is the same as that in people who do not have headache. There is also significant potential for uncovering incidental findings which may not require further management but can cause increased anxiety.

As a result, routine CT scanning of patients with facial pain and headaches is not advised. A thorough history can reduce the need for imaging in most patients.

Guidelines

Due to the wide range of conditions that cause headache and facial pain there is not one set of clinical management guidelines. There are, however, several useful clinical guidelines. These include the British Association for the Study of Headache guidelines, National Institute for Health and Care Excellence guidelines for diagnosis and management of headaches, and more specific condition-based guidelines such as SIGN 155 on the pharmacological management of migraine. The EPOS2020 guidelines provide useful strategies for the clinical management of CRS and acute rhinosinusitis (**Box 31.3**).

Conclusion

Headaches and facial pain can be a diagnostic challenge due to the wide range of possible diagnoses and the often-false belief of patients that it is due to their sinuses. A structured approach including a detailed history, nasendoscopy, and imaging in selected cases should enable a diagnosis to be established and appropriate management implemented.

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32

Epiphora

Rohit D. Gohil and Ashok Rokade

Introduction

Epiphora—or the symptom of excessive tearing—is often a troublesome and sometimes upsetting symptom for patients. It can affect one or both eyes with or without other symptoms. It is estimated that nasolacrimal obstruction can affect 10% of the population at 40 years of age, rising to approximately 40% at 90 years. Studies have shown a female preponderance in epiphora (55% of patients).

These patients have historically been managed in the first instance by ophthalmology colleagues, where an assessment is performed and a diagnosis is given for the cause of epiphora. In the situation where the cause is blockage of tear transit from the surface of the eye to the nose, then the otolaryngologist may be consulted for their input in surgical management.

Recently many centres have adopted a combined multidisciplinary approach in managing patients suffering from epiphora. They can be seen, examined, and potentially listed for surgery in a one-stop setting. Often the surgery too is carried out jointly. This allows for increased productivity, while encouraging an open forum for combined decision-making in difficult cases.

In this chapter we outline the basic science of lacrimation, the causes of epiphora, its assessment and subsequent management, and finally controversies in the surgical management of epiphora.

Basic science

Tears initially form within the lacrimal gland situated in the orbit—anteriorly in the superolateral quadrant. The gland occupies the lacrimal fossa, a depression in the orbital plate of the frontal bone. It is approximately 2 cm in length and is bilobed: the *orbital* lobe is the larger of the two and contacts the lateral margin of the levator palpebrae superioris muscle; the *palpebral* lobe occupies the inner surface of the eyelid.

The gland itself receives its vascular supply from the lacrimal artery—a branch of the ophthalmic artery which branches from the internal carotid. Its venous drainage is via the superior ophthalmic vein to the cavernous sinus via the superior orbital fissure.

The stimulation of lacrimal fluid secretion is via the parasympathetic nervous system. Preganglionic fibres run from the nervus intermedius onto the greater superficial petrosal nerve followed by the nerve of the pterygoid canal where they synapse at the pterygopalatine ganglion. Postganglionic fibres travel with the maxillary nerve and then the zygomatic nerve. A communicating branch to the lacrimal nerve ends at the gland itself. The inhibition of fluid secretion is mediated by sympathetic control, with fibres arising from the superior cervical ganglion, travelling in the internal carotid plexus and thereafter the deep petrosal nerve. The deep petrosal and greater superficial petrosal nerves combine to form the nerve of the pterygoid canal, where sympathetic fibres follow the same route as parasympathetic fibres (**Figure 32.1**).

The gland produces lacrimal fluid in acinar cells that travel to the surface of the eye via excretory ducts that open at the superior conjunctival fornix—a reflection of conjunctiva back upon itself from the surface of the eye to the eyelid. The fluid is then distributed over the surface through blinking—this mechanism is referred to as the 'lid pump'.

Once the tear film is distributed over the eye, it consists of three distinct layers: lipid, aqueous, and mucus. The lipid layer is outermost and produced by the Meibomian glands of the eyelid—it serves to reduce the rate of tear film evaporation and provides surface tension to the film. The aqueous layer is produced by the lacrimal gland itself and forms the majority component of the tear film. The innermost mucus layer allows the film to be effectively distributed by blinking and allows efficient movement of the tear film medially—the mucus is produced by the conjunctiva (**Figure 32.2**).

The tear film constantly migrates medially and accumulates as the lacrimal lake at the medial canthus. The lacrimal lake drains via two puncta, medially with one at each eyelid, 6 mm lateral from the canthus itself (**Figure 32.3**). The superior and inferior puncta open into their respective canaliculi that drain perpendicular to the free edge of the lid, before turning medially and running parallel to the margin. The inferior canaliculus drains 60% of the fluid of the lacrimal lake. After 10mm and now medial to the medial canthus, the superior and inferior canaliculi unite to form the common canaliculus. The common canaliculus is approximately 4 mm in length and opens into the lacrimal sac at its posterolateral surface. The lacrimal sac itself sits within a bony fossa of the medial orbital wall composed of lacrimal bone and frontal process of maxilla and is bounded anteriorly and posteriorly by the respectively named bony lacrimal

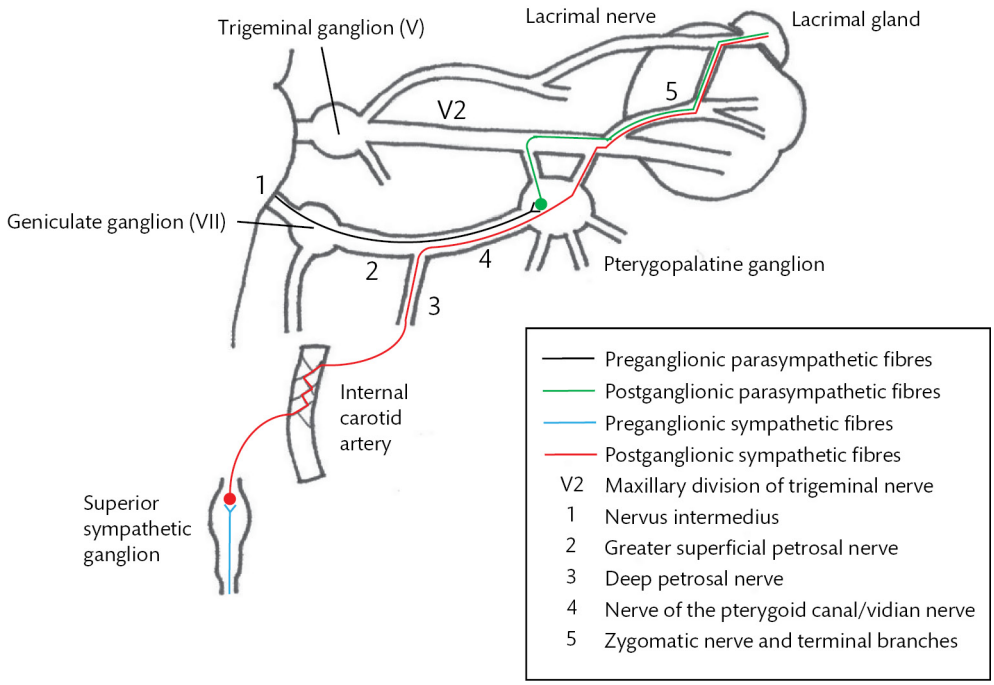


Figure 32.1 Autonomic nervous supply to the lacrimal gland.

crests. The lacrimal sac then descends inferiorly as the nasolacrimal duct—again shielded by the frontal process of maxilla and anterior to the leading edge of middle turbinate. The duct is finally described as taking a sharp turn medially to open into the inferior meatus, approximately 12–15 mm posterior to the leading edge of inferior turbinate—via the valve of Hasner (or plica lacrimalis). The tear fluid thereafter transits posteriorly along with native nasal mucus via mucociliary action (Figure 32.4).

Causes of epiphora

An imbalance in the system of production and drainage may lead to epiphora. The aetiology of epiphora can be divided into whether the patient is suffering from reflex tearing or has reduced tear drainage.

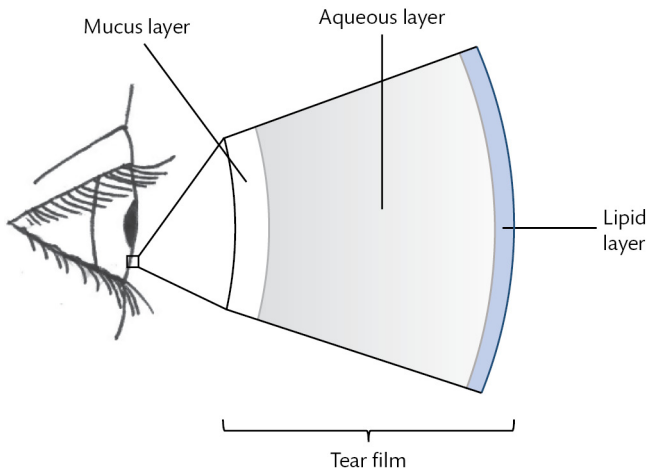


Figure 32.2 The layers of the tear film.

Greater production compared to loss directly results in tearing; however, loss in excess of production can lead to dryness and reflex tearing, the reflex being mediated by the Vth and VIIth cranial nerves.

Reflex tearing may be a consequence of dry eye, but also occurs in inflammation, allergy, tear film problems, or disorders of the surface of the eye. Dry eye has been found to be the issue in 40% of patients referred with epiphora. Issues relating to the tear film can result in hypersecretion—such as over-evaporation and tear film instability due to a poor lipid layer contribution. The use of topical lubricants can improve symptoms in the majority of these cases.

Reduced tear drainage can be a consequence of lid pump dysfunction—such as eyelid laxity, or an obstruction at any point in the nasolacrimal drainage system. Obstruction at the level of the nasolacrimal duct can be acquired or congenital and there may be a single cause, or multifactorial. Therefore, adequate expert assessment is necessary to discern the aetiology of the epiphora and either parallel or sequential treatment may be necessary to address the problem effectively.

Functional epiphora is a term given to the complaint of excessive tearing due to lacrimal drainage dysfunction with other causes being ruled out, but upon testing the system is patent.

Figure 32.5 illustrates the causes of epiphora with respect to the anatomy and physiology of the lacrimal system.

Assessment and investigation

The initial assessment of a patient complaining of excessive tearing is usually undertaken by an ophthalmologist; however, there is great value in the otolaryngologist being involved in the process when undertaking combined cases and clinics in managing epiphora.

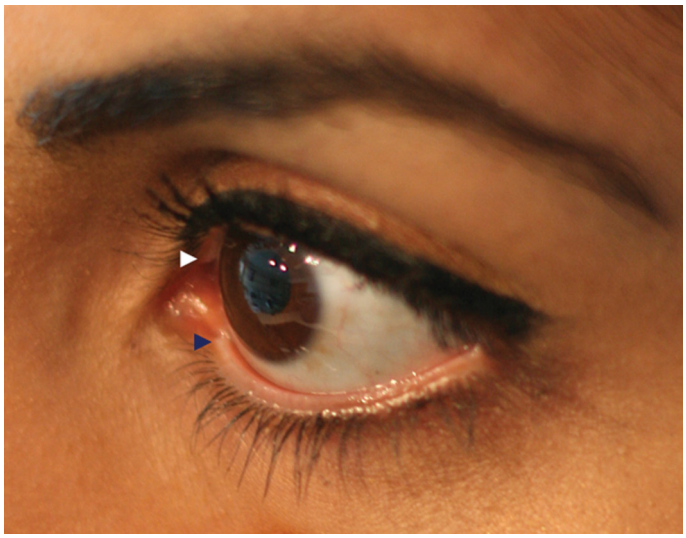


Figure 32.3 Clinical photograph illustrating the positions of the superior (white arrowhead) and inferior (navy blue arrowhead) lacrimal puncta of the left eye.

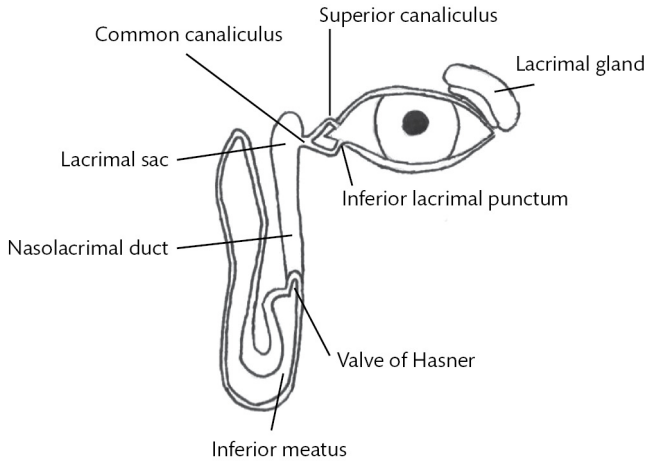


Figure 32.4 A schematic diagram of the lacrimal system.

A thorough history covering the points mentioned in Table 32.1 is essential in helping to determine the underlying cause for a patient's epiphora.

The history can also be complemented with a symptom-reporting questionnaire which can be useful in giving a numerical score that can be repeated after intervention to exhibit and document benefit. The authors use the Lac-Q questionnaire which enquires into eye-specific and social impact factors. Thereafter, the total score (the Lac-Q score) is recorded. This has been shown to be a useful tool in assessing outcomes after nasolacrimal surgery with regard to validity and reliability (Figure 32.6).

Thereafter the assessment requires examination and further investigation if deemed necessary and is again usually undertaken by the ophthalmologist, if not jointly.

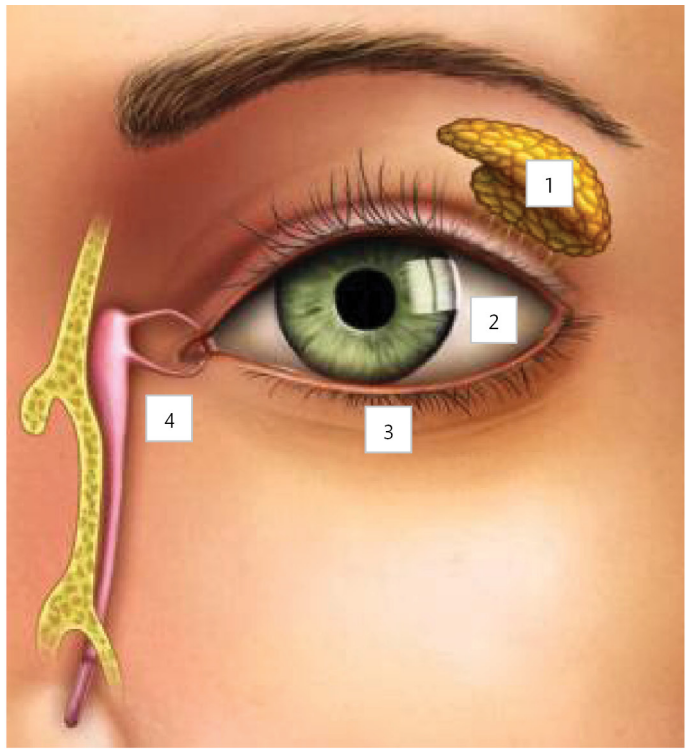
Initially inspection is necessary to observe any facial/periorbital asymmetry, especially at the eyelids, any discharge, fistulae, or scars. The surface of the eye is assessed along with the blink reflex—looking especially for lagophthalmos (incomplete closure of the eye). Slit lamp examination allows for detailed assessment of the surface along with appreciation of the size of the tear meniscus that can indicate nasolacrimal duct obstruction. Additionally, the *tear break-up time* can be assessed at the time of slit lamp examination. Here, the patient has fluorescein instilled into the eye such that the visible ocular surface appears green/yellow—the patient is then asked to blink and the time between blinking and a black spot to appear on the surface is recorded. The black spot represents drying of the ocular surface and regression of the fluorescein. The average time is quoted to be between 10 and 35 seconds—any sooner can indicate tear film instability.

Palpation of the lacrimal sac region may be fluctuant and produce pus/mucus from the puncta or a fistula and will indicate dacryocystitis. Any firmness or irregularity can indicate a neoplasm. Lid laxity is determined by pulling the lid laterally—it is deemed to be excessively lax if stretched more than 8 mm and/or it takes longer than 8 seconds to return to its normal position. However, the lid may not naturally oppose the eye at rest in severe cases.

Obstruction can be tested for through a number of techniques. The dye disappearance test involves the instillation of 2% fluorescein onto the surface of the eye and inspection after 5 minutes. An excess of fluorescein at the lacrimal lake or asymmetric clearance indicates an obstruction. This test is more sensitive in diagnosing persistent or severe obstruction but may not pick up intermittent or distal/nasal causes for obstruction. The *Jones 1* test observes fluorescein clearance under physiological conditions administered as described above, but with a cotton bud placed in the inferior meatus—where the aim is for fluorescein to be recovered after 3–5 minutes. The *Jones 2* test is non-physiological and is usually performed after a failed Jones 1 test. The test involves washing away residual fluorescein and irrigating saline into the inferior canaliculus. A positive test shows dye at the cotton bud and indicates partial nasolacrimal duct obstruction. A negative test does not recover any dye and indicates total obstruction or a defective pump mechanism. Finally, lacrimal system irrigation may yield useful information on its own. This involves direct canalicular irrigation, with or without dilatation of the puncta. Resistance, reflux, delay, or lack of clearance into the nasal cavity (reported by the patient) allows the assessor to determine the level of any obstruction. Resistance indicates the severity of obstruction; reflux through the same canaliculus suggests canalicular obstruction, while reflux through the opposing puncta indicates either common canalicular or more distal obstruction.

In the authors' experience, simultaneous nasal endoscopy improves diagnostic accuracy of nasal irrigation with further help in the diagnosis of any coexistent or causative nasal problems that may be contributing to the epiphora. Rigid nasal endoscopy will also prove useful in determining access to the level of the nasolacrimal duct in cases where endonasal dacryocystorhinostomy (DCR) is being planned. Findings such as nasal polyps, masses, or septal deviations will have an impact on the type and timing of management.

Additional investigations include contrast dacryocystography and dacryoscintigraphy that are useful in evaluating anatomy along with function of the lacrimal system. Dacryocystography involves the injection of dye into the lacrimal system followed by imaging and digital subtraction analysis. Dacryoscintigraphy gives physiological information by tracking the transit of radionucleotide drops using a scintigram. These techniques are used less now due to the



Cause	Neural/hypersecretion (1)	Tear films/surfaces causes (2)	Lid appositional problems (3)	Distal obstruction (4)
Comment	Unusual or rare causes	Many reasons, some examples below	Good contact between lid and globe required to achieve balance in controlling evaporation, ensuring tear migration and direction to puncta	Obstruction occurring at any point between puncta to nasal cavity
Examples	Parasympathetic compression/local tumour growth	Chronic blepharitis	Ectropion	Iatrogenic (surgery, radiotherapy)
	Aberrant cranial nerve VII regeneration following trauma	Keratoconjunctivitis sicca	Entropion	Trauma
	Dacryoadenitis	Trichiasis	Floppy eyelid syndrome	Acute dacryocystitis
	Cholinergic agonists	Allergic conjunctivitis	Punctual eversion	Chronic infection (e.g. *Actinomyces*)
		Exposure keratopathy	Trauma	Autoimmune (e.g. pemphigoid)
		Iatrogenic (medicine side effects)	Post-surgical resection	Granulomatous disease
			Cranial nerve VII palsy	Chronic nasal disease/inflammation
				Neoplasia
				Involutional stenosis
				Idiopathic

Figure 32.5 The causes of epiphora.

Table 32.1 Points to be covered when taking a history from a patient complaining of epiphora

Age	The reporting of epiphora is known to increase with advancing age, however it can be present from birth. The latter represents congenital epiphora and is present in 20% of the neonatal population. Of these, the overwhelming majority will resolve in the first year of life. The minority of children for whom this persists will need further investigation and management
Laterality	Does tearing only affect one eye, or both? In unilateral cases, one should be alerted to ruling out sinister underlying causes for epiphora either primarily, within the lacrimal system, or within the nasal cavity. Red flag symptoms include blood-stained nasal discharge and bloody tears with or without pain. Bilateral epiphora is often more likely to be associated with a systemic cause
Continuous or intermittent?	Continuous tearing is more likely to be associated with a severe obstructive cause, and the intermittent reporting of symptoms may represent a functional problem. The severity of symptoms reported will be worse in colder climates and seasons due to increased evaporation and reflex tearing phenomena
Where do the tears form?	If tears are collecting at the medial canthus, this is more likely to represent an obstructive cause, whereas if tears are forming more laterally, this may indicate an issue with lid apposition, tear film, or surface abnormality
Ocular symptoms	This will help to deduce whether epiphora is due to reflex tearing or lid problems. Questioning includes enquiring about eye trauma; irritant exposure to the eyes; itching; sticky/purulent discharge; burning and pain; crusting and redness. Questions about swellings next to the lateral nasal wall may suggest dacryocystitis or a dacryocystocele
Nasal symptoms	In particular, symptoms such as nasal obstruction, congestion, rhinorrhoea, anosmia or hyposmia, and episodes of acute sinusitis should be enquired about. The presence of nasal disease can represent distal obstruction of the lacrimal system, particularly that affecting the inferior meatus. Again, caution must be taken if the patient complains of unilateral nasal symptoms along with unilateral epiphora—this should be investigated urgently
Previous surgery	Previous intervention either of the lacrimal system or surrounding structures, such as septorhinoplasty, functional endoscopic sinus surgery, craniofacial or maxillofacial surgery should be enquired about. The relative proximity of the lacrimal system to these areas means that postoperative swelling may cause either temporary epiphora or persistent tearing in the case of inadvertent trauma
Previous trauma	Previous trauma to the facial skeleton may result in epiphora. This is usually short-lived and resolves with conservative management. However, for those where it persists, intervention is required. In these cases, definitive management of epiphora should be delayed while the initial injury heals in case of spontaneous resolution
Systemic symptoms	Epiphora may be a presenting complaint for an underlying systemic condition such as granulomatosis with polyangiitis. Therefore, local symptoms of nasal crusting, pain, discharge, and bleeding, along with systemic features, such as pulmonary, renal, urinary, and cutaneous symptoms, should be enquired about in order to guide diagnosis
Full medical history	A full medical history including medication, smoking, and occupational history will also be helpful in determining aetiology

advent of alternative imaging techniques and relative usefulness and simplicity of lacrimal irrigation.

Imaging using either computed tomography (CT) or magnetic resonance imaging (MRI) is useful in determining anatomy following surgery or trauma, or in congenital deformity, or if concurrent sinonasal disease is diagnosed. The modality is also essential in investigating potential neoplasia. CT gives useful information on bony anatomy and surrounding structures including the anterior skull base, orbit, and frontal sinuses. MRI is superior in soft tissue delineation. CT or/and MRI evaluation may also be useful in the management of concomitant sinonasal disease.

Treatment

In order to successfully manage a patient with epiphora, the clinician needs to accurately determine the underlying cause or causes. Alongside a diagnosis, an active discussion with the patient about the underlying cause but also the expectations and limitations, including risks of potential interventions, should be had.

The simplest management is the conservative approach. This may prove to be the best approach in patients that have fairly mild symptoms and just require an answer to the cause of their complaint. Other reasons for adopting a conservative approach would be the relative risk of alternative treatments, especially if general anaesthetic is required in a patient with multiple comorbidities. An intervention under local anaesthetic may be an option in such patients however.

The use of medications to address excessive tearing may be initiated by the ophthalmologist depending on the underlying diagnosis. This is especially the case in non-obstructive diagnoses and goes beyond the scope of this chapter. If, however, underlying nasal pathology is contributing to the patient's complaint of tearing, treatment may be necessary including the use of a steroid nasal spray, drops, an antihistamine, and/or a saline nasal rinse.

The surgical management of epiphora is usually reserved for obstructive diagnoses and is determined by the level of obstruction. However, the insertion of punctal plugs may be performed by the ophthalmologist for managing some cases of dry eye. Punctal plugs are a reliable and safe method of preserving the tear film on the ocular surface. They can be either absorbable or non-absorbable (requiring removal if necessary), but also come in many varieties, occupying the punctum or the canaliculus, with either complete or partial occlusion depending on the indication. The applicability of punctal plugs in the setting of dry eye is also beyond the scope of this chapter. Otherwise, the surgical procedure usually required to relieve obstruction is called a DCR.

DCRs were previously performed by an ophthalmologist through an external approach involving an incision on the face lateral to the

Lac-Q - The Lacrimal Symptom Questionnaire **Name:** **Number:** **Date:**

Social and lifestyle impact of tear duct problem

Which of these five statements is true about the tear duct problem overall in the last eight weeks?
Please tick the box next to any true statement.

- Friends or family have commented about the watery eye problem. ☐
- The watery eye problem has caused embarrassment in company. ☐
- The watery / sticky eye problem has interfered with everyday activity, for example (underline each that applies):
  Reading .. Driving .. Wearing make-up
  Wearing glasses .. Hobbies
  Other activity (specify): ☐
- The vision is sometimes blurred because of the watery / sticky eye problem. ☐
- Medical attendance: visit to the family doctor's surgery, or the hospital eye clinic, because of tear duct problem. ☐

(Scoring: score one point for each box ticked, maximum score =5)

Total score for social impact: ☐

Problems with each eye separately

For each of the four problems (watery eye, pain, sticky eye or swelling), put a tick in the box next to the statement which best describes the situation over the last eight weeks.
Use the left hand column for the left eye, and the right hand column for the right eye.

	Left		Right
• Watery eye			
No watery eye problem	☐	0	☐
The eye waters occasionally, mainly outdoors	☐	1	☐
Troublesome watering of the eye, indoors and outdoors, some days	☐	2	☐
Troublesome watering of the eye most days	☐	3	☐
Troublesome watering of the eye every day	☐	4	☐
• Pain in or around the eye; soreness of eyelids			
No pain	☐	0	☐
Some pain or soreness, but has not sought medical advice or treatment	☐	1	☐
Pain or soreness, has used prescription eyedrops	☐	2	☐
Painful and swollen (lacrimal abscess), requiring antibiotics or surgical drainage	☐	4	☐
• Sticky eye			
No problem with sticky eye	☐	0	☐
The eye is sometimes sticky in the mornings	☐	1	☐
The eye is sticky every day in the mornings	☐	2	☐
The eye has sticky or mucous discharge throughout the day	☐	3	☐
There is infected discharge leaking through the skin of the lower eyelid (fistula)	☐	4	☐
• Swelling or lump at the medial canthus (mucocoele)			
No swelling or lump	☐	0	☐
Swelling present, but only intermittently	☐	1	☐
Swelling present all the time	☐	2	☐
(Scoring: use numbers in central column)	**Total scores for each eye:**	☐	☐

Lac-Q score (sum of three total scores): ☐

Figure 32.6 The Lac-Q questionnaire.
Reproduced from Mistry N, Rockley TJ, Reynolds T, Hopkins C. Development and validation of a symptom questionnaire for recording outcomes in adult lacrimal surgery. *Rhinology*. 2011;49(5):538–545.

the attendant facial scar was less appealing. There has been a recent trend towards performing endoscopic or endonasal DCR, in conjunction with the otolaryngologist, thus avoiding the skin incision, but with the advantage of being just as successful as the external approach. Table 32.2 outlines the complications of endonasal DCR that the patient should be counselled on prior to undergoing surgery.

Figure 32.7 illustrates the authors' preferred method for performing endonasal DCR. In some centres it is usual practice to place nasolacrimal tubes (O'Donoghue tubes) from the canaliculi through the rhinostomy and secure them intranasally. These are kept *in situ* for between 2 and 6 weeks before being removed in clinic as part of a routine multidisciplinary follow-up.

Postoperatively, the patient will be encouraged to rinse their nose with saline along with topical eye drops, such as chloramphenicol or Betnesol 0.1%, which is determined by the ophthalmologist.

At follow-up the patient's puncta and rhinostomy will be assessed and tubes may be removed if they are *in situ*. Confirmation

Table 32.2 List of immediate, early, and late complications of endonasal dacryocystorhinostomy

Immediate/operative	Early: up to 4 weeks postoperatively	Late: up to 6 months postoperatively
Bleeding	Pain	Rhinostomy scarring
Orbital fat exposure	Adhesions	Rhinostomy stenosis
Periorbital bleeding	Nasal crusting	Persistent nasal crusting
Lacrimal sac trauma	Infection	Nasal adhesions
	Scarring/fibrosis of rhinostomy	Lacrimal sump syndrome
	Granulations at rhinostomy	
	Subluxation of tube into eye	
	Migration of tube into nasal cavity	
	Corneal abrasion	
	Punctal cheese-wiring	

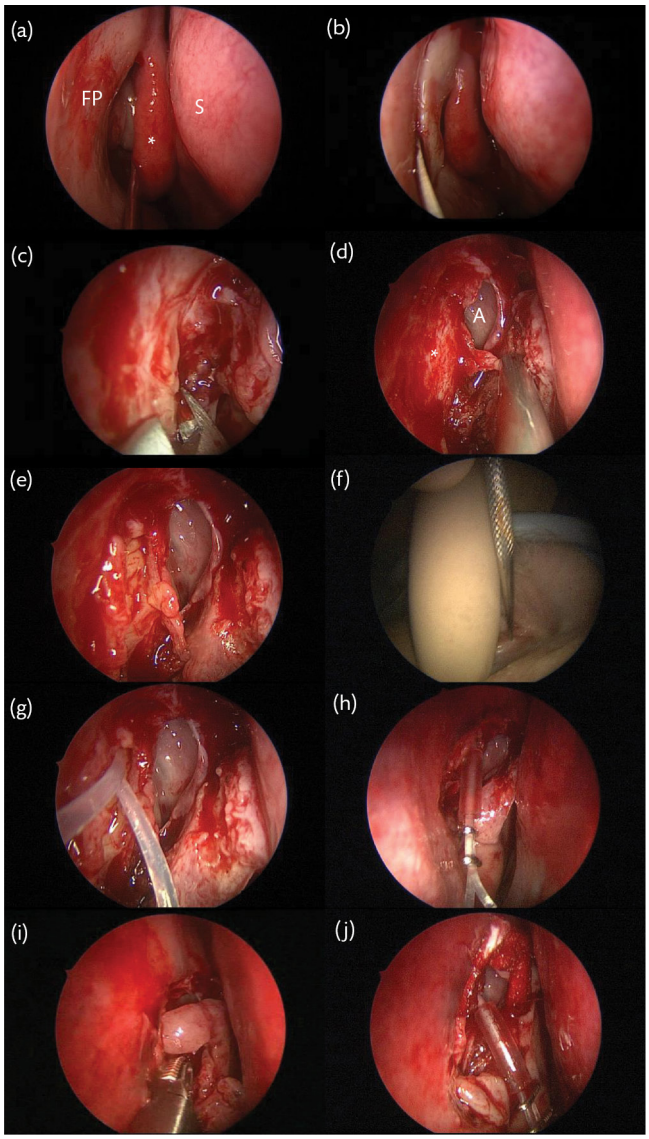


Figure 32.7 Intraoperative images of a right-sided endonasal DCR. (a) Normal intranasal anatomy. *, middle turbinate; FP, mucosa overlying frontal process of maxilla; S, nasal septum. (b) A 'C'-shaped posteriorly based nasal mucoperiosteal flap is raised over the frontal process of maxilla region. (c) The bone of the frontal process is then removed using a Kerrison rongeur and a bur. (d) The nasolacrimal duct has been exposed (*) and the associated agger nasi cell has been opened (A). (e) A vertical incision into the nasolacrimal duct exposes its lumen; this is formalized with anterior and posterior release incisions to define the rhinostomy further. (f) The superior and inferior lacrimal puncta are cannulated with O'Donoghue nasolacrimal tubes after being dilated with a Nettleship dilator. (g) DCR tubes are delivered into the nose. (h) The tubes are secured with a Watski sleeve and two liga-clips. (i) Before replacement the nasal mucosal flap has its central portion removed to mirror the size of the nasolacrimal duct opening. (j) Once positioned, the flap is overlain with Gelfoam dressing.

of nasolacrimal patency can be assessed by application of topical fluorescein eye drops and observation of its appearance through the rhinostomy.

In some cases, it may not be possible to perform a conventional endonasal DCR. This may be due to severe canalicular stenosis or that the quality of the anatomy following trauma, surgery, or radiotherapy, for example, will not allow for it. In these cases, an alternative procedure can be performed to bypass the lacrimal drainage system via a conjunctivo-dacryocystorhinostomy. In these cases, a fistulous tract is created from the caruncle at the medial canthus to the region of the axilla of the middle turbinate. Thereafter, a ceramic tube (a Lester Jones tube is used by the senior author) is placed through the fistula. In order to aid positioning, the leading edge of the middle turbinate may be resected. The appropriate size and shape of prosthesis is selected to ensure the tube is flush at the conjunctiva and does not contact the septal mucosa endonasally. Some varieties of tube are bevelled to achieve this; others are flanged to avoid excessive movement (**Figure 32.8**).

Similarly, in cases where epiphora is caused by punctal stenosis, an oculoplastic surgeon may consider a procedure to improve punctal patency. This can be by simply dilating the punctum itself by cannulating the proximal drainage system with probes of enlarging calibre, or by incising into and enlarging the orifice using the two-snip/three-snip punctoplasty technique (**Figure 32.9**). Similarly, lid repositioning surgeries, details of which are beyond the scope of this chapter, can be offered in appropriate cases by the oculoplastic team.

Controversies

Surgery of the nasolacrimal system is constantly evolving and, therefore, a number of inconsistences exist in practice for which there is a paucity of evidence. We will outline below some of these controversies.

Open versus endonasal dacryocystorhinostomy

DCRs were originally performed through an external approach, which was considered the gold standard. Although this is an effective procedure for addressing obstructive epiphora, it does involve a scar on the face that may be unappealing for the patient. The advent of endonasal DCR meant that in many centres it became the preferred approach—however, enthusiasm for the external approach persists in some centres. The benefits of the external approach mean

Figure 32.8

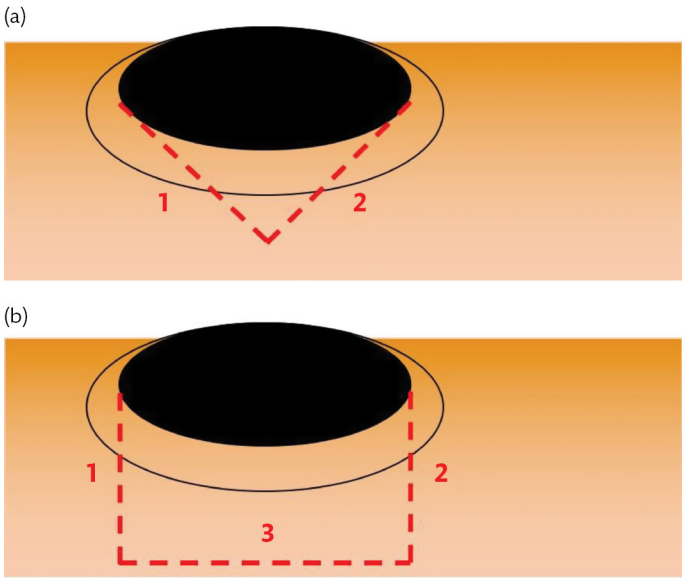


Figure 32.9 Diagrams of the punctum of the inferior lid illustrating two punctoplasty techniques. (a) Two-snip punctoplasty—using Vannas-type microscissors, two diagonal conjunctival incisions are placed, one at the lateral and one at the medial extent of the ampulla aiming to join inferiorly at a median axis. The two cuts therefore join and a 'V' of tissue is excised reducing the amount of canaliculus disruption. (b) Three-snip punctoplasty—two cuts are made vertically at the medial and lateral extent of the punctum and a third incision employed to remove intervening scarred tissue. There are a number of variations in the three-snip technique; however, this rectangular snip approach is the most widely practised.

that no endoscopic skills or separate surgical teams are required. However, along with the potential scar, it is not possible to address any coexisting sinonasal pathologies through this method. Table 32.3 lists the advantages and disadvantages of both techniques. In most settings endonasal DCR requires input from two teams with expertise in endoscopic surgery and lacrimal probing.

Mitomycin C

Mitomycin C is an antiproliferative drug that has been used as a chemotherapeutic agent both intravenously and topically in the management of various cancers of the gastrointestinal tract, bladder, and breast, among others. It has also been used in non-oncological settings to limit scarring, such as in glaucoma and after subglottic stenosis surgery. The topical application of mitomycin C to the rhinostomy following DCR has been shown to improve functional outcomes and patency rates in a number of studies. However, these studies were confounded by patient demographics, case selection, revision cases, surgical expertise/technique, and drug dosage. Though data do point in favour of mitomycin C safely improving outcomes, there is not enough evidence that this is achieved independently of all other factors and, therefore, more research is required to answer this question.

Tubes or tubeless?

The placement of silastic tubes through the lacrimal drainage system following DCR is commonplace in many centres. The rationale is that even the temporary placement of tubes will promote healing through scarring such that a more patient fistula is achieved before removal. However, others argue that the foreign body itself is the cause for failure. Many patients undergo DCR without the placement of tubes and similar rates of control are achieved for their epiphora. Studies have found that there was no perceived benefit for tube placement in primary DCR. By avoiding tube placement, surgical time is reduced and the risks of having a tube *in situ* and the need for its removal are avoided. There is similarly a lack of evidence for the benefit of tube placement in revision cases, but it is the senior author's preference to do so in this setting.

There is also no evidence to suggest benefit for tube use in cases with concomitant sinonasal disease.

Preservation of mucosal flaps

A number of techniques exist in performing endonasal DCR. One point of difference is either the preservation or resection of mucosa when fashioning a rhinostomy. As outlined previously, the senior author adopts a mucosa-preserving technique for both the nasal and lacrimal mucosal flaps. This has been shown to reduce the granulation rate in the healing phase and may be associated with better functional outcomes. Reviews have shown that there was no difference in complication rate between the two techniques but again, there is only limited literature available at present and this is a point for further research.

Tips and pearls

- The causes of epiphora are numerous and it may be due to single or multiple factors including concomitant nasal disease.
- Epiphora may be due to reflex tearing/dry eye or obstruction of lacrimal drainage.
- A multidisciplinary approach involving both ophthalmology and otolaryngology specialists improves efficiency and decision-making, and may enhance patient experience.
- Along with appropriate history and examination, imaging has a role in select cases in achieving a diagnosis.
- Treatment is tailored to the underlying diagnosis and is conservative, medical, or surgical.

Table 32.3 Advantages and disadvantages of external and endonasal dacryocystorhinostomies

	Endonasal DCR	External DCR
Advantages	No external scar Can concurrently manage nasal pathology No disturbance to canthal anatomy Combined approach in complex cases	No endoscopic skills needed Single operator (ophthalmologist)
Disadvantages	Endoscopic skills Lacrimal probing skills	Facial scar Nasal issues cannot be addressed

- Surgical management usually involves a DCR that is performed endoscopically, often as a combined case between ophthalmology and otolaryngology.
- Many differences of opinion and controversies exist in the surgical management of epiphora and should be the subject of further research.

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33

Nasal obstruction

Peter Andrews and Alfonso Luca Pendolino

Definition and prevalence

Nasal obstruction (NO), also described as 'nasal blockage' or 'nasal stuffiness', is a subjective feeling of discomfort caused by difficulty breathing through the nose. It represents one of the commonest presenting ear, nose, and throat (ENT) symptoms in primary care and one of the commonest ENT symptoms referred into secondary care. It affects over 30% of the adult population in the UK and is estimated to have an annual National Health Service and societal cost of £18 billion.

Physiology of nasal airways

Functions of the nose

The nose has two main, interlinked, functions: air conditioning and olfaction. The important function centred around 'air conditioning' has evolved and adapted over millennia and involves air filtration, humidification, and warming during a single breath. This enables inspiratory air to reach the lungs in an optimal state and maximizes the process of respiration. In addition, some of the inspired air is directed towards the roof of the nose to facilitate olfaction. In humans, olfactory epithelium is located superiorly within the left and right nasal cavities, divided by the midline nasal septum, in two narrow areas named olfactory clefts. They contain the olfactory receptor cells, specialized bipolar neurons which are responsible for the detection of odorants. These cells have a long central process that form the fila olfactoria, which pass through the anterior skull base and directly connect to the olfactory bulb within the anterior cerebral cortex.

As to why we have two nasal cavities or nostrils is open to debate; however, it offers the ability to localize smell through left and right differentiation. This is not dissimilar to having two eyes and two ears, which enable distance and direction calculation, thereby locating the 'source of smell' and perhaps providing an evolutionary survival advantage. An additional hypothesis focuses on the need to rest one side of the nasal cavity while the other functions, which ensures regular recovery and repair of the respiratory nasal mucosa in keeping with the natural nasal cycle (see below). This is particularly beneficial in heavily polluted environments.

Nasal airflow route

The nose plays an important role in breathing by providing an airflow resistance which is essential for good pulmonary function. Particularly, the nose is responsible for almost two-thirds of this resistance with most of it occurring in two main areas of the nose, called nasal valves, that act as a flow-limiter. These are divided into an internal (INV) and external nasal valve (ENV)—the former is a space made up of the nasal septum medially, upper lateral cartilages superolaterally, floor of the nose, and head of the inferior turbinate inferiorly which fluctuates in size during the day; the ENV, although strictly not a true 'valve', is a cross-sectional area of the nasal ala or opening of the nasal vestibule (**Figure 33.1**). The INV, in particular, plays an important role in controlling nasal airway resistance (NAR).

Nasal airflow during normal nasal breathing passes through the ENV in a laminar flow pattern with an approximate velocity of 2–3 m/s. Air then passes through the INV, the narrowest part of the nasal cavity, and speeds up significantly to 12–18 m/s whereby it becomes more turbulent. The speed then reduces to 2–4 m/s. The majority of inspired nasal airflow runs inferiorly along the nasal cavity with 10% of flow directed superiorly towards the olfactory region with each breath under resting conditions. The middle turbinate acts as an air foil: changing direction of airflow and also splitting the airstream superiorly into the olfactory cleft. Sniffing increases the percentage of inspired air directed towards the olfactory area by up to 20% (250 mL), making the perception of smell more appreciable. Air turbulence is key to the 'air conditioning' function of the nose which enables lateralization of air onto the nasal turbinates. Gaseous interaction between air and nasal mucosa ensures the air is warmed, humidified, and filtered through interaction with ciliated respiratory mucosa.

Autonomic regulation and perception of nasal airflow

Nasal mucosa consists of a vast network of venous sinusoids which form a dense subepithelial plexus throughout the nasal cavity and provide a reservoir of capacitance vessels. These are able to hold a large volume of blood during the *congestion* phase and, equally, are able to expel this large volume of blood during the *decongestion* phase. The process of congestion and decongestion is under autonomic control and ensures constant total NAR and facilitates the important action of turbulent airflow.

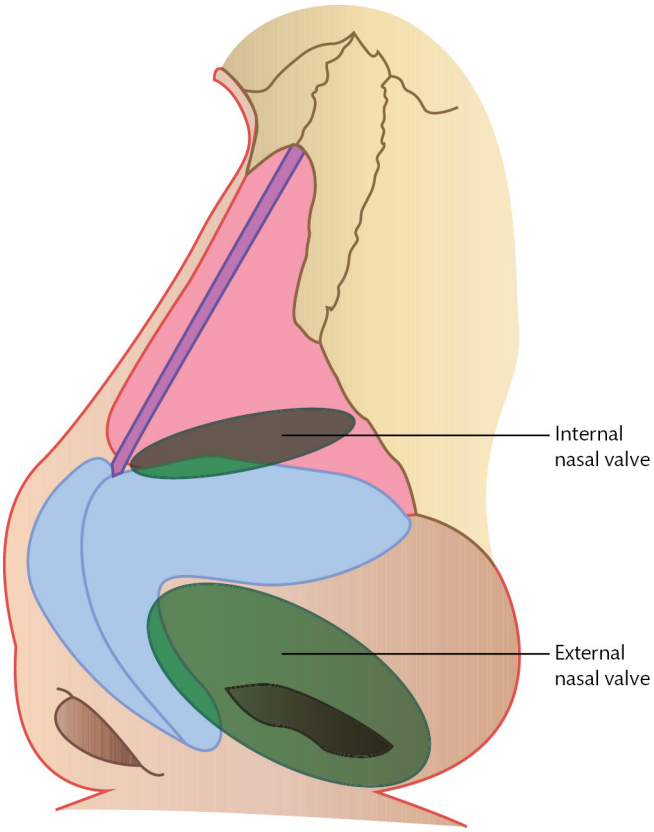

Figure 33.1 Internal and external nasal valves.
Image courtesy of Mr Alfonso Luca Pendolino (Consultant Rhinologist and ENT Surgeon, Imperial College Healthcare NHS Trust).

Arteries and veins of the nasal mucosa have a rich nerve supply made up of both sympathetic and parasympathetic termination. Preganglionic sympathetic fibres originate in the thoracolumbar region of the spinal cord. They pass into the vagosympathetic trunk to relay in the superior cervical ganglion, through the internal carotid plexus. From here, some postganglionic fibres form the deep petrosal nerve, which joins the nerve of the pterygoid canal and passes through the pterygopalatine ganglion without any relay thus reaching the nasal mucosa through the caudal nasal nerve. Other postganglionic fibres may reach the nasal mucosa via the sphenopalatine nerve and the major palatine divisions of the maxillary nerve. Preganglionic parasympathetic fibres arise in the superior salivatory nucleus in the brainstem and follow the greater superficial petrosal nerve. They then course with the vidian nerve to relay in the pterygopalatine ganglion, from which postganglionic fibres are distributed to the nasal mucosa with the posterior superior, nasopalatine, posterior inferior, and greater palatine nerves.

Sympathetic input causes vasoconstriction and empties the venous sinusoids thereby increasing nasal airflow, whereas stimulation of the vidian nerve causes an increase in NAR. However, physiologically there is a predominant sympathetic control of the capacitance vessels and nasal mucosa normally receives a tonic sympathetic discharge, which has led to the assumption that mucosal congestion may be more related to a withdrawal of sympathetic discharge rather than to an overactivity of parasympathetic nerves.

The perception of nasal breathing or airflow is mediated and controlled through somatosensory afferents of the trigeminal nerve. As resistance and pressure increase, somatosensory afferents are triggered resulting in the sensation of NO. However, these afferent fibres may become neuralgic or dysfunctional, perhaps secondary to surgery or trauma, resulting in abnormal impulse production even in the presence of a normal nasal airway.

There is an important requirement to maintain nasal resistance as this provides the desired feedback during inspiration and expiration. It is very important to avoid over-resection of sinonasal anatomy, mainly of the nasal turbinates, thereby creating the 'empty nose syndrome'. Patients with large, typically postoperative nasal cavities may still perceive a sensation of nasal blockage. Equally, patients with a significant deviated nasal septum (DNS) sometimes do not complain of NO.

The ability of nasal mucosa to respond to a nasal decongestant is profound and has an immediate effect on improving NO, which is what patients are particularly keen on although it comes at a cost. Interestingly, the response of nasal mucosa to decongestant is dependent upon whether it is diseased or not and aids in the diagnosis of NO. Recent evidence has demonstrated that allergic rhinitis has the most profound decongestant response to a nasal decongestant, whereas chronic rhinosinusitis (CRS) has little response owing to this being a more chronic condition.[1]

Nasal cycle

Nasal mucosa spontaneously congests and decongests during the day, whereas congestion of one cavity is accompanied by reciprocal decongestion of the contralateral side. This phenomenon, first described by Kayser in 1895, is called the 'nasal cycle'. It is present in about 80% of normal individuals and usually lasts between 2 and 6 hours with the overall resistance remaining the same. The cycle diminishes with age.[2,3] The nasal cycle ensures a constant total NAR with increased resistance on the congested side and reduced resistance on the decongested side.

The exact role of the nasal cycle is not clear. Over the years, several authors have tried to explain its function: perhaps for production of nasal secretions, maintaining humidification of inspired air, respiratory defence, modulation of epithelial function and antimicrobial features, or just as an example of dynamic lateralization of the autonomic nervous system.

Aetiology and pathophysiology

The three common causes for NO include rhinitis (either in the form of non-allergic or allergic rhinitis), CRS, and DNS. The estimated prevalence rate in Europe of allergic rhinitis is 25% and for CRS is 11%. The true prevalence of a DNS is unknown but is estimated to be between 26% and 97% globally.[4]

What makes a nose obstructed?

The sensation of NO is triggered when NAR increases with a subsequent reduction in nasal airflow. However, the perceived severity of NO does not always correlate with clinical or objective findings. In fact, the symptom of NO is dependent upon the site, the grade of blockage, and subsequent increased nasal resistance. For instance, there is a greater density of proprioceptors around the INV; hence, obstruction around this region in particular will be perceived as more severe by the subject.

Box 33.1 Ohm's law

Ohm's law originally referred to an electrical circuit relationship whereby electric current = voltage/resistance. However, this principle can also be applied to nasal flow:

$$\text{Nasal flow} = \frac{\textit{pressure gradient}}{\textit{nasal resistance}}$$

Box 33.2 Poiseuille's law

Poiseuille's law describes the relationship between airway resistance and the radius of the airway and the equation is given below, where R = resistance, μ = substance viscosity, l = length of tube and r = radius of tube:

$$R = \frac{8\mu l}{\pi r^4}$$

The two mathematical laws which best describe NAR and its relationship between nasal airway diameter, nasal pressure differential, and flow rate are Ohm's law and Poiseuille's law (Boxes 33.1 and 33.2).

Resistance is inversely proportional to the radius of the nasal airway whereby the smaller the radius, the larger the resistance.

Aetiology of nasal obstruction

The aetiology of NO can be broadly divided into structural and inflammatory causes. When the cause is structural, the sensation of NO is more permanent and often one-sided whereas when caused by inflammation, the feeling of NO is more fluctuating during the day and less permanent.

Inflammatory causes of nasal obstruction

The inflammatory causes for NO are divided into two common diseases: rhinitis (either non-allergic or allergic rhinitis) and CRS. Not rarely, NO is caused by an overlapping of the two forms. Non-allergic rhinitis, allergic rhinitis, and CRS are inflammatory nasal pathologies causing nasal congestion which results in a consequent sensation of NO. These two causes of NO are generally considered as reversible forms as long as the underlying disease is treated or the inflammatory trigger is eliminated. Non-allergic rhinitis and allergic rhinitis particularly affect the mucosa of the inferior turbinates while CRS particularly affects the middle turbinate and, consequently, both have different responses to nasal decongestion.

Structural causes of nasal obstruction

The structural causes for NO commonly involve three main areas: the nasal septum, the INV and the ENV. A severely DNS as well as an insufficiency of the INV or the ENV may cause nasal blockage.[5,6] Clearly, a combination of these structural causes may coexist. A DNS occurs when the septum is deviated either into the left or right nostril or a combination of both (S-shaped deviation). The site of the DNS as well as the degree of deviation is key to the perception of severity of NO. Cottle's classification for nasal septum deviation (Figure 33.2) is probably the most clinically relevant grading system and divides the nose into five segments.

A level 1 DNS involves the ENV region (caudal septum). Level 2 is in an area between the ENV and INV. A level 3 DNS is a high septal deviation at the level of the INV region, and levels 4 and 5 are posteriorly within the nasal cavity and adjacent to the inferior turbinate. Levels 4 and 5 are distal to the INV and run parallel to the inferior turbinate. As alluded to, levels 1–3 correspond to areas around the INV and ENV regions, which represent key areas of minimal cross-sectional area within the nose. When the septum is deviated in these areas, this will therefore result in greater resistance according to Ohm's law. Moreover, these areas control overall nasal resistance and are very important for NO perception. Therefore, surgical procedures on these areas greatly influence the dynamics of nasal airflow.

The most common causes of obstruction at the INV are either due to a high septal deviation, typically Cottle's areas 2 or 3, or due to weakening/deformity of the upper lateral cartilages. Similarly, ENV insufficiency can be caused by an anterior septal deviation (Cottle's area 1/2) or by a weakening/deformity of the lower lateral cartilages.

Nasal obstruction assessment

Clinical diagnosis of nasal obstruction

The clinical diagnosis of NO is based on a thorough clinical history, nasal examination (either by means of anterior rhinoscopy or nasal endoscopy), and/or imaging (computed tomography or magnetic resonance imaging scan) if other methods are not feasible or sufficient. NO is a common complaint in many nasal conditions and significantly impacts quality of life; therefore, its objective measurement is of paramount importance. Generally, NO can be assessed by means of a validated questionnaire, thus evaluating the reported subjective sensation of NO or objectively, either by evaluating nasal airflow and/or nasal resistances during inspiration/expiration or by calculating the nasal cavity area/section as an indirect index of nasal patency. However, it has been widely reported that there is a lack of reliable correlation between subjective and objective measurements of NO. As a consequence, the inherently subjective diagnosis of NO, based solely on history and examination, can present challenges when attempting to correlate it with objective assessments.

Subjective evaluation of nasal obstruction

Today, several questionnaires are validated for the measurement of subjective NO. The Sino-Nasal Outcome Tests 22 and 23 (SNOT-22 and SNOT-23) and the Nasal Obstruction Symptom Evaluation (NOSE) questionnaires provide a disease-specific outcome measure. Moreover, the visual analogue scale (VAS) for NO represents an easy and rapid tool to assess NO. Conversely, generic tools of health status, like the 36-item Short Form Health Survey and the abbreviated variant called SF-6D, are considered too generic and are not usually used in clinical practice when severity of NO needs to be evaluated. Interestingly, a survey conducted in 2017 in UK showed that less than 30% of British ENT surgeons utilize a validated questionnaire to assess NO.[7]

Objective assessment of nasal obstruction

NO can be measured by assessing nasal airflow, nasal resistance, or the cross-sectional area of the nasal cavities. The most commonly used tools to evaluate NO in clinical practice include rhinomanometry (RMM), acoustic rhinometry, and peak nasal inspiratory flow (PNIF).

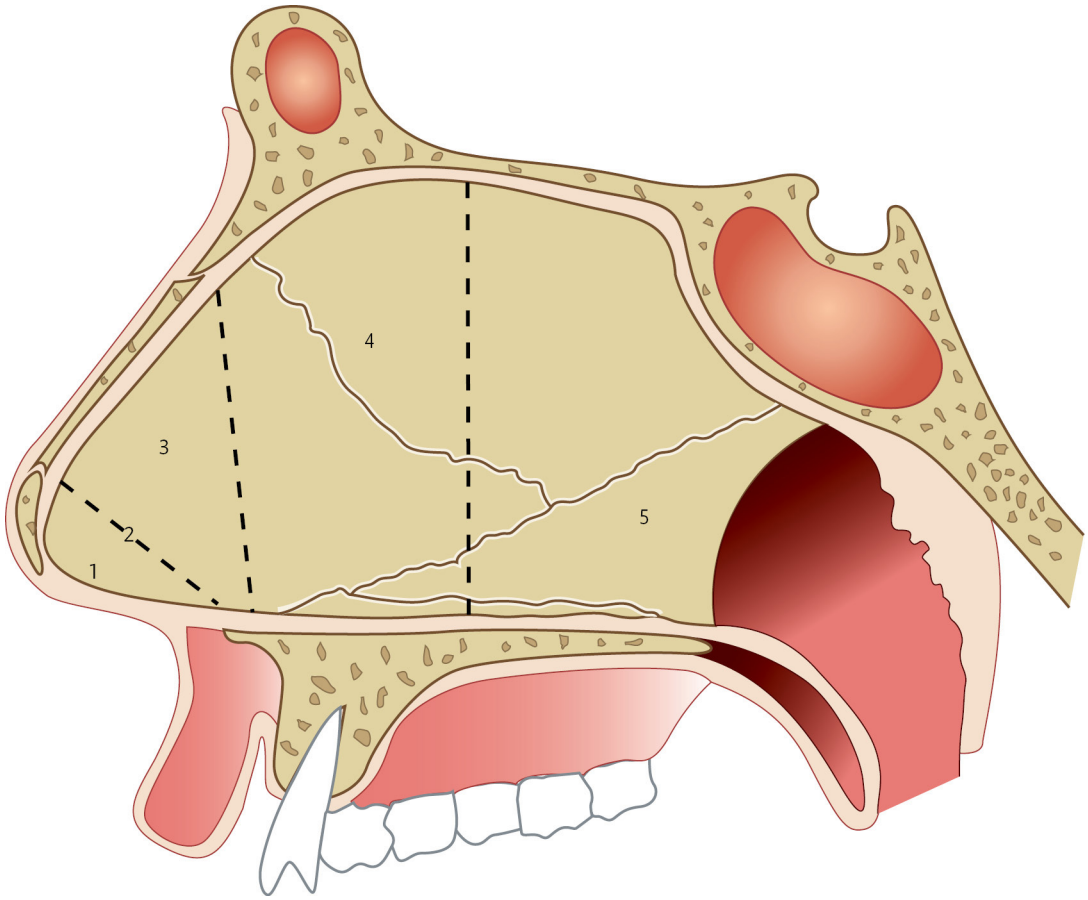

Figure 33.2 Cottle's classification for nasal septum.
Image courtesy of Mr Alfonso Luca Pendolino (Consultant Rhinologist and ENT Surgeon, Imperial College Healthcare NHS Trust).

Rhinomanometry

RMM is considered the 'gold standard' objective evaluation for NAR and NO. It simultaneously measures nasal airflow and the pressure gradient required to achieve that flow from which NAR can then be calculated. It can be anterior or posterior, and performed passively or actively. Anterior RMM is performed unilaterally with the pressure probe inserted into the contralateral nostril which enables unilateral evaluation. Posterior RMM enables bilateral NAR measurement with the probe inserted via the mouth into the postnasal space. Moreover, in active RMM the patient breathes and generates a natural nasal airflow whereas in passive RMM the airflow is generated externally.

Anterior active rhinomanometry (AAR) is the most commonly used RMM nowadays. In AAR a mask is attached to a device that measures transnasal pressure and flow, and interfaces with a computer. The pressure sensor is placed in one nostril with a total seal created by taping off the nostril. The patient is then asked to nasally breathe, and a simultaneous display of the pressure–flow curve is provided. NAR is then calculated utilizing Ohm's law and reported in units of $Pa/cm^3/s$. The same procedure is then performed with the contralateral nostril. The more obstructed the airway, the greater the pressure required to generate a certain flow. Even though nasal resistance can be calculated at each point of the pressure–flow curve, conventionally, to allow comparison between subjects and studies, the International Committee on Rhinomanometric Standards has described calculating NAR in classic AAR at a standardized pressure of 150 Pa. Normal values for AAR are reported in Table 33.1.

Acoustic rhinometry

Acoustic rhinometry is an objective measurement of the cross-sectional area and volume of the nasal cavity at different points which are obtained by using reflected sound pulse technology. The size and pattern of the reflected sound waves provide information on the structure and dimensions of the nasal cavity, with the time delay of reflections correlating with the distance from the nostril. This information is recorded by software and then converted using

Table 33.1 Normal values for anterior active rhinomanometry in adults and children

Measurement	Nasal resistance at 150 Pa
Normal values for adult males (Caucasian)	0.24 $Pa/cm^3/s$ (95% CI 0.09–0.39)
Normal values for adult females (Caucasian)	0.26 $Pa/cm^3/s$ (95% CI 0.08–0.44)
Normal values for children (8 years)	0.24 $Pa/cm^3/s$ (95% CI 0.11–0.37) (after nasal decongestion)

CI, confidence interval.

mathematical calculations and theoretical assumptions in order to obtain nasal volume and area calculations. The measurements are done with the patient in the upright position and performed during breath holding.

Acoustic rhinometry measurement divides the nasal cavity into anterior, middle, and posterior segments. The anterior segment represents the first 3 cm of the nasal cavity and corresponds to the nasal vestibule and internal nasal valve with limited congestive capacity. The INV represents the first minimal cross-sectional area (MCA1) and the narrowest part of the nose. The middle segment measures from 3 to 5.2 cm (MCA2) and contains the inferior and middle turbinate areas, which represent the main congestive region and the most responsive region to decongestant. The upper limit of normal for MCA1 is 0.60 ± 0.04 cm^2 and for nasal volume it is 8.25 ± 5.23 cm^3. The decongestability component can be measured using the nasal congestion index (NCI): (decongested MCA) – (base MCA)/baseline MCA = %change. The higher the NCI (>25%), the higher the decongestible component of the NO and the stronger the association with the subjective feeling of nasal blockage. In this situation, the cause of NO is more than likely allergic rhinitis. Conversely, a NCI less than 15% is more than likely within normal limits. While primarily a research tool for nasal cavity measurements, acoustic rhinometry is also frequently employed in rhinology laboratories to assess changes in nasal cavity volume and MCA after a decongestant test. This allows it to aid in the differential diagnosis of reversible (i.e. mucosal disease) versus irreversible (i.e. structural deformities) causes of NO.

Peak nasal inspiratory flow

The PNIF is a cheap and quick method for the objective assessment of nasal airway obstruction, also unilateral, and has been demonstrated to be reproducible in the evaluation of nasal airway obstruction and as good an indication of objective nasal patency as formal RMM.[8] It is a very simple tool made of a flow meter connected to a facemask. To obtain the PNIF, the patient, sitting upright, is first asked to exhale fully and then, once the mask is in place and well-sealed to the face, is asked to inhale as hard and fast as they can while keeping the mouth closed. Usually, three satisfactory maximal inspirations are obtained and the highest of these results is taken as the PNIF value. PNIF is highly influenced by pulmonary function and thus, its value decreases with age, is higher in males, and increases with height.[5,6] Normal values for both children and adults are reported in Table 33.2.

PNIF is a portable, quick, and easy-to-use tool and probably, for these reasons, it is the commonest method used by ENT doctors for NO assessment. However, it is based on high flow rates and potentially may cause valve collapse, especially when unilateral measurements are taken which may influence the results obtained.

Table 33.2 Normal values for peak nasal inspiratory flow in adults and children

Measurement	Maximal nasal inspiratory flows
Normal values for adult males (Caucasian)	143 ± 48.6 L/min
Normal values for adult females (Caucasian)	121.9 ± 36 L/min
Normal values for children (8 years)	80 ± 25 L/min

Treatment of nasal obstruction

Inflammatory diseases treatment

Treatment of inflammatory causes of NO (allergic rhinitis, non-allergic rhinitis, and CRS) is mainly medical whereas surgery only plays a role when control of symptoms has not been achieved after optimal medical treatment. The mainstay of treatment is nasal douching and topical corticosteroids, either in the form of spray or drops. Avoidance or management of the triggering causes—for instance, allergen avoidance, pollution control, or treatment of underlying laryngopharyngeal reflux—must be adopted in order to achieve control of rhinitis. Depending on the type of nasal disease, additional medical treatments can include antihistamines (local or systemic), antibiotics (local or systemic, short or long term), immunotherapy, and, more recently, biologics.

Surgical options for allergic rhinitis and non-allergic rhinitis rely mainly on inferior turbinate surgery—by cauterization (by means of submucosal diathermy or bipolar cautery), turbinoplasty, or simple outfracture—and aims to improve the nasal airway by reducing inferior turbinate engorgement. However, it is important to bear in mind that inferior turbinate surgery effects are usually not long-lasting and, thus, cannot be considered as an absolute alternative to medical treatment.

Surgical treatment of CRS involves mainly endoscopic sinus surgery, which aims to open up the sinuses, clear disease, and aid to optimize steroid delivery. Therefore, also for CRS, surgery cannot be considered the cure for disease but more as a key step to improve delivery of medical therapy and to control disease relapse. More recently, balloon dilation of sinuses has been introduced as a minimally invasive surgical treatment for CRS by allowing the opening of natural sinus ostia without damaging nasal mucosa with the added benefit of being predominantly performed under local anaesthesia.

Structural diseases treatment

Treatment of structural causes of NO aim to correct an anatomical nasal deformity which creates an impairment in nasal patency.

Deviations of the nasal septum not involving the INV and ENV can easily be corrected by a septoplasty. In this operation mucoperichondrial flaps are gently elevated bilaterally and the deviated part of the septum—cartilaginous and/or bony portion—is excised or repositioned. The incision is later closed, and the two raised flaps are quilted together in order to prevent a haematoma.

However, if a high septal deviation or a very anterior septal deviation is present and compromises the INV or ENV, a conventional septoplasty will not suffice and a functional septorhinoplasty—either a closed or an external approach—should be performed instead to ensure best functional outcomes. The main aim of this procedure is to improve nasal breathing focusing on both the septum and the nasal valves. A functional septorhinoplasty, in fact, will also allow the positioning of grafts which are used to augment or reinforce the nasal valves. The indications for septoplasty very much focus on correcting mid to posterior septal deviations, whereas the indications for functional septorhinoplasty focus more on correcting anterior and high septal deviations as well as augmenting the nasal valves. Moreover, extremely severe caudal septal deviations may require complete removal of the septum through a so-called extracorporeal septoplasty approach. This should only be performed by expert nasal surgeons and the decision should not be taken lightly.

In many cases, especially following trauma, conventional septoplasty surgery alone is unlikely to achieve a straight septum unless correcting the nasal pyramid is also addressed, which includes osteotomies to the nasal bones.

Controversies

National and international guidelines are widely accepted and used when treating inflammatory causes of NO. However, controversies still exist when considering best practice for correcting a structural cause of NO. Even though a recent randomized controlled trial has demonstrated that septal surgery with or without turbinate surgery is more effective than medical management for treating NO due to a deviated septum, level 1 evidence remains lacking for ESS, turbinate surgery, and septorhinoplasty surgery.

In the opinion of the authors, the planning of septal surgery with regard to the site of the septal deviation, as well as INV and ENV function and whether turbinate disease exists, all need to be assessed and quantified. This will better steer decision-making and streamline surgery inclusion criteria. Equally, a consensus does not exist on how best to approach a functional septorhinoplasty, through either an external/open or internal/closed approach.

An external approach involves making a transcolumellar incision which facilitates access to the ENV and INV and anterior septum, but is not the case when performed through an internal approach. An internal approach is performed within the nostril and, arguably, has less access to the ENV and INV although this is very much surgeon dependent. The external approach enables easier access to place spreader grafts, pieces of cartilage that are placed in between the septum and the upper lateral cartilages to augment the INV, without violating nasal mucosa. Similarly, an external approach will allow a complete evaluation of all the structures necessary to guarantee the strength of the ENV (i.e. integrity of the lower lateral cartilages, their attachment to the scroll area, and medial crural footplate attachment to the septum) and permit a much easier placement of struts and/or grafts.

Conclusion and the future

In the UK, patients with persistent NO can wait up to 5 years before they receive a definitive diagnosis in secondary care. This causes a significant delay in tailored treatment with a subsequent bottleneck in the patient's journey. Consequently, there is a need to improve diagnostics in primary care and reduce treatment delay. For conditions like CRS, evidence suggests that delaying vital surgery can lead to reduced postoperative quality of life, with increased postoperative healthcare needs and reduced olfactory improvement. A 'pure tone audiometry' equivalent in rhinology does not yet exist. Progress is being made with the development of new medical devices to better diagnose the cause of NO through harnessing computational fluid dynamics as well as nasal disease-specific acoustic signatures. Nasal acoustic analysis is based on technology analogous to a stethoscope which captures the acoustic sound of nasal airflow and creates disease-specific acoustic signatures. It relies on the nasal decongestant response which eliminates the natural nasal cycle and identifies the location of nasal disease (inferior, middle turbinate, or septal deviation). The nasal acoustic device shows promising results in helping to diagnose the cause of NO and could be used in primary care.[1]

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34

Nasal crusting

Catherine Rennie

Introduction

Nasal crusting is a common symptom presenting to ear, nose, and throat (ENT) clinics and is often caused by local infections or trauma; however, in a small number of cases it may be associated with systemic disease or even malignancy. Therefore, it is important to take a comprehensive history, perform a full examination, and direct investigations according to medical history. A detailed understanding of the underlying causes of nasal crusting will guide the history, examination, and investigations of this condition. Screening tests for inflammatory conditions may be useful but care should be taken when interpreting them as many are non-specific. A biopsy should be taken if there is a suspicion of malignancy and if needed to support a suspected diagnosis of vasculitis. The treatment will depend on the underlying cause of nasal crusting and the severity of the patient's symptoms.

History

Local versus systemic

Nasal crusting can be caused by local or systemic disease. Local causes include infections and trauma whereas systemic causes may be associated with an underlying infection or vasculitis.

Duration

The duration of symptoms will help narrow down the long list of differential diagnoses (Table 34.1). Chronic nasal symptoms without any systemic features suggest a local cause such as chronic rhinosinusitis, whereas chronic nasal symptoms with systemic features would raise suspicion of granulomatosis with polyangiitis (GPA), sarcoidosis, eosinophilic granulomatosis with polyangiitis (EGPA), or other systemic causes (Table 34.2) and would therefore warrant further investigation.

Associated symptoms

Nasal crusting is frequently associated with other nasal symptoms, most commonly with epistaxis and nasal obstruction but also rhinorrhoea and hyposmia. These symptoms often occur in patients with rhinosinusitis along with facial pain, headache, and/or nasal discharge. Nasal symptoms can be the first presentation of systemic diseases such as GPA and it can initially be difficult to differentiate GPA from other causes of rhinosinusitis, therefore further investigations are necessary to rule out systemic causes of nasal crusting. Unilateral nasal symptoms should raise suspicion of sinonasal malignancy—red flag symptoms include unilateral nasal obstruction, unilateral epistaxis, epiphora, paraesthesia, dental pain, unilateral facial swelling, and unilateral otitis media with effusion. Ear symptoms are often associated with nasal conditions. Nasal obstruction can result in Eustachian tube dysfunction and otitis media with effusion as is often seen in EGPA. Hearing loss both mixed and sensorineural is a feature of autoimmune disease and vasculitis (rheumatoid arthritis and GPA). In immunocompromised patients, fungal infection should be considered as a cause of nasal crusting.

Table 34.1 Causes of nasal crusting

	Acute	Chronic
Traumatic/ iatrogenic	Nasal surgery	Nose picking Foreign body
Nasal drugs		Nasal decongestants Nasal corticosteroids Cocaine
Occupational exposure	Chromium fumes Sulphuric acid Swarf Powdered glass	
Inflammatory	SLE (acute exacerbation)	GPA Sarcoidosis EGPA SLE Crohn's/ulcerative colitis Rheumatoid arthritis Relapsing polychondritis
Infectious	Bacterial (*Staphylococcus aureus*)	Tuberculosis Atrophic rhinitis Rhinoscleroma Syphilis Leishmaniasis Leprosy
Neoplastic		Carcinoma

Table 34.2 An overview of systemic disorders causing nasal crusting

Disorder	History and examination	Laboratory tests	Histopathology	Treatment
GPA	Respiratory symptoms, upper respiratory tract infection, cough, haemoptysis, progressive renal involvement	Full blood count, ↓ haemoglobin, Urea and electrolytes, creatinine, +ve c-ANCA, proteinase-3, urinalysis with 24-hour urinary protein	Vasculitis of medium and small vessels with intramural eccentric necrotizing granulomatous lesions	Steroids Cyclophosphamide Methotrexate Mycophenolate mofetil Rituximab Azathioprine
Sarcoid	Respiratory involvement, anterior uveitis, erythema nodosum, lupus pernio, polyarthralgia, myopathy, lymphadenopathy	↑ serum or urinary calcium, ↑ angiotensin-converting enzyme levels Chest X-ray and CT: reveals hilar lymphadenopathy/ pulmonary fibrosis	Multiple non-caseating granulomas	Steroids Methotrexate Hydroxychloroquine Anti-tumour necrosis factor-α
EGPA	Allergic rhinitis, nasal polyps, asthma, pneumonia, gastroenteritis	Eosinophilia, −ve c-ANCA, +ve p-ANCA, myeloperoxidase	Necrotizing vasculitis of small and medium-sized vessels with necrotizing extravascular granulomas and prominent eosinophilia	Steroids Cyclophosphamide
SLE	Photosensitivity, arthralgia, oral ulcers, malar/discoid rash Other: renal/neurological (e.g. seizures)	Antineutrophil antibodies, anti-double-stranded DNA, anti-Sm, LE cell, full blood count, rhesus factor, urinalysis with 24-hour urinary protein	Necrotizing vasculitis of small and medium-sized vessels with necrotizing extravascular granulomas and prominent eosinophilia	Steroids
Tuberculosis (TB)	Chronic productive cough, low-grade fever, weight loss, lymphadenopathy, TB contact	Microscopy (Ziehl–Neelsen stain), Mantoux, polymerase chain reaction, chest X-ray, biopsy and culture	Granuloma with central caseating necrosis.	Anti-TB therapy
Syphilis	Mucosal involvement is seen in sexually active patients, secondary syphilis- dermatitis, and lymphadenopathy Nasal involvement is seen also in congenital syphilis and gummatous stage of syphilis	VDRL, fluorescent treponemal antibody absorption test, *Treponema pallidum* hemagglutination assay	Perivascular infiltrate of plasma cells and lymphocytes Increased angiogenesis	Penicillin

Nasal surgery or trauma

Nasal crusting is also a common feature of septal perforation (see Chapter 28), other features are nasal obstruction, epistaxis, and whistling, especially if the perforation is small. Septal perforations are often iatrogenic as a result of nasal procedures such as septoplasty, bilateral nasal cautery, and endoscopic sinus surgery. Some studies have shown septal perforation rates of up to 5% following septoplasty. Septal perforations can also be traumatic secondary to a septal haematoma which compromises the blood supply to the septum. Other causes of septal perforation include nose picking, nasal piercings, corrosive foreign bodies, repeated nasogastric tube insertion, radiotherapy, cocaine abuse, and systemic conditions such as GPA. Persistent crusting can also occur after surgery even in the absence of a perforation. Overly aggressive nasal surgery (inferior turbinectomy) is associated with secondary atrophic rhinitis (empty nose syndrome); this is a clinical syndrome in which people who have wide, clear nasal passages experience nasal obstruction, nasal dryness, and crusting, and a sensation of being unable to breathe. The nasal cavity functions as a very efficient filter and humidifier for the air we breathe, the large surface area and excellent blood supply provide a warm moist mucosal layer over which heat and temperature exchange can occur. Nasal surgeries which significantly reduce the mucosal surface area risk disrupting this fine balance within the nasal cavity, this can lead to nasal dryness and crusting.

Drugs

The use of some intranasal medications can lead to nasal crusting or even septal perforations (Figure 34.1). Prolonged use of topical nasal decongestants such as xylometazoline can result in rhinitis medicamentosa, a feature of which can be nasal crusting as well as nasal obstruction. Long-term use of intranasal corticosteroids has been linked to septal perforation although this is an exceptionally rare complication. Nasal sprays can cause epistaxis, often due to mechanical trauma through touching or abrading the septum with the hard plastic applicator or repeated spraying at the same location which may lead to erosions or even ulcerations on the septum. Therefore, when prescribing nasal sprays, it is good practice to demonstrate best spray technique.

The use of illicit drugs, particularly cocaine, has been associated with nasal crusting and septal perforations. Cocaine not only causes vasoconstriction but also produces local anaesthesia, so the user does not feel pain and trauma to the nose. Also, cocaine is frequently 'cut' with known irritants such as borax and talcum powder. These additions may also contribute to the mucosal destruction. Chronic cocaine abuse in conjunction with infection worsens the condition to such an extent that membrane, bone, and cartilage necrosis takes place and leads to perforation, nasal collapse, intranasal stenosis, and saddling. This clinical picture can progress to become a 'cocaine-induced midline destructive lesion'. A study in 2014 revealed that

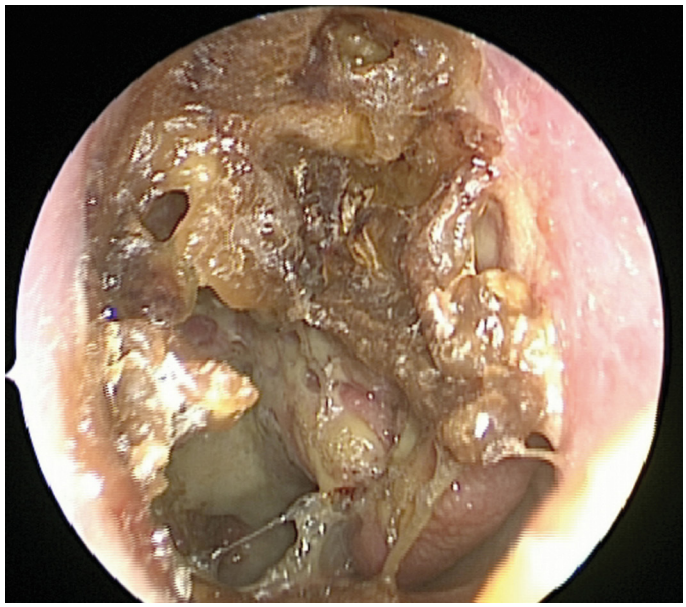

Figure 34.1 Nasal cavity following years of cocaine abuse.

is a veterinary antiparasitic medication; it was previously used in humans as an anticancer agent but has since been banned as it can cause agranulocytosis.

Levamisole can also induce a vasculitis in humans which can result in midline destructive lesions in cocaine users with similar features to GPA, the clinical manifestations of cocaine–levamisole use are multiple and can be local or systemic. The most important serological study is antineutrophil cytoplasmic antibodies (ANCA), perinuclear ANCA (p-ANCA), and/or cytoplasmic ANCA (c-ANCA) positivity, which may have multiple antigenic specificities, such as myeloperoxidase and proteinase-3.

Systemic symptoms

A number of systemic diseases can cause nasal septal crusting and perforation like GPA (Figure 34.2), systemic lupus erythematosus (SLE), and sarcoidosis (Table 34.2). Clinical features are often non-specific and include obstruction, epistaxis, postnasal discharge, whistling, and crusting. The presence of other associated features relating to the respiratory, cardiovascular, gastrointestinal, musculoskeletal, skin, and ocular systems can guide further investigations. It is important to have an awareness of these conditions, as they can have serious consequences for the patient if left untreated.

Travel

In endemic areas tuberculosis (TB), leishmaniasis, rhinoscleroma (Figure 34.3), and leprosy are common causes of nasal crusting but are rare in the developed world. The incidence of extrapulmonary TB is increasing, it now constitutes 40% of the total disease occurrence, with many cases involving the ENT region. Nasal secretions and swab specimens have a very low yield and should not be used to rule out nasal TB. Definitive diagnosis requires histopathology with culture. Primary atrophic rhinitis (also known as ozaena) is usually found in the tropics, in rural areas of low socioeconomic status, and is more common in women than men. It is a progressive chronic nasal disease characterized by mucosal atrophy with resorption of the underlying bone, formation of thick crusts, and a distinct foetor. The destructive process is accompanied by nasal cultures growing *Klebsiella ozaenae*.

Occupation

A history of any exposure to chemicals such as chrome, agricultural dust, or chemical dust should also be elicited. There is a well-recognized association between ulceration and perforation of the nasal septum and occupations involving chrome compounds. Industrial and agricultural aerosolized dust, as found in grain and rice elevators and cement, glass, and lime factories, can cause perforations. Individuals working with swimming pool chemicals or in chemical factories are also at high risk. Sinonasal malignancy has been related to occupational exposures in the leather tanning, nickel, and wood industries. Workers in these industries should wear properly fitted filter masks during exposure to the irritant.

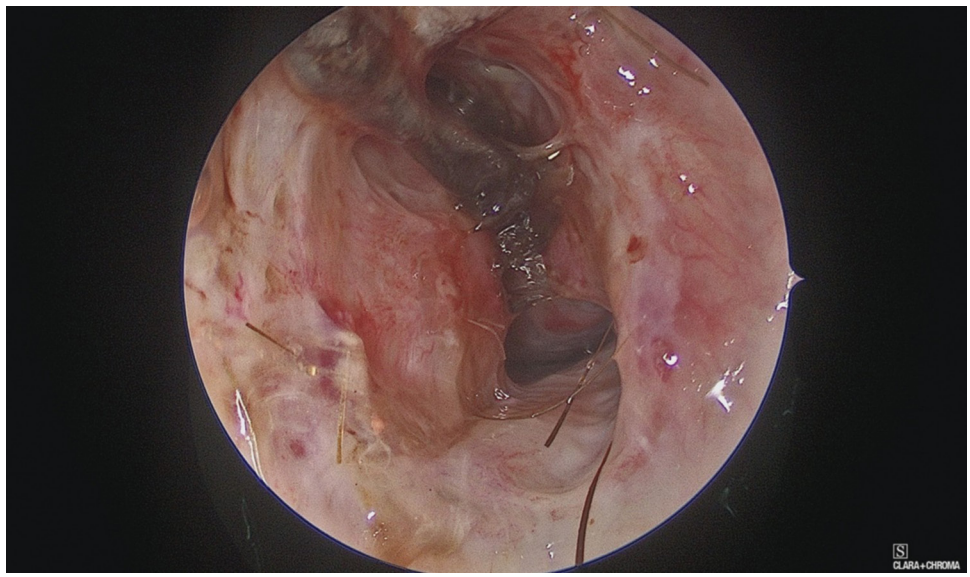

Figure 34.2 GPA.

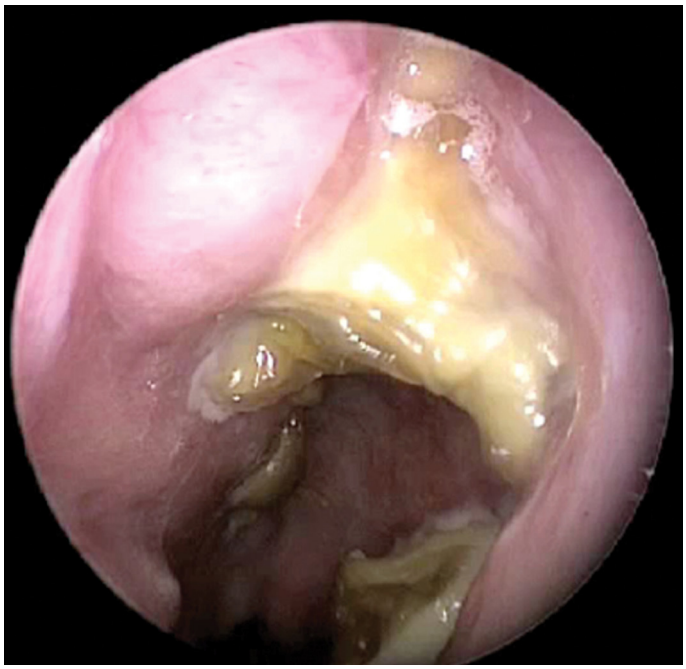

Figure 34.3 Rhinoscleroma.

Neoplasia

Lethal midline granuloma is a rare entity that usually arises in the nasal cavity, exhibits a male preponderance, and has a wide age range. The majority of lethal midline granulomas are T-cell lymphomas; they usually present with nasal obstruction followed by purulent rhinorrhoea and serosanguinous discharge. As symptoms progress, usually unilateral mucosal ulceration with extension into the palate, maxillary sinus, and upper lip may occur. Mucosa is often pale and friable, and extensive crusting is often present. Oronasal fistulas as well as nasal septal perforations may occur.

Examination

Examination will also help to direct the investigations for nasal crusting. Inspection of the external nose may reveal a saddle deformity which could be suggestive of trauma, GPA, relapsing polychondritis, or intranasal cocaine use, whereas the classic appearance of lupus pernio is indicative of sarcoidosis. Anterior rhinoscopy should be performed as it will reveal the site of the crusting and identify a septal perforation or any further intranasal destruction. Extensive nasal destruction of septum and turbinates may suggest cocaine use. If a perforation is identified, the site, size, and health of the perforation edges should be recorded. Irregular, bleeding, or ulcerated edges may suggest a more sinister cause of the perforation. On anterior rhinoscopy, polyps, nasal masses, nasal discharge, or even foetor can also be identified.

It is important to perform nasal endoscopy to examine the rest of the nasal cavity as well as the postnasal space. Nasal polyps associated with EGPA may not be identified on anterior rhinoscopy and the full extent of cocaine midline destructive lesions will not be determined with anterior rhinoscopy alone. Since many of the causes of nasal crusting have other ENT manifestations it is important to carry out a full ENT examination. Systemic examination should be targeted depending on the symptoms elicited in the history.

Investigations

The investigations will depend on the history and examination. The majority of cases with nasal crusting will not need further investigation; however, in order not to miss a serious underlying condition, the presence of red flag symptoms or associated features relating to the respiratory, cardiovascular, gastrointestinal, musculoskeletal, skin, and ocular systems should guide further investigations (Table 34.2).

Laboratory tests for nasal crusting include full blood count, urea, electrolytes and creatinine, erythrocyte sedimentation rate, angiotensin-converting enzyme, antineutrophil antibodies, antineutrophil antibodies, double-stranded DNA, anti-Sm, rheumatoid factor, anti-Ro and anti-La antibodies, VDRL, *Treponema pallidum* hemagglutination assay, fluorescent treponemal antibody absorption, ANCA, p-ANCA, c-ANCA, proteinase 3, and myeloperoxidase:

- Urine screen and hair test for cocaine abuse.
- Urinalysis.
- Biopsy and cultures for fungal, mycobacterial, and bacterial species.
- Chest X-ray, Mantoux test.
- Skin testing for allergy.
- Computed tomography scan of the nose and paranasal sinuses.
- Biopsy of the perforation.

Careful interpretation is required when reviewing screening blood tests as sensitivity can be low. If there is high clinical suspicion, patients should be referred for specialist opinion even in the absence of positive screening tests.

ANCA are markers of systemic vasculitis and show predominantly c-ANCA in GPA whereas p-ANCA is more often associated with EGPA. Elevated c-ANCA has 99% specificity for GPA, but sensitivity is related to disease activity, being 91% in active disease and 63% in inactive disease. Typically, a positive c-ANCA on immunofluorescence is followed by a confirmatory proteinase-3-ANCA on enzyme immunoassay, whereas in EGPA approximately 50% of patients have a positive ANCA which is usually perinuclear on immunofluorescence and with myeloperoxidase specificity on enzyme immunoassay.

The criteria for the classification of GPA are:

1. oral ulcers or purulent or serosanguinous nasal discharge
2. abnormal chest radiograph (presence of nodules, fixed infiltrates, or cavities)
3. urinary sediment, microhaematuria, or red cell casts in urine sediment
4. granulomatous inflammation on biopsy.

For purposes of classification, a patient is said to have GPA if at least two of these four criteria are present. The presence of any two or more criteria yields a sensitivity of 88.2% and a specificity of 92.0% (American College of Rheumatology (1990) criteria).

The criteria for the classification of EGPA are:

1. asthma (wheezing, expiratory rhonchi)
2. eosinophilia of more than 10% in peripheral blood
3. paranasal sinusitis
4. pulmonary infiltrates (may be transient)
5. histological proof of vasculitis with extravascular eosinophils
6. mononeuritis multiplex or polyneuropathy.

The presence of four of the above six findings results in a diagnostic sensitivity of 86%, with a specificity of 99% according to the American College of Rheumatology (1990) criteria.

Elevated serum angiotensin-converting enzyme and calcium levels are suggestive of sarcoidosis but the sensitivity and specificity are low; however, they are used in combination with a chest X-ray as a screening tool.

SLE is unlikely if the antinuclear antibody test is negative but false positives are common, particularly in elderly females.

In the past, nasal biopsy was considered a mandatory part of investigation of nasal septal ulceration and perforation. It has been found that histological results are most of the time non-specific and not useful in further management of patients. In most cases of rheumatic diseases or vasculitis, biopsy shows only chronic inflammation. Septal biopsy may be helpful in preventing the need for more invasive biopsy (i.e. a renal biopsy) when positive and ruling out malignancy.

Treatment

Topical intranasal treatments are often required to treat nasal crusting. Saline nasal irrigation is an important part of treatment and is well tolerated by patients with crusting from all causes. Irrigation can be done multiple times a day as required. Regular steaming or humidification can also be beneficial in loosening crusts. Other agents that can help prevent nasal dryness and crusting are sesame oil (Nozoil), water-soluble nasal gels formulated with sodium hyaluronate (NasoGel), petroleum jelly, and 25% glucose in glycerol and may be used to lubricate and moisten the nostril. Topical steroids are part of the treatment algorithms for allergic rhinitis, chronic rhinosinusitis, EGPA, and nasal sarcoidosis.

Mupirocin, Fucidin, and Naseptin are nasal creams that can be used to treat bacterial colonization of the nasal vestibule and vestibulitis; persistent cases that do not improve after one course of nasal cream can be treated by rotating the nasal creams with 2 weeks of each, three times a day.

If rhinitis medicamentosa is the cause of crusting, the causative nasal decongestant should be stopped. However, a troublesome rebound swelling and congestion may arise—steroid nasal drops and antihistamines can help alleviate this.

In systemic disease the nasal symptoms will improve when the disease is in remission and surgery should not be considered unless there is remission. The treatment for systemic diseases causing nasal crusting is usually monitored by physicians from the appropriate medical team (e.g. renal, respiratory, rheumatology) (Table 34.2).

When nasal crusting is extensive, general anaesthesia may be required to clear the crusts in order to permit irrigation of the nasal cavity. Procedures to reduce the size of the nasal cavity have been used in patients with empty nose syndrome with variable results. Some cases of atrophic rhinitis have been successfully treated with nasal obturators or a Young's procedure. However, medical management remains the mainstay of treatment. Endoscopic sinus surgery is performed in the treatment of chronic rhinosinusitis, fungal sinusitis, and nasal polyps. However, care should be taken in patients with systemic disease as surgery may exacerbate the symptoms.

Nasal crusting often presents as a diagnostic dilemma due to an extensive differential diagnosis (Table 34.1) and vague presenting signs and symptoms. Differentiating between the multiple varied aetiologies can be challenging for clinicians and a multidisciplinary approach is often required.

Tips and pearls

- Nasal crusting is a common presenting symptom.
- Most nasal crusting can be managed conservatively with nasal irrigation and topical emollients.
- A small number of cases may be systemic disease or malignancy.
- Systemic symptoms should prompt the clinician to investigate for an underlying systemic disease.
- Screening tests should be performed in patients with systemic features or where there is a high clinical suspicion.

FURTHER READING

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35

Rhinoplasty

Fazil Apaydin

Introduction

Rhinoplasty is aesthetic surgery of the nose to give a better shape in order to fulfil the requests of the patient. It is widely regarded as the most challenging procedure of facial plastic surgery, balancing expectations with outcomes. Patients seem to be becoming more demanding with the increasing use of social media. Therefore, there is a need to be very thorough during each step of rhinoplasty. In this chapter the three stages of rhinoplasty will be detailed: (1) preoperative preparation, (2) surgery, and (3) postoperative care.

Preoperative preparation for rhinoplasty

History taking

When a patient attends for rhinoplasty, it is good practice to collate data regarding general health, a patient-reported outcome measure, a body dysmorphic syndrome questionnaire, and a few general questions to help better understand what the requirements are. After a review of these documents open-ended questions such as 'How can I help you?' or 'What can I do for you today?' are directed to the patient. During this conversation, the surgeon starts to develop a feeling about the patient's motivations for surgery, thought processes, and also whether they are a good candidate for surgery with reasonable expectations. A very important question at the end of this conversation is, 'Why now?' This question often helps the surgeon to predict the true requirements of the patient (Box 35.1).

Informing the patient

It may be very helpful to ask your patient to read brochures and watch videos of rhinoplasty before attending for a consultation. After taking a history and examining the patient, the surgeon should then discuss the surgery in detail including the grafts to be taken or used, the admission to hospital, and the postoperative period. It should be mentioned that the goal of surgery is to make the aesthetic appearance better, not perfect. The main goals of rhinoplasty should be reflected back to the patient: a nose that is in harmony with the rest of the face, and to draw attention to the beauty of the eyes.

Ear, nose, and throat examination

The nose is initially carefully inspected, which is vital for rhinoplasty assessment as photographs are usually insufficient in reflecting the true concerns of the patient. The dynamic situation of the nose is observed when the patient is talking, smiling, and breathing. If the tip droops a lot during smiling or laughing, the surgical plan may be adjusted accordingly. The second crucial step is palpation. First the bony pyramid is palpated to find out the length and shape of the nasal bones and the connection of the nasal bones with the ascending process of the maxilla. Next, the middle vault and then the tip are also palpated. A 'recoil' test is very useful to reveal the strength of the lower lateral cartilages and septal support. The anterior nasal spine and the caudal septum are palpated with a gloved hand.

The patient then goes through a functional examination of the nasal airways (gentle inspiration and gentle expiration), followed by examination of the internal nasal valve region and the nasal passages using both a nasal speculum and an endoscope.

Photography

Standardized photography for rhinoplasty is crucial to validate results after surgery. Ideally, the patient is photographed in a studio setting with two flashes (strobes or LED lights) to reduce shadowing, using a blue or black background. At least six standard views are recommended (Box 35.2). A ruler is used for frontal, lateral, and basal views to facilitate further measurements. Sky, frontal, and lateral smiling views may also be taken to furnish information about the dynamic behaviour of the nose. A single-lens reflex camera (full frame, mirrorless, APS-C or compact) with a portrait lens seems to be the ideal and cost-effective solution for obtaining 1:1 life-size photographs. The equipment and software for three-dimensional photography is still very expensive. Although the use of mobile phones may appear very attractive and practical, a lack of standardization remains the main problem.

Facial analysis

See Box 35.3.

The surgeon should have a standard regimen for aesthetic analysis of the standard views. We have been using software called Rhinobase, which we developed in 2000 with the aim of standardizing photometric and aesthetic analysis:

Box 35.1 Rhinoplasty self-evaluation form

Name: Date: / /

Over the past month, how much of a problem was the following:	No problem					Extreme problem
1. Having a blocked or obstructed nose	0	1	2	3	4	5
2. Getting air through my nose during exercise	0	1	2	3	4	5
3. Having a congested nose	0	1	2	3	4	5
4. Breathing through my nose during sleep	0	1	2	3	4	5
5. Decreased mood and self-esteem due to my nose	0	1	2	3	4	5
6. The shape of my nasal tip	0	1	2	3	4	5
7. The straightness of my nose	0	1	2	3	4	5
8. The shape of my nose from the side	0	1	2	3	4	5
9. How well my nose suits my face	0	1	2	3	4	5
10. The overall symmetry of my nose	0	1	2	3	4	5

11. Are you very worried about your appearance in any way?
[ ] Yes [ ] No

12. Have you had nose surgery before?
[ ] Yes [ ] No

13. Does this concern/preoccupy you? That is, do you think about it a lot and do you wish you could worry about it less?
[ ] Yes [ ] No

14. Have you had facial trauma before?
[ ] Yes [ ] No

15. Did these concerns cause you a lot of distress, torment, or pain? (Circle the best answer)

1	2	3	4	5
No	Mild, not too disturbing	Moderate, disturbing but still manageable	Severe, very disturbing	Extreme, disabling

16. Did these concerns cause you impairment in social, occupational, or other important areas of functioning? (Circle the best answer)

1	2	3	4	5
No	Mild, not too disturbing	Moderate, disturbing but still manageable	Severe, very disturbing	Extreme, disabling

17. Did these concerns often significantly interfere with your social life? (Circle the best answer)

1	2	3	4	5
No	Mild, not too disturbing	Moderate, disturbing but still manageable	Severe, very disturbing	Extreme, disabling

18. Did these concerns often significantly interference with your schoolwork, job, or ability to function in your role? (Circle the best answer)

1	2	3	4	5
No	Mild, not too disturbing	Moderate, disturbing but still manageable	Severe, very disturbing	Extreme, disabling

19. Are there things you avoid because of these concerns?
[ ] Yes [ ] No

20. What are the three most important things that you would like to have with this surgery? (Please list in order of importance):
1.
2.
3.

Box 35.2 Standard rhinoplasty photographs, taken preferably in a studio with two flashes

1. Frontal.
2. Lateral right.
3. Lateral left.
4. Oblique right.
5. Oblique left.
6. Basal.

Box 35.3 Facial analysis key points

Frontal view

Brow-tip aesthetic lines.
Deviation.
Skin thickness.
Width of the upper, middle, and lower thirds of the nose.
Tip bulbosity.

Lateral view

Nasal length.
Projection.
Rotation.
Profile changes, such as hump or saddle.
Retraction of the ala or columella.
Nasolabial angle.
Chin.
Submental region.

Basal view

Shape.
Deviation.
Width of the columella and alae.
Caudal septal position.
Height of the lobule.
Columella.

- *Frontal view*: the brow-tip aesthetic lines, deviation, skin thickness, width of the upper, middle, and lower thirds of the nose, and tip bulbosity are evaluated.
- *Lateral view*: nasal length, projection, rotation, profile changes, such as hump or saddle, retraction of the ala or columella, nasolabial angle, and chin and submental region are all assessed and noted.
- *Basal view*: the basal view is evaluated in terms of shape, deviation, width of the columella and the alae, caudal septal position, height of the lobule, and columella.

Imaging

Photographs may be loaded into a computer to show the possible patient profile, projection, and rotation changes using software such as Photoshop or AlterImage. Most surgeons, however, will base their discussion around the standardized views. The author finds morphing software very useful in planning surgery with the patient.

Informed consent

The surgeon must explain all aspects of surgery and possible complications before surgery. This should include a detailed discussion around the intended approach, graft materials, and a thorough discussion around complications, preferably quoting their own figures. The patient should be provided with a copy of the correspondence upon which to reflect, and an information leaflet pertaining to the surgery. It is good practice to see the patient for a second visit prior to surgery allowing for a period of reflection. Anything said before the operation is information, and the things said after the operation are excuses. A consent form must be signed by the patient and surgeon prior to surgery.

Surgery

Anaesthesia

Surgery may be performed under local anaesthetic, intravenous sedation, or general anaesthesia. The majority of rhinoplasty surgeons would prefer a general anaesthetic because it is more comfortable both for the patient and for the surgeon. The operative plan should be marked on the nose using ink, including the incision, dehumping, and osteotomy lines, particularly when the surgeon is first starting their rhinoplasty career. The nose is infiltrated with a mixture of 2% lidocaine and 1:80,000 adrenaline. The incision lines, the osteotomy tracks, the nasal dorsum, and the nasal septum are all injected and before starting surgery. It is suggested to wait 15–20 minutes for better haemostasis. Some surgeons will perform their infiltration in the anaesthetic room. A final facial analysis is performed and the operative plan reviewed before commencing surgery.

Surgical approaches and incisions

There are broadly two approaches: endonasal and external. In the endonasal approach, if the lower lateral cartilages are not disturbed it is called non-delivery but if they are exposed to make changes to the tip, it is called a delivery technique. In the external approach, an additional incision on the columella is used. It has been a debate for decades on which approach is better. There are certain indications, advantages, and disadvantages for each of these approaches (Box 35.4). What matters to the patient is the result. Hence, surgeons should choose the approach that best helps them to obtain an optimal surgical outcome. Rather than being polarized to one approach, a good rhinoplasty surgeon should be familiar with both approaches and use them accordingly.

Profile alignment

In reduction rhinoplasty, getting rid of a hump is an important part of the procedure. Currently there are three main ways for hump removal: (1) simply resecting the hump in one piece (Figure 35.3); (2) separating the upper lateral cartilages from the nasal septum and then resecting the bony-cartilaginous hump from the nasal septum; and (3) resecting the nasal septum under the nasal dorsum by preserving the nasal dorsum and pushing the nasal dorsum down.

When the dorsal hump is removed in one piece, using the Joseph technique, the unique transition between the nasal bones, the upper lateral cartilages, underlying septal cartilage, and ethmoid bone is destroyed. Some of the functional and aesthetic problems which may occur subsequently, such as middle vault collapse, inverted V deformity, and internal nasal valve stenosis, have led surgeons to find solutions, such as spreader graft and flap insertion (Box 35.5).

Box 35.4 The incisions that can be used in rhinoplasty

1. Septal:
   - **a.** Hemitransfixion incision (Figures 35.1a, b).
   - **b.** Partial transfixion incision.
   - **c.** Killian incision.
   - **d.** Complete transfixion.

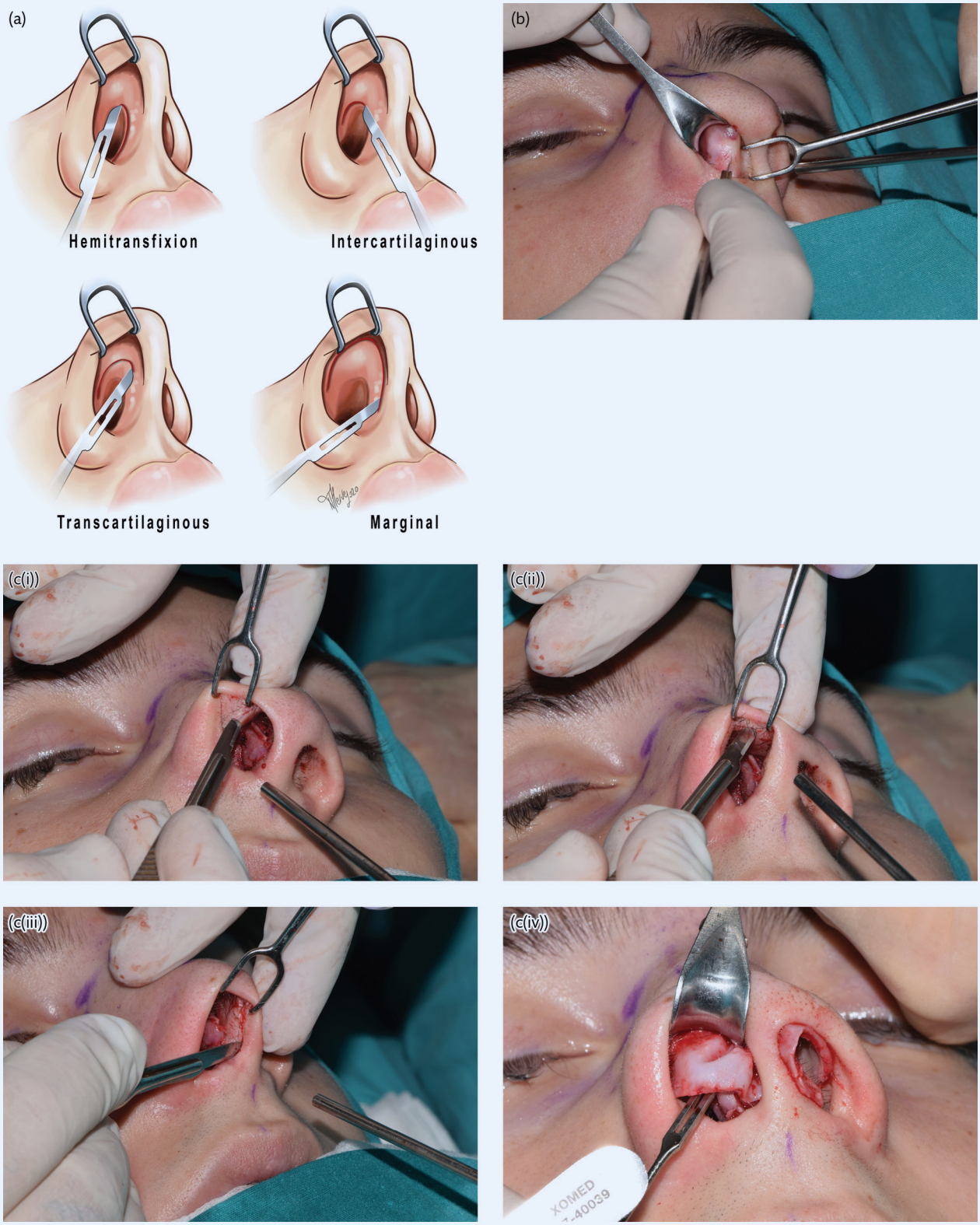

Figure 35.1

2. Alar cartilage:
   a. Intercartilaginous (Figure 35.1a).
   b. Transcartilaginous (intracartilaginous, cartilage splitting) (Figure 35.1a).
   c. Marginal (Figure 35.1a, c).
3. Transcolumellar:
   a. Midcolumellar (Figure 35.2a):
      i. Inverted 'V' incision (Figure 35.2a, b, c, d).
      ii. Stairstep incision.
      iii. 'V' incision.
   b. Lower columellar.

Figure 35.2

e(iii) Wedge resections completed, and the nostrils are more proportional in size and shape.

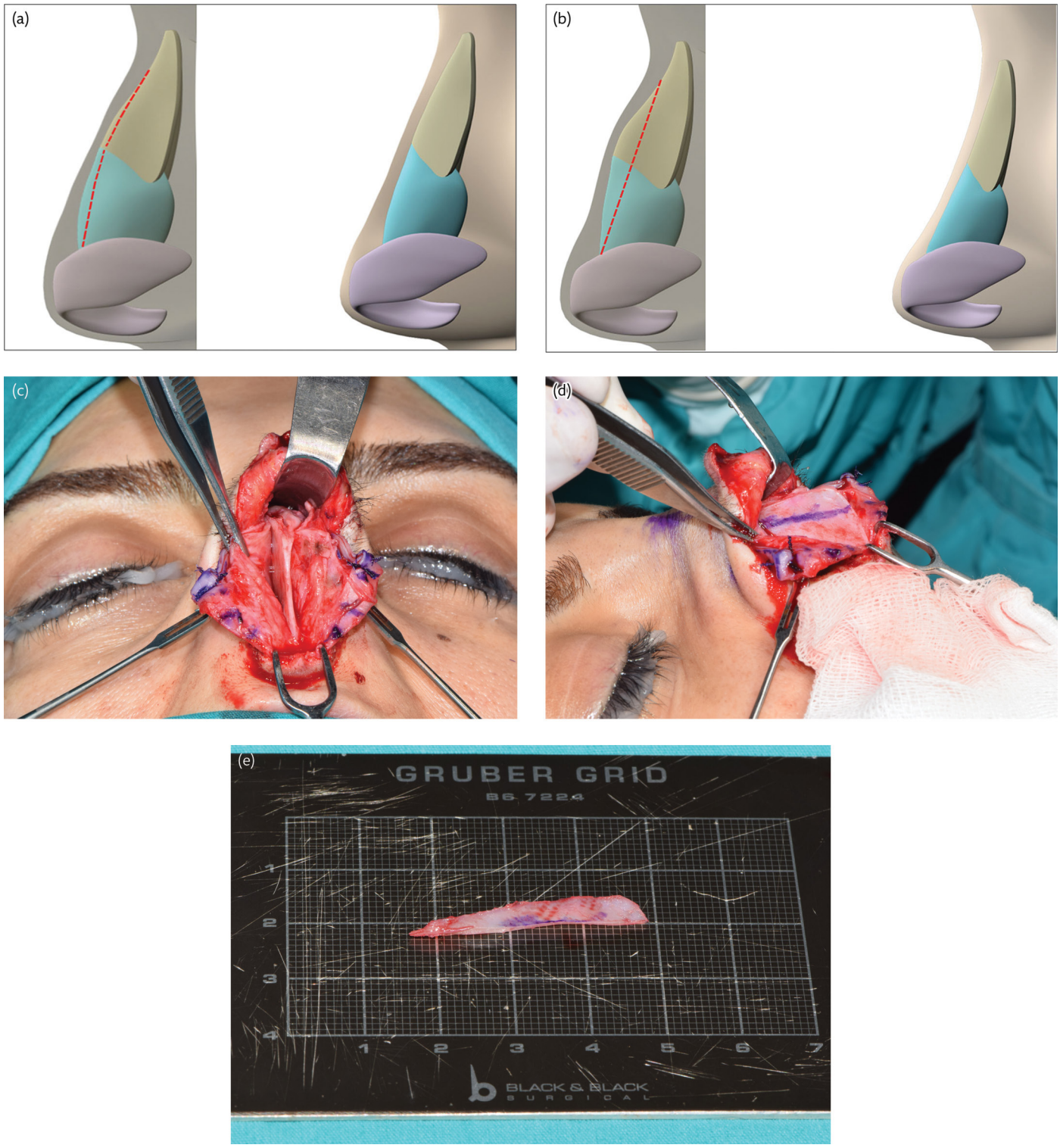

Figure 35.3 Hump removal: (a) and (b) two different approaches to hump removal; (c,d) interoperative photograph of hump removal; (e) excised slice of hump cartilage.

'Preservation' rhinoplasty techniques have been revived over the last 5 years to try to prevent functional problems.

Hump removal may be performed in three main ways:

1. *Composite resection*: the bony-cartilaginous hump is resected in one piece. It is better to detach the mucoperichondrium under the nasal dorsum in order not to sacrifice it because this soft tissue can act as a soft tissue spreader to keep the nasal valves open after hump removal.
2. *Component resection*: the bony cap is rasped and then the upper lateral cartilages are separated from the nasal septum. The septal hump is resected first as required. Then the upper lateral cartilages can be used as a spreader flap or simply resected less than

Box 35.5 Intraoperative reasons for using spreader grafts or spreader flaps

After hump removal, the main reasons to use spreader grafts and/or spreader flaps are as follows:

1. Reconstitute the anatomy of the middle vault.
2. Avoid middle vault collapse.
3. Restore, widen, or maintain the internal valve.
4. Recreate the brow-tip aesthetic lines.
5. Create a smooth transition at the key area.
6. Splint deviated dorsal segment of the nasal septum.

Spreader grafts

Originally described by Sheen, the spreader graft was inserted submucosally in a pocket next to the upper part of the dorsal septum (Figure 35.4). Following popularization of external structure rhinoplasty, spreader grafts were more commonly sutured to the dorsal segment of the cartilaginous septum. In revision cases, they invariably form part of the procedure unless already used in the primary surgery.

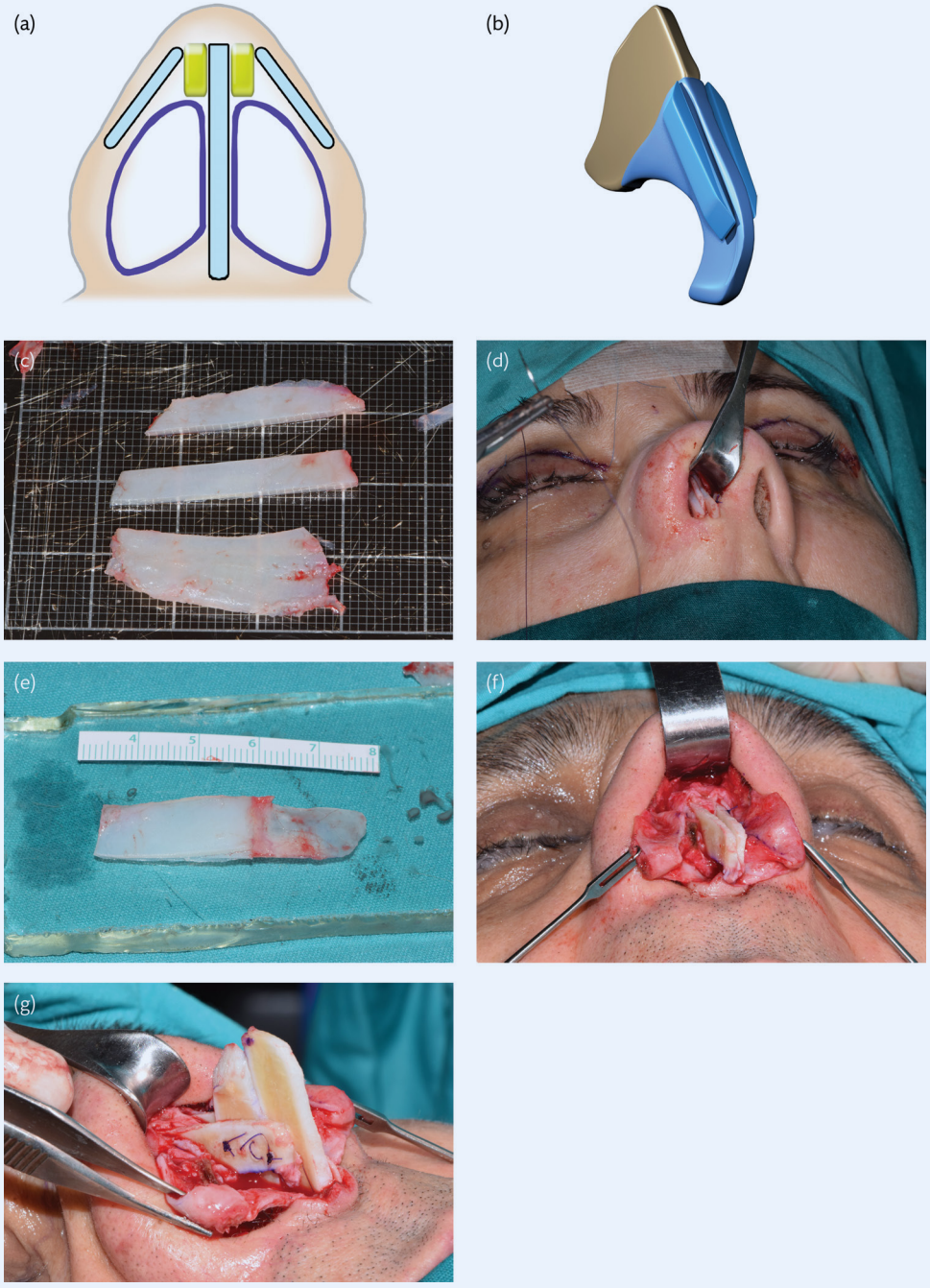

Figure 35.4 Spreader grafts: (a) and (b) diagrammatic illustration of spreader graft placement. (c) septal cartilage spreader graft; (d) spreader graft in situ; (e) bony-cartilagenous spreader graft; (d) rib spreader graft; (e) cartilaginous spreader graft; (f) revision spreader graft (using rib) in situ; (g) revision spreader graft placement.

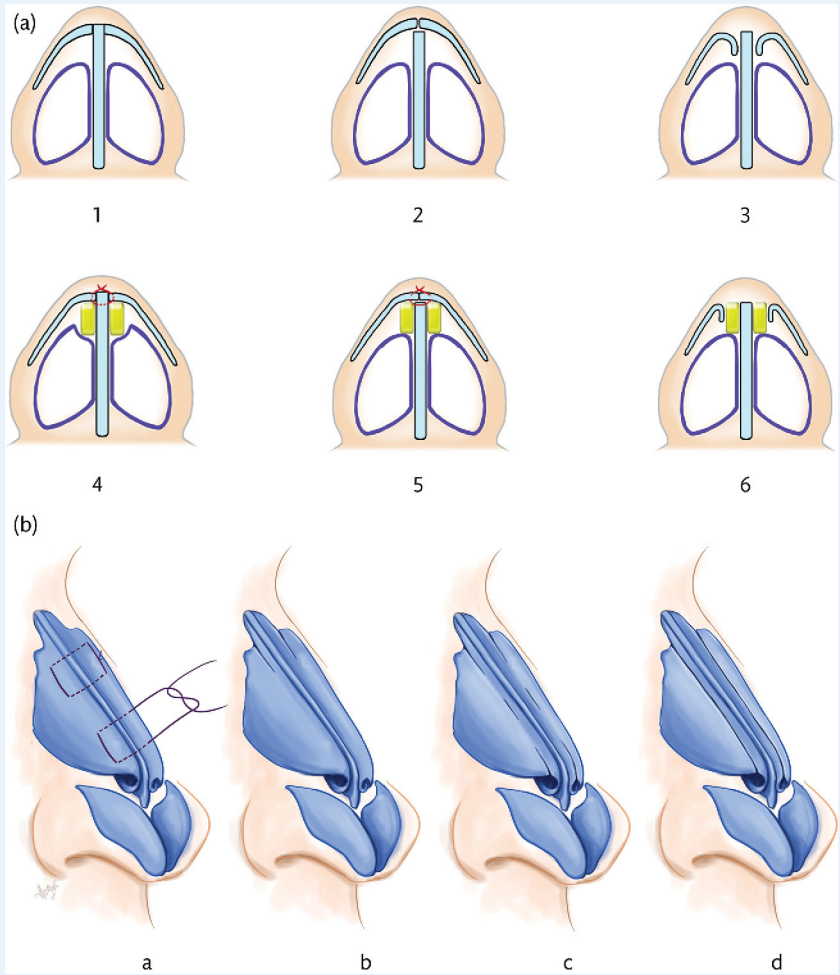

Figure 35.5 Spreader flaps: (a) classification of spreader flaps; (b) different types of spreader flap.

Spreader flaps

Spreader flaps are designed from the upper lateral cartilages dissected away from the septal quadrangular cartilage. They have proven to be versatile and efficient in middle vault reconstruction since their advent over the last decade. They are a very important alternative to spreader grafts. Their major advantage over spreader grafts is that there is simply no need to use an additional graft obtained from anywhere else. They can be used alone or together with a spreader graft (Figure 35.5).

the amount taken from the septal cartilage and spreader grafts are used (Figure 35.3).

3. *Preservation rhinoplasty*: preservation rhinoplasty is a term which has become known within the last few years. It is described by Daniel to have three components: subperichondrial elevation of the canopy of the nose, preservation of ligaments, and preservation of the nasal dorsum. Although the first two have been heavily discussed but not gained wide acceptance, preservation of the nasal dorsum is found to be very interesting by many surgeons and may be used in certain select cases: if the nasal dorsum looks close to ideal but there is a hump, then a dorsum preservation technique may be considered. There are currently three main techniques:
   a. High septal strip resection.
   b. Intermediate strip resection.
   c. Low septal strip resection.

Osteotomies

During rhinoplasty, osteotomies are performed on the bony pyramid for the following reasons:

- Close the open roof.
- Narrow the bony pyramid.
- Correct a deviated or asymmetric bony pyramid.
- Shape the bony pyramid.

Depending on where the bones are cut, there are different osteotomy types (Box 35.6, Figure 35.6):

1. *Medial*: these can be done in two ways: vertically just 1–2 mm away from the nasal septum up to the level of an imaginary line between the medial canthi, called paramedian osteotomies; or the more popular one is the fading medial osteotomy, which starts adjacent to the nasal septum vertically and then directed at an angle of 15–20° laterally and stopping at the same

Box 35.6 Osteotomy types

1. Medial:
   a. Fading oblique.
   b. Paramedian.
2. Lateral:
   a. Low–low.
   b. High–low–high.
   c. Low–high.
3. Intermediate.
4. Transverse.

imaginary line. The main aim of medial osteotomies is to create a certain backfracture line after lateral osteotomies. Thus, if a large hump is removed, especially in females, medial osteotomies may not always be necessary since the roof is already open.

2. *Lateral*: these are usually performed at the level where the bony pyramid joins the cheek. These osteotomies are usually undertaken after the medial ones, and are crucial to mobilize and narrow the bony pyramid to close an open roof. Depending on the dimensions of the piriform aperture, nasal bones and frontal process of the maxilla, the length of a lateral osteotomy varies from 12 to 20 mm. They are classified according to their start, course, and ending as follows:
   a. Low–low: this osteotomy follows a straight line where the bony pyramid joins the maxilla and is an osteotomy through the frontal process of the maxilla.
   b. High–low–high: this osteotomy starts at the upper part of the inferior turbinate to prevent its medial migration after osteotomy so as not to narrow the internal nasal valve and nasal airway. The mid-portion of the osteotomy follows the same trajectory as the previous type, and then is directed high. This is the most popular type of osteotomy.
   c. Low–high: this is a much more rarely used type. As name implies, it starts low and is then directed high.
3. *Intermediate*: when the bony pyramid is too wide and the bony sidewalls are asymmetric as in a crooked nose, an osteotomy between the lateral osteotomy track and the superior margin of the nasal bones may be performed.
4. *Transverse*: transverse osteotomies are performed on a horizontal imaginary line connecting the medial canthi. The aim is to connect the osteotomy lines of the paramedian and lateral osteotomies with a greenstick fracture, which facilitates mobilization of the bony plates.

Lateral osteotomies may be performed externally/percutaneously or internally/intranasally. During an internal lateral osteotomy, the lateral periosteum is left intact if possible because many surgeons use micro-osteotomes of 2–3 mm width with a guide. However, there was a time when larger osteotomes were used and it was necessary to specifically elevate the lateral periosteum to keep it intact. For those using powered instrumentation, such as burs and microsaws, or Piezoelectric osteotomes for osteotomies, the periosteum all over the bony pyramid is elevated for wide exposure and the osteotomies are performed under direct vision. Micro-osteotomes are still the most often used instruments for osteotomies for two main reasons: they are good enough to help the surgeon to make precise osteotomies and they are much cheaper. Powered instruments are more expensive, need personnel for the set-up, and also need expensive consumables.

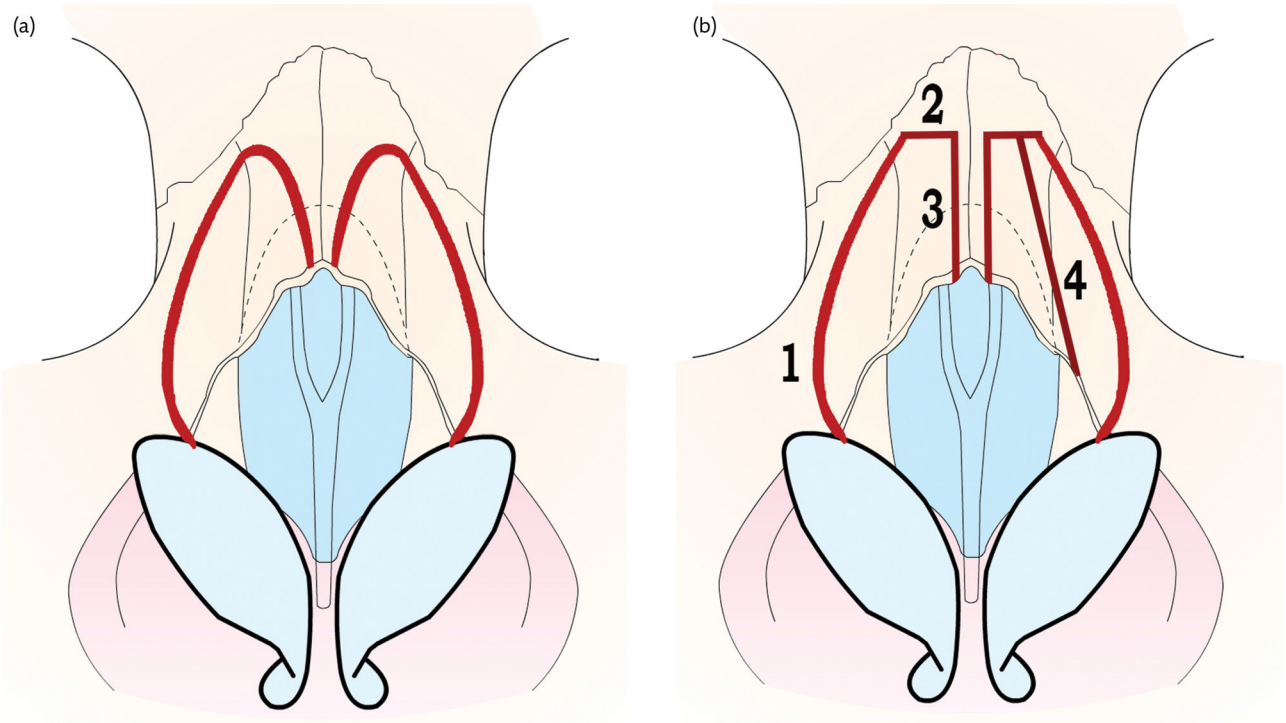

Figure 35.6 Osteotomies: (a) typical position of medial and lateral osteotomies; (b) position of osteotomy variations.

As alluded to above, osteotomes can be used intranasally or percutaneously. The guided osteotomes for medial fading and lateral osteotomies are more frequently used intranasally by experienced rhinoplasty surgeons. For beginners, lateral osteotomies should be done percutaneously for precision.

When deciding on osteotomies, there are some important points to remember and discuss with the patient. It should be borne in mind that the nasal airway will be narrowed, which is a usual end product of reducing the size of a big nose. Furthermore, there will be swelling and possibly bruising around the nose for 7–10 days. Rarely, ecchymoses are seen and may last up to 2 weeks.

Tip surgery

The tip is considered the most difficult aspect of rhinoplasty. There are many techniques to shape and refine the nasal tip:

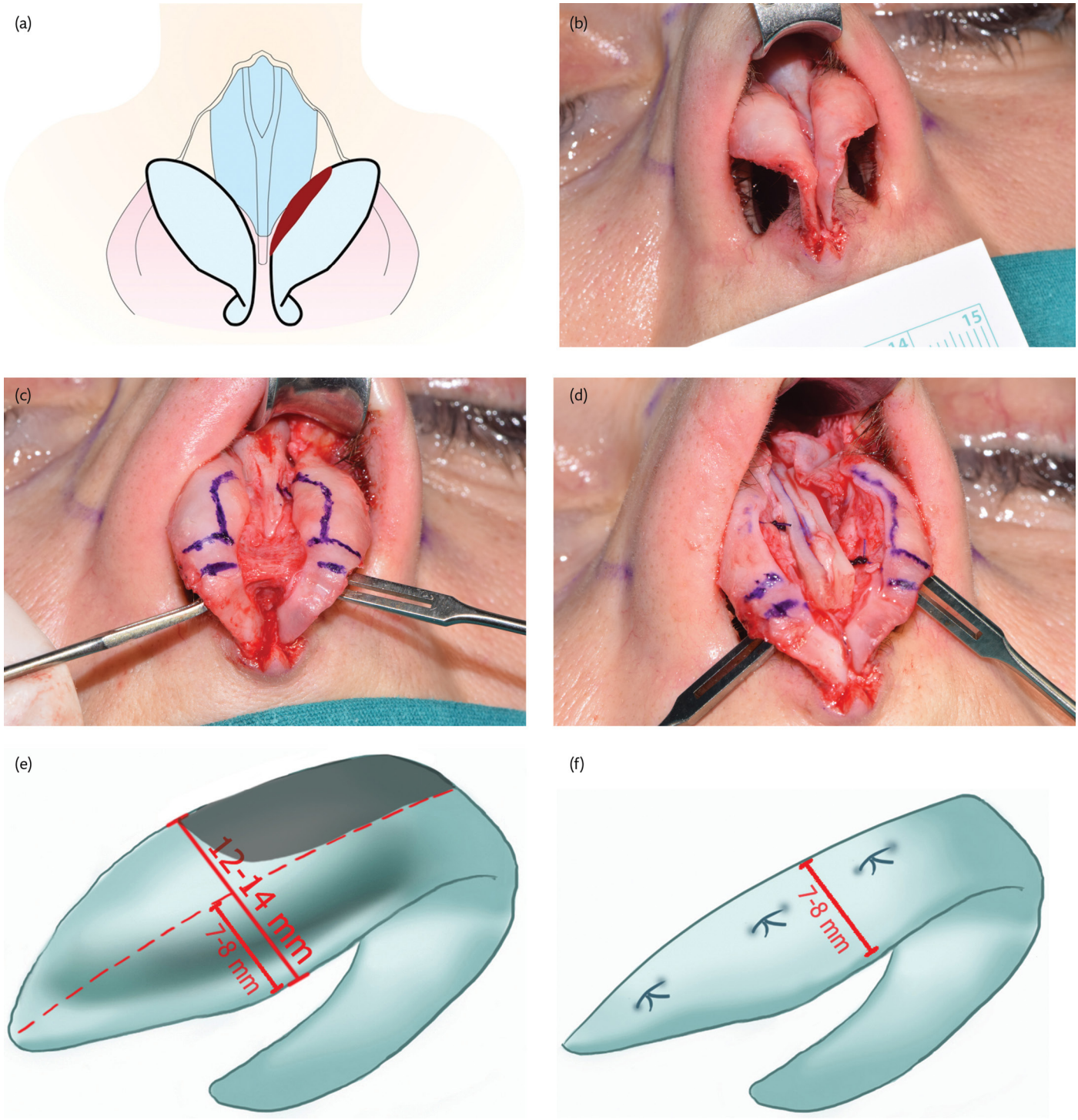

Figure 35.7 (a–l) Sequence of diagrams and interoperative photographs to illustrate the process of devolumising the lateral nasal crus by performing a cephalic trim.

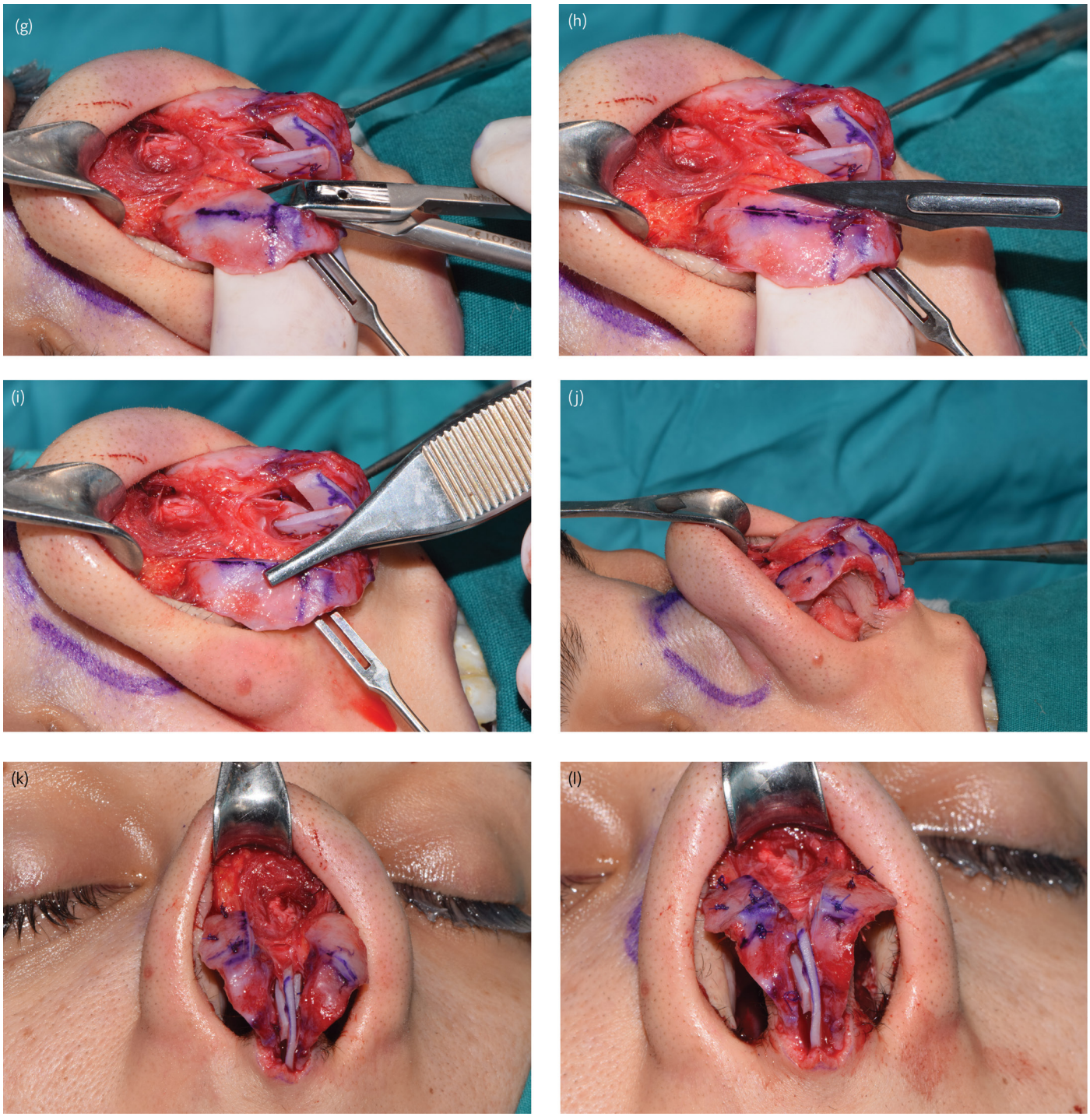

Figure 35.7 Continued

1. *Devolumization of the lateral crus*: a common manoeuvre to make the ala more aesthetic is cephalic trim (Figure 35.7). In order not to inadvertently cause alar collapse, 7–8 mm of the lateral crus must be left in place. A good alternative to cephalic trim is to turn the cephalic part of the lateral crus under the remaining 7 mm lateral crus and suture the two cartilages like a sandwich. By doing so, the lateral crus is reshaped more favourably and is also supported (Figure 35.7).

2. *Suture techniques*: the most popular method to reshape the nasal tip is to use sutures (Box 35.7 and Figure 35.8). The most often used sutures are described below (Box 35.8):

   a. Domal suture: this is the most popular suture to reshape the dome. It narrows the domal arch and pulls the lateral crura medially. If 2–4 mm of lateral crus is 'stolen' from laterally while using this suture, a new dome is formed. Projection and rotation are increased at the same time. This so-called

Box 35.7 Indications for tip sutures

1. Narrowing of a bulbous nasal tip with an obtuse domal angle.
2. To decrease the interdomal angle (bifid tip).
3. To increase tip projection.
4. To increase tip rotation.
5. In patients with thin skin and strong cartilages.

Box 35.8 Tip sutures

1. Domal suture.
2. Dome-equalizing suture.
3. Transdomal suture.
4. Septocolumellar suture.
5. Tongue-in-groove.
6. Vector suture (modified lateral crural spanning suture).

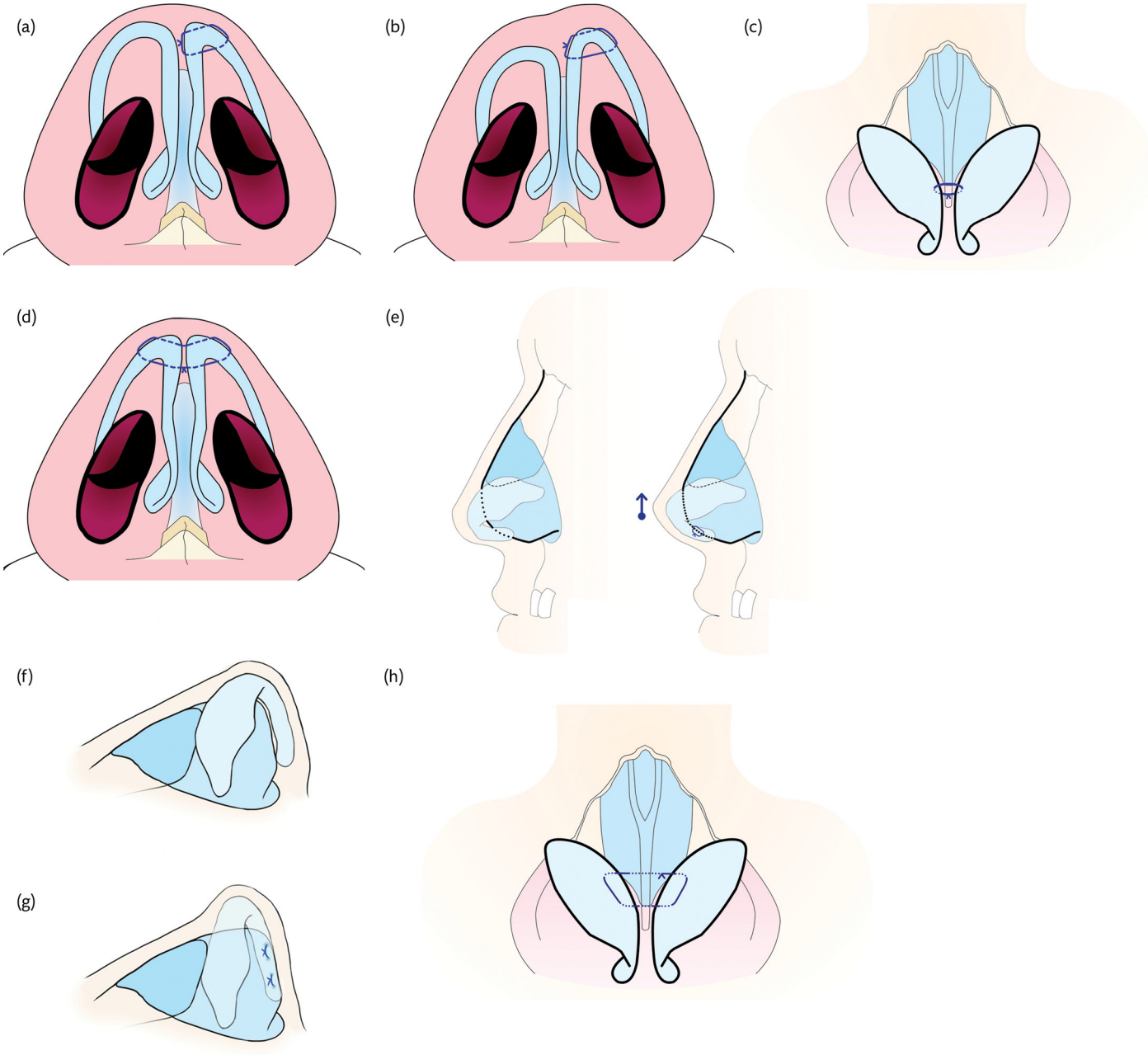

Figure 35.8 Suture techniques to reshape the nasal tip: (a) domal suture; (b) lateral domal steal suture; (c) domal equalising suture; (d) transdomal suture; (e) septocolumellar suture; (f) and (g) tongue-in-groove suture; (h) vector suture.

lateral crural steal suture is very popular in patients that have a droopy tip. Hemitransdomal, orient dome, cranial tip, and high domal sutures are all variations of this suture to prevent pinching of the domal angle and to change the longitudinal axis of the latera crus to a more favourable position.

b. Dome-equalizing suture: this suture passes from the cephalic edges of the both domes to approximate and equalize the domes.
c. Transdomal suture: a mattress suture passing from the midportion of both domes to approximate the domes.
d. Septocolumellar suture: a mattress suture passing between the septum and medial crura in order to make changes in projection and rotation of the tip. This is a powerful suture.
e. Tongue-in-groove: the medial crura are sutured to the caudal septum in such a way that the caudal margins of the medial crura and the caudal margin of the caudal septum are on the same plane. This is a very powerful suture only used when the caudal septum is long enough or when there is a caudal septal extension graft. There is a risk of stiffness of the tip, a compromise while obtaining a more predictable projection and rotation of the tip.
f. Vector suture (modified lateral crural spanning suture): if the lateral crura are convex and far from the caudal margin of the upper lateral cartilages, this mattress suture passing from the lower part of the nasal dorsum and from the cephalic margin of the lateral crura helps to flatten the lateral crura and approximate the cephalic margins to the midline. This is a very powerful suture and may inadvertently elevate the nostril arches.

3. *Tip grafts*: these are used to treat tip asymmetry, increase projection, change rotation, add structure, and reshape the lateral crura. The most popular tip grafts in use are described below (**Box 35.9** and **Figure 35.9**):

a. Columellar strut: a rectangular or slightly curved graft sutured between the medial crura. It is used to support the tip, to increase projection, and to correct medial crural anomalies. In external rhinoplasty, it is routinely used to support the weakened tip cartilages in order not to lose projection and rotation. When a larger graft is sutured to the caudal end of the septum, either end-to-end or in an overlapping fashion, it is called a caudal septal extension graft.

Box 35.9 Tip grafts

1. Columellar strut.
2. Shield graft.
3. Tip (Peck) graft.
4. Buttress graft.
5. Lateral crural graft.
6. Lateral crural strut graft.
7. Alar batten graft.
8. Alar rim graft.

b. Shield graft: a graft in the form of a shield to give better tip definition, to camouflage tip asymmetry, and to increase projection. It is most often used in revision surgeries.
c. Tip (Peck) graft: a graft sutured onto the domes to increase projection and to give better tip definition.
d. Buttress graft: a graft sutured just above a shield graft to prevent its cephalic migration and to obtain an aesthetic transition to the supratip.
e. Lateral crural graft: used when the lateral crura are partially missing or to obtain a smooth transition to the alae when a shield graft is used to increase projection.
f. Lateral crural strut graft: a graft sutured under the lateral crus for a better shape, to support if there is a need to reposition, or sometimes to reconstruct the lateral crus.
g. Alar batten graft: described to support external and internal valves when there is alar collapse or internal valve stenosis. Their use for internal valve problems is limited because the patient can often see and palpate the graft in the longer term, which may become aesthetically unacceptable. However, they are very helpful to support and reshape the alae.
h. Alar rim graft: alar rim grafts are inserted in a pocket along the alar rim when there is an alar retraction, alar contour anomaly, or alar collapse. They can be used as a floating graft or sutured to the lateral crura as articulated grafts.

4. *Alar repositioning*: when the lateral crura are cephalically oriented, the alar compartments are empty and can collapse. In cases such as these, they may be elevated and repositioned in a pocket directed towards the lateral epicanthi.

Closure of incisions, packing, and splints

At the end of surgery, all incisions should be sutured. The endonasal incisions are usually closed with absorbable suture material, such as 5-0 Vicryl Rapide. External incisions can be sutured with 6-0 Prolene or Vicryl Rapide. A quilting suture to close dead space and prevent haematoma formation, using 4-0 Vicryl Rapide on a 13 mm Keith needle, is very helpful. If the surgeon feels that a quilting suture is good enough at bringing the mucoperichondrial flaps together, an intranasal splint or packing is not mandatory. However, in crooked or difficult noses, it may be better to insert intranasal Doyle splints to stay in place for 2–7 days, depending on how extensive the septal surgery was.

After applying Steri-Strips with great care and attention on the skin, a thermoplastic splint is usually applied to stay in place for 7 days. In a crooked nose correction, keeping the splint *in situ* for 10 days may be a better option to help with stability.

Special situations

Crooked nose

Usually due to trauma, less often congenital. It may be linear, C-shaped, or S-shaped (**Figure 35.10**). In the case of a hump, there should be less rasping on the deviated side. Deviations of the bony pyramid can be corrected by double osteotomies (intermediate and lateral) or wedge resection on the non-affected side. Very rarely, a

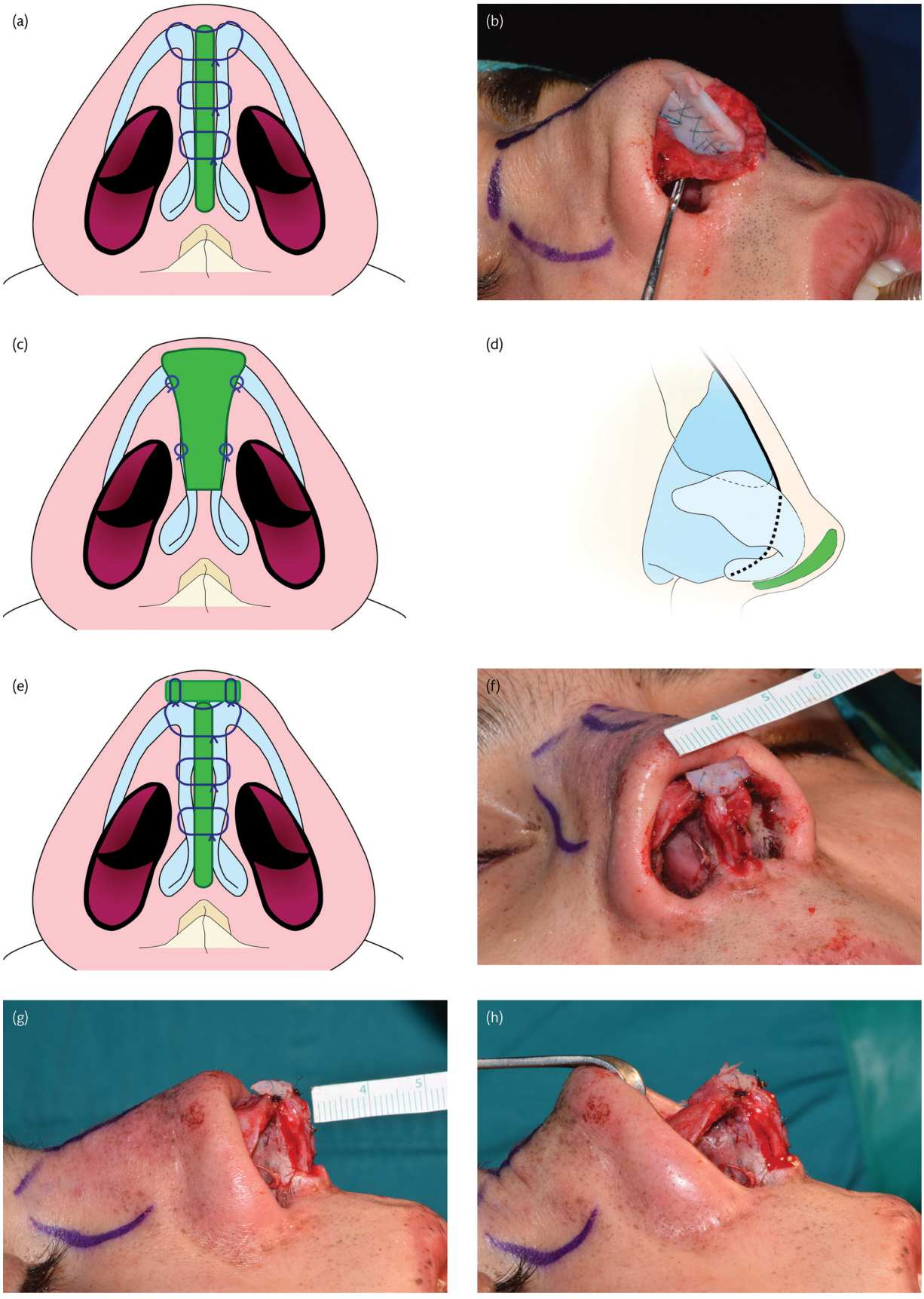

Figure 35.9 Tip grafts: (a) and (b) columellar strut; (c) and (d) shield graft; (e), (f), (g), and (h) tip graft; (i) buttress graft; (j), (k), and (l) lateral crural graft; (m) alar batten graft; (n) and (o) alar rim graft; (p) and (q) preparation and placement of cartilage chips for camouflage.

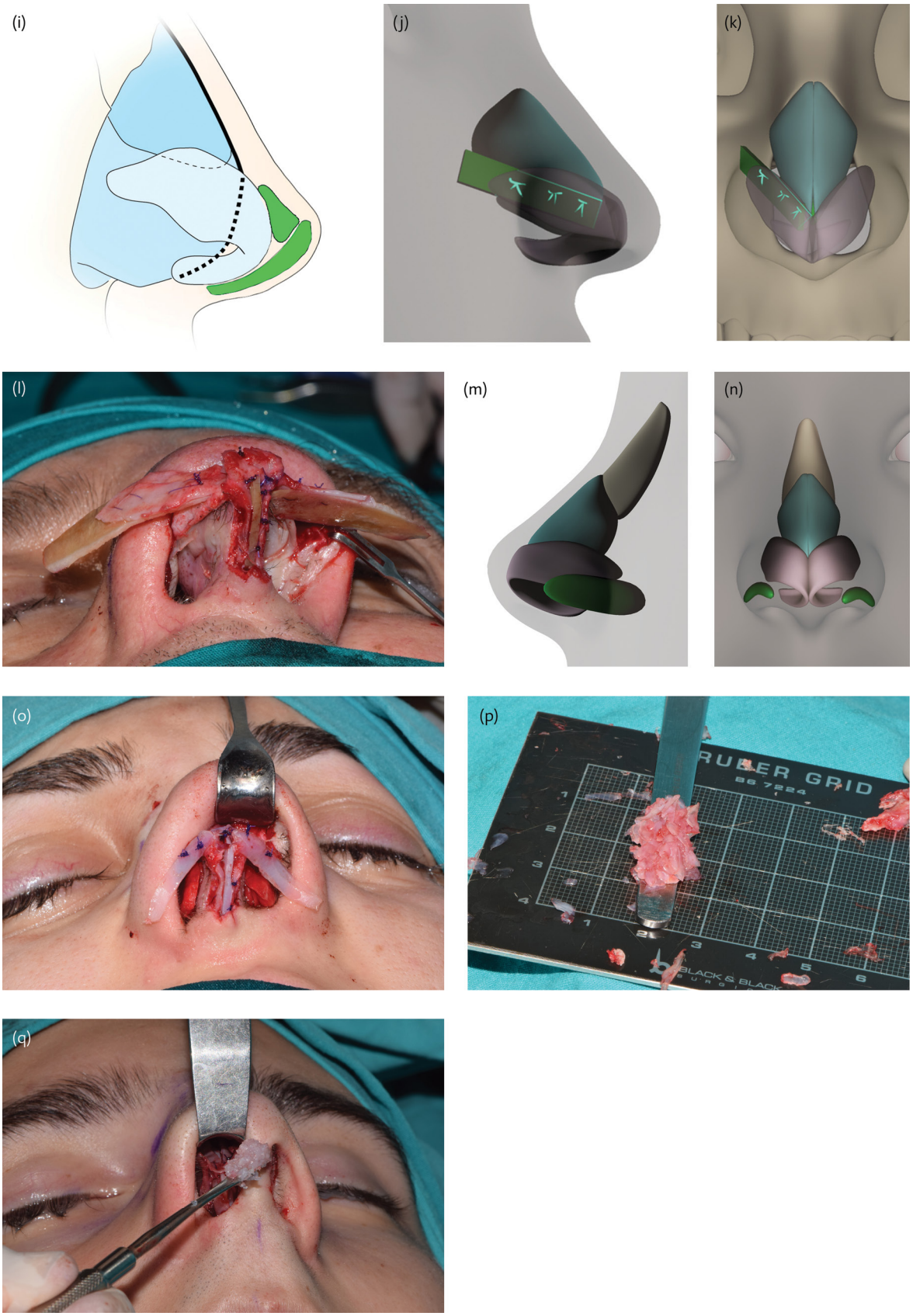

Figure 35.9 Continued

root osteotomy may be needed at the radix to mobilize the midline anatomy.

A difficult challenge in the crooked nose is correction of the deviated nasal septum. In such cases, 'segmental septal reconstruction' may guide correction:

1. *Dorsal segment*: deviations of the dorsal segment cause a crooked nose such as a C-shaped or S-shaped deformity. The workhorse of reconstruction of these deviations is the use of splinting spreader grafts, which also aid in the reconstruction of the middle vault.
2. *Caudal segment*: problems that require correction are subluxation, concavity, retraction, or a fractured segment. There are usually two approaches: splint the existing caudal segment using a batten or caudal septal extension graft, or replace it with a straighter graft.
3. *L-strut framework*: in the case of a severely traumatized nose and/or a crooked nose, the septal cartilage may be very severely deviated. The dorsal segment, caudal segment, and their junction may be very concave. To help, after cross-hatching, the concave side of an L-strut graft of perpendicular plate of ethmoid lamina, vomer, or cartilage may be a good option to splint the L-strut framework.
4. *Subtotal reconstruction*: in severely traumatized noses, very rarely the L-strut cannot be left in place due to multiple fractures. Most of the cartilaginous and bony septum is removed during surgery. The key point here is to leave part of the dorsal segment adjacent to the key area intact as the dorsal segment of the newly created L-strut is sutured to this remaining cartilage for adequate fixation. Two options exist for a subtotal septal reconstruction: either to reconstruct the nasal septum as one piece, which is rare, because it is difficult to obtain such a substantial graft; or, to use two splinting spreader grafts coupled to a caudal septal replacement graft reconstructing the L-strut. In case of cartilage deficiency, cartilage from the ear concha or rib can be used.

Saddle nose

This can be iatrogenic or, rarely, congenital. In mild to moderate cases, augmentation can be performed using septal and/or ear cartilage. In more severe cases, cartilage implants obtained from rib cartilage should be used. An oblique split technique enables the surgeon to obtain multiple implants of various length and of varying thicknesses. They are first used to reconstruct the nasal septum and then build the nose onto the underlying scaffold. Dorsal augmentation can be performed using one solid graft, suturing the two or three laminae of implants, and diced cartilage in fascia (**Figure 35.11**). In the Far East, the use of allografts is widespread and with success. This situation is not widely accepted by facial plastic surgeons in Europe, America, and the Middle East.

Endoscopic rhinoplasty

The endoscope should be available during rhinoplasty for the purpose of intranasal examination, for recording procedures performed inside the nose, and for operating on concurrent pathology if reasonable. Could the whole rhinoplasty procedure be performed endoscopically? Yes, it is possible. However, it is neither necessary nor is it practical. There are perhaps less than a handful of surgeons in the world performing endoscopic rhinoplasty.

Revision surgery

All surgeons try the best for their patients but, unfortunately, surprises related to wound healing, accidental small mistakes, and missed deformities during the primary surgery may necessitate revision surgery. Approximately 10–12% of rhinoplasty patients may need a revision procedure. Bear in mind that you are dealing with patients who are disappointed, sometimes angry, asking, 'Why me?' They may even accuse you that it is your fault, a fact that is actually true. The first challenge in dealing with a dissatisfied patient is to try to cool the emotions involved in the consultation and then, prepare him/her for a revision operation. Good documentation and photography are essential. Never abandon your dissatisfied patient, especially in the early postoperative period. Listen to the patient very carefully to understand the reasons for their frustrations. Often it is just a minor defect which can be solved very easily, either non-surgically or surgically.

Revision rhinoplasty is inherently difficult because the surgeon has to deal with scarred tissues. In addition, if it is not your case and patient primarily, there are usually not any operation notes regarding the first operation. Your patient should be referred for a second opinion if you are not experienced enough. Some patients may be in a hurry to correct the problem and may put pressure on you. You should tell them that the best time for revision is at least 6 months and preferably 12 months after surgery.

The main reasons for revision are the deviated nasal septum, depressions or residual bumps on the nasal dorsum, loss of tip projection, and under-/over-rotation. In order to correct these problems, there may be a need for cartilage, which is obtained from the ear or rib. Detailed information must be provided on these potential donor sites, and informed consent *must* be taken before surgery. Revision operations usually take longer and are more complex operations than the primary operations, and are less likely to have a satisfactory outcome—a situation that must be discussed with the patient before surgery. In addition to the use of structural grafting techniques, camouflage techniques using soft tissues, such as perichondrium or fascia (temporalis, rectus abdominis), cartilage dust, and/or chips, may be needed.

Postoperative follow-up

During the immediate postoperative period, analgesia, such as paracetamol, and antiemetic agents, such as ondansetron, should be used to diminish patient anxiety. Cold compresses applied around the nose can start immediately after surgery.

During the first postoperative week the patient is prescribed a prophylactic antibiotic due to the grafts placed at the time of surgery, such as amoxicillin/clavulanate, an oral decongestant, such as pseudoephedrine, and analgesia for 3–4 days, such as paracetamol or paracetamol with codeine. If there is no packing inside the nose, topical use of decongestant sprays (oxymetazoline) is recommended. Cool compresses to prevent or reduce swelling and ecchymosis should be used for 3–4 days intensively.

Absorbable sutures are very effective in rhinoplasty surgery. In some selected cases of revision rhinoplasty, or when there is slight

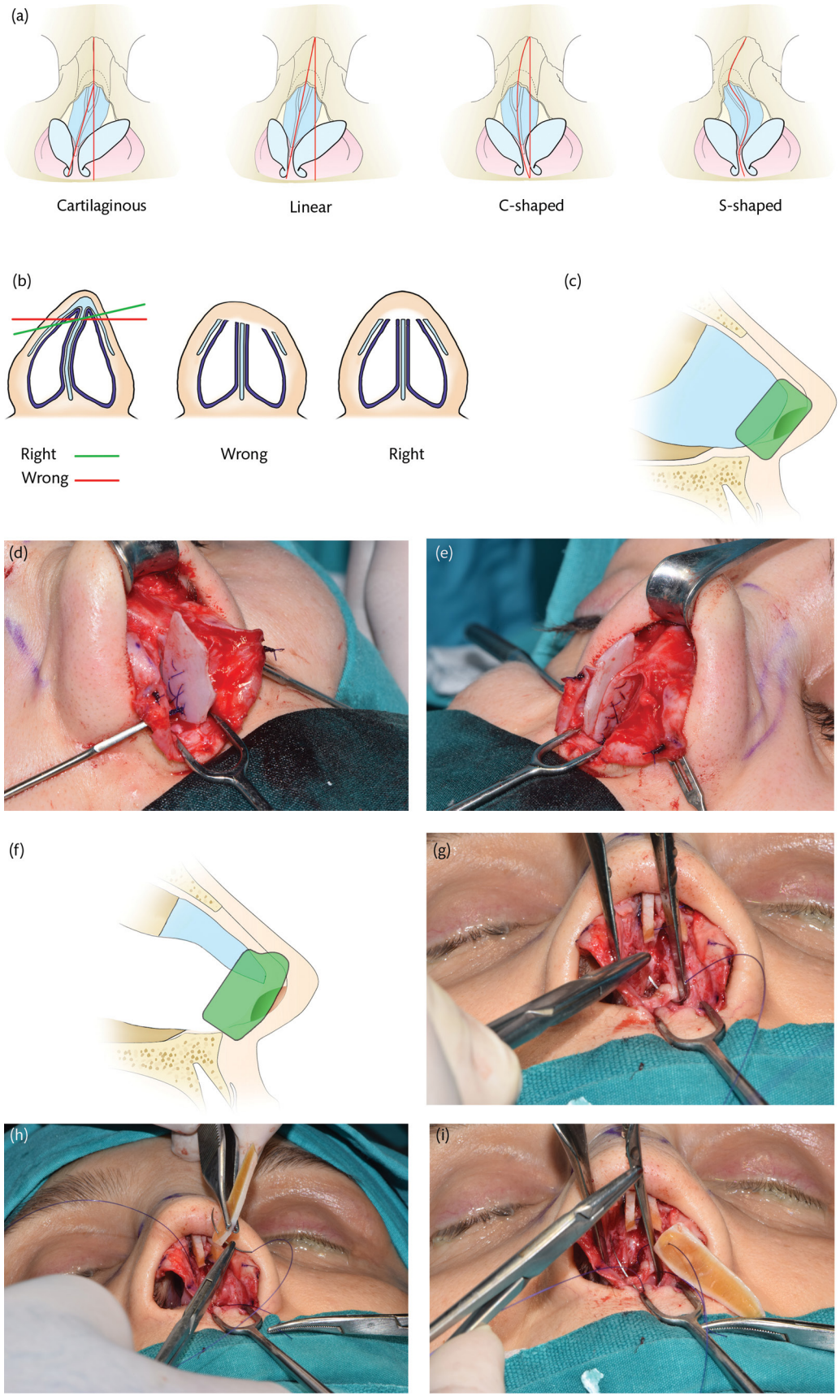

Figure 35.10 The crooked nose: (a) types of crooked nose; (b) dehumping technique; (c), (d) and (e) technique for septal extension; (f), (g), (h), (i), and (j) technique for septal replacement; (k), (l), (m), (n), (o), and (p) technique for placing a septal strut graft; (q) subtotal reconstruction of the nasal septum.

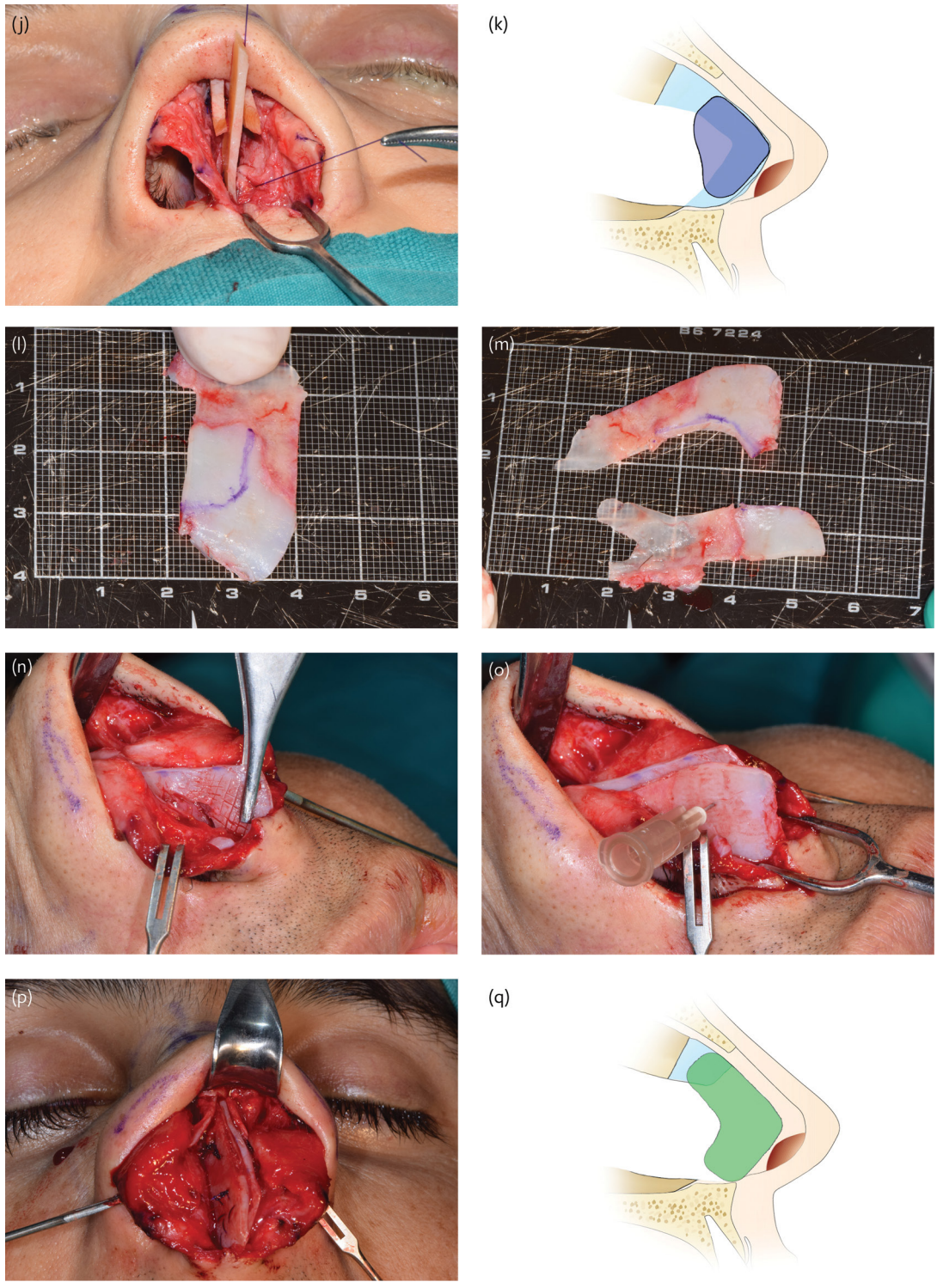

Figure 35.10 Continued

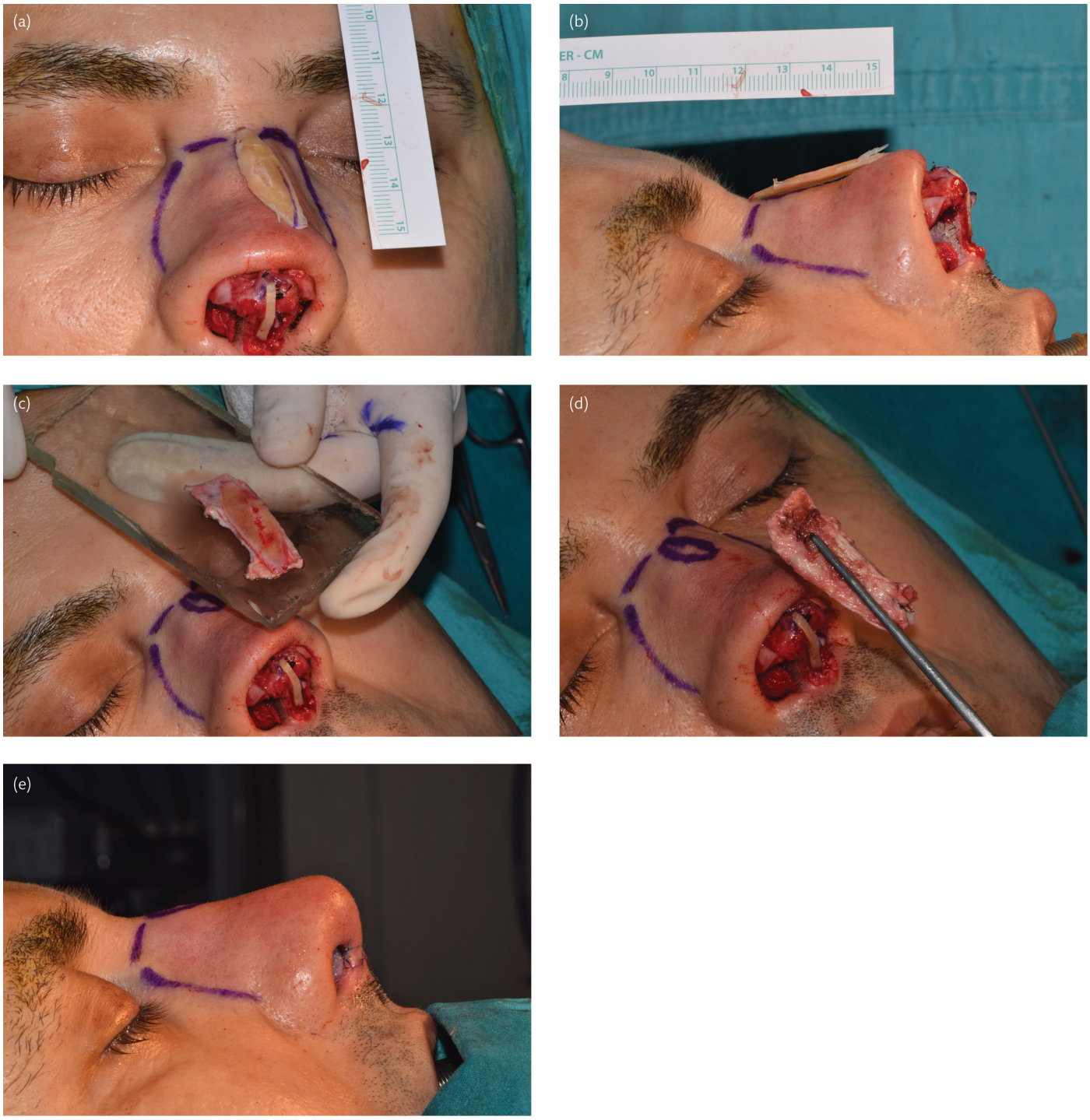

Figure 35.11 (a)–(e) Technique for correct a saddle nose deformity by dorsal augmentation.

tension at the wound edges after closure, 6–0 Prolene is helpful, but this needs to be removed after a week. This can be uncomfortable for the patient.

Instructions at home

Most septorhinoplasty surgery is performed as a day case procedure but patients require somebody to stay with them for the first night and require an adult to escort them home. They should stay off work for at least 2 weeks, avoid strenuous exercise for 4 weeks, and avoid all contact sports for at least 6 weeks.

At home, patients are advised to sleep with their head elevated with at least two pillows for the first week. Postoperative bruising and swelling is a frequent concern for patients and reassurance is provided that their nose will continue to remain swollen for several weeks and up to a few months after surgery; the nasal tip in particular can take around 12 months to refine, longer in revision cases.

Follow-up

The first postoperative appointment usually takes place at 2–7 days depending on whether intranasal splints were inserted. The splint is usually removed 1 week after surgery and an additional taping of 1 more week can be used to minimize swelling. The patient is scheduled to come back to clinic at 2 weeks, 6 weeks, 3 months, 6 months, and 12 months after surgery. The patient should be reminded that swelling steadily dissipates over 6–12 months (Figure 35.12). Expectations can be difficult to manage.

At the 6-month visit, all patients are asked to complete a patient-reported outcome measure (**Box 35.1**) again. Postoperative clinical photography is undertaken at 3, 6, and 12 months and then every year after surgery if the patient is motivated to return.

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36

Acute rhinosinusitis

Isam Alobid

Introduction

Acute rhinosinusitis (ARS) is a symptomatic inflammation of the mucosal lining of the nasal cavity and paranasal sinuses, where clinical symptoms have been present for less than 4 weeks. It can be caused by either a viral or a bacterial infection.

Rhinosinusitis is one of the most common health complaints leading to a physician visit in the US, as well as one of the leading causes of antibiotic prescriptions. More than 20 million cases of acute sinusitis of viral or bacterial aetiology are diagnosed in the US each year, affecting an estimated 16% of the adult population and causing up to 73 million restricted activity days in 1 year in the US due to rhinosinusitis. The estimated annual cost of treatment in the UK is £10 million (€14.7 million, US$20 million).

Bacterial rhinosinusitis is greatly overdiagnosed with concomitant overuse of both diagnostic tools and antibiotics, with up to 60% receiving a course of antibiotics on day 1 of an event. Primary care physicians often tend to think of sinusitis as an acute bacterial infection and antibiotics are prescribed in 92% of patients in the UK and 85–98% of sinusitis patients in the US. Furthermore, early administration of antibiotics appears to have little or no bearing on the development of complications of ARS. Thus, an accurate diagnosis depends on a thorough clinical assessment of patients, as commonly performed in the primary care settings by a general practitioner in the UK.

Classification of acute rhinosinusitis

Rhinosinusitis is usually classified as acute (≤4 weeks or less), subacute (4–12 weeks), recurrent ARS (four or more episodes per year lasting at least 7 days each, in any 1-year period), and chronic rhinosinusitis (≥12 weeks). However, the European Position Paper on Rhinosinusitis and Nasal Polyps 2020 (EPOS2020) group felt that the term subacute rhinosinusitis used to describe patients with prolonged ARS is not necessary because of the small number of patients that comprise this group, and hence the very little data on which to offer evidence-based recommendations on how to manage these patients.

Aetiology

The most common cause of ARS is a viral infection. The majority are self-limiting and can be classified as common colds. Common colds are defined as acute viral rhinosinusitis with a duration of symptoms of less than 10 days. When symptoms increase after 5 days, or when symptoms are persistent for more than 10 days, with less than 12 weeks' duration, then EPOS2020 classifies this as acute/subacute postviral rhinosinusitis.

Following an episode of viral sinusitis, 0.5–2% of cases will progress to acute bacterial sinusitis. The three most common bacteria are *Streptococcus pneumoniae* (20–43% of cases), *Haemophilus influenzae* (22–35% of cases), and *Moraxella catarrhalis* (2–10% of cases). The latter is less common in the adult population. In a study done in a single centre in the UK, *Streptococcus anginosus* was the microorganism most often involved in complicated acute bacterial rhinosinusitis (ABRS) (61%).

Although the bacterial pathogens have not changed over time, their antibiotic-resistance patterns have changed. The prevalence of penicillin-resistant *S. pneumoniae* has increased from 24% to 35%, and the prevalence of macrolide-resistant *S. pneumoniae* has increased 9% to 39%, both over a 10-year period. Penicillin resistance is much higher for *H. influenzae* and *M. catarrhalis*, sometimes approaching 100%.

Pathophysiology

ARS is most likely to be caused by the combination and interaction of environmental and host factors (**Table 36.1**) linked to a viral infection. This results in an inflammatory response, oedema, and mucus production that obstructs sinus drainage pathway and blocks normal ventilation. With this decreased mucociliary clearance and mucus plugging, a secondary bacterial infection can take place.

Diagnostic approach step by step

- Diagnosis of ARS is based on history and physical examination.
- Most acute viral upper respiratory tract infection (common colds) are self-limiting, so postviral ARS should not be diagnosed before 10 days of symptoms unless there is a clear worsening of symptoms after 5 days.

Table 36.1 Predisposing factors for acute rhinosinusitis

Anatomic defects	Septal deviation, polyps, conchae bullosa, trauma, and fractures involving the sinuses or surrounding structures
Impaired mucous transport	Cystic fibrosis, ciliary dyskinesia
Immunodeficiency	Chemotherapy, HIV, and diabetes mellitus
Intensive care unit	Body positioning compromising mucociliary clearance, prolonged oxygen use causing mucosal lining drying, or a foreign body like a nasogastric or nasotracheal tube
Chronic inflammation or drug abuse	Rhinitis medicamentosa, toxic rhinitis, nasal cocaine abuse, and barotrauma
Environmental factors	Allergens (animal dander, polluted air, smoke, and dust), irritants, viruses, fungi, and bacteria

Step 1: history

A history of an upper respiratory tract infection or exacerbation of a known allergic rhinitis is generally the main complaint. It is important to take into account that ARS may cause acute exacerbation of asthma or migraine in patients with these conditions.

The subjective assessment of ARS in adults is based on the presence and severity of symptoms, mainly:

- nasal blockage or congestion
- nasal discharge or postnasal drip
- facial pain/pressure or headache
- reduction/loss of smell.

The first and most important initial approach is a thorough evaluation of the symptom duration and trajectory in order to differentiate viral from bacterial sinusitis (Table 36.2) This will determine the next steps and management. Viral infection tends to peak early and gradually resolve in the next 5–10 days. So as a first general rule, symptoms present for less than 10 days are characteristic of a viral infection, whereas symptoms present for more than 10 days without any improvement suggest a bacterial infection. Symptoms that worsen after an initial improvement (called 'double sickening') also suggest secondary bacterial infection.

Not only duration but other specific symptoms help distinguish between viral and bacterial sinusitis.

Table 36.2 Clinical characteristics of viral and bacterial acute rhinosinusitis

Acute viral rhinosinusitis	Fever, sore throat, myalgia, or a clear nasal discharge
Major factors in ABRS	Purulent nasal discharge, nasal obstruction, facial pain/pressure, facial congestion/fullness hyposmia, and fever (in the acute period)
Minor factors in ABRS—diagnostically significant only with one or more major factors	Dental pain (unilateral facial or dental pain have been found to be predictors of acute maxillary sinusitis), headache, halitosis, fatigue, malaise, cough, and otalgia

It is important to consider that the colour of the mucus as a sole indicator does not distinguish bacterial from viral aetiology. Cough is a common symptom in acute viral and bacterial sinusitis and may occur secondary to postnasal drainage or as a consequence of asthma exacerbation.

Step 2: physical examination

A complete head and neck examination is mandatory. Complications in ARS are uncommon but need to be suspected and identified early as they may initially present with non-specific symptoms or can go unnoticed if not intentionally searched for: periorbital or malar oedema, orbital proptosis, visual disturbances, abnormal extraocular movements, or abnormal neurological signs may suggest complications, and an urgent otolaryngology consultation should be undertaken.

Generally, the presence of facial tenderness with gentle percussion or palpation, postnasal pharyngeal secretions or exudate, nasal mucosa with erythema and purulent discharge, and cervical adenopathy are seen.

Optimal examination is performed before and after topical decongestant spray, with either a nasal speculum and head light or an endoscope (Figure 36.1). Although anterior rhinoscopy alone is very limited, it should be performed in the primary care setting when suspecting ARS as it may reveal supportive findings and important information such as polyps, anatomical abnormalities, or suspicious lesions that allow early specialist referral for further treatment. Endoscopy can provide excellent visualization of the nasal cavity and sinus drainage pathways. Rigid nasal endoscopy has superior resolution, uses one hand, and allows for cultures. Flexible nasal endoscopy is more comfortable for patients but requires both hands. Both can be used indistinctly.

Step 3: laboratory testing

No laboratory tests are indicated for acute uncomplicated sinusitis. Although in ABRS demonstration of a bacterial infection in the paranasal sinuses is the 'gold standard' for the diagnosis (sinus washout or puncture and culture), it is not easy or widely available so a clinical diagnosis is made and initial empirical antimicrobial therapy is started based on local guidelines and experience. In the

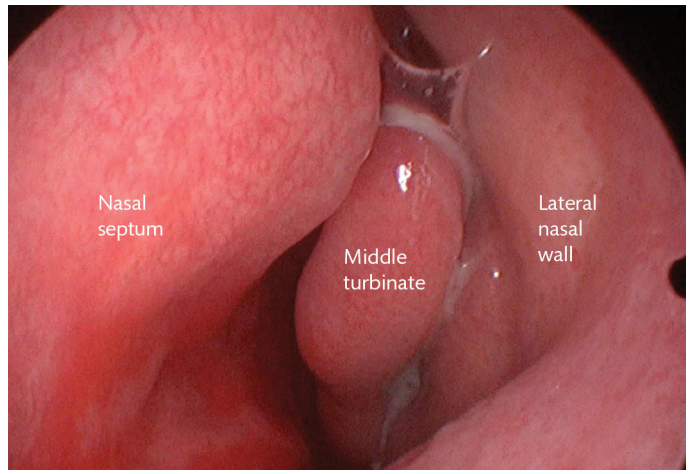


Figure 36.1 Endoscopic view of a left nasal cavity with purulent discharge from the middle meatus.

acute scenario, culture and biopsy are indicated in the immunocompromised patient or as an important aid in antibiotic selection for refractory cases after empirical antibiotic therapy.

C-reactive protein and procalcitonin are haematological biomarkers raised in bacterial infection and are used specifically in respiratory tract infections, the intention being to limit unnecessary antibiotic usage. A low or normal C-reactive protein level correlates with a low likelihood of bacterial infection and unlikely benefit from antibiotics.

Erythrocyte sedimentation rate (ESR) is raised in ABRS and may reflect disease severity, indicating the need for more aggressive treatment. ESR levels have been correlated with computed tomography (CT) changes in ARS; ESR greater than 10 is predictive of sinus opacity on CT scans.

Step 4: radiographic studies

Not recommended in the evaluation of a routine acute sinusitis as cannot differentiate viral from bacterial sinusitis. Recommended when complications are suspected, such as facial cellulitis, orbital or intracranial infection, in the case of recurrent episodes of sinusitis, anatomical abnormalities, or when an alternative diagnosis is suspected.

Non-contrast coronal CT at a thickness of 3–4 mm is the modality of choice (Figure 36.2). Contrast studies are saved for when a complication is suspected in order to differentiate cellulitis from an abscess. Classical findings consistent with, but not diagnostic of, ARS are sinus opacification, air–fluid level, or severe mucosal thickening.

Magnetic resonance imaging (MRI) is useful if extra-sinus complications are suspected, especially central nervous system or cavernous sinus involvement, or a fungal aetiology (Figure 36.3).

X-rays should be supplanted by CT scanning if imaging is required.

Differential diagnosis

Differential diagnoses include allergic rhinitis (the most common mistake), non-allergic rhinitis, maxillary toothache that can also mimic the pain caused by maxillary sinusitis, odontogenic sinusitis (Figure 36.4), adenoiditis, tension headache, migraine or vascular headache, foreign body, brain abscess, epidural abscess, meningitis, and subdural empyema. All can be mistaken for acute sinusitis.

Treatment approach step by step

- Treatment usually aims to resolve and relieve symptoms while the infection resolves itself.
- Antibiotic use must be justified and empirically commenced when indicated.
- Bacterial infection may occur in ARS but, in most cases, antibiotics have little effect on the course of the illness.
- A universal treatment protocol is hard to achieve because it depends on the antibiotic resistance and local availability which varies according to geographic location.
- Poor patient selection increases the risk of unwanted antibiotic resistance and later complications.
- Antibiotics are effective in well-selected patients when symptoms and signs suggest ABRS. Amoxicillin/penicillin (beta-lactams) are effective and clinical improvement is usually evident by day 3.

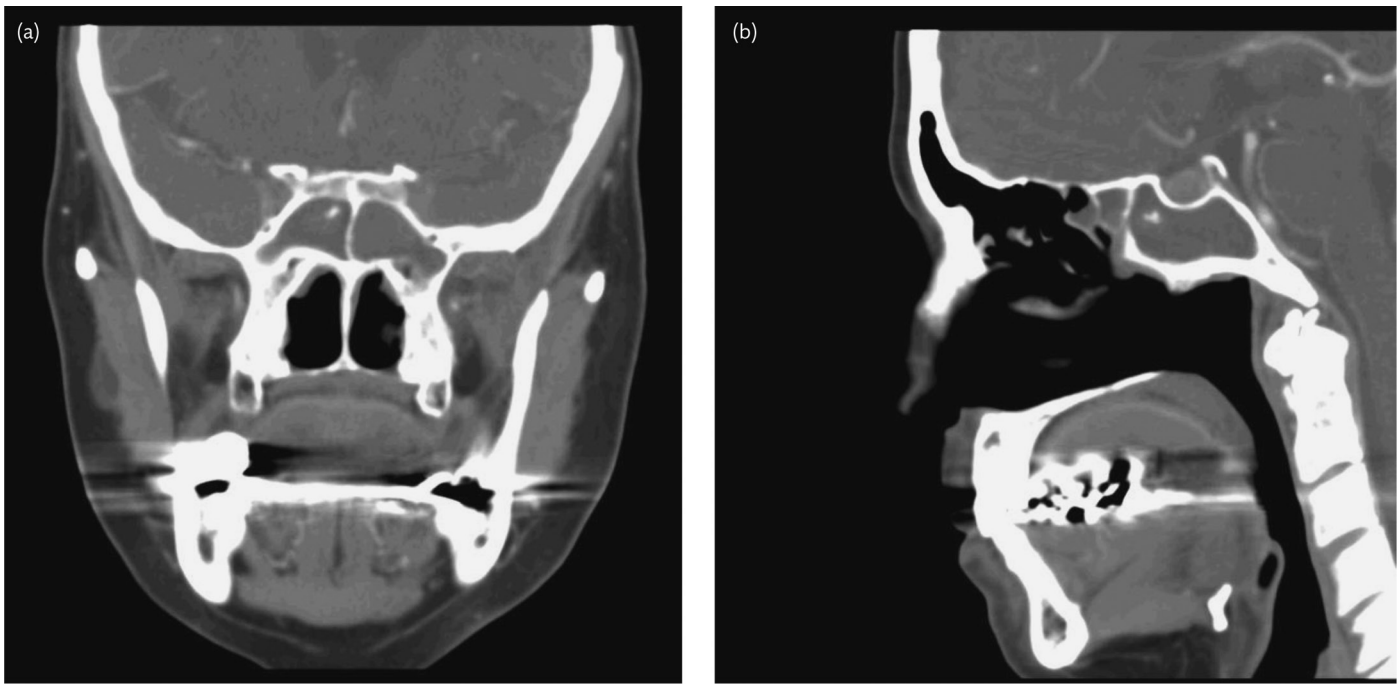
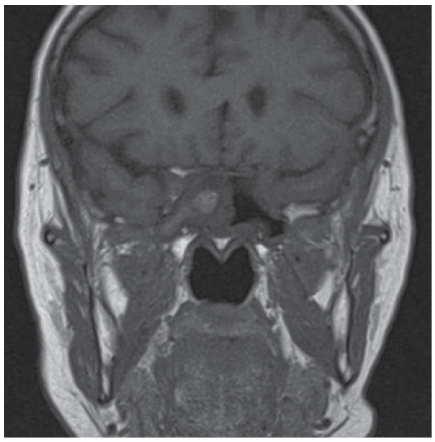
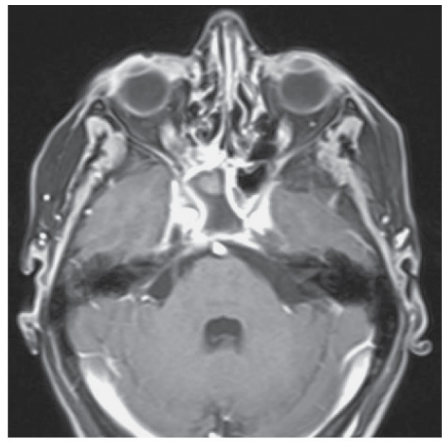


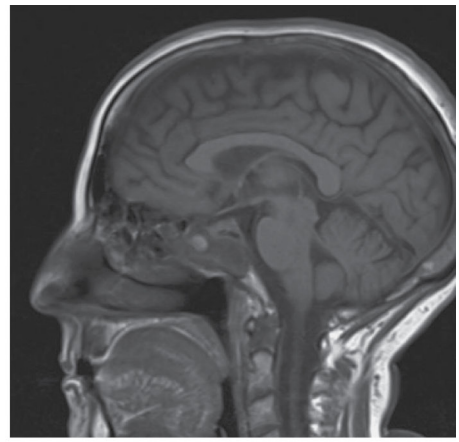
Figure 36.2 Axial (a) and sagittal (b) contrast-enhanced CT scan showing a sphenoid sinusitis. Bilateral soft tissue attenuation and a high attenuation material seen at the right sphenoidal sinus can suggest fungal infection (fungus ball).



Coronal T1-weighted MRI



Axial T1-weighted gadolinium contrast MRI



Sagittal T1-weighted MRI

Figure 36.3 T1 simple and gadolinium contrast MRIs with a heterogeneous right sphenoidal sinus opacification and a central enhancing mass in contrast and non-contrast images, with calcification, suggesting a fungal aetiology that is an indication for surgery.

Figure 36.4 Coronal CT scan of a left odontogenic maxillary sinusitis. The arrow points to an oroantral fistula.

Treatment varies depending on whether the aetiology is viral or bacterial.

Acute viral sinusitis treatment

As this is generally a self-limiting disease, treatment is primarily symptomatic. Adequate rest and hydration, steam inhalation, and room humidification are recommended, as well as use of over-the-counter medications, depending on the specific symptoms. Treatments should be tried for 5–10 days before reassessing the patient. Antibiotics are not recommended in patients with suspected acute viral sinusitis. Antihistamines have not been shown to be useful and can lead to impaired drainage and consequent worsening of symptoms and bacterial infection.

Symptomatic treatment can be divided into the following:

- *Analgesics and antipyretics* (paracetamol, ibuprofen, or paracetamol/codeine) are recommended for pain and/or fever. Codeine should only be used for acute pain that has not been successfully managed with other analgesics and for the shortest period of time possible. Contraindicated in children under 12 years, and not recommended in patients with obstructive sleep apnoea or severe lung disease.
- *Decongestants* (topical oxymetazoline is preferred over systemic pseudoephedrine because of less risk of adverse effects) are thought to restore sinus ostial patency with consequent symptomatic relief but still no strong evidence exists. It is important to inform the patient that topical agents should only be used for up to 3–5 days, to prevent rebound congestion.
- *Intranasal corticosteroids* are recommended in patients with congestion, with no systemic adverse effects. In allergic patients it may decrease the allergic response and swelling within the sinuses. One month of therapy is usually the minimum time recommended but treatment will depend on disease course. However, the clinical effect is small and has not shown an effect on quality of life.
- *Systemic corticosteroids* with or without antibiotics do not have a positive effect on recovery. A small but significant effect has been reported versus placebo on facial pain at days 4–7 after initiating treatment but the EPOS2020 steering group advises against the use of systemic corticosteroids in acute postviral rhinosinusitis.
- *Topical anticholinergics* (ipratropium) are recommended in adults with marked rhinorrhoea. But they have no effect on nasal congestion and have been associated with more side effects compared with placebo or no treatment. However, these side effects appear to be well tolerated and self-limiting.
- *Intranasal saline irrigations/sprays* are useful in relieving congestion and may reduce medication usage. They have an important role in relieving nasal symptoms. The following instructions for a home-prepared saline irrigation may be helpful for patients (University of Michigan Health System: saltwater washes (nasal saline lavage or irrigation) for sinusitis):

  - Add 240 mL (1 cup) of distilled water to a clean container. If using tap water, boil it first to sterilize it, and then let it cool down.
  - Add 2.5 g (half a teaspoon) of salt to the water.
  - Add 2.5 g (half a teaspoon) of baking soda.
  - This solution can be stored at room temperature for 3 days.
  - To use the home-made solution, fill a large medical syringe, squeeze bottle, or nasal cleansing pot (such as a neti pot) with the solution, insert the tip into the nostril, and squeeze gently.
  - Aim the stream of solution towards the back of the head.
  - The solution should go through the nose and out of the mouth or the other nostril.
  - Gently blow the nose after using the solution, unless instructed otherwise.
  - Repeat several times every day.
  - Clean the syringe or bottle after each use.
- *Mucolytics*: there is currently insufficient evidence to support their use.
- *Probiotics* have shown better results than placebo in reducing the episodes and mean duration of acute upper respiratory tract infection as well as antibiotic use and school absence. Thus they may be used as a preventive measure rather as a treatment.
- *Antibiotics*: there is no benefit in prescribing antibiotics for post-viral ARS. There is no effect on cure or duration of disease, and there are more adverse events.

Acute bacterial sinusitis treatment

It is important to evaluate the risk/benefit of antibiotic use: a high rate of spontaneous resolution without antibiotic treatment versus a shortened duration of symptoms with less need for follow-up but an increased risk of bacterial resistance, higher cost, and adverse effects.

Only a minority of ARS cases are from a bacterial origin. It should be suspected when symptoms are severe or start worsening, moreover if symptoms have lasted for more than 10 days.

ABRS is still defined by EPOS2020 by at least three of the following five symptoms/signs:

- Discoloured discharge.
- Severe local pain (often unilateral).
- Fever greater than 38°C.
- Raised C-reactive protein/ESR.
- 'Double' sickening.

In the case of an immunocompetent patient with non-severe symptoms, two approaches may be taken. Some guidelines recommend 'watchful waiting' for up to 10 days with only symptomatic therapy, leaving antibiotic therapy for deteriorations. The majority of non-severe cases will resolve on their own. Alternatively, immediate antibiotic therapy when started 72 hours after the onset of symptoms can shorten the duration of symptoms and the need for follow-up.

Ten to 14 days of amoxicillin or amoxicillin–clavulanate is first-line treatment for non-severe disease in immunocompetent people. High-dose amoxicillin–clavulanic acid is recommended as first-line therapy for patients who have severe disease or are immunocompromised. This is based on UK centres where the predominant organism isolated from complicated ABRS samples (61.3% of paranasal sinus pus samples, 83.3% of orbital complications, and 77.8% of intracranial complications) was *Streptococcus anginosus*. All isolates were sensitive to penicillin without any penicillin-resistant organisms. This suggests that penicillin can be used as an appropriate first-line empirical antibiotic for uncomplicated ABRS. Guidance from the UK's National Institute for Health and Care Excellence recommends phenoxymethylpenicillin as a first-line option unless patients have severe symptoms or no improvement of symptoms after 10 days, after which the recommendation is amoxicillin–clavulanic acid.

Other antibiotic options can be second- or third-generation cephalosporins alone or in combination with clindamycin for penicillin-allergic patients with known cross-sensitivity risks.

Fluoroquinolones should only be used when no other treatment options are available because of important adverse effects like tendinitis and tendon rupture, neuropathies, and other musculoskeletal or nervous system effects. Co-administration of a fluoroquinolone and a corticosteroid should be avoided. The UK and the US have issued a safety communication restricting the use of fluoroquinolones in acute sinusitis because of an increased risk of aortic dissection, significant hypoglycaemia, and mental health adverse effects in patients taking fluoroquinolones.

When no symptom improvement occurs after 3–5 days of antibiotic treatment, an alternative antibiotic should be considered. Symptomatic therapy is the same as used for acute viral sinusitis.

Specialist referral

Referral to an otolaryngologist is recommended when the patient is immunocompromised, has asthma, or has nasal polyps which can complicate management. Complications include periorbital oedema/erythema, displaced globe, ophthalmoplegia, reduced visual acuity, cranial nerve deficit, frontal swelling, or signs of meningitis/reduced consciousness. Recurrence of symptoms or failure to respond to antibiotic treatment also requires referral.

When acute invasive fungal rhinosinusitis is suspected, it must be referred immediately. It is a rare life-threatening condition with a high mortality rate. Surgical debridement, systemic antifungal therapy, and correction of predisposing conditions are needed promptly.

Complications of acute bacterial rhinosinusitis

Complications are uncommon but potentially life-threatening events. They can be grouped into orbital, endocranial, and osseous. In hospitalized patients with ABRS, complications vary from 3% to 20% with the highest number of admissions from January to March.

Orbital complications (~60–80%) include preseptal cellulitis, orbital cellulitis, and subperiosteal and intraorbital abscesses; rapid diagnosis and treatment (intravenous antibiotics and/or surgical drainage) is imperative to avoid long-term morbidity.

Endocranial complications (~15–20%) include epidural and subdural empyema, brain abscess, meningitis, encephalitis, and superior sagittal and cavernous sinus thromboses. Endocranial complications can present with non-specific signs and symptoms, requiring a high clinical suspicion.

Finally, osseous complications (~5%) result from osteomyelitis and may present as a subperiosteal frontal bone abscess (Pott's tumour) or a frontocutaneous fistula.

Chronic sinusitis is often considered as a complication but the microbial pathogens responsible for chronic sinusitis are different, so the empirical antibiotic of choice changes.

Orbital complications of acute bacterial rhinosinusitis

Orbital complications are the most common. Infection can spread directly through the lamina papyracea or through venous return. Orbital complications are more common in paediatric patients. Management is strongly encouraged to be multidisciplinary: otolaryngology, oculoplastic/ophthalmology, microbiology, and paediatric when appropriate. An abscess will require drainage if there is an inadequate response to antimicrobial therapy or worsening eye/orbital signs.

Chandler's classification still remains the most commonly used although some authors have discredited it for two reasons. Firstly, since the orbital septum is defined as the anterior limit of the orbit, a preseptal cellulitis is specifically an eyelid infection and it not only has a totally different clinical picture, management, and prognosis from all other orbital infections, but it is more associated with a complicated upper respiratory tract infection, dacryocystitis, or skin infection, while rhinosinusitis is an uncommon cause. Orbital involvement (postseptal cellulitis) presents with swelling, exophthalmos, and impaired, painful extraocular eye movements. Secondly, the final stage of the Chandler classification is cavernous sinus thrombosis, which has been suggested to be an intracranial complication rather than the end stage of orbital infection, associated more with sphenoid rather than orbital infection. In either case, consultation with an ophthalmologist should always be sought, both for clinical and for medicolegal reasons, and for objective assessment of the orbital pathology. Despite this, the classification remains widely used.

According to Chandler's classification, orbital complications may be divided into five stages based on their clinical and radiological findings:

Stage 1: preseptal cellulitis

Inflammation of the eyelid and conjunctiva anterior to the orbital septum, clinically diagnosed but if a CT/MRI scan is ordered a soft tissue swelling is present. Orbital pain, eyelid oedema, erythema, and fever with no proptosis or eye movement limitation. It normally responds to oral antibiotic therapy but if not timely and appropriately treated, it can spread beyond the orbital septum.

Stage 2: orbital cellulitis

Orbital cellulitis, subperiosteal abscess, and orbital abscess are all complications more commonly associated with ARS. The clinical scenario can overlap but orbital cellulitis classically presents with proptosis, some limitation of ocular movement (diplopia can present in extreme gaze), chemosis (conjunctival oedema), ocular pain, and tenderness. This clinical scenario requires immediate and aggressive treatment with intravenous antibiotics and a contrast-enhanced CT scan becomes mandatory in order to distinguish between cellulitis or abscess (intraorbital or subperiosteal). Orbital cellulitis generally resolves with antibiotics but can evolve to an abscess when suboptimal treatment is implemented, where there is antibiotic resistance or as part of disease evolution. An abscess generally requires surgical drainage, so close monitoring and a low threshold for surgical intervention when no response to antibiotic after 24–48 hours is extremely important.

If a concomitant intracranial complication is also suspected, MRI scan with contrast is better at diagnosing orbital complications.

All three complications (orbital cellulitis, subperiosteal abscess, and intraorbital abscess) cause globe proptosis and limit ocular movement.

The indications for surgical intervention are:

- evidence of subperiosteal or intraorbital abscess
- reduced visual acuity/reduced colour vision/affected afferent pupillary reflex
- no orbital or general condition improvement after 48 hours of intravenous antibiotics.

Stage 3: subperiosteal abscess

Located in the periorbita outside the ocular muscles (extraconal). It presents with the common symptoms of orbital cellulitis plus limitation of ocular motility and as a consequence of extraocular muscle paralysis (ophthalmoplegia). Visual acuity diminishes. High fever and raised leucocyte count in most cases. Contrast-enhanced CT scan shows oedema of the medial rectus muscle, lateralization of the periorbita, and lateral downward displacement of the globe.

Stage 4: orbital abscess

An orbital abscess is between the extraocular muscles and the eyeball (intraconal) and generally occurs as a result of diagnostic and optimal treatment delay, or an immunosuppressed status. Contrast-enhanced CT reveals a loss of definition in the extraocular musculature and optic nerve, and a confluent mass with peripheral enhancement. The diagnostic accuracy of clinical examination alone to determine an orbital abscess has been reported to be approximately 80% and rises to approximately 90% with the addition of CT, which emphasizes the importance of a thorough clinical examination.

Stage 5: cavernous sinus thrombosis

Considered by many authors as an endocranial complication, cavernous sinus thrombosis is rare (<10% of intracranial complications) and is caused by a retrograde spread of infection to the cavernous sinus through the venous system of the paranasal sinuses causing sepsis with multiple cranial nerve involvement.

Patients can present with orbital symptoms when involvement of the eye is present or in the classical clinical presentation with meningeal irritation, papilloedema with associated spiking fevers, and prostration. An MRI in the venous phase shows absence of venous flow in the affected cavernous sinus which is the 'gold standard', although high-resolution CT scanning with contrast can also show filling defects. Immediate endoscopic drainage of the affected sinus (almost always the sphenoid) is mandatory. Mortality in the past used to be as high as 30% in the adult population but recent articles report mortality rates of 0%.

Endocranial complications of acute bacterial rhinosinusitis

Endocranial (or intracranial) complications of ABRS are epidural or subdural empyema, brain abscess, meningitis, cerebritis, and superior sagittal and cavernous sinus thromboses. Usually associated with frontoethmoidal or sphenoid rhinosinusitis and haematological spread or by direct continuity to endocranial structures.

It is important to be aware that non-specific signs and symptoms are common in the initial stages, such as high fever, headache, and lethargy, or it may be clinically silent. Generally, specific central nervous system signs are present due to focal neurological damage or increased intracranial pressure, such as nausea and vomiting, neck stiffness, and an altered mental state.

To differentiate and diagnose these compilations it is necessary to perform an MRI, the 'gold-standard' investigation, or a contrast-enhanced CT scan at a minimum. Clinical suspicion should be high. Intracranial abscess may be relatively asymptomatic or cause subtle affective and behavioural changes (signs of increased intracranial pressure). Subdural empyema often presents with neurological symptoms as an emergency case while an epidural empyema is more often diagnosed on imaging studies.

Current treatment involves a multidisciplinary approach from an otolaryngology and neurosurgical team for both intracranial and paranasal drainage procedures, most often of the frontal sinus which is generally performed endoscopically. The lack of early sinus drainage has been associated with the need for repeated craniotomies. The prognosis of intracranial complications in ABRS depends on the severity of neurological signs and the delay in diagnosis and management. Mortality rates vary from 0% up to 19%.

Osseous complications of acute bacterial rhinosinusitis

Osteomyelitis and subperiosteal abscess constitute osseous complications that can eventually involve the brain and the nervous system. The most frequent intracranial route of spread is the frontal sinus and the most common osseous complications is osteomyelitis of the maxillary sinus, which is more common in the paediatric population. Frontal sinusitis can cause osteitis and osteomyelitis of the frontal sinus walls, which anteriorly may cause a subperiosteal (Pott's puffy) abscess or a sino-cutaneous fistula (rare) or posteriorly, through the diploic veins, to cause meningitis, epidural or subdural empyema, and brain abscess. Contrast-enhanced CT scanning or MRI is needed to confirm the diagnosis.

Prognosis

Most cases of uncomplicated acute bacterial sinusitis may be treated on an outpatient basis with excellent prognosis. Frontal or sphenoid sinusitis with air–fluid levels may require hospitalization with intravenous antibiotics. Patients that are immunocompromised or are septic require admission also. Invasive fungal sinusitis is associated with high morbidity and mortality.

Tips and pearls

- ARS is a common disease that is diagnosed clinically (based on history and physical examination), treated empirically, but with important complications that must always be suspected and positively excluded.
- It is always best managed by a multidisciplinary team that includes an otolaryngologist, ophthalmologist, neurosurgeon, microbiologist, haematologist, paediatrician when indicated, specialist nurses for monitoring, and pharmacists when complicated.
- Prevention may be helped by reducing triggers, such as quitting smoking or optimizing patient control of allergic rhinitis and/or asthma.
- The majority of cases are of viral aetiology, so indiscriminate use of antibiotics should be avoided. The condition is usually self-limiting so symptomatic therapy is usually the only treatment required.
- Imaging is not required for diagnosis unless complications are suspected.
- The outcome depends on the cause but regardless of treatment, recurrences are common and may lead to a poor quality of life.

FURTHER READING

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37

Endoscopic sinus surgery

Claire Hopkins

Introduction

Endoscopic approaches to the paranasal sinuses have become the standard of care in most cases of sinonasal disease since their introduction in the 1980s. Early work by Messerklinger was further refined and developed by Heinz Stammberger and others, and initially focused on re-establishing sinus drainage at the ostiomeatal complex, the 'functional' end point of the anterior sinuses, leading to the term 'functional endoscopic sinus surgery'—a paradigm shift to the inferior meatal, but non-functioning, antrostomies, and Caldwell–Luc approaches that had preceded. However, many extended applications, and some minimally invasive approaches, have followed, such that the term 'functional endoscopic sinus surgery' is perhaps best replaced with endoscopic sinus surgery (ESS) to encapsulate the variety of techniques. The basic concept remains that of correcting obstruction to sinus drainage, but surgery also aims to allow better access to topical treatments, particularly with patients with polyps/inflammatory disease.

Indications for endoscopic sinus surgery

The most common indication for ESS is chronic rhinosinusitis. The widely held dogma that 'sinus surgery should be considered when medical treatment has failed' leads to wide practice variation as both optimum medial therapy and adequate disease control have until recently been poorly defined.

Rice reported 'Indications for endoscopic sinus surgery' in a paper in the *Ear, Nose & Throat Journal* in 1994:

> If a patient has appropriate history, physical findings and CT [computed tomography], typically ESS will be indicated and beneficial. One should be wary of the patient who does not fit these criteria. A cause for concern is those patients whose pain is out of proportion to the disease found. This is doubly true if their pain does not respond to medical treatment. These people will usually not be helped by operative intervention.

Indeed, severe facial pain has since been shown to be a negative predictive factor for the presence of chronic rhinosinusitis, and alternative diagnoses such as migraine should be considered prior to surgery in such cases (see Chapter 31).

There have been few attempts to elaborate on indications for surgery in the setting of chronic rhinosinusitis since, but widespread usage and publication of patient outcomes have highlighted that as many as one in three patients fail to achieve clinically meaningful improvement in symptoms after ESS. Rudmik et al. therefore utilized a RAND consensus process, based on best available advice, and defined a minimum baseline, below which ESS should be performed only in carefully considered circumstances, as it would be less likely to be beneficial; ESS could be considered appropriate if there was confirmatory evidence of disease on CT, persistent symptoms measured by the 22-item Sino-Nasal Outcome Test (SNOT-22) to be greater than 20, and that a trial of appropriate medical therapy had been undertaken to include a trial of intranasal and oral corticosteroids for chronic rhinosinusitis with nasal polyps (CRSwNP), or a trial of intranasal corticosteroids and antibiotics for chronic rhinosinusitis without nasal polyps (CRSsNP). Not all patients above this threshold require surgery, but equally if medical therapy has been undertaken and failed to achieve adequate symptom improvement, repeated rounds of medical therapy and a delay in surgery may be detrimental to the final outcome.

Less frequently, ESS may be considered in recurrent acute rhinosinusitis (with at least one episode confirmed on endoscopy or imaging), mucocoeles, or in the presence of complications of acute infection, such as in orbital cellulitis. Both benign and malignant tumours may be approached endoscopically in selected cases. The sinuses may also be used as a corridor to approach the orbit, skull base, pituitary, and beyond.

The decision to proceed to surgery must be made in conjunction with the patient with an informed discussion regarding likely risks, benefits, and need for further treatment.

Preoperative preparations

All patients require a preoperative CT scan to confirm the presence and extent of disease and to plan the extent of surgery and approach required. There still remain indications for an external approach, such as very laterally based frontal sinus pathology.

The CT scan must be carefully studied immediately prior to surgery and be available throughout. Important features to note to minimize the risk of complications may be easily summarized by the

Table 37.1 Checklist for preoperative computed tomography analysis

C	Cribriform niche—depth and asymmetry should be evaluated
L	Lamina papyracea—examined for dehiscence
O	(Onodi) Sphenoethmoidal cells
S	Sphenoid sinus—examined for dehiscence of bone overlying the optic nerve and carotid artery Skull base—examined for dehiscence, and asymmetry in height
E	Ethmoidal arteries—the position of the anterior and posterior ethmoid artery relative to the skull base should be assessed. The anterior artery is more likely to be exposed in a mesentery in the presence of a supraorbital ethmoid cell

mnemonic 'CLOSE'—see Table 37.1. Anatomical variants are commonplace, and extra care is required in revision cases or in extensive disease where landmarks such as the middle turbinate may have been resected or distorted. The complexity of the anatomy, particularly in the frontal sinus, may dictate the need for onward referral in some cases. Lateralization of the uncinate should alert the surgeon to use a retrograde approach to prevent orbital penetration.

There is evidence from clinical trials that preoperative oral steroids may achieve statistically significant reductions in intraoperative blood loss although it does not appear to be clinically relevant and should not be used routinely. There may be benefit in selected cases of extensive nasal polyps, but there are alternatives to optimize the surgical field with fewer side effects. There is no evidence that preoperative antibiotics are routinely helpful.

Patients on low-dose aspirin may be at a higher risk of thromboembolic events than the risk of bleeding, and surgery can be performed on such patients without discontinuing. However, all patients on high-dose aspirin or other anticoagulants should normally be withheld with bridging therapy if required.

Consent

An informed consent procedure should cover alternatives to surgery, surgeon-specific risks of complications, and any relevant features that may increase risks in the individual (see 'Complications'), likely benefits (see 'Outcomes'), the need for further treatment or revision rates, as well as risks of surgery in general. The death of Elaine Bromiley in 2005 during a 'routine' sinus operation due to failed intubation serves as an important reminder that all procedures carry risks.

Perioperative interventions

A skilled anaesthetist is likely to be the greatest determinant of the surgical field. Mild hypotension in the absence of rebound tachycardia can be achieved with a variety of agents but there is some evidence to suggest that total intravenous anaesthesia may be more effective in this regard. Intubation and use of a throat pack may often be avoided by the use of a laryngeal mask. There is increasing use of local anaesthesia, with or without sedation, to facilitate 'office/clinic-based' procedures and enhanced recovery.

Topical agents may be placed in the nose to achieve vasoconstriction, either using an aerosol device or delivered on neuropatties or ribbon gauze. 'Moffett's' solution was once widely used, but has a risk of cardiotoxicity and so alternatives such as decongestants have become more popular, particularly if there are known cardiac risk factors. Our current recipe for a modified Moffett's solution contains 100 mg cocaine, 1 mL of 1 in 1000 adrenaline, 3 mL of 8.4% sodium bicarbonate, and 5 mL of normal saline, delivered as an aerosolized solution.

Tranexamic acid may be given intravenously on induction and has been shown to achieve clinically relevant reductions in intraoperative blood loss. Hot saline (49°C) irrigation may also be beneficial in enhancing the surgical field.

Surgical technique

Basic sinus surgery typically involves removal of the uncinate process, exposing or enlarging the natural ostium of the maxillary sinus, and removing at least some of the partitions of the ethmoid sinuses (Figure 37.1). A wide variety of through-cutting, grasping instruments are available. Balloon dilatation was introduced to address frontal, maxillary, and sphenoid disease but cannot address ethmoidal pathology, and is probably best reserved for limited CRSsNP.

Removal of frontoethmoidal cells is challenging, requiring experience and specialist instruments. Powered microdebriders may enhance surgical efficiency and maintain a clear visual field but will

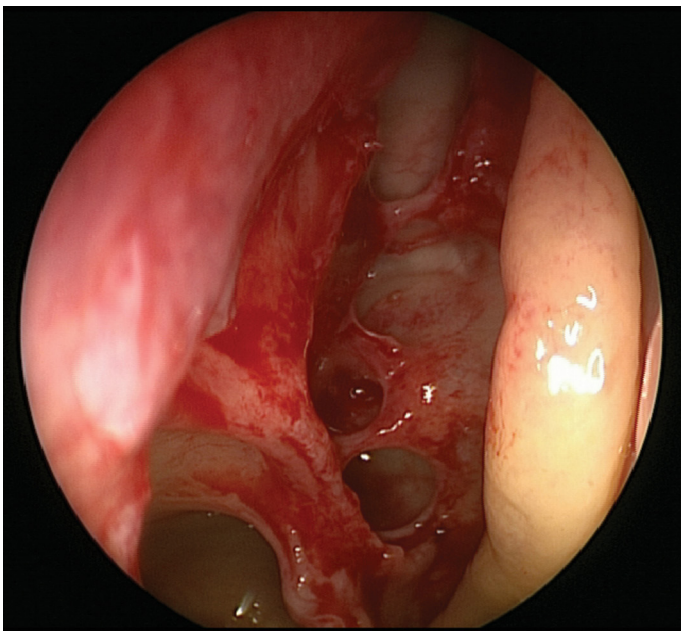

Figure 37.1 Basic ESS. Right nasal cavity demonstrating complete removal of the uncinate process, exposure, and enlargement of the maxillary sinus ostium. The anterior ethmoid cells have been opened widely, revealing the anterior ethmoid artery running in a small mesentery below the skull base. The frontal recess has been exposed by 'uncapping the egg', resecting the agger nasi cell. The basal lamella has been partially opened to allow access to the posterior ethmoid cells. Care has been taken to avoid trauma to the middle turbinate in order to reduce the risk of adhesions.

likely increase the severity of complications arising as a result of orbital or intracranial penetration.

The extent of surgery is usually tailored both to the extent of disease on radiological imaging and the underlying pathology. Where topical access to medical treatments to prevent recurrence of inflammatory mucosal disease, in type 2 chronic rhinosinusitis/CRSwNP, is important in reducing risk of recurrence, more extensive surgery may be undertaken, including larger antrostomies, complete removal of all ethmoidal partitions, and extended approaches to the frontal and sphenoid sinuses. While preservation of mucosa is considered a basic tenet of sinus surgery in CRSsNP, there are now some advocates for removal of diseased mucosa in CRSwNP, or 'rebooting' the sinuses.

There remains huge variation in both surgical practice and the terminology applied to it. The choice of surgical approach, instrumentation, and extent of surgery is not supported by any significant evidence base, and surgical trials are required to better address this topic. The current evidence base supports the use of more extensive or 'complete' surgery in CRSwNP.

However, the most common findings in revision cases are a residual uncinate process (Figure 37.2), creation of an accessory sinus ostium, and residual anterior ethmoid cells, suggesting that if greater care is applied to undertaking and performing the more basic steps of ESS alone, the need for further surgery may be reduced (Figure 37.3).

A septoplasty may be required for access to the sinuses during the procedure, and one recent study suggested it may improve symptomatic benefit in patients undergoing ESS who are found to have a deviated septum. Turbinate hypertrophy may also be addressed concurrently.

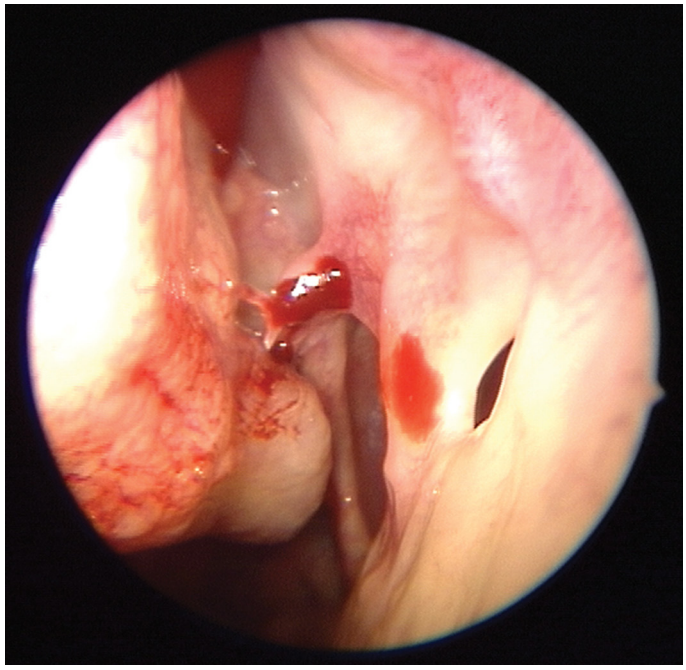

Figure 37.2 'Missed ostium': when viewed with a 30° scope it is possible to identify the natural os, partially concealed by the incomplete uncinectomy, in front of the large antrostomy created in the posterior fontanelle. This is a common finding in revision cases and may allow recirculation of mucous and persistent symptoms.

Image guidance

Image guidance may act as a useful adjunct to a thorough knowledge of sinus anatomy and the careful study of multiplane imaging, and may be particularly useful in training and in the presence of complex anatomical variants or in revision cases. The ability of image guidance to reduce the risk of complications remains controversial—enhanced confidence may lead the surgeon to take risks beyond their surgical capability and over-reliance may lead to an increased risk of error if the limitations of image guidance and accuracy are not fully appreciated; for example, with surface mapping accuracy may deteriorate deeper into the nasal cavity. Augmented reality now facilitates preoperative surgical planning and may even help direct dissection and warn of the proximity of high-risk structures. As image guidance becomes more widely available, there is a risk that undertaking surgery without such equipment becomes considered unacceptable, despite the lack of evidence to support improved outcomes or lowered risks.

Packing and stents

Routine postoperative packing with removable dressings and overnight stay were considered standard practice when ESS first became widely used. However, the majority of patients are now managed on an ambulatory basis, facilitated by changes in anaesthetic agents and techniques and recognition that packing is not essential in all cases; avoidance will also reduce early postoperative discomfort. There are a large number of absorbable dressings that may be used as alternatives to removeable packs, and may act as temporary spacers to reduce the risk of lateralization of the middle turbinate, to directly promote haemostasis or to act as a reservoir for topically applied medications.

A recent development has including the use of steroid-eluting stents. There is some evidence that these may be beneficial in selected cases when placed postoperatively, and different types are being produced that may also be placed in the middle meatus in office/clinic as an alternative to surgery. They are currently expensive, and cost-effectiveness must be established before routine use. Steroid-soaked absorbable dressings may be used as an alternative, although this is likely to achieve only short-term steroid delivery.

Complications

Complications may be categorized according to their severity and the site of injury (Table 37.2). Minor complications are reported in approximately 5% of cases but major complications are fortunately rare, occurring in less than 1 in 1000 cases.

Exposure of orbital fat, as a result of a breach of the lamina papyracea, may be managed conservatively; the surgery can be completed if safe to do so, and the patient must be instructed not to blow their nose to avoid surgical emphysema. Orbital haematoma, which most commonly results from retraction of an injured anterior ethmoid artery, needs urgent decompression, and all surgeons undertaking ESS must be trained to be able to undertake a lateral canthotomy and cantholysis if required (Figure 37.4). Injury to the

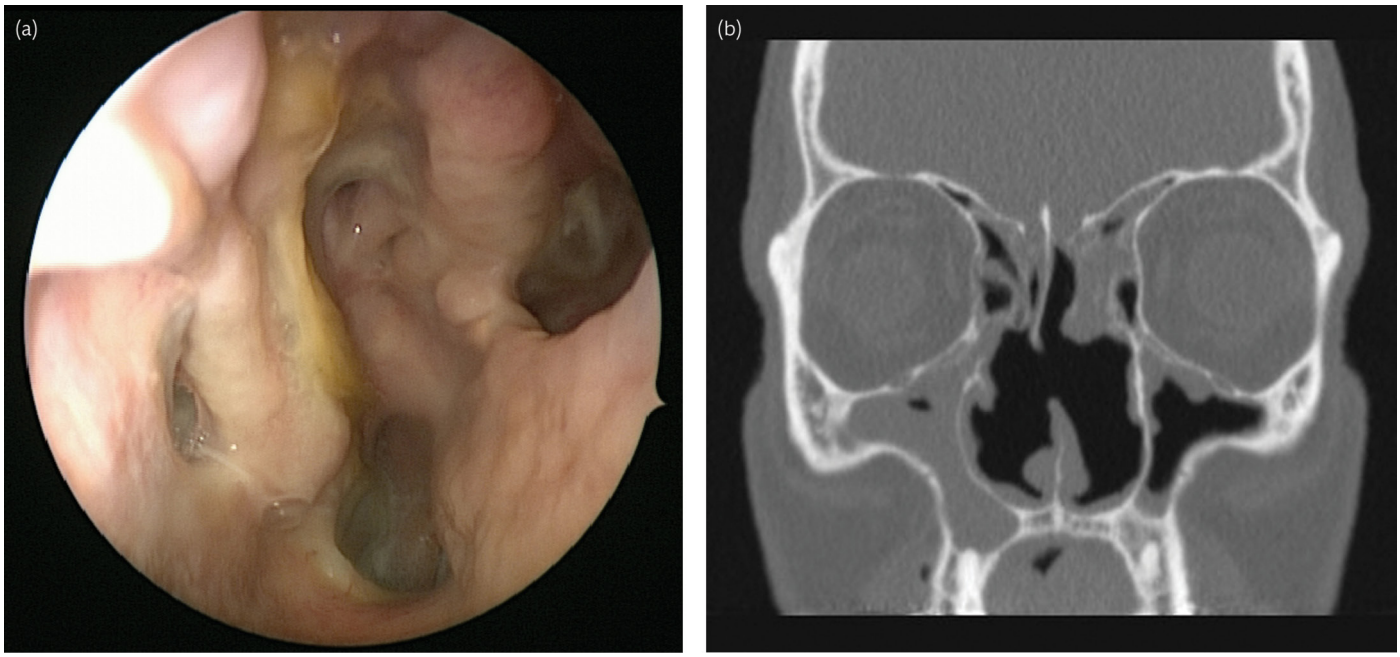

Figure 37.3 'Failed surgery': this patient with recalcitrant chronic rhinosinusitis has undergone multiple procedures which have left him with a large septal perforation, complete removal of all turbinate tissue, and inferior meatal antrostomies, yet have failed to identify the natural ostium, and there is a residual uncinate and incomplete dissection of the anterior ethmoid cells. Careful execution of the basic steps of ESS may have avoided this poor outcome.

globe or optic nerve is irretrievable, while medial rectus trauma is likely to lead to diplopia, but urgent ophthalmological input may allow repair.

Mild postoperative bleeding is commonplace, and can usually be managed conservatively, although patients may occasionally require readmission and packing. Major bleeding may require ligation of the bleeding vessel, or in the case of internal carotid artery injury, interventional radiology for embolization.

Any evidence of cerebrospinal fluid leakage at the time of surgery should prompt a repair at the same sitting. If there is uncertainly, a postoperative CT scan may demonstrate a breach in the skull base and pneumocephalus. Fluorescein cannot be used at the time of an unexpected cerebrospinal fluid leak as it requires consent for off-licence use, but it can be helpful if a cerebrospinal fluid leak is confirmed postoperatively by testing fluid for TAU/beta-2-transferrin protein, but the site cannot be identified.

Adhesions may not cause symptoms, so can often be managed conservatively. While many patients have surgery with the hope of improving their sense of smell, in some cases it may deteriorate, caused by scarring of the olfactory cleft or due to topical medications applied during surgery.

In addition, complications common to all procedures may occur, and standard protocols should be followed for the prevention of thromboembolic disease and other complications of surgery and anaesthesia.

Table 37.2 Complications of endoscopic sinus surgery

	Minor	Major
Orbital	Exposure of orbital fat Orbital emphysema Bruising	Orbital haematoma or optic nerve injury leading to visual loss Medial rectus injury or enophthalmos leading to diplopia Nasolacrimal duct injury leading to epiphora
Bleeding	Bleeding controlled with conservative measures, packing	Bleeding requiring return to theatre, ligation of bleeding vessel, or transfusion Carotid artery injury
Intracranial complications		Cerebrospinal fluid leak Meningitis and other intracranial infections Tension pneumocephalus Injury to brain parenchyma
Other	Adhesions Readmission Acute infection	Loss of sense of smell

Postoperative medication

Many patients report postoperative congestion as more bothersome than pain itself, and generally the need for opioid-based medications can be avoided with regular use of simple analgesia such as paracetamol and non-steroidal anti-inflammatory drugs. Patients with aspirin-exacerbated respiratory disease, who may undergo repeated ESS for recurrent nasal polyps, can usually tolerate celecoxib, and a test dose may be given in hospital to help manage their postoperative pain control.

Sinus irrigation may help reduce the need for postoperative debridement and can usually be safely commenced on the first postoperative day for routine sinus procedures. The need for regular postoperative debridement remains controversial; it is routinely used in North America, but rarely performed in the UK, with little difference in reported outcomes. It is more important after extended procedures such as a Draf III frontal drill-out.

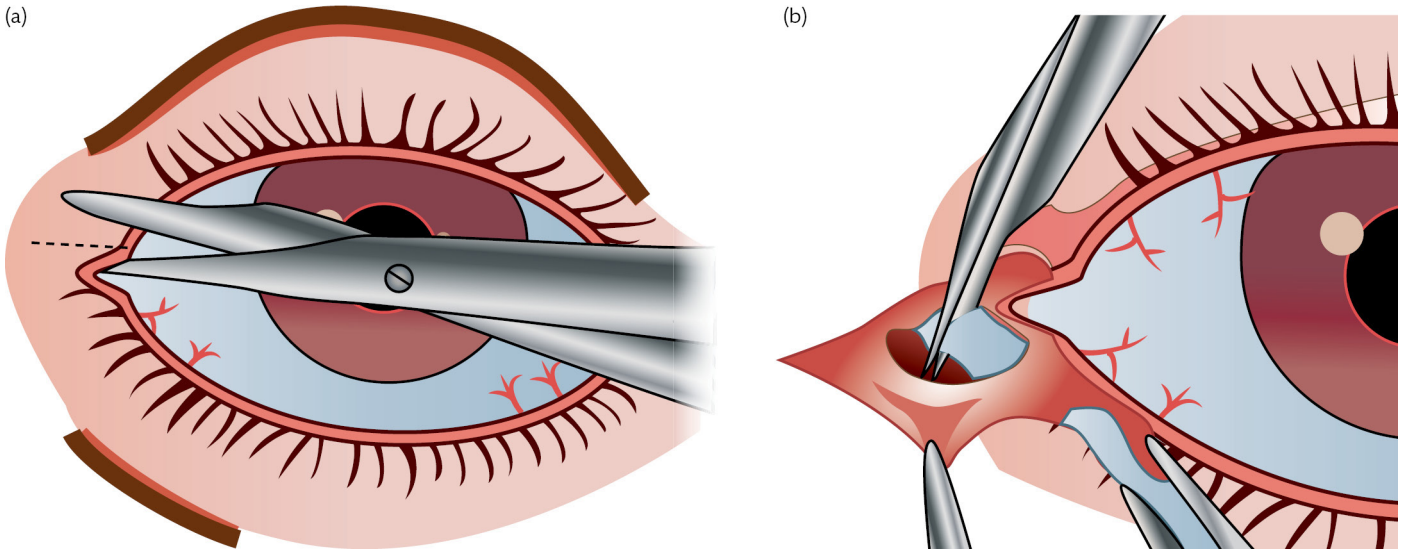

Figure 37.4 (a) A lateral canthotomy and cantholysis is performed to relieve intraorbital pressure in the setting of orbital compartment syndrome. Significant proptosis, occurring during or after ESS, should prompt urgent intervention without delay for measurements of intraocular pressure, although an assessment of vision and pupillary reflexes should be performed where possible. In an awake patient, local anaesthetic with adrenaline is first injected into the incision site, and a haemostat can be used to then crush the tissue to reduce bleeding. Iris scissors are used to cut from the lateral canthus to the orbital rim for the lateral canthotomy. (b) For the cantholysis, the inferior limb of the lateral canthal tendon can be identified by retracting the lower lid (it can often be felt by 'strumming' with the scissors) and is then cut. The incision is not sutured, and the eye should be protected with lubricant. Ophthalmological assessment should be performed as quickly as possible to confirm that intraocular pressure has been adequately controlled, but if there is any ongoing concern, an endoscopic medial orbital decompression may be performed.

Topical steroids may promote postoperative healing and form the mainstay in ongoing medical care, and are particularly important in patients with type 2 chronic rhinosinusitis/CRSwNP. They may usually be started immediately postoperatively. Sinus delivery may be achieved in patients with open sinus cavities by adding topical steroids to sinus rinse solutions and irrigating into the nose; both Pulmicort Respules or Flixonase Nasules may be used. Ongoing use of intranasal steroids has been shown to reduce risk of recurrence, and steroid irrigation has been further shown to achieve better endoscopic control of disease than corticosteroid sprays.

There is little evidence that routine use of postoperative antibiotics helps to reduce the relatively high rate of postoperative infections, and these should be treated as needed rather than using antibiotics prophylactically.

Recovery

ESS can usually be performed on an ambulatory basis, and most patients will be sufficiently recovered to allow same-day discharge. A 3–4-hour period of observation is recommended to allow bleeding to settle. All patients must be told how to access postoperative care in the case of visual change, severe headache, neurological symptoms, or heavy bleeding, prior to discharge. Most patients will require at least a week of postoperative recovery and on average return to full activity 10–14 days after surgery. They may not feel the full benefit from surgery until 4–6 weeks after.

Outcomes of endoscopic sinus surgery

Patient-reported outcome measures are usually considered the most important outcome in surgery performed with the aim of improving symptomatic control and the impact of disease on quality of life. The SNOT-22 is the most widely used, with a range in scores from 0 to 110. The UK audit of sinonasal surgery was one of the first large, multicentred, prospective cohort studies to use patient-reported outcome measures to measure the effectiveness of surgery. This study reported a statistically significant, large improvement in disease-specific quality of life, maintained over a 5-year period. A recent publication has shown this improvement to be durable for more than 10 years in patients available for long-term follow-up. A systematic review of outcomes in 40 unique cohorts has shown a mean improvement of 24.4 points in the SNOT-22.

To better understand a 'successful' outcome at an individual level, the concept of a 'minimal clinically important difference' is usually applied, that is, the smallest change in patient-rated outcome scores that a patient can perceive as a real change; for the SNOT-22 it is 8.9 points. Applying this to cohort series suggests that only 65–75% of patients achieve a successful outcome from surgery. Baseline preoperative SNOT-22 score is the best predictor of achieving improvement in health-related quality of life greater than the minimal clinically important difference, and therefore case selection is key to improving outcomes. Generally, patients achieve a 50% improvement from their baseline score; patients with more severe symptoms will achieve a greater absolute reduction in symptom scores, but may still have higher postoperative scores than the normal population. This is important when managing patient expectations in preoperative discussions.

Revision surgery rates

Revision surgery should not necessarily be considered a treatment failure in the setting of chronic sinus disease. However, it is important that patients are made aware of the chance of subsequent

revision surgery. Prospective cohort studies report revision surgery rates of 11% at 3 years, 19% at 5 years, and 17% at 10 years. Patients with CRSwNP report higher rates of revision surgery (11%, 21%, and 25% at 3, 5, and 10 years, respectively) than CRSsNP (10%, 15%, and % at 3, 5, and 10 years, respectively). A population-based study using electronic healthcare records found long-term revision rates of 15.9% with a mean follow-up of 9.7 years, with female sex, older age at first surgery, presence of nasal polyps, comorbid asthma and allergy, and a family history of chronic rhinosinusitis all being associated with a higher risk of requiring revision. Revision rates in CRSwNP were 29.9% compared with 9.8% in CRSsNP.

Tips and pearls

- Routine use of patient-reported outcome measures in clinical practice will help improve outcomes through regular self-audit.
- Lateralization of the uncinate should prompt a retrograde approach to uncinectomy in order to avoid orbital penetration.
- If orbital fat is seen during the procedure, do not resect or try to replace it, but cover with some Surgicel, ribbon gauze, or equivalent to act as a marker and complete the surgery if it is safe to do so. The patient must be told not to blow their nose for 2 weeks to avoid surgical emphysema.
- Changing to a 30° scope after performing the uncinectomy may help identify the natural maxillary sinus ostium, which sits at a 45° angle to the vertical plane, if it is not immediately visualized, and may help reduce the risk of a missed ostium.
- Frontal sinus anatomy is highly variable. Understanding the anatomy is essential for safe dissection, but also allows surgeons to better plan which cases fall within their area of expertise and those that may benefit from onward referral. In such cases, use of balloon dilatation is no substitute for anatomical knowledge and surgical ability.
- A wide range of instruments are available to facilitate safe and careful dissection. In particular, frontal sinus surgery requires specialist instrumentation and curved suction should not be used as a 'dissecting tool' in the frontal recess.
- The floor of the orbit, visualized through an antrostomy, and the skull base identified in the sphenoid are invaluable fixed anatomical landmarks that may be of use in revision cases where the middle turbinate has been resected and the ethmoid anatomy has been distorted. Image guidance may be helpful in these cases but should not be relied on completely, and accuracy regularly checked by returning to these fixed and visible landmarks.
- In the setting of inflammatory disease, it is always wiser to stop the surgery if bleeding and poor field prevent clear visualization rather than risk complications. Applying vasoconstriction topically and using warm water rinses may help improve the field during the procedure.
- Surgery is unlikely to achieve durable benefits in disease-specific quality of life in inflammatory sinus disease without ongoing postoperative topical treatments. It is essential to stress to patients both before and after surgery that the choice is between ongoing medical treatment or surgery *and* ongoing medical treatment.

Controversies

Given the paucity of randomized trials to inform decision-making in ESS, almost all aspects may be considered controversial and could generate lengthy debate.

In particular, the following topics merit further evaluation:

- The optimum timing of surgery, with respect to whether early intervention achieves better long-term outcomes or risks overtreatment of patients who may have improved with medical treatment alone. This is particularly relevant with the promotion of minimally invasive office/clinic techniques, such as balloon dilatation, in this context.
- Moving to an office/clinic setting and performing surgery in an awake patient is becoming increasingly popular but it remains unclear whether the drive is supported by improved patient outcomes or enhanced remuneration. Certainly, studies are required to compare both early symptomatic outcomes but also longer-term revision rates.
- The optimum extent of surgery in patients with inflammatory sinus disease, particularly those with type 2 disease, has yet to be defined. In particular, there is a need to study the effectiveness of extended approaches including Draf III frontal sinus procedures and 'rebooting' and whether these should be performed at the primary procedure, or only in cases of multiple revisions.
- The relative benefits of surgery versus biological treatments in these type 2 patients will need both clinical and cost-effectiveness studies to determine at what point the patient pathway, and in which patients, biologics should be introduced.
- As image guidance usage increases, there is a concern that surgeons trained only where this is available become overly dependent upon the equipment, and that if complications occur in a case where image guidance was not used, this may become a focus of medicolegal complaint.
- Use of robotic surgery is currently limited due to the size of the instruments, and therefore its potential in the nose and sinuses has not yet been fully explored. Other novel innovations, such as three-dimensional endoscopy, have yet to have the impact on the field achieved by the initial pioneering work of Messrs Messerklinger, Hopkins, Draf, and Stammberger, to whom we remain indebted.

FURTHER READING

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38

Chronic rhinosinusitis

Richard J. Harvey and Janet Rimmer

Introduction

The proposed aetiological and disease-modifying factors related to chronic rhinosinusitis (CRS) have expanded over the last decade resulting in a plethora of potential treatment strategies. Potentially, advancement of knowledge in this field has developed more than any other in otolaryngological science. However, most treatment concepts lag behind in general practice.

Trainees, students, and residents may find the heterogeneous group of pathophysiological mechanisms difficult to apply in clinical decision-making. However, identifying the inflammatory, microbial, and/or mucociliary factors for a specific patient allows tailoring of treatment critical to achieve disease remission. A logical strategic process, rather than using one broad, ill-defined approach for every CRS patient, is a more successful approach.

Contemporary research in CRS points to a central failure of mechanical and innate immunity with a subsequent proinflammatory immune response and although simplistic in description, it is the key issue that needs to be resolved in most patients (Figure 38.1).

Three main driving forces exist in CRS. There is an interacting triad of intrinsic mucosal inflammation, local microbial dysbiosis, and mucociliary dysfunction. However, each individual patient will have a greater contribution from one of these factors dominating the process while the other factors are secondary disease-modifying events.

Mucosal inflammation, the microbial community, and mucociliary dysfunction are interdependent, with positive feedback mechanisms between them (Figure 38.1). For many patients, all three of these factors rapidly become involved in the disease process, regardless of the driving factor that initiated the CRS state. Defining

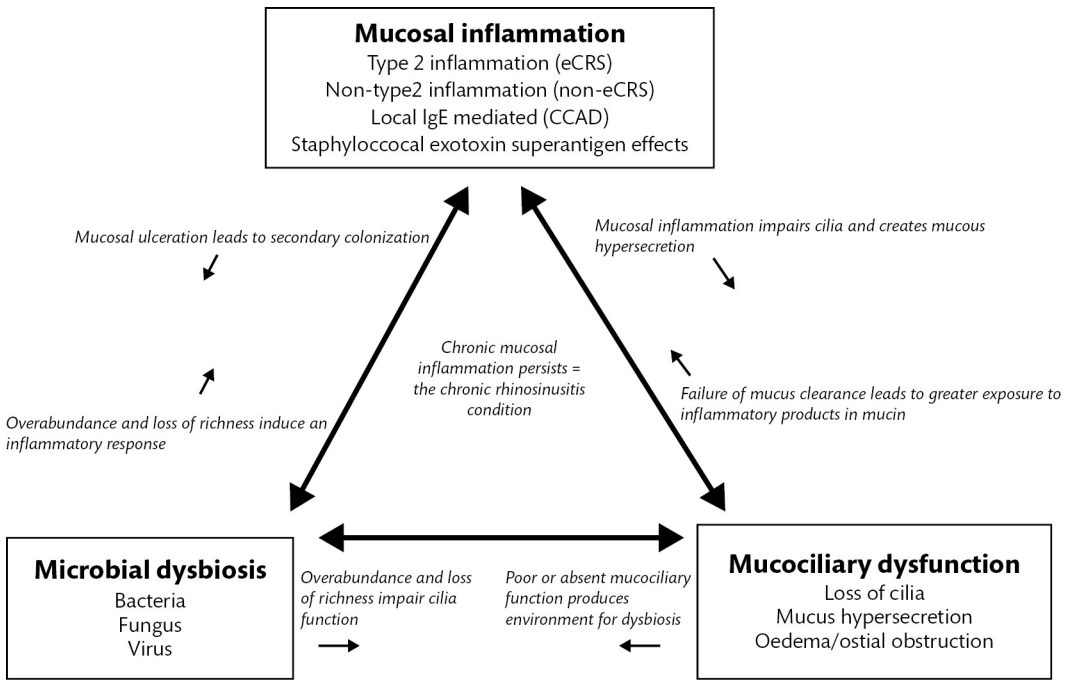

Figure 38.1 The interplay of pathophysiological factors that lead to chronic inflammation in the paranasal sinuses. CCAD, central compartment atopic sinus disease; eCRS, eosinophilic chronic rhinosinusitis.

the predominant driving factor, however, is critical in treatment strategy.

Currently, those patients with discrete mucociliary dysfunction or simple untreated infection (primary localized non-type 2 CRS) appear to have the best prognosis. In contrast, those with intrinsic mucosal inflammation (eosinophilic CRS or primary diffuse type 2 CRS) require greater care as they present a broader inflammatory airway disorder.

In treating CRS, all three factors should be considered. The relative weight of importance of the driving factor should be targeted both before and after surgery.

Basic science: anatomy and physiology

The paranasal sinuses represent some of the most variable human anatomy. Knowledge of the developmental and functional anatomy is critical to understand patterns of disease and avoiding injury to critical structures during surgery. Other chapters cover specific anatomy in more detail but the relevant anatomical factors to CRS are discussed below.

Functionally, the paranasal sinuses are divided into anterior, posterior, and sphenoid functional units These functional units are based on mucociliary or 'drainage' pathways. The basis of these three functional units is important, as those patients with CRS changes restricted to one functional unit, or perhaps two adjacent functional units, are defined as having 'localized' CRS and usually have a local anatomical basis for the disease process. In contrast, those patients with multiple units, generally bilateral disease, and changes that do not follow the known 'drainage pathways' have 'diffuse' CRS and will often reflect an underlying inflammatory disorder rather than a simple structural issue.

Anterior functional unit

Comprised of the maxillary, anterior ethmoid, and frontal sinuses which drain into the nose through the ostiomeatal complex. Disease can be limited to just these three sinuses as they share this common outflow in the ostiomeatal unit (Figure 38.2). CRS that limits itself to these functional units is referred to as 'primary localized CRS'.

Maxillary sinus

The maxillary sinus is the largest sinus and drains via an ostium that lies in the superior and anterior part of the lateral nasal wall. This ostium opens into a small cleft referred to as the infundibulum and which cannot be seen on endoscopy as it is covered by the uncinate bone medially. The anterior ethmoid and frontal sinuses drain into this area and the region is referred to as the ostiomeatal complex.

Ethmoid sinus

The ethmoid 'bulla' refers to the largest and most consistent anterior ethmoid air cell. It attaches to the lamina papyracea laterally and has variable attachments to the skull base and basal lamella creating a series of clefts and spaces that are well described. An infraorbital anterior ethmoid cell can pneumatize into the maxillary sinus as a normal variant called a Haller cell.

The complex shape of the middle turbinate is divided into three segments according to the sagittal, coronal, and axial planes. The sagittal segment attaches to the skull base at the lateral lamella. The coronal segment creates the basal lamella which separates anterior and posterior ethmoid cavities. Although often referred to as a single 'ethmoid' unit, the anterior and posterior ethmoid cavities have different drainage pathways and embryological origins. The axial segment attaches to the lateral nasal wall at the sphenopalatine artery terminal branch point.

Frontal sinus

The frontal recess is not a simple door opening but a transitional space between the superior aspect of the ethmoid and the true frontal sinus. There are structures that may encroach this space from anterior, posterior, and medial and lateral directions.

- *Anterior*—agger nasi, posterosuperior uncinate process, and frontal ethmoid cells:
  - Agger nasi refers to the anterior-most ethmoid cell and is bound medially by the uncinate process. The degree of agger pneumatization influences the superior uncinate position and nasofrontal beak thickness and varies greatly between patients. The uncinate process can insert into the medial orbital wall, skull base, or middle turbinate and has multiple attachments in more than 50% of cases.

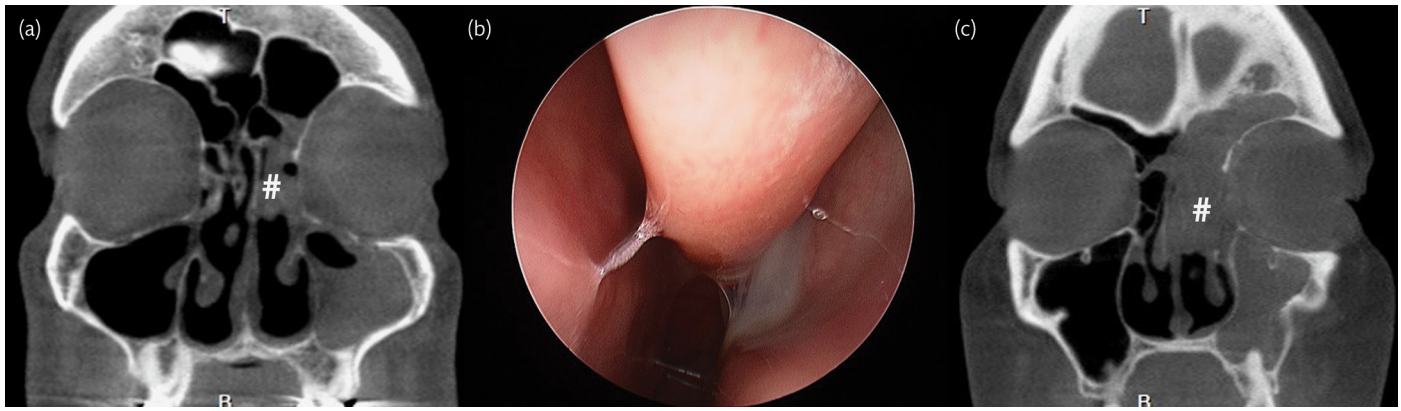

Figure 38.2 Primary localized CRS of the left anterior functional unit sinuses (a, c) (anterior ethmoid, maxillary, and frontal) which all drain mucous from a common area: the ostiomeatal complex (#). On endoscopic view, purulent secretions can be seen coming from the middle meatus (b).

- Uncinate inserts onto the medial orbital wall in 85% of cases resulting in a frontal recess drainage pathway medial to the uncinate. An uncinate with an isolated attachment to skull base or middle turbinate occurs in 15% of cases leading to drainage lateral to the uncinate.
- Frontal cells pneumatize above the agger towards the frontal sinus. According to contemporary classifications, a type 1 frontal cell is a single frontal ethmoidal cell above the agger and below the frontal sinus floor; type 2 is a tier of cells above the agger; type 3 fills less than 50%; and and type 4 fills greater than 50% of the frontal sinus. Such classifications are defined to ensure that a surgeon correctly enters the frontal sinus.

- *Posterior*—supraorbital ethmoid and suprabulla cells:
  - Supraorbital ethmoid cells are anterior ethmoid cells extending superiorly and laterally over the orbital roof. They are attached to the basal lamella rather than the frontal process of the maxilla (cf. agger nasi). Suprabulla cells are similar pneumatized extensions above the ethmoid bulla along the skull base and frontal sinus posterior table, but do not contact with the orbital wall or roof.
- *Medial and lateral*: medially, these include frontal intersinus septal cells and a medially inserting uncinate. Intersinus septal cells represent pneumatization of the frontal sinus septum. Lateral encroaching structures include frontal cells, agger nasi, and a lateral uncinate attachment.

Posterior functional unit

Comprised of the posterior ethmoid cells which drain into the superior meatus. An Onodi cell is a posterior ethmoid cell that pneumatizes laterally and posteriorly over the optic nerve and potentially exposes it to injury during surgery (Figure 38.3).

Sphenoid functional unit

The sphenoid sinus drains via a small ostium into the sphenoethmoidal recess between the superior meatus and septum. The supreme turbinate may be seen here. The sphenoid ostium opens 'behind' the superior turbinate as it is a separate embryological bone from the ethmoid. Structures associated with the sphenoid sinus include the optic nerve, internal carotid artery, and sella turcica. Primary localized CRS can occur here as it has its own discrete drainage pathway (Figure 38.4). Isolated sphenoid sinus changes often require surgical intervention as non-inflammatory conditions, such as neoplasia, fungal ball, and encephalocoeles, are common.

Assessment

Symptoms

The presenting symptoms of CRS are diverse and overlap with rhinitis and cephalgia disorders (see Chapter 31). However, four cardinal features are often present: nasal congestion, nasal discharge (anterior), facial pressure, and loss/distortion of smell. These are the 'local' symptoms of chronic sinonasal disease. Regional symptoms can include Eustachian tube dysfunction, cough, postnasal drip, and dysphonia. Although cough, throat, and lung symptoms can be a result of direct irritation from secretions, they are more likely to represent broader involvement of the airway in the associated disease process. When it comes to primary diffuse disease, symptoms in only one compartment of airway symptoms (upper, lower, middle) are the exception rather than the rule. Systemic features include malaise and minor depression, but fever is uncommon.

The local features are important and current diagnostic criteria need to include the presence of nasal congestion or discharge with

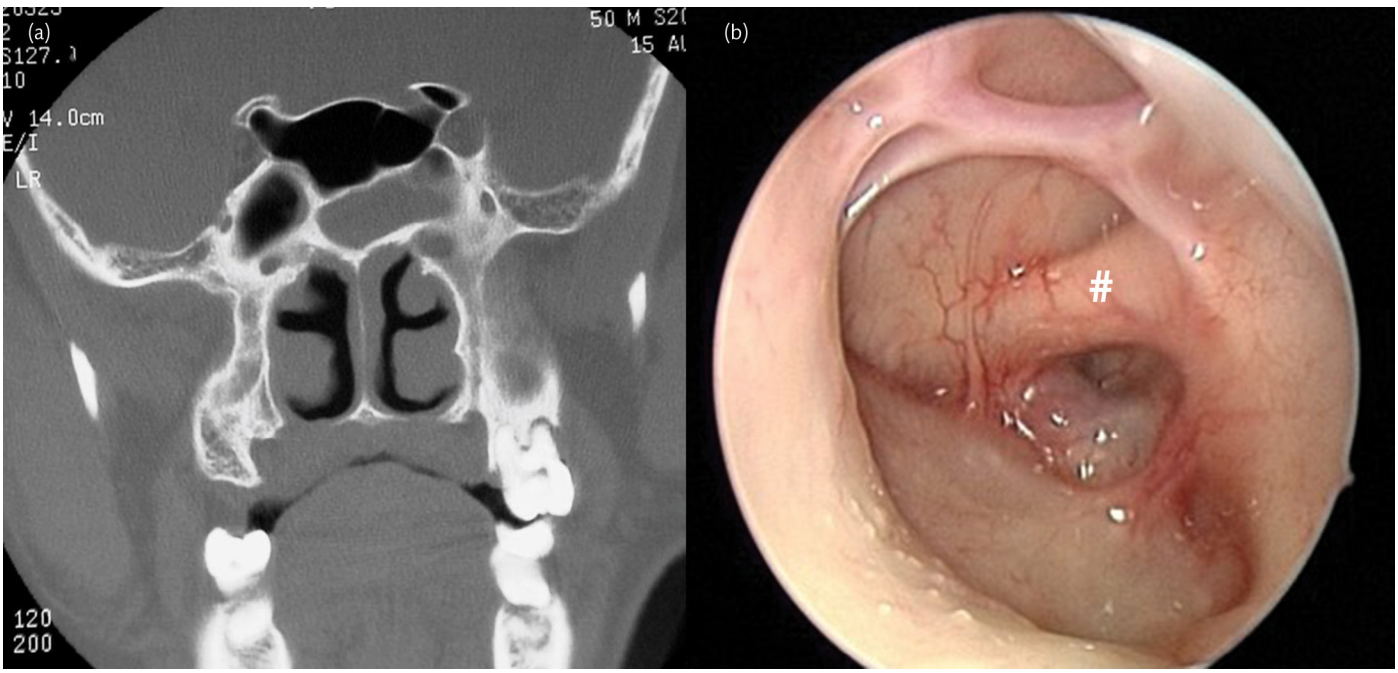

Figure 38.3 The left sphenoid cell lies below a sphenoethmoidal (or Onodi) cell on computed tomography scanning (a) and displaces the optic nerve (#) into the 'ethmoid' on endoscopic view (a). The Onodi cell is one of the many anatomical variants of the paranasal sinus system.

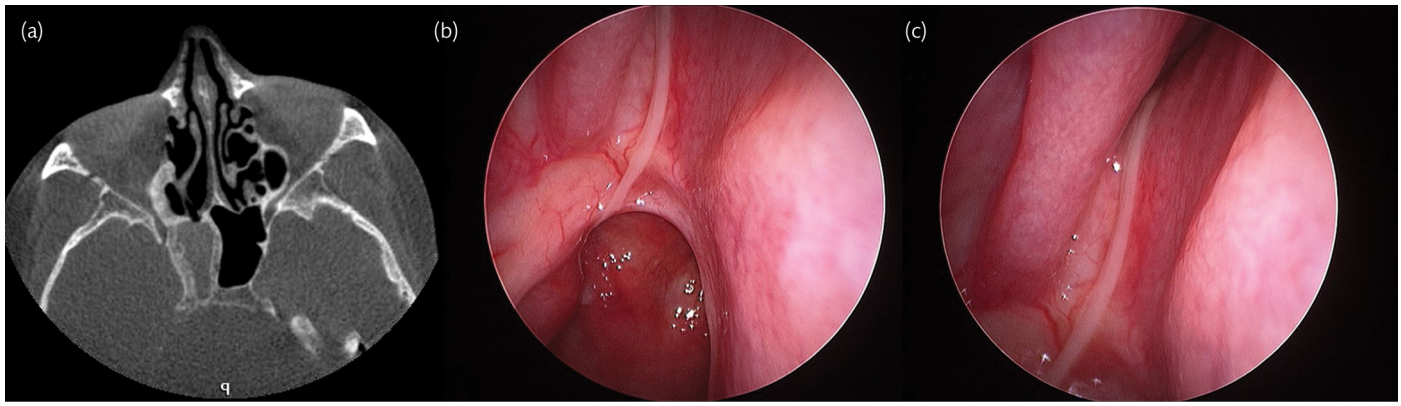

Figure 38.4 Primary localized non-type 2 CRS or chronic isolated sphenoid sinusitis. The left sphenoid is involved on axial computed tomography (a), the bone changes are a reactive response to chronic inflammation. Purulent secretions are seen coming from the sphenoethmoidal recess on straight (b) and angled (c) endoscopy.

either pressure or smell loss in the setting of endoscopic or radiological evidence of mucosal inflammation.

The presence of nasal congestion is challenging as the inflammatory condition is primarily in the sinuses. Although concomitant rhinitis can exist, this nasal congestion is seen even in simple localized forms of CRS (infected fungal ball, odontogenic CRS). Along with the vasodilation that can occur from nearby inflammation, there is an ipsilateral nasal reflex with induced vascular dilation in response to sinus stimulation. Thus, congestion is often used to describe this symptom in CRS, although direct involvement of the nasal cavity with nasal polyps can also cause nasal congestion.

Nasal discharge is a common feature in the setting of CRS. Anterior mucoid discharge is common in inflammatory disease and is often green in appearance. The green pigment of mucous comes from the iron-containing haem groups of myeloperoxidase. Myeloperoxidase comprises 5% of the dry weight of polymorphonuclear cells and it accumulates in the absence of bacteria, reflecting inflammation and not necessarily infection. Sticky or 'chewing gum' mucous is a common feature of eosinophilic conditions that are associated with degranulated eosinophil products in mucin. The presence of fungal material in mucin often gives a 'peanut butter' appearance. Postnasal mucous may just represent a symptom due to the presence of extensive inflammation rather than mucous production. Thus, those patients in whom 'postnasal drip' is a dominant feature, especially in the absence of anterior discharge, suggests comorbid conditions such as extra-oesophageal reflux. Cacosmia associated with discharge is a common feature of CRS and is often of odontogenic origin and a true bacterial causation.

Although facial pressure is defined as a major feature of CRS, it is rarely true facial pain. CRS is inherently a mucosal inflammatory condition and the presence of facial pressure (pain) needs to be proportionate to other symptoms. If mucosal inflammation was causing significant pain, then other mucosal-based symptoms would be present (congestion, discharge). This is the basis for the thought that patients with facial pain as their main symptom are not likely to have CRS as the cause. Similar to isolated 'postnasal drip', patients with facial pain as their main or sole complaint rarely have CRS as the cause.

Loss or distortion of smell is a common finding in CRS but not in rhinitis. Olfaction is sensitive to mucosal inflammation and is a strong predictive factor for CRS. The opposite is true for facial pain.

Many CRS patients have a disease burden and the impact on quality of life is high. CRS has been linked to depressive illness and lethargy. Although not life-threatening, it causes loss of productivity, accounts for a significant proportion of presentations to primary care physicians, and has a large health expenditure cost.

CRS has the following characteristic symptoms:

- Loss of smell is a strong positive predictive factor for the presence of mucosal inflammation in CRS and is uncommon in simple rhinitis.
- Significant facial pain as a dominant presentation is rare. Inflammation of mucosal surfaces that produces pain also produces other significant sinonasal symptoms.
- Infective exacerbations in CRS usually last at least 1–2 weeks. Flare-ups that last only several days are likely to be rhinitis and not rhinosinusitis.
- Cough and other regional symptoms such as dysphonia are more likely to represent broader airway involvement in a chronic inflammatory process, or associated comorbidities such as extra-oesophageal reflux, rather than the direct effects of sinus disease alone.

Endoscopy

Endoscopy is essential in defining CRS and assisting in identifying those patients that have a unilateral or localized disease process as well as any secondary process. For primary CRS patients, there are three characteristics of inflammation described on endoscopy; mucosal oedema/polypoid change, characteristics of the discharge seen (clear, purulent, tenacious), and the anatomical distribution.

Atopy as a comorbid condition in CRS or in the diagnosis of central compartment atopic disease (CCAD) can manifest as middle turbinate oedema, which can be viewed on nasal endoscopy (Figure 38.5).

The middle turbinate is representative of ethmoid mucosa without the vascularity and fibrous stroma that is seen in the inferior turbinate. In patients with allergic rhinitis, inhaled allergens are deposited on the head of the middle turbinate, leading to inflammation and oedema of the mucosa. Patients with diffuse or polypoid oedema of the middle turbinate, on endoscopy, had a higher likelihood of presence of inhalant allergy (positive predictive value = 91.7% and 88.9%, positive likelihood ratios = 8.0 and 6.2, for diffuse and

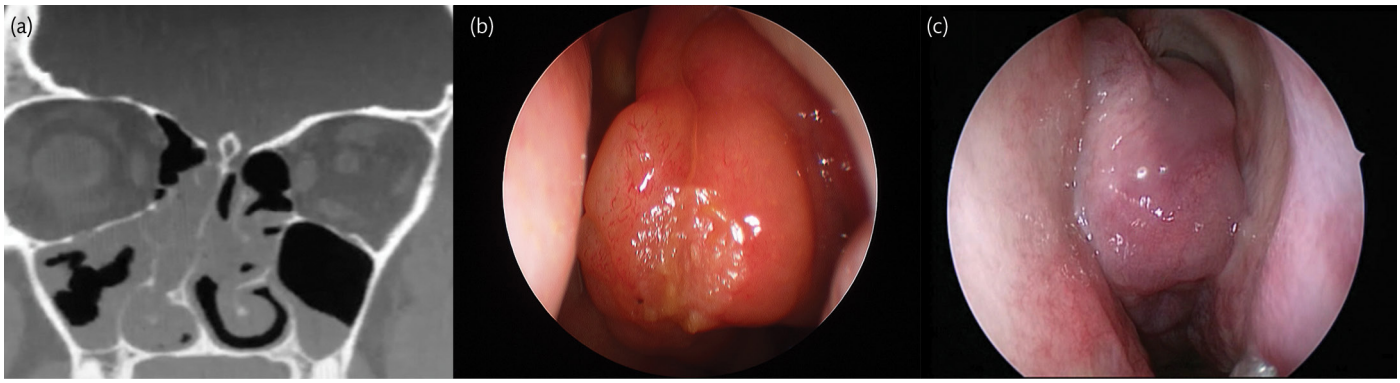

Figure 38.5 The allergic oedema that occurs in the 'central' part of the nose, giving the characteristic radiological appearances (a). These appear due to polypoid mucosal degeneration that occurs from inhalant allergy on the middle turbinate (b) and can appear as 'polyps' arising from either the turbinate or septum (c).

polypoid oedema, respectively). In more severe cases, middle turbinate oedema can progress to involve the superior turbinate and posterior nasal septum and potentially cause obstruction to the 'central' sinus cavity. The sinus ostial occlusion produces barotrauma and trapping of mucin. This condition is referred to as CCAD and is a specific phenotype of CRS (Figure 38.6), and is a sequelae of long-standing advanced allergic rhinitis.

CCAD patients will not exhibit the thick eosinophilic mucin that is noted in eosinophilic CRS or allergic fungal rhinosinusitis. Acute mucosal oedema and 'peau d'orange' appearance is more common in adult eosinophilic CRS, especially in the early phase of the condition. The polypoid changes that arise from within the middle meatus in eosinophilic CRS or allergic fungal rhinosinusitis are true polyps arising from the sinus cavity rather than simply redundant changes from the turbinates as in the allergy-based CCAD.

Secondary CRS, an inflammatory state due to another pathology, is occasionally identified on endoscopy, such as neoplasia. Occasionally, a unilateral medialized lateral nasal wall might correlate with focal symptoms in primary localized CRS, or an odontogenic cause for secondary focal CRS.

Contemporary classification of chronic rhinosinusitis

The presence or absence of polyps offers little insight into the pathophysiology of CRS and such historical classification systems have been replaced by a schema that follows the known pheno/endotypes of CRS that have been described in recent literature. This can be summarized as (Figure 38.6):

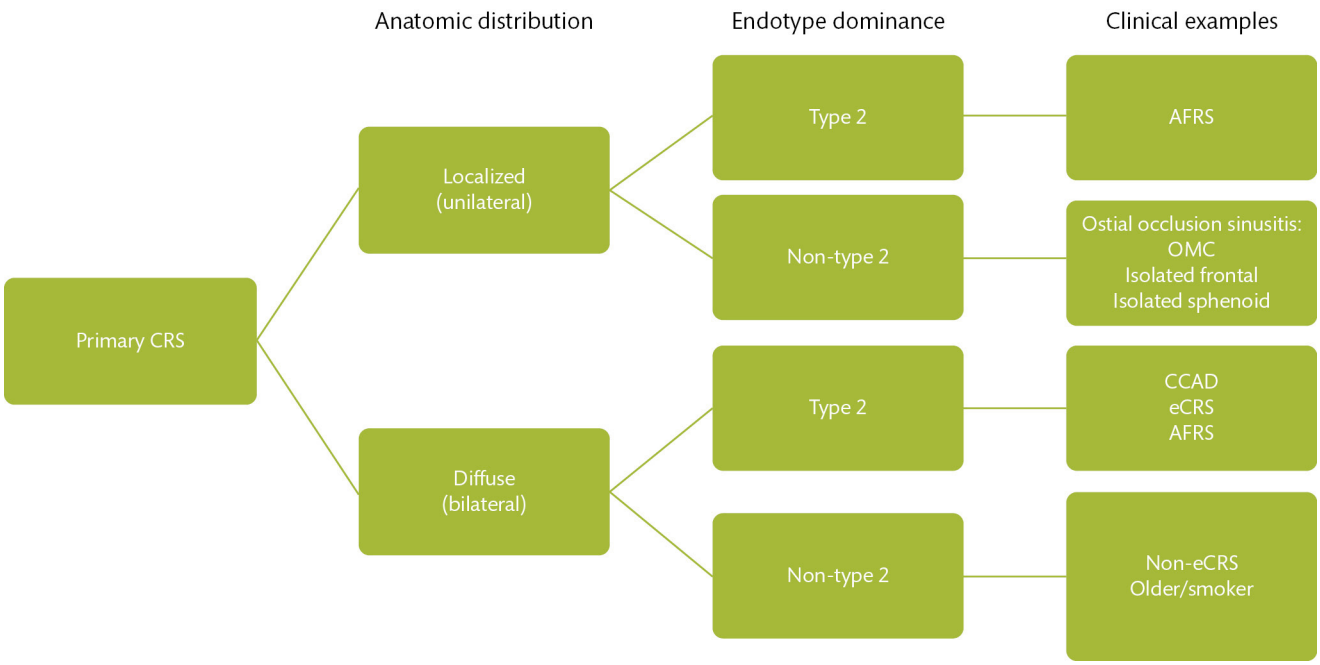

Figure 38.6 The current classification of the CRS condition. This is 'primary' CRS as those patients that have another pathology that produces mucosal inflammation of the airway are considered to be 'secondary' CRS patients. AFRS, allergic fungal rhinosinusitis; CCAD, central compartment atopic sinus disease; eCRS, eosinophilic chronic rhinosinusitis; OMC, ostiomeatal complex.

- primary CRS is a respiratory disorder whereas secondary CRS is either part of local pathology or a broader systemic condition (primary or secondary)
- the anatomical distribution (localized or diffuse)
- the endotype or inflammatory dominance (type 2 or non-type 2).

Primary versus secondary chronic rhinosinusitis

Primary CRS is contemporarily identified as a primary inflammatory disorder of 'the airway' or respiratory system. Patients are defined as having 'primary' CRS if they have a disorder that is 'limited' to their airway/respiratory system *only*. For example, patients that have CRS in the setting of systemic disorders such as immunodeficiencies (e.g. selective immunoglobulin deficiency), autoimmune conditions (e.g. eosinophilic granulomatosis with polyangiitis (EGPA)), odontogenic sinusitis, or local neoplasm *do not* have primary CRS.

Anatomical distribution

Localized

The key feature here is that the sinus cavities involved with disease are 'anatomically discrete' and almost always follow known ostial or functional drainage pathways. Examples are isolated sphenoid sinusitis, isolated frontal sinusitis, or the 'ostiomeatal complex' group of sinuses: maxillary, anterior ethmoid, and frontal. These are examples of anatomically focal or 'localized' disease process (**Figures 38.2 and 38.4**). Usually, these are unilateral conditions, although there may be circumstances in which a localized or discrete process may be occurring synchronously on both sides, although this would be very uncommon. Concomitant lower airway symptoms are also uncommon.

Diffuse

Diffuse airway inflammation describes a broader inflammatory disorder that may affect both the upper and lower airways. Diffuse CRS is usually patchy and diffuse on radiological assessment and does not imply complete opacification of all sinuses, but is not restricted to functional anatomical compartments. There is variability in the degree to which the upper and lower airways are involved as one site may dominate the clinical presentation but usually some degree of disease process exists across the airway.

Endotype dominance

Currently, much is known about the eosinophilic or type 2 inflammatory response in CRS. This inflammation is classically responsive to corticosteroid medications and is the target of biological therapies such as antibodies to interleukin (IL)-4/IL-5/IL-13 receptors. Although several patterns of inflammatory immune response have been described, the most commonly recognized are the type 1, type 2, and type 3 immune responses. These are historically referred to as Th1, Th2, and Th17 immune responses.

Investigations

Radiology

A clinical diagnosis of CRS is not always straightforward, due to the range of symptoms overlapping with other differential diagnoses such as allergic rhinitis, non-allergic rhinitis, primary cephalgia, and extra-oesophageal reflux. The European Paper on Rhinosinusitis and Nasal Polyps (EPOS) 2005 was the first to recommend the use of sinus imaging with or without nasal endoscopy to confirm the diagnosis of CRS. To date, sinus imaging is not a requirement for the diagnosis of rhinosinusitis in primary care; however, computed tomography of the sinuses has become the gold standard when clinical symptoms and endoscopy are not confirmatory. Imaging of the sinuses excludes other pathology and provides additional information, such as the distribution of disease (localized or diffuse) and sinonasal anatomy, which is essential in planning of endoscopic sinus surgery (**Figure 38.7**).

Many previous studies have reported on various incidental sinus abnormalities such as thickening or sinus opacification, with the prevalence of incidental sinus findings common in the normal population with mucous extravasation cysts (10–15%) and mucosal thickening (25% of 'normal' patients have a Lund–Mackay (LM-CT scoring system) score <4). The range of 'normal' LM score in a general population is yet to be determined. A recent systematic review explored the range of reported 'normal' LM scores of between 1 and 4 and thus the EPOS2020 consensus suggested a LM score of greater than 5 has an excellent positive predictive value for CRS.

The hallmark of inhalant/immunoglobulin (Ig)-E-driven CRS, or CCAD, is the central thickening of the turbinate and septum with near-normal peripheral sinus mucosa. On computed tomography scanning, the sinus changes will appear as soft tissue thickening in the central portion of the nasal cavity with sparing of the roof and lateral walls of the sinus cavity (**Figure 38.5**).

Other modalities of imaging, such as magnetic resonance imaging (MRI), are used mainly to assess the characteristics of extensive soft tissue changes, where fungal or neoplastic conditions may coexist. MRI mucosal thickness is weakly correlated with endoscopic assessment ($r = 0.267$, $p < 0.001$). MRI mucosal thickness less than 4 mm is unlikely to represent inflammation (77.0% negative predictive value). One large-scale MRI study reported MRI mucosal thickening in 49% of the general population. Similar studies of inpatients without sinonasal pathology have reported the prevalence of MRI mucosal thickening to be as high as 60% and suggested that such radiographic changes may not be associated with symptoms, thus making MRI an imaging modality with poor specificity for confirmation of CRS.

Blood

Eosinophilia is important to measure in patients with diffuse CRS as it defines a type 2 endotype with greater than 0.25×10^9 cell/L, and confers a poor prognosis when greater than 0.45 cells $\times 10^9$/L. It may also be a marker for an underlying autoimmune condition. Inflammatory markers, such as C-reactive protein and erythrocyte sedimentation rate, are rarely raised in primary CRS. Immunoglobulins are rarely assessed as the presence of immunodeficiency plays a minor role in CRS. Unless there is evidence of broader disease at other non-respiratory sites, minor immunodeficiencies are unlikely to be of major clinical relevance. Isolated IgA deficiency, the commonest form of immunodeficiency with a prevalence of 1 in 600, has been shown to be more prevalent in the refractory CRS cohort than compared to the general population.

Allergy testing

Patients that suffer from allergic airway inflammation have higher serum-specific IgE when compared with those that are suffering from other subtypes of respiratory disease. These patients

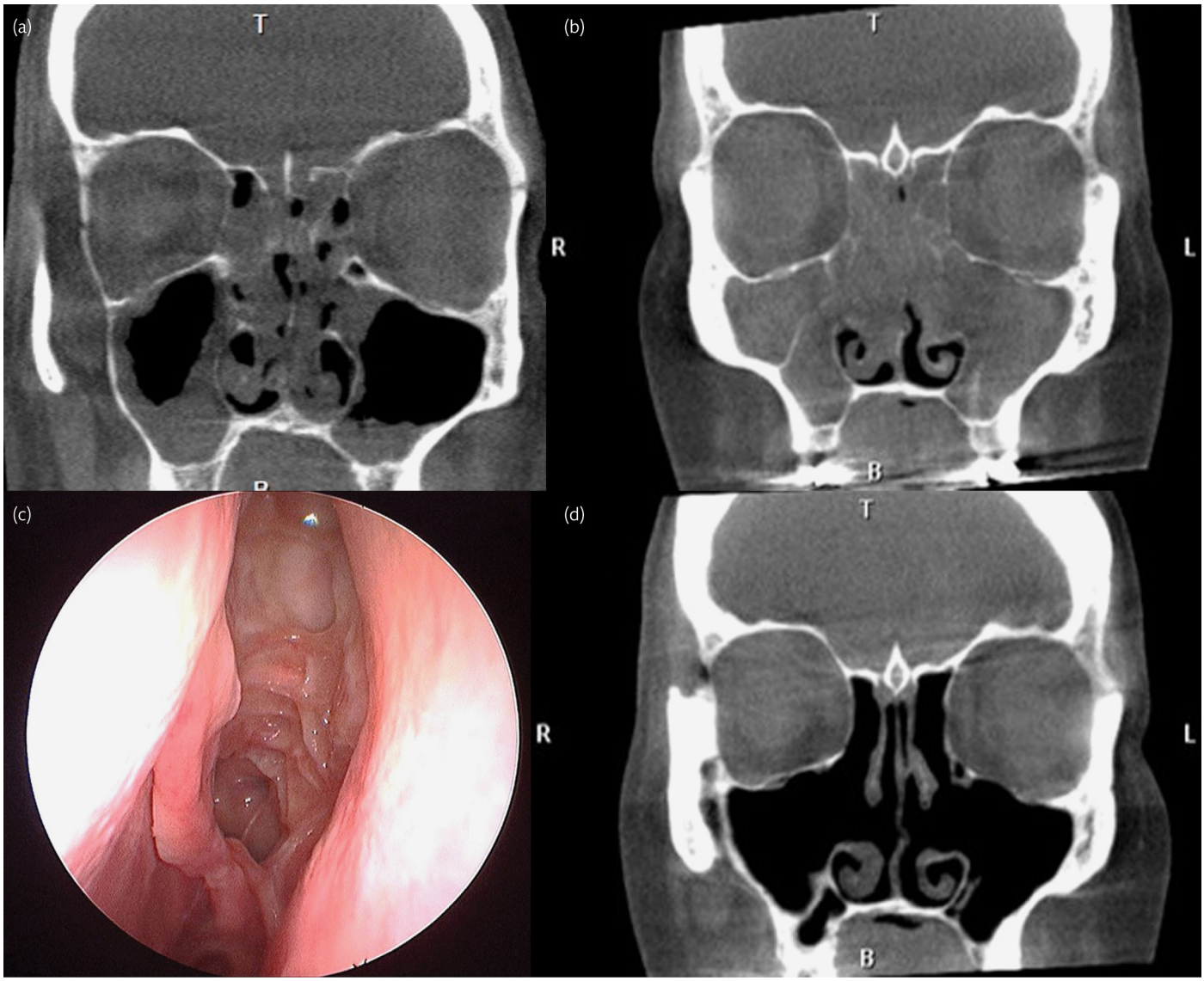

Figure 38.7 Primary diffuse CRS can be either patchy in distribution (a) or very dense (b), but does not imply 'pansinusitis', but simply that the inflammation is not restricted to any one anatomically functional area. Endoscopic sinus surgery in primary diffuse disease achieves a cavity that is treatable with local therapies (c). The subsequent 'neo-sinus' cavity overcomes mucous trapping and pressure phenomenon, and allows for easier delivery of topical therapies, and differs to the surgery applied to primary localized CRS (d).

will have evidence of atopy on skin prick test or immunoassay/radioallergosorbent test. Their clinical history should confirm clinically relevant trigger factors. Persistent allergen exposure and inflammation are normally required to produce the CCAD-phenotype. Hence, seasonal allergens are less likely to cause CCAD compared with perennial allergens such as dust mite.

Differentials diagnoses/associated conditions

Extra-oesophageal reflux disease

While gastro-oesophageal reflux disease is an increasingly prevalent condition that affects up to 20% of the population in the Western world, extra-oesophageal reflux disease (EORD) has been proposed to contribute to the pathogenesis of CRS by causing sinonasal congestion and mucosal inflammation. It is postulated that direct chemical inflammation occurs secondary to contents of reflux, such as pepsin/pepsinogen, entering the sinus cavities. Gastro-oesophageal reflux disease and CRS are both common conditions, so it is not unlikely that they can often coexist. However, there is significant evidence that the association is more than by chance alone. Either EORD alone or in conjunction with minor sinus inflammatory changes can mimic significant CRS symptoms. Just as EORD can make asthma patients appear 'poorly' controlled, such as unresponsive cough and bronchospasm, so too might EORD contribute to 'poorly' responsive CRS symptoms, such as postnasal drip, cough, and congestion.

Ciliary dysfunction disorders (a secondary chronic rhinosinusitis)

Cystic fibrosis is a chronic genetic multisystem disorder characterized by recurrent endobronchial infections, progressive obstructive

pulmonary disease, and pancreatic insufficiency with intestinal malabsorption. Almost 100% of cystic fibrosis patients will have radiological evidence of sinus disease, with up to 67% having nasal polyposis. The diagnosis is often made earlier in life due to the pulmonary manifestations. However, in the low-penetrance cohort the diagnosis may rarely be delayed with presentation as an undiagnosed condition in an uncontrolled CRS patient.

Primary ciliary dyskinesia (PCD) is a genetically heterogeneous recessive disorder characterized by abnormal structure and/or function of motile cilia and sperm flagella. The incidence is estimated at 1:15,000 but likely underestimated due to difficulties in diagnosis. The extra-sinonasal manifestations in PCD include neonatal respiratory distress (>80%), chronic wet-sounding cough since childhood, recurrent middle ear disease, 'atypical' asthma that does not respond to standard treatment, and reduced fertility rather than infertility. In 50% of PCD patients, the defective function of embryonal nodal cilia leads to laterality defects including situs inversus. Due to the non-specific presentation of PCD and varying severity of the disease, patients are often misdiagnosed as uncontrolled primary diffuse CRS and then subsequently reclassified as a secondary diffuse mechanical CRS.

Common autoimmune conditions (a secondary chronic rhinosinusitis)

EGPA is a granulomatous necrotizing small vessel vasculitis, initially described by Churg and Strauss in 1951. EGPA presents with upper and lower airway inflammation with rhinitis, CRS, and asthma. Other autoimmune conditions that predispose towards CRS are granulomatosis with polyangiitis (GPA) and sarcoidosis. Although GPA has universal sinonasal involvement, it typically presents with nasal crusting, obstruction, epistaxis, and septal perforation and is only rarely an underlying pathology in secondary diffuse CRS.

Treatment

Primary chronic rhinosinusitis

Medical

The traditional concept of 'ventilating' a sinus cavity or simply 'draining' does not reflect accurately the pathological processes that produce CRS either in animal models or in humans. The complete physical 'obstruction' of a sinus ostia does not necessarily lead to CRS, as mucocoele formation and atelectasis are also common features of ostial occlusion. Most patients with primary localized CRS will require a surgical intervention as there is invariably an identifiable local anatomical factor contributing to the pathophysiology. Medical therapy can occasionally resolve this but the need for surgical intervention is high. In primary diffuse disease, although surgery may be used at some stage, treating the three processes (see below: mucosal inflammation, microbial dysbiosis, mucostasis) that lead to chronic mucositis are essential.

Treatment targeting intrinsic mucosal inflammation

Mucosal inflammation is the defining feature of CRS. It may be intrinsic or occur as a result of local infection or environmental exposure. Glucocorticoids have formed the mainstay of medical therapy for CRS, while long-term macrolides, doxycycline, and other novel approaches also target the inflammatory pathway. The main drawback for glucocorticoids is that simple intranasal sprays do not deliver medication to the sinuses and distribute primarily to the nasal cavity only. Systemic therapy is required to adequately reach the sinuses in the unoperated state. Corticosteroids are most effectively delivered via larger volumes and in the postsurgical sinus cavity.

New biological therapies targeting IgE (omalizumab), IL-5 (benralizumab, mepolizumab), and IL-4/IL-13 (dupilumab) show promise for the treatment of upper airway inflammation although at present their cost precludes widespread use for this purpose.

Treatment aimed at reducing microbial load

Treatment aimed at reducing microbial load or eradicating pathogens from the sinuses assumes that these play a role in causing or propagating CRS. Bacteria may play a role in acute infective exacerbations. Bacterial exotoxin may trigger an inflammatory response, while superantigens (predominantly staphylococcal) and fungi may induce eosinophilic inflammation. The organisms isolated from patients with CRS differ from those in acute rhinosinusitis, with *Staphylococcus aureus*, *Staphylococcus epidermidis*, and anaerobic and Gram-negative bacteria predominating. The bacterial colonization, however, is often an opportunistic infection in the setting of an inflamed mucosal barrier or mucostasis and often not the primary factor contributing to the CRS state. Antimicrobial therapy alone rarely resolves CRS.

Treatment aimed at improvement in mucociliary clearance

Normal mucociliary clearance is dependent upon both ciliary function and the mucous blanket. Saline irrigation can improve mucociliary clearance by the removal of abnormal mucous, infected crusts, and proinflammatory agents. This has been confirmed by a Cochrane meta-analysis, which included eight studies and demonstrated benefit from saline irrigation both when used as sole modality treatment and as an adjunct, albeit less efficacious than intranasal steroid. Large-volume irrigation appears to have a greater impact on mucous management than simple sprays on randomized controlled trials. This mucous-clearing effect is amplified postsurgery.

Surgical

Contemporary and historical surgical interventions for the paranasal sinuses range from dilation of the natural ostia to interventions that completely remodel the anatomy and drainage pathways. This also includes procedures that are intended to leave behind a 'functional' sinus cavity and other surgical interventions designed to 'obliterate' the neo-sinus cavity. Endoscopic sinus surgery is an important component in the management of medically refractory CRS, to facilitate the long-term application of topical therapies, for the prevention of mucostasis, and restoration of sinus ventilation (**Box 38.1**). It has been demonstrated that more extensive sinus surgery results in better distribution of topical irrigation therapy, which is the mainstay of long-term management of CRS.

Essential elements in surgery

- An analysis of preoperative imaging to recognize variants of normal anatomy and areas for potential complication in order to perform safe and comprehensive surgery.

Box 38.1 The goals of endoscopic sinus surgery in the setting of chronic rhinosinusitis

- Improved ventilation.
- Facilitation of drainage/mucociliary clearance.
- Improving the nasal airway.
- Minimizing damage to uninvolved structures.
- Providing access for topical therapy.
- Reduced mucosal surface area.
- Improving odorant delivery to the smell cleft.
- Ability to survey the sinus cavity over time.

- Identification of the anatomical limits that delineate the surgical boundaries of dissection, applying the concept of the paranasal box (with its vertical 'frontal' component), provides a basic framework for endoscopic sinus surgery.
- Early identification of key anatomical landmarks of the maxillary sinus roof (orbital floor), medial orbital wall, and sphenoid sinus (thus posterior skull base) guides the surgeon through the variable and potentially previously modified sinus anatomy.
- The ultimate goal of surgery is to create a functional cavity. Removing all bony partitions within a sinus functional unit/space and ensuring that all natural ostia have been incorporated into the surgical cavity will prevent mucous recirculation/trapping.
- The ability to view the limits of dissection with a single position of the endoscope avoids leaving behind disconnected cells from the surgical cavity preventing mucocoele formation and mucous re-circulation, and enables maximal delivery of topical therapy in the postoperative setting.

Current teaching has shifted away from defining the pneumatization clefts to identifying the important (or fixed) anatomical landmarks and thus making removal of the partitions (or variable anatomical components) safe and quick. This is not a novel concept. Many temporal bone surgeons have shifted from concepts of the 'antral air cell and short process of incus' or 'retrofacial air cell tract' to more fixed anatomy (dura, sigmoid sinus, sinodural angle, vertical facial nerve), and, in the process, acknowledged that many of these previous surgical features were variable and often heavily affected by pathology.

There are two key fixed spaces: the maxillary and sphenoid sinuses. Both ethmoid and frontal sinus cavities can be highly variable and often affected by previous surgery or pathology. Additionally, knowing the exact location of the orbit or orbital axis (i.e. visualizing the entire length of the orbit from the medial orbital wall, to the apex, and then the optic nerve canal) is critical for more extended procedures. The maxillary sinus or orbital floor is found early to identify the medial orbital wall. This allows a safe level to proceed through the basal lamella to enter the sphenoid. The sphenoid sinus provides the surgeon with the level of the skull base. The 'corner pocket' of the sphenoid also serves to identify the orbital apex and skull base junction point, and with the anterior landmarks forms the basis of defining the orbital axis. The skull base can then be defined from known-to-unknown through to the posterior table of the frontal sinus if required (Figure 38.7).

Secondary chronic rhinosinusitis—specific approaches

Identifying and managing the underlying disease process is essential, whether this is 'localized' pathology (neoplasia, odontogenic, or fungal ball) or a 'diffuse' process, such as mechanical (cystic fibrosis/PCD), inflammatory (GPA, EGPA), or immune-related (common variable immunodeficiency, IgA deficiency, diabetes mellitus). For those secondary localized CRS patients, surgery is required in cases of fungal ball, odontogenic disease, or sinonasal tumour. For those with secondary diffuse CRS, much greater care needs to be taken to identify and optimize any underlying mechanical/mucociliary defect, broader inflammatory disorder (autoimmune conditions, GPA, or EGPA), and those with immune deficiencies.

Tips and pearls

Common pitfalls for refractory disease:

- Wrong diagnosis: treated as CRS when there is a broader systemic condition occurring, including gastro-oesophageal reflux disease.
- Ineffective delivery of therapy (reliance on sprays or topical therapy in non-operated patients).
- Inappropriate therapy: incorrect endo/phenotyping (i.e. corticosteroid for non-eosinophilic/non-type 2 disease).
- A failed sinus: lack of mucociliary return and 'bronchiectasis-like' state to the upper airway.
- Severe form of disease not controlled with topical therapies alone even with surgery.

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39

Sinonasal tumours

Carl H. Snyderman and Eric W. Wang

Introduction

Sinonasal tumours include a wide variety of benign and malignant tumours with different biological behaviours. The differential diagnosis is aided by advances in pathological testing and reclassification of tumours. Sinonasal tumours require different treatment strategies, depending on their biological behaviour. Dogmatic approaches are being replaced by new surgical techniques and treatment paradigms. Endoscopic surgery enables oncological resection of most sinonasal tumours with less potential morbidity and equivalent oncological outcomes. Parallel advances in medical and radiation oncology are also providing new treatment options.

Anatomy

The paranasal sinuses are bounded by the anterior cranial base, orbit, and middle cranial fossa. A basic understanding of surface anatomy of the skull base is essential for any sinus surgeon. The anterior cranial base extends from the frontal sinuses to the optic canals and laterally to the orbital roof. The ethmoidal arteries are branches of the ophthalmic artery and diverge as they cross the cranial base from the orbit. The anterior ethmoid artery is between the second and third lamellae of the ethmoid sinus, just posterior to the nasofrontal recess. Radiographically, it is situated in the coronal plane tangential to the posterior surface of the globe. The frontal sinuses and cribriform plates are separated anteriorly by the crista galli. The attachment of the middle turbinate separates the olfactory sulcus (cribriform plate) from the ethmoid sinus.

The most important real estate is in the region of the sphenoid sinus: optic canals, internal carotid arteries (ICAs), and pituitary gland (Figure 39.1). Key landmarks include the planum sphenoidale, optic canals, lateral optic-carotid recesses, sella, clival recess, and parasellar and paraclival segments of the ICA. The lateral recess of the sphenoid sinus is bounded by the maxillary nerve in the foramen rotundum superolaterally and the vidian nerve in the pterygoid canal inferomedially. The vidian nerve is an important surgical landmark for exposure of the petrous segment of the ICA and Meckel's cave.

The lacrimal sac is located in the lacrimal fossa adjacent to the axilla of the middle turbinate. The nasolacrimal duct runs within the bone of the medial maxillary wall at the anterior limit of the middle meatus, entering the nasal cavity deep to the anterior attachment of the inferior turbinate.

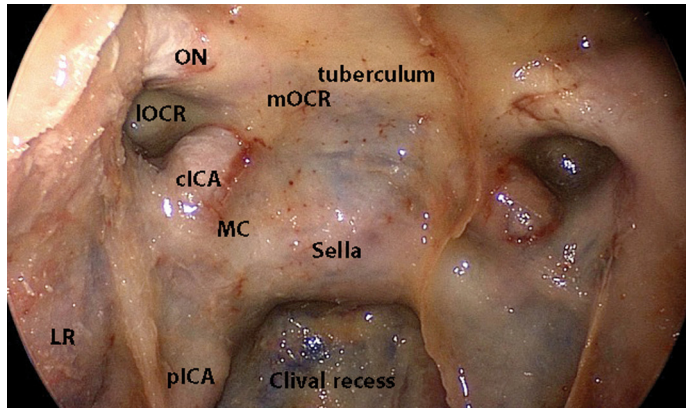


Figure 39.1 Anatomical landmarks of the sphenoid sinus. cICA, cavernous internal carotid artery; lOCR, lateral optic-carotid recess; LR, lateral recess; MC, middle clinoid; mOCR, medial optic-carotid recess; ON, optic nerve; pICA, paraclival internal carotid artery.

Assessment

Sinonasal tumours often present late due to non-specific symptoms that are confused with inflammatory sinus disease. Atypical symptoms such as persistent pain and epistaxis, or signs of tumour extension beyond the paranasal sinuses (orbital symptoms such as diplopia, visual loss, proptosis, epiphora; trigeminal nerve dysfunction including hypoaesthesia and weakness of muscles of mastication), are suggestive of a tumour diagnosis. Nasal endoscopy is an essential part of the examination to determine the location and extent of the tumour. In particular, questions to consider include: what is the site of origin? Is the tumour unilateral or bilateral? Is it invading or displacing the nasal septum? Does it extend to the skull base? Does it invade the orbit? Does it involve the palate? Is it hypervascular?

The examination should also address extranasal features of tumour extension/invasion: vision loss, proptosis, paresis of

extraocular muscles, trigeminal nerve deficits, trismus, and facial swelling. Lacrimal swelling may be due to obstruction of the nasolacrimal duct by intranasal tumour or tumour growth within the duct. Examination of the neck for cervical lymphadenopathy is important for staging of malignant tumours. Metastatic lymphadenopathy at presentation is associated with high-grade malignancy such as squamous cell carcinoma (SCC) or sinonasal undifferentiated carcinoma (SNUC).

Although biopsy of the tumour at presentation is desirable to establish a diagnosis and guide management, it may not be feasible due to inaccessibility of the tumour or risk of haemorrhage. In particular, hypervascular tumours such as angiofibromas should not be biopsied due to the risk of brisk haemorrhage. Be wary of a smooth pulsatile mass high in the nasal vault—it could be a meningoencephalocoele of the anterior cranial fossa.

Investigations

Radiological assessment of sinonasal tumours provides essential information regarding the extent of the tumour, its relationship to critical structures, stage, and composition. Tumours such as angiofibroma have a characteristic appearance and diagnosis can usually be established without biopsy. For other tumours, the differential diagnosis can be narrowed with imaging.

Computed tomography (CT) and magnetic resonance imaging (MRI) provide complementary information. CT is superior for demonstration of bony anatomy and is usually the first choice for imaging. The CT scan should be carefully studied to identify anatomical variations or areas of bone erosion that may increase surgical risks. For planning purposes with surgery, a CT angiogram with navigation protocol is obtained for improved identification of the ICA during surgery. MRI is superior for delineation of soft tissues, especially when the tumour extends beyond the boundaries of the sinuses into the cranial cavity or orbit. Unlike CT, T2-weighted MRI sequences can help differentiate between mucous or tumour within an opacified sinus. With large tumours, both CT and MRI are obtained and can be fused for intraoperative navigation.

Positron emission tomography (PET) is used to assess metabolic activity of malignant neoplasms and can be fused with CT or MRI to screen for distant metastases and differentiate inflammatory from neoplastic lymphadenopathy. It is also useful to establish a baseline of metabolic activity prior to chemotherapy or radiotherapy (RT) for comparison. For neuroendocrine tumours such as olfactory neuroblastoma or neuroendocrine carcinoma that express somatostatin receptors, imaging with ^{64}Cu DOTATATE PET-CT provides greater specificity and may prove to be superior for detection of metastases and recurrent disease.

Angiography is generally reserved for highly vascular tumours such as angiofibroma. It is both diagnostic and useful in preoperative planning. Embolization of the tumour's extracranial blood supply (internal maxillary artery) may be performed at the same time in preparation for surgery. The ethmoidal arteries cannot be embolized safely due to the risk of blindness from loss of the ophthalmic artery. When surgery requires dissection within the masticator space, preoperative sacrifice of the internal maxillary artery also facilitates dissection without concern for vascular injury.

Table 39.1 UPMC staging system for angiofibroma

Stage I	Nasal cavity, medial PPF
Stage II	≥1 sinus, lateral PPF; no residual vascularity
Stage III	Skull base erosion, orbit, ITF; no residual vascularity
Stage IV	Skull base erosion, orbit, ITF; residual vascularity
Stage V	Intracranial extension, residual vascularity; M: medial extension; L: lateral extension

ITF; infratemporal fossa; PPF; pterygopalatine fossa.

Staging

A variety of staging systems have been proposed for angiofibroma. The most common systems focus on tumour location and extent. In contrast, the University of Pittsburgh Medical Center (UPMC) staging system incorporates the vascularity of the tumour and provides superior prognostication regarding intraoperative blood loss and risk of residual/recurrent disease (Table 39.1).

The TNM (tumour, node, metastasis) staging system can be used for most malignant sinonasal tumours although alternate staging systems have been proposed for specific tumour types. The Kadish staging system is the most widespread staging system for olfactory neuroblastoma but is imprecise and provides poor discrimination of prognostic groups. It has been modified to include staging of the neck and dural infiltration. A modification of the TNM staging system has been proposed by Dulguerov.

Differential diagnoses/ associated conditions

One of the challenges of managing sinonasal tumours is the diversity of pathologies. Proper diagnosis requires a skilled head and neck pathologist. Although the differential diagnosis can often be narrowed based on demographic and radiological features alone, differentiation of tumours with similar histological appearance requires the use of immunohistochemistry and molecular markers. In particular, 'small blue cell' tumours are often misdiagnosed and include both benign and malignant tumours with different treatment strategies. The fourth edition of the World Health Organization classification of head and neck tumours provides a completely revised classification of sinonasal tumours with the inclusion of several new entities. Some of the most relevant sinonasal tumours are discussed here.

Benign sinonasal tumours

Inverted papilloma

Although these tumours are benign, they are locally aggressive with an infiltrative growth pattern. Cure requires complete resection. An endoscopic endonasal approach may be supplemented with a Caldwell–Luc approach if the tumour is extensive or laterally based. Drilling of the bone at the site of origin decreases the risk of recurrence. Tumours should be carefully examined histologically for evidence of dysplasia or SCC in approximately 5% of patients.

Angiofibroma

Angiofibromas are rare benign tumours found almost exclusively in adolescent males. They arise at the basisphenoid in proximity to the sphenopalatine foramen. These highly vascular tumours derive most of their blood supply from branches of the internal maxillary artery but can recruit significant contribution from the ICA as they enlarge. Angiofibromas follow pathways of least resistance, growing through fissures and foramina to gain access to the nasopharynx and paranasal sinuses, masticator space, infratemporal fossa, orbit, and middle cranial fossa.

The preferred management of angiofibroma is surgery. Early-stage tumours are readily excised using endoscopic techniques, often without the need for preoperative embolization. Advanced stage tumours with residual vascularity from the ICA require preoperative embolization of external carotid artery branches. Surgery may need to be staged based on tumour extent and intraoperative blood loss. Giant tumours that extend intracranially or fill the infratemporal fossa may be safely removed using endoscopic-assisted or completely open approaches, depending on the experience of the surgical team. The bone of the pterygoid base is often infiltrated by tumour and drilling of this bone is necessary to remove all remnants.

Recurrence rates are similar for open and endoscopic approaches. Residual tumour may be observed for growth prior to committing to additional therapy. For accessible tumour recurrence, repeat endoscopic surgery is preferred; high-risk areas such as the cavernous sinus may be observed or treated with radiation (stereotactic radiosurgery). Patients should be followed for at least 3 years with imaging to detect recurrence.

Glomangiopericytoma

Glomangiopericytoma is a highly vascular tumour of borderline or low malignant potential that can occur in the nasal cavity. These indolent tumours should not be confused with solitary fibrous tumour, previously called haemangiopericytoma, which has greater malignant potential. Surgical excision is the treatment of choice.

Malignant sinonasal tumours

Squamous cell carcinoma

SCC may arise anywhere in the sinonasal tract, either *de novo* or from inverted papilloma. Roughly half arise within the nasal cavity; the maxillary sinus is the most common paranasal sinus of origin followed by the ethmoid sinus. Unlike SCC arising in the oral cavity, pharynx, or larynx, there is not a strong association with tobacco use. SCC includes multiple subtypes: keratinizing, non-keratinizing, spindle cell, and lymphoepithelial. In contrast to keratinizing SCC, the non-keratinizing type is associated with human papillomavirus but the prognostic significance of this is not well defined. Cervical metastases at presentation are associated with advanced stage tumours, particularly the maxillary sinus.

The treatment strategy is similar to SCC arising in other sites. Combination therapy with complete surgical excision followed by RT provides the best outcome. Oncological outcomes are comparable with open and endoscopic approaches. Treatment of the neck is dependent on preoperative imaging. Advanced tumours that involve the orbit or skull base may be treated with induction chemotherapy followed by RT or surgery, depending on the response. A good response to induction chemotherapy is a favourable prognostic indicator and may allow preservation of the orbit. Overall, the prognosis for advanced SCC of the nasal cavity and sinuses remains poor with a significant risk of local recurrence. In comparison to other paranasal sinuses, the ethmoid region has a better prognosis, probably due to its greater resectability, even in the presence of local recurrence. SCC arising from inverted papilloma has a similar prognosis to *de novo* SCC.

Olfactory neuroblastoma

Olfactory neuroblastoma, otherwise known as esthesioneuroblastoma, is a malignant neuroectodermal tumour arising from the olfactory epithelium. It is characteristically situated between the nasal septum and middle turbinate in the olfactory sulcus. Because of its proximity to the cribriform plate, skull base involvement is common with risk of dural invasion and intracranial spread. Tumour within the olfactory bulb may have a characteristic cystic appearance on imaging. Olfactory neuroblastoma may occur in young adults; paediatric tumours are often more advanced at presentation and may have more aggressive biological behaviour. Cervical metastases may develop in up to 20% of patients.

Most patients with olfactory neuroblastoma will require resection of the anterior cranial base (see Chapter 44). Superior results are observed with a combination of surgery and postoperative RT.

Neuroendocrine carcinoma

Within the new classification system, neuroendocrine carcinoma consists of two types, large cell and small cell. It has an aggressive biological behaviour with increased risk of both lymphatic and distant metastases. For these reasons, combined modality treatment including RT and chemotherapy is usually recommended. Surgery is an option for early-stage tumours but is more often used as salvage treatment following radiochemotherapy.

Sinonasal undifferentiated carcinoma

SNUC is an aggressive neoplasm that often presents in an advanced stage with intracranial spread and cervical metastases. Good responses to induction chemotherapy followed by RT have been observed. Surgery is usually reserved for evidence of residual tumour following radiochemotherapy.

NUT carcinoma

NUT carcinoma is a poorly differentiated carcinoma, often with squamous differentiation, that typically afflicts young adults. The NUT carcinoma shares a rearrangement of the nuclear protein in testis (*NUTM1*) gene with testicular cancer. It is an aggressive cancer with a very poor prognosis and does not respond well to standard therapy. Some clinical response to bromodomain inhibitors underscores the importance of a proper diagnosis.

Adenoid cystic carcinoma

Adenoid cystic carcinoma is a malignant tumour of minor salivary gland origin. It has a predilection for perineural spread and frequently involves branches of the trigeminal nerve. Proximal perineural growth along branches of the maxillary nerve may extend to Meckel's cave and then follow the course of the mandibular nerve distally. The orbital apex and cavernous sinus are often involved. For these reasons, complete surgical excision is rarely possible. Adenoid cystic carcinoma with a predominantly solid growth pattern has a

worse prognosis compared to the cribriform and tubular patterns. The most common site for metastasis is the lung and patients should be screened with a CT scan of the chest.

When feasible, radical excision followed by RT offers a survival advantage. Orbital exenteration remains controversial. Although it may improve local control, it may not alter long-term prognosis. Patients can live for many years with persistence of local disease or pulmonary metastases. In the presence of skull base involvement, maximal resection with minimal morbidity is the goal with preservation of the orbit, cranial nerves, and ICA. Residual tumour is treated with RT. Alternatively, good responses have been observed with RT, in particular proton beam therapy, as the primary treatment.

Adenocarcinoma

Sinonasal adenocarcinoma is divided into two types: intestinal type and non-intestinal type. The intestinal type is associated with the woodworking industry (exposure to hardwood dust). Although it has the histological appearance of adenocarcinoma of the intestines, it does not represent a metastasis and evaluation of the gastrointestinal tract is not mandatory in the absence of symptoms. Intestinal type adenocarcinoma is often multifocal and wide excision of sinus mucosa is recommended. The non-intestinal adenocarcinoma is further classified as low grade or high grade. Surgery alone with good margins may be sufficient for low-grade adenocarcinoma whereas adjunctive RT is recommended for high-grade adenocarcinoma.

Sarcoma

Sarcomas of the sinonasal region are extremely rare. A new entity in the World Health Organization classification of sinonasal tumours is the biphenotypic sinonasal sarcoma (formerly low-grade sinonasal sarcoma with neural and myogenic features). Local recurrence is common following treatment, but distant metastases have not been reported.

Box 39.1 Management paradigm for malignant sinonasal tumours

Surgical resection
- Low-grade chondrosarcoma.
- Some low-grade adenocarcinomas.
- Some low-grade sarcomas.

Surgical resection and postoperative radiation therapy
- Olfactory neuroblastoma.
- Adenocarcinoma.
- Adenoid cystic carcinoma.
- SCC.
- Most metastases.
- Some low-grade sarcomas.

Pre- and postoperative chemotherapy, surgical resection, and postoperative radiation therapy
- SCC.
- High-grade sarcoma.
- SNUC.
- Neuroendocrine carcinoma.
- Melanoma.

Chemotherapy and radiation therapy
- Lymphoma.
- Ewing's sarcoma.
- Rhabdomyosarcoma.
- Malignant peripheral nerve sheath tumour.
- Some patients with SNUC and other neuroendocrine carcinomas.

Chemotherapy, radiation therapy, surgical resection, and stereotactic radiosurgery
- SCC, especially with perineural extension.
- Adenoid cystic carcinoma.
- Some high-grade sarcomas, SNUC.

Adapted from Demonte F. Management considerations for malignant tumors of the skull base. *Neurosurg Clin N Am*. 2013;24(1):1–10.

Treatment

The choice of treatment depends on a multitude of factors including diagnosis, stage, patient comorbidities, surgical team experience, and patient values. Surgery is utilized for benign tumours. Malignant tumours can be grouped according to biological behaviour. Resectability depends on proximity to critical structures and the ability to obtain clear margins. Generally, low-grade malignancies are treated with surgery; adjunctive RT depends on resection margins and stage of the tumour. With high-grade malignancies, treatment options include (1) surgery followed by radiochemotherapy, and (2) induction chemotherapy followed by surgery or RT (**Box 39.1**).

Surgery

The majority of sinonasal tumours are now managed using endoscopic surgical techniques when surgery is indicated. The endonasal corridor may be supplemented with an endoscopic-assisted transmaxillary corridor (transoral sublabial approach). Endoscopic approaches to the anterior cranial base are covered in Chapter 44. Relative contraindications to endoscopic surgery include tumour extension beyond the paranasal sinuses and involvement of superficial soft tissues. The surgeon's experience with endoscopic techniques is also a factor.

The principles of endoscopic resection include dissection around the periphery of the tumour to identify key anatomical landmarks and devascularize the tumour. Large obstructive tumours often require debulking to improve access. The area of attachment or invasion can then be removed en bloc. Abundant literature confirms that en bloc excision is an outdated concept that cannot be realistically applied to most sinonasal tumours; equivalent oncological results are achieved with piecemeal resection as long as final resection margins are negative. For malignant tumours, oncological resection should include a layer of uninvolved tissue beyond the margin of the tumour. For example, the medial wall of the orbit (lamina papyracea) is resected for tumours of the ethmoid sinus; if the bone is eroded, the adjacent periorbita is also resected. Malignant tumours such as olfactory neuroblastoma that involve the olfactory sulcus may have intracranial spread along olfactory filia and resection of the anterior cranial base should be considered even in the absence of skull base erosion.

One of the greatest challenges intraoperatively is dealing with tumour haemorrhage. Haemostatic strategy starts with preoperative planning. Highly vascular tumours such as angiofibroma and some

high-grade malignancies benefit from preoperative embolization of feeding vessels. Even if the tumour is not hypervascular, dissection in the masticator space is facilitated by angiographic sacrifice of the internal maxillary artery.

Intraoperatively, elevation of the head of bed (reverse Trendelenburg position) by 20° decreases bleeding due to venous congestion. If embolization is not feasible, the vascular supply can be sacrificed intraoperatively. The branches of the internal maxillary artery can be ligated in the retromaxillary space using an endoscopic transantral or anterior transmaxillary approach. The anterior and posterior ethmoidal arteries can be cauterized at the skull base where they exit the orbit by decompressing the medial orbit; rarely, an external approach is used.

A variety of haemostatic tools are useful intraoperatively to coagulate the tumour surface. With endoscopic surgery, an endoscopic bipolar electrocautery device is essential for cauterization of small arteries and tumour surface. The Aquamantys (Medtronic) is a flexible bipolar electrocautery device with a fixed distance between the tips and is well suited for cauterization of mucosa and tumour surface. The Harmonic scalpel (Ethicon) uses ultrasonic energy to cauterize and transect tumour tissue simultaneously. It is used to transect angiofibroma tumour tissue when the tumour is too large to be removed in one piece and to improve access. Coblation (Smith & Nephew) uses plasma technology to cauterize and ablate tumour tissue. Other haemostatic adjuncts include a haemostatic matrix (Floseal (Baxter), Surgiflo (Ethicon)) for venous bleeding and warm saline irrigation at 40°C.

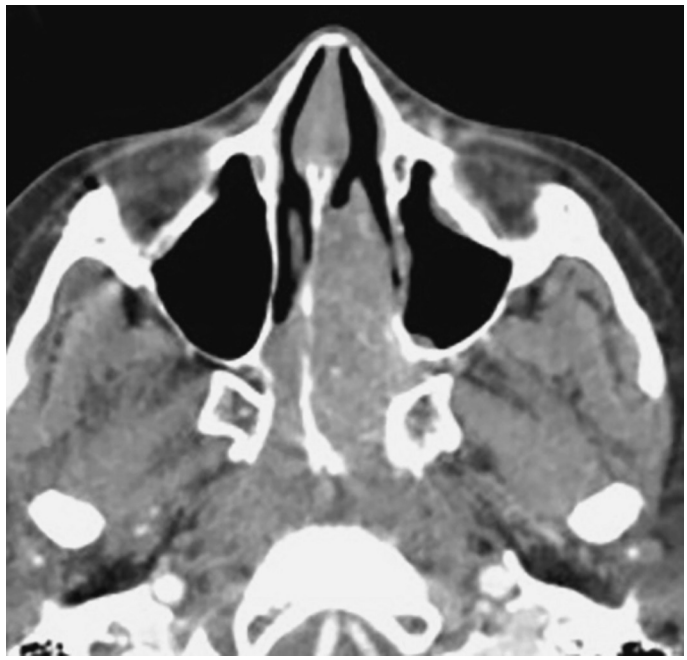
Large vascular tumours such as angiofibromas are removed in vascular units corresponding to their blood supply. For example, the extracranial segments would be removed first on each side, followed by the intracranial segment. Careful dissection along the course of the ICA allows selective cauterization of feeding branches from the ICA. If intraoperative blood loss is excessive (>50% total blood volume), it is advisable to place packing for haemostasis and stage the surgery, especially if there is intracranial tumour remaining.

CASE EXAMPLE 1: ANGIOFIBROMA

A 14-year-old male presented with left nasal obstruction. Nasal endoscopy demonstrated a hypervascular tumour consistent with angiofibroma. This was confirmed by CT imaging (Figure 39.2) with UPMC stage I. Preoperative embolization of the left internal maxillary artery successfully devascularized the tumour. An endoscopic left transpterygoid approach was used to remove the tumour. Sacrifice of the vidian nerve with drilling of the pterygoid base assured complete removal of tumour.

CASE EXAMPLE 2: ADENOCARCINOMA

An 80-year-old female presented with left nasal obstruction and mild epistaxis. Nasal endoscopy revealed a large obstructive tumour. Radiological imaging (Figure 39.3) demonstrated a unilateral tumour with extension to the skull base. The site of origin was indeterminate. Biopsy of the tumour confirmed a low-grade adenocarcinoma. Surgery was recommended as the primary treatment. Endoscopic debulking of the tumour was necessary to reveal a small site of origin on the nasal septum with no evidence of skull base involvement. En bloc excision of the area of septal invasion with negative frozen sections of margins was performed. Final pathology confirmed complete excision. She declined postoperative RT.



Complications of surgery

Complications of surgery for sinonasal tumours are similar to those encountered with endoscopic sinus surgery for inflammatory disease. Good control of haemorrhage throughout surgery is important to maintain good visualization and avoid complications. Intraoperative navigation using preoperative CT or MRI is routinely used to help identify key anatomical structures and confirm margins of the tumour. Team surgery (with one surgeon driving the endoscope) is also an asset, especially when dissecting large hypervascular tumours, with two hands available for haemostasis and manipulation of tumour tissue.

Vascular complications include injury to branches of the external carotid artery. Inadequate cauterization of the sphenopalatine artery can result in delayed epistaxis which can be severe. Injury to the ethmoidal arteries can result in retraction into the orbit with development of a retrobulbar haematoma. This needs to be promptly recognized and treated to prevent visual loss. A carotid injury can result from instrumentation of the sphenoid sinus. The anterior genu of the parasellar segment of the ICA is the most frequent site of injury. Bleeding is controlled with focal packing and the patient is transferred for angiographic evaluation and treatment (stenting or carotid sacrifice).

Inadvertent injury to the medial wall of the orbit results in herniation of orbital fat. It is important to avoid further instrumentation of the orbit with potential orbital haemorrhage or injury to the extraocular muscles. The optic nerve and ophthalmic artery can be injured

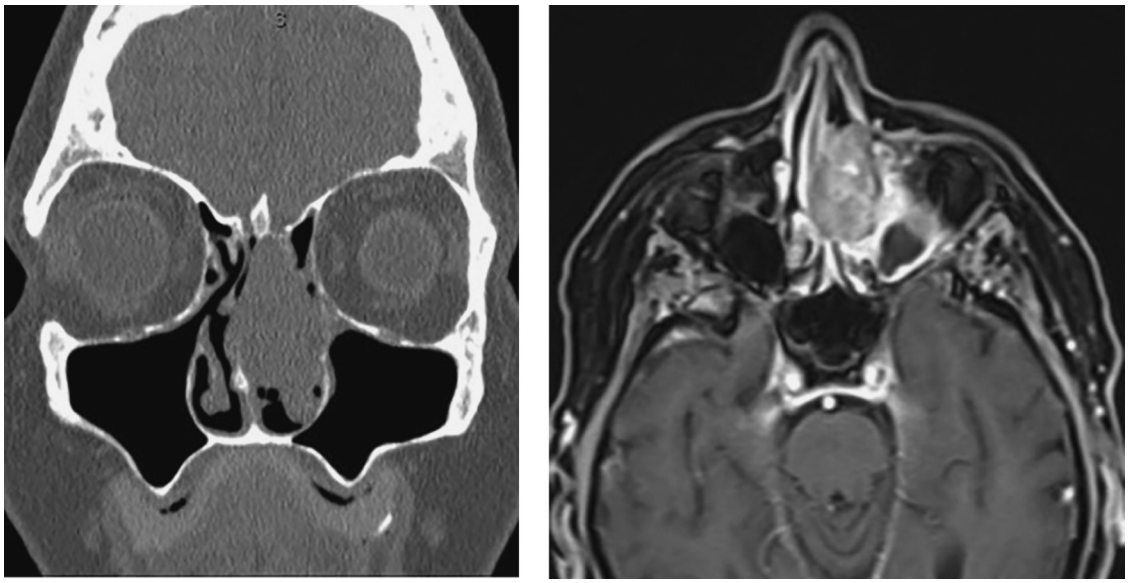


Figure 39.3 Coronal CT (a) and axial MRI (b) views of a sinonasal malignancy of the left nasal cavity. The tumour abuts the anterior cranial base.

in the sphenoid sinus, especially when the bone is eroded by tumour. The presence of an Onodi cell (posterior ethmoid pneumatization) increases the risk of optic nerve injury.

The risk of a cerebrospinal fluid leak is greatest along the anterior cranial base in the region of the cribriform plate. This may be due to fracture of the bone from instrumentation or tearing of the dura around the olfactory nerves. The risk of a fracture of the lateral lamella of the cribriform plate is increased in patients with a low cribriform plate on preoperative imaging (Keros classification). Small cerebrospinal fluid leaks can be repaired using small inlay or onlay tissue grafts.

Radiochemotherapy

As discussed previously, RT is an adjunct to most surgery for sinonasal malignancy and is associated with improved disease-free survival. Although intensity-modulated RT is the standard, treatment with heavy particle radiation (proton beam therapy) may be superior for certain tumours such as adenoid cystic carcinoma. Focused stereotactic radiosurgery is generally reserved for focal recurrence or treatment of high-risk margins following conventional RT.

Chemotherapy is also used as an adjunct to surgery and RT. Chemotherapy regimens will vary based on the tumour type. Induction chemotherapy is increasingly used for high-grade sinonasal malignancy (**Box 39.1**). If there is no response to chemotherapy, surgery is performed if the tumour remains operable. A response to chemotherapy is a favourable prognostic indicator and can be followed by surgery or RT. Good results have been observed with SNUC and neuroendocrine carcinoma with induction chemotherapy and consolidation radiochemotherapy. Surgery is reserved for salvage of residual/recurrent tumour.

Tips and pearls

- Establish a proper diagnosis, especially when the pathology report does not match the biological behaviour of the tumour. Be wary of 'small blue cell' look-alikes.
- Have a strategy for haemostasis that includes preoperative embolization, positioning, haemostatic tools and techniques, and warm saline irrigation.
- Devascularize the tumour before dissection of the tumour by cauterizing or ligating feeding vessels (sphenopalatine, ethmoidal arteries) when possible.
- Debulk the tumour if necessary, to allow early identification of key anatomical landmarks.
- Confirm negative margins with frozen sections; be prepared to combine surgical approaches in order to achieve negative resection margins.
- Choose a treatment algorithm based on the biological behaviour of the tumour.

Controversies/areas of interest for the future

The treatment of sinonasal malignancy is rapidly evolving. Improved diagnosis and classification of tumours promotes standardization of treatment and identifies rare histological types that benefit from new treatments. New molecular markers and genomic analysis offer the potential for new therapeutic approaches using specific chemotherapy and immunotherapy drugs.

New imaging modalities such as ^{64}Cu DOTATATE PET for neuroendocrine tumours offer improved diagnosis and staging as well as a new radiotherapeutic approach. Lutathera (lutetium-177 dotatate) is a radiopharmaceutical that has been successfully used for treatment of gastrointestinal tumours; there is limited experience with olfactory neuroblastoma. The proliferation of proton beam centres has increased the opportunity for proton beam therapy with more effective dosing of tumour margins in critical areas and less dosing of normal tissues.

The biggest paradigm shift for the treatment of sinonasal malignancy is the role of induction chemotherapy. Although there is an inherent treatment bias in the selection of therapy, a growing body of

literature supports the concept of induction therapy as a prognostic marker to guide further therapy.

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40

Surgical facial rejuvenation

Ozcan Cakmak and Mahmoud Daoud

Introduction

Human ageing is defined as a process involving physical, psychological, and social changes. While a youthful appearance is valued in society, an aged appearance can cause the perception of being unattractive, and undesirable.

Ageing changes in the face occur at different rates for each individual.

The theories of facial ageing revolve around changes occurring in the soft tissues, with atrophic laxity leading to tissue descent. Loss of thickness and elasticity, due to loss of collagen and elastin, creates sagging and a relative excess of skin, together with wrinkling associated with a decrease in cross-linking between fibroblasts and collagen fibrils. The attenuation of support of the retaining ligaments with ageing leads to the downward displacement of facial fat compartments into the plane between the superficial and deep facial tissues of the face, and is responsible for many of the stigmata that occur with ageing (**Figure 40.1**).

Concurrent lipoatrophy and selective bone resorption in specific areas of the facial skeleton also contribute to the features of the ageing face, such as flattening or indentation of normally convex contours and volume loss.

The ultimate goal of surgical rejuvenation is improving the undesired deformities of ageing to bring out the beauty in the face that existed during youth. Therefore, the surgeon should understand facial soft tissue anatomy and the changes associated with ageing to achieve successful long-term results.

'Ideal' surgical rejuvenation is only possible when these changes are adequately and harmoniously addressed, as well as attaining a natural appearance and durable results without compromising patient safety. Since each individual has unique facial features, a patient-customized treatment plan is applied according to aesthetic needs and desires.

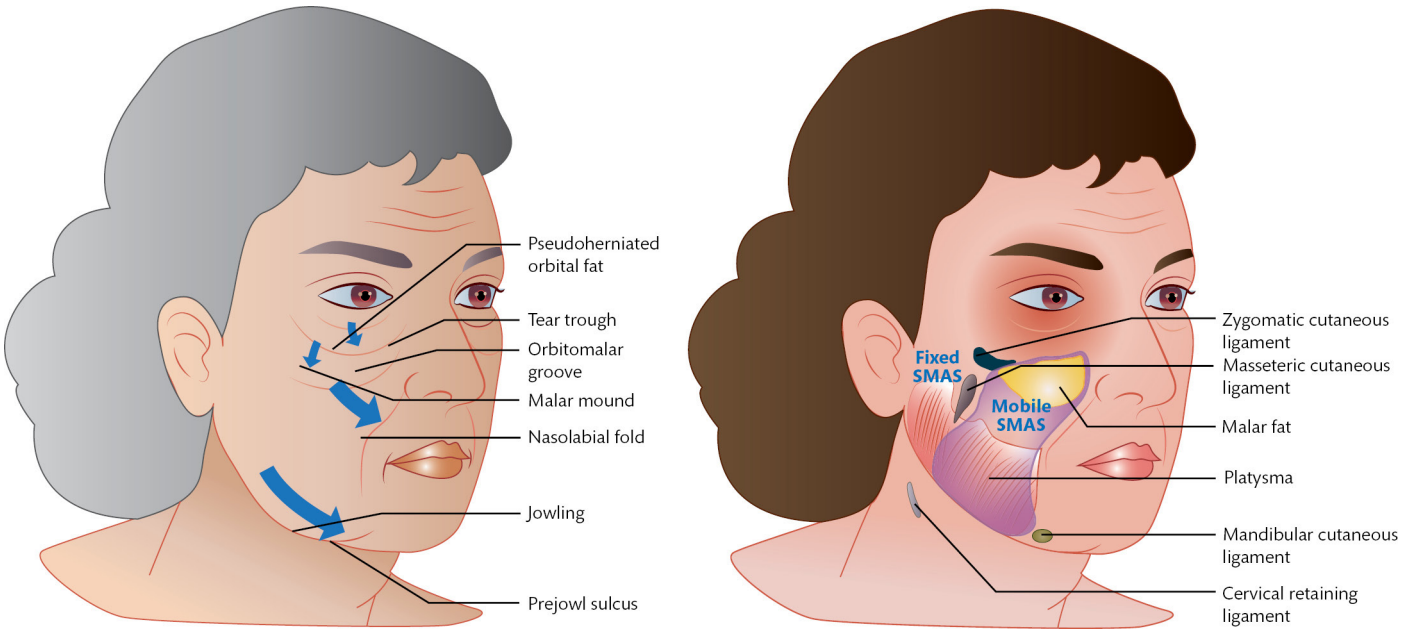

Figure 40.1 Ageing changes, ligaments.

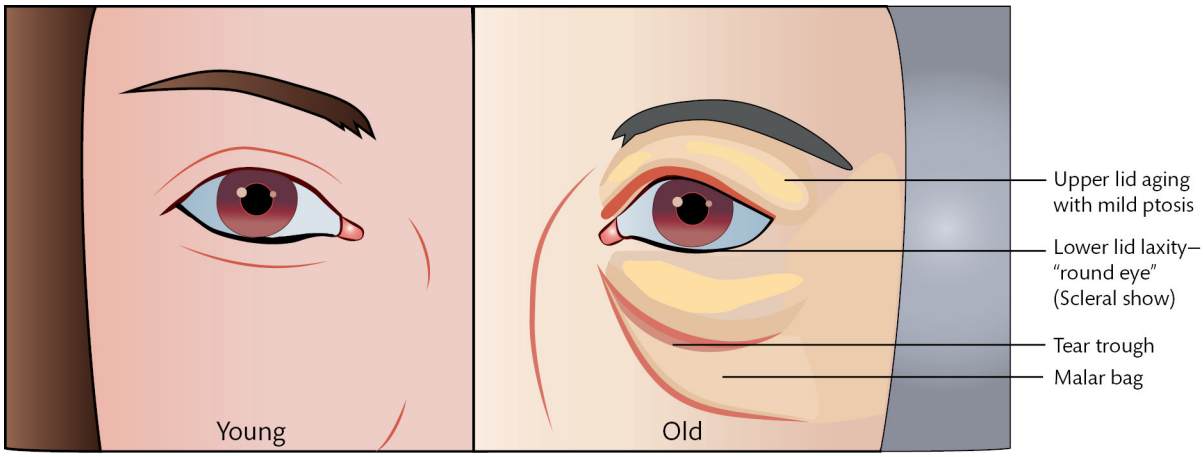

Figure 40.2 Youthful/aged-looking eyebrow complex. Left side: youthful eyebrow complex including higher eyebrow, pretarsal space, and smooth lid–cheek junction. Right side: old-looking eyebrow complex including more puffiness, more wrinkled, tea-trough deformity (puffiness, hollowness below the puffiness), descendent brows, lax lower lid, and obscured upper eyelid crease (lateral hooding).

Upper face

Forehead

The ideal brow shape and position vary depending on gender, age, culture, ethnicity, orbital shape, eye prominence, facial proportions, and current aesthetic trends.

A normal eyebrow sits at or above the superior orbital rim. A youthful female brow arches superiorly at least 1 cm above the supraorbital ridge. Male eyebrows tend to be lower and flatter than female eyebrows and located just over the supraorbital ridge (Figure 40.2).

Ageing changes in the forehead region manifest as ptosis of the eyebrows, the development of dynamic and static rhytids, changes in the hairline, and a concave appearance of the temporal area. The eyebrow complex becomes ptotic, and upper lid skin becomes redundant, which may obscure the supratarsal crease, and lead to temporal hooding.

Patients may chronically elevate their brows to reduce visual field obstruction from excess upper eyelid skin. Rhytids (wrinkles) develop from repeated contraction of mimetic muscles. Horizontal lines on the forehead give an expression of worry or concern; vertical lines give the expression of dissatisfaction or even anger. Typically, patients have more temporal drooping than the medial portion of the brow (Figure 40.2).

Prominent brow ptosis may give the appearance of significant dermatochalasis ('baggy eyes'). If the brow is stabilized and lifted into an appropriate position manually, the clinician can differentiate true dermatochalasis from that confounded by brow ptosis.

Although brow lifting can be performed as a single procedure, it is commonly combined with an upper eyelid blepharoplasty to restore younger-looking periorbital and forehead regions.

Brow lift

The objectives of any brow lifting procedure are to eradicate the depressing effect of the musculature of the glabellar region and to lift up the lateral aspect of the brow.

Several approaches have been utilized in the treatment of brow ptosis. The main techniques used for rejuvenation are the coronal, trichophytic, and endoscopic brow lifts (Figure 40.3).

Coronal brow lift

For many years, the workhorse approach for forehead lifting was the *coronal* approach. This approach is well suited for patients that have significant brow ptosis with a normal to low hairline. The coronal lift is still a reliable method to address this.

The coronal forehead lift is performed with a bevelled incision parallel to the hair shafts, 4–6 cm behind the hairline in a gull-wing fashion extending inferiorly to the temporal sideburn area (Figure 40.3). It allows excellent exposure for precise correction of problems, and long-lasting results without needing overcorrection.

A subgaleal dissection (between the galea aponeurotica and the periosteum) is performed over the frontal bone to develop a coronal flap (Figure 40.3).

In patients with deep forehead creases, frontalis muscle myotomies are performed, remaining medial to the mid-pupillary line to avoid injury to the temporal branch of the facial nerve.

This open-approach permits excellent visualization of the depressor brow muscles. Myotomies of the corrugators, procerus, and depressor supercilii can be performed accurately under direct vision.

Corrugator muscle resection and procerus muscle transections are performed under direct vision with care to preserve branches of the supraorbital and supratrochlear neurovascular structures (Figure 40.3).

Laterally, subgaleal blunt dissection is proceeded above the superficial layer of the deep temporal fascia beneath the temporal branch of the facial nerve. Dissection inferiorly over the zygomatic arch is unnecessary for forehead rejuvenation. It puts the temporal branch of the facial nerve at risk of injury.

After following the dissection to the orbital rims, the flap is completely released from the supraorbital ridge. The flap is advanced superiorly and posteriorly, the excess scalp skin is resected incrementally, and the scalp is closed in layers.

Trichophytic brow lift

Patients with very high foreheads that are considering a brow lift should be offered a trichophytic brow lift. This approach follows a similar dissection to a coronal brow lift without elevating the hairline.

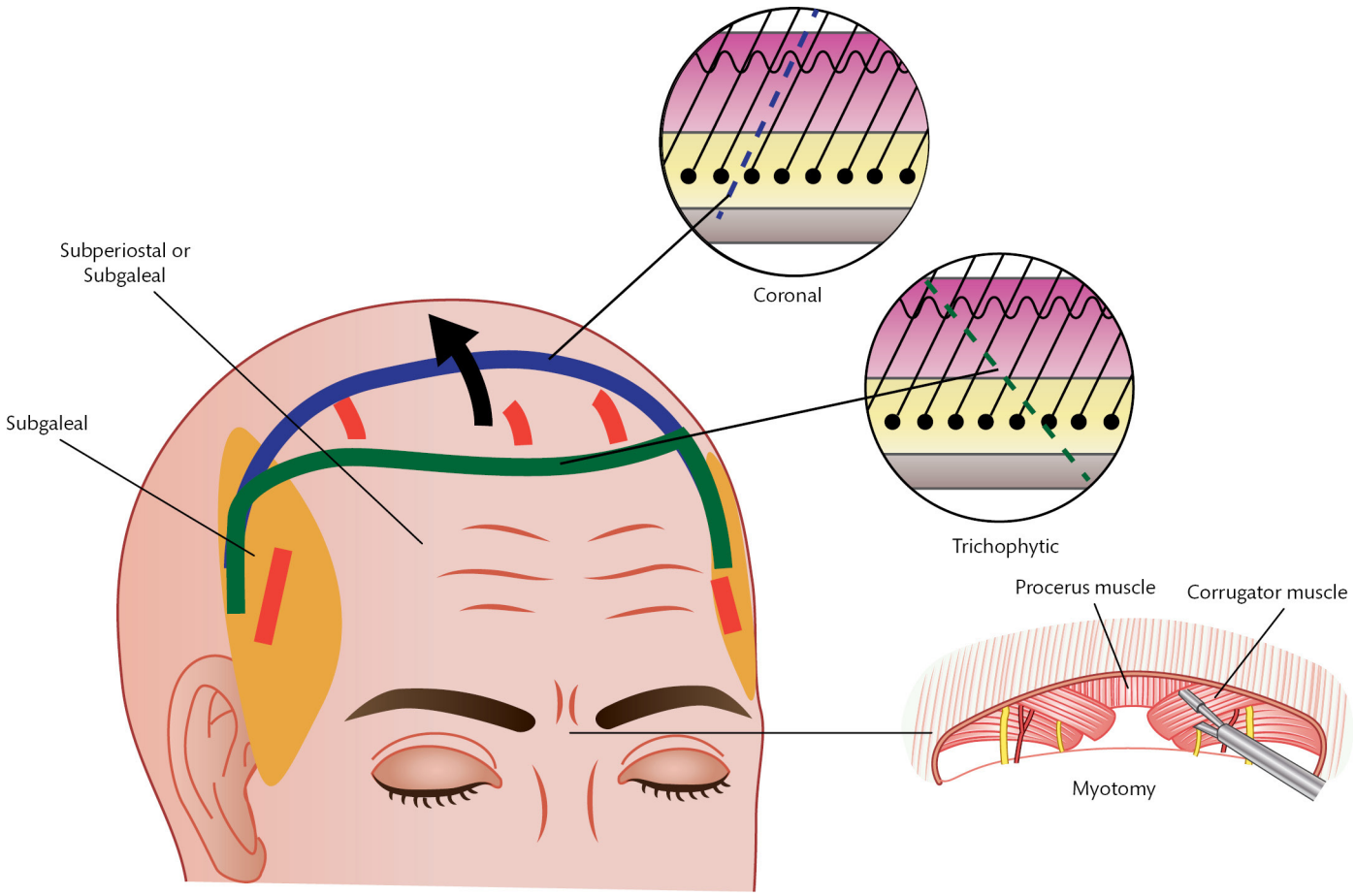

Figure 40.3 Browlift approaches: the coloured lines correspond to trichophytic (green), coronal (blue), and endoscopic (red) approaches.

An irregular-fashion incision, bevelled perpendicular to hair shafts, is placed two to three hair follicles behind the anterior hairline (Figure 40.3). This allows hair growth through the forehead flap from preserved hair follicles and future disguise of the incision.

The incision is extended laterally in the temporal region posterior to the hairline as parallel to hair shafts as possible. The flap is elevated in a similar fashion to the coronal lift with similar options for performing myoplasties if desired.

After removal of excess skin from the forehead flap, it is advanced with a bevel matching the scalp flap.

Endoscopic brow lift

As a more recent technique, endoscopic brow lift is now the favoured approach for most surgeons. Compared to open approaches, having multiple small skin incisions concealed posterior to the hairline reduces the risk of prominent scarring, alopecia, and numbness.

The ideal patient for this approach has a mild to moderate brow ptosis with minimal skin excess, relatively symmetric brows, and a normal to low frontal hairline.

Using a 30° endoscope, structures such as the supraorbital and supratrochlear neurovascular bundles, the sentinel vein, as well as the temporal branch of the facial nerve can be protected.

Five small (each approximately 2–3 cm) incisions are used: three vertical scalp and two temporal incisions. These are a vertical midline incision, placed 1–2 cm posterior to the frontal hairline; two paramedian incisions are placed approximately 5 cm on either side of the midline incision; and the bilateral temporal incisions placed parallel and 1–2 cm posterior to the temporal hairline (Figure 40.3). Temporal dissections are performed in the subgaleal plane (superficial to the superficial layer of the deep temporal fascia).

Inferiorly, the dissection is stopped 1 cm above the zygomatic arch so as not to endanger the temporal branch of the facial nerve. Forehead dissection through the central and paramedian incisions is carried out in a subperiosteal plane, typically in a blunt fashion, to a level 1–2 cm above the supraorbital rim.

Once the initial dissection pockets are created, the surgery continues under the guidance of a 30° endoscope.

The sentinel vein, which is a reliable landmark for the temporal branch of the facial nerve, is identified 1 cm lateral to the zygomaticofrontal suture. Ligation of the vein may increase postoperative oedema. However, if it is necessary, this should be undertaken as deep as possible to avoid injury to the facial nerve.

The lateral (temporal) subgaleal and the central (forehead) subperiosteal pockets are connected by dividing the conjoint tendon at the superior temporal line from a lateral to medial direction. The myotomies are performed through the corrugator and procerus muscles using electrocautery after identification and preservation of the supraorbital neurovascular bundle (Figure 40.3).

The arcus marginalis is completely released in order to achieve adequate brow lift.

The transposed forehead flap is repositioned superiorly via one of the paramedian incisions and is anchored to the cranium using a fixation technique described in the literature, such as microscrew placement in the paramedian incision and fixation with skin staples posterior to the microscrew, or sutures secured through cortical bone tunnels, or absorbable cortical anchors.

The lateral suspension is directed superiorly and posteriorly, and secured with sutures to the deep temporal fascia.

Upper eyelid

The attributes of a youthful upper eyelid area include a partially visible pretarsal skin with a well-defined upper eyelid (supratarsal) crease and smooth transition between the upper eyelid and lateral temporal skin (**Figure 40.2**).

The main anatomical consideration in upper blepharoplasty is the upper eyelid crease, which divides the upper lid into tarsal and orbital portions.

The levator aponeurosis attaches to the anterior surface of the tarsus and sends slips of tissue forward to the skin through orbicularis muscle, which creates an upper eyelid crease. Attachment position of the levator aponeurosis to the anterior surface of the tarsus determines its location.

A youthful eyelid crease is usually located 8–10 mm above the lid margin in non-Asian women whereas in men, the distance is shorter, measuring 6–8 mm.

With advancing age, upper lid skin becomes redundant (dermatochalasis), visibility of the upper eyelid may be effaced ('hooding') due to skin laxity and/or brow ptosis, and lateral hooding may obstruct the visual field. Also, fat may protrude above the palpebral fold due to weakening of the orbital septum combined with bone resorption at the superomedial orbital rim, or there may be loss of fat from the upper eyelid sulcus.

Other findings of ageing, such as eyelid ptosis by dehiscence or weakening of the levator aponeurosis and lateral canthi descent, may contribute to a sad-eyed look (**Figure 40.2**).

The upper eyelid examination usually focuses on the level of brow ptosis, dermatochalasis, and extent of lateral hooding.

The surgeon should always assess brow position before attempting blepharoplasty: a brow lifting procedure may obviate the need for blepharoplasty (**Figure 40.2**). The brow should be elevated manually so that the patient may fully understand its contribution to upper lid dermatochalasis.

If a forehead lift is indicated at the same time, the brow lift is performed first before an upper lid blepharoplasty, as the amount of upper lid redundancy is significantly reduced after correction of the brow position. This minimizes the risk of aggressive removal of redundant skin which may cause lagophthalmos, and also permits for a more accurate upper lid blepharoplasty.

Evidence of other conditions, such as periorbital hollowing and eyelid ptosis (blepharoptosis), should also be excluded to be able to reproduce as natural a facial ideal of youth as is possible.

Upper blepharoplasty

Upper blepharoplasty is a procedure performed to restore a youthful, attractive upper eye appearance involving one or a combination of resection of excess skin, removal of bulging fat and muscle, aesthetic placement of the supratarsal crease, and filling/contouring of a deep upper orbital sulcus with fat injection or transfer.

It is essential to avoid over-resection of skin, muscle, and fat, which can distort normal anatomy, ocular function, and interferes with closure of the eyelids as well as creating an unnatural hollowed appearance.

The design of the upper lid skin incision is critical to avoid postoperative lagophthalmos, particularly when an upper lid blepharoplasty is combined with a brow lift.

The upper mark of the incision is determined when the patient is in the upright sitting position and by asking the patient to look straight ahead to help factor the role of gravity on eyelid and brow position (**Figure 40.4**).

The upper mark should be at least 10 mm from the inferior border of the brow so as not to include any brow skin, which could pull the brow down.

The lower mark is placed below the supratarsal crease, which avoids injury to the levator insertion and helps to create a well-defined upper lid crease.

The distance from the lid margin to the lid crease at the mid-pupil level should be 8–9 mm in women, and it should be placed closer (7–8 mm) in men to avoid a 'feminized' appearance. The marking follows the natural curve of the upper lid, and the distance from the lid margin to both the medial and the lateral canthi should be approximately 4 mm.

Medially, the upper eyelid crease marking is tapered slightly upward and terminated 2–3 mm before the medial canthus since extending to the orbital nasal depression may lead to an unsightly webbed scar.

Laterally, at the level of lateral canthus, the skin marking is curved superiorly towards the orbital rim. This allows maximal skin removal to eliminate lateral hooding without pulling down the upper eyelid–eyebrow complex (**Figure 40.4**).

During upper eyelid surgery in men, the lateral extension of the incision must be shorter because the resulting scar may not be covered with makeup, and scarring also tends to be more visible.

After skin incision and subcutaneous dissection, marked skin is removed.

The orbicularis muscle is dissected to reach the fat pads.

If definition of the supratarsal fold is desired, a small strip of preseptal orbicularis muscle is resected.

Attention is then directed to the removal of pseudoherniated fats The orbital septum is divided to access the nasal and central fat compartments. The globe can be gently pushed into the orbit for identification of fat pads.

The nasal fat pad is denser and paler than the central fat pad. It is critical to avoid injury to the superior oblique tendon and trochlea, which separate the fat pads of the upper lid (similar to how the inferior oblique muscle separates the nasal and central fat pads of the lower lid). The bulging fat is gently teased out, cauterized with bipolar diathermy, and resected (**Figure 40.5**).

Excess traction/manipulation of orbital fat may result in avulsion of deep orbital vessels and should be avoided.

After judicious haemostasis is attained, the skin is closed using 6.0 sutures.

Lower eyelid

A young-looking lower eyelid has a characteristic of smooth contours blending with the overall facial shape without any redundancy, bags, or depressions (**Figure 40.2**).

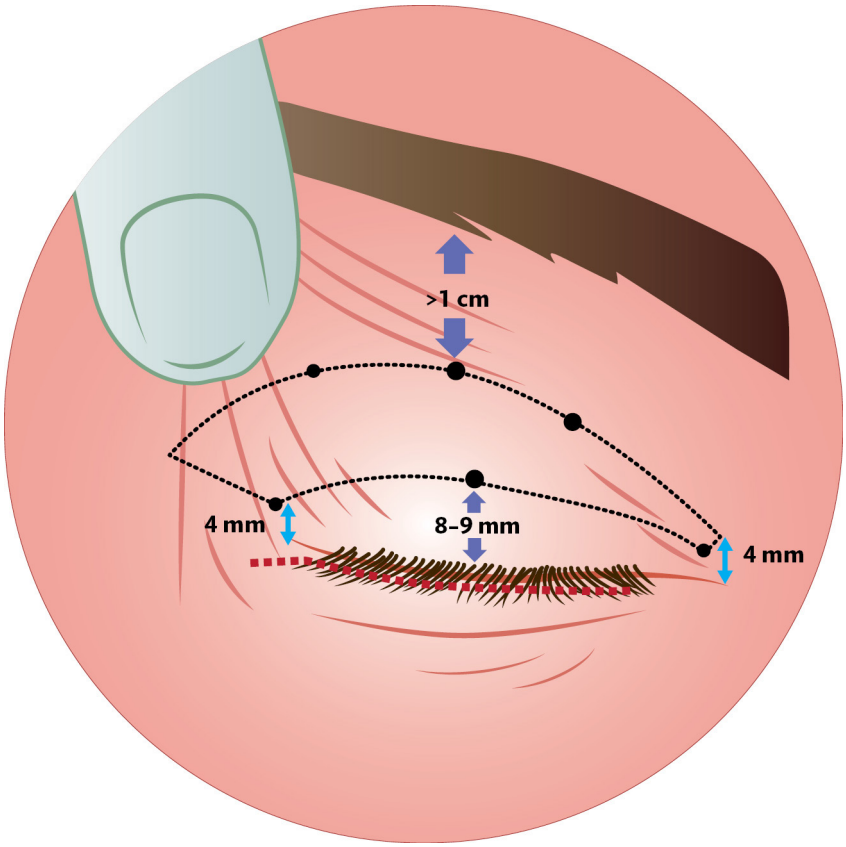

Figure 40.4 Blepharoplasty incisions.

Age-related changes in the lower eyelid area are multifactorial and include the result of cumulative changes in skin texture, loss of elasticity, attenuation of retaining ligaments, volume depletion due to lipoatrophy/maxillary resorption, and descent of the malar fat pad (Box 40.1).

Lower eyelid laxity must be addressed with *distraction* and *snap* tests to maintain eyelid function, appearance, and to prevent postoperative eyelid malposition (Figure 40.6).

The lid distraction test is performed by grasping the lower lid with the thumb and index finger and displacing anteriorly. If the lid is distracted more than 8 mm from the globe, the lid is lax.

The snap test is performed by pulling the eyelid inferiorly to the level of the inferior orbital margin, then releasing it. Typically, within 1–2 seconds the eyelid should snap back to its normal position. If there is a relative delay or failure to regain its normal anatomical position, the lid is lax.

When lid laxity is noted, lateral canthopexy or canthoplasty should be performed as part of the blepharoplasty procedure to prevent eyelid malposition.

Lower eyelid blepharoplasty

In the past, blepharoplasty was an operation to remove 'excess' fat, skin, and muscle that often led to a hollow appearance and lower eyelid malposition. Nowadays, lower blepharoplasty has evolved significantly to achieve fuller, natural, and unoperated-looking eyelids with a gradual blending of the lower lid–cheek junction and overall facial shape (Figure 40.7, Box 40.2).

There are mainly two approaches to treating the lower eyelid. Each has its advantages and disadvantages.

Transconjunctival approach

The traditional transcutaneous approach disrupts the orbicularis muscle and the orbital septum that may lead to lower eyelid malposition and rounding of the canthal angle. Though the evolution of additional canthal suspension techniques has reduced the occurrence of these complications, they can still occur at an unacceptably high rate.

The transconjunctival approach (also referred to as muscle-sparing lower blepharoplasty) is a relatively new technique that provides immediate access to the deep layers avoiding the division/denervation of the orbicularis oculi muscle (Figure 40.8).

Besides the proposed benefits of minimizing complications, such as scleral show and ectropion, this approach also eliminates a visible scar related to an external incision.

The transconjunctival approach is ideal for young patients that have pseudoherniated fat without excess skin. It may be combined with adjunctive techniques, such as skin-pinch excision, laser resurfacing, or a chemical peel to address associated minimal skin laxity.

Transconjunctival blepharoplasty can be performed in either the preseptal (requiring septal division to access fat) or postseptal (direct approach to fat with preservation of orbital septum) planes (Figure 40.8).

The lower lid is inferiorly retracted with simultaneous globe retropulsion to expose the conjunctiva.

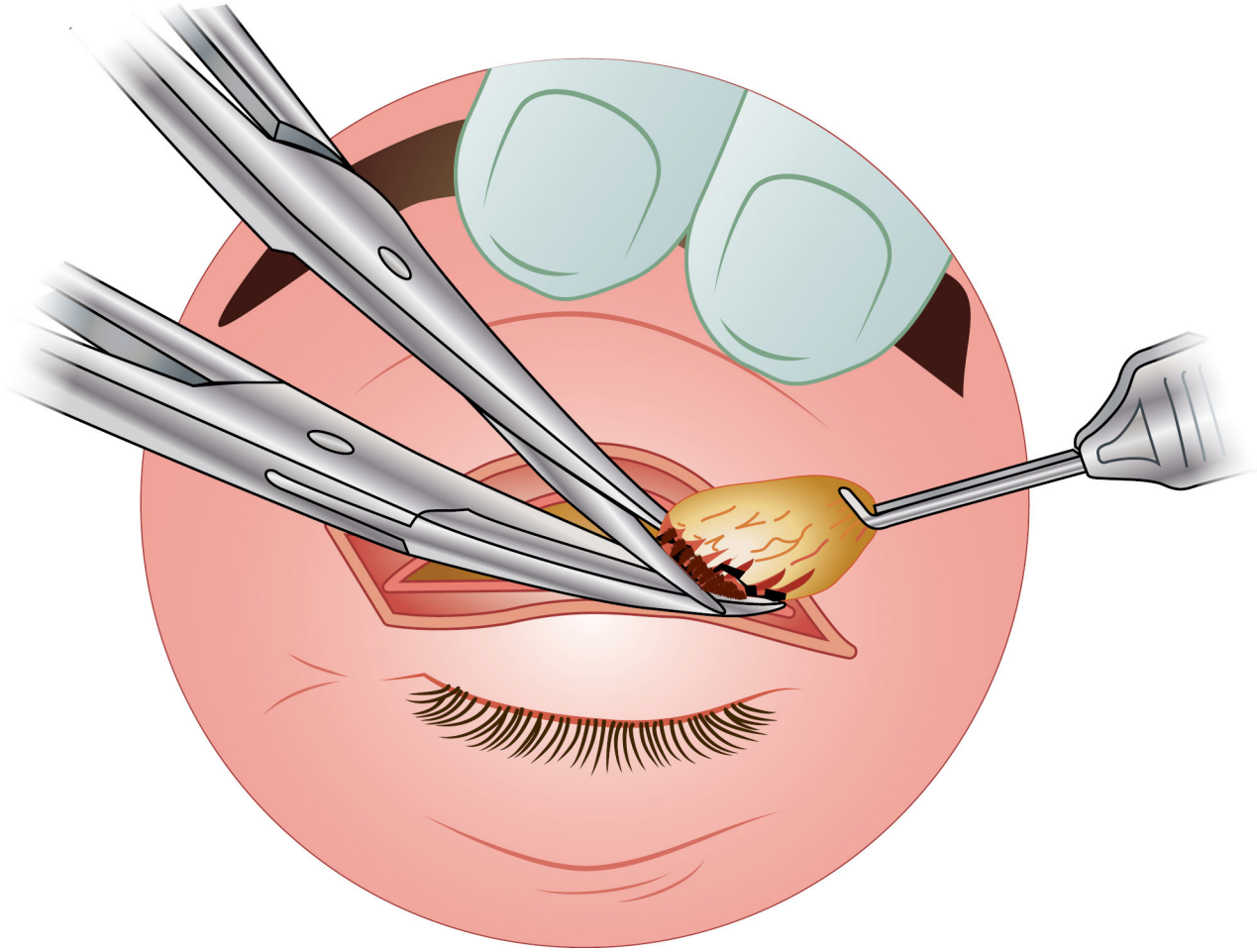

Figure 40.5 Orbital fat removal. Note: the dark line at to the bottom of the fat (just superior to the edge of clamp) is to indicate that this area has been cauterized before trimming to avoid bleeding.

Box 40.1 Characteristics of ageing changes

- Dermatochalasis.
- Pseudoherniation of orbital fat.
- Orbital hollow.
- Depression caudal to orbital fat (tear-trough deformity).
- Horizontal lower eyelid laxity.
- Scleral show.
- Cascades of lax skin and orbicularis muscle over the malar eminence (festoon).
- Chronic soft tissue swelling over the malar eminence (malar mound).
- Descent of the lid–cheek junction with a distinct demarcation line (Figures 40.1 and 40.2).

For the preseptal approach, an incision is made just below the tarsus (seen as a greyish edge containing a vascular arcade) (Figure 40.8). Following blunt dissection between the orbital septum and orbicularis oculi muscle, the orbital septum is divided to access fat.

For the postseptal approach, an incision is made 4–5 mm posterior to the tarsus (halfway between the lower edge of the tarsus and the fornix) through the conjunctiva and the lower eyelid retractors (Figure 40.8).

The postseptal approach allows direct entry to fat while the septum is left intact.

Although all three fat compartments are exposed, the inferior oblique muscle between the medial and central fat pads is protected. The lateral fat pad is commonly encased in a dense fibrous network. Prolapsing fat is either cauterized and removed or transposed into a subperiosteal or supraperiosteal plane to improve any tear trough deformity.

Over resection of fat should be avoided to prevent hollowness of the lower eyelids.

After meticulous haemostasis the conjunctiva and lower eyelid retractors are returned to their normal anatomic position, and the conjunctival wound is allowed to heal by secondary intention.

Transcutaneous (subciliary) approach

The main advantage of a transcutaneous approach is to allow management of excess skin and orbicularis muscle that requires redraping for adequate lid recontouring. It enables broad exposure

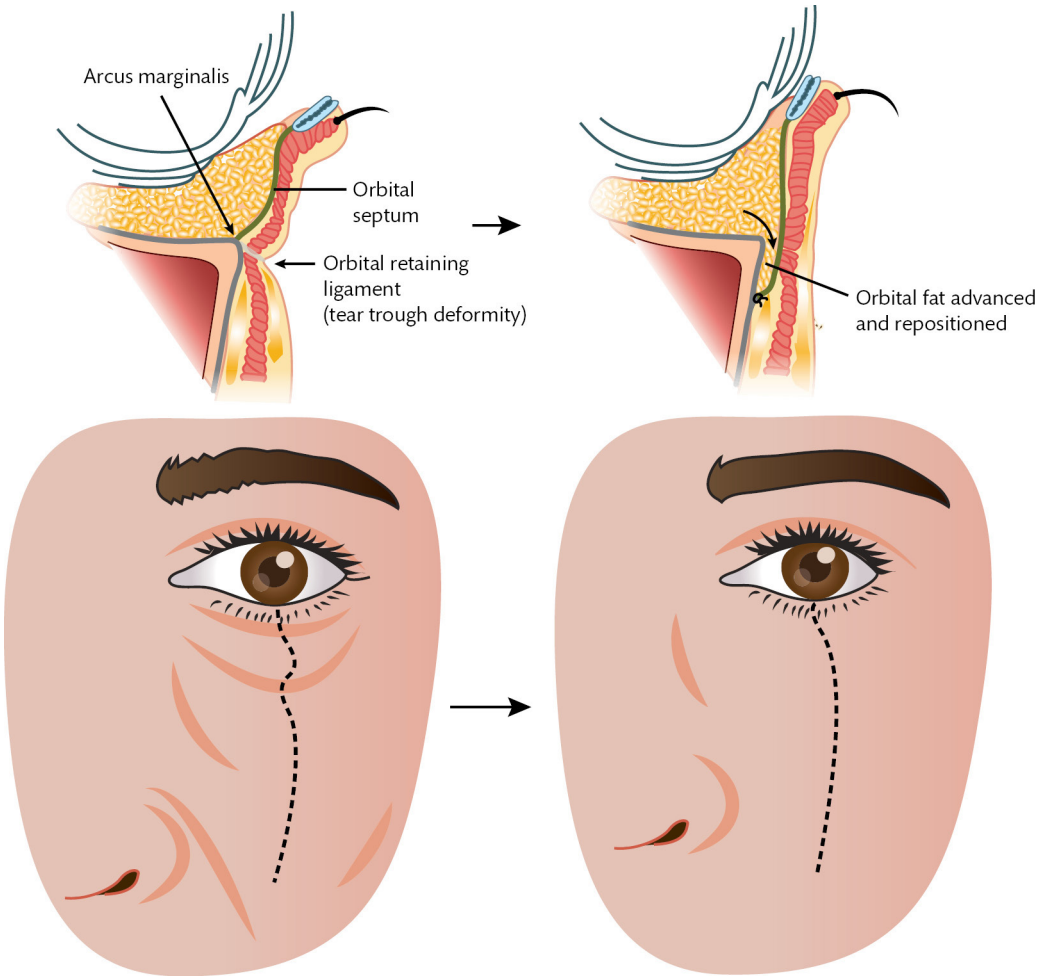

Figure 40.6 Fat transposition (repositioning).

to extend the dissection for a wide release of retaining ligaments and precise correction of problems such as inferior rim hollowing, malar mounds, and festoons (Figures 40.1 and 40.2).

Due to damage to and denervation of orbicularis muscle and septal (middle lamella) scar contraction—with or without the combination of excess skin and/or muscle excision—the transcutaneous approach involves a high risk of:

- lower eyelid malposition
- lower eyelid retraction
- lateral rounding
- scleral show
- ectropion.

This approach requires prophylactic lower-lid reinforcing manoeuvres, such as orbicularis muscle suspension, lateral canthopexy, or canthoplasty for establishing/maintaining lower lid support, creating appropriate postoperative contour/position, and minimizing the risk of lid malposition.

The skin is incised 1 mm below the lash line starting medially from 2–3 mm lateral to the punctum and ending laterally at 1–1.5 cm lateral to the lateral canthus (Figures 40.4 and 40.8).

First, a skin flap is elevated inferiorly and a stairstep incision is made in the orbicularis muscle to preserve an intact pretarsal sling of about 5 mm (Figure 40.8).

Then, a composite skin–muscle flap is elevated up to the orbital rim.

Nowadays, repositioning—rather than removing—the pseudoherniated fat beyond the infraorbital rim is preferred in order to blend the lower eyelid–cheek junction and prevent hollowness.

The orbital retaining ligaments are released beyond the inferiormost aspect of the marked tear trough, and a pocket is created inferior to the orbital rim either in the supraperiosteal or subperiostal plane.

The orbital septum is next opened, and the prolapsing nasal and central fat pads are advanced and repositioned into the pocket to camouflage the lower orbital rim depression and smooth the lower eyelid–cheek contour (Figure 40.7).

Repositioned fat is fixed either internally, to the underlying periosteum with absorbable buried sutures, or externally, to the skin with percutaneous sutures which are removed a few days after surgery.

The pseudoherniated lateral fat pad is removed. If there is lower lid laxity, lateral canthopexy or lateral canthoplasty are performed.

After meticulous haemostasis, the skin–muscle flap is redraped superolaterally and the redundant skin and orbicularis muscle are very conservatively removed so as not to cause postoperative ectropion or lower eyelid malposition.

An orbicularis muscle suspension suture is placed between the superior cuff of the orbicularis muscle edge of the lower eyelid flap and the lateral orbital rim periosteum to reinforce the lower eyelid

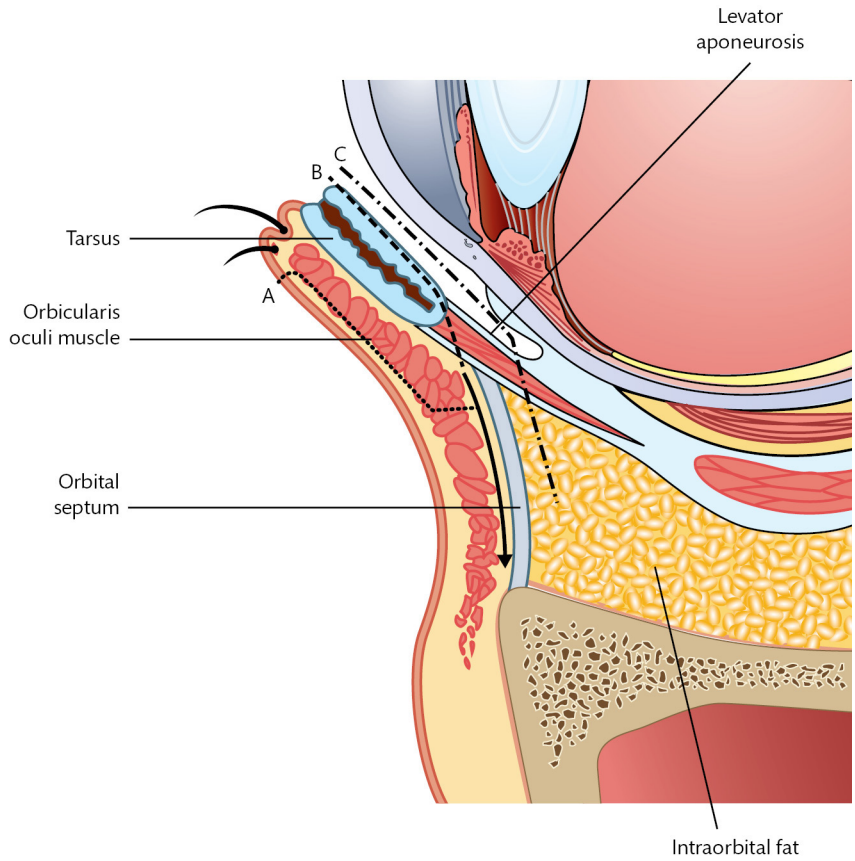

Figure 40.7 Blepharoplasty approaches. (a) Subciliary incision. (b) Transconjuctival preseptal incision. (c) Transconjuctival postseptal incision.

Box 40.2 Contemporary lower eyelid rejuvenation

The goal of contemporary lower eyelid rejuvenation is to recreate the characteristics of a youthful eyelid by:

- re-establishing a smooth contour at the lower eyelid–cheek junction
- eliminating visible fat
- effacing depressions inherent to their transition
- achieving even, wrinkle-free skin.

This is while maintaining/restoring normal lower lid support, position, and function.

support, to help to blend the lid–cheek junction, and also to reduce the risk of postoperative eyelid malposition.

After final trimming of excess skin, the incision is closed with 6.0 sutures.

Lower face and neck

A youthful face typically has a smooth blending of contours between the aesthetic subunits of the face with full and well-supported fat. With ageing, the attenuation of ligamentous support and the deflation of facial fat leads to platysma, fat, and skin descent (Figure 40.1).

Volume deflation and facial fat descent may cause demarcation lines with facial fat volumetrically becoming situated anteriorly and inferiorly. These changes contribute to the characteristic signs of an older-looking face with:

- jowls
- loss of definition of the jaw line
- marionette lines
- upper lip lengthening (Figure 40.1).

Similarly, accumulation of submental fat, platysmal banding, diastasis of medial platysmal edges, pseudoherniation of subplatysmal fat, loss of the cervicomental angle, and drooping of redundant skin lead to a characteristic, old-looking neck appearance (Figure 40.1).

The goal of facelifting is to create a harmonious, natural, and younger-looking face. Understanding the anatomy of the SMAS (superficial musculoaponeurotic system), facial nerve, and retaining ligaments is of utmost importance for adequate and safe release of descended tissues, producing the most natural result.

The SMAS is an extension of the superficial cervical fascia cephalad into the face and forms a continuous layer throughout the head and neck to form a single anatomic and functional unit that gives the human face its unique mobility (Figure 40.9).

The SMAS continues as the frontal muscle at the forehead; temporoparietal fascia at the temporal region; platysma at the lower face and neck; firmly attaches to the zygomatic arch; and invests the superficial mimetic muscles at the cheek (Figure 40.9).

Similarly, the corresponding deeper layer of cervical fascia (the superficial layer of the deep cervical fascia) continues into the face and has been termed the parotidomasseteric fascia or deep facial fascia (Figure 40.9).

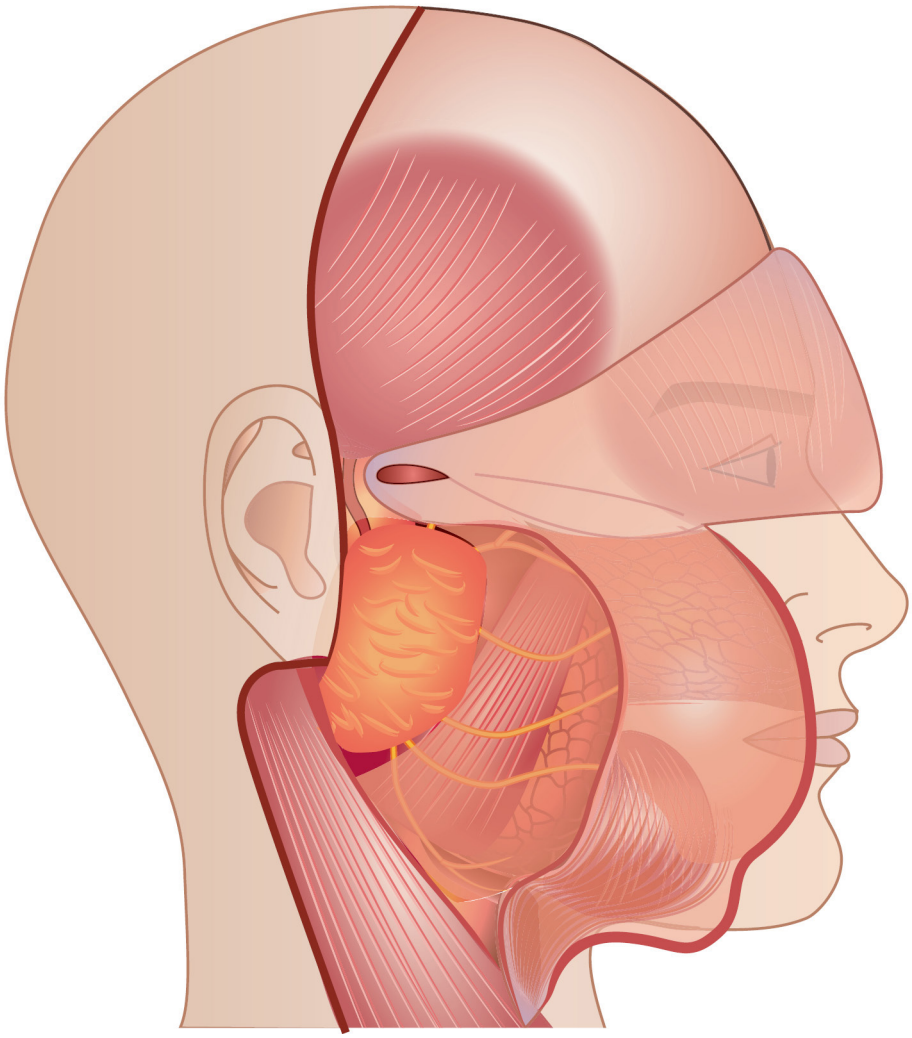

Figure 40.8 SMAS, deep fascia, facial nerve.

As a discrete fascial layer, the SMAS separates overlying subcutaneous fat from the underlying parotidomasseteric fascia and facial nerve branches. Since the facial nerve branches travel beneath the parotidomasseteric fascia for a variable distance after exiting the parotid gland, maintaining the integrity of parotidomasseteric fascia during sub-SMAS dissection protects the facial nerve from injury (Figure 40.9).

The malar fat pad is subcutaneous fat superficial to the SMAS layer investing zygomaticus major, zygomaticus minor, and levator labii muscles. Retaining ligaments anchor the dermis, subcutaneous fat, and SMAS to the bony periosteum or deep fascia.

The zygomatic cutaneous, mandibular cutaneous, and orbitomalar ligaments are examples of osteocutaneous ligaments that originate from the periosteum and insert directly into the dermis.

The masseteric cutaneous and parotid cutaneous ligaments are formed as condensations between the superficial and deep fascia. The attenuation of support of these ligaments with ageing leads to downward displacement of facial fat compartments into the plane between the superficial and deep tissues of the face responsible for many of the stigmata that occur with ageing (Figure 40.1).

Attenuation of zygomatic ligaments leads to descent of the malar fat inferomedially along with the midface and accentuates the nasolabial folds. Weakening of masseteric ligaments attributes to jowl formation. Jowling occurs posterior to the mandibular ligament (Figure 40.1).

Facelift

Facelift can be defined as applying lifting forces in the opposite direction to ageing vectors for the adjustment of changes caused by ageing. Since the advent of facelift surgery, there has been a continuing evolution of techniques. There are now mainly three facelift techniques concerned with the depth and extent of dissection.

Subcutaneous (skin-only) lift

The subcutaneous facelift involves dissecting a variably sized skin flap, excising redundant skin, and closing the incisions. Since increased skin redundancy is not the only factor causing ageing, the result tends to be unnatural and short-lasting without any additional repositioning of deeper tissues. Furthermore, the skin flap is placed under tension when closing, which may lead to excessive scarring at the incision line.

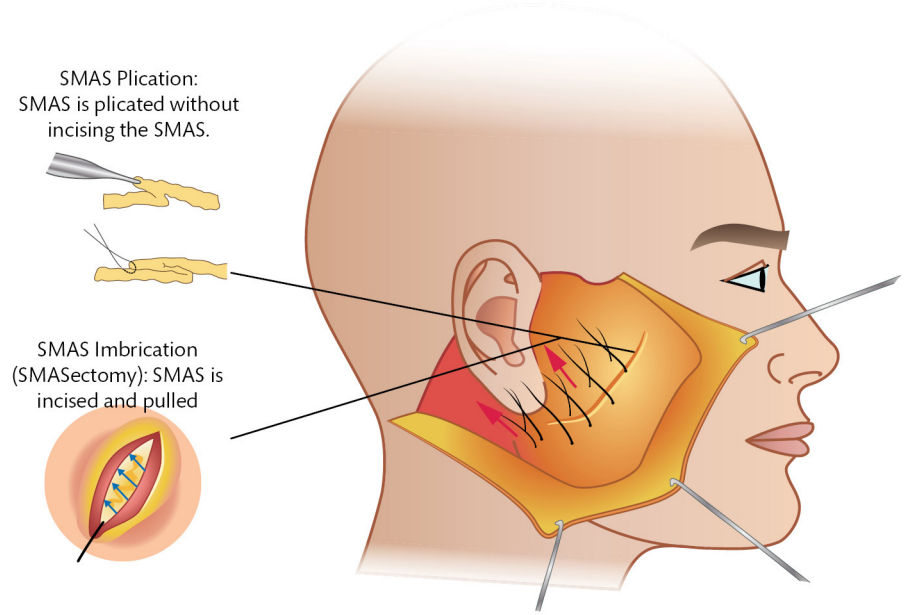

Figure 40.9 Facelift incision, and SMAS techniques.

This technique is typically used in very select situations where skin laxity is the only issue.

SMAS lift techniques

Lifting and suspending the SMAS in vector (superior-lateral direction) opposite to the ageing vectors can be applied by different methods.

Imbrication and plication of the SMAS are the most common facelift techniques.

Plication involves in-folding of the SMAS and suture suspension without any SMAS incision (Figure 40.10). The SMAS of the lower face is pulled, plicated, and anchored to the more immobile SMAS overlying the parotid (Figure 40.10).

Imbrication techniques involve a SMAS incision and a portion of SMAS is either removed or transposed with or without limited sub-SMAS dissection (Figure 40.10).

Ageing changes in the lower face and neck may be successfully treated with a SMAS-lift technique. However, there is no lifting effect at the midface and nasolabial fold, since the retaining ligaments in the midface are not released.

Extended facelift techniques

Extended facelift techniques involve surgical release of retaining ligaments in the midface (zygomatico-cutaneous and masseterico-cutaneous ligaments) that prevent the transmission of traction to the malar portion of the facelift dissection. SMAS dissection is advanced

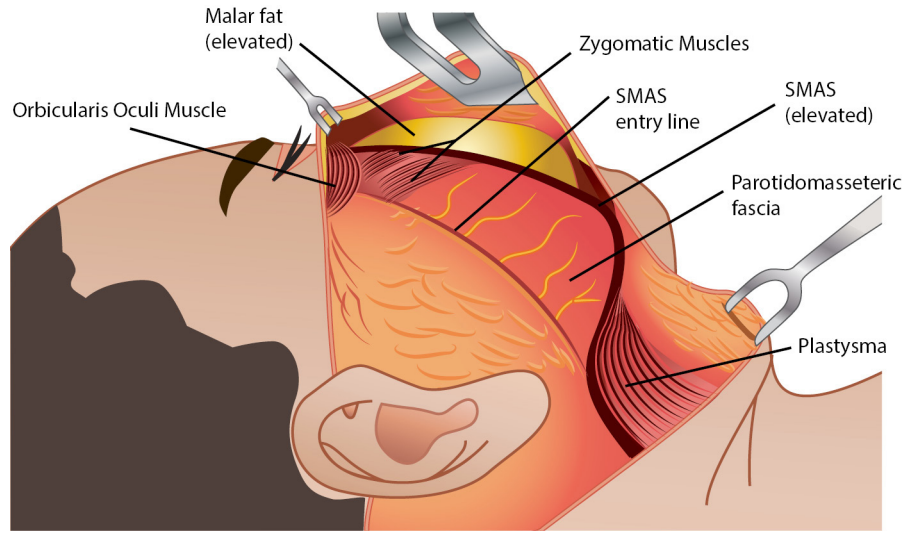

Figure 40.10 Deep plane facelift.

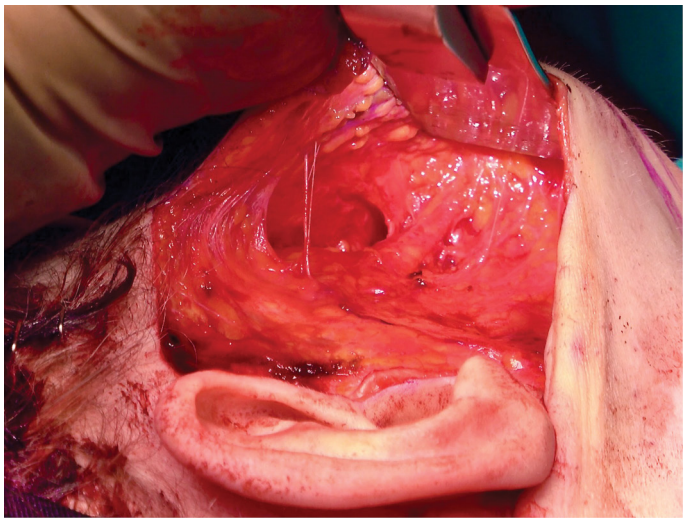

Figure 40.11 Intraoperative photograph demonstrating an extended facelift dissection towards the midface along the superficial surface of the zygomaticus major muscle.

towards the midface along the superficial surface of the zygomaticus muscles to allow repositioning of the ptotic malar fat, which diminishes the nasolabial folds (Figure 40.11).

Extended facelifts produce combined, balanced, and harmonious rejuvenation of the midface, cheek, and lower face without requiring a separate midface lift procedure.

There are different techniques with similar extended midface dissection, each with some variation:

- An extended SMAS technique involves a long skin flap and a distinct SMAS flap dissected and pulled separately.
- A high SMAS technique involves a similar dissection but utilizes a higher SMAS flap along the superior border of the zygomatic arch.
- A deep plane facelift involves undermining of a skin–SMAS flap as a single unit following a more limited subcutaneous dissection (Figure 40.11).
- A composite plane facelift, in addition to the deep plane facelift dissection, the lower part of the orbicularis oculi muscle is also dissected and included in the flap.

Since deep and composite flap facelifts involve a single unit, they allow excellent blood supply to overlying skin which is important, especially in smokers, to prevent vascular compromise and subsequent skin necrosis.

A facelift incision begins horizontally below the temporal hairline, then carries on inferiorly in a native preauricular crease (Figure 40.10).

The incision follows a post-tragal course in females, and a pretragal course in males so as not to move bearded skin into the external ear canal. It continues around the ear lobule and is carried on superiorly onto the posterior surface of the concha. It is then curved posteriorly and inferiorly at the level of the helical insertion into or just anterior to the hair-bearing portion of the scalp behind the ear (Figure 40.10).

After wide skin undermining, mobile medial SMAS is pulled and fixed to the relatively immobile lateral SMAS overlying the parotidomasseteric fascia.

If additional midface rejuvenation is required, zygomaticocutaneous and masseterico-cutaneous ligaments are released and SMAS dissection is advanced towards the midface along the superficial surface of the zygomaticus muscles to allow repositioning of the ptotic malar fat, thus diminishing the nasolabial folds.

In order to obtain a tighter neck, a sub-SMAS dissection is extended inferiorly below the angulus mandible, and the cervical retaining ligaments (between sternocleidomastoid muscle and platysma) are released. The SMAS flap of the face and neck is pulled and suspended in a direction opposite to the ageing vectors.

If necessary, concurrent liposuction is performed to remove excess subcutaneous fat.

If there are platysma bands, dehiscence, and redundancy, a corset platysmaplasty is performed through a horizontal submental incision.

The central components of the deeper neck layers (i.e. subplatysmal fat, anterior digastric muscles, and submandibular glands) may also be reshaped to obtain a flat submental contour especially in patients with a heavy neck.

Preservation of the facial nerve is crucial in facelift. There are certain danger areas during dissection that can compromise the nerve. These include the areas where the nerve branches become more superficial in the dissection plane, and where they traverse between the retaining ligaments of the face. Due to risk of facial nerve injury much care is needed when attempting to release these ligaments. Many facelift surgeons are rightly hesitant in transecting these ligaments during extended facelift dissections. However, most recent comprehensive systematic reviews and meta-analysis have shown the opposite: extended facelift techniques which identify and preserve the anatomy often lead to lower complication rates than the techniques requiring working blindly around vital structures.

Conclusion

The creation of the American Academy of Facial Plastic and Reconstructive Surgery (AAFPRS) in 1964 officially marked the beginning of modern facial plastic surgery as a subspecialty of otolaryngology.

For those interested in rejuvenation of the ageing face, we recommend finding a mentor early in your training/residency, who can subsequently help and guide you through your training.

We also recommend attending courses, seminars, and virtual webinars to improve both theoretical and practical skills and knowledge.

After finishing your otolaryngology training, we strongly recommend completing a facial plastic surgery fellowship. Various organizations offer fellowships. We advocate the fellowship of the European Academy of Facial Plastic Surgery (EAFPS).

After completing the fellowship, you can apply to the European Board for certification in Facial Plastic and Reconstructive Surgery (EBFPRS).

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41

Non-surgical facial rejuvenation

Alwyn R. D'Souza and Chew Lip Ng

Introduction

The outward manifestations of facial ageing are a result of three-dimensional changes in the craniofacial skeleton, as well as skin and soft tissues changes influenced by extrinsic and intrinsic factors.[1] Extrinsic factors are primarily sun exposure leading to cutaneous photoageing, and smoking leading to skin ageing. Intrinsic factors contributing to skin ageing are shown in **Box 41.1**.

Skeletal remodelling leads to a decrease in height and prominence of the maxilla, widening of the orbits, and reduction in mandibular volume and prominence. The loss of bony support causes soft tissues to descend leading to mid-cheek droop, jowl formation, orbital hollowing, and appearance of eye bags.

A wide range of non-surgical options for facial rejuvenation have been developed to address facial ageing (**Table 41.1**). They include botulinum neurotoxins (BoNT), fillers, chemical peels, and energy devices such as lasers and radiofrequency devices.[1] They target different facial structures and are frequently employed in combination. **Figure 41.1** demonstrates the causes and features of facial ageing, and the target of interventions described in this chapter. **Figure 41.2** demonstrates the effects of ageing.

Botulinum toxin

BoNTs are protein toxins derived from anaerobic bacteria of the genus *Clostridium*, such as *Clostridium botulinum*.[1] There are seven serotypes (A–G), of which type A is most commonly used in clinical practice. BoNT inhibits the release of acetylcholine from presynaptic neurons, leading to flaccid muscular paralysis and reduction in glandular secretion.[2] The effects of BoNT injections last from 3 to 12 months.

Box 41.1 Intrinsic factors contributing to skin ageing

- Skin atrophy, leading to thinner, dryer skin prone to rhytid formation.
- Fat atrophy, leading to devolumization.
- Laxity of ligaments and fascia, leading to soft tissue descent.
- Bony resorption and remodelling of the craniofacial skeleton.

The applications of BoNT in facial rejuvenation are as follows[1,2]:

Reduction of facial rhytids

Mimetic facial muscles cause skin wrinkling and folding when they contract. They are a hallmark feature of ageing. Rhytids can be classified into dynamic and static rhytids. Dynamic rhytids are seen only during active muscular contractions and not during muscular relaxation while static rhytids, which are seen as a progression from dynamic rhytids, are seen even during muscular relaxation. The hypothesized effects of BoNT on dynamic rhytids are shown in **Box 41.2**. BoNT injections may lead to a reduction in dynamic rhytids and delay their progression into static rhytids.

Common areas treated, shown in **Figure 41.2**, are:

- forehead horizontal rhytids formed by contractions of the vertically oriented frontalis muscles
- glabellar rhytids or 'frown lines' formed by contractions of the corrugator supercilii, depressor supercilii, and procerus muscles
- lateral orbital rhytids or 'crow's feet' formed by contractions of the orbicularis oculi
- perioral rhytids formed by contractions of the orbicularis oris muscle.

Brow lift

Brow ptosis is common as the face ages. Brow position is controlled by opposing muscles that elevate and depress the brow. Injecting BoNT into brow depressors, primarily the procerus and corrugator supercilii, leads to their weakening and the consequent elevation of the brow. Conversely, when injecting the frontalis (the major brow elevator) for horizontal rhytids, one must be careful not to overweaken the muscle which can lead to the undesirable consequence of brow ptosis. **Table 41.2** shows the elevator and depressor facial muscles.

Reduction of muscle bulk

Hypertrophic muscles can lead to facial contours that are considered non-aesthetic. A prime example is masseter hypertrophy, which can occur in conditions like bruxism. A bulky masseter leads to a strong and prominent angle of the jaw, rendering the appearance of a 'square-face'. This can be an attractive masculine feature in males but

Table 41.1 Options for facial rejuvenation

Signs of ageing	Tissue components involved	Rejuvenation options
Superficial fine lines Pigmentary changes Telangiectasia	Dermal	Carbon dioxide laser, pigmentary laser, chemical peel, intense pulsed light, sun protection, moisturization, and other skin care products
Rhytids (dynamic and static) Devolumization and soft tissue descent	Deep dermal, subcutaneous, and deeper soft tissues, ligaments, and fat Bone	Botulinum toxin, fillers
Deep folds (nasolabial, marionette lines, deep neck folds)	Deep dermal, subcutaneous, and deeper soft tissues, ligaments, and fat	Lasers, botulinum toxin, fillers Surgery—face lift, blepharoplasty, neck lift may be required

may not be desirable in females. BoNT injection into the masseters leads to atrophy of the muscles, and a softer and less angular jaw line and facial profile.

Platysmal banding

Platysma is a paired flat muscle in the neck which is deficient near the midline. The anterior edges of platysma form prominent bands as one ages. BoNT injection into platysmal bands causes weakening and atrophy and leads to less prominent bands and a smoother neck contour.

Reduction of salivary gland bulk

Prominent parotid glands can give the same effect to the lateral facial contours as masseteric hypertrophy. Hypertrophic or ptotic submandibular glands can be noted as bulging lumps in the submandibular region, particularly as someone ages, which is consider non-aesthetic. Injection of BoNT directly into the glands has been shown to reduce the parasympathetic stimulation of glandular activity and, hence, induces atrophy and reduction in the size of the glands.

Other cosmetic indications

Include bunny lines, marionette lines, a gummy smile, a cobblestone chin, and a droopy nasal tip.

Complications of botulinum toxin injection

BoNT injections may lead to complications.[3] Common ones include bruising, asymmetry, and unintended paralysis of adjacent muscles. Rarely, allergic reactions to the toxin can occur. The injector must be well acquainted with the anatomy of facial muscles, including the location and depth of the target and surrounding muscles, preventing paralysis of adjacent muscles. Sometimes, diffusion of BoNT to adjacent muscles can occur even if accurate injection into the target muscle is performed.

The most obvious complications are those that result in asymmetry. Examples are blepharoptosis from involvement of the superior portions of the orbicularis oculi muscle during BoNT injections at the glabellar region, and drooping of the angle of the mouth from involvement of the lip elevators from paranasal injections. Dysfunctional animation of perioral muscles leading to oral incompetence and drooling can occur from depressor anguli oris, mentalis, and orbicularis oris injections. Lateral rectus palsy may occur following lateral orbicularis oculi injection. Injection several millimetres lateral to the lateral orbital rim is advised to prevent this.

Fillers

Fillers are materials that are injected into the facial soft tissues for the correction of rhytids and folds, facial contouring, sculpting, and volumizing.[1,4] They can be classified as permanent, semipermanent, and temporary fillers.

Temporary fillers may last up to 12 months, while semipermanent fillers can last for several years and permanent fillers are not resorbed or broken down. Temporary fillers are most commonly used as their effects can generally be reversed and they have good safety

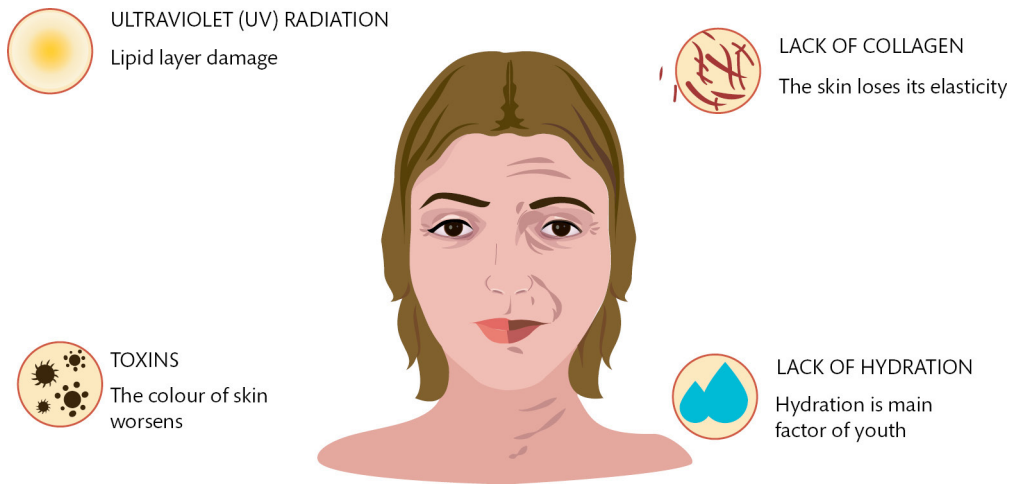

Figure 41.1 Causes and features of facial ageing.

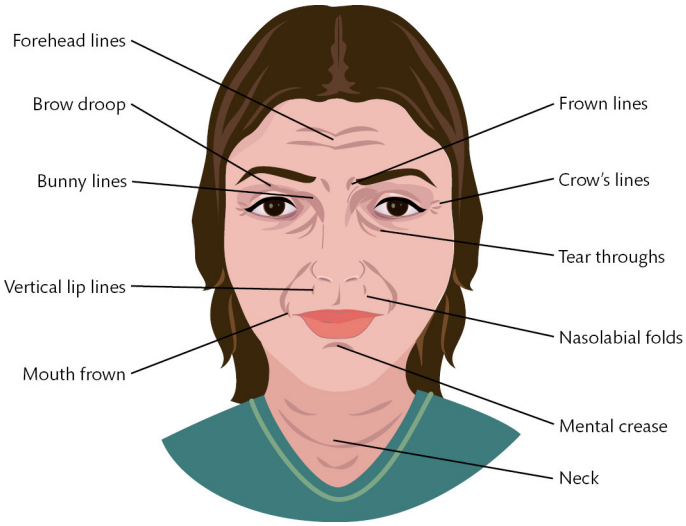

Figure 41.2 The most common types of wrinkles.

Box 41.2 Hypothesized effects of botulinum neurotoxin on dynamic rhytids

- Weakening of mimetic muscles, reduces skin creasing.
- Weakening of muscles leads to impairment of lymphatic drainage from the skin and soft tissues, leading to mild skin oedema and improvement in rhytid.
- BoNT's reduction of sweat gland activity, leading to improved skin quality.

profiles. Hyaluronic acid (HA) fillers can be dissolved by injecting hyaluronidase. The main disadvantage is that repeat injections are required to maintain their effects.

Permanent fillers are less popular today as they are irreversible and can stimulate foreign body reactions and scar deposition leading to palpable nodules, granuloma formation, and infections, which may require surgical excision or drainage. Table 41.3 shows the classification of various filler materials.

The physical properties of fillers vary to suit different functions[4,5]:

- *Elastic modulus (G′ or G prime)*: the G′ is a measure of the ability of the filler to retain its three-dimensional structure and resist deformation under stress. Products with a higher G′ provide greater structural support and volumization but may be more palpable while those with a lower G′ disperse more and are better suited to smoothening of superficial irregularities and fine lines.
- *Cross-linkage*: HA is a polysaccharide which lasts for 4 days in its natural state. Cross-linking of HA molecules enables HA to remain stable for longer periods, increasing the longevity and firmness of the filler. The type and degree of cross-linkage and proportion of cross-linked versus non-cross-linked HA differ across products.
- *Biostimulatory effects*: some fillers such as PMMA and polycaprolactone induce neocollagenesis, leading to a volumizing effect that lasts beyond the filler lifespan.
  - *Other properties*: the particle size, degree of hydration, concentration and combination of filler materials, presence of local anaesthetic, and combination with vitamin C, collagen, or other ingredients can all influence the physical features and functions of the product.

Table 41.2 Elevator and depressor facial muscles

	Facial elevator muscles	Facial depressor muscles
Periorbital	Frontalis	Procerus Corrugator supercilii Orbicularis oculi Depressor supercilii
Perioral	Levator labii superioris Levator labii superioris alaeque nasi Levator anguli oris Zygomaticus major Zygomaticus minor	Depressor anguli oris Depressor labii inferioris Platysma

Table 41.3 Classification of filler materials

Temporary	Semipermanent	Permanent
Hyaluronic acid	Calcium hydroxyapatite	Polymethylmethacrylate (PMMA)
Collagen	Poly-L-lactic acid	Polyacrylamide
	Polycaprolactone	Silicone

Properties of an ideal filler

- Easy to administer.
- Painless to administer.
- Non-migrating.
- Long-lasting.
- Non-inflammatory/non-allergenic.
- Minimal adverse effects.
- Affordable.
- Reproducible results.
- Reversible.

Common treatment sites using a filler

Upper third of face

- Filling of forehead depressions.
- Volumization of temporal hollowing.

Middle third of face

- Glabellar lines.
- Nasal radix and dorsal augmentation.
- Nasal tip projection and refinement.
- Orbital hollowing.
- Tear troughs.
- Lower eye bags.
- Cheek and zygomatic volumization and projection.
- Perialar depressions.
- Nasolabial folds.

Lower third of face

- Lip volumization and lifting.
- Marionette lines.
- Chin augmentation.
- Jaw line augmentation and definition.

Whole face

- Superficial fine rhytids (e.g. lateral orbital and perioral lines).
- Deep rhytids.

Complications

Common early reactions

These include swelling, bruising, and presence of needle marks, which usually resolves within days with conservative management such as icing.[6]

Product visibility

The plane of injection and the quantity injected must be accurately controlled. Injecting too superficially or in excessive quantities into a region with thin skin may lead to a translucent light-blue or grey appearance in the area called the 'Tyndall effect' with HA, or with whitish or palpable lumps with opaque products. Massaging to disperse the product, drainage of the product, or, in the case of HA, injection of hyaluronidase to help break down the product can facilitate a more cosmetic outcome.

Allergic reaction

Allergic reaction to filler materials are rare but may happen with bovine collagen or HA. A history of allergy to medications, HA, bovine collagen, and other constituents of fillers should be obtained prior to injection. Management is with antihistamines, systemic corticosteroids, and/or hyaluronidase.

Infection

Infection may be early or delayed. Prevention is paramount. Thorough antiseptic cleansing of the treatment area is essential. It is prudent to pretreat patients with previous herpes simplex infection or active acne with antiviral and/or antibiotics pre- and post-injection. Biofilm formation may lead to latent infections that can surface months to years after filler injection, especially in permanent fillers. Treatment of infections is with antibiotics or antivirals. Drainage of abscesses and surgical removal of infected permanent filler may be necessary.

Vascular occlusion

The face is very well vascularized with veins and arterial branches from the external and internal carotid arterial systems. The most feared complication of filler injection is vascular occlusion. This occurs when filler material is inadvertently injected intravascularly, or when filler material compresses an adjacent vessel cutting off blood supply to distal tissues. Filler embolization may occur and lead to subsequent distal tissue ischaemia.

When vascular occlusion occurs, skin blanching takes place almost immediately in the angiosome of the affected vessel. It can be accompanied by pain. The skin then turns dusky and necrosis may ensue leading to loss of skin and soft tissues, which can be very distressing to both the patient and the injector. Superimposed infection may also occur. The affected area can become irregular and depressed when healed.

Immediate response can limit the extent of damage. Injection of hyaluronidase directly into the site of filler administration, massaging the area to disperse the filler, application of nitroglycerine paste, and warm compresses may help. Other possible treatments include systemic antiplatelet therapy using aspirin, local or systemic corticosteroids such as prednisolone, and hyperbaric oxygen therapy is also described. Hyaluronidase has no effect if a non-HA filler is used.

If orbital pain and/or visual disturbance occurs following injection, filler embolization into the ophthalmic artery circulation must be suspected. Blindness following filler injection is a known, but thankfully rare, complication. It is postulated that filler material is embolized into the ophthalmic and/or retinal artery via anastomoses between the external and internal carotid arterial systems in the perinasal and periorbital regions.

Vascular events may be prevented by the following:

- Good knowledge of the vascular anatomy of the face.
- Use of a local vasoconstrictor (e.g. lidocaine with adrenaline) to reduce the diameter of vessels.
- Low-pressure, slow injection of small aliquots of filler.
- Pulling back the plunger for several seconds after insertion to aspirate for blood (that indicates that the needle or cannula is intravascular) before injecting filler.
- Injecting with a wide-bore, blunt-tip cannula to reduce the risk of vascular penetration.

Other modalities

Other somewhat more invasive yet non-surgical modalities include the use of lasers, chemical peels, and other energy-based devices, such as radiofrequency which may be monopolar, bipolar, or microneedling, and high-intensity ultrasound.[1,7] Radiofrequency and ultrasonic devices heat up subdermal tissues to a temperature of approximately 42°C, which results in fibroblast stimulation and eventually neocollagenesis.

Laser and chemical peels achieve similar effects, but there is more epidermal injury. Lasers may be ablative (carbon dioxide and erbium YAG) or non-ablative. Ablative lasers vaporize the epidermis, heat up the dermis, and induce healing and neocollagenesis, leading to smoother and tighter skin. They are suitable for moderate to deep rhytids and scars. Chemical peels produce similar effects as ablative lasers. Non-ablative lasers heat up the dermis to induce neocollagenesis without removing the epidermis. They are suitable for mild to moderate rhytids and improve skin tone. Other devices include intense pulsed light which work on the principle of lasers but are multifrequency by nature. The authors generally do not recommend intense pulsed light for facial rejuvenation in a medical setting. Cutaneous pigmentary lesions, such as solar lentigos, appear as one ages. Pigmentary lasers are absorbed by melanin in skin. Laser energy is converted to heat which leads to the destruction of melanocytes and improvement or resolution of pigmented lesions.

Regulations

The US Food and Drug Administration considers fillers as a medical device[8] and as potential adverse outcomes from the use of BoNT and fillers can be significant when administered inappropriately, their use is regulated in most countries, albeit to varying degrees.

While their administration is restricted to qualified medical practitioners in some countries, in others, nurses and aestheticians are also licensed to administer these products.

In many countries, medical practitioners have to complete training approved by the relevant health authorities to administer BoNT and fillers. In the UK, Health Education England has standardized the training of non-surgical cosmetic procedures as qualifications accredited by the Office of Qualifications and Examinations Regulation, with standardized graduated theory and practical modules. Practitioners in the UK have to attain these qualifications before they are allowed to perform these procedures, but this is not strictly enforced. Despite this regulatory pathway, the illegal use of these products is widespread.

The authors recommend that interested practitioners familiarize themselves with local and national regulations, attend courses accredited for training, and gain adequate experience before embarking on practising non-surgical rejuvenation.

Health Education England's training regulations can be found online (https://www.hee.nhs.uk/news-blogs-events/hee-news/new-qualifications-unveiled-improve-safety-non-surgical-cosmetic-procedures).

Additionally, cosmetic fellowships in ear, nose, and throat, plastic surgery, and dermatology that provide advanced training opportunities are available in many countries through educational bodies such as the European Academy of Facial Plastic Surgery (https://eafps.org/). The authors encourage the reader to visit the relevant websites for further information.

Conclusion

Non-surgical facial rejuvenation is a significant industry today and continues to grow at a rapid rate across the globe. The products and techniques have improved tremendously in terms of outcomes and safety profile over the past few decades and continue to do so.

BoNT and dermal fillers have always been at the core of non-surgical facial rejuvenation, and will continue to be so, because of the safety profile and excellent outcome.

While their administration has been largely safe in the hands of well-trained practitioners, inappropriate administration has led to significant complications on occasions and should be avoided. As such, standardized accredited training is key for safe and regulated practice of non-surgical facial rejuvenative procedures.

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42

Allergy of the nose

Richard Douglas

Introduction

The most common clinical manifestation of allergy of the nose is allergic rhinitis, and the pathogenesis, diagnosis, and management of this condition will be the focus of this chapter.

Allergic rhinitis is an extremely common chronic disease with a prevalence of around 20% in Western societies. It is the most commonly occurring of all allergic diseases and is associated with a reduced quality of life, decreased productivity at work, and impaired learning at school.

Pathogenesis

Allergic rhinitis develops as a consequence of the production of allergen-specific immunoglobulin (Ig)-E in response to exposure to aeroallergens. The aeroallergens towards which these specific IgE molecules are directed are both ubiquitous (grass pollen, house dust mite, cat dander) and inherently harmless. Why certain antigens provoke an allergic reaction is largely unknown. Whether an individual produces specific IgE to an aeroallergen on exposure to that allergen is a reflection of the cytokine response of that individual's T-helper lymphocytes. A predominantly Th1 cytokine response tends to discourage the production of specific IgE in response to aeroallergen exposure whereas a predominantly Th2 cytokine response (characterized by the release of IL-4, IL-5, and IL-13) tends to encourage specific IgE production. Whether an individual tends towards one response or the other is a reflection of their genetic inheritance, as well as their exposure to infections and other antigens in early childhood. Individuals who develop specific IgE to aeroallergens are termed atopic. However, many atopic individuals do not develop allergic diseases (allergic rhinitis, allergic asthma, atopic dermatitis, and food allergies). This fact must be remembered in the interpretation of skin prick and radioallergosorbent test (RAST) results.

Specific IgE molecules are synthesized by plasma cells in regional lymph nodes. They are released into the circulation and are taken up by basophils and predominantly by mast cells in tissues. Aeroallergens are inhaled and come into contact with sinonasal mucosa. An aeroallergen molecule causes cross-linking of two adjacent IgE antibodies with specificity to that aeroallergen and that triggers the release of inflammatory mediators such as histamine. These mediators result in tissue oedema, sneezing, itch, and excessive mucus production that characterize allergic rhinitis.

Differential diagnosis

Chronic nasal obstruction is most commonly caused by inflammatory conditions, such as rhinitis (confined to the nasal cavity) or rhinosinusitis (extending to involve the mucosa of the sinuses), and anatomical problems, such as septal deviation. In childhood, adenoidal hyperplasia is an important differential diagnosis. This section will outline an approach to the patient with nasal congestion and/or rhinorrhoea.

Chronic rhinitis and rhinosinusitis often present with similar symptoms. Obstruction and rhinorrhoea are usually a feature of both. The rhinorrhoea of chronic rhinosinusitis (CRS) is more likely to be viscous, discoloured, and posterior. CRS is more frequently associated with hyposmia than is rhinitis.

Nasal endoscopy and computed tomography (CT) sinus scans objectify the diagnosis, and one or both of these are required in cases that do not respond readily to medical therapy.

Not all cases of rhinitis have an allergic pathogenesis and distinguishing the various forms of rhinitis is not always an easy task. Chronic rhinitis can be divided into three types: allergic rhinitis, non-allergic rhinitis, and vasomotor rhinitis.

Allergic rhinitis

Allergic rhinitis rarely occurs in children younger than 2 years because individuals require low-dose exposure to the causative allergen for a prolonged period before sensitization develops. Its age of onset is typically about 3 years, which is later than the age of onset of atopic dermatitis but younger than is usual for allergic asthma (these being the three conditions of the 'allergic march'). A family history of one or more of these conditions is often reported by patients with allergic rhinitis.

Grass pollen hypersensitivity causes seasonal allergic rhinitis, reflecting the increased exposure to pollen during the spring and summer months. Airborne tree pollen counts, and particularly birch pollen in Northern and Central Europe, are typically highest in late winter and early spring.

Patients with sensitivity to perennial allergens, such as dust mite, cat dander, and mould spores, tend to have symptoms that are relatively constant throughout the year. Atopic patients are frequently sensitive to both seasonal and perennial allergens and as a result complain of perennial symptoms that worsen during the pollen season.

Non-allergic rhinitis

The symptom profile of non-allergic rhinitis is similar to that of perennial allergic rhinitis but has the two important differences of onset in early adulthood and an association with adult-onset asthma.

Histopathologically, there is no difference between allergic and non-allergic rhinitis, and unless allergen-specific treatment is contemplated, both the medical and surgical treatment of these two conditions are very similar.

Vasomotor rhinitis

Vasomotor rhinitis manifests quite differently from allergic and non-allergic rhinitis. It typically begins during middle age or later in life and is characterized by clear anterior rhinorrhoea. The rhinorrhoea is usually not associated with nasal obstruction and is characteristically worsened by exposure to cold air and to hot or spicy food. It is thought to be caused by an imbalance between the sympathetic and parasympathetic nerve supply to the nasal mucosa in which the parasympathetic innervation becomes predominant, encouraging the excess production of nasal mucus. Unlike both allergic and non-allergic rhinitis, vasomotor rhinitis is not associated with asthma.

Examination findings in allergic rhinitis

Examination of the face may show dark periorbital skin and puffiness of the eyelids, usually the result of venous stasis caused by chronic nasal obstruction. This appearance is often referred to as 'allergic shiners'. A horizontal supratip crease at the junction of the upper and lower lateral cartilages is the result of frequent pushing upward of the nasal tip by the back of the hand to wipe away mucus, the so-called allergic salute.

There are several different approaches to the examination of the nasal passages but rigid nasal endoscopes offer the best view. Allergic inferior turbinates are swollen and have a pale blue hue (Figure 42.1). The middle turbinate is either normal or its anterior aspect may be oedematous.

In order to allow evaluation of the reversibility of nasal congestion, a topical decongestant such as oxymetazoline can be applied, and the nasal cavity examined a few minutes later. This step also facilitates examination of the middle meatus.

Investigations

Very many cases of uncomplicated rhinitis and sinusitis require no specific investigations, particularly if they respond to simple therapeutic interventions. If, however, symptoms persist despite the first level of medical intervention, then some investigations are warranted.

The investigations for rhinitis have two goals: the first is to determine whether the rhinitis has an allergic basis (aeroallergen skin prick test or RAST) and the second is to confirm the diagnosis by excluding concomitant sinusitis (sinus CT scan; Figure 42.2).

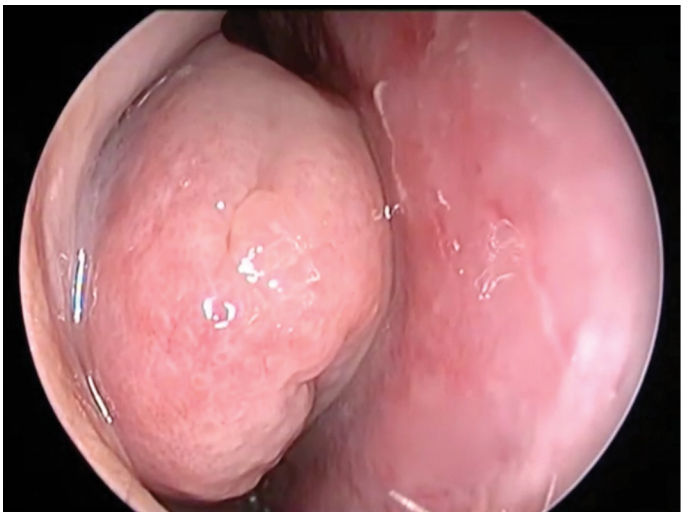

Figure 42.1 A patient with severe rhinitis. The inferior turbinates are very large and pale.

Skin prick testing

Allergy testing can be useful to identify a potential allergic trigger. Aeroallergen skin prick testing is safe, rapid, and relatively inexpensive. A patient, with presumed allergic rhinitis, has placed on his or her volar wrist a very small amount of aeroallergen suspended in glycerol. Next, a lancet with a 1 mm tip is pushed through the droplet and into the upper layers of the skin. If the patient has mast cells within the skin that have specific IgE to the allergen, the mast cells will degranulate and the histamine released will form a wheal. A wheal more than 3 mm in diameter is considered positive (Figure 42.3).

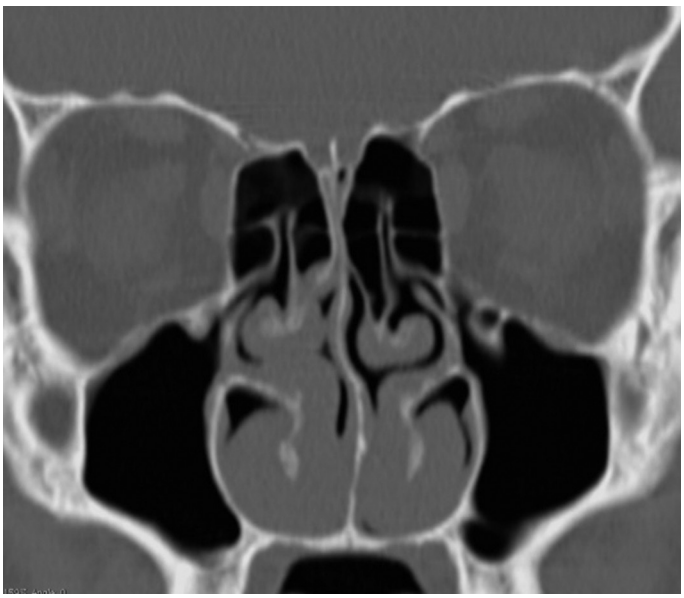

Figure 42.2 CT scan of the same patient as Figure 42.1. Scan shows very enlarged inferior turbinates but there is no evidence of sinusitis.

Figure 42.3 Skin prick tests are easily performed. A small lancet is placed through a drop of allergen suspension. The resultant wheal is measured after 15 minutes. Full cardiorespiratory resuscitation equipment should be available if performing skin prick testing. A comprehensive and safe protocol should be developed.

Most allergic rhinitis is caused by a small number of aeroallergens (house dust mite, grass pollen mix, tree pollen mix, weed mix, cat dander, dog allergen, and mould mix) so testing for a very large number of allergens is rarely productive. Others may be tested for if the patient gives a history of reaction to specific allergens.

Although skin prick testing can determine whether a patient is atopic or not, it does not confirm an allergic illness. Only about half of all subjects with positive skin prick tests have an allergic disease (allergic rhinitis, allergic asthma, or atopic dermatitis). This implies there are a number of aeroallergens to which sensitization occurs but this immunological reaction results in no symptoms.

In general, a skin prick test panel should be requested only when allergen-specific treatments (immunotherapy or specific allergen avoidance) are being considered. This is because the non-allergen specific treatments for allergic and non-allergic rhinitis are essentially the same.

Antihistamines should be stopped for a few days before testing is performed as patients taking them may have false-negative results.

Radioallergosorbent test

RAST detects the presence of allergen-specific IgE circulating in the serum. The level of tissue-specific IgE is very much higher than the serum level, so that RAST tests are generally considered a little less sensitive than skin prick tests. They have a significant advantage in that the patient simply needs to have a venepuncture and does not need to attend a laboratory or allergy clinic to have the skin prick testing performed, and the results are unaffected by taking antihistamines.

Radiology

CT scans of the paranasal sinuses and nasal cavity are extremely helpful in the management of patients with allergic rhinitis that is resistant to medical therapy. CT scans distinguish between rhinitis, in which the soft tissue changes are confined to the turbinates and nasal cavity and sinusitis, where mucosal thickening within the sinuses is the key feature (**Figure 42.2**). CT scans also clearly reveal deviation of the nasal septum and other anatomical features (such as conchae bullosae) which may predispose the patient to nasal obstruction.

Treatment

There are four types of treatment for allergic rhinitis: drugs, allergen avoidance, immunotherapy, and surgery. These options can be used according to the patient's response to more simple therapies and their personal preference.

Oral antihistamines and topical steroids

Patients with symptoms that are only intermittent can often be managed by taking oral antihistamines as needed. Oral antihistamines are effective in controlling symptoms such as sneezing, itching, rhinorrhoea, and lachrymation but these drugs are less effective in improving nasal congestion. Second-generation antihistamines, such as loratadine and fexofenadine, possess a very benign side effect profile, causing no sedative effect while delivering excellent symptomatic relief for patients with mild disease.

Topical corticosteroid sprays are the preferred medication for patients with persistent congestion and/or rhinorrhoea. Occasional bloodstained nasal discharge can occur as a side effect, but it can usually be managed by reducing the dose and frequency of application. Directing the spray directly against the septum may worsen this side effect.

It is important to highlight that intranasal steroid sprays have a slow onset of action. They take several days to start working and do not reach a plateau of clinical effectiveness until they have been administered for at least 2 weeks. Second-generation intranasal steroids are preferred among the others due to their very low bioavailability and long-term safety. There is some evidence that beclomethasone (first generation), with its higher bioavailability, may result in growth retardation if used twice daily in children. Daily intranasal corticosteroid sprays and oral antihistamines can be used very effectively in combination. However, by no means do all patients achieve adequate symptomatic relief from these medications used alone or in combination.

Very severe cases of allergic rhinitis may be improved rapidly with a short course of prednisone. This has the effect of shrinking the size of the inferior turbinates, allowing better access for intranasal corticosteroid sprays. Intramuscular depot corticosteroid injections work for a number of weeks, but only because they deliver a significant dose of corticosteroids over this time. If patients require or request this form of treatment, they can nearly always be managed as effectively with topical treatment, immunotherapy, or surgery, thereby reducing the patient's exposure to systemic corticosteroid side effects and the risk of lipoatrophy at the site of the injection.

Surgery for resistant cases

Surgical treatment for allergic rhinitis is a good option if patients fail to improve with simple medical therapy. This usually requires a short general anaesthetic and involves reducing the size of the inferior turbinates. Septal deviation may be addressed at the same time. Some in-office/outpatient techniques for inferior turbinate reduction have been developed, such as coblation, radiofrequency, and cauterization. Their long-term efficacy is less than operative techniques, but logistically they have advantages in that the patient does not have to go to an operating theatre.

Randomized trials of the efficacy of inferior turbinate reduction techniques suggest that the key manoeuvre to achieve a long-lasting improvement is to reduce the inferior conchal bone.

Immunotherapy for allergic rhinitis

Allergen-specific immunotherapy involves the repeated administration of antigen extract, either sublingually or subcutaneously, in an attempt to alter patients' immunological responses and improve their symptoms.

The key to successful immunotherapy is the correct identification of the relevant allergen and then administering it at high doses for a prolonged period. This is relatively straightforward for seasonal allergic rhinitis. The problem is not quite so easily solved for perennial allergens as several may be involved although house dust mite is widely thought to be the most important in most temperate countries. The majority of immunotherapy regimens are based on this allergen.

It is crucial to establish that the patient has specific IgE against the allergen to which he or she is going to be desensitized. This can easily be achieved with skin prick testing or RAST. Immunotherapy may be administered subcutaneously, sublingually, or orally. The advantage of sublingual and oral administration is that injections are avoided and the possibility of anaphylaxis (very small after subcutaneous injections) is close to zero. Surgery and immunotherapy should not be regarded as mutually exclusive. Some patients will prefer one modality to the other and if one fails to generate sufficient improvement of symptoms, the other can be tried.

Allergen avoidance

Aeroallergens tend to be ubiquitous and so avoidance can be difficult to achieve. Exposure usually has to be reduced to a very low level to have a clinically meaningful effect. The best-studied interventions are those which reduce house dust mite exposure, such as the wrapping of pillows and mattresses in allergen-impermeable covers. Although some patients derive benefit from such measures, the overall response rates in clinical trials have not been impressive.

Allergic rhinitis and adenoidal hyperplasia

It is not uncommon for a 4- or 5-year-old child to present with nasal obstruction and snoring. Often there is a family history of allergic illness and the inferior turbinates appear pale and swollen. Posterior nasendoscopy reveals adenoids filling much of the posterior choanae. In such cases it is difficult to determine the relative contribution of these two factors to the impediment of airflow. Combining an inferior turbinate reduction procedure with adenoidectomy is often the best course of action for these young patients. Surgical reduction of inferior turbinates is often an effective intervention in children with severe allergic rhinitis as younger patients are frequently reluctant to use sprays regularly. A 9-year-old child with severe nasal obstruction due to allergic rhinitis simply mouth breathes as he or she has done since early childhood, seeing no advantage in medical treatment.

Allergic rhinitis and chronic rhinosinusitis

The prevalence of atopy is now around 40% in Western societies. Accordingly, it is not uncommon for patients with CRS to have positive skin prick tests for aeroallergens. The effect that atopy has on the pathogenesis and severity of CRS has been debated extensively in the rhinology literature. CRS is generally a disease that occurs in adulthood, whereas an individual's atopic status is determined in the first few years of life. If either atopy or allergic rhinitis had a significant role in the pathogenesis of CRS, then it would be not unreasonable to expect a much higher prevalence of CRS in children than that observed.

However, over the last few years a condition termed central allergy syndrome has been described in a subgroup of CRS patients. These patients are generally young adults and have oedematous inferior and middle turbinates and polyps in the middle meatuses, but on CT scanning have little mucosal inflammation elsewhere in the sinuses. Patients with this pattern of CRS tend very frequently to be atopic, and it is likely that the atopic status is associated pathogenically with this condition.

Allergic rhinitis and asthma

The upper and lower airways are closely linked from histological and immunological perspectives. The great majority of patients with allergic asthma have allergic rhinitis. The contribution that concomitant allergic rhinitis makes to the severity of asthma is often underestimated. Treating the allergic rhinitis of patients with difficult-to-control asthma often results in an improvement of both conditions. Asthmatics do better if they can breathe freely through their noses. Inferior turbinate reduction and/or immunotherapy can be considered at an earlier stage in patients with the combination of allergic rhinitis and asthma, as both the upper and lower airways may benefit from treatment with these modalities. Anecdotally, one often sees a reduction in topical and systemic corticosteroid use.

Tips and pearls

- Allergic rhinitis begins in early childhood.
- Children often do not complain of nasal obstruction but simply breathe through their mouths.
- Adenoidal hyperplasia is an important differential diagnosis in the paediatric population.

- Non-allergic rhinitis and CRS are key differential diagnoses in adults.
- A positive skin prick test or RAST to an aeroallergen demonstrates atopy but does not confirm an allergic condition, as many people have asymptomatic sensitization.
- Most patients with allergic asthma have allergic rhinitis, and treatment of their rhinitis often improves their asthma control.

Summary

Allergic rhinitis is highly prevalent and significantly reduces the quality of life of those who suffer from it. The mainstay of medical therapy for persistent sufferers is topical corticosteroid sprays, supplemented by oral antihistamines. Both surgery and immunotherapy are effective options for patients who have troublesome symptoms despite medical therapy, or for those who do not tolerate medical therapy.

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43

Hereditary haemorrhagic telangiectasia

Simon Gane

Introduction

Hereditary haemorrhagic telangiectasia (HHT), or Osler–Weber–Rendu syndrome, is a rare multisystem autosomal dominant genetic syndrome which often presents first to the ear, nose, and throat (ENT) surgeon as epistaxis is a cardinal symptom. The optimal management of the acute presentation has significant differences to the usual strategies outlined in Chapter 27, and the longer-term prevention of epistaxis often requires a combined surgical and medical approach.

Nomenclature and demographics

Although the term HHT was coined in 1909, it was first described by Sutton in 1864 as 'Curious cases of recurrent epistaxis associated with internal haemorrhages and telangiectases of the skin' followed by Rendu in 1896, Osler (1901), and Weber (1907). Prevalence in the UK is not known but is probably in the order of 10/100,000, given data from other European countries.

Diagnosis

Diagnosis is usually clinical and based on the Curaçao criteria (**Box 43.1**). The presence of any three is considered diagnostic or 'definite HHT', while the presence of two criteria is 'possible HHT'.

Box 43.1 Curaçao criteria for hereditary haemorrhagic telangiectasia

1. Recurrent spontaneous epistaxis.
2. Multiple telangiectases at characteristic sites (lips, oral cavity, nose, and fingers).
3. Visceral lesions such as gastrointestinal telangiectases or arteriovenous malformations in the brain, lung, spine, or liver.
4. First-degree relative with HHT.

Source data from Shovlin CL, Guttmacher AE, Buscarini E, et al. Diagnostic criteria for hereditary hemorrhagic telangiectasia (Rendu–Osler–Weber syndrome). *Am J Med Genet*. 2000;91(1):66–67.

Telangiectasia is the characteristic abnormality of the syndrome: small arteriovenous malformations arising in the oral and nasal mucosa as well as lips and fingers (**Figures 43.1 and 43.2**). The same pathophysiological process, probably an imbalance in angiogenic factors, is likely to cause arteriovenous malformations (AVMs) in the solid organs.

For most patients, clinical diagnosis is sufficient and genetic testing is not required, but there may be a role for screening suspected family members or *de novo* presentations. Currently, genetic testing requires referral to a specialist centre.

The underlying genetic abnormalities for almost 90% of criteria-diagnosed HHT sufferers have been elucidated. The majority of these are mutations in the *ACVRL1*, *ENG*, and *SMAD4* (associated HHT and juvenile (colonic) polyposis) genes. Recently, three other genes or loci have been described, as summarized in **Table 43.1**.

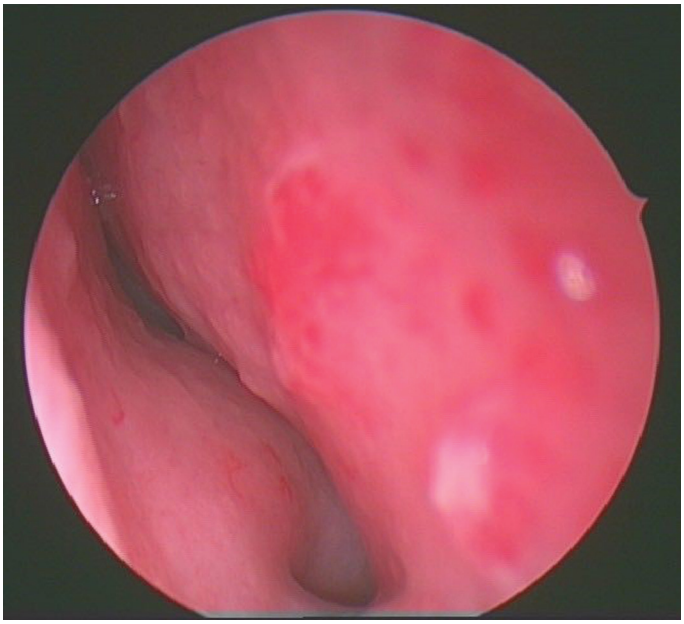

Figure 43.1 Endoscopic view of the right nasal septum and lateral nasal wall showing the small AVMs, telangiectasia, the characteristic abnormality of HHT.

Figure 43.2 Clinical photograph showing telangiectasia of the lips and skin in HHT.

Exactly why these mutations result in macro- and micro-AVMs is still unknown. Endoglin and ACVRL1 are both part of the transforming growth factor beta pathway, but why this should result in these vascular malformations is still not clear.

Differential diagnosis

Other causes of epistaxis are discussed in Chapter 27. Telangiectases may be found in other conditions such as scleroderma, CREST (calcinosis, Raynaud phenomenon, (o)esophageal dysmotility, sclerodactyly, and telangiectasia) syndrome, and in ataxia–telangiectasia, but all of these are even rarer than HHT.

Management

The evidence base for most surgical interventions in HHT is scanty. Most of these interventions have, at best, level 3 (case series) evidence. A combined approach with both medical and surgical interventions is required as outlined in Figure 43.3.

Surgical management

The flowchart for HHT management is outlined in Figure 43.4. The first surgical intervention is almost always to perform an examination under anaesthetic with focal ablation such as KTP laser to stage the disease. If this fails to control the epistaxis for a significant time (>3 months), consider septodermoplasty or medical intervention. Severe disease, indicated, for instance, by need for regular transfusions, may not be controlled by laser and may result in serious bleeding, so consider other options early in these cases. As this is an incurable and progressive disease, management success is characterized by a significant reduction in nosebleeds, for months in the case of focal ablation, years with septodermoplasty, and permanently with nasal closure.

Anaesthesia for patients with hereditary haemorrhagic telangiectasia

Anaesthesia for HHT surgery should preferably be undertaken by an anaesthetist with experience of the condition. These patients are often anaemic and have a higher risk of arteriovenous shunts. All should have antibiotic prophylaxis on induction, guided by local policy, and it is the author's practice to give 1 g tranexamic acid intravenously at the same time to reduce the risk of significant and prolonged bleeding. Mechanical deep vein thrombosis prophylaxis is also preferred as most patients are suitable for early postoperative mobilization, reducing the need for chemoprophylaxis.

Focal ablation of lesions

The mainstay of management of severe epistaxis in HHT is focal ablation of mucosal lesions. There are several options for topical control and the choice should be guided by local availability and an effort to minimize associated damage of normal tissue surrounding the lesions. To this end, a laser, such as KTP (532 nm) or argon (488 nm and 514 nm) green or green-blue coloured lasers utilizing frequencies absorbed preferentially by haemoglobin, is the modality of choice. Diathermy is also described although it, and chemical cautery with silver nitrate, should be avoided if possible due to the possibility of damage to surrounding tissue and risk of septal perforation. Due to the need for specialized equipment and a consistent multidisciplinary approach, it may be best if regional HHT care is centralized if feasible.

Laser surgery should only be performed in a laser-safe operating theatre by appropriately trained staff. Every staff member should have appropriate eye protection, as should the patient (a damp operative swab placed over the eyes is acceptable) and the endoscopic camera will need an appropriate filter for the laser wavelength. A laser-safety checklist is included as part of the preoperative pause.

The nose is prepared by the operator with gentle application of ribbon gauze or pledgets soaked with Moffett's solution (1 mL 10% cocaine, 1 mL 1:1000 adrenaline, 2 mL 5% sodium bicarbonate, 6 mL 0.9% saline) or adrenaline 1:10,000 into each nostril. The laser fibre is deployed in a laser handpiece with a suction channel for smoke clearance. A 0° 4 mm endoscope is used to visualize the nose and aim the laser via a visible aiming beam.

Each lesion is identified and circumscribed with the laser beam before ablating the central component. While ideally every lesion should be coagulated while attempting to minimize collateral damage to the septum, this is not always possible in severe cases. In most noses the lesions will be limited to the anterior septum and

Table 43.1 Genes and loci causing hereditary haemorrhagic telangiectasia

Subtype	Gene/locus	Prevalence	Phenotype
HHT1	*ACVRL1*	40–50%	Pulmonary and cerebral AVMs
HHT2	*ENG*	40–50%	Hepatic AVMs
HHT3	5q31.3–32	?	
HHT4	*RASA1*	?	
HHT5	*GDF2* (*BMP9*)	?	
Juvenile polyposis and HHT	*SMAD* 4	1%	Aortic dilatation

Adapted from Kroon S, Snijder RJ, Faughnan ME, Mager HJ. Systematic screening in hereditary hemorrhagic telangiectasia: a review. *Curr Opin Pulm Med*. 2018;24(3):260–268.

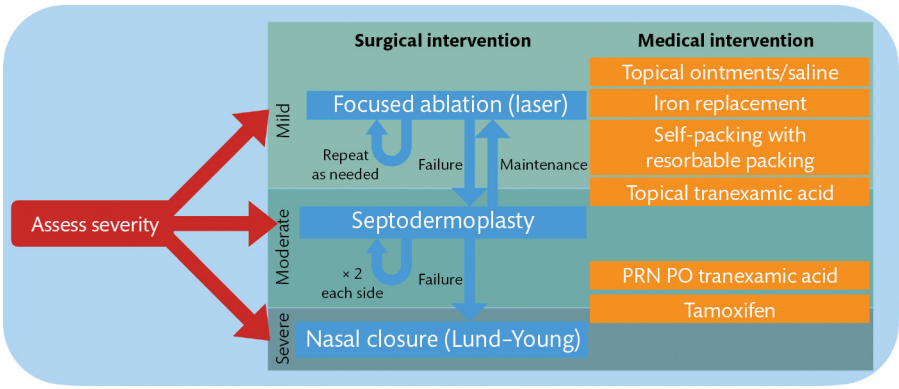

Figure 43.3 Overview of medical and surgical interventions with progression by severity in HHT. PO, orally; PRN, as needed.

lateral wall anterior to the head of the middle turbinate, although the floor may also be involved. The sill and valve areas require particular attention. Once completed, approximately 2 mL of chloromycetin ointment can be instilled into the nostrils, but a nasal pack is not necessary. A typical postoperative appearance is shown in Figure 43.5.

The main risks are summarized in Box 43.2, but the most severe is (rare) sudden profuse nasal haemorrhage, filling the nose and obscuring the endoscopic view. This can be challenging.

The first step is to use two large suckers, such as a Rowbotham, to remove blood while attempting to identify the source. Endoscopic bipolar diathermy may be more effective in a bloody environment than the laser which is absorbed by excess haemoglobin and eschar. If this is not effective, a matrix haemostatic foam such as Floseal may be instilled into the area. If this is still not effective, a fully opened-out 4 × 4 operative gauze soaked in 1:10,000 adrenaline and wrung out is a very effective temporary pack.

Successful laser ablation can result in months of epistaxis control, but patients often require repeated interventions.

Septodermoplasty

First described by William Saunders in the late 1960s as 'replacing the fragile respiratory epithelium of the anterior part of the nose with a split thickness skin graft. The skin graft, being tough, resists trauma and thereby prevents bleeding.' It is a reliable method for managing the anterior septal area which is almost always the site of the majority of telangiectases as described above.

This endoscopic technique involves preparation of the nose with topical anaesthetic and decongestant (usually Moffett's solution in the author's unit) followed by submucosal injection of local anaesthetic (1% lidocaine and 1:80,000 adrenaline).

Remove the septal nasal mucosa using a 4 mm microdebrider, preserving the white, fibrous perichondrium. The full height of septum anterior to the middle turbinate to the mucoepidermal junction can be prepared, ideally from posterior to anterior. Place a pledget with topical adrenaline or Moffett's into the nose while harvesting the graft.

The graft is an appropriately sized (about 5 × 5 cm) 0.5 mm thick split-skin graft, usually from the upper lateral thigh, using a powered dermatome if available. Place the graft skin side down on a square of Jelonet and fenestrate with a scalpel to prevent haematoma. Trim the Jelonet to size, slightly larger than the graft, and place the two layers onto the prepared graft site, Jelonet side up. Carefully free the Jelonet and remove using a Cottle's speculum which stabilizes the graft in position. Once the graft is placed satisfactorily, a small amount of fibrin glue can be placed under the anterior edge to maintain position. Pack the nose carefully without disturbing the graft position using a resorbable dressing such as Spongostan and dress the donor site with an occlusive waterproof dressing. The nasal dressing does not

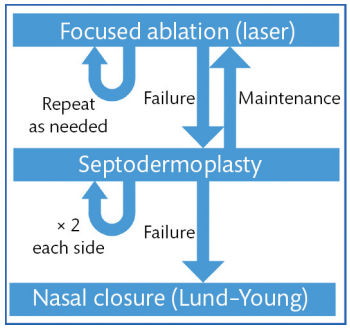

Figure 43.4 Algorithm for surgical management of HHT.

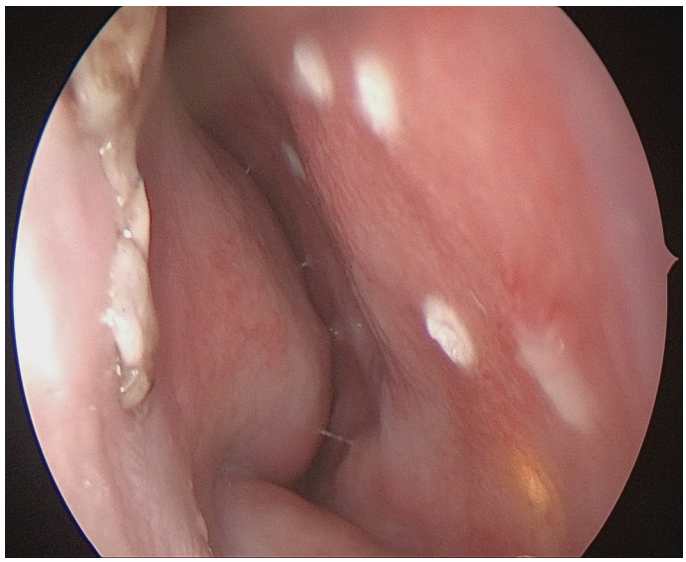

Figure 43.5 Intranasal view of right septum, with small telangiectases already lasered (white areas).

Box 43.2 Risks of laser ablation of nasal telangiectasia

- Bleeding intra- and postoperatively.
- Nasal obstruction.
- Synechiae.
- Laser injury to nose and other structures.
- Septal perforation and change of nasal shape.
- Recurrence of epistaxis.

need to be removed and a prophylactic course of a broad-spectrum antibiotic, such as co-amoxiclav, is given for 1 week.

This procedure may result in up to 3 or 4 years of epistaxis control on the treated side and can be performed twice on each side. The grafts usually shrink with age and new 'crops' of lesions occur at their margins.

Nasal closure

Nasal closure is the ultimate surgical treatment for severe and persistent epistaxis in select patients. The complete obturation of the nasal airway carries a large morbidity in terms of loss of ortho- and retronasal olfaction and the sequelae of obligate mouth breathing but for those with transfusion or infusion-dependent anaemia or significant quality-of-life loss, this trade-off may be worth it. Many end-stage HHT patients have permanent nasal obstruction due to blood clot and crust and can accept this trade-off.

Nasal closure is best performed in a centre with experience in the procedure as even the smallest fistula can result in persistent postnasal epistaxis. The Lund-modified Young's procedure using a three-flap technique gives reliable results.

Prepare the nose by instilling Floseal to reduce bleeding into the operative site. After local injection of 1% lidocaine and 1:80,000 adrenaline to the area, incise a circumferential transcutaneous incision along the mucocutaneous junction. From this incision make three anterograde incisions, one at the apex and one on either side of the vestibular base to divide this flap into three parts. Raise these flaps carefully to form three anteriorly based skin flaps. Each flap should be raised enough to allow tension-free apposition to the other two as shown in Figure 43.6. Then pack the nose with a resorbable dressing, such as Spongostan and meticulously suture the flaps to their neighbours using 4-0 PDS suture material in a horizontal mattress suture. The trifurcation is closed with a purse-string stitch. It is vital that these sutures are airtight (but not too tight!), as any leak will allow fistula formation and failure. A typical postoperative appearance is shown in Figure 43.7.

Acute management of hereditary haemorrhagic telangiectasia epistaxis

Even more than usual, an assessment using an ABC approach and early intravenous access is vital. Send a full blood count and crossmatch, these patients are often chronically anaemic and can be further compromised by multiple shunts in AVMs. HHT patients are generally experts in managing their own epistaxis and tend only to present as emergencies in extremis, usually after bleeding for a prolonged time.

A lot of the management advice in Chapter 27 applies, especially the need for personal protective equipment and the utility of a bright light, good suction, an endoscope, and an experienced assistant.

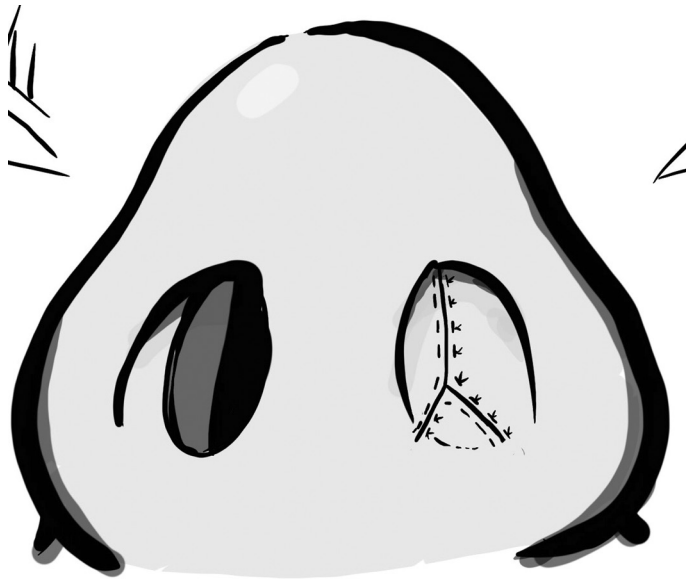

Figure 43.6 Diagram of basal view of nose with left nostril closed.

Although the evidence base is poor, the overriding principle of management in HHT is to be gentle, avoiding worsening of the situation by disrupting more fragile telangiectases:

1. Clear the blood clot with suction or nasal lavage and apply pressure by pinching the nostrils. Most patients will have already performed this manoeuvre before reaching hospital. Consider applying topical ointment such as Vaseline.
2. If that fails, pack gently with resorbable packing, such as Surgicel or Nasopore. The bleeding is almost certainly from the anterior third of the nose and posterior packing is not usually needed.

Figure 43.7 Basal view of successful nasal closure with three-flap technique.

Non-resorbable packing is to be avoided if possible but may be the only option in some settings. The hydro-colloid matrix covering of dressings such as Rapid Rhino enables smooth insertion and removal. It may improve intranasal coagulation and is to be preferred over firmer packing devices, such as Merocel, which can tear other telangiectases on insertion and removal. Any non-resorbable packing can be challenging to remove without restarting the bleed.

3. Floseal haemostatic matrix or similar can be injected into the nose if nasal packing fails. These can be very useful in controlling bleeds of unknown origin.
4. Any packing (even *in situ* for <48 hours) should be covered with prophylactic antibiotics, such as amoxicillin, or according to local guidelines.
5. If local packing fails to control the bleeding, interventional radiological embolization of the feeding arteries (facial and maxillary) can be undertaken, avoiding coils. This does not control nosebleeds for anything other than the short to medium term as this is a mucosal disease and the nose has an excellent collateral blood supply.
6. Consider iron replacement and check haemoglobin level in a few days.

Medical management

As a rare disease, HHT management suffers from a paucity of evidence. Most of the medical options are off licence and their efficacy is anecdotal. The sensible and evidence-based interventions are summarized here.

Iron supplementation

HHT patients are often chronically anaemic and their haemoglobin level should be checked regularly. Iron fumarate 300 mg orally once a day is a sensible first-line therapy iron supplementation and will often need to be given over several months.

Education and self-management

All patients with HHT should, of course, be counselled about the disease and its genetic origin. Ideally all patients should receive written information about epistaxis management and the need for antibiotic prophylaxis when undertaking dental or other procedures. Patients should also be counselled as how to best manage their nosebleeds themselves, and to be aware of red flags for significant blood loss and anaemia. Many find self-packing with a small patch (about postage stamp size) of Kaltostat helpful in controlling minor nosebleeds.

Topical ointments

Topical interventions such as humidification, gentle lavage with normal saline, and topical lubricants are low risk.

Topical ointments, such as a series of antibiotic creams rotating every 2 weeks (**Box 43.3**), are commonly prescribed in the author's unit.

Hormonal treatment

Tamoxifen, a partial oestrogen agonist, has been shown to be effective in improving bleeding, haemoglobin, and quality of life scores in one randomized controlled trial (RCT) and several case reports. At 20 mg orally, once a day, it is usually well tolerated. In women, side effects include menstrual irregularity, hot flushes, and vaginal discharge. Rare risks include benign endometrial hyperplasia which can concern their gynaecologists, deep venous thrombosis, stroke, cataracts, and uterine carcinoma. A transvaginal ultrasound scan should be performed prior to commencing treatment and patients are best managed jointly. Men may have gynaecomastia, headaches, nausea, and rashes, reduced sexual interest, and impotence.

Box 43.3 Example of nasal topical cream regimen

- Bactroban top to both nostrils twice daily for 2 weeks, then
- Naseptin top to both nostrils twice daily for 2 weeks, then
- Fucidin cream top to both nostrils twice daily for 2 weeks, then
- Restart Bactroban.

Tranexamic acid

As an antifibrinolytic agent, long-term systemic tranexamic acid (1 g orally three times a day) has been shown to improve self-reported epistaxis, without significant adverse events in RCTs. Short-term use to improve control for travel or social events, or in the event of a prolonged bleed, seems sensible, but long-term use in other diseases has been associated with thromboembolic events.

Tranexamic acid can also be used intranasally. In the UK it is available in a 5% solution as a mouthwash, or patients can dissolve one 500 mg tablet in 10 mL of sterile normal saline for the same dose. This can be instilled with a syringe or on dressings either regularly as prophylaxis or in response to a bleed. One RCT of a 10% tranexamic acid spray showed no superiority over topical bevacizumab, estriol, or placebo; however, a case report of 0.25 mL of 100 mg/mL tranexamic acid reported significant benefits and anecdotally, many patients report a reduction in incidence and severity of their epistaxis.

Obturation

Although cosmetically unsatisfactory, some patients prefer the reversible nasal obstruction of an obturator which can be custom-made using the same method as a hearing aid mould. In the author's experience, these do not tend to be tolerated by many.

Controversies

Screening

Screening for most solid-organ AVMs is recommended as outlined in Table 43.2, but advice on screening cerebral AVMs is mixed.

Table 43.2 Visceral arteriovenous malformation screening recommendations

Organ	Screening
Pulmonary AVMs	High-resolution CT chest/bubble echocardiography 5-yearly, also after puberty, pre- and postpregnancy
Hepatic AVMs	Not recommended
Cerebral AVMs	Not recommended (see 'Screening' in main text)

Pulmonary AVMs result in a right-to-left shunt and an increased risk of paradoxical thrombotic and septic emboli. Their natural history in this disease is still poorly understood and it is not known whether they arise *de novo* in adults or merely become detectable with growth of smaller pre-existing lesions from childhood. It is, however, well established that they cause morbidity and mortality, can evolve with time, and can be safely treated and are therefore ideal for targeted screening.

Currently, guidelines recommend an initial investigation with bubble echocardiography or high-resolution computed tomography scanning of the chest on first presentation and then every 5–10 years. Additional scans are required after puberty and pregnancy and before a pregnancy if one has not been performed within 5 years. Treatment consists of interventional radiological transcatheter embolization of a lesion. If even a small lesion is detected in HHT, patients should have written advice on antibiotic prophylaxis prior to non-sterile invasive procedures such as dental and endoscopic interventions.

Hepatic AVMs are associated with a lower morbidity but are very common (about 74% in one study). Current advice is not to screen for hepatic AVMs as they are rarely symptomatic, but some may occasionally require transplant.

Cerebral and spinal AVMs are detectable in approximately 10% of individuals with HHT and are a source of significant neurological morbidity due to rupture and intracerebral haemorrhage. Screening is controversial, with proponents suggesting one magnetic resonance imaging (MRI) scan of the brain for every HHT patient, and others pointing to the increased risk of stroke in interventionally managed unruptured cerebral AVMs, as demonstrated in the ARUBA trial. Current UK guidelines are to offer all asymptomatic patients an informed choice, but routine MRI screening is not recommended.

For a good overview of current screening evidence, see the review by Kroon and colleagues (see 'Further reading').

The author of this chapter arranges a high-resolution computed tomography scan of the chest for all HHT patients on first presentation, but no other imaging unless clinical signs or symptoms suggest it is needed. Genetic testing is not routinely helpful as it does not change management. The author discusses the evidence around cerebral AVM screening and recommends against it but will offer MRI scanning to those that are concerned.

Sphenopalatine artery/anterior ethmoidal artery ligation or embolization

Many HHT patients have undergone arterial ligation for management of their epistaxis. In the author's experience the procedure is rarely successful over the medium term. It is important to remember that this is a mucosal disease caused by abnormal arteriovenous communications and, thus, reducing regional arterial supply is unlikely to help. The same is true for endovascular approaches, although both may have a rare role in the emergency management of an uncontrollable epistaxis.

Potential future medical therapies

Bevacizumab

The monoclonal antibody vascular endothelial growth factor-inhibitor bevacizumab (Avastin) has been used both systemically and topically in HHT. A systematic review has concluded that local bevacizumab therapy may not be effective in the treatment of HHT but that more evidence is needed. It may be applied topically or submucosally, with submucosal injection associated with an increased risk of septal perforation, which can worsen epistaxis in HHT patients.

Intravenous bevacizumab is still considered experimental, with a recent review recognizing a lack of evidence but noting in their assessment of current publications (mainly case reports and case series) that 'patients experience a clinically relevant reduction in bleeding, reducing the need for transfusions, and elevating hemoglobin level'. However, at least one patient death out of the 177 cases was thought to be linked to the treatment, which should give pause to its use in a non-fatal disease.

Timolol

A recent double-blinded RCT of this topical beta-blocker droplet delivered over 4 weeks showed no change to epistaxis or quality-of-life measures, although the drug was well tolerated. A small Japanese trial showed some benefit in patients who had previously undergone septodermoplasty.

Thalidomide

A recent systematic review of thalidomide suggested it might have a place in the management of refractory epistaxis in HHT, although it is associated with a significant risk of adverse events including arterial thrombosis and deep vein thromboses.

Guidelines

In the absence of reliable evidence, a great deal of management advice is in the form of expert opinion and guidelines. The French have guidelines for acute management and are the most recent. These are available in English. The advice given in the above section 'Acute management of hereditary haemorrhagic telangiectasia epistaxis' is based partially on these.

VASCERN, the European research network on rare multisystemic vascular diseases, published a set of five patient outcome measures to maximize excellent patient care and minimize morbidity and mortality in 2018. These are summarized in **Box 43.4**.

Conclusion

HHT is a rare genetic disease which can be difficult to manage. It requires a multidisciplinary approach to manage the whole patient, but the ENT surgeon has a vital role to play. HHT patients are often experts in the management of their own disease and only usually

Box 43.4 VASCERN patient outcome recommendations

1. Pulmonary AVM screening.
2. Written nosebleed advice.
3. Assessment of iron deficiency.
4. Antibiotic prophylaxis prior to dental and surgical procedures for patients with pulmonary AVMs.
5. Written advice on pregnancy.

present late for emergency management. There are significant differences in management with the acute management of epistaxis, of which all ENT surgeons should be aware.

Acknowledgements

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FURTHER READING

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44

Endoscopic approaches to the anterior skull base

Haissan Iftikhar and Shahzada Ahmed

Introduction

The past two decades have seen a sharp increase in the number and types of endonasal endoscopic procedures. This in particular is attributed to advances in anterior skull base approaches for lesions both benign and malignant, which would once require open, transcranial and transfacial procedures. With greater understanding of endoscopic nasal and paranasal anatomy, surgeons ventured beyond the confines of sinuses to eradicate disease using endoscopic visualization. One such paradigm shift is seen in sellar pathologies, in particular pituitary gland lesions, which over the past decade or so have been almost universally approached endoscopically. Similarly, anterior skull base lesions which were previously only considered to be managed with open craniotomies are now being performed endoscopically. This has also been in part due to advances in reconstructive options. Both neurosurgeons and rhinologists have made great strides in managing even more complex cases beyond the boundaries of the nose and paranasal sinuses.

The sella can be taken as the epicentre of endoscopic skull base surgery as it is usually the starting point of many endoscopic skull base procedures and has a number of critical structures in its vicinity. Moreover, pituitary lesions outnumber any other skull base pathology. Endoscopic cerebrospinal fluid (CSF) leak repairs have also seen a steep rise due to enhanced visualization around corridors that modern angled endoscopes offer. This chapter will focus on various anterior skull base approaches, and will highlight the common pathology that can be addressed endoscopically.

Approaches

Anterior skull base approaches can be broadly classified into two planes, sagittal and coronal (**Table 44.1**). The anterior skull base extends from the sella to the frontal sinus in the sagittal plane and roofs of the orbits in the coronal plane (**Figures 44.1 and 44.2**).

The approach to the skull base is through the sinuses. The cribriform plate can be accessed by going through the ethmoidal air cells. Similarly, the sella is located beyond the sphenoid sinus. Therefore, a thorough knowledge and understanding of the nose and paranasal anatomy is paramount.

Sagittal plane

Transcribriform

Location/anatomy

The cribriform plate forms the roof of the nasal cavity. It is part of the ethmoid bone which hosts ethmoidal air cells inferiorly. The olfactory fila pass through the small foramina within the plate which mediate olfaction (**Figure 44.3**).

Pathologies

Both benign and malignant lesions can be found in the region of the cribriform plate. These include meningocele, meningoencephalocele, meningioma, esthesioneuroblastoma, and a range of sinonasal malignancies to name a few. The most common place of iatrogenic injury leading to CSF leak is the lateral lamella of the cribriform plate. Similarly, spontaneous CSF leaks are also commonly located in this area.

Boundaries

The transcribriform approach is bounded anteriorly by the frontal sinus, posteriorly by the planum sphenoidale, laterally by the orbits, and medially hosts the crista galli. Ethmoidal air cells have to be completely removed to provide access to this region. The cells make up a bony labyrinth and are divided into anterior and posterior air cells by the basal lamella of the middle turbinate. Some ipsilateral tumours such as olfactory groove meningiomas can be resected endoscopically via this approach, preserving the contralateral olfactory fila to avoid permanent anosmia.

Transsphenoidal

The sphenoid sinus is located posteriorly in the skull base. It hosts the sella and the roof of the sinus forms the planum sphenoidale, also referred to as the planum.

Table 44.1 Anterior skull base approaches with common indications

	Approaches	Common indications (pathology)
Sagittal plane	Transcribriform	Meningocele Meningoencephalocele Meningioma Esthesioneuroblastoma Sinonasal malignancies CSF leak
	Transsphenoidal:	
	Transplanum	Suprasellar meningioma Craniopharyngioma
	Pituitary lesion	Pituitary micro- or macroadenomas Rathke's cleft cyst Craniopharyngioma
	Transclival	Chondrosarcoma Chondroma
Coronal plane	Transorbital	Sphenoid wing meningioma CSF leaks Sinus tumours Thyroid eye disease Optic nerve decompression
	Transpterygoid	CSF leaks from the lateral recess of the sphenoid sinus Cholesterol granuloma Petrous apicitis Mucocoele Chondroma Chondrosarcoma Meningioma Trigeminal schwannoma Extensive juvenile angiofibroma
	Infratemporal fossa	Haemangioma Lymphoma Neurofibroma Malignancies of somatic origin
	Transodontoid	Basilar artery, vertebral artery, anterior inferior cerebellar artery, posterior communicating artery aneurysms Pontine cavernous malformation Meningioma Fibrous dysplasia Angiosarcoma Endodermal cysts and bulbus cavernoma

Source data from Snyderman CH, Gardner PA, Fernandez-Miranda JC, Wang EW. Extended anterior skull base approaches. In: Watkinson JC, Clarke RW, eds. *Scott-Brown's Otorhinolaryngology and Head and Neck Surgery*. 8th ed. CRC Press; 2018:1289–1304.

Transplanum

Location/anatomy

The planum sphenoidale forms the roof of the sphenoid sinus. It is a relatively horizontal piece of bone. Complete access to the planum can be achieved after removing posterior ethmoid air cells along with wide sphenoidotomies right up to the skull base (Figure 44.4).

Pathologies

A transplanum approach may be required for giant (>4 cm) pituitary adenomas compressing the optic nerves. Similarly, suprasellar meningioma and craniopharyngioma may also be approached via this route. Moreover, ophthalmic artery aneurysm can be clipped after dissecting the bone forming the planum.

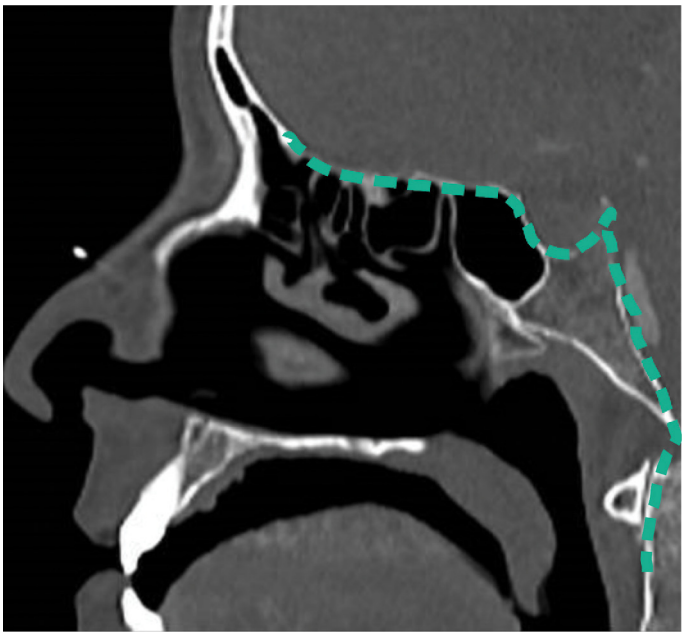

Figure 44.1 Skull base approaches in the sagittal plane.

Boundaries

The planum sphenoidale is bound anteriorly by the cribriform plate and the posterior ethmoidal artery, posteriorly by superior intercavernous sinus, posterolaterally by the optic canals, and anterolaterally by the orbital apex. After the region has been identified intraoperatively, the mucosa is stripped off the bone. The bone is then thinned out with a small diamond burr in the shape of a triangle with the base just behind the posterior ethmoid arteries and the medial border of the optic canals as the two sides. Copious

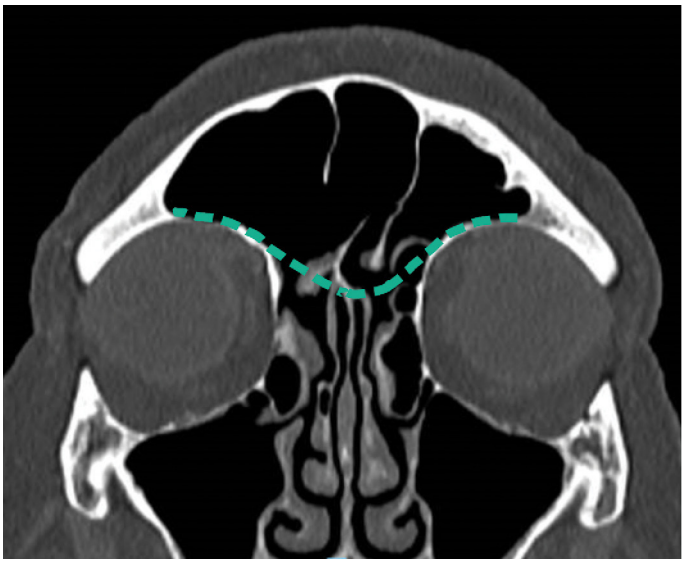

Figure 44.2 Skull base approaches in the coronal plane.

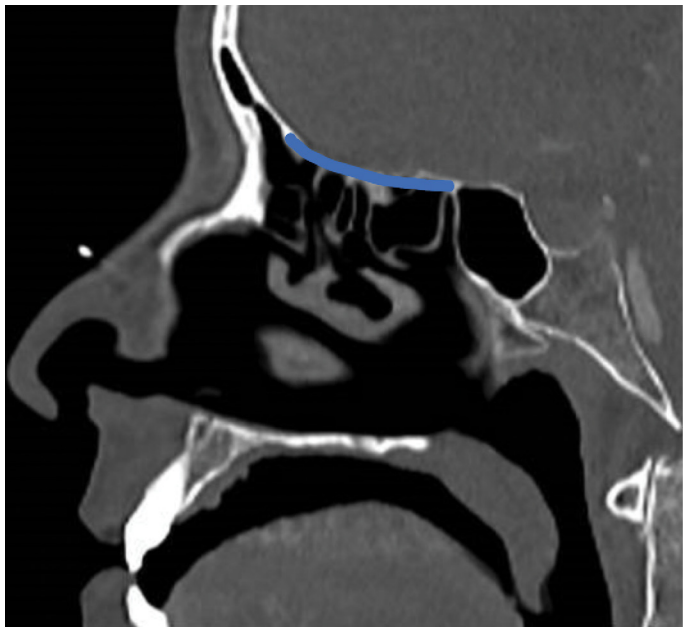

Figure 44.3 Transcribriform approach to skull base.

Figure 44.5 Transsellar approach to skull base.

irrigation is used to prevent thermal injury to the optic nerves in close proximity. Care has to be taken anteriorly to avoid inadvertent injury to the cribriform plate which can lead to a CSF leak.

Pituitary lesions

Location/anatomy

Pituitary gland tumours are found in the sella where the pituitary gland hangs below the diaphragm. The sella is formed by a bony covering that separates the sphenoid cavity from the pituitary gland. It often also has an anterior projection which can be appreciated within the sphenoid sinus (Figures 44.5 and 44.6).

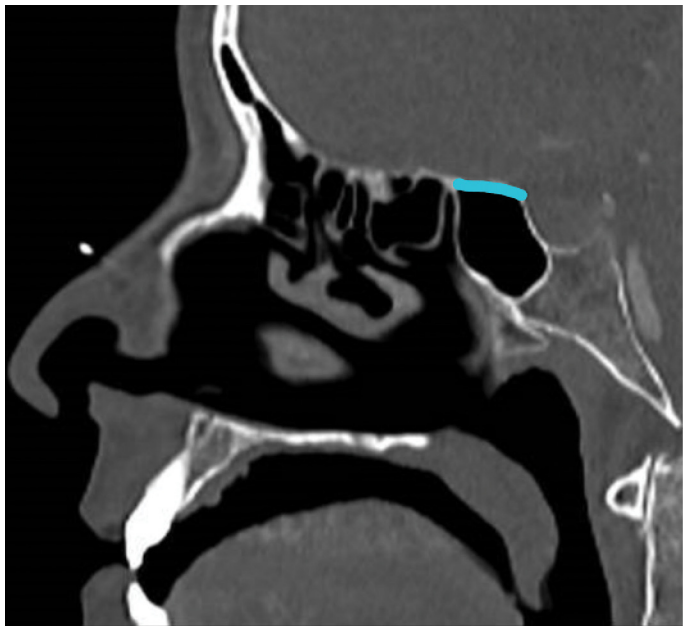

Figure 44.4 Transplanum approach to skull base.

Pathology

A transsellar approach is utilized for pituitary micro- or macroadenomas. Other pathologies include Rathke's cleft cyst and craniopharyngioma.

Boundaries

It is bounded anteriorly by the planum sphenoidale, posteriorly by the pituitary stalk, and laterally by the paraclinoid and paracavernous internal carotid arteries. The transsellar approach can be combined with transplanum and supra sella approaches for some large macroadenomas and suprasellar meningiomas. After wide sphenoidotomy and removal of the intersinus septum, a window is created in the anterior sella fossa. This is often deficient in

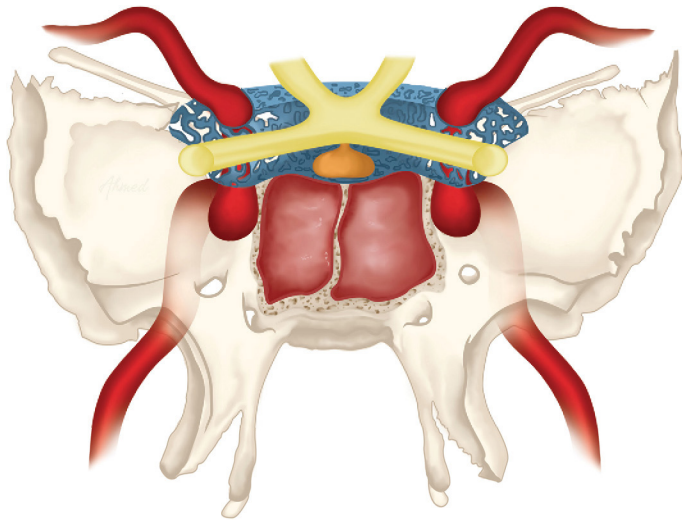

Figure 44.6 Coronal view of sphenoid and relationship to pituitary gland, optic nerves, and carotid arteries.

Figure 44.7 Transclival approach to skull base.

macroadenomas and care should always be taken when opening out this window laterally.

Transclival

Location/anatomy

The clivus (Latin for slope) is a bone at the skull base which slants posteriorly. It starts at the depression inferior to the dorsum sellae. It is made up of the posterior aspect of the sphenoid bone (basisphenoid) and a part of occipital bone (basiocciput). It is further classified as upper, middle, and lower. The upper part is at the level of the sella turcica. The middle part extends from the sella floor to the floor of the sphenoid sinus and the lower part is from the sphenoid floor to the caudal end of the clivus (Figure 44.7).

Pathology

The most common pathologies that are reached using the transclival approach include chondrosarcomas and chordomas. Chondrosarcomas are slow-growing malignant lesions of chondroid cell origin arising from the axial and appendicular skeleton. Chordomas are a rare type of bone tumours arising from transformed notochord remnants.

Boundaries

The clivus is bounded anteriorly by the body of the sphenoid bone and posteriorly by the basilar artery. The lateral limits are formed by the foramen lacerum. The superior limit is marked by the planum. A lower transclival approach may also be utilized to reach the odontoid process and the arch of the second cervical vertebra, to gain access to foramen magnum. Cranial nerves lie lateral to the clivus, therefore care must be to taken while addressing tumours which might be lying in close proximity. Cranial nerve III is at risk in the superior clivus, cranial nerve VI in middle clivus, and XII in lower clivus approaches.

Coronal plane

Transorbital

Location/anatomy

The orbital components include the eyeball along with extraocular muscles encased in periorbita. The orbit posteriorly has bony deficiencies which form the superior and inferior orbital fissures through which various neurovascular bundles traverse. These include the optic nerve and ophthalmic artery along with branches of trigeminal nerves. The orbital contents are sealed off anteriorly by the conjunctiva that is continuous with the upper and lower eyelids. The eyeball along with its extraocular muscles form the intraconal component of the orbit whereas structures surrounding them form the extraconal component.

Pathology

Transorbital surgery can further be classified into extradural and intradural. Extradural pathologies which can be address by transorbital surgery include sphenoid wing meningioma, CSF leaks, sinus tumours, thyroid eye disease, and optic nerve decompression. Intradural transorbital surgery, also referred to as TONES (TransOrbital NeuroEndoscopic Surgery), can address the pathologies of the anterior cranial fossa, middle cranial fossa, and cavernous sinus.

Boundaries

Transorbital surgery is bounded anteriorly by the conjunctiva, posteriorly by the orbital fissures, and by the bony wall forming the orbit. Endoscopic transorbital surgery can utilize four ports, namely superior, medial, inferior, and lateral, depending on the site of pathology. The surgery is undertaken with multiple teams involved including otolaryngologists, ophthalmologists, and neurosurgeons. Moreover, transorbital surgery can also be a part of multiportal surgery where the nose as a conduit is utilized for additional instrumentation and control. Surgery usually begins with a precaruncular incision and the eyeball with or without extraocular muscles are retracted. Care has to be taken with continuous measurement of intraocular pressure in order to avoid permanent loss of vision.

Transpterygoid

Location/anatomy

The transpterygoid approach is utilized to gain access to the pterygopalatine fossa, infratemporal fossa, middle and posterior cranial base, and lateral recess of the sphenoid sinus. The pterygoid body and lateral and medial pterygoid plates form the posterior border of the pterygopalatine fossa. The infratemporal fossa is located lateral to the pterygoid body and lateral pterygoid plate. The vidian canal and foramen rotundum can be approached via this route.

Pathology

A transpterygoid approach can be utilized to treat CSF leaks from the lateral recess of the sphenoid sinus, tumours in that region, cholesterol granuloma, petrous apicitis, mucocoele, chondroma, chondrosarcoma, meningioma, trigeminal schwannoma, extensive juvenile angiofibroma, and malignancies with perineural invasion of

Table 44.2 Carrau–Prevedello–Kassam classification of endoscopic endonasal transpterygoid approaches

Class	Region removed	Approach to
A	Partial removal (thinning) of pterygoid process	Pterygopalatine fossa (extended)
B	Anterior aspect of the base of the pterygoid process	Lateral recess of the sphenoid sinus
C	Base of the pterygoid process with dissection of vidian canal	Petrous apex/Meckel's cave
D	Partial or complete removal of the pterygoid plates with dissection of the petrous internal carotid artery	Extensive lesion requiring access to the infratemporal fossa and control of the petrous carotid artery
E	Partial or complete removal of the pterygoid plates with dissection of the petrous internal carotid artery and removal of the Eustachian tube	Nasopharyngeal malignancies, extensive tumours of the middle and posterior skull base (e.g. chordomas, chondrosarcomas)

Source data from Kasemsiri P, Solares CA, Carrau RL, et al. Endoscopic endonasal transpterygoid approaches: anatomical landmarks for planning the surgical corridor. *Laryngoscope*. 2013;123(4):811–815.

cranial nerve V_2. Nasopharyngeal malignancies extending far lateral into the pterygopalatine fossa and infratemporal fossa can also be approached via transpterygoid/infratemporal fossa routes.

Boundaries

Endoscopic endonasal transpterygoid approaches have been classified into five types (Carrau–Prevedello–Kassam class A–E) (Table 44.2). The boundaries of this approach are defined posteriorly by the VELPPHA area, which includes the vidian canal, Eustachian tube, foramen lacerum, petroclival fissure, and pharyngobasilar fascia. Anteriorly, this approach is limited by the pterygoid plates and the ascending process of the palatine bone. The lateral boundaries are composed of the pterygomaxillary fissure and the posterior wall of the antrum. The middle cranial fossa and greater sphenoid wing define the superior boundary.

Infratemporal fossa

Location/anatomy

The infratemporal fossa is located below the zygomatic arch, lateral to pterygopalatine fossa and deep to masseter muscle. It acts as a conduit for nerves arising from the cranial cavity, namely foramen ovale and spinosum, and supplying various structures in the face. The maxillary artery, a terminal branch of the external carotid artery, gives off the middle meningeal artery while passing through the fossa. It also hosts the pterygoid venous plexus and the maxillary vein. The nerves traversing through the infratemporal fossa include mandibular, auricular temporal, buccal, lingual, and inferior alveolar nerves.

Pathology

The pathologies associated with the infratemporal fossa include haemangioma, lymphoma, neurofibroma, and malignancies of somatic origin. Furthermore, extension of tumours from surrounding structures such as the postnasal space, orbit, palate, ear, and sinuses can occur. Metastasis from distant tumours can also be found in this region.

Boundaries

The infratemporal fossa is bound laterally by the ramus of the mandible, medially by the lateral pterygoid plate, and anteriorly by the posterior border of the maxillary sinus. The carotid sheath is located posteriorly. The roof is formed by the greater wing of sphenoid and the inferomedial floor by the lateral pterygoid plate and medial pterygoid muscle. An endoscopic approach is made via performing a wide medial maxillectomy followed by removal of the posterior wall of the maxillary sinus. The maxillary artery is then ligated exposing the infratemporal fossa. Additionally, for further exposure the lateral and medial pterygoid plates can also be drilled.

Transodontoid

Location/anatomy

The odontoid process is an upward bony projection off the axis, the second cervical vertebra. The atlas, the first cervical vertebra, rotates over it, enabling the head to rotate either to the right or the left. It is a dense part of bone which is somewhat constricted at its neck and attaches to the main body of the axis by its base. The dens is oval anteriorly at the attachment of the anterior arch of atlas. The apex is pointed, which gives attachment to the apical ligament (Figure 44.8).

Pathology

Numerous vertebrobasilar pathologies which once required open craniotomy or disfiguring transfacial approaches can now be addressed via the expanded transclival transodontoid route. These

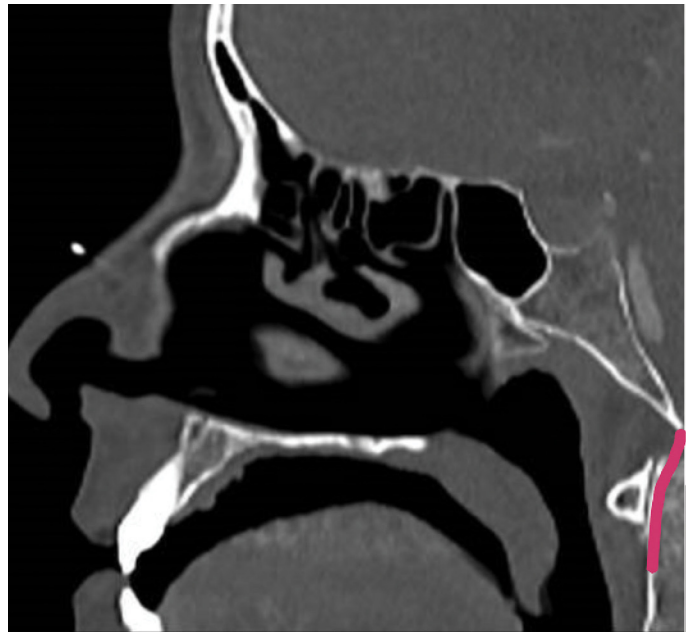

Figure 44.8 Transodontoid approach to skull base.

include, but are not limited to, basilar invagination and aneurysms of the basilar, vertebral, anterior inferior cerebellar and posterior communicating arteries, as well as pontine cavernous malformation, meningioma, fibrous dysplasia, angiosarcoma, endodermal cysts, and bulbus cavernoma.

Boundaries

The expanded transclival transodontoid approach requires first cervical vertebra anterior arch and odontoid process resection. After wide sphenoidotomy and resection of the rostrum of the sphenoid sinus is carried out, the Vidian canals are identified as the lateral limits of the surgical corridor. Within the sphenoid sinus the lateral limits are marked by the opticocarotid recess bilaterally. The posterior pharyngeal mucosa, atlanto-occipital membrane, and the longus muscles are divided. The clivus is then drilled using a high-speed drill. The apical, alar, and transverse ligaments are further resected. The first cervical vertebra anterior arch and odontoid are resected to complete the transclival transodontoid approach to reach the craniovertebral junction. The inferior aspect to this approach is limited by the hard palate. The endoscopic endonasal approach has less morbidity as it gives direct access to this region, avoiding splitting the palate or mandible compared to the transoral route.

Reconstructive options

Recent years have seen significant advances not just in greater endoscopic reaches beyond nasal sinuses but also in reconstructive options. These include local, regional, and free flaps along with synthetic dural patches and haemostatic agents which complement the reconstruction of skull base.

Local flaps

Various local flaps have been described in the literature. Here we will discuss a few commonly used flaps which have gained popularity in the past decade.

Nasoseptal flap, inferior, middle turbinate, and lateral nasal wall flaps are all based on branches of the sphenopalatine artery emerging from the sphenopalatine foramen. The nasoseptal flap is a versatile flap with superior coverage and reach. It is harvested off the nasal septum such that incisions are made 1 cm inferior to and parallel to the skull base, the second incision parallel to the floor of the nose, and varied according to the predicted size of the skull base defect. They are joined by a vertical incision anteriorly at the squamous mucosal junction at the most caudal end of the nasal septum. This flap is harvested at the onset of the surgery and placed in the nasopharynx until the reconstruction to avoid damage to the flap during the main resection.

Similarly, mucosa over the inferior and middle turbinate can be harvested and rotated on a pedicle to fill a skull base defect.

Regional flaps

These include pericranial and temporoparietal fascial flaps. These are vascular flaps which provide for larger defects not being covered by local flaps. A bicoronal scalp incision is utilized and the flap is harvested with an island of contralateral temporalis muscle. A cuff of temporalis muscle provides additional vascularized tissue that helps seal the skull base defect. The flap is then mobilized from underneath the zygomatic arch and tunnelled through the infratemporal fossa, posterior wall of maxillary sinus, and into the nasal cavity. For the pericranial flap, once raised and pedicled anteriorly, a slot along the superior aspect of the frontal sinus can be drilled creating a window for the flap to re-enter the sinonasal cavity and then be positioned in the defect to be reconstructed. Care has to be taken to ensure that there is still enough space for the pathway for the frontal sinus to drain. Further support is often given with a Foley catheter, and Surgicel along with tissue glue can be utilized to hold the flap in position.

Free flaps

Free tissue transfer, most commonly a radial forearm microvascular flap, can be utilized for multiple revision cases where previous pedicled flaps have failed. The vascular anastomoses are usually performed with the vessels in the neck.

Haemostasis

As more advances were made in reaching the far recesses in skull base surgery, so too has been seen with haemostatic agents and methods. Prior to any reconstruction after tumour dissection, maintaining haemostasis is paramount. The predominant blood supply in the nose is from the sphenopalatine artery and anterior and posterior ethmoid arteries. The sphenopalatine artery can be easily controlled in anticipation of bleeding as can the ethmoidal arteries either endonasally or via an external lynch incision. Synthetic agents such as Surgifoam or Floseal can help in haemostasis from the cavernous sinus. Bipolar cauterization can be applied to dural edges which contain small vessels. Oozing from the surface of the brain can be stopped by copious irrigation with warm saline at 49°C.

Complications

A thorough understanding of anatomy, cadaveric dissections, and purposeful preoperative planning, for example, to reduce intraoperative bleeding, are imperative to reduce the chances of complications. Recent advances in telescopic lens and better illumination and magnification have led to far fewer complications as were previously reported. Complications can be classified as intraoperative and postoperative. Intraoperative complications with significant morbidity include damage to optic nerves or major blood vessels such as the carotid arteries. Loss of vision could be either due to direct trauma to the nerve or injury to branches of the superior hypophyseal arteries supplying it.

Postoperatively, CSF leak remains the most common complication. Meticulous reconstruction in layers with the use of vascularized flaps can help reduce the leaks. Avoiding the routine use of lumbar drains unless there is a high-flow leak or in redo surgery results in better outcomes. Lumbar drains can lead to infections and need more careful postoperative ward care and observation and are therefore not used in every CSF leak repair. Other complications include nasal synechiae formation, crusting, epistaxis, and loss of olfaction. Crusting and synechiae can be reduced by saline irrigations postoperatively and regular nasal toilet as well as a more mucosal preserving surgical technique. Epistaxis is usually from branches of the sphenopalatine arteries and is fairly easy to manage. Reduction in sense of smell is due to altered nasal airflow and sometimes destruction of the olfactory epithelium by the tumour or by surgery

itself. Saddle nose deformity can be due to excessive cartilage excision and can be avoided by leaving a minimum of 1 cm strip on the dorsum and caudally.

Controversies

Open versus endoscopic approach

Some controversy exists for certain pathologies for a purely endoscopic resection versus open resection. One such example is olfactory groove meningioma—proponents of endoscopic endonasal resection advocate the minimally invasive procedure. This approach avoids extensive scars and complex craniotomies with a higher morbidity and longer hospital stay. The endoscopic approach has its caveat that is anosmia although there is a recent trend to consider a unilateral skull base resection to preserve olfaction. In contrast, the open approach can sometimes avoid olfactory dysfunction and is reported to have lower CSF leak rates compared to endoscopic approaches.

Vascular versus non-vascular reconstruction

Great strides have been made to reconstruct the skull base with and without the use of vascularized tissue. The most commonly used nasal flap for skull base reconstruction is the nasoseptal/Hadad flap (vascularized flap). There are excellent and comparable outcomes in using both non-vascularized and vascularized reconstruction of skull base defects. The only possible advantage of using a vascularized flap is considered in patients who are to have radiotherapy. Meticulous attention to detail with non-vascularized layered tissue reconstruction gives comparable CSF leak rates.

Conclusion

Endoscopic endonasal approaches to the skull base have gained widespread acceptance for pathologies involving the skull base. This is in part due to higher magnification and better visualization and also because of lower morbidity and comparable outcomes compared to conventional open procedures. With high-resolution preoperative imaging and meticulous planning, the rates of complications have significantly reduced. The cornerstone of advances in endoscopic skull base surgery is an understanding of the anatomy of the neurovascular bundles surrounding the sinonasal corridors.

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45

Anterior skull base reconstruction

Caroline Smith, Omar N. Pathmanaban, and Rajiv Bhalla

Introduction

Endoscopic endonasal techniques have evolved greatly over the past 20 years with improved instrumentation and advanced technology such as angled endoscopes, high-definition monitors, and image guidance. This enabling technology has been accompanied by an impressive expansion of endoscopic skull base operations with increased breadth and complexity of cases. Endoscopic anterior skull base reconstructions are therefore required for a wide range of anterior skull base pathologies, from small idiopathic cerebrospinal fluid (CSF) leaks to large iatrogenic defects after tumour excision, with comparable outcome rates and decreased patient morbidity when compared to open approaches. This has led to ever greater reconstructive challenges.

The use of vascularized flaps, first described by Hadad and colleagues,[1] revolutionized endoscopic anterior skull base reconstruction, decreasing the rate of CSF leak and making the successful reconstruction of larger defects possible.

This chapter will outline the principles of anterior skull base reconstruction, techniques, and commonly used grafts and vascularized flaps.

Principles of reconstruction

The aim of reconstruction is to achieve a barrier between the sinonasal cavity and the intradural space to prevent CSF leak, pneumocephalus, and, ultimately, meningitis. See **Figures 45.1 and 45.2**.

Multilayer closure has become recognized as a successful method of reconstruction for anterior skull base defects, that is, inlay graft and onlay graft/flap with obliteration of dead space if necessary. The purpose of the inlay graft is to create a water-tight seal and serve as a direct dural repair. The edges of the inlay are apposed to the inner layer of dura with a tissue sealant and long applicator, such as fibrin glue. The inlay graft has tended to be autograft, such as fascia lata plus or minus fat. This is not without donor site morbidity though, often overlooked as unimportant. The onlay may be a free mucosal graft or more commonly in the authors' practice, a vascularized flap, which is usually also secured with a suitable sealant and then supported with dressings, packing, or Foley catheter balloons depending on the site of the defect.

Decision-making

The choice of graft and/or flap depends on the size and site of the defect, low- versus high-flow intraoperative CSF leak, and patient factors including previous surgery, radiotherapy, and tissue availability. Gravitational forces anteriorly and posteroinferiorly must be meticulously considered.

Some papers quote low-flow CSF leaks are those that occur after dural opening but do not involve an opening into the ventricle or arachnoid cistern, whereas high-flow CSF leaks are defined as a defect that violates a cistern or ventricle.

Various grading systems exist. One commonly used by Esposito and colleagues[2] for transsphenoidal surgery may be applied to other ventral skull base defects (**Table 45.1**). In the senior authors' opinions, intraoperative visual assessment of the CSF leak and taking account of the other factors alluded to above aids decision-making and this becomes easier with greater experience.

Generally speaking, small defects with low-flow CSF leak can be managed successfully with free grafts such as the 'bath-plug technique' described by Wormald and McDonogh[3] using autologous fat inlay and free nasal mucosa graft onlay.

For larger defects and high-flow CSF leak, fascia lata inlay is ideally tucked 5–10 mm beyond the edges of the dural defect and the use of a vascularized flap onlay is recommended (**Figure 45.1**a–f). This is also recommended for sites where there is a high risk of CSF leak (**Box 45.1**) and in patients where healing may be an issue, for example, in patients with immunocompromise such as long-term steroid use, malnutrition, or previous radiotherapy.

Mucosa should be denuded from bone where the onlay graft will be placed to ensure a secure seal and to prevent mucocoele formation.

Free grafts

See **Table 45.2** for commonly used free grafts.

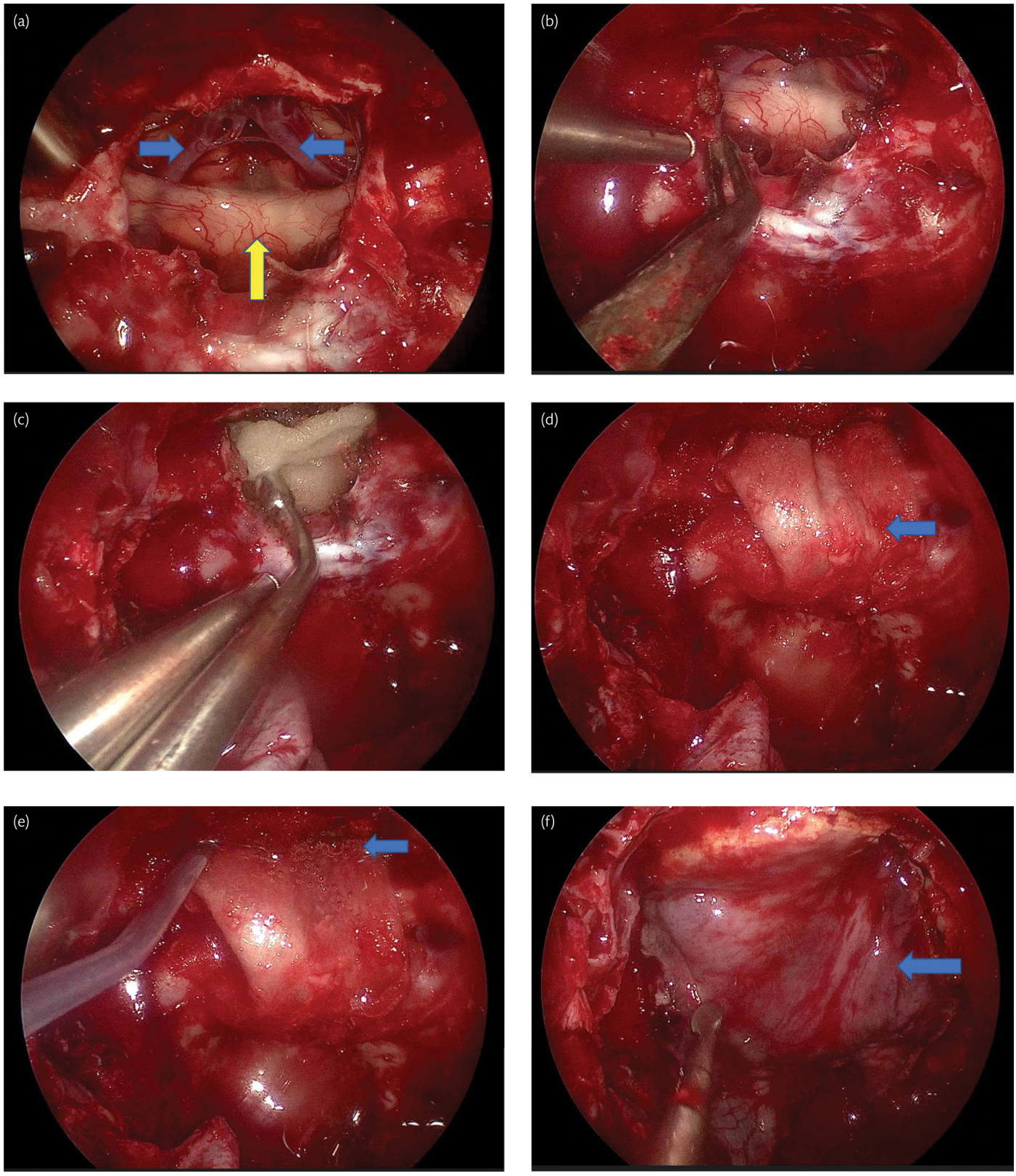

Figure 45.1 (a) Endoscopic view with 30° rigid endoscope showing large dural defect after excision of a transplanum and transtuberculum meningioma. Optic chiasm (yellow arrow) and anterior cerebral arteries (blue arrows) are visible. (b) Bipolar cautery to dural edges to achieve absolute haemostasis. (c) Floseal applied intracranially. (d) Tutoplast fascia lata (arrow) inlay. (e) Tutoplast fascia lata inlay secured with Tisseel tissue sealant (arrow). (f) Nasoseptal flap onlay (arrow) also subsequently secured with Tisseel. (g) Surgicel stamps to edges of nasoseptal flap. (h) Non-adherent Gelfoam covers the flap to prevent nasal packing from sticking. (i) Foley catheter (arrow) used to bolster the reconstruction.

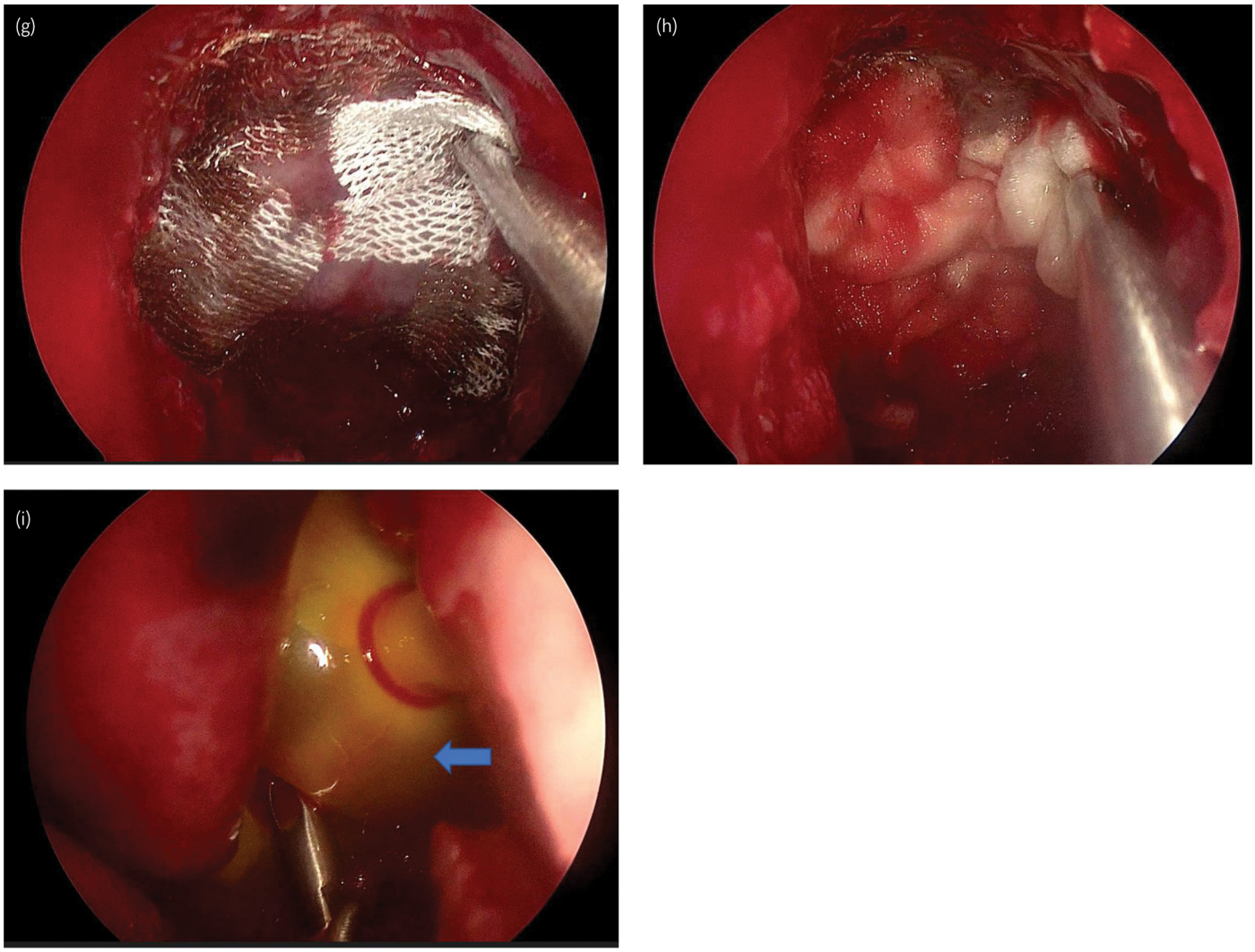

Figure 45.1 Continued

Autografts

Autografts provided the early options for skull base reconstruction and remain an excellent choice. Fat is generally harvested from the abdomen (or thigh if also harvesting fascia lata) and is useful to obliterate dead space and prevent pooling of CSF. Fat for a bath-plug technique can be harvested from the ear lobe utilizing a laterally placed incision.

Free nasal mucosal grafts can be harvested from the lateral nasal wall, posterior bony septum, floor of nose, or from the middle or inferior turbinates. It is recommended the free graft be 25% larger than the defect as there will usually be roughly 20% shrinkage.

Free bone grafts may be harvested from the vomer and perpendicular plate of ethmoid or as split calvarial grafts. They are useful when rigid repair is required, such as in morbidly obese patients with risk of brain or meningeal herniation over time. However, bone tends to resorb over time, and there is a risk of osteoradionecrosis and breakdown of the repair if the patient requires postoperative radiotherapy.

Cartilage used for the same purpose retains its integrity and longevity, and is less likely to resorb over time than bone.

Allografts and xenografts

Alternative dural substitutes exist such as the collagen matrix DuraGen (Integra Life Sciences) or Durepair (Medtronic). Both are bovine xenografts.

Alloderm (Life Cell) and Tutoplast fascia lata (RTI Surgical Holdings Inc.) are both allografts. These tissue grafts act as a scaffold for the ingrowth of fibroblasts and gradually integrate into dura mater. They have been used in reconstruction with excellent results. The senior authors use Tutoplast fascia lata routinely for reconstruction of transcribriform and transplanum/tuberculum approaches and most transclival approaches.

Prosthetic materials

Garcia-Navarro and colleagues[4] report excellent results using a gasket seal repair with Medpor porous polyethylene or similar

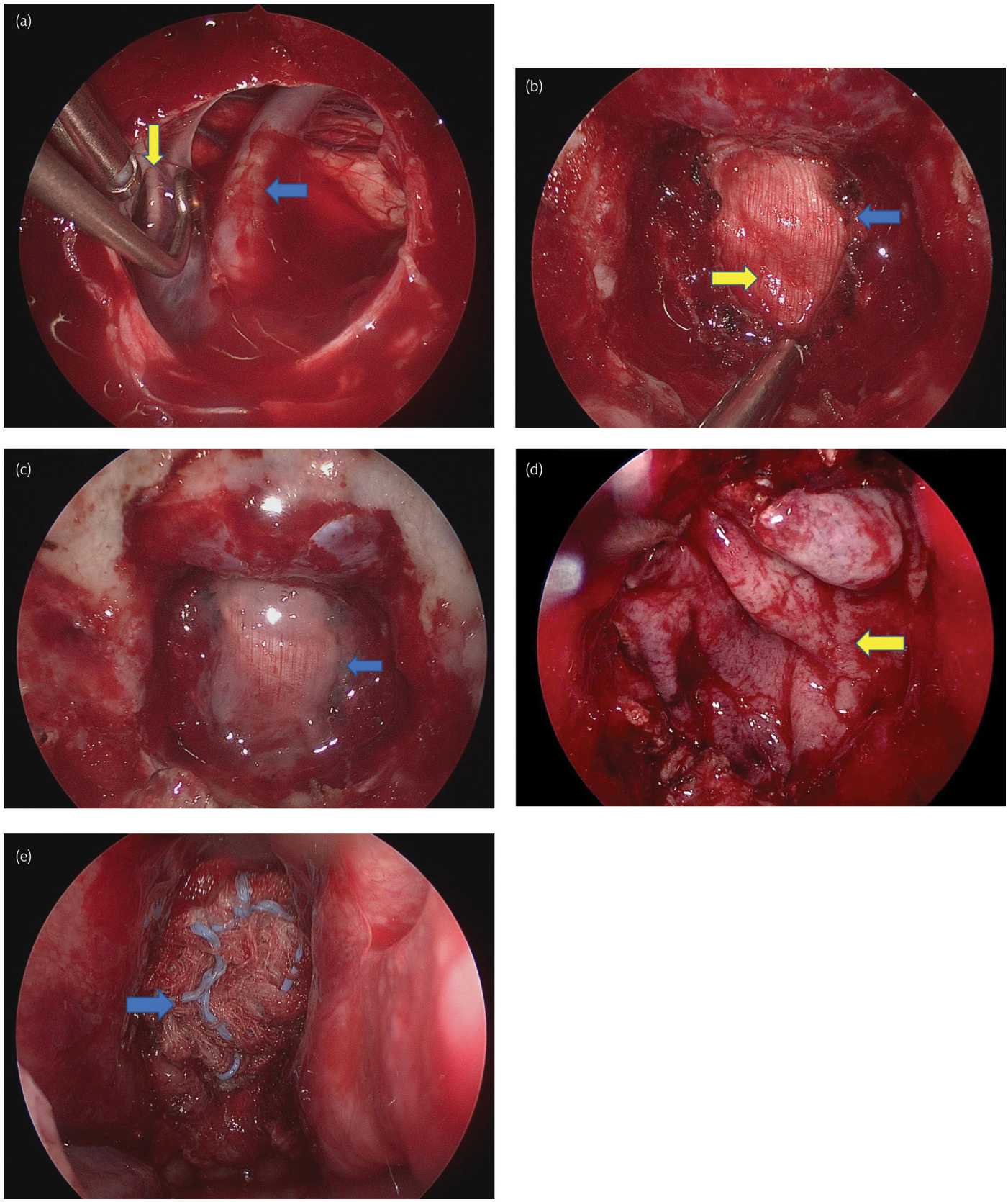

Figure 45.2 (a) Endoscopic view with 0° rigid endoscope showing large dural defect post excision of clival chordoma with basilar artery (blue arrow) and cranial nerve VI visible (yellow arrow). (b) Floseal is again applied intracranially, dural edges bipolared (blue arrow) to achieve haemostasis, and Tutoplast fascia lata inlay graft placed (yellow arrow). (c) Tutoplast fascia lata is secured with Tisseel (arrow). (d) Nasoseptal flap onlay (arrow) subsequently secured with Tisseel. (e) Ribbon gauze (arrow) in Bactroban ointment to clival defect to support nasoseptal flap (after application of Surgicel stamps to flap edge and Gelfoam to cover flap).

Table 45.1 Cerebrospinal fluid leak grading system

Grade 0	Absence of CSF leak, confirmed by Valsalva manoeuvre
Grade 1	Small 'weeping' leak confirmed by Valsalva manoeuvre without obvious or with only small diaphragmatic defect
Grade 2	Moderate CSF leak, with obvious diaphragmatic defect
Grade 3	Large CSF leak, typically created as part of extended transsphenoidal approach through the subdiaphragmatic or clival dura for tumour access

Source data from Esposito F, Dusick JR, Fatemi N, Kelly DF. Graded repair of cranial base defects and cerebrospinal fluid leaks in transsphenoidal surgery. *Oper Neurosurg (Hagerstown)*. 2007;60(4 Suppl 2):295–303.

Box 45.1 Indications for vascularized flap

- Large defect (>2 cm).
- High-flow CSF leak.
- Immunosuppression.
- Clival defect.
- Revision procedure.

products, although others are concerned about the risk of extrusion and infection.

Vascularized flaps

Intranasal flaps

Pedicled nasoseptal flap (Hadad–Bassagasteguy flap)

The workhorse flap for endoscopic anterior skull base reconstruction. This is a pedicled flap of the nasal septum mucoperiosteum and mucoperichondrium based on the nasoseptal artery, a branch of the posterior septal artery.

It is highly reliable and versatile with a wide arc of rotation, lending itself to reconstruction of extensive defects of the anterior, middle, clival, and parasellar skull base. It can be extended to involve the floor, lateral wall, and inferior turbinate as necessary.

The flap is harvested at the beginning of the procedure and is stored in the nasopharynx or maxillary antrum until the reconstructive phase of surgery.

Posteriorly pedicled middle turbinate flap

This pedicled flap is based on a branch of the sphenopalatine artery, the posterior lateral nasal artery. Mucoperiosteum is dissected from both the medial and lateral surfaces of the thin conchal bone of the middle turbinate, and the bone is subsequently removed using through-cut forceps, a back biter, and a fine ball probe.

This flap is ideally utilized for small anterior cranial fossa defects and most commonly the authors use this for repair of cribriform meningoencephalocoeles. However, these flaps may also be utilized in the planum sphenoidale and sella when other intranasal flaps are not feasible or to augment an alternative primary flap. Bilateral flaps can complement each other.

Middle turbinate flip flap

A modification of the posteriorly pedicled middle turbinate flap, mucoperiosteum is excised from the medial surface of the middle turbinate with the microdebrider and the thin conchal bone dissected and removed leaving the lateral mucoperiosteal flap. Again, this is useful for cribriform and ethmoid defects.

Posteriorly pedicled inferior turbinate flap

This is based on the inferior turbinate artery, a posterior branch of the posterior lateral nasal artery that originates from the sphenopalatine artery.

Similar to the middle turbinate flap, mucoperiosteum is dissected from the medial and lateral surfaces of the inferior conchal bone of the turbinate, and the bone is removed.

Due to the limited arc of rotation, it is most suited to posterior sella floor, parasellar, and clival defects.

Posteriorly pedicled lateral nasal wall flap (Carrau–Hadad flap)

This is based on branches of the sphenopalatine artery and encompasses the mucoperiosteum of the inferolateral wall and floor of the nasal cavity. Once elevated from the lateral nasal wall and inferior concha, the bone is excised.

This flap is suitable for reconstruction of defects extending from planum sphenoidale to the nasopharynx anteroposteriorly, and from orbital apex to orbital apex or from internal carotid artery canal to internal carotid artery canal.

Extranasal pedicled vascularized flaps

These are a good option when intranasal flaps are not feasible, for example, due to previous surgery, cancer margins, or tumour infiltration of prior reconstructive flap, or where multiple flaps are required. They are a good option in salvage surgery to repair a persistent CSF fistula, particularly when tissues have been treated with radiotherapy.

Table 45.2 Commonly used free grafts

Autograft	Allograft	Xenograft	Prosthetics
Fat	Alloderm	DuraGen	Titanium
Fascia lata	Tutoplast fascia lata	Durepair	Medpor
Nasal mucosa (onlay only)		Duraform	Silicone
Temporalis fascia		Duramatrix	
Muscle			
Bone			
Cartilage			

Transfrontal pericranial flap

This is another regional vascularized pedicled flap based on the supraorbital and supratrochlear neurovascular bundles. It is harvested via a bicoronal incision or, less commonly, by smaller step incisions with the assistance of a subgaleal endoscope, though this is of questionable advantage.

The flap is transposed into the nasal cavity via an osteotomy at the glabella just above the nasion. It is important not to twist the pedicle as the flap is delivered into the sinonasal cavity.

This flap is excellent for large defects extending anteroposteriorly from crista galli to tuberculum sella, and laterally from orbit to orbit. It is extremely robust, reliable, and is likely to have been out of a previous radiotherapy field, which can be extremely useful in revision cases.

Longer flaps will reach down to the craniocervical junction, although this is rarely required; the authors find a temporoparietal fascial flap optimal for the clivus down.

Transpterygoid temporoparietal fascial flap

This is a regional vascularized pedicled flap based on the anterior branch of the superficial temporal artery and can cover large defects.

It is harvested via a hemicoronal incision and transposed into the nasal cavity via a transpterygoid tunnel.

There is a risk of injury to the temporal branch of the facial nerve and care should be taken to elevate this in the superficial fat pad.

This flap is particularly useful for ventral defects in the central and posterior skull base.

Pedicled free flaps

These may need to be utilized when other pedicled reconstructive options are exhausted and where extensive reconstruction is required.

An adipofascial-only flap is used for endoscopic reconstruction.

Microvascular anastomosis is usually to facial vessels, with tunnelling to the operative site via an infratemporal fossa, retroantral, retropharyngeal corridor.

Radial forearm fasciocutaneous flap

This is the go-to free flap for complex endoscopic skull base reconstruction in the authors' practice. Vascular supply is the radial artery and vein.

This flap is a pliable piece of tissue with a long pedicle, allowing usage of neck vessels as recipients.

Radial artery occlusion testing should be carried out preoperatively to avoid the complication of hand ischaemia.

Rectus abdominis myocutaneous flap

Vascular supply is the epigastric artery and vein. The major disadvantage is the bulkiness due to the width of subcutaneous tissue. There is also a risk of hernia due to abdominal wall weakness.

Anterolateral thigh fasciocutaneous flap

Vascular supply is the lateral femoral circumflex artery and vein. This also has the advantage of a long pedicle. However, this can also be a very bulky flap.

Securing the reconstruction

Fixation of the tissue against the defect is critical to its healing. Each layer is secured in place with sealant or fibrin glue such as Tisseel (Baxter), DuraSeal (Integra), or Adherus (Stryker). The authors also use stamps of Surgicel along the edges of the flap.

Larger reconstructions using vascularized flaps may require bolstering with nasal packing to prevent gravitational pull and early flap migration.

It is important to cover the flap with absorbable non-adherent material such as Gelfoam (Pfizer) to prevent removable nasal packing from sticking to the flap and to distribute any localized pressure from packing. The senior authors use ribbon gauze in Bactroban nasal ointment for sphenoid and clival defects, and Foley catheters with Rapid Rhinos (Arthrocare) for cribriform defects (**Figures 45.1 and 45.2**).

Postoperative care

The authors rarely use lumbar drainage at the outset, but prefer to use this if there is a subsequent leak. For high-risk defects, Rapid Rhinos are removed after 72 hours, Foley catheters after 1 week, and ribbon gauze 4 weeks postoperatively. This is tailored and reduced for each individual patient's defect.

For high-risk patients, the authors may also cover with acetazolamide to reduce CSF production, particularly in patients with raised body mass index or idiopathic intracranial hypertension.

The patient is advised to avoid bending, lifting, and straining for 6 weeks, and to avoid blowing their nose for the same time. It is also recommended that they sneeze with their mouth open. Codeine analgesics are avoided, and a stool softener is prescribed.

Saline nasal spray is prescribed regularly to help with nasal hygiene, crusting, and outpatient debridement. The authors feel this is very important for comfort, to reduce the risk of infection (either local osteitis or from sinus ostial obstruction), and to facilitate the action of topical preparations, such as saline douche and topical anti-inflammatories.

The nasal cavity is examined in the outpatient department 3–4 weeks postoperatively with flexible or rigid nasendoscopy to ensure healing is progressing as expected, absence of flap necrosis particularly at the margins, absence of CSF fistula, and to allow debridement as alluded to above if necessary.

Complications

Cerebrospinal fluid leak

Prior to the use of vascularized flaps, the postoperative CSF leak rate in expanded endoscopic endonasal procedures was as high as 50%. Now the rate in the literature varies from 5% to 12% with an average of 9%.

If CSF rhinorrhoea is suspected, a sample should be sent for beta-2 transferrin and the patient treated as for a CSF leak if there will be a delay in obtaining the result.

The authors' stepwise management for a low-flow early CSF leak is as follows:

1. Exclude hydrocephalus, CSF diversion if necessary.
2. Lumbar drainage (10 mL/hour) for up to 7 days with challenge at 3–5 days.
3. With or without acetazolamide orally.
4. Re-exploration and repair if ongoing leak despite lumbar

If it is a high-flow CSF leak, consider early re-exploration as this is more likely to be a flap complication. Possibilities include the following:

- Non-adherence—haematoma formation, inadequate bolstering, gravitational pull, tension at the origin of the pedicle, or inadequate bone drilling are all possible causes.
- Flap necrosis—due to a compromised pedicle or poor healing secondary to radiotherapy, immunocompromise, malnutrition.

Meningitis

Usually associated with CSF leak, but may occur without a leak or with an occult leak. There should be a low threshold for contrasted brain imaging and diagnostic lumbar puncture.

Treat with appropriate antibiotics in line with institutional antimicrobial policy and monitor inflammatory markers and radiological changes.

Brain abscess and subdural empyema are very unusual, but must be excluded.

Pneumocephalus

Small volume pneumocephalus is normal after major skull base procedures and even moderate volume bifrontal pneumocephalus can be seen, without major concern. High-flow oxygen may expedite resorption.

Tension pneumocephalus is very rare but may occur and requires urgent neurosurgical management followed by re-exploration of the skull base repair.

Controversies

Lumbar drains

Lumbar drains reduce intracranial pressure by continuous CSF drainage and take the CSF pulsation off the skull base repair. In the setting of postoperative skull base reconstruction, this is thought to facilitate wound healing due to decreased tension and may improve success rate by decreasing the chance of persistent CSF fistula.

With the increased dependability of multi-layered closure and vascularized flaps, the need for routine lumbar drainage is often a point of debate.

The senior authors do not routinely use lumbar drainage but, they are used in the following settings:

- Treatment of early postoperative CSF leak (days 1–7).
- High-flow CSF leak with high risk of failure due to clinical or anatomical reasons.
- To permit use of intraoperative intrathecal fluorescein or contrast for cisternogram, although this is usually done with a lumbar puncture needle unless lumbar drainage is anticipated afterwards.
- Known or suspected intracranial hypertension.
- High risk defects with raised body mass index.

Operating in infection

On the rare occasion where a prospective endoscopic skull base patient has concomitant acute or chronic rhinosinusitis, the authors would recommend managing this in advance of the skull base procedure medically and/or surgically if time permits. If, however, the pathology is time dependent, the senior authors have proceeded to manage both simultaneously without adverse outcome.

Autograft versus allograft

The use of autografts versus allografts in anterior skull base reconstruction will often come down to the surgeon's/patient's preference and availability, as both have been used frequently with low rate of CSF leak.

Autografts have their advantages—they are readily available, cheap, no risk of host immune response, and there are rarely any issues with consent. However, there is potential morbidity of the donor site, which should be enquired after. For fascia lata this includes pain or awareness during exercise or climbing stairs, muscle prolapse or herniation, and lateral cutaneous thigh hypoaesthesia which can be very bothersome for the patient and prolong inpatient stay. Perioperative haematoma is also not unusual and fascia harvest should include meticulous haemostasis and repair of the fascial defect using absorbable suture. Temporalis fascia or muscle harvest can cause awareness during chewing for some time after.

The advantages of using dural substitutes derived from allografts or bovine xenografts mentioned previously are they are 'off the shelf', saving operating time required for graft harvest, and there is zero donor site morbidity. There is, however, additional cost but this may be offset by a reduced inpatient hospital stay.

It is very important to be aware of the origin of the tissue used. Some patients may not agree to the use of bovine or cadaver-derived grafts due to religious reasons, and this should be respected.

Whichever technique is employed, this should be discussed fully with the patient and documented in the notes and on the consent form.

Tips and pearls

- Multilayer closure combined with a vascularized flap is the gold standard for large anterior skull base defects.
- Ensure as large a flap as possible is harvested—this can be trimmed if necessary.
- Protect the flap pedicle particularly when drilling the sphenoid rostrum.
- Ensure the pedicle is not twisted to ensure flap viability.
- Ensure mucosal surface of flap is correctly orientated away from the brain.
- Denude mucosa from surrounding bone to achieve a good seal and prevent mucocoele formation.
- Use nasal packing to bolster flaps and avoid early migration.

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SECTION 4

Head and neck surgery

46. **Principles of clinical assessment and management of the head and neck patient** *343*
Jemy Jose and B. Y. Winson Wong

47. **Dysphonia** *348*
Yakubu Gadzama Karagama

48. **Dysphagia** *356*
Somiah Siddiq

49. **Sore throat** *368*
Jenny Montgomery

50. **Clinical assessment of neck lumps** *375*
David Walker and Cameron Davies-Husband

51. **Snoring and sleep apnoea** *386*
Khalid Ghufoor and Jahangir Ahmed

52. **Acute and subacute airway obstruction** *391*
Navin Mani, Amy Teasdale, and Sofia Clegg

53. **Head and neck trauma** *397*
Jaiganesh Manickavasagam and Kishore Shekar

54. **Oral ulceration: a pragmatic guide to diagnosis and management** *406*
Stefano Fedele and Stephen Porter

55. **Laryngeal precancer and cancer** *411*
David Hamilton and Anna Thompson

56. **Oropharyngeal cancer** *419*
Zaid Awad and Dorothy M. Gujral

57. **Oral precancer and cancer** *428*
Deepa Nair, Florida Sharin, and Sarbani Ghosh Laskar

58. **Salivary gland tumours** *438*
Khanh K. Nguyen and Nitin A. Pagedar

59. **Thyroid cancer** *444*
Hannah Nieto and Neil Sharma

60. **Hypopharyngeal cancer** *450*
Laura Warner and Kee Wong

61. **Nasopharyngeal cancer** *456*
Shreerang A. Bhide and Pablo Nenclares

62. **Adult deep neck space infections** *465*
Zi-Wei Liu and Abdul Ahmed

63. **Parathyroid disorders** *470*
Ekpemi Irune, Brian Fish, and Ruth Casey

64. **Principles of head and neck reconstruction** *477*
Vijay V. Haribhakti

65. **Non-surgical treatments for head and neck cancer** *491*
Muhammad Shahid Iqbal and Charles Kelly

66. **Rehabilitation of the head and neck cancer patient** *498*
Sarah Stephen and Joanne Patterson

67. **Benign neck lumps** *508*
Samantha Hassid, Georges Lawson, and Sébastien Van Der Vorst

68. **Head and neck pathology** *520*
Max Robinson and Selvam Thavaraj

69. **Head and neck radiology** *527*
Derfel ap Dafydd

70. **Tracheostomy** *540*
Jack Faulkner, Aleix Rovira, and Asit Arora

71. **Phonosurgery** *547*
Kate Heathcote

72. **Movement disorders of the larynx** *554*
Julia Selby

46

Principles of clinical assessment and management of the head and neck patient

Jemy Jose and B.Y. Winson Wong

Introduction

The word 'cancer' is in most clinicians' minds when a referral is made for a head and neck problem. This chapter offers a high-level view of the clinical assessment and management of head and neck patients. We will discuss the presenting symptoms, the relevant history including assessment of comorbidity, focused clinical examination, order of investigations in different scenarios, and new evolving ways that patients might be assessed. The chapter will then focus on some of the models of care for diagnosis and treatment and emphasize the importance of collaborative and multidisciplinary working.

Symptoms

In the UK, there is a list of symptoms that could be red flags for head and neck cancer (**Box 46.1**).

A short duration of symptoms and progressive worsening could be considered as main features of a serious pathology; however, several other benign and recalcitrant conditions of the upper aerodigestive tract and neck share these symptoms. The art of good clinical assessment is in differentiating between the two.

Box 46.1 Symptoms that could be red flags for head and neck cancer

- Persistent hoarseness for more than 3 weeks.
- Persistent sore throat for more than 3 weeks.
- Progressive dysphagia.
- Referred otalgia, especially associated with painful swallowing.
- Lump in the neck, new or changing over past 3–6 weeks.
- Persistent swelling of submandibular or parotid gland.
- Unilateral nasal obstruction along with bleeding and/or unilateral hearing loss.
- Recent-onset thyroid lump.

Urgent referral pathway

Following assessment by the primary care practitioner, patients with symptoms included in **Box 46.1** are referred to a specialist in secondary care. The referral guidelines mandate that these patients are seen within 2 weeks from the date of referral, that all investigations are performed, and a cancer diagnosis is established or ruled out within 28 days. With increasing demand for finite resources, be it clinic appointments to see the specialist, or for imaging, or for biopsies and specialist reporting of these procedures, achieving these targets can be challenging for several reasons: (1) the 'cancer' may not be obvious at the outset on clinical examination or indeed it might have to pass through a longer diagnostic route of an unknown primary with metastatic cancer; (2) poor specificity of the referral criteria leading to excessive referrals that overwhelm capacity. Ongoing research will target this need, leading to a better triaging system.

Clinic set-up in secondary care

Patients will need to be assessed in a set-up where a comprehensive examination of the head and neck area can be performed. Adequate instrumentation, lighting, suction, examination chair, flexible and rigid endoscopes, clinical photo documentation, and archiving system are mandatory. Access to adjunctive methods such as chromoendoscopy and stroboscopy is desirable. The services of other professionals, such as dietitians, speech and language therapists, and clinical nurse specialists, are often required for this patient group.

History

A detailed but focused history taking should narrow down the following factors:

- Subsite (nasal/naso/oro/hypopharyngeal/laryngeal/salivary/thyroid etc.) of the upper aerodigestive tract to focus one's attention.

Box 46.2 Commonly used indices for estimation of comorbidity and performance status

- World Health Organization performance status.
- Adult Comorbidity Evaluation 27 score (ACE-27).
- Washington University Head and Neck Comorbidity Index.
- Elixhauser Comorbidity Index.
- Royal College of Surgeons Charlson score.
- Combined Comorbidity Index.
- World Health Organization fitness score.
- Karnofsky Performance Scale.

- The risk/suspicion of a serious diagnosis.
- Whether to look for an infraclavicular cause or a systemic cause presenting as an ear, nose, and throat (ENT) symptom.

Onset, duration, and rate of progression of symptoms are important. Personal habits, especially the aetiological associations of smoking and alcohol use, need to be documented objectively—for example, as pack-years for smoking and units/week for alcohol use. Sexual history is relevant if human papilloma virus-related malignancy is suspected. Past history and family history of cancers (especially thyroid) will need to be explored in addition to detailed medical history and list of medications. Systemic symptoms related to non-head and neck regions should be elucidated; examples include B symptoms in patients with multiple, bilateral lymph nodes, and chest, breast, and abdominal symptoms in patients with left supraclavicular lymphadenopathy.

It is important at this early stage to assess and document:

- the patient's general health and activity (both work and leisure related), as this can influence the decision to offer treatments associated with significant side effects
- the patient's nutritional status as dietician input may be required from the first appointment to support and improve nutrition
- their ability to make an informed choice if given a choice of treatment options
- their family/social support network.

In the event of a cancer diagnosis, it is considered good practice to objectively score performance status and comorbidity using one of the instruments listed in **Box 46.2**. Comorbidity and performance status has been established as an independent predictor of overall survival in head and neck cancer patients. All the above information will influence the decision-making process, which should be centred around the patient.

Additional notes on detailed evaluation of the above-mentioned symptoms (dysphagia, hoarseness, neck lump, etc.) can be found in the respective chapters.

Examination

Detailed head and neck assessment involves examination of the entire upper aerodigestive tract, palpation of neck nodes, examination of nodes in other regions and hepatosplenomegaly where appropriate, recording of cranial nerve function, and examination of salivary glands and thyroid glands. Flexible nasendoscopy will be necessary for the majority of these patients. Increasingly, the following additional services are being offered in many units:

- *Virtual chromoendoscopy*: an endoscopic imaging technology that enhances visualization of the abnormal vasculature pattern seen in dysplastic and cancerous lesions, distinguishing these lesions from the surrounding normal tissue, a facility not afforded by conventional white light endoscopy. These are available in most modern imaging stack systems.
- *Clinic-based endoscopic biopsy*: biopsy forceps that can be passed through the side port of a flexible channelled nasendoscope is used for this intervention. The obvious advantage is procurement of a specimen for histology in the outpatient setting, which is valuable for patients who are not suitable for a general anaesthetic procedure, and rapid diagnostic throughput without delays in awaiting an operating theatre slot. Disadvantages include inadequate assessment of the tumour extent and inability to assess suitability for transoral surgery.
- *Transnasal oesophagoscopy*: up to a third of patients who present with throat symptoms can have pathology within the oesophagus. Depending on logistics in the clinical setting, this procedure can be offered at the same visit or later for the relevant patient group.

Many patients referred with a head and neck symptom can be discharged with a definitive diagnosis or assurance after a single consultation. Few might require a telephone call or a letter with imaging results. Those patients suspected of a serious pathology should be referred on to a specialist or a specialist clinic to be reviewed with investigations. Depending on the symptom or the working diagnosis, several service models of joint working exist to offer excellent patient care, some of which are described below.

For obvious reasons, patients with a clearly malignant-appearing lesion or one that is highly suspicious for malignancy should be prioritized to the next step of investigation. Depending on the local policy, early referral to or seeking advice from the head and neck multidisciplinary team is recommended.

One-stop neck lump clinic

This is a specialized set-up aimed at arriving at a definitive or a most likely diagnosis in patients presenting with a neck lump. The clinical team usually comprises an ENT surgeon, an ultrasonographer/radiologist, and a cytopathologist/laboratory technician working jointly in a clinic that should be streamlined to see exclusively these patients. After a thorough ENT examination, an ultrasound scan is performed to determine the origin of the lump and its character. Ultrasound-guided needle cytology is performed from an area that is deemed most representative of the lesion. A laboratory technician or a cytopathologist can confirm adequacy of the sample for a diagnosis, and if inadequate, sampling can be repeated. This is particularly relevant in swellings that are primarily cystic (e.g. thyroid lumps, branchial cysts, and cystic/necrotic lymph nodes), wherein a sample needs to be obtained from the capsule of the cyst. Often a confirmatory diagnosis can be reached at the same sitting. The obvious advantage of this set-up is the rapidity of the diagnostic process. Several models exist, including where the specialists are in adjacent clinical rooms to a flexible association wherein patients move from

one department to another, while the principle of reaching a provisional diagnosis in one visit is met. More recently, and with core biopsies of lumps garnering momentum, adequacy of sample has become less of an issue. However, cytology is still a good adjunct to thyroid clinics where this remains the mainstay for diagnosis.

Other joint clinic models

Some of the common clinic models for onward referral include the following:

- *Joint voice clinics*: a team of a laryngologist, a speech and language therapist, and often a vocal coach. This is particularly useful for patients with voice and undifferentiated throat symptoms, where a benign diagnosis is suspected. Examples include vocal nodules, vocal cord polyps and other benign-appearing lesions of the voice box, and intractable globus pharyngeus symptoms.
- *Chronic cough clinic*: this model will usually comprise a chest physician, an ENT surgeon, a speech and language therapist, and a chest physiotherapist. Patients with recalcitrant cough leading to voice, throat, or chest symptoms may benefit from oesophageal manometry, pH studies, and upper gastrointestinal endoscopy to evaluate reflux and a chest-related cause.
- *Transnasal oesophagoscopy service*: usually performed by an ENT surgeon and sometimes accompanied by an upper gastrointestinal physician. Useful to rule out an oesophageal lesion in a small but significant number of patients presenting with globus-like symptoms.

Imaging sequence

Imaging protocols vary in individual units depending on the resources available locally and the preferences of the radiology team. Imaging should preferably be performed prior to biopsy to prevent post-intervention oedema and inflammation that makes interpretation of images difficult and can potentially upstage the tumour. The imaging modalities are discussed in greater detail in Chapter 69.

The multidisciplinary team and the cancer patient

Multidisciplinary working confers significant advantages in the management of complex clinical problems, with our patients being the ultimate beneficiaries. In addition to coordination of care, this also serves as a governance mechanism ensuring delivery of high-quality care.

This is especially true in the management of head and neck cancer patients, who need the expertise from several professional groups (**Box 46.3**) for optimal outcomes. However, multidisciplinary working is increasingly seen in the provision of laryngology and dysphagia services.

Head and neck cancer treatment will have a significant impact on the lives of patients and their carers/family, in addition to affecting their social and work environments. Except for the earliest stages of glottic or oral cancer treated with upfront surgery, the diagnostic journey, treatment period, and recovery phase for most head and neck malignancies takes a minimum of 12 weeks. For patients with advanced cancer who need multimodality therapy (e.g. surgery followed by adjuvant radiation), this can often mean 6 months for recovery from the acute side effects.

Box 46.3 Professional groups in the head and neck cancer multidisciplinary team

Core team

- Surgeons (ENT, oral and maxillofacial, plastic surgery).
- Clinical oncologists.
- Restorative dentist.
- Pathologists.
- Radiologist.
- Clinical nurse specialists.
- Speech and language therapist.
- Senior nursing staff from the head and neck ward.
- Palliative care specialist (doctor or nurse), who should work with palliative care services in the community.
- Dietitian.
- Team secretary.
- Data manager.
- Multidisciplinary team coordinator.
- Endocrinologist.

Extended team

- Other specialist surgeons.
- Anaesthetist.
- Gastroenterologists, radiologists, surgeons, and other health professionals with expertise in gastrostomy creation, feeding tube placement, and support for patients who require tube feeding.
- Ophthalmologist.
- Pain management specialist.
- Nuclear medicine specialist.
- Therapeutic radiographer.
- Maxillofacial/dental technician.
- Dental hygienist.
- Social worker.
- Benefits advisor.
- Liaison psychiatrist.
- Clinical psychologist.
- Counsellor.
- Physiotherapist.
- Occupational therapist.

Given the complexity of diagnosis and treatment, the information needs of head and neck cancer patients are significant, both to consent to any planned treatment and to understand the short- and long-term implications of cancer therapy. It is impossible for such volumes of information to be absorbed in one session, and there is an ongoing need for information provision.

Furthermore, once treatment is complete, monitoring for late effects and follow-up often leads to several years of specialized clinical care and imaging during clinical visits for this patient group. With the advent of immunotherapy regimens that some patients will receive for years, head and neck cancer patients need prolonged clinical input. Such treatment, by necessity, will be led by several professional groups with exclusive expertise within their fields. At all stages, it is vital that the multiprofessional group is transparent about the processes of care, the side effects and toxicity of treatment, and the outcomes from cancer therapy; this often leads to a patient who is much more satisfied with the processes of care and engenders

Box 46.4 An example cancer patient pathway

Nutritional evaluation by specialist dietitians with an interest in head and neck oncology should be carried out prior to initial treatment taking into account the patient's pretreatment nutritional status, disease site and extent, and radiotherapy target area volume. In the treatment for locally advanced head and neck cancer, it is very likely that the patient will require tube feeding during and soon after the course of treatment. Options include either having nasogastric tube feeding when swallowing becomes problematic, or proactive placement of a gastrostomy tube, before any treatment begins. This should be discussed in detail and patients should be involved in this decision-making process. Even after insertion of a feeding tube, patients should be encouraged to continue to swallow liquids and soft diet for as long as possible to reduce the rate of late swallowing dysfunction. Nutritional status should be monitored throughout the course of therapy. Oral care and pain management should be sought actively in order to address expected symptoms of pain and mucositis. It is not uncommon for these patients to need hospital admission for symptom management and support, especially in the second half of their treatment course.

trust in the professionals looking after them. Two exemplar patient pathways, from diagnosis to follow-up, are set out in Box 46.4 and Table 46.1, with the various professionals involved in care as shown in Box 46.3.

Diagnostic stage

This stage is beset by anxiety for the patient and the carers. The need for investigations and the likely wait time for the diagnostic tests to be reported should be explained to the service user. In some complex diagnostic pathways (e.g. the unknown primary cancer) significant diagnostic delays can occur owing to the number of tests and the need for specialized assessment under general anaesthesia (see Chapter 3), and the rationale for such tests should be conveyed to the patient. It is important to relay any suspicions of malignancy sensitively to the patient as soon as possible. Most patients will be expecting this news, particularly if there is significant symptomatology. Being honest about the clinical assessment, findings, and suspicions allows open discussion to build a rapport between patient and clinician, improves patient compliance with investigations, and enables early management of symptoms and modification of risk factors such as smoking and alcohol use.

Treatment stage

Involving patients in the decision-making process is vital and it is useful for clinicians to discuss likely treatment options with patients in advance of multidisciplinary team meetings so that the patients' preferences are known, as this frequently influences discussion. While most patients will accept the expert decision for cancer treatment, it is not uncommon for some to have very firm views, which, even if contrary to best practice, must be respected and the treating team will have to alter treatment processes accordingly.

Once a multidisciplinary team decision for treatment has been made, often the clinician involved in the first-line therapy will see the patient. Based on the treatment process, the toxicity profile, and the patient's functional status and needs, additional professional groups will often see the patient at the same sitting or at a later stage. In instances where more than one option for treatment is available, the patient should be seen by all treating teams, and full and frank discussions should be had about the pros and cons of each treatment modality. Patient preferences must be elicited to try and understand their values and the importance they ascribe to oncological and functional outcomes. This often provides clarity for the treatment team, who can then tailor treatment accordingly.

Post-treatment and follow-up stage

In the early months after treatment, be it surgery or radiation therapy, follow-up is directed at symptom relief and reassurance. Based on well-defined protocols, additional imaging is performed at predetermined times. Often, for rehabilitation, specialist professional groups

Table 46.1 Example of a cancer patient journey from diagnosis to follow-up (55 years old with T3N1 human papillomavirus-positive oropharyngeal squamous cell carcinoma having chemoradiotherapy)

Phase of journey	Professional seen	Reasons
Diagnosis	• Surgeon • Macmillan nurse	• Full clinical assessment and biopsy to be taken by surgeon to achieve a diagnosis • Cancer nursing support begins when diagnosis is given
Treatment	• Oncologist • Dietician • Speech and language therapist • Restorative dentist • Macmillan nurse	• Oncologist to discuss treatment details with patient • Dental assessment and treatment to be carried out to prevent dental complications • Assessment of nutrition and swallow is provided pretreatment with support continued throughout cancer treatment
Follow-up	• Surgeon • Dietician • Speech and language therapist • MacMillan nurse	• Post-treatment scans to be performed • Patient to be followed up by surgeon after treatment completed to detect any cancer recurrence
Recurrence of nodal disease in neck at 18 months	• Radiologist • Surgeon • Oncologist • Dietician • Speech and language therapist • Macmillan nurse	• Ultrasound assessment and biopsy might be performed by radiologist following cross-sectional imagining • Following discussion in multidisciplinary team meeting, if appropriate, salvage neck dissection to be offered by surgeon • If salvage surgery is not appropriate, palliative systemic treatment can be considered by oncologist

will need to offer more input. For instance, patients who have had radiation therapy with concomitant chemotherapy and poor swallow may experience dysphagia and will need the input of a swallowing rehabilitation expert. After neck dissection, an individual with a sore shoulder will need an expert physiotherapist. Patients with cancer that recurs and requires long-term immunotherapy will need a clinical or a medical oncologist's input.

Future perspectives

In a world with increasing demand and short supply of specialists, use of a triaging process that will identify patients with higher risk to see a cancer specialist is being investigated. An audit of telephone triage of referrals using a risk calculator on patients with the red flag symptoms listed in **Box 46.1** has shown promising results. Further studies are awaited to trial whether this process can be facilitated in primary care or indeed by patients themselves.

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47

Dysphonia

Yakubu Gadzama Karagama

Introduction

Dysphonia can be defined as a change in voice as a result of anatomical, pathological, functional, or physiological abnormalities in the larynx, which can affect the patient physically, emotionally, and functionally. Patients and physicians sometimes describe dysphonia as hoarseness or as a husky, croaky, or gravelly voice. The anatomy and physiology of the larynx is one of the most complex among body organs, and thus understanding this will help with a thorough clinical and objective assessment to establish a firm diagnosis and plan effective management strategies. This chapter discusses the basic anatomy of the larynx and assessment of patient with dysphonia and the differential diagnoses.

Anatomy of the larynx

The larynx comprises three unpaired and three paired cartilages, and extrinsic and intrinsic muscles, with their unique neural and vascular network.

Unpaired cartilages

Thyroid cartilage

The thyroid cartilage is the largest of all the laryngeal cartilages and has two parts: the left and right thyroid laminae that are attached to each other in the midline to form a prominence in front of the neck, often referred to as 'Adam's apple'. Posteriorly, the laminae have free borders but extend superiorly as the superior thyroid horn (cornua) and inferiorly as the inferior thyroid horn. The superior thyroid horn is attached to the hyoid bone through the thyrohyoid ligament and the inferior horn articulates with the cricoid cartilage to form a synovial joint called the cricothyroid joint. The cricothyroid joint is responsible for the rocking movement between the cricoid and thyroid to produce an increase and decrease in vocal cord length and tension, thereby causing changes in pitch during speech and phonation.

Cricoid cartilage

The cricoid cartilage is a hyaline cartilage and is the only complete cartilaginous ring in the larynx, broader posteriorly, giving it the appearance of a signet ring. Posteriorly, the superior border hosts the paired arytenoid cartilages to create the cricoarytenoid joints, and on its side to the thyroid cartilage, where the articulation creates the cricothyroid joints. These are synovial joints and are prone to inflammatory diseases such as rheumatoid arthritis. These joints can dislocate following trauma to the neck resulting in dysphonia and in some cases stridor. The cricoid cartilage marks the inferior border of the larynx.

Epiglottis

The epiglottis is a leaf-shaped elastic cartilage that is attached to the internal surface of the thyroid cartilage in the midline through a narrow stalk. It has a superior broader that overhangs the laryngeal inlet and protects the inlet from aspiration during deglutition. The epiglottis marks the superior border of the larynx.

Paired cartilages

Arytenoid cartilages

These are pyramidal in shape, consisting of the apex, base, vocal process, and muscular process. The vocal process is attached to the vocal ligament anteriorly while the muscular process provides the attachment for the lateral and posterior cricoarytenoid muscles. The arytenoid articulates with the superior border of the cricoid cartilage forming a synovial joint, which allows movement and rotation of the arytenoid over the cricoid in both anteroposterior and lateral directions with the aid of the corresponding intrinsic muscles of the larynx, namely the lateral and posterior cricoarytenoid muscles. The interarytenoid muscles also play a part in adductor movement of the arytenoids.

Corniculate cartilages

These are small cartilages that attach to the apex of the arytenoids and have no defined function.

Cuneiform cartilages

The cuneiform are minor cartilages embedded within the aryepiglottic fold, that may add strength to the aryepiglottic fold.

Muscles of the larynx

Extrinsic muscles of the larynx

These are called extrinsic because of their attachment to the external surface of the laryngeal cartilages. The thyrohyoid attaches the thyroid cartilage superiorly (laryngeal elevators) to the hyoid bone and

the sternothyroid (laryngeal depressors) attaches the thyroid cartilage inferiorly to the sternum.

Intrinsic muscles of the larynx

The intrinsic muscles of the larynx are attached to the cartilages within the laryngeal cartilage framework. These are the adductors (lateral cricoarytenoid and inter/oblique arytenoid muscles and the thyroarytenoid muscles) and the abductors (posterior cricoarytenoid muscles).

Laryngeal nerves

The recurrent laryngeal nerve, a branch of the vagus nerve, supplies all the intrinsic muscles of the larynx apart from the cricothyroid muscles (which tense the vocal cords), which are supplied by the superior laryngeal nerve, also a branch of the vagus nerve.

The recurrent laryngeal nerve also provides sensory supply to the laryngeal mucosa inferior to the vocal cord and the superior laryngeal nerve supplies sensation above the vocal cord level.

Blood supply

The blood supply of the larynx arises from the superior and inferior laryngeal arteries; while the former arises from the external carotid system, the latter is a branch of the thyrocervical trunk, which arises from the subclavian system. Superior and inferior laryngeal veins drain into the internal jugular and the brachiocephalic veins respectively.

Histological layers of the vocal cord

The vocal cord is made up of five histological layers:

- Epithelium (mucosa).
- Lamina propria comprising of three layers:
  - Superficial layer (jelly-like substance that makes up the Reinke's space).
  - Intermediate layer (elastic, fibrous substance like rubber bands).
  - Deep layer (thread-like collagenous layer).
- Vocalis muscle: stiff and provides support for the vibrating vocal cords.

The epithelium and superficial layers are referred to the cover, the intermediate and deep layers the transition, and the vocalis muscle the body. The jelly-like superficial layer allows frictionless motion of the mucosal layer during vocal cord vibration, often referred to as the mucosal wave. This is only visible with the aid of a stroboscope or a high-speed fibre endoscopic camera as the vocal cords vibrate 100–150 times per seconds in males and 150–250 times per second in females, which cannot be captured by the naked eye.

Physiology of phonation

Phonation occurs through various stages and functional interactions between the respiratory system involving the lungs, trachea, larynx, pharynx, oral cavity, and nasal cavity. The lungs provide the airflow which makes the vocal cords vibrate and generate the sound. This sound is then articulated to form speech by the articulators in the oral cavity (tongue, teeth, palate, buccal cavity) while the pharynx, nasal cavity, and sinuses provide the vocal resonance. While the recurrent laryngeal nerve is responsible for the adduction and abduction of the vocal cords, the air flow from the lungs is necessary to create subglottic pressure on the adjacent vibratory edges of the vocal cords, which generate the mucosal wave responsible for phonation. The pitch is regulated by changes in the length and tension in the vocals by the action of the cricothyroid muscles on the cricothyroid joint.

Clinical assessment of dysphonia

Clinical assessment of dysphonia is best achieved in a multidisciplinary voice clinic set-up consisting of the otolaryngologist and the speech and language therapist. An extended voice clinic set-up will also be helpful, and this may comprise of a singing coach, an osteopath, and a clinical psychologist. A detailed history of the onset, triggers, relieving factors, occupation, drug history, allergy, and other existing or past health issues and lifestyle are essential in establishing the diagnoses and for treatment planning.

It is useful to listen and record the patient's voice while reading a short and phonetically balanced passage that reflects the variety of sounds and mouth movements used in normal, unscripted English speech. Examples are the 'Rainbow passage' or 'Arthur the rat'.[1] Other standard tasks include sustained vowels at level, rising, and falling pitch, voiceless and voiced fricative pairs (e.g. s/z), counting, a short sample of connected speech, and increased loudness on a word such as 'hey' or 'taxi'. Repetition of a sniff followed by the vowel /i/ is useful in assessing the degree of vocal fold abduction. Specific voicing tasks to differentiate muscle tension dysphonia from laryngeal dystonia may be included, as described in Chapter 72.

The larynx should be examined using a high-definition fibreoptic endoscope and stroboscope, with the patient sitting and physician sitting or standing in a comfortable position to look for vocal cord mobility problems and any mucosal lesions.

Palpation of the neck might be necessary to examine for areas of muscle tension or neck masses.

Flexible video laryngostroboscope versus a rigid video laryngostroboscope

Flexible fibreoptic video laryngostroboscopy

Distal chip fibreoptic cameras available of late have greatly enhanced the image quality for laryngeal examination. This has made it possible to perform a flexible laryngeal stroboscopy with an image quality close enough to the rigid 10 mm rigid laryngostroboscope. The advantage of the flexible video stroboscope is its ability to allow assessment of the vocal cords during connected speech without triggering the gag reflex, thus making it easier for the patient to tolerate a full assessment.

Rigid videolaryngostroboscopy

The rigid videolaryngostroboscopy has a slightly better image quality than its flexible counterpart. There are different sizes ranging from 4 mm to 10 mm diameter and 70° or 90° angled tip to allow visualization of the larynx by placing the tip of the telescope in the

oropharynx above the epiglottis without touching it. This way, the gag reflex is minimized. Occasionally, the oropharynx may need to be sprayed with a topical anaesthetic spray to reduce the gag reflex. The other drawback of the rigid stroboscope is its inability to allow examination of the vocal cords using connected speech.

Subjective assessment

There are several questionnaires available to assess a patient's report of voice symptoms. These include the following:

- Voice handicap index (VHI).[2]
- Auditory perceptual evaluation using the grade of hoarseness, roughness, breathiness, asthenia, strain (GRBAS) scale.[3]
- Consensus Auditory-Perceptual Evaluation of Voice (CAPE-V).[3]
- Voice Scoring System (VoiSS).[4]

The VHI and VoiSS questionnaires are self-administered by the patient. The VHI is available in two forms, VHI 10 or VHI 30, with the numbers indicating the items in the questionnaire. In routine clinical settings, the VHI 10 is considered to offer equivalent information to VHI 30.

The GRBAS[3] and CAPE-V involve assessment of the voice by an expert listener, usually a speech pathologist, phoniatrician, or laryngologist.

GRBAS

The patient is asked to read a phonetically balanced passage while listening and assessing the overall grade of hoarseness G, roughness R, breathiness B, asthenia A and strain S. This is score on a scale of 0 to 3, with 0 being normal voice and 3 worse voice.

Objective measures

The objective assessment of dysphonia is performed by recording the voice which is then analysed using voice software.[5] There are several parameters that can be measured. The most common software analysis is the Multidimensional Voice Programme (MDVP) acoustic analysis—it is recorded using a continuous voice 'aaa' which is analysed using the software to provide several parameters. This is then compared against standard normal values for men and women. It provides objective calculations of several vocal parameters such as the fundamental frequency, shimmer, jitter, noise–harmonic ratio, and so on. These parameters are of more importance in research to demonstrate objective changes in voice before and after treatment. The disadvantage of objective voice assessment is that the results do not always correlate to clinical findings or patient symptomatology, and hence the results should be interpreted in isolation. It is, however, more useful if interpreted along with clinical and subjective voice assessment.

Causes of dysphonia

Dysphonia can be caused by organic and functional issues intrinsic to the larynx, or other systemic causes (Box 47.1). Additionally, cord palsy, a major cause of dysphonia, can be the result of several other pathological processes (Box 47.2).

The following section discusses the various causes of dysphonia arising from a primary laryngeal organic cause.

Box 47.1 Hoarseness due to systemic causes

Neurological
- Parkinson's disease.
- Motor neuron disease.
- Multiple sclerosis.
- Stroke.

Chronic inflammatory disorders
- Sarcoid.
- Amyloid.
- Granulomatosis with polyangiitis.
- Rheumatoid arthritis.
- Sjögren's syndrome.

Congenital

Hoarseness might be caused by a congenital vocal cord paralysis, cyst, or haemangioma on the vocal cords. This is usually diagnosed soon after birth due to poor cry from the neonate. The management at this stage is usually conservative unless there is airway compromise. These are discussed in Chapter 79.

Reinke's oedema

Reinke's oedema is due to excessive accumulation of fluid in the superficial layer of lamina propria, otherwise known as Reinke's space of the vocal cord. It is usually as a result of vocal cord irritants like smoke, acid reflux, and infection. The treatment should be aimed at removing the offending agents first to increase chances of surgical success. The surgery may be by cold steel or laser drainage and excision of any redundant epithelium. Care must be taken not to damage the vocal ligament otherwise this could result in a permanent scar that is more difficult to treat.

Vocal cord scar

Vocal cord scar is commonly iatrogenic but can also be caused by acid reflux, trauma, and infection. Initial treatment should be conservative, with speech therapy initially. Surgery via a suspension micro-laryngoscopy might be necessary to excise the scar, combined with injection of a long-acting steroid to prevent recurrence. Vocal cord augmentation with autologous fat or hyaluronic acid might improve voice quality.

Box 47.2 Causes of vocal cord palsy

Iatrogenic
- Thyroid surgery.
- Cervical discectomy.
- Cardiothoracic surgery.
- Neck dissection.

Neoplastic
- Lung cancer.
- Thyroid/parathyroid cancer.
- Laryngopharyngeal cancer.
- Neck metastasis.

Others
- Idiopathic.

Figure 47.1 Vocal cord leucoplakia.

Vocal cord leukoplakia

This is characterized by white patches on the vocal cords due to metaplasia of the epithelium (Figure 47.1). It is caused by smoking, acid reflux, and infection. Biopsy is required to exclude cancer. The treatment is to stop smoking and treat the reflux. Surgical excision with cold steel or laser may be helpful for focal lesions. Care must be taken not to excise the vocal ligament as this may result in a permanent scar and worsening of the dysphonia.

Vocal cord cyst

Vocal cord cysts are benign fluid-containing lesions, arising from a blocked mucus gland (retention cyst) (Figure 47.2a) or subepithelial cysts arising from an epithelial sac (Figure 47.2b). They are usually single, can affect any part of the vocal cord but most commonly on the medial edge of the membranous vocal cord. Often the friction from the cyst may cause a reaction damage to the opposite vocal cord epithelium and this may be confused with a vocal cord nodule. However, close examination with a high-definition videostroboscopy will usually reveal the cystic nature of the swelling. The treatment is surgical excision. The whole cyst should be removed, avoiding injury to the vocal cord ligament as this may result in a permanent scar.

Vocal cord bamboo nodes

These are inflammatory swelling in the mid part of the vocal cords bilaterally (Figure 47.3). It is often associated with chronic inflammatory or autoimmune diseases. Blood test such as rheumatoid factor and autoantibody tests may lead to a diagnosis of autoimmune disease. Management includes treating the underlying autoimmune disease. This may involve use of steroids or immunosuppressive drugs. Speech therapy or intralesion injection of a long-acting steroid such as Depo-Medrone may be helpful. These lesions may recur after treatment.

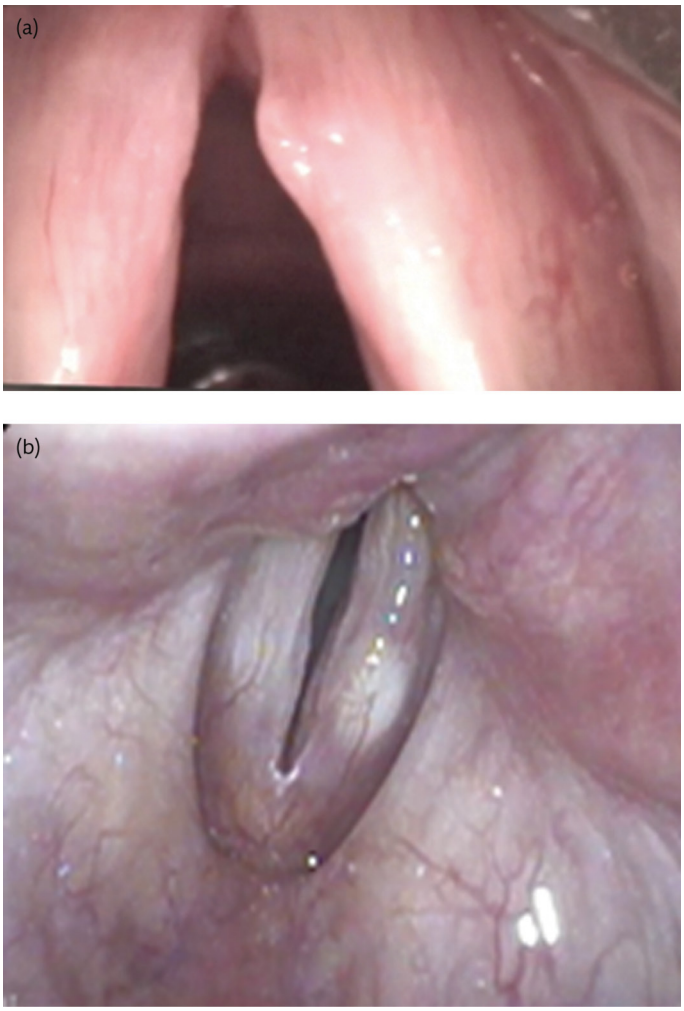

Figure 47.2 (a) Vocal cord cyst. (b) Subepithelial cysts.

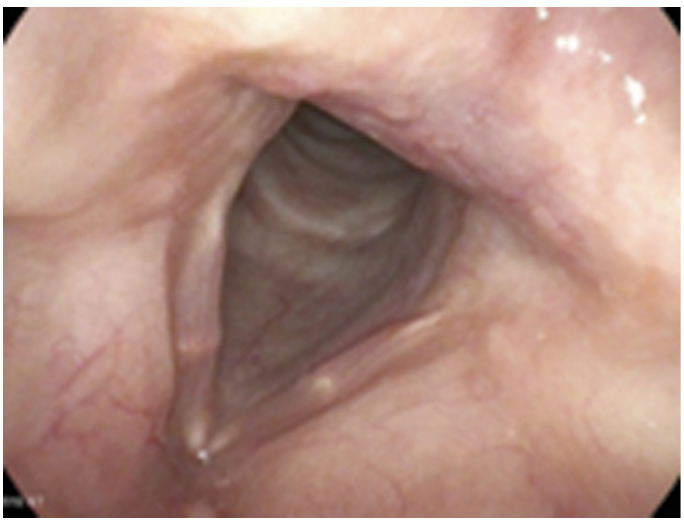

Figure 47.3 Vocal cord bamboo nodes.

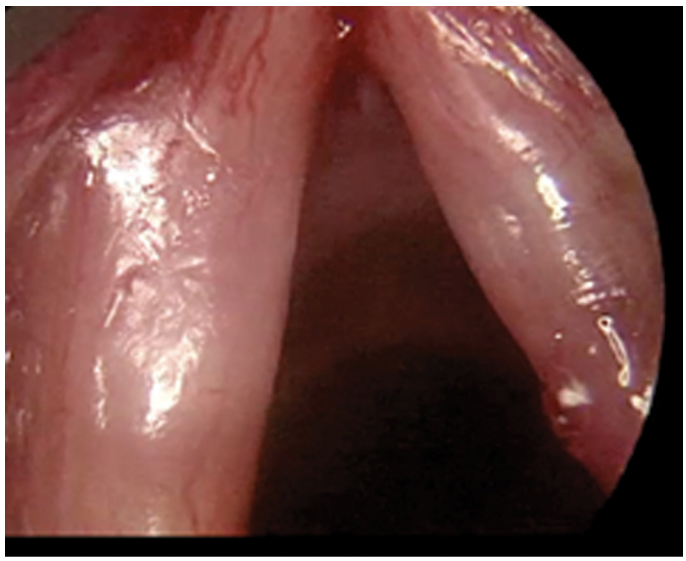

Figure 47.4 Sulcus vocalis.

Figure 47.5 Vocal cord nodules.

Sulcus vocalis

Sulcus vocalis (Figure 47.4) is a rare cause of hoarseness, caused by a groove or infolding of mucosa along the surface of the vocal fold, which can be seen on one or both vocal cords. Unilateral sulcus is usually acquired from a burst vocal cord cyst, vocal trauma, or following an incomplete cyst excision, or from trauma, infection, acid reflux, or smoke irritant.

Bilateral sulcus is often due to congenital absence of Reinke's space or the epithelium, or both, in the medial edge of the vibratory membranous vocal cords. The voice has usually been hoarse since childhood.

There are different grades of sulcus, categorized by the severity. Type 1 is referred to as physiological sulcus, where a minor abnormality of the vocal cord epithelium leads to a small reduction in mucosal waves. The voice is only mildly affected. In type 2, the epithelium of the vocal cord is adherent to the vocal ligament. There is loss of the superficial lamina propria and the voice is moderately affected. Type 3 is the most severe form with either complete loss of the epithelial lining or invagination of part of the epithelium into the vocal ligament (blue arrow). The voice is significantly dysphonic.

The treatment involves speech therapy and surgery. Surgery may involve excision of the scar and injection of steroid. Any phonatory gap may be closed by vocal cord augmentation using hyaluronic acid or autologous fat.

Vocal cord nodules

These are callus-like opposing pair of hypertrophied benign epithelial swellings that occur at the mid part of the medial edge of the membranous vocal cords (Figure 47.5). They are due to excessive friction between adjacent sides of the epithelium of the vocal cords from vocal misuse. They are most seen in professional voice users. Vocal rest and speech therapy are effective in their treatment. Rarely, suspension phono-microlaryngeal surgery may be employed to excise them. This can be either with the use of a laser or cold steel technique. Care must be taken not to excise the vocal ligament as this will lead to a permanent scar formation and worseness of the voice.

Vocal cord polyps

Vocal cord polyps (Figure 47.6) are different from nodules because they can occur on one or both vocal cords. They usually present as larger lesions, with more vascularity and mucinous content. They tend to arise deeper than the epithelium, causing bulging of the surface of the vocal cord. Vocal cord polyps can be caused by any laryngeal irritant like smoking, acid reflux, or infection. Voice abuse can also cause vocal cord haematoma which may later develop into a haemorrhagic polyp. The treatment is aimed at removing the causative agent, vocal hygiene, vocal rest, and speech therapy as the first line of management. In some cases, surgery may be the first line of treatment especially if the patient is severely dysphonic or if biopsy is required to exclude malignancy.

Recurrent respiratory papilloma

The is a benign lesion caused by the human papillomavirus (HPV) (Figure 47.7). It can occur anywhere in the respiratory

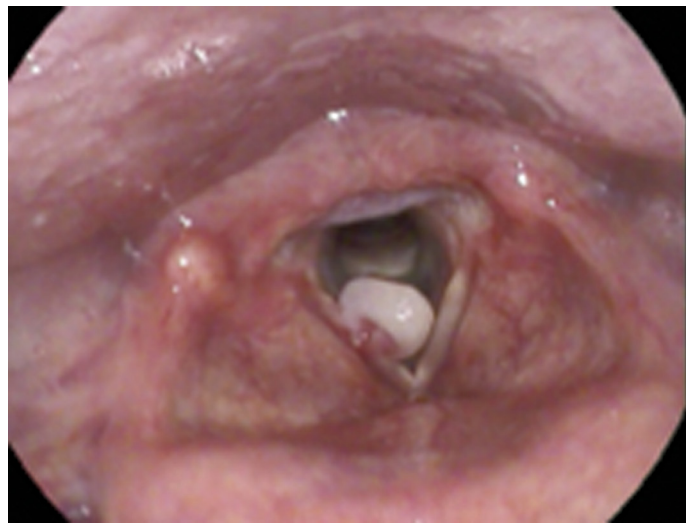

Figure 47.6 Vocal cord polyps.

Figure 47.7 Recurrent respiratory papilloma.

system and is associated with a 1% risk of malignant transformation. There are over 150 types of HPV, but the ones that are more associated with recurrent respiratory papilloma are HPV6 and HPV11. These types are similar to the genital HPV and the mode of transmission is via the orogenital route. HPV vaccines are effective in preventing transmission.[6] For existing disease, the treatment is palliative debulking to improve voice or breathing. Several surgical techniques are available and there is no convincing evidence that one technique is better in preventing recurrence than the other. These techniques include suspension microlaryngoscopy and cold steel using microdebrider, or lasers (carbon dioxide laser, KTP or blue laser). The procedure may also be performed in the office using a fibre-guided laser via a channelled nasal endoscope (see Chapter 71). Medical treatment with injection of cidofovir injection,[7] Gardasil,[8] or bevacizumab (Avastin)[9] have been used with some promising reduction of recurrence of disease.

Laryngopharyngeal reflux

Reflux of gastric acid can cause inflammation of the vocal cord. The larynx may exhibit some signs of reflux-like posterior commissure oedema (also known as pachydermia due to its appearance resembling elephant skin), inflamed arytenoids or granulomas of the vocal process (see 'Laryngeal granuloma'), obliteration of the laryngeal ventricles due to oedema, subglottic oedema, and excessive laryngeal mucus. The management involves lifestyle and dietary management, combined with drug therapy as appropriate (proton pump inhibitors and antacids).

Laryngitis

Inflammation of the vocal cords is most commonly viral, usually self-limiting, and resolves within a few days.

Laryngeal granuloma

This is chronic inflammation on an exposed surface of the arytenoid vocal process from excess deposits of giant cells on an exposed and inflamed cartilaginous surface of the arytenoid (Figure 47.8). The most common causes are prolonged intubation or laryngopharyngeal reflux. The treatment involves voice rest, antireflux medication, excision, and injection of long-acting steroid at the base of the lesion. Botulinum toxin injection might be necessary in some resistant cases.

Figure 47.8 Laryngeal granuloma.

Vocal cord paralysis (left)

Vocal cord paralysis (Figure 47.9) may be caused by a peripheral lesion or trauma that affects the recurrent laryngeal nerve along its course, or a central lesion that affects the vagus nerve and recurrent laryngeal nerve at its nucleus. Symptoms may vary depending on the nature of the paralysis: unilateral versus bilateral, and adductor or abductor. In an adductor paralysis the voice is usually affected due to glottic incompetence whereas in abductor paralysis breathing is affected predominantly. The management of a unilateral vocal cord adductor paralysis includes voice therapy (see Chapter 72) to medialize the vocal cord using one of several techniques: injection of

Figure 47.9 Left vocal cord paralysis.

bulking agent[10] like calcium hydroxyapatite, hyaluronic acid fat, or fascia into the affected paraglottic space, Isshiki type 1 medialization thyroplasty,[11] or recurrent laryngeal nerve reinnervation,[12] discussed in greater detail in Chapter 71.

Laryngeal dystonia

Previously known by the term spasmodic dysphonia, this is a neurological disorder of unknown cause. There is involuntary contraction of the laryngeal muscles resulting in voice breaks and strain. It is thought to be due to abnormal release of acetylcholine at the neuromuscular junction. There are three types of laryngeal dystonia: adductor, abductor, or mixed dystonia, depending on which muscles are affected. The standard treatment is by injection of botulinum toxin into the affected muscle group (thyroarytenoid for adductor type and posterior cricoarytenoid muscle for abductor type).[13] This provides a temporary improvement of voice that lasts up to 3 months before the injection is required again. There are recent advances of surgical management of adductor spasmodic dysphonia using carbon dioxide laser to excise the thyro-arytenoid muscles,[14] thyroplasty type 2 lateralization,[15] or selective denervation reinnervation of the laryngeal muscles,[16] discussed in detail in Chapter 71.

Laryngeal cancer

This is discussed in Chapter 55.

Psychogenic/functional dysphonia

This is a voice disorder due to a non-organic cause[17] and is discussed in Chapter 72.

Puberphonia

This is a mutational voice disorder seen in teenage boys whose voice fails to deepen. The line of treatment is usually with speech therapy.[18] In older patients who fail to respond to speech therapy, surgery might be required. Thyroplasty[19] is performed in such cases under a local anaesthesia to relax the vocal cord with a resultant lowering of the pitch.

Presbyphonia/presbylaryngis

This is due to loss of muscle tone and atrophy from ageing.[20] The voice is weak and breathy and tires easily during phonation. The treatment is by speech therapy. Patients who do not respond to speech therapy or who cannot have speech therapy for any reason may benefit from vocal cord injection of a filler. Injection augmentation using autologous fat or calcium hydroxyapatite or hyaluronic may provide a temporary improvement in voice. In severe cases with significant phonatory gap, a type 1 medialization thyroplasty using a silicon implant might provide a longer-lasting improvement in voice.

Essential tremor

A generalized neurological tremor may affect the laryngeal muscles and present with a tremulous voice and breaks. The treatment might be a combination of speech therapy, beta blockers, and benzodiazepines. However, the risk and side effects of taking these drugs must be carefully assessed against the benefit.

Controversies

- *Role of objective voice analysis*: the role of objective voice analysis in clinical practice continues to be controversial owing to a lack of correlation with the patient's perception of their voice. However, it is a useful tool when used in conjunction with other voice assessment measures.
- *Voice rest following surgery on vocal cords*: there is no fixed period for voice rest after phonosurgery. It will depend on the extent of the surgery and the profession of the patient. The average is from 3 days to 2 weeks with gradual return to normal speaking. Concern exists over prolonged voice rest in professional voice users, which may cause disuse atrophy of the vocal cords.
- *Timing for surgical intervention in management of vocal cord nodules*: the timing for surgery for vocal cord nodules varies according to the severity of the voice and how long the patient is prepared to wait for speech therapy to resolve the nodule.
- *Cold steel versus laser surgery on the vocal cords*: there is debate whether cold steel or laser is better to excise lesions of the vocal cords. Both techniques have their advantages and disadvantages, and the expertise of the treating team should influence the decision.

Guidelines

Useful websites

- British Voice Association: https://www.britishvoiceassociation.org.uk/
- British Laryngological Association: https://www.britishlaryngological.org/
- European Laryngological Society: http://www.elsoc.org/
- American Laryngological Association: https://alahns.org/
- ENT UK: https://www.entuk.org/

Conclusion

Assessment of dysphonia is best performed in a multidisciplinary set-up comprising an otolaryngologist and a speech and language therapist. History taking might provide the diagnosis but an endoscopic examination of the larynx is invaluable in confirming the majority of the diagnoses. The equipment should include, as a minimum, a high-definition videolaryngoscope and stroboscope. Additional equipment may be needed for voice analysis or laryngeal electromyography may be needed in special situations in the outpatient setting.

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48

Dysphagia

Somiah Siddiq

Introduction

Dysphagia is the subjective sensation of impairment of the passage of swallowed material. The patient typically reports dysphagia as a sensation of food 'getting stuck', or passing down too slowly. Food bolus obstruction is when the obstruction lasts for more than 1 minute. Dysphagia is an uncomfortable sensation when present, and should be differentiated from odynophagia which is pain experienced during swallowing. Dysphagia should also be differentiated from a range of other swallow-related symptoms. Globus pharyngeus is the commonest swallow-related symptom and is defined as the painless sensation of a lump or constriction in the throat which does not cause actual impairment to the passage of swallowed material, is present at times other than during swallowing, and may improve on swallowing.

The historical approach to classifying dysphagia (**Figure 48.1**) has been to consider it as *oropharyngeal dysphagia* (dysphagia caused by impairment of processes that affect oral preparation of a bolus and its transfer through the pharynx and the upper oesophageal sphincter into the oesophagus), and *oesophageal dysphagia* (impairment to the passage of bolus through the oesophagus and the lower oesophageal sphincter into the stomach). This differentiation conveniently

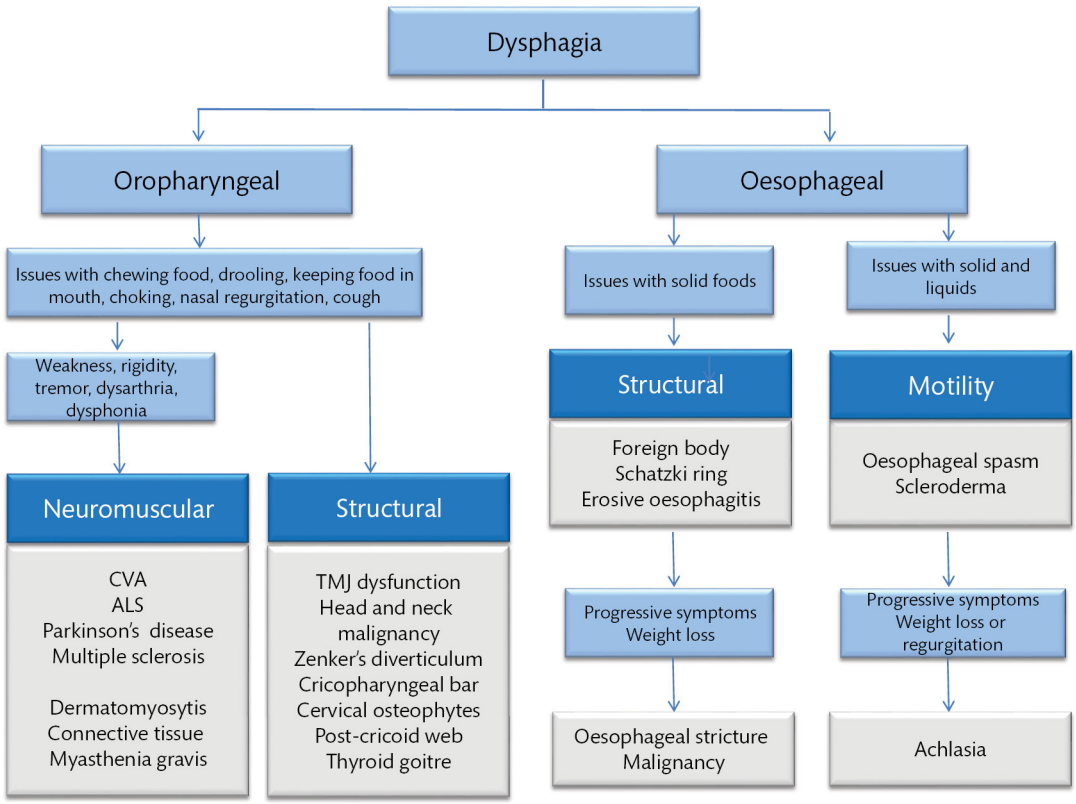

Figure 48.1 Dysphagia history algorithm. ALS, amyotrophic lateral sclerosis; CVA, cerebrovascular accident; TMJ, temporomandibular joint.

Table 48.1 Red flag symptoms

Head and neck	Gastroenterology*
Any of the following persisting for >3 weeks: • Odynophagia • Unilateral throat pain/otalgia • Difficulty swallowing/obstruction • Neck lump • Hoarseness	>55 years with dysphagia >55 years with weight loss and any of the following symptoms: • Upper abdominal pain • Reflux • Dyspepsia

* Direct access 2-week-wait endoscopy.

divides the symptom between otolaryngology/speech therapy and gastroenterology and is embedded in many diagnostic referral pathways including the 2-week-wait suspected cancer pathways (Table 48.1). In many cases, this distinction is fairly clear on the basis of a good clinical history but inter-specialty dysphagia referrals are responsible for delayed diagnosis and 2–3% of head and neck and oesophageal cancers being missed.[1] As such, a clinician evaluating dysphagia should be familiar with the diagnostic evaluation of the symptom as a whole and to work within a multidisciplinary setting (Figure 48.2).

Anatomy and physiology

The anatomy of the pharynx is described in detail in Chapters 56 and 60. The swallowing mechanism may be considered as a series of dynamic chambers and muscular sphincters that provide closure, and by coordinated neuromuscular contraction and relaxation, create zones of high pressure on the bolus and negative pressure inferior to the bolus that propel it from the oral cavity to the stomach. Swallowing is the most stereotyped and the most complex motor activity mammalian species undertake and requires closely timed coordination of breathing, speech, and swallowing across multiple cranial nerve nuclei, highly coordinated motor commands sent via multiple cranial nerves, and precise execution across 25 pairs of muscles.

Swallowing physiology can be classified into four phases based on the location of the bolus (Figure 48.3). Any disorder of swallowing can be considered as a combination of derangements in one or more of four basic processes of *preparation*, *perception*, *protection*, and *propulsion*.

Preparatory phase

The voluntary process of mastication breaks down food, mixing it with saliva to form a bolus. The orbicularis oris (*first sphincter*) and buccinators maintain food bolus position. Chewing is undertaken by the muscles of mastication.

Movement and position of the bolus within the oral cavity is orchestrated primarily by extrinsic and intrinsic tongue muscles. The palate (*second sphincter*), by contraction of the palatoglossus muscles approximates to the posterior tongue and closes the oral cavity. Masticatory efficiency will be impacted by reduced occlusal contact secondary to missing dentition, loose-fitting dentures, or temporomandibular dysfunction.

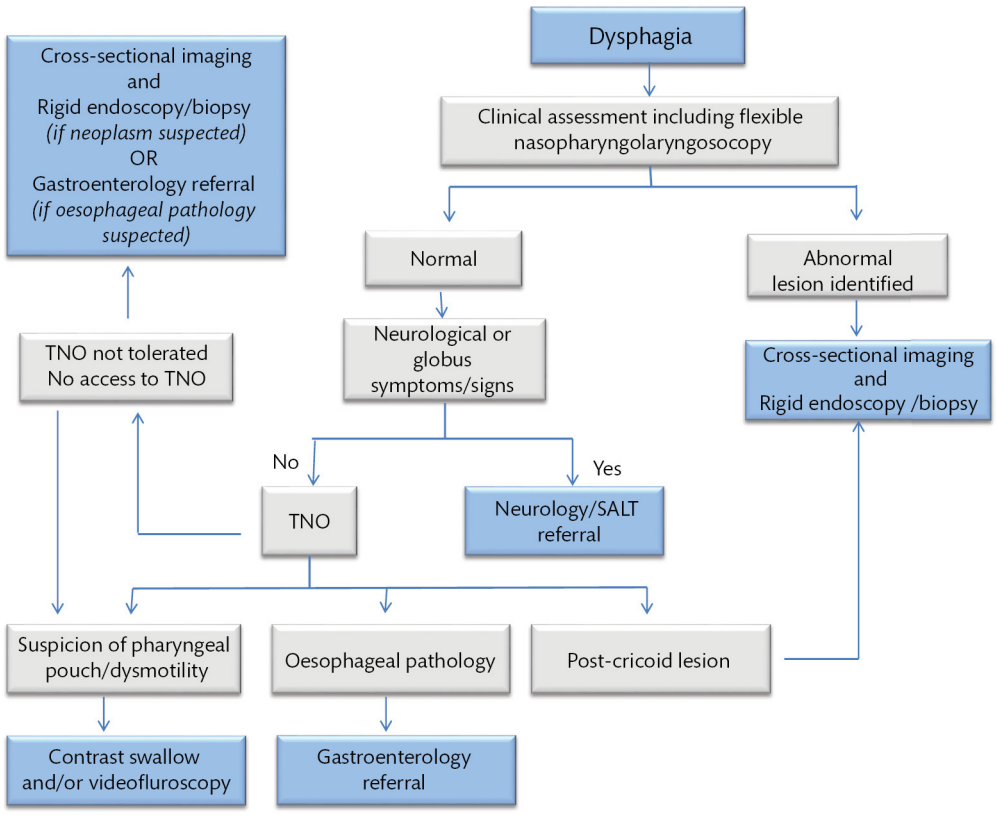

Figure 48.2 Suggested diagnostic pathway in patients presenting with dysphagia to the ENT clinic. (SALT, Speech and Language Therapy.)

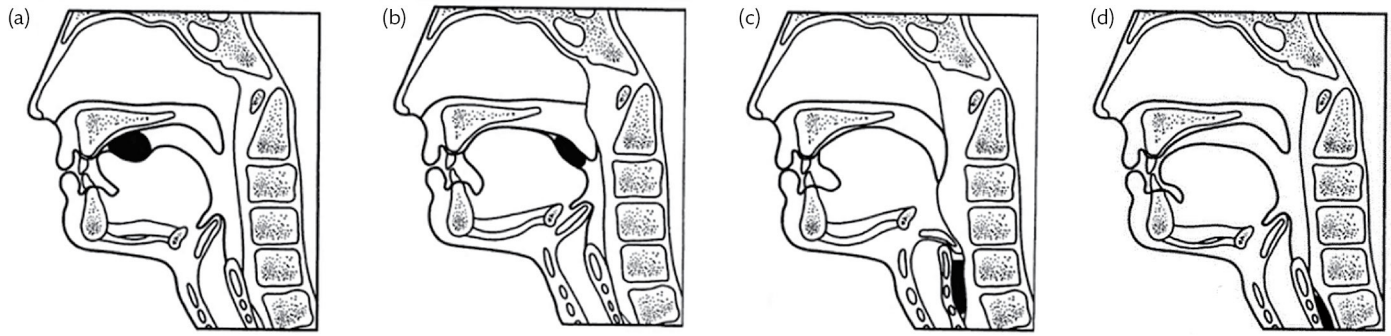

Figure 48.3 (a) Preparatory phase, (b) oral phase, (c) pharyngeal phase, and (d) oesophageal phase of swallowing.

Reproduced from Sivan M, Phillips M, Baguley I, Nott M, eds. *Oxford Handbook of Rehabilitation Medicine*. 3rd ed. Oxford University Press; 2019 with permission from Oxford University Press.

Oral phase

This is a voluntary phase under cortical control. Soft palate elevation (contraction of the veli palatini muscles) and depression of posterior tongue causes bolus propulsion from the oral cavity into the pharynx. Contraction of the orbicularis oris and buccinator muscles prevent bolus pressure escape anteriorly and laterally. On full contraction of the palate, contact with the adjacent pharyngeal wall (superior constrictor, motor innervation (cranial nerve X) via the pharyngeal plexus) seals the nasopharynx preventing bolus/air escape.

Early elevation of the hyoid bone results from mylohyoid contraction and subsequent elevation of the hyoid and larynx by the anterior and posterior belly of the digastric muscles.

Sensory receptors within the oral cavity and the oropharynx sense the location, volume, and consistency of the bolus and finely adjust the force of contraction of pharyngeal muscles to enable safe transfer. Abnormalities in the sensing of the type and location of a bolus (e.g. radiotherapy effect on the sensory nerves of the pharynx) cause abnormalities in this process and typically lead to 'premature bolus spillage' into the pharynx and reduced coordination of pharyngeal peristalsis.

Pharyngeal phase

This is an involuntary irreversible motor event and lasts around 1 second. Respiration stops through coordination at brainstem level (swallow-related apnoea). Backflow into the oral cavity is prevented by contraction of the base of tongue and tonsillar pillar muscles. Backflow into the nasopharynx is prevented by the action of palatal muscles, principally tensor and levator veli palatini, and in some patients the superior-most aspect of the superior pharyngeal constrictor also contributes to concentric nasopharyngeal closure.

Elevation and contraction of the pharynx result in a descending peristaltic wave, forcing the bolus inferiorly. The hyoid and larynx simultaneously rise and are pulled forward (this protects the larynx from bolus penetration and expands the hypopharyngeal chamber) under the root of the tongue by contraction of the suprahyoid muscles. The epiglottis folds over the larynx and vocal folds close with contraction of intrinsic laryngeal muscles. The bolus is kept cohesive and is transferred, by the action of the base of tongue and the circular and longitudinal pharyngeal constrictor muscles.

The pharyngo-oesophageal segment (*third sphincter*) is closed at rest by tonic contraction of cricopharyngeus muscle. Opening involves inhibition of tonic contraction and consequent relaxation, and laryngeal elevation pulls the cricoid lamina away from the posterior pharyngeal wall with the resultant decreased pressure driving the bolus through the pharynx into the upper oesophagus. The pharyngeal phase is complete once the soft palate returns to its normal position and the larynx reopens for respiration.

Oesophageal phase

This is involuntary phase with peristaltic wave contraction to facilitate bolus transport down the oesophagus to the stomach. Lower sphincter tonic contraction of smooth muscles is inhibited, allowing relaxation for bolus passage into stomach.

Clinical assessment

History

Dysphagia versus swallow-related symptom

As many as 60% of patients referred for dysphagia evaluation via the suspected cancer pathway do not have dysphagia and the commonest dysphagia mimic is globus pharyngeus. Whether or not there is an actual report of impairment to the passage of swallowed material should be carefully ascertained. Swallow-related symptoms are managed according to their own pathways.

Speed of onset

An acute history suggests a foreign body and may be mechanical or due to a neurological cause if timing is related to an acute cerebrovascular accident. Acute food-bolus obstruction, especially in a male patient, is cancer or eosinophilic oesophagitis until proven otherwise. In both cases, the full length of the oesophagus and the cardio-oesophageal junction need to be directly visualized, and if no malignant lesions are seen, biopsies from different regions of the oesophagus need to be taken to exclude eosinophilic oesophagitis.

Duration and nature of symptoms

A short duration (<8 weeks) is more likely to represent malignancy; a longer history and intermittent dysphagia is more likely to be due to benign pathology than progressive dysphagia.[2] However, when the

patient's differential diagnosis includes cancer, the duration and nature of symptoms should be used to guide urgency of investigations rather than whether or not investigations need to be performed. Although intuitive, it is important to challenge the underlying assumption that progressive lumen obstruction clinically translates to progressive symptom development. It does not necessarily, and as many as half of patients with malignancy reported non-progressive dysphagia symptoms.[3] Neither intermittent nor stable dysphagia rule out malignancy.

Consistency and dysphagia-related decompensation

Dysphagia to both solids and liquids is more likely to be related to neurological and motility causes, and solid food dysphagia is considered more likely to be related to structural pathology. Swallowed materials may be divided into dry solids (breads, cakes, or biscuits), wet solids (meat or fruit), soft solids (custard), thickened fluids (soup), liquids, or pills.

The history needs to ascertain whether dysphagia includes dysphagia to liquids, which type(s) of solid foods are affected, and whether the patient permanently avoids certain foods. Thus, dysphagia can be characterized into four types: absent, present without the need for compensatory strategies (chewing more carefully, eating more slowly, dietary modification), present and compensated for (typically through diet modification), and present and uncompensated (dysphagia associated with unplanned weight loss).

Anatomical localization

Given the wide sensory receptive field of the vagus nerve, dysphagia symptom localization is poor. Both balloon distension of the oesophagus or duplication of symptoms by radiographic impaction of marshmallow in the distal oesophagus have shown where the patient feels symptoms are not reliable to determine the location of oesophageal dysfunction.

Symptom-based localization

By contrast, the presence of co-localizing symptoms provides a reasonable indication of the underlying location of the pathology.

- *Head and neck symptoms*: presence of throat pain, especially if lateralizing and particularly if associated with referred otalgia, swallow-related pain (odynophagia), hoarseness, atypical oral pain, globus (in addition to true dysphagia), a post-swallow wet voice, and a history of pneumonia point towards possible head and neck cancer (Table 48.1).
- *Neurological symptoms*: presence of nasopharyngeal regurgitation, dysarthric voice patterns, and post-swallow coughing and choking point towards a neurological cause. Other neuromuscular symptoms and signs include diplopia, muscle weakness, vertigo, tremor, and ataxia.
- *Oesophageal symptoms*: gastro-oesophageal reflux typically presents with heartburn and acid regurgitation but may also cause atypical chest pain, especially in women, and lead to reflux-related lung disease that presents with wheezing and chronic cough. A specific warning sign is sudden resolution of long-standing reflux symptoms as this may indicate malignant oesophageal lumen obstruction causing a reduction in oesophageal acid exposure.
- *Gastroduodenal symptoms*: these symptoms include dyspepsia, nausea and vomiting, early satiety, and epigastric pain. Presence or absence of these symptoms should be documented in all patients.
- *Regurgitation*: halitosis and regurgitation may indicate Zenker diverticulum or achalasia.

Medical and drug history

Assessment should encompass associated systemic conditions that may result in swallowing difficulties including collagen vascular diseases, such as scleroderma or systemic lupus erythematosus. Various antipsychotics may impact swallow by causing xerostomia, direct oesophageal mucosal injury, or decreased lower oesophageal sphincter tone to cause reflux. Patients on long-term immunosuppressants with pain may suggest underlying infective oesophagitis. Both anticholinergic and antimuscarinic drugs affect smooth muscle, reducing oesophageal motility and tone of the lower oesophageal sphincter precipitating reflux. Those on centrally acting medication can cause tardive dyskinesia with difficulty in mastication and swallowing. Medication-induced oesophagitis may result secondary to non-steroidal inflammatory drugs, ferrous sulphate, tetracycline, alendronate, and clindamycin resulting in oesophageal ulceration and possible stricture-related complications. Opiates can alter oesophageal motor function resulting in reduced motility.

A history of atopy, asthma, or food allergies may be suggestive of eosinophilic oesophagitis.

Clinical examination

A stepwise approach to clinical examination should be undertaken bearing in mind the phases of swallow with simultaneous neurological assessment of cranial nerve function (Table 48.2) during the examination.

Two tongue blades are used to examine the oral cavity. Assess lips for oral competence and signs of salivary leak, interincisal distance for trismus, absent dentition, malocclusion or ill-fitting dentures, and xerostomia (spatula sticking to tongue/cheek). Next, assess tongue movement (both protrusion and side to side movement),

Table 48.2 Cranial nerves to be assessed during clinical examination

Cranial nerve	Function
V	Muscles of mastication (masseter, temporalis, and medial and lateral pterygoids) Sensation to the anterior two-thirds of the tongue and buccal cavity
VII	Nerve to supply the orbicularis oris muscle (lip closure/seal), buccinator muscle Taste to the anterior two-thirds of the tongue
XI	Taste and sensation to the posterior third of the tongue and buccal cavity Sensation to the tonsils, laryngeal mucosa, and soft palate Involved in the gag (soft palate elevation) and cough reflexes
X	Similar to cranial nerve IX Involved with phonation and vocal cord closure in the larynx Sensation and motility of the oesophagus
XII	Tongue movement

noting any weakness, tethering, atrophy, or signs of fasciculation (an early sign for amyotrophic lateral sclerosis). Tongue strength can be assessed by asking the patient to push up against the tongue blade. Grasping the tongue with a swab and moving it to the side can offer a good view of the posterior aspect of the floor of mouth, an area that can be difficult to examine. The oropharynx should be assessed ensuring symmetrical elevation of the soft palate when the patient is asked to say 'ah' and gentle touch of anterior fauces to elicit the gag reflex.

Neck examination should assess for any obvious cervical lymphadenopathy, thyroid goitre, kyphosis, and appropriate laryngeal elevation on swallow and lateral displacement of the larynx to ensure no loss of crepitus or tethering.

Flexible nasendoscopy assessment should include a stepwise review of the postnasal space, nasal surface of the soft palate, tonsil, and tongue base for asymmetry or obvious tumour. Ask the patient to undertake manoeuvres (tongue protrusion, lateral head turn, chin up, Valsalva manoeuvre) to ensure the valleculae and hypopharynx respectively are more clearly visualized and inspected (Figure 48.4). Vocal fold mobility is assessed by asking the patient to say 'ee' and a more forceful 'eee' brings in the side walls of the pharynx to allow assessment of pharyngeal closure. The interarytenoid region may evidence signs of reflux with erythema or hypertrophy of mucosa. Pharynx sensation can be assessed by gentle touch of the tip of the flexible scope to tongue base, posterior pharyngeal wall eliciting a gag reflex, and laryngeal surface of the epiglottis, lateral pharyngeal walls, and vocal folds eliciting a cough or swallow. Subtle signs of pooling of saliva to valleculae or pyriform sinuses and any impeded movement and shape of the epiglottis should be noted.

Visually assess the swallow by asking the patient to take a sip of water and hold in their mouth, to ensure adequate lip competence and no obvious drooling and then on swallow note signs of nasal regurgitation, immediate cough or choking, need for multiple swallows, or change in phonation 'wet' voice. If there are no overt concerns for frank aspiration then the 100 mL water test screen may be conducted to further assess oropharyngeal dysphagia.

Investigations

Patient-reported outcome measures

Numerous patient-reported outcome measures exist and attempt to quantify the impact of dysphagia on patients' health-related quality of life, assess improvements following intervention, or aid in treatment decision-making. Validated measures for structural and neuromyogenic oropharyngeal dysphagia include the Swallowing Quality of Life (SWAL-QOL) questionnaire, Sydney Swallow Questionnaire (SSQ), and Swallowing Quality of Care (SWAL-CARE) questionnaire.[4] Broader dysphagia self-questionnaires include the Dysphagia Handicap Index (DHI) and Eating Assessment Tool (EAT-10). The EAT-10 comprises ten questions and can be completed in 2 minutes with a higher score associated with risk of aspiration. Condition-specific measures, such as the MD Anderson Dysphagia Index (MDADI), are validated for dysphagia related to upper aerodigestive cancers. Appropriate selection of self-questionnaires must bear in mind many are validated in English with specific literacy level expectations.

Reliance on questionnaires alone may underestimate aspiration and hence clinical screening assessment such as the water swallow test, or to minimize risk of large amounts of liquid and aspiration concerns, the volume-viscosity swallow test may be considered.

Post-stroke dysphagia screening has been advocated particularly in view of the significant morbidity and mortality related to aspiration-induced pulmonary complications, although is based on limited evidence.

Barium swallow versus endoscopy

Endoscopy as a first-line investigation provides a high diagnostic yield (>75%) in patients presenting with dysphagia.[5] This is particularly pertinent to eosinophilic oesophagitis, a diagnosis based on histopathology even in the setting of a normal-appearing oesophageal mucosa on endoscopy. In addition, diagnostic endoscopy combined with therapy is more cost-effective than barium swallow followed by therapeutic endoscopy.[5] A barium swallow may be arranged following this to rule out underlying motility disorders. If

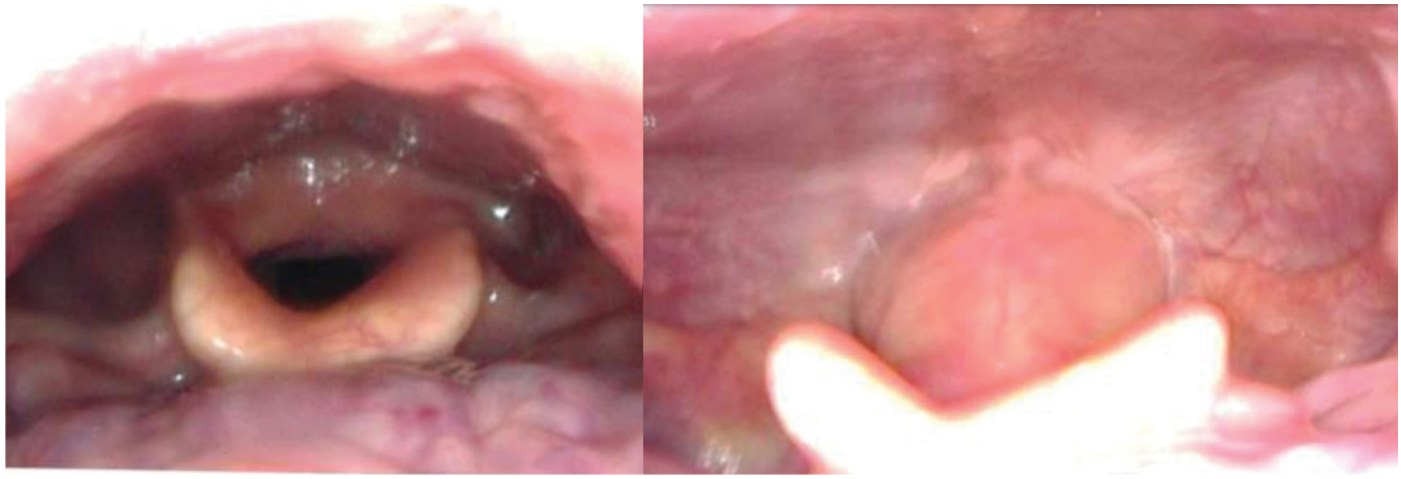

Figure 48.4 Flexible nasendoscopy view of pharynx and larynx. Significantly improved view and thereby inspection of the hypopharynx (post-cricoid, pyriform fossae, and posterior pharyngeal wall) during Valsalva.

suspecting a complex stricture (post radiation or caustic injury), a barium swallow in the first instance to establish the location, length, diameter, and number of strictures is recommended prior to endoscopic dilatation.[6]

Traditional oesophagogastroduodenoscopy performed by the transoral route requires sedation in the majority of patients with risk of cardiopulmonary events and the requirement for additional monitoring, nursing staff, and recovery area.

With the advent of transnasal oesophagoscopy (TNO) in the early 1990s, performed with an ultra-thin scope via the nasal passages, endoscopy is better tolerated (based on visual analogue scales) with equivalent diagnostic yields for the detection of Barrett's oesophagus, gastric cancer, and gastro-oesophageal reflux disease (GORD). The avoidance of sedation is particularly advantageous in the elderly and those with multiple comorbidities. TNO lends itself to therapeutic procedures, especially negotiating strictures for balloon dilatation, insertion of nasoenteral/percutaneous endoscopic gastrostomy feeding tubes and tracheo-oesophageal puncture for speech rehabilitation. This facilitates the option of a bedside procedure and avoids further sedation and (compared to fluoroscopy-assisted insertion) radiation exposure.

High-resolution manometry

Oesophageal high-resolution manometry has revolutionized the evaluation of oesophageal motility disorders. High-resolution manometry incorporates up to 36 pressure sensors spaced 1 cm apart along a catheter in contrast to conventional manometry using a few (three to five sensors) widely spaced sensors. Diagnosis of oesophageal motility disorders is based upon the international consensus Chicago classification, a manometry-based diagnostic algorithm used in clinical practice which is now in its third iteration.[7]

Akin to high-resolution oesophageal manometry, high-resolution pharyngeal manometry is a catheter-based method for recording the pressures and bolus flow generated by pharyngeal muscles during swallow.[8] Currently, in the early phase of protocol and diagnostic standardization, the role of high-resolution pharyngeal manometry to tailor management, assess therapy, monitor disease recovery or progression, or facilitate biofeedback training is yet to be fully determined.[9] In cricopharyngeal dysfunction, high-resolution pharyngeal manometry in conjunction with a videofluoroscopic swallow study (VFSS) can assess pharyngeal strength, cricopharyngeal muscle, pharyngeal coordination, and upper oesophageal sphincter relaxation. Cricopharyngeal dysfunction is likely in normal pharyngeal contraction with elevated upper oesophageal residual pressure during swallow and reduced relaxation times.

Videofluoroscopy swallow study versus flexible endoscopy evaluation of swallowing

VFSS, also known as the modified barium swallow, is considered a gold standard for dysphagia assessment providing a dynamic assessment of oral, pharyngeal, and oesophageal phases of swallow. The technique involves fluoroscopic images taken and recorded during dynamic swallowing. Radio-opaque material is administered over a spectrum of both varying bolus textures and volumes. Standardization in VFSS to minimize inter- and intraobserver variability is achieved through use of pre-made barium sulfate containing contrast agent consistencies (e.g. Varibar series) and methods of assessment and reporting. The severity of swallow dysfunction may be classified using the eight-point Penetration–Aspiration Scale (PAS) or Modified Barium Swallow Impairment profile (MBSImp). A distinct advantage of VFSS is the detection of silent aspiration (**Figure 48.5**) which can be present in 40–60% of patients with neurogenic dysphagia. However, VFSS is both labour and equipment intensive and an invasive examination due to radiation exposure limiting repeated assessment.

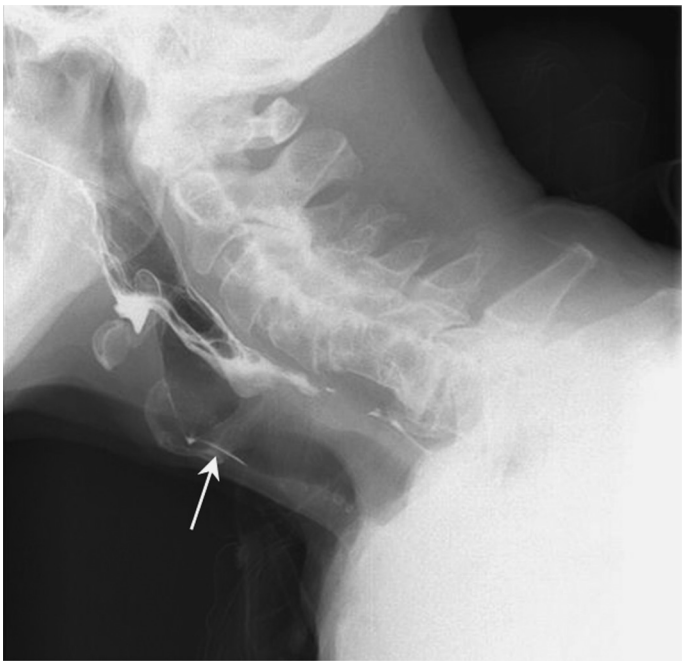

Figure 48.5 VFSS demonstrating post-swallow silent aspiration and penetration (marked by arrow) of contrast, moderate cricopharyngeal hypertrophy and osteophytes at level of C6–C7 indenting posterior wall of oesophagus.

Flexible endoscopic evaluation of swallowing (FEES) can be conducted at the bedside or in the clinic setting using a fibre- or videoscope inserted transnasally (**Figure 48.6**). FEES compared to VFSS demonstrates slightly increased sensitivity for aspiration, penetration, and laryngopharyngeal residue (significant for secondary swallow aspiration) with similar specificity.[10] Hence, it is recommended that FEES be prioritized in centres lacking both VFSS and FEES.[11] The FEES assessment involves non-swallow and voicing tasks conducted prior to trial of food and liquids. Various consistencies of bolus are trialled with the scope in place and assessment made of spillage into the larynx and laryngopharyngeal residue (this may be used in conjunction with the PAS). A normal FEES examination may avoid the need to conduct a VFSS. However, FEES may not be tolerated in some patients, may miss silent aspiration during white out on swallow, and does not evaluate the oral and oesophageal phases of swallow.

Causes of oropharyngeal dysphagia

Globus pharyngeus

Globus (Latin for sphere or globe) pharyngeus is a sensation of a lump or fullness to the throat. It often presents with other associated

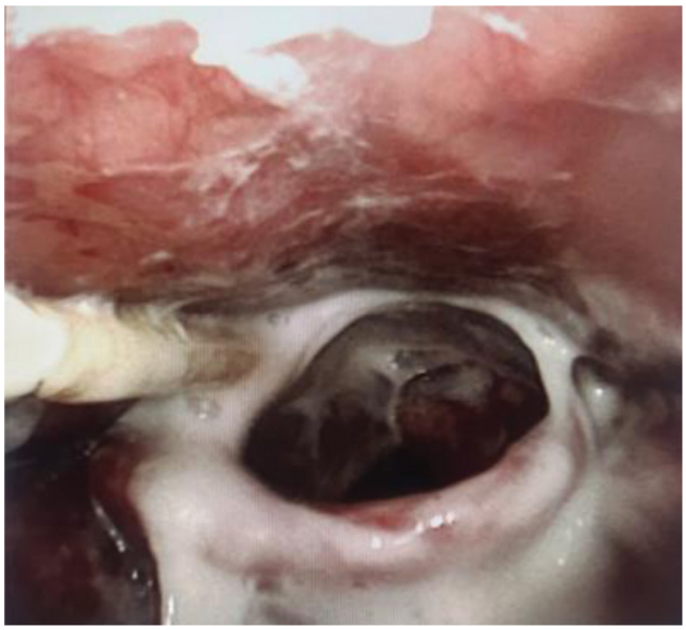

Figure 48.6 FEES demonstrating post-swallow laryngeal overspill and silent aspiration and residue pattern related to reduced epiglottis deflection and tongue base elevation.
Courtesy of Grace Herrity, SALT.

throat symptoms including repeated throat clearing and a mucous/phlegm/catarrh sensation to the throat. It is frequently referred to as a dysphagia mimic, but the sensation is present in the absence of swallowing and may be relieved with the ingestion of liquids/solids. Diagnosis is based on the Rome IV criteria where symptoms should be present for at least 12 weeks with absence of GORD or other major oesophageal motor disorders. In a significant proportion of patients, symptoms may be present for years.

The diagnosis of globus is clinical. Crucially, any associated red flag symptoms (Table 48.1) indicative of potential underlying upper aerodigestive malignancy should be ruled out. A subset of patients may have associated GORD symptoms and if present, severity should be assessed (e.g. reflux symptom index). GORD may directly cause dysphagia by causing oesophageal dysmotility and is a risk factor for Schatzki ring formation, benign peptic strictures, and malignant oesophageal obstruction as secondary causes of dysphagia. Globus is classified as a somatoform disorder and has been associated with anxiety and depression and these should be elicited during the history.

In the presence of typical symptoms with no concerning red flag symptoms and normal flexible nasoendoscopy findings, further investigations result in a very low diagnostic yield for additional pathology and may exacerbate underlying anxiety (see 'Controversies'). If there is significant associated GORD then onward gastroenterology referral for ambulatory pH monitoring and impedance may be considered in a small proportion of patients.

The aetiology and pathophysiology of globus pharyngeus remain unclear with paucity of evidence for effective treatments and strategies. Reassurance and explanation of the benign nature of the condition is the first line and often the mainstay of management.

There is no evidence that the traditional empirical use of high-dose proton pump inhibitors for treatment of globus and its related throat symptoms show benefit compared to placebo[12] and should be avoided. The use of alginates is an area of future research in pepsin-related pharyngeal hypersensitivity.

Speech and language therapy has been shown to improve globus pharyngeus symptom scores[13,14] after 3 months compared to control groups receiving reassurance only. Strategies include voice hygiene instructions, relaxation techniques, and exercises to relieve pharyngolaryngeal tension.

Approximately 50% of patients will still be symptomatic on long-term follow-up, in a cohort with the longest mean follow-up of 7 years[15] none developing an upper aerodigestive malignancy.

Referral for mental health assessment and support may be warranted in a subset of patients identified to have a psychogenic basis or who require psychotherapeutic treatments for their globus symptoms. Little evidence exists for the use of anxiolytics or antidepressants but cognitive behavioural therapy or hypnosis-induced relaxation techniques may be helpful with intractable symptoms.

Pharyngeal pouch

See Chapter 60.

Cricopharyngeal dysfunction

The upper oesophageal sphincter is composed of the inferior pharyngeal constrictor, the cricopharyngeus muscle, and proximal part of the cervical oesophagus, also referred to as the pharyngo-oesophageal segment. The cricopharyngeal muscle (composed of both skeletal muscle and connective tissue) allows passage of the food bolus by passive stretching without active relaxation or inhibition. Cricopharyngeal dysfunction results from incoordination and reduction in the maximal opening of the upper oesophageal sphincter. Patients may report a lump-like sensation, throat clearing, and, in more severe cases, dysphagia, cough, and aspiration.

An initial clinical assessment of swallow (presence of throat clearing and repeated swallow) followed by a combination of FEES, electromyography, manometry, and VFSS may be required to assist in diagnosis and likely management. VFSS may identify a cricopharyngeal bar (bar-like protrusion) in the lumen at the level of C5–C6 secondary to spasm or failure of the inhibition of tonic cricopharyngeus muscle contraction. However, a cricopharyngeal bar may be present in a third of patients with no associated symptoms. Hence, complete assessment of the remaining oesophagus is required to exclude other causes of dysphagia.

Therapeutic interventions for cricopharyngeal dysfunction depend upon severity of symptoms and impact on quality of life. In impaired upper oesophageal sphincter opening, both normal laryngeal elevation and adequate oral and pharyngeal propulsion should be present for treatment consideration.

Broadly, management strategies targeting the cricopharyngeal muscle include balloon dilatation, electromyography-directed chemical denervation (Botox injection), and permanent surgical management via endoscopic (laser) or open cricopharyngeal myotomy.

Neuromuscular disorders

The poor coordination and weakness of facial, oral, and pharyngeal musculature in neuromuscular disorders gives rise to oropharyngeal dysphagia.

Neuromuscular disorders may be broadly divided into:

- neurological disorders: stroke, motor neuron disease, multiple sclerosis, bulbar and brainstem tumours
- striated muscle disorders: myasthenia gravis, muscular dystrophies, and dermatomyositis.

Stroke is the leading cause of oropharyngeal and oesophageal dysphagia with the incidence approaching 50% with persistent dysphagia in a quarter of patients.[16] Pneumonia occurs in 10–40% of stroke patients and is associated with death or significant morbidity.

Amyotrophic lateral sclerosis accounts for 80% of adult-onset motor neuron disease and approximately a third of patients present with bulbar symptoms. The most common bulbar symptoms are dysarthria, dysphonia, and dysphagia and hence patients may be referred in the first instance to an otolaryngologist.[17] Spastic dysphonia can resemble adductor spasmodic dysphonia resulting in misdiagnosis and delayed neurology referral until the development of additional symptoms.

Instrumental assessment of swallow is warranted with a modified contrast swallow/VFSS, or if patient mobility issues exist, a FEES may be undertaken.

Dysphagia rehabilitation is focused towards compensatory strategies and interventions to ensure safe oral intake. These include:

- postural changes in the body and head or specific swallowing manoeuvres: chin-down position or head turned to the weakened side to redirect the bolus and reduce the risk of aspiration, also includes cervical collar or modified utensil use
- behavioural: scheduling meal times when patient most alert, smaller more frequent meals
- dietary: adjusting bolus volume or viscosity with thickeners, increased calorie dense supplement/foods.

In some cases, enteral feeding options (e.g. gastrostomy) may be required if there is inadequate oral intake or a significant aspiration risk is determined. Transcutaneous neuromuscular electrical stimulation has some limited benefit in stroke patients and surgery may have a role if there is evidence of pharyngo-oesophageal dysfunction.

Spinal pathology

Kyphosis or lordosis, previous spinal surgery, and cervical anterior osteophytes may result in dysphagia. Anterior osteophytes occur in 20–30% of the elderly population and are usually asymptomatic (Figure 48.7). Most commonly, they occur in diffuse idiopathic skeletal hyperostosis, spondyloarthritis, post trauma, or are congenital. Dysphagia is the most common symptom as osteophytes are located between C4 and C7 and as a result, constriction or stenosis of the retropharyngeal space and cervical oesophagus occurs either secondary to direct compression or inflammation (direct friction of bone spurs and soft tissue). The normal epiglottis tilt lies over the laryngeal inlet at this level.

The history will suggest dysphagia to solids then progressing to liquids. Aspiration arising from restricted movement of epiglottis and the larynx should be excluded. On flexible nasendoscopy a narrowing or bulge of the posterior pharynx wall may be evident with impedance of complete epiglottis closure and aspiration on swallow assessment from bolus deflection towards the larynx.

Conservative measures include anti-inflammatory or analgesic medication, muscle relaxants (if associated with osteophyte-induced inflammation, pain, and muscle spasm), or appropriate swallowing strategies and adaptations to diet. In cases where there is a significant impact on quality of life (significant weight loss, malnutrition, and/or aspiration), a referral for surgical resection of osteophytes may be considered. However, case series are small in number and the surgery in itself is associated with significant risks and has the potential to worsen dysphagia. In elderly patients with severe dysphagia and multiple comorbidities, enteral feeding options may be more appropriate.

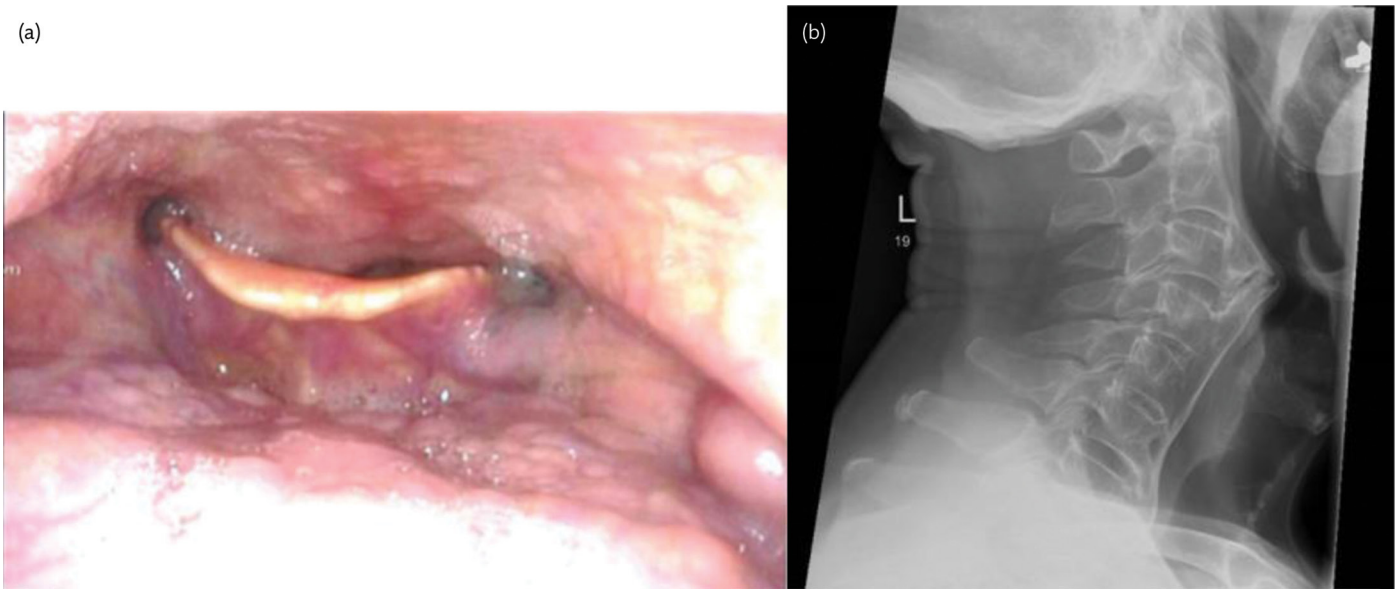

Figure 48.7 Flexible nasendoscopy view (a) demonstrating smooth asymmetrical prominence of posterior pharyngeal wall preventing dorsiflexion of epiglottis and partial occlusion of right pyriform fossa and glottis. Plain lateral cervical X-ray confirming the presence of significant C3/C4 osteophytes in same patient (b).

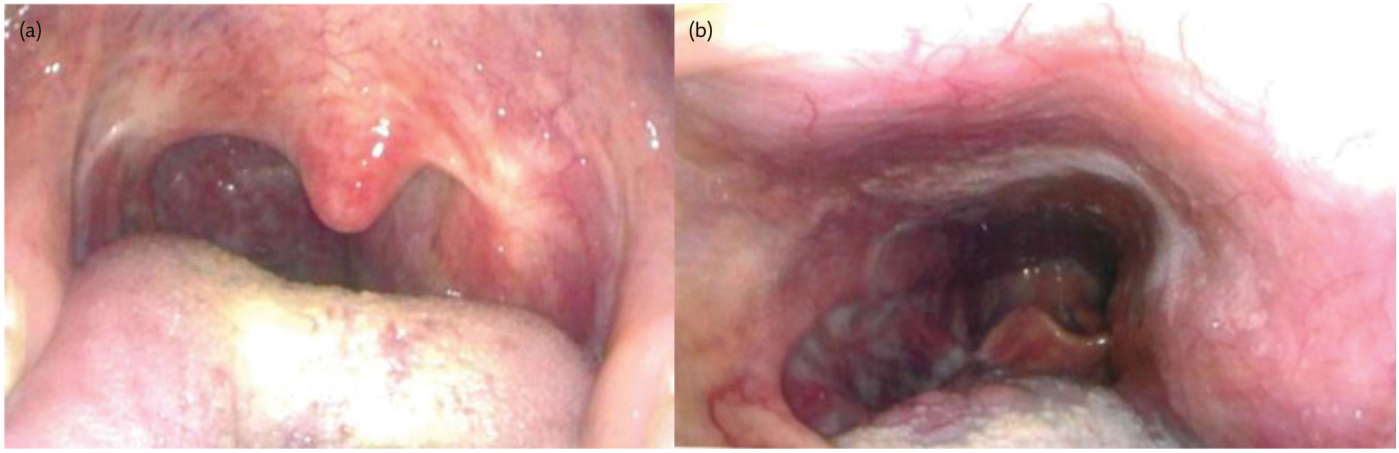

Figure 48.8 Intraoral view of fullness (a) oropharynx (patient had undergone a tonsillectomy in childhood). Asymmetry of left pharyngeal wall noted on flexible nasendoscopy representing underlying parapharyngeal mass in same patient (b).

Head and neck tumours

Tumours of the upper aerodigestive tract may affect swallow due to mechanical obstruction, infiltration resulting in reduced pliability of soft tissue, direct invasion of underlying musculature and fixation, nerve involvement, and loss of sensation/function and pain.

Clinical history and examination including flexible nasoendoscopy will elucidate common head and neck malignancy presentations.

Rarer parapharyngeal tumours (Figure 48.8) on intraoral examination should be assessed. The commonest initial presentation symptoms include an intraoral or cervical mass, otalgia, and dysphagia.[18]

The management of hypopharyngeal, oropharyngeal, nasopharyngeal, and laryngeal cancer and neck lumps are discussed elsewhere in this textbook.

Vocal cord palsy

The cricopharyngeus muscle receives innervation from the recurrent laryngeal nerve and the pharyngeal plexus bilaterally. Sensation is supplied by the recurrent laryngeal nerve to the ipsilateral subglottis and glottis and a branch of the internal superior laryngeal nerve to the supraglottis.

In unilateral vocal cord palsy (Figure 48.9a), symptomatic dysphagia occurs in up to 69% and aspiration in 20–50% of cases (Figure 48.9). Central lesions and acute-onset unilateral vocal cord palsy are associated with more severe dysphagia.[19] Glottis incompetence (Figure 48.9b) but also pharyngeal weakness and decreased sensation may contribute to swallowing dysfunction.

Laryngeal electromyography may assist in predicting prognosis in the preserved nerve setting and deciding upon treatment options. Primary intervention is usually to address voice concerns with either injection laryngoplasty (temporary) or type 1 thyroplasty (permanent). Equivocal results are found with respect to impact on dysphagia outcomes. These laryngeal entities are described in detail in Chapters 47, 71, and 72.

Post-cricoid web

Oesophageal webs are most commonly found along the anterior wall of the lower hypopharynx and upper cervical oesophagus. Although

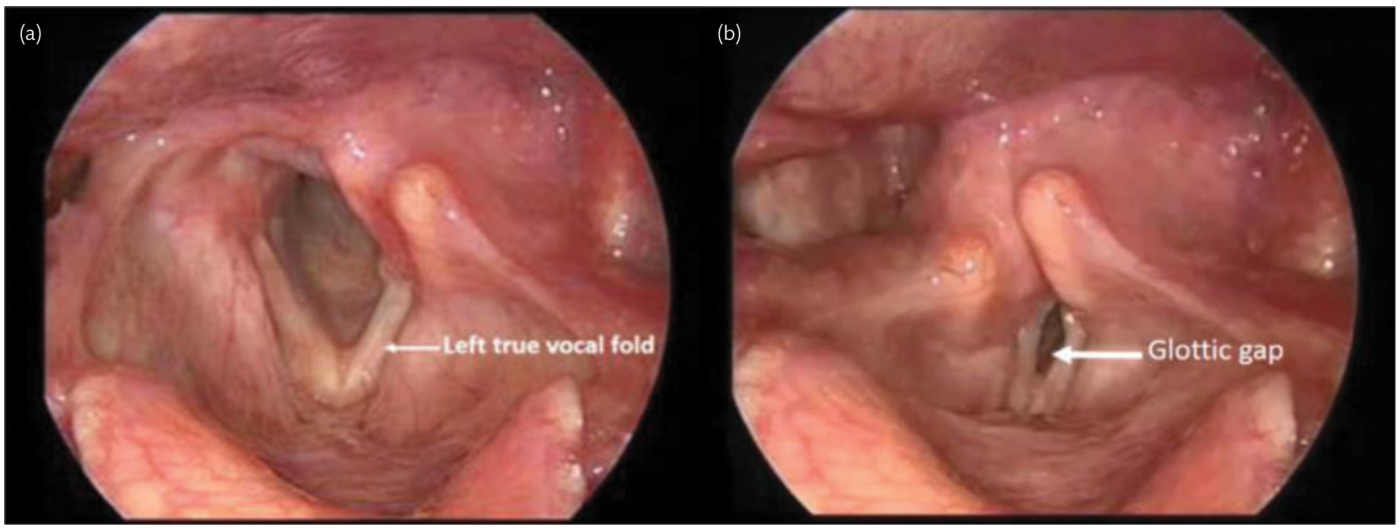

Figure 48.9 Left true vocal fold paralysis shows lateral position and foreshortening of vocal cord (a) and a glottic gap during phonation (b).

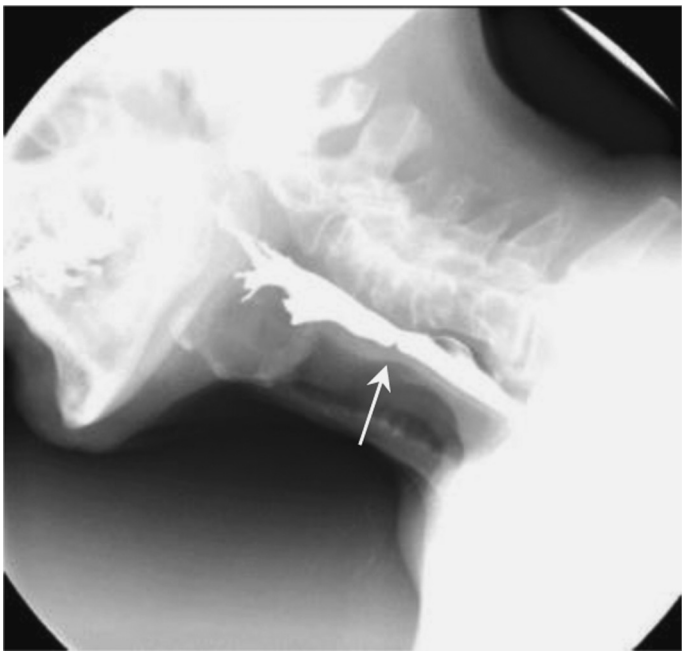

Figure 48.10 Incidental finding of anterior wall indentation (marked by arrow) on barium swallow suggestive of post-cricoid web.

largely incidental findings and asymptomatic, they may cause intermittent dysphagia to solids with a 'lump'-like sensation in the throat. In combination with iron deficiency anaemia and glossitis this is characterized as Plummer–Vinson syndrome. The condition is mainly reported in multiple case series and presents with intermittent dysphagia and signs of anaemia on oral examination (angular cheilitis, stomatitis) and if severe anaemia, koilonychia.

Post-cricoid webs may be missed on barium swallow but more clearly defined on VFSS as 1–2 mm-wide shelf-like lumen filling defects (**Figure 48.10**).

Assessment under endoscopy often results in simultaneous treatment as the scope ruptures the web as is passed through or may be addressed with dilatation. Endoscopic surveillance in Plummer–Vinson syndrome in view of the association with post-cricoid squamous cell carcinoma is recommended.

Thyroid goitres

Goitre-related oesophageal compression and swallowing dysfunction is evidenced in both objective and subjective measures. Increased transit and motility disturbance is evident on scintigraphy and manometry and improved cross-sectional area of the cervical oesophagus on magnetic resonance imaging post thyroidectomy.[20]

Patient-based questionnaires are heterogeneous and difficult to compare. However, studies that have used the goitre symptom scale of the Thyroid-Specific Patient-Reported Outcome (ThyPRO), SWAL-QOL, and Glasgow–Edinburgh Throat Scale have shown improved swallow following uncomplicated thyroidectomy. This tends to be more common after 3 months with no difference noted between hemithyroidectomy versus total thyroidectomy.[21]

Dysphagia related to subcervical goitre is commoner than cervical goitres and may not be apparent on neck examination (**Figure 48.11**). Subtle signs of laryngeal deviation or even vocal fold immobility may be seen on flexible nasendoscopy. Clearly, a malignant cause for the latter should be excluded but may be secondary to underlying compression of the recurrent laryngeal nerve. Previous chest X-ray images, if available, should be reviewed for signs of a goitre not evident on clinical assessment. Ultimately, cross-sectional imaging may be required and, in some cases, a modified contrast

Figure 48.11 Coronal contrast-enhanced computed tomography image of a retrosternal thyroid goitre to level of aortic arch giving rise to symptoms of compression and dysphagia (a). Photograph of thyroid goitre specimen removed through a cervical approach (b).

swallow to fully assess the dynamic impact on swallow. Careful evaluation of the remaining oesophagus is required to ensure no other cause exists for dysphagia. The management of thyroid disorders is set out in Chapter 59.

Controversies

High dysphagia

Patients are often referred with 'high' localized dysphagia for suspected head and neck cancer and this should be considered a misnomer. The assessment of a patient presenting with dysphagia should rule out potential causes from the oral cavity to the stomach. In patients reporting localization to the suprasternal notch, over 50% is due to lower oesophageal disease.[3] Three of the largest TNO series to date[22–24] have demonstrated that positive pathology is present in 44–52% of patients with the majority being oesophageal related (oesophagitis, gastritis, hiatus hernia, Barrett's oesophagus, candidiasis, stricture, or carcinoma).

Endoscopy assessment of the oesophagus therefore should be considered in the setting of a normal flexible pharyngolaryngoscopy assessment in patients referred with dysphagia. This is easily facilitated in centres where TNO is available and if not, onward referral to gastroenterology should be considered.

Contrast swallow

There are few indications for contrast swallow without prior endoscopy. There is high reporting variability between radiologists in the incidence of dysmotility and reflux.[25] Therefore, the role of contrast swallow is limited to failed or limited access to endoscopy, complementary to endoscopy for preoperative assessment of Zenker's diverticulum, or to assess for motility disorders if manometry is not available/possible.

Rigid panendoscopy

With the advent of high-resolution cross-sectional imaging, the role of rigid panendoscopy is to assess disease resectability and perform a simultaneous biopsy where office/clinic-based biopsy is not available or feasible. Rigid oesophagoscopy, as conducted in otolaryngology practice fails to detect up to 40% of lower oesophageal cancer (beyond 24 cm from incisors) in patients presenting with pharynx localized dysphagia.[3] Flexible oesophagoscopy versus rigid oesophagoscopy offers several advantages including reduced complication rates of oesophageal perforation[26] and ameliorating anatomical issues (limited neck extension, unfavourable dentofacial parameters, oesophageal strictures). Although overall secondary incidence of oesophageal malignancy is low in patients with head and neck squamous cell carcinoma, in certain primary subsites (e.g. oropharynx, hypopharynx) flexible endoscopy assessment[27] is best placed following anatomy-altering surgery and/or chemoradiotherapy with minimal risk.

Tips and pearls

- Closer assessment with cross-sectional imaging (magnetic resonance imaging) is essential for suspected deep seated or midline tongue base lesions (with no significant asymmetry apparent) and infiltrative tonsillar lesions which may be easily missed.
- Record and playback during TNO scope removal will aid closer review and inspection of the post-cricoid region.

Guidelines

Dysphagia assessment should include a comprehensive assessment of aetiology from the oral cavity to the stomach. Although oropharyngeal or 'high' dysphagia is often referred to otolaryngologists, the subjective feeling of symptoms to the throat/neck as alluded to earlier in this chapter may relate to oesophageal pathology.

To this end, the reader is recommended to review the World Gastroenterology Organization guidance,[28] which provides a practical approach to dysphagia.

Guidance based upon a systematic review of evidence rated according to the Grading of Recommendation Assessment, Development, and Evaluation (GRADE) approach has been published by the Canadian Association of Gastroenterology.[29] This consensus document summarizes 13 statements focused on the role of strategies for dysphagia assessment.

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49

Sore throat

Jenny Montgomery

Introduction

Throat pain is one of the most common presentations to ear, nose, and throat (ENT) departments. It is estimated that a third of the population will have a severe episode of throat pain, and just over a third of those will seek medical help for this reason.[1] ENT practitioners will attend to sore throats in every aspect of practice, from outpatients to emergency work. The causes of throat pain are varied. It is important to be able to filter these into benign and malignant causes while simultaneously managing patient expectations.

The anatomy of throat pain

The pharynx is a muscular tube that has breathing and swallowing functions. It also serves to aerate the middle ear through the Eustachian tube and assist with speech. It runs from the skull base to the inferior border of the cricoid ring anteriorly and to the inferior edge of the sixth cervical vertebra posteriorly. The posterior wall of the pharynx rests against the prevertebral fascia. It can help to think of the pharynx in layers as shown in Table 49.1.[2]

The pharynx is compartmentalized into the nasopharynx, oropharynx, and hypopharynx (laryngopharynx). These divisions are useful for describing sites of pathology and are shown in Table 49.2, along with the anatomical boundaries and contents.[3,4] The subsites used to describe the site and stage of cancers are discussed further in Chapters 56 and 60.

The outer circular muscle layer is comprised of the constrictor muscles as shown in Figure 49.1. The constrictor muscles are the superior, middle, and inferior. They are paired skeletal muscles that contract involuntarily during swallowing. They blend into each other in the posterior midline at the pharyngeal raphe. The inner longitudinal muscles are the palatopharyngeus, salpingopharyngeus, and stylopharyngeus. These muscles elevate the larynx when swallowing. They also shorten the pharynx to assist with swallow and speech. The constrictor muscles are all supplied by the pharyngeal branch of the vagus (cranial nerve X) nerve and the pharyngeal plexus which receives input from the vagus and glossopharyngeal nerves, as well as the sympathetic nerve supply. Palatopharyngeus and salpingopharyngeus are both supplied by the pharyngeal branch of the vagus nerve and the pharyngeal plexus. The stylopharyngeus is supplied by the glossopharyngeal nerve.

Table 49.1 Fascial layers of the pharynx

Depth of layer	Fascial layer
Innermost	Mucous membrane of pharynx of which the submucosa contributes to *pharyngobasilar fascia*
Middle	*Muscular layer* with inner longitudinal muscles and outer circular muscle fibres
External	*Buccopharyngeal fascia* which coats the muscular layer. This fascial layer blends with the pretracheal fascia

The pharynx also contains a rich supply of lymphatic tissue, which forms an incomplete circle. This is known as Waldeyer's ring or the pharyngeal lymphatic ring, and consists of the adenoids (pharyngeal tonsils), the tubal tonsils (sited in the nasopharynx, see Figure 49.2), the palatine tonsils, and lingual tonsils. The lymphatic drainage as relevant to neoplasms is discussed in Chapters 56 and 60.

Also relevant to potential causes of throat pain, the ear is uniquely supplied by four cranial nerves (branches of the trigeminal, facial, glossopharyngeal, and vagus nerves) as well as branches C2 and C3 of the cervical plexus. This complex innervation can allow for throat pathology to create non-otogenic sources of otalgia. An example of referred otalgia occurs with supraglottic cancer due to activation of the internal branch of the vagus nerve.

Considering the anatomy of the pharynx may also help to locate the aetiology of the pain. Figure 49.2 shows a cross-sectional of the pharynx at the level of the palatine tonsils. Given the close anatomical relationship, it is easy to see why musculoskeletal pain can manifest as throat pain. Similarly, parapharyngeal pathology or dental issues can create throat pain, as can the rich amount of lymphatic tissue within the oropharynx.

Assessment

Assessment of throat pain begins with history taking. A thorough history usually points the clinician in the right direction early in the consultation. Some simple questions related to persistence,

Table 49.2 Pharyngeal contents and boundaries

Pharyngeal section	Boundaries	Contents
Nasopharynx	Superior: skull base Inferior: just superior to roof of soft palate Lateral: medial pterygoid plates and superior constrictor Posterior: posterior pharyngeal wall (superior constrictors) Anterior: right and left nasal cavities	Eustachian (pharyngotympanic) tube Torus tubarius (formed by cartilage of the Eustachian tube) Salpingopharyngeal fold (formed by muscle) extends inferiorly from torus tubarius Fossa of Rosenmüller (pharyngeal recess) posterior and lateral to torus tubarius Adenoids (pharyngeal tonsils) Tubal tonsils (located inferior to torus tubarius in submucosa)
Oropharynx	Superior: roof of soft palate Inferior: soft tissue tip of epiglottis, bony superior surface of hyoid bone Lateral and posterior: formed by superior and middle constrictor muscles Anterior: palatoglossal arch	Palatine tonsils Lingual tonsils
Hypopharynx (laryngopharynx)	Superior: inferior to tip of epiglottis Inferior: inferior border of cricoid cartilage Lateral and posterior: formed by middle and inferior constrictor muscles externally and palatopharyngeus and stylopharyngeus internally Anterior: larynx	Muscles surround distinct areas of pyriform fossae bilaterally and post-cricoid mucosa

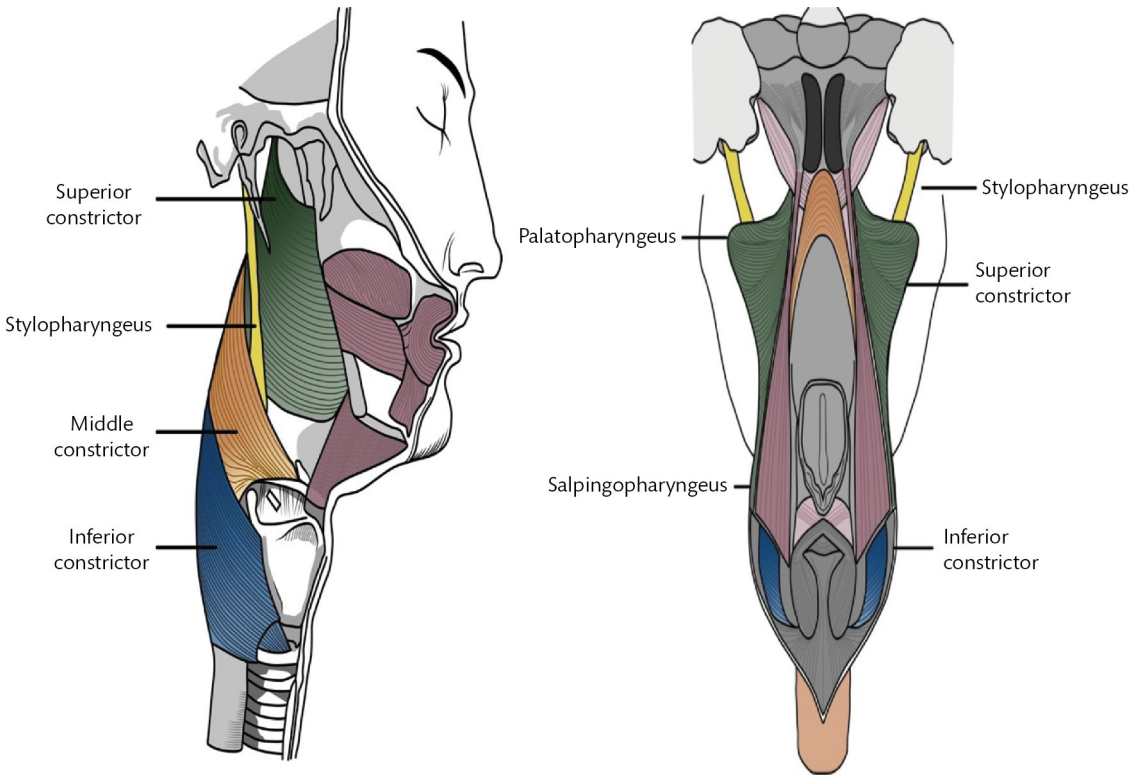

Figure 49.1 The circular and longitudinal muscles of the pharynx.

intermittence, severity, character, and location of pain, including whether the pain is central or lateralized, can help guide with causation. The onset and duration will also help to determine the underlying problem. Focused history relating to demographic information including smoking, alcohol, and drug use status can help to determine a patient's risk of having different head and neck pathologies. Occupational factors such as an air-conditioned working environment are relevant to their throat health. Other useful past medical history includes steroid inhaler use, gastro-oesophageal reflux disease, regular use of mouthwashes, or whether there is any weight loss.

Most clinicians will be worried about missing an underlying throat cancer in a patient with throat pain; however, it is relatively unusual for early-stage cancers to cause pain. Accordingly, throat pain is absent from the National Institute for Health and Care Excellence guidelines for symptoms suggestive of suspected cancer.[5] The Scottish referral guidelines for suspected head and neck cancer do include throat pain as persistent throat pain or pain on swallowing

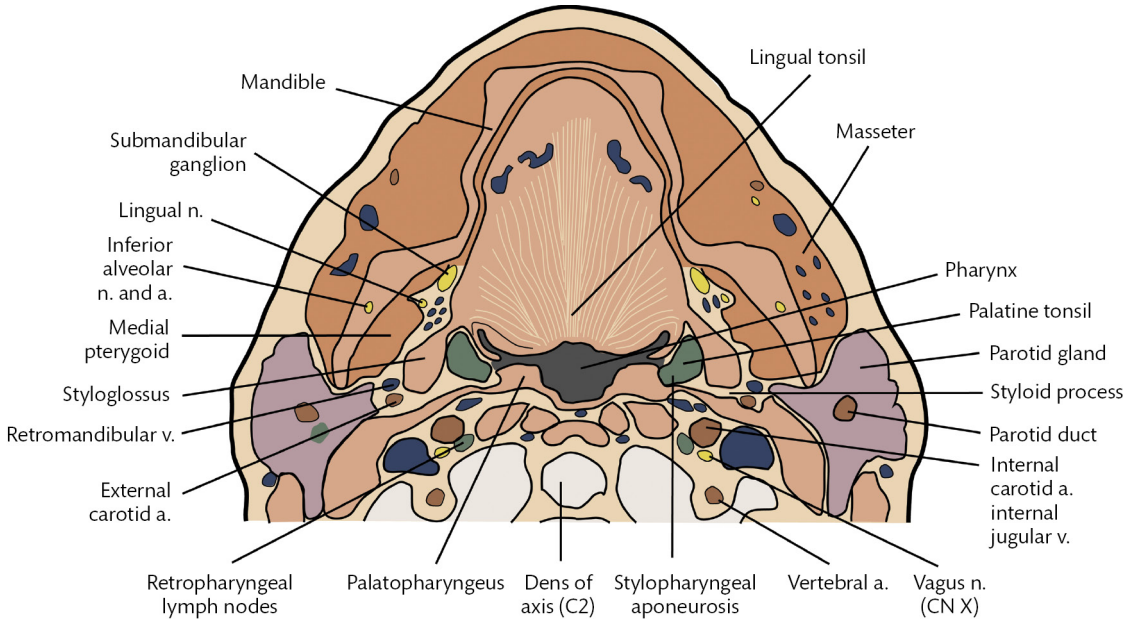

Figure 49.2 Cross-sectional anatomy of the pharynx at the level of the tonsil.

lasting longer than 3 weeks.[6] Another useful tool when considering throat pain, and the additional symptoms that patients can present with, is the UK-developed head and neck risk calculator, currently in its second version.[6]

Examination should include comprehensive assessment of the upper aerodigestive tract mucosa and the neck; principles of clinical assessment and detailed description of examination of these areas can be found in Chapters 46–48 and 50. Some points relevant to sore throat are identified here.

In the nasopharynx, it is not always easy to inspect the pharyngeal recess (fossa of Rosenmüller). Looking at this area is crucially important in patients with a unilateral ear effusion, as this can imply that there is an obstructive lesion in the nasopharynx. Instructing the patient to breathe through their nose will separate the soft palate from the posterior pharyngeal wall. Taking time to inspect the vallecula by asking the patient to stick out their tongue is important, as this is another hidden area until the tongue is protruded. Similarly, inspecting the hypopharynx fully by asking the patient to say 'ee' or 'hay' will give a better view. The last tip to open the hypopharynx for inspection is to pinch the nose gently with the examiner's hand that is holding the scope and ask the patient to close their mouth and puff out their cheeks. This inflates the pyriform fossae with air and will allow a better surface mucosal assessment.

Parapharyngeal pathology or infection may be seen as a bulge in the lateral pharyngeal wall, which is best investigated further with axial imaging. The only area of the hypopharynx that cannot be seen with flexible laryngoscopy is the post-cricoid mucosa. Pathology in the post-cricoid area could be suggested by secretions in the hypopharynx that do not clear away with swallowing.

Acute throat pain—infective causes

Throat pain can be characterized as acute or chronic. Recurrent acute throat pain in the context of other infective parameters such as fever, painful odynophagia, and tender cervical lymphadenopathy is usually tonsillitis. This tends to present as an emergency admission via primary care, when being unable to swallow becomes the criteria for hospital admission. The differential diagnosis for this type of patient is acute bacterial tonsillitis, infectious mononucleosis, peritonsillar abscess, deep neck space infection, or supraglottitis. Clinical examination can help further, with infective exudate seen on the tonsils in bacterial tonsillitis and infectious mononucleosis. Peritonsillar abscess tends to be associated with asymmetrical soft palate swelling, deviation of the uvula to the contralateral side, and trismus. Deep neck space infection also tends to be associated with trismus but also decreased neck mobility. Supraglottitis tends to be associated with pain, drooling, and stridor. Intraoral examination, neck palpation, and flexible laryngoscopy for all these patients is important.

Investigations in acute infective episodes include measurement of routine blood tests to obtain inflammatory markers including C-reactive protein and the white cell count. In cases of infectious mononucleosis, a monospot can be sent to the haematology laboratory. A temporary transaminitis is also frequently seen. Throat swabs do not tend to alter management of throat pain and are not recommended. It is common for sepsis bundles to be performed in the emergency department, of which the Sepsis Six is the most common.[7] This includes serial monitoring of lactate. Although its use is commonplace, and for other organ systems is linked to decreased mortality, it has not been widely used in neck space infection. If neck space infection is suspected, obtaining a contrast-enhanced computed tomography scan of the neck can help to determine the extent of any abscess and facilitate surgical planning.

Management of straightforward tonsillitis leads to rapid resolution, and often patients can go home after around 12 hours of rehydration and intravenous antibiotics. Choice of antibiotic is dictated by local microbiology protocols, but generally penicillin is used (unless contraindicated due to allergy). Drainage of peritonsillar abscess is usually one of the earliest skills learned in otolaryngology training and is easily done in the treatment room. Gathering all equipment and setting up prior to beginning aspiration of the abscess makes it easier for the patient and yourself. Local anaesthetic spray, a sick

bowl, and a drink of water for the patient to rinse their mouth afterwards will give the patient some comfort. Use of a 22-gauge large-bore needle or intravenous cannula is preferred to accommodate the viscosity of any purulent material aspirated. Drainage of the abscess needs to be lateral to the tonsil, and in the most dependent or inferior area to perform the most effective aspiration. Often patients feel instantly better following this and it is possible for them to start swallowing again. When discharging patients from hospital, it can be useful to have a conversation about indications for tonsillectomy; these may include five episodes of tonsillitis per year, for 2 consecutive years. Other pragmatic indications to be discussed with patients are more than one episode of peritonsillar abscess or more than one episode of hospitalization due to tonsillitis.

Rates of adult supraglottitis are around 5/100,000 population.[8] The management of paediatric supraglottitis is different and is dealt with in Chapter 76. Adults present with throat pain, painful dysphagia, drooling saliva, and stridor. A useful rule of thumb is that if a patient has a painful throat and is septic, but there is no examination evidence of oropharyngeal sepsis, supraglottitis should be considered. Patients will have raised inflammatory markers but the main management issue at presentation is the airway. Flexible laryngoscopy will establish whether intubation of the airway is feasible or whether local anaesthetic tracheostomy should be planned. Decision-making requires involvement of anaesthetic staff. Depending on the degree of oedema seen on the epiglottis and surrounding supraglottic tissues, patients can be managed conservatively with antibiotics and steroids. If a patient is a smoker, it is worthwhile reviewing them in the outpatient clinic after the acute inflammation has settled in case there has been an infection surrounding a cancer. This could be difficult to see in the context of inflammation, and may only become apparent once the acute phase has settled.

Deep neck space infection rates are increasing,[9] and successful drainage is the key to management. This is, to some degree, thought to be due to more stringent criteria for performing tonsillectomy.[10] Deep neck space infection can be highly morbid resulting in multispecialty care including ENT, oral maxillofacial surgery, and intensivists, and is discussed in detail in Chapter 62. Patients can require tracheostomies, feeding tubes, and anticoagulation if there is evidence of venous thrombosis and if subjected to more than one episode of surgery. Commonly, deep neck space infection is related to dental sources and intraoral examination will make this clear. Another common site for neck space infection to originate is the tonsil, resulting in parapharyngeal abscesses (**Figure 49.3**) or retropharyngeal abscesses. These sources of infection can sometimes be managed with intraoral drainage. If there are signs of a collection lateral to the carotid artery then management via a neck incision will avoid a sump of fluid collecting within the neck. Perhaps future iterations of tonsillectomy guidelines will include less strict surgical criteria in an effort to combat rates of deep neck space infection.

Fungal infections such as those caused by *Candida* spp. can cause pain and discomfort. Candidiasis presents as several clumps of white material that can be scraped away with a spatula. Unexplained candidiasis should prompt checks for immunosuppressive conditions such as diabetes mellitus or human immunodeficiency virus. It is also seen in patients with xerostomia and when there has been previous radiotherapy to the neck. Conservative advice can include meticulous hygiene measures for dentures, as they harbour fungal colonies.

Figure 49.3 Parapharyngeal abscess, extending posterolateral to neck vessels. Scan was obtained 48 hours after intraoral drainage, when the patient was found to be worsening.

Medical therapy includes nystatin drops dosed as 100,000 units four times a day for a week. If the infection involves the pharynx, is recalcitrant or widespread, oral fluconazole can be used at 50 mg daily for 7–10 days. This can be prescribed in capsules or liquid form, which is useful if the patient has a nasogastric tube. If the oral surfaces are painful, then miconazole gel can be sparingly applied to the tongue or gingivae. Small amounts of this can be used to brush dentures with if they are suspected as the source of the infection.

Throat pain related to malignancy

Throat pain attributable to head and neck cancer is seen with advanced disease. Early-stage oropharyngeal cancer can be remarkably pain free, with patients only reporting vague and inconsistent throat symptoms. Since human papillomavirus-related oropharyngeal cancer was recognized more than 20 years ago, the most common presenting symptom is the presence of neck adenopathy. This contrasts with supraglottic cancer, which may lead to odynophagia even in the earliest stages. Persistent odynophagia is an ominous symptom, second only to unexplained neck lump in terms of positive predictive value in the red flag symptoms of head and neck cancer.[11] Patients can be seen visibly wincing even when swallowing saliva, and even a negative outpatient flexible laryngoscopy should prompt additional investigations with narrow band imaging assessment,[12] cross-sectional imaging, or examination under anaesthesia. Symptom control can be overlooked at the initial visit and requires to be addressed at the outpatient appointment. The pain caused by such a tumour will also prevent a patient from eating properly, thus requiring early input from the dietetic team (see Chapter 46).

Pain associated with late-stage head and neck cancer is the symptom that requires our utmost attention. It can be clearly seen when patients are in discomfort, and this can be a difficult symptom

to bring under control. Pain can be related to the cancer itself or associated with oncological or surgical treatment. It may be opioid responsive or neuropathic in nature, requiring medication such as amitriptyline or gabapentinoids as an initial consideration. When prescribing opioids, it is important to consider the high rates of co-existent pulmonary disease seen, to avoid respiratory depression. Optimization of analgesia is often expertly performed by clinical nurse specialists and colleagues in palliative medicine and a team-based approach to complex analgesia requirements will be associated with better management.

Foreign bodies and trauma

These are common emergency presentations—foreign bodies in adults tend to be food boluses, bones from food, and occasionally dentures. In children, this is more varied. The most important childhood foreign body to remove immediately due to its corrosive properties is the button battery. Many of these are radio-opaque and will be visible on soft tissue neck X-rays or chest X-ray. Fish bones are not always visible on X-rays and a good clinical examination is all that is required. If examination is normal, the patient can be reassured that the pain should resolve over coming weeks and counselled to return if worsening. The history will usually offer the diagnosis in terms of the onset in relation to eating. Removal can be performed with a headlight and forceps with the patient awake or in theatre if not possible.

Chronic throat symptoms

Tonsilloliths

These can also be referred to as tonsilloliths or tonsil stones. They are a hard accumulation of calcium salts within the crypts of the palatine tonsils. They are thought to be a mixture of trapped debris and *Actinomyces* infection. *Actinomyces* bacteria are seen in a higher proportion of patients undergoing surgery for tonsilloliths compared to tonsillectomy for recurrent tonsillitis.[13] Patients tend to present with throat discomfort and halitosis, indicating there is a quality-of-life issue associated with tonsilloliths. Treatment within the NHS tends to be conservative, with advice to gargle salt water after meals with the aim of preventing build-up of debris. Other conservative advice can involve commercially available probiotic drinks; however, no tangible evidence exists to support their use. Patients will manually try and remove them. Surgical treatment may include cryptolysis using either laser or coblation. Tonsillectomy prevents tonsilloliths from forming.

Eagle syndrome

Also known as stylohyoid syndrome, this is a controversial condition—with some doubting its existence.[14] Pain can be felt unilaterally, with radiological bilateral findings as in Figure 49.3. It is associated with pain and thought to be secondary to either elongation or deformity of the styloid process or calcification of the stylohyoid ligament (Figure 49.4). Presentation is persistent unilateral throat pain—usually prompting axial imaging, where it is identified. The differential diagnosis includes malignancy or neuralgia of the trigeminal or glossopharyngeal nerve. Medical management includes trialling low-dose amitriptyline or gabapentinoid medication and titrating to effective dosages. Surgery for this condition is performed rarely, but, in appropriate cases, appears to offer good symptom relief. Surgery can be performed intraorally or via an external approach to the styloid process.

Figure 49.4 Bilateral stylohyoid calcification.
Image courtesy of Dr Andrew Hunter.

First bite syndrome

First bite syndrome is characterized by intense pain or cramping in the region of the parotid. It is associated with the first bite of the meal, or by anticipating the first bite. The pain tends to lessen with each subsequent bite but will reoccur at the next meal. The pain is perceived in the parotid or towards the angle of the mandible but may be referred into clinic as throat pain. It is thought to occur from disruption of the sympathetic innervation of the parotid gland. Generally, it is seen in two groups of patients. Any type of surgery that may disrupt the sympathetic chain in the upper neck can be responsible. Examples include prophylactic ligation of the external carotid artery branches for transoral surgery, and removal of parapharyngeal space tumours.

If a patient presents with these symptoms and does not have a background of neck surgery, imaging (either computed tomography scan or magnetic resonance imaging of neck) should be performed to investigate for potential malignant causes—primarily of the parotid. Spontaneous resolution is often the rule. In recalcitrant cases, analgesia can include neuropathic agents such as gabapentinoids or botulinum toxin injected into any residual parotid tissue.

Other general irritants

Mouthwashes are sold commercially with the aim of reducing dental debris and gingival health but are frequently employed for gargling.

Anecdotally, many patients present with burning throat discomfort after using these. Advice can be given to switch to saltwater rinses or soda bicarbonate rinses, which tend not to cause the same level of irritation.

Vapes and e-cigarettes are increasing in popularity and are often successfully used by patients as a means of smoking cessation. Often these products are coloured and flavoured. Flavours tend to be made using food products which are meant to be ingested rather than inhaled. In addition to this, the amount of nicotine within them can also vary which is another irritant. When the fluid level within the vape is too low or empty but they are still used it leads to a steam inhalation, which can also cause pain. This is sometimes termed 'a dry hit'. Advice can be given regarding avoidance of coloured, flavoured vaping liquids. An opportunity for general health advice can also be given regarding avoiding cannabis oil vaping. Oils do not dissolve in water, and this can potentially be damaging to the lungs. This should not detract from the fact that vapes and e-cigarettes do help people to stop smoking. Advice should be given to use a vape where it is possible to titrate down the amount of nicotine contained towards stopping completely.

General management of chronic throat symptoms

Chronic throat symptoms are usually termed as discomfort, globus sensation ('a feeling of something in the throat'), catarrh, mucus, and throat clearing (see Chapter 48). This is sometimes termed troublesome throat awareness[15] rather than globus with the aim of improving patient understanding. Management of patient expectations for these symptoms is crucial, as medical interventions have no role or benefit. A common strategy previously was to treat these patients indiscriminately with proton pump inhibitors with the aim of treating silent reflux disease. The landmark and recently published TOPPITS trial has shown that this has no benefit over placebo.[16]

Useful advice can include that these symptoms are common after an infection and can take several months to settle. Patient will often attribute this to catarrh, but a normal clinical examination can provide assurance that this is not the case, and in fact that this has become habitual. Advice to take a small sip of water each time they feel like they are about to clear their throat instead is a gentler method of removing secretions. Simple conservative measures such as avoidance of throat clearing and increasing oral hydration can be recommended. The presence of symptomatic heartburn and reflux can also be addressed. Lifestyle advice on weight loss and avoidance of eating in the evenings is useful. If patients are exposed to irritants at work, advice on wearing of protective masks should be given. Ultimately this management will be in the domain of the general practitioner, rather than the ENT surgeon. Finally, a discussion about stress and anxiety to explain that sometimes this can manifest in throat symptoms can provide reassurance. Time is what patients should be counselled over, explaining that throat symptoms will diminish over the coming months (rather than weeks). Clearly, the ENT clinic is not the ideal place to discuss psychological concerns, but sensitivity is needed as many patients acknowledge this as a cause for their symptoms. Some patients will require additional reassurance, and contact numbers to facilitate patient-initiated review can help. Generally, time spent providing explanations and advice, will help to close the care episode and prevent re-referral patterns.

The difference between discomfort and actual throat pain is subjective. If genuine throat pain is reported then a focused history surrounding the nature of the pain, timings, relationship to eating, severity, and laterality should be recorded. If examination is fruitless and clinical index of suspicion is low, then strategies could be patient-initiated review after an interval or consideration of axial imaging to exclude a submucosal lesion or parapharyngeal pathology. At this juncture, if imaging is negative, patients can be further reassured.

Tips and pearls

- If patients don't have red flag symptoms, they can be confidently reassured that their throat symptoms should settle with time.
- Trials of proton pump inhibitors for non-reflux symptoms are not indicated.
- During flexible laryngoscopy, pay special attention to the nasopharynx, vallecula, and laryngeal surface of epiglottis.
- If your outpatient stack has the capability, check mucosal surfaces with narrow band imaging.

Conclusion

Throat pain is common and having a strategy for managing patients in a reassuring manner is helpful. Persistent unilateral pain is worth further cross-sectional imaging. Early oropharyngeal cancers may not be painful, but supraglottic cancers can be painful. Thorough history taking and examination is important and is all that will be required for most patients. Most chronic throat symptoms will settle conservatively. A flow chart for suggested management is shown in Figure 49.5.

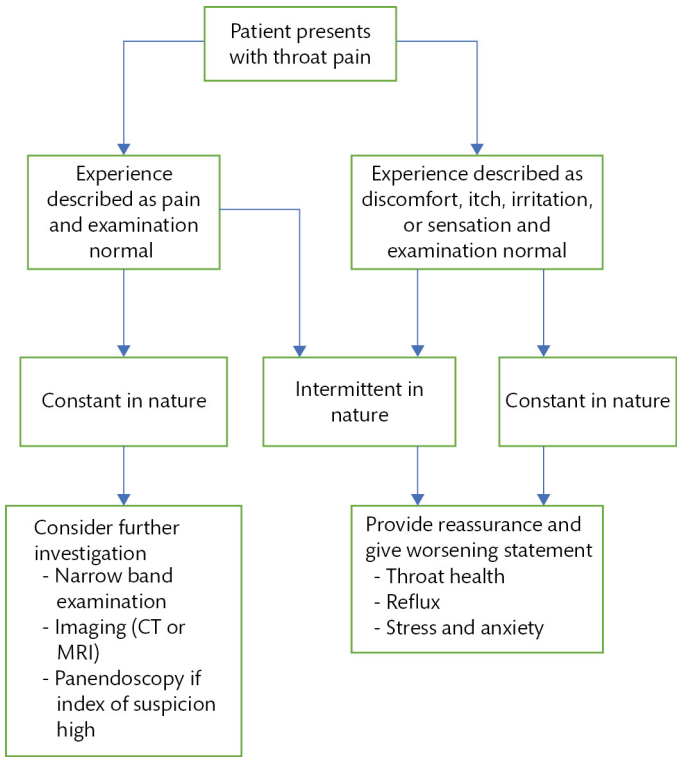

Figure 49.5 Suggested management algorithm for throat pain. CT, computed tomography; MRI, magnetic resonance imaging.

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50

Clinical assessment of neck lumps

David Walker and Cameron Davies-Husband

Introduction

The opportunity to assess a patient presenting with a head and/or neck symptom is to be considered a privilege. The combination of an insightful, empathetic manner, coupled with a sound knowledge base and grounding in clinical acumen are concurrently required to bring the surgeon swiftly to the correct diagnosis. The assessment of the head and neck patient is a sequential, stepwise process, with the success of each action being dependent on the former. Obtaining a comprehensive history is essential, though doing so without establishing rapport would be irksome. Similarly, performing a systematic examination is mandatory, but without the patient providing key information as to a rare eponymous syndrome, an otherwise-invisible clinical sign may be missed. The aphorism 'a careful history will lead to the diagnosis 80% of the time' is well indoctrinated into medical practice, seemingly originating from a paper by Hampton et al.[1] who investigated the impact of history-taking, clinical examination, and investigations in reaching the correct diagnosis. While there have been significant advances in medical diagnostics over the last two decades, including narrow band imaging, magnetic resonance imaging (MRI), positron emission tomography (PET)/computed tomography (CT), and high-definition endoscopy, the authors are of the opinion that a detailed clinical assessment is still among the most valuable of tools in the head and neck surgeon's armamentarium. As with all aspects of medicine, the Hippocratic oath of 'do no harm' remains a fundamental theme throughout the process and a thorough patient assessment ensures this promise, all the way to the operating theatre and beyond.

General appearance and first impressions

From an early age, we are encouraged 'not to judge a book by its cover'. Contrariwise, patient profiling, in the absence of prejudice or judgement, is a valuable skill, with the most experienced head and neck surgeons attesting to beginning the clinical assessment from the moment the patient enters the room. Naturally, the reader is urged to remain open-minded to the full range of diagnoses and not to jump to conclusions, for exceptions can (and do) occur. However, if one were to consult an 85-year-old, presenting with a parotid lump and multiple cutaneous scalp lesions, Occam's razor would implicate a metastatic squamous cell carcinoma over a pleomorphic adenoma. Similarly, a well-kempt, 25-year-old female, non-smoker, professional runner, is less likely to have serious underlying pathology compared to a dishevelled, 65-year-old male patient wheeled into the clinic on a chair, smelling strongly of cigarette smoke. Assessment of the patient's affect may provide valuable cues to their mental state, or if under the influence of alcohol. Similarly, tar-stained teeth or fingernails allude to heavy tobacco consumption. Oversized trousers tightened up incongruently around a cachectic waist with a belt may suggest rapid weight loss. These anecdotes emphasize the importance of the assessing clinician having a sound knowledge of disease epidemiology and aetiology, allowing them to prepare to ask the most useful questions in each case. Without such insight, the assessment process will seem laborious. Nothing inspires confidence more in a patient when the surgeon seems to understand exactly what the patient is experiencing through the questions they pose.

Obtaining a patient history

The consulting surgeon should make every effort to ensure that the patient's previous medical records are available for reference. Many prior head and neck cancer patients will have had multiple investigations and procedures/operations, the details of which will provide insight into their presenting complaint. Previous imaging and histology results should also be reviewed. Taking 2 minutes to familiarize oneself with the patient's case before the consultation allows the surgeon to begin conjecturing potential diagnoses and significantly abbreviates the time taken to obtain the most pertinent information once the consultation begins.

Most patients presenting with neck lumps will be apprehensive about their diagnosis, for it represents the quintessential manifestation of a potentially serious problem. 'Doctor, I have a lump …' is an opening statement to a subsequent death sentence in some patients' minds and as such, the surgeon's initial priority should be in gauging their anxiety and establishing rapport. If the patient appears particularly distracted, it is crucial to invest time to address their fears before proceeding, else the history may become hampered from their intrusive, negative thoughts. Once a non-emotive forum for discussion has been established, the surgeon should focus on the neck lump specifically, enquiring about its onset, duration, location, change in size, associated

pain, and exacerbating or alleviating factors. Associated symptoms, which may suggest serious upper aerodigestive tract pathology, include odynophagia, dysphagia, otalgia, haemoptysis, and hoarseness. Such 'red flag' symptoms, if elicited in the history, should be questioned about in detail, as above. Constitutional symptoms such as weight loss, pruritus, night sweats, malaise, and change in appetite should also be explored. At this point, the surgeon may be forming a provisional diagnosis, or at the least, begin to appreciate the risk of an underlying malignancy.

Next, the surgeon should address the patient's past medical history (**Box 50.1**). If the patient ultimately requires surgery, a full knowledge of such is crucial when considering such a procedure. General anaesthesia carries significant risk in the presence of cardiorespiratory comorbidities. Similarly, impaired surgical wound healing may be predicted in patients with diabetes mellitus (particularly in smokers) and free flap reconstruction may be impossible in the presence of peripheral vascular disease. A thorough knowledge of the patient's past surgical history is also required, including difficulty in intubation or adverse reactions to anaesthetic medications. All patient medications and allergies should also be accurately recorded.

Once a complete medical history is obtained, it is beneficial to ascertain risk factors pertaining to specific diseases (**Table 50.1**). Tobacco use is of paramount relevance, being the single most contributory factor to the development of cancers of the upper aerodigestive tract.[2] The patient's predilection for chewing tobacco, cigars, or cigarettes should be explored, for it may delineate the primary site of disease. Similarly, concurrent alcohol consumption has a multiplicative effect on carcinogenesis, so frequency, type, and duration should be recorded. Increasingly, the human papillomavirus has been implicated in the rise in observed frequency of oropharyngeal squamous cell carcinoma[3] and current guidelines have suggested exploring the patient's sexual history and practices, including frequency and modes of intercourse, in addition to number of sexual partners. Similarly, risk factors for communicable diseases such as the human immunodeficiency virus (HIV) and hepatitis virus should be confirmed. A history of radiation exposure should also be determined, for it is a risk factor for the development of numerous malignancies.

Box 50.1 Summary of key points when obtaining a history from a patient presenting with a neck lump

- Obtain previous notes, investigations, results.
- First patient impressions.
- Self-introduction:
  - Name, title, job desolation.
  - Establish rapport.
  - Put patient at ease.
- Presenting complaint:
  - Onset.
  - Duration.
  - Location.
  - Change in size (i.e. increasing, fluctuating).
  - Pain.
  - Exacerbating/alleviating factors.
  - 'Red flag' symptoms.
  - Constitutional symptoms.
- Past medial history.
- Past surgical history.
- Current mediations; drug allergies.
- Risk factors:
  - Environmental.
  - Occupational.
  - Recreational.
- Systems review.
- Performance status, frailty score.

The patient's social history and occupation will provide occasionally confirmatory, but always useful information in the context of a neck lump. Spot-diagnostic case presentations include the carpenter presenting with nasal obstruction and bilateral cervical masses (i.e. sinonasal carcinoma); the retired publican presenting with a chronic cough, hoarseness, and a supraclavicular mass (i.e. bronchial carcinoma); and the veterinary nurse presenting with cat scratches and painful cervical adenopathy (i.e. bartonellosis). Naturally this list is

Table 50.1 Examples of different presentations of neck lumps, due to varied associated symptoms and extrinsic risk factors; 'Occam's razor' diagnoses refer to the most *likely* diagnosis, according to the clinical presentation

Neck lump(s), with	Extrinsic risk factors	Occam's razor diagnosis
Haemoptysis, weight loss	Smoking, alcohol	Bronchogenic carcinoma
Stridor, hoarseness, dysphagia	Smoking, alcohol	Laryngeal squamous cell carcinoma
Dysphagia, odynophagia, otalgia	Multiple sexual partners, oral sex, smoking, alcohol	Oropharyngeal squamous cell carcinoma
Dysphonia, dysphagia	Radiation exposure	Poorly differential thyroid carcinoma
Fevers, night sweats	Radiation exposure	Lymphoma
	Endemic geography	Tuberculosis
	Cat scratches	Bartonella
Nasal obstruction, epistaxis	Carpenter, smoking, alcohol	Nasopharyngeal squamous cell carcinoma
Blood expectoration, tongue pain	Cigar smoking, alcohol	Oral cavity squamous cell carcinoma
Odynophagia, dysphagia, stertor, pyrexia, malaise	Newly enrolled university student	Infectious mononucleosis
Ecchymosis, cervical pain, torticollis	Professional kickboxer	Haematoma/carotid artery dissection
Fevers, night sweats, weight loss, odynophagia	Multiple sexual partners, intravenous drug usage	HIV
Scalp lesions	Farmer	Cutaneous squamous cell carcinoma

Table 50.2 Summary of steps performed in the examination of a head and neck patient presenting with a neck lump

Facies
- Facial nerve paralysis
- Lipodystrophy
- Oedema
- Eye changes

Skin
- Face
- Neck
- Scalp

Ears
- Auricle
- External auditory canal
- Tympanic membrane

Nose
- Anterior rhinoscopy
- Nasopharynx

Oral cavity
- Lips
- Hard palate
- Anterior (two-thirds of) tongue
- Buccal mucosa
- Alveolus
- Retromolar trigone
- Floor of mouth

Oropharynx
- Tonsillar pillars/palatine tonsils
- Soft palate
- Posterior (third of) tongue
- Posterior/lateral pharyngeal mucosa
- Vallecula

Larynx
- Supraglottis
- Glottis
- Subglottis*

Hypopharynx
- Posterior hypopharyngeal wall*
- Piriform fossa
- Post cricoid*

Neck
- 1a (submental)
- 1b (submandibular (gland))
- 2a, 2b (jugulodigastric)
- 3, 4 (carotid/muscular triangle)
- 5 (posterior, transverse cervical, supraclavicular)
- 6 (pretracheal, paratracheal, perithyroidal, thyroid (gland))
- Parotid gland
- Preauricular
- Retroauricular
- Suboccipital

Neurological
- I (sense of smell to substances)
- II (visual acuity, fields)
- III (extraocular movements, pupillary reflex)
- IV (superior oblique muscle)
- V (facial sensation/corneal reflex)
- VI (lateral rectus muscle)
- VII (facial symmetry at rest/during movement)
- VIII (hearing assessment)
- IX, X (gag reflux; vocal cord movement)
- XI (trapezius muscle movement/atrophy)
- XII (tongue movement/fasciculations/atrophy)

*Views of selected subsites are expected to be limited in the clinic setting and may require general anaesthesia for definitive inspection.

not attempting to be exhaustive, but to stress the importance of comprehensively exploring the patient's environmental risk factors. On a side note, taking time to ask what someone does for a living is an extremely humanizing question and helps the consulting surgeon both to build rapport and gain trust.

Finally, a brief, but succinct 'systems review' should be performed, detailing symptoms that may be extremely relevant, but not immediately so to the lay person. These questions should explore changes in the patient's cardiac, respiratory, gastrointestinal, endocrine, neurological, urological, musculoskeletal, and cutaneous systems.

Clinical examination

Once the surgeon has obtained a comprehensive history, specific areas may then be sequentially examined (Table 50.2). It is important to gain informed consent. The patient may feel confused or alienated if you ask to examine anything other than their neck lump (i.e. the reason they believed they were there). Accordingly, take a moment to explain that the examination involves multiple components.

Begin with inspecting the patient's face for asymmetry, examining the facial skeleton and soft tissues. Anti-retroviral HIV medications may result in lipodystrophy, presenting a 'gaunt' appearance with exaggeration of the muscles of expression and mastication. Hypothyroidism may present with periorbital oedema, coarse, dry skin with ptosis of the upper eyelids, and an archetypal 'blank' expression. Malignant tumours of the parotid gland may compromise the facial nerve, leading to ipsilateral facial muscle paralysis.

Next, the skin of the scalp and neck should be examined. These areas are particularly susceptible to sun exposure and the presence of cutaneous lesions should be ascertained, including squamous cell carcinoma, basal cell carcinoma, and malignant melanoma. The skin of the auricles, external auditory canal, and tympanic membranes are inspected under direct vision, then otoscopy, respectively. The postauricular region is an area that may be overlooked, harbouring discrete lesions or scars from previous surgery.

The examination should then proceed to inspection and palpation of the neck (Figure 50.1). Here, appreciation of anatomy

Figure 50.1 A photograph to demonstrate correct clinician–patient orientation during palpation of the neck. The clinician should position themselves behind the patient, placing both hands on the neck simultaneously.

is crucial to achieving an accurate diagnosis. While there is crossover-terminology concerning neck zones, anatomical triangles, and levels, the latter, used in oncological surgery, is a particularly useful method of dividing the neck into subunits and will be described for the purpose of the chapter. With concurrent appreciation of the regional anatomy for each level (Figure 50.2), the clinician will rarely be surprised by the diagnosis when confirmed (Figure 50.3).

Level Ia pathology

The submental triangle is bounded by the anterior bellies of the digastric muscle and forms a central compartment, extending from the mandibular symphysis to the hyoid bone. Simple skin or subcutaneous lesions such as epidermoid cysts, lipomas, or dermoid cysts are common in this region. A deeper neck lump is likely to be related to oral pathology.

Lymphadenopathy may be reactive, secondary to dental or floor of mouth pathology, or malignant, secondary to oral cavity cancer. Primary malignancies (i.e. lymphoma) should also always be considered. Careful assessment of the anterior floor of mouth, mobile tongue, buccal mucosa, and dentition is mandatory. Ludwig's angina

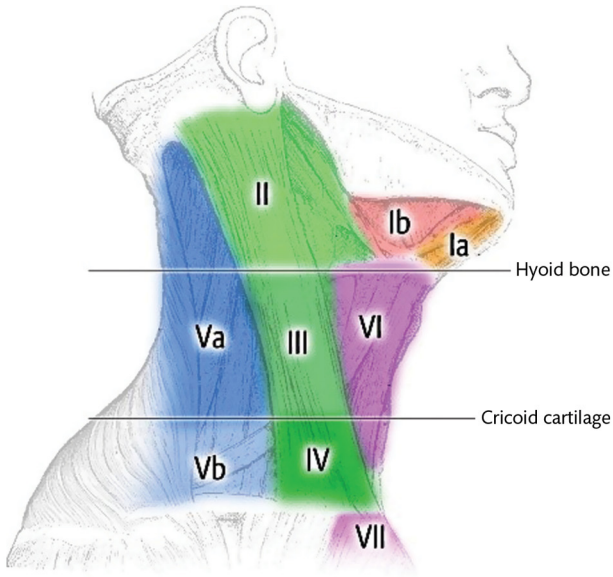

Figure 50.2 An illustration to demonstrate lymph node levels of the right neck.

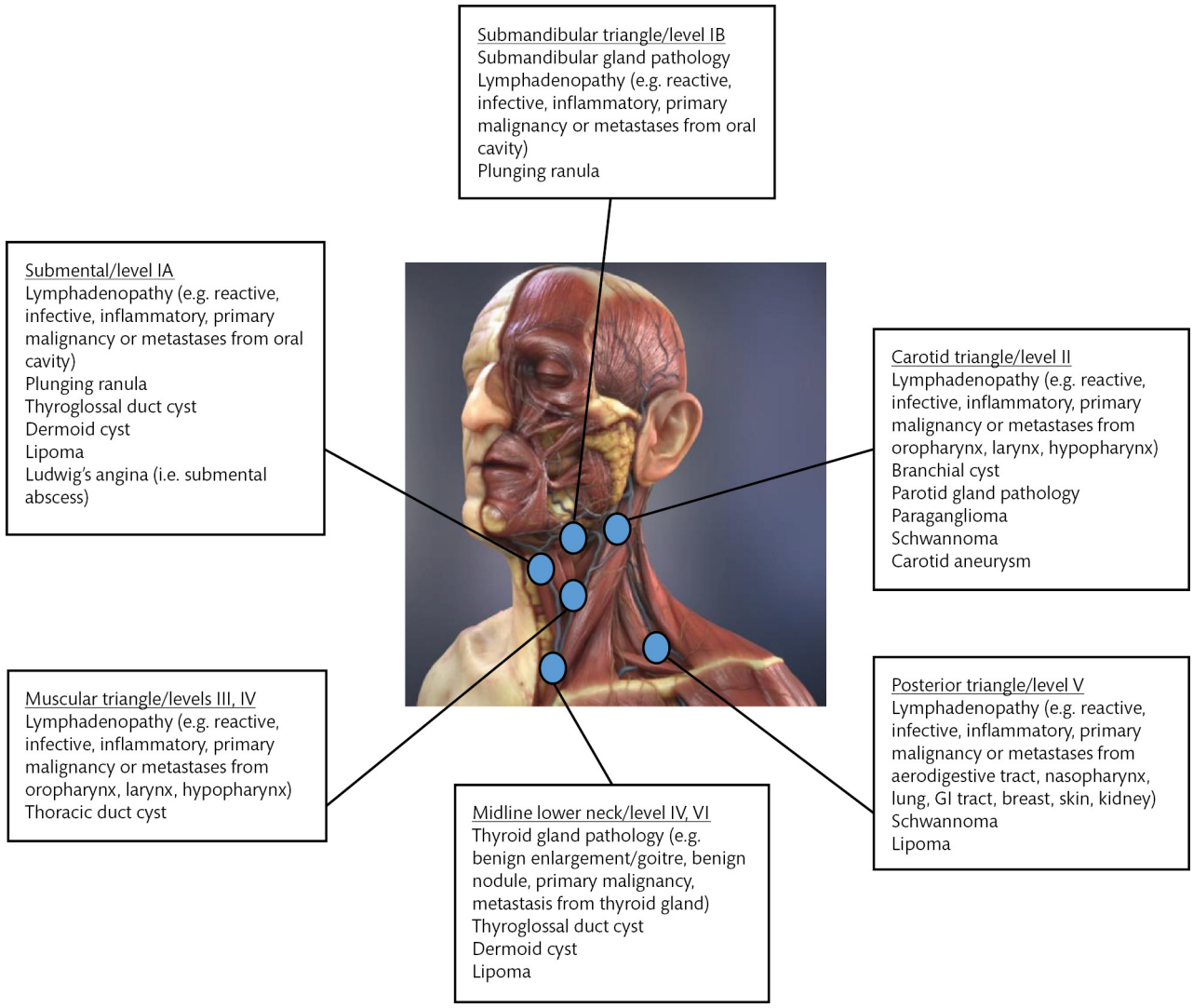

Figure 50.3 An anatomical prosection demonstrating nodal levels and common pathologies encountered in each.

(representing a floor of mouth abscess), a plunging ranula, and sublingual gland disease arise in this subsite.

Radiology pearls

If the lesion arises from dermis/subdermis, imaging is unlikely to be necessary. Deeper lesions may require ultrasound for characterization, with biopsy if required. Masses directly related to intraoral structures commonly require cross-sectional imaging for elucidation. Oral examination confirming bone involvement may necessitate an orthopantomogram and/or CT scan for treatment planning. Confirmation of malignant disease in the oral cavity requires staging imaging, including MRI/CT of the primary lesion and CT of the thorax, respectively.

Level Ib pathology

Level Ib corresponds to the submandibular triangle, bounded by the edge of the submandibular gland posteriorly, the mylohyoid muscle superiorly, and the hyoid bone inferiorly. Neck lumps in this area usually represent submandibular salivary gland pathology (see Chapter 58), lymphadenopathy, or oral cavity disease.

Palpation should aim to differentiate between normal anatomy and pathology. The submandibular gland commonly becomes ptotic with age and can frequently prompt a referral to secondary care. Rarely, immunoglobulin G4 disease (chronic sclerosing sialadenitis), a fibroinflammatory process, can involve the submandibular gland (i.e. Kùttner's tumour).

As with level Ia, this anatomical triangle comprises a number of lymph nodes, for which the differential diagnosis remains broad. In the presence of a level Ib mass, the floor of mouth should be bimanually palpated to assess for the presence of salivary calculi within Wharton's duct, predisposing to obstructive sialadenitis.

Radiology pearls

Ultrasound provides the highest utility for assessment of the submandibular space (Figure 50.4). Primary salivary pathology can be delineated extremely efficiently, while lymph node architecture can be simultaneously assessed. Similarly, strictures or stones within Wharton's duct can be commonly diagnosed. Ultrasound-guided core biopsy is a sensitive modality for the diagnosis of submandibular gland lesions, which have an inherently increased risk of malignancy compared to lesions within the parotid gland. MRI should be requested concurrently if there is suspicion of primary malignant submandibular gland disease, though delineation between benign and cancerous lesions may not be possible clinically without confirmatory core biopsy.

Level II pathology

Level II represents an area responsible for an extremely varied list of differential diagnoses. Accordingly, patients with masses in this region frequently present during Objective Structured Clinical Examinations.

The anatomical borders extend from the skull base superiorly to the level of the hyoid bone inferiorly, with the posterior edge of the sternocleidomastoid muscle forming the posterior border and the posterior border of the submandibular gland representing the anterior border. Deeply, this anterior limit corresponds to the anterior border of the stylohyoid muscle. The spinal accessory nerve divides the region into IIb above and IIa below. Though irrelevant in the clinic setting, this is an important landmark for dissection intraoperatively.

Examination of a neck lump in this area should include detailed examination of the oropharynx for primary malignancy (see below).

Figure 50.4 A photograph demonstrating correct patient–clinician positioning during ultrasound assessment of the anterior neck in the clinic setting.

Lesions of the parapharyngeal space may present with a level II mass, but with medialization of the lateral pharyngeal wall should there be deep neck space extension. The integrity of the accessory, hypoglossal, and facial nerves should be assessed, all of which traverse this region. Paresis may represent a benign (e.g. schwannoma) or malignant nerve sheath tumour, or compression secondary to a carotid body tumour (e.g. paraganglioma). The latter may exhibit mobility on displacement horizontally, but not in a craniocaudal plane (i.e. Fontaine's sign). It is useful to feel for pulsation while palpating the mass, as a prominent carotid bifurcation is a common reason for neck lump referral, particularly in the older patient with a slender neck.

Defining the surface marking of the parotid gland is a common exam question and its overall size is frequently underestimated. The superior border of the gland can be imagined as occupying two-thirds of an imaginary line, traced from the ipsilateral tragus to the modiolus of the lips. From here, the line turns 90° to travel inferiorly to bisect the mandible. Importantly, the gland extends beyond the inferior border of the mandible, within a range between 1 and 5 cm. From the submandibular region, the line then courses to the tip of the mastoid bone posterior to the mandibular ramus and, finally, curves underneath the lobule of the ear lobe, to rejoin the tragus. The implication from this description is that parotid gland lesions can (and frequently do) arise from the 'tail' of the superficial component of the gland, *within the neck*. Masses within the parotid gland may be benign cysts (e.g. HIV), inflammatory (e.g. Sjogren's disease), lymph node metastases (e.g. cutaneous squamous cell carcinoma), or benign or malignant lesions.

Key learning point—assessment of the cystic level II neck lump

Differentiating a branchial cleft cyst from a squamous cell carcinoma cystic metastasis can be challenging, particularly with the rise in incidence of p16-positive, human papillomavirus-implicated oropharyngeal cancers, for which cystic nodal metastases are phenotypical.[4] The previous rhetoric of non-smoking patients under 40 years old being unlikely to harbour malignancy in a cystic mass is increasingly antiquated, with reports of healthy, non-smoking patients of ever diminishing age presenting asymptomatically with a neck lump, subsequently proven to represent a nodal metastasis. In the event of ultrasound-guided fine-needle aspiration confirmation, the absence of primary disease following comprehensive examination of the aerodigestive tract (including nasolaryngoscopy) confers the diagnosis of 'carcinoma of unknown primary'. Dedicated cross-sectional imaging with MRI and subsequently PET/CT is recommended to allow for primary tumour localization, which, if p16 positive, has a predilection for the oropharynx in over 80% of cases.[5] If the primary tumour is identified, focused biopsies can be performed at the time of pharyngo-oesophagoscopy. However, should both MRI and PET/CT be negative, UK head and neck guidelines recommend proceeding to bilateral tonsillectomy and tongue base mucosectomy, the latter of which should be performed in a specialist centre.[5] Mucosectomy has superseded blind biopsies of the tongue base, with current data demonstrating an increase in primary cancer identification from 8% to 90% of cases, in selected case series.[5] Mucosectomy may be performed robotically (i.e. TORS), with carbon dioxide laser (i.e. TOLS) or endoscopically with monopolar electrocautery,[6] with comparable sensitivity rates (see Chapter 56).

A conundrum may arise when ultrasound-guided biopsies are unable to demonstrate malignant cells. In this setting, excision of the cyst for diagnosis may reveal a metastasis, thus requiring radiotherapy (with the addition of concurrent chemotherapy if extracapsular spread is present). When the diagnosis is at equipoise, many multidisciplinary teams would recommend a selective neck dissection (see Chapter 56). UK guidelines recommend that if pathological analysis of the dissection specimen were to confirm the presence of a single metastasis, without extracapsular spread, that the neck may be considered treated, without need for adjuvant radiotherapy.[7] Attention then reverts to the upper aerodigestive tract, and tonsillectomy/mucosectomy may be considered. Numerous case series have demonstrated complete excision of microscopic tumours, leading to surgical 'cure' of select patients in this cohort. Obviating the long-term sequalae of radiotherapy in a younger patient demographic is highly advantageous, particularly in the context of xerostomia, dysphagia, dysphonia, and the development of second-primary cancers. Long-term survivorship data in the carcinoma of unknown primary p16-positive cohort is currently lacking and the role of mucosectomy for this patient group continues to evolve.

In summary, metastatic p16-positive oropharyngeal cancer should *always* be considered in the presence of an isolated, cystic, lateral neck mass. This scenario is an exception to the theme of the chapter, benefitting hugely from developments in cross-sectional imaging, high-resolution endoscopy, narrow band imaging, and transoral surgical techniques.

Radiology pearls

Ultrasound is highly effective as a first-line investigation for level II masses and remains the most sensitive modality for demonstrating features indicative of pathological (i.e. enlarged, rounded, loss of fatty hilum, intranodal necrosis, calcification), rather than physiological/reactive lymph nodes. Ultrasound-guided biopsy is usually requested concurrently, unless there is suspicion of a bleeding diathesis or vascular lesion, respectively. Exquisite pain during biopsy may alert the clinician to the diagnosis of a nerve sheath tumour, such as a schwannoma or post-traumatic neuroma. MRI is indicated to provided complementary cross-sectional images of the upper aerodigestive tract, should malignancy be suspected. In the presence of biopsy-proven carcinoma of unknown primary (as above), MRI is indicated in the first instance, proceeding to PET/CT should the primary disease fail to be identified. Contrast CT has utility in the characterization of carotid body tumours, with splaying of the internal and external carotid arteries being highly indicative (i.e. Lyre's sign).

Level III pathology

Level III is bounded by the inferior border of the hyoid bone to the inferior border of the cricoid cartilage, and comprises lymph nodes that drain the oral cavity, oropharynx, hypopharynx, and laryngopharynx. Thyroglossal duct cysts, while typically a midline mass, may be lateralized and lie within these boundaries. Laryngoceles, which represent herniation of laryngeal mucosa through an acquired defect in the thyrohyoid membrane, may also be palpable at this subsite. When attempting to differentiate between such pathologies, assessing the consistency of the neck lump is extremely useful. A cyst or laryngocele is likely to be compressible or reduceable, respectively, compared to the solid architecture of a pathological node. Nasolaryngoscopy is indicated should a

laryngocele be suspected, for it may represent a secondary manifestation of an obstructing primary tumour of the larynx.

Radiology pearls

Ultrasound is a faithful first-line investigation to delineate between a lateralized thyroglossal duct cyst, pathological lymph node, or laryngocele. In cases where the latter is suspected, a CT scan has superior spatial resolution compared to MRI and is usually confirmatory. Similarly, a laryngocele may spontaneously reduce with the patient's natural breathing cycle, so a prolonged MRI reduces the modality's sensitivity compared to an expedient CT scan. In the presence of a pathological lymph node and should ultrasound-guided biopsy confirm lymphoma, a PET/CT scan should be arranged for formal whole-body staging, according to the Ann Arbor classification.

Level IV pathology

Level IV is demarcated from the inferior border of the cricoid cartilage superiorly to the clavicle inferiorly and includes the medial supraclavicular lymph nodes. During neck dissection, the omohyoid is used as a surgical landmark separating levels III and IV. Anterior to both the phrenic nerve and anterior scalene muscle, the thoracic duct courses through level IV in the left neck to drain into the junction of the left subclavian and internal jugular veins.

Examination of a level IV neck lump inexorably involves assessment of the thyroid gland (see level VI below), representing a common location of malignant nodal metastases of thyroid origin. Similarly, metastases from numerous primary subsites may present with a pathological node in this region, including carcinoma of the larynx, hypopharynx, oesophagus, lung, breast, and kidney. As such, these systems should be addressed in turn through focused history and examination.

Radiology pearls

Ultrasound remains useful in establishing the characteristics of pathological lymph nodes, which is particularly important in the context of possible metastases from numerous primary sites. Rarely, a thoracic duct cyst or lymphocele may be encountered, for which lymphoscintigraphy can be helpful, usually in conjunction with MRI to allow for preoperative planning.

Level V pathology

Level V is delineated by the posterior border of the sternocleidomastoid, the anterior border of the trapezius muscle, and the medial third of the clavicle, inferiorly. The level is further divided into 'Va' and 'Vb', relating to above or below the traversing accessory nerve. Up to 75% of neck lumps in this region will represent malignancy.[8]

On discovery of a level V neck lump, traditional teaching mandates examination of the nasopharynx to exclude a primary malignancy, though the risk of metastasis from this area, while significant, is actually in the order of 27% compared to higher rates of metastases in level II (70%) and level III (45%).[9] Lymphoma is the most common malignant pathology in this level in certain case series, though metastases from lung, breast, and viscera are important differentials.[8] Granulomatous disease, Castleman's disease, lipomas, and a variety of non-malignant conditions may present as a neck mass in this level. A considered systems review, with request of pertinent blood tests, may help secure the diagnosis.

Radiology tips

In all but the most benign of clinical presentations, masses within level V should be investigated aggressively and assumed to represent malignancy until proven otherwise. A supraclavicular mass in a patient with 'red flag' symptoms and concurrent historical risk factors should prompt the clinician to avoid delay in requesting imaging sequentially. Ultrasound-guided biopsy is an expedient method to obtain the pathological diagnosis. However, *do not wait for the results before requesting further investigations in the presence of ominous sign/symptoms*. The presence of a chronic cough mandates urgent cross-sectional imaging of the thorax. Concurrent symptoms involving the laryngopharynx, nasopharynx, or hypopharynx require MRI of the aerodigestive tract, with CT imaging of the thorax if disease is confirmed on examination. Widespread axillary/inguinal lymphadenopathy with a deranged full blood count would suggest lymphoma, necessitating PET/CT. Similarly, the presence of a keratotic, ulcerative skin lesion, ipsilateral to the mass, would suggest metastatic squamous cell carcinoma and staging imaging is required. Aim to avoid delay when requesting multiple investigations. Endeavour to arrange appropriate imaging in parallel, rather than in series, wherever possible. This will result in shortened patient pathways and reduce the risk of disease upstaging in the context of malignancy.

Level VI pathology

Level VI, or the 'central compartment' of the neck, is a common subsite for benign disease and, as such, is frequently encountered in Objective Structured Clinical Examinations. The examining clinician will have the opportunity to demonstrate their understanding and prowess in a variety of medical specialties, including embryology, anatomy, and clinical examination technique.

The central compartment of the neck extends from the hyoid bone superiorly to the suprasternal notch inferiorly, bounded laterally between the common carotid arteries. The thyroid gland itself resides in this level, inferior to the cricoid cartilage, deep to the overlying strap muscles. Neck lumps in this area are relatively unique, in that there is the possibility for diagnosis and differentiation based solely on clinical examination. Three which may be distinguishable are benign thyroid nodules, thyroglossal duct cysts, and dermoid cysts.

With the clinician placing their hands on the lump and paratracheal area from behind the patient, asking them to swallow and protrude their tongue in turn, three combinations of outcomes may occur as shown in Table 50.3.

Thyroglossal duct cysts represent a remnant of the embryological thyroglossal duct and may develop anywhere between its origin at the foramen caecum of the tongue, to the pyramidal lobe of the thyroid gland. As the duct is in continuity with the tongue, protrusion

Table 50.3 Summary of features exhibited by common level 6 neck masses during clinical examination

	Thyroid nodule	Thyroglossal duct cyst	Dermoid cyst
Moves on swallowing	Yes	Yes	No
Moves on tongue protrusion	No	Yes	No

will typically result in movement of the lump in a cranial direction. Similarly, as the duct is adherent to the strap muscles, swallowing will also be associated with lump movement. Contrariwise, thyroid nodules reside within the substance of the gland, which is anatomically separated from the thyroglossal duct. The gland itself is adherent to the laryngeal cartilaginous framework via the ligament of Berry and, accordingly, thyroid nodules will move on swallowing. However, being separate from the thyroglossal duct, no movement should be observed during tongue protrusion. Finally, dermoid cysts archetypally reside in the subdermal plane, superficial to the strap muscles and are not related to the duct. As such, they should not move in response to these manoeuvres, though exceptions can occur.

A cautionary note: careful palpation of a suprasternal notch lesion for pulsation should be made to identify any uncommon but critical vascular anomalies, such as an innominate artery aneurysm,[10] though extremely rare.

Radiology tips

Ultrasound with or without biopsy is the gold standard for delineation of thyroid nodules and their subsequent classification via 'U grading', discussed in detail in Chapter 59. Thyroglossal ducts extending from a presumed thyroglossal duct cyst are readily visualized, while simultaneously the presence of an ectopic/absent cervical thyroid gland can be confirmed or refuted. In this scenario, cross-sectional imaging should be employed to exclude a lingual thyroid gland. CT/MRI may also be used in conjunction with ultrasound for further characterization of the level VI mass, particularly if there is evidence of thoracic extension, concurrent dysphagia, lymphadenopathy, or vocal cord palsy to suggest retrosternal goitre, extrinsic oesophageal compression/invasion, cervical metastases, or recurrent laryngeal nerve palsy due to thyroid malignancy, respectively. Oesophagoscopy and/or bronchoscopy should be performed prior to any definitive procedure, should there be even the remotest suspicion of malignant invasion of the oesophagus/trachea.

Examination of the upper aerodigestive tract

Assessment of the neck lumps is incomplete without assessment of the upper aerodigestive tract mucosa including flexible nasolaryngoscopic examination (**Figure 50.5**). Detailed descriptions on examination of the oral cavity, pharynx, and larynx can be found in other chapters.

Blood/pathology tests

At this stage of the consultation, the clinician should already have a strong suspicion as to the diagnosis. The investigations requested are aimed to confirm that which the surgeon is already suspecting and should be focused, rather than comprehensive (**Table 50.4**).

Blood tests can implicate or, in certain situations, confirm the diagnosis of a patient presenting with a neck lump. A full blood count is a useful screen to establish inflammatory or infectious pathology. Significant derangement in the patient's white cell count in the presence of constitutional symptoms may be indicative of a haematological malignancy. In such scenarios, a blood film may demonstrate blast cells to suggest acute leukaemia, for example. Similarly, an elevated lactate dehydrogenase level may be seen in patients with lymphoma. Concurrent C-reactive protein levels may be used to add credence to the diagnosis of an inflammatory/infective aetiology, such as that found in benign inflammatory salivary gland disease, which may be infective, obstructive, or autoimmune. Autoantibody

Figure 50.5 Demonstration of a clinician performing flexible nasoendoscopy on a patient in the clinic setting.

Table 50.4 Table summarizing the differential diagnosis for a patient presenting with a cervical neck lump, with key investigations relevant to securing the diagnoses

Benign	Key investigations	Malignant	Key investigations
Infectious		**Primary**	
Viral		*Haematological*	
CMV	Serology	Leukaemia	FBC, blood film, LDH, USS core, PET
HIV	Serology	Lymphoma	FBC, blood film, LDH, USS core, PET
EBV	FBC, CRP, LFT, Monospot	*Salivary gland*	
Paramyxovirus	Serology	Mucoepidermoid	USS core, MRI, CT
HSV	PCR	Adenoid cystic	USS core, MRI, CT
Bacterial		Acinic	USS core, MRI, CT
Streptococcal (etc.)	MC&S, CRP, LFT	*Thyroid*	
Bartonella	USS, serology	Papillary	TFT, USS core, MRI, CT
Toxoplasmosis	USS, serology	Follicular	TFT, USS core, MRI, CT
Tuberculosis	USS core, MC&S, CT	Medullary	USS FNA, calcitonin, TFT, MRI, CT
Atypical mycobacteria	USS core, MC&S	Anaplastic	TFT, USS core, MRI, CT
Actinomycosis	USS core, MC&S	*Musculoskeletal*	
Neurovascular		Sarcoma	USS core, MRI, CT
Paraganglioma	USS, CT/MRI	**Secondary**	
Schwannoma	USS, CT/MRI	*Skin*	
Carotid aneurysm	USS, MRA/CTA	Squamous cell carcinoma	USS core, MRI, PET
Haemangioma	USS, MRI	Basal cell carcinoma	USS core, MRI, CT
Inflammatory		Melanoma	USS core, MRI, PET
Reactive	Reassurance (FBC), (serology), (USS)	*Salivary gland*	
Thyroid		Mucoepidermoid carcinoma	USS core, MRI, CT
Nodule	TFT, USS FNA/core	Adenoid cystic carcinoma	USS core, MRI, CT
Multinodular	TFT, thyroid antibodies, USS	Acinic cell carcinoma	USS core, MRI, CT
Physiological	TFT, thyroid antibodies, USS	*Thyroid*	
Inflammatory	TFT, thyroid antibodies, FBC, CRP USS	Papillary	TFT, USS core, MRI, CT
Congenital		Follicular	TFT, USS core, MRI, CT
Branchial cyst	USS FNA, MRI	Medullary	USS FNA, calcitonin, TFT, MRI, CT
Thyroglossal duct cyst	USS, MRI	Anaplastic	TFT, USS core, MRI, CT
Dermoid cyst	USS	*UADT*	
Salivary gland		Squamous cell carcinoma	USS core, MRI, CT
Pleomorphic adenoma	USS FNA/core, MRI	*Alternate*	
Warthin tumour	USS FNA/core, MRI	Breast	USS core, CT
Sialadenitis	FBC, CRP, autoantibodies, USS, MRI	Lund	USS core, CT
Obstructive	USS, sialography	Kidney	USS core, CT
Sialadenosis	FBC, fasting glucose, LFT, vitamin B_{12}/folate, USS	Gastrointestinal	USS core, CT
Cyst	USS (HIV serology)		

CMV, cytomegalovirus; CRP, C-reactive protein; CT, computed tomography; CTA, computed tomography angiography; EBV, Epstein–Barr virus; FBC, full blood count; FNA, fine needle aspiration; HIV, human immunodeficiency virus; HSV, herpes simplex virus; LDH, lactate dehydrogenase; LFT, liver function tests; MC&S, microscopy culture and sensitivity; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging; PCR, polymerase chain reaction; PET, positron emission tomography; TFT, thyroid function tests; USS, ultrasound scan.

screening may help confirm the diagnosis in the latter. Sialadenosis is common in patients with diabetes mellitus, or those with a history of either a formal eating disorder, or excessive alcohol intake; accordingly, fasting glucose, vitamin B_{12}/folate deficiencies, and liver function tests can be supportive of the clinician's interpretations. Positive serological tests of distinct pathologies in the presence of associated historical risk factors/examination findings (e.g. bartonella/cat scratches; paramyxovirus/bilateral parotid swelling) are usually sufficient to conclude the diagnostic pathway. Bilateral parotid cysts in the presence of associated risk factors should prompt the clinician to request HIV serology.

Ironically, thyroid function tests, routinely comprising free T4 and thyroid-stimulating hormone, may commonly be within normal range in the presence of a thyroid mass. Indeed, the majority of isolated thyroid nodules are non-secretory. However, the presence of thyroid peroxidase antibodies may be seen in patients with hypothyroidism and an enlarged thyroid goitre, implicating Hashimoto's disease. Patients with this diagnosis are at increased risk of developing lymphoma.[11]

Numerous viral infections may result in the development of cervical adenopathy. Bacterial infections will usually present with acute, concurrent throat symptoms, in the context of pharyngitis, tonsillitis, or peritonsillar abscess. The diagnosis may be confirmed on swabbing the throat. Herpetic ulceration in the presence of painful cervical adenopathy may be diagnosed in a similar fashion. Inpatient admission may be required in the presence of sepsis, for both intravenous antibiotics and fluid resuscitation. Atypical bacterial infections, contrariwise, have a more indolent presentation, often without pyrexia. The diagnosis is usually via a sample of tissue (e.g. a pathological lymph node), demonstrating granulomatous inflammation and culture confirming the causative organism. Cross-sectional imaging of the thorax is similarly required once established. A surgical 'sieve' is useful to help stratify the diagnosis, along with pertinent investigations that may be required to secure the diagnosis unequivocally. For patients presenting with clinical features of a malignant lesion, the mainstay of investigative workup will comprise an initial ultrasound scan/core biopsy, MRI of the neck and aerodigestive tract, and CT staging of the thorax (and abdomen), as stipulated by UK national multidisciplinary guidelines.[12]

The vast majority of diagnoses are confirmable on either fine needle aspiration cytology or core biopsy. The former is recommended for all thyroid nodules graded U3 or above, in accordance with British Thyroid Association guidelines.[13]

The efficacy of fine needle aspiration cytology is improved when performed under ultrasound guidance with significant benefit to both sensitivity and specificity.[14] Blind biopsies for thyroid lumps are not recommended, due to the higher rates of inadequate samples, lower sensitivity,[13] and the need for radiological concordance.

Diagnostic difficulty can arise in cytology obtained from branchial cysts[15] (as above) and Warthin tumours[16] (papillary cystadenoma lymphomatosum). Ultrasound-guided core biopsy has gained favour over fine needle aspiration cytology for salivary gland tumours, with significant improvements in sensitivity and specificity over the latter, without appreciable complications (most notable, iatrogenic injury to the facial nerve). Systematic review and meta-analysis has been shown to detect malignancy in 96% of cases.[17] The larger sample size presented to pathology affords improved microscopic examination but also immunohistochemistry evaluation. Notably, in the context of metastatic squamous cell carcinoma, this can allow for detection of p16 status, a surrogate marker for human papillomavirus-related disease, which has implications with regard to prognosis and indications for treatment.

Core biopsy can also prove useful in the diagnosis of lymphoma, obviating the requirement for incisional/excisional biopsy of a pathological lymph node. Subtype diagnosis of lymphoma often requires a range of immunohistochemistry techniques which can frequently, but not always, be ascertained from a core biopsy specimen. If the diagnosis is still in doubt, an open biopsy may be required. A larger sample of continuous tissue enables the cellular pathologist greater freedom in performing specific techniques, such as fluorescence *in situ* hybridization and polymerase chain reaction.

Conclusion

This chapter demonstrates a practical approach to neck lump assessment. A patient presenting with a neck lump may do so in isolation of any other concerns. Alternatively, the presence of a concurrent myriad of symptoms, spanning multiple organ systems, may be reflective of serious underlying pathology. This emphasizes the importance of a comprehensive and multidisciplinary approach to the patient's assessment, implementing the full armamentarium of the treating otolaryngologists' knowledge base and the recruitment of a complement of allied professionals, including anaesthetists, intensivists, and subspecialist consultants. Patients will be inherently apprehensive about their ensuing diagnosis and subsequent treatment, such that the treating physician should make every effort to provide comprehensive psychosocial support, while also ensuring expediency and good communication throughout the assessment and investigative period. Patient trust is attainable when a relationship has a history of reliability, advocacy, beneficence, and good will. The authors entrust the reader with the opportunities to demonstrate such qualities through the processes described.

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51

Snoring and sleep apnoea

Khalid Ghufoor and Jahangir Ahmed

Introduction

Snoring and sleep apnoea are presenting symptoms for sleep-related breathing disorders (SRBDs), which are a spectrum ranging from occasional snoring to severe obstructive sleep apnoea (OSA). They often occur in overweight or obese populations, but a significant minority of patients have a low body mass index (BMI).

In addition to the inherent work and societal problems consequent to daytime fatigue and mood change from sleep deprivation, there is an increasing recognition of causal links to adverse chronic medical conditions. Untreated sleep apnoea has been correlated with hypertension, type 2 diabetes, metabolic syndrome, reduced sexual function, increased stroke risk, cardiomyopathy, arrhythmias, and myocardial infarct. A holistic approach to treatment therefore necessitates a diverse set of healthcare professionals, creating a unique multidisciplinary sub-specialism for SRBDs. The SRBD clinical team may therefore include physicians from the following specialties: respiratory medicine, otolaryngology, oral and maxillofacial surgery, bariatric surgery, cardiology, neurology, psychiatry, anaesthetics, and intensive care medicine. This broad group have separately and collectively helped to provide a multifaceted approach to managing these conditions.

The majority of airway opening surgery is conducted by ear, nose, and throat (ENT) surgeons, ideally with a specialist interest and will be discussed later in this chapter; although for select cases orthognathic surgery may also be appropriate.

Clinical assessment and suitability for surgical intervention

History

This will typically include snoring, sleep deprivation, and daytime fatigue. Repeated choking or gasping episodes, worse while lying on the back, are usually noted by the patient's partner. Other nocturnal symptoms include palpitations, sweating, and nocturia. Pre-existing, or symptoms of potentially undiscovered correlated, comorbidities, particularly cardiovascular symptoms, should be noted.

A general nasal and airway history is relevant. Preoperative identification of swallow, voice, and palatal dysfunction should lead to a more detailed speech and swallow assessment before consideration of any surgical intervention. A formalized validated questionnaire such as the Epworth Sleepiness Scale may indicate the likelihood of OSA, with scores over 10 usually being clinically significant.

Examination

The upper airway consists of both fixed structural (such as a deviated nasal septum/inferior turbinate hypertrophy, enlarged adenoid and tonsils) and dynamic components. Failure to appreciate and treat the dynamic elements that contribute to obstruction usually leads to a disappointing surgical outcome.

The general ENT examination should take note of any sources of nasal obstruction, retrognathia, and the degree of mouth opening. The Friedman mouth opening score[1] (measured with mouth open with the tongue kept inside the oral cavity) is a useful predictor of the likely contribution of obstruction from the tongue base. A high Friedman score, such as an inability to visualize the soft palate upon mouth opening, predicts difficult surgical access as well as a large, muscular tongue base contribution. This is a difficult situation that will influence surgical decision-making and may preclude surgery at multiple oropharyngeal subsites simultaneously (see below). The Friedman tonsil grade[1] should be documented, and the palate and uvula should be inspected for length and redundancy. An endoscopic view of the tongue base, lymphoid tissue in the vallecula (lingual tonsil), and the supraglottic appearance should ideally be photo documented. A dynamic view of the pharynx while performing Müller's manoeuvre (inhaling against a closed nose and mouth) has traditionally been used to try to predict dynamic surgical target(s); however, this does not represent the physiological state during sleep.

Assessment for suitability of transoral surgery

For optimal surgical success, patients should have a BMI of less than 35 kg/m^2 and ideally closer to 30 kg/m^2.[2] The interincisal distance should be at least 2.5 cm to accommodate the arms of the Da Vinci or other robotic system. It is important to establish normal laryngeal, pharyngeal, and palatal function. If in doubt, a formal clinical,

instrumental, and radiological assessment of palate and swallow function should be performed.

Investigations

The sleep study

To ensure a favourable cost/benefit ratio for advanced transoral surgery, a firm diagnosis of OSA should be established. In most cases, a domiciliary sleep study alone usually suffices, but a more comprehensive in-patient sleep study should be requested if there is doubt regarding the nature of the nocturnal sleep disturbance.

An elevation of the Epworth Sleepiness Scale score can also occur due to insomnia, nocturnal epilepsy, parasomnia, movement disorders, and central sleep apnoea. In such cases, an in-patient multichannel polysomnographic study will help distinguish between these diverse conditions. A similar in-patient polysomnograph should also be obtained prior to revision surgical cases.

For OSA, an apnoea episode in adults is defined as a cessation of breathing for more than 10 seconds and hypopnoea refers to a reduction of tidal volume of over 50%. The number of episodes of apnoea or hypopnoea per hour, the apnoea–hypopnoea index (AHI), defines the severity of the condition. Up to five episodes of apnoea per hour is deemed normal, 5–15 mild, 15–30 moderate, and greater than 30, severe.

In OSA, apnoeas and hypopnoeas are accompanied by respiratory effort, in contrast to central apnoeas which occur as a consequence of respiratory muscle hypotonia secondary to a central neurological aetiology.

Imaging

Plain radiography

Imaging is not usually required but can sometimes be helpful in anticipating difficult cases or when contemplating orthognathic surgery. The lateral plain X-ray can provide information on the anteroposterior (AP) airway. An important anatomical landmark is the distance between the hyoid bone and the mandibular plane, the hyomandibular plane. This indicates the vertical height of the tongue which in turn correlates with upper airway collapse, when this figure is greater than 25 mm.

Cross-sectional imaging

Cross-sectional imaging (magnetic resonance imaging/computed tomography) can be useful in delineating the nature and extent of the obstructing tissue in the postnasal space, tongue base, or larynx. It is mandated in patients where the obstructive tissue looks suspicious for neoplasm. A computed tomography scan of the mandible will demonstrate a retrognathic jaw and airway dimensions can be calculated, although these will vary depending on the phase of respiration or swallow. Very importantly, aberrant vascular anatomy can be delineated, and this may influence the decision to perform invasive transoral tongue base surgery.

Dynamic cross-sectional imaging has been used to demonstrate changes in cross-sectional airway at the level of the retrolingual space in both AP and lateral dimensions throughout the sleep cycle, although its utility in the assessment and surgical management of OSA has yet to be defined.

Treatment

The mainstay of OSA management is through lifestyle modification (dietary and alcohol restriction, weight loss regimens) and continuous positive airway pressure (CPAP). The latter is very effective in maintaining oxygenation but needs to be used daily for at least 7 continuous hours. Long-term compliance, however, can be a problem, compounded by the side effect profile of the device. CPAP-associated rhinitis or epistaxis, especially with use of the nasal mask, can sometimes be alleviated with humidification and topical emollients. Those needing high pressures may also develop skin pressure changes including ulceration and pain. Many patients also complain of claustrophobia and swallowed air (aerophagy). The patient's partner may have trouble sleeping due to noise from the device.

Prosthesis and devices

Other conservative means to open the airway include nocturnal use of nasal valve strips, chin straps (to encourage mouth closure), and mandibular advancement splints. The latter are the most effective devices in appropriately selected individuals but they are not for ubiquitous use as the patient must have healthy dentition with no temporomandibular joint dysfunction or other contraindications such as epilepsy. They work on the premise of enhancing the AP dimension at the level of the retrolingual space. Relative mandibular advancement will enhance this dimension due to the insertion of the geniohyoid muscle into the central anterior mandible. Long-term use, however, may lead to bite change and temporomandibular joint dysfunction.

In isolation, devices are not effective in moderate or higher degrees of OSA and many patients are not able to tolerate usage.

Surgery

At present, the National Institute for Health and Care Excellence in the UK suggests that surgery has a limited role to play in patients with OSA. However, a significant number of patients do fail to respond to or tolerate non-operative approaches and are refractory to lifestyle-altering measures. These patients are often referred to otolaryngologists for an assessment of their upper aerodigestive tract with a view to seek out potential airway-opening surgical targets.

Surgical intervention can be divided into indirect and direct airway-altering procedures. The former include bariatric surgery and nasal airway opening operations such as septoplasty, turbinate reduction, and adenoidectomy. The latter include facial skeletal expansion surgery and endoluminal expansion mostly through tissue reduction (see below).

Although a reduction in the BMI is probably the most effective indirect target, particularly for those with a very high starting BMI, a significant minority (up to 30%) of patients are already within or close to their ideal BMI.

Nasal surgery not only facilitates mouth closure, with consequent tongue protrusion, but also reduces the work of breathing from consequent reduction in the negative intraluminal pressure, and can facilitate nasal CPAP. It is, however, uncommon for nasal surgery alone to cure OSA, except occasionally in those with mild severity disease.

Orthognathic surgery in carefully selected patients with relative midface and mandible hypoplasia can be extremely effective

at opening the airway and reversing OSA. Such procedures require careful preoperative workup, dental optimization, and counselling. The surgery can lead to altered cosmesis, which may indeed be of secondary or even primary benefit to the patient. However, airway obstruction often occurs at multiple levels and orthognathic surgery alone may not address particular airway blocks such as lateral pharyngeal wall collapse and epiglottic obstruction.

Expansion of the airway calibre by endoluminal means of tissue reduction, framework expansion, and stiffening collapsible tissue is currently the state of the art in effective airway surgery. The remainder of the chapter will focus on the most effective of these techniques in the authors' opinion.

Presurgical considerations

Historical surgical protocols relied predominantly on two operations: tonsillectomy and uvulopalatopharyngoplasty. The latter entailed removing a 1 cm strip of the convergent palatal arches as well as total uvulectomy, but was associated with a 3% rate of palatal dysfunction including nasal speech and nasal regurgitation. This combination of procedures carried a long-term 'success' of only 50%.

The key to improving successful surgery is to ensure that the precise anatomical problem(s) causing airway obstruction are addressed. This can be achieved by performing a sleep nasal endoscopy. In essence, the patient is sedated using standard intravenous anaesthesia titrated to achieve spontaneously ventilated 'sleep' and a nasal endoscope is introduced to video document the pattern(s) of obstruction/noise generation.

A number of drug-induced sleep endoscopy (DISE) classification systems have subsequently been described. The European position paper in 2014[3] describes the anaesthetic technique and types of airway occlusion patterns commonly seen. At each anatomical subsite (palate, lateral pharyngeal wall, tonsil, and tongue base) the pattern of compression (AP/lateral/concentric) is recorded. At the epiglottic level, the compression pattern does not apply and the nature of the epiglottic anatomy is described instead (retroverted, trap door, etc.).

An important observation is the variable nature of airway occlusion seen on DISE, often at variance to the awake airway assessment. Airway obstruction at multiple levels is usually the rule and warrants methodical observation at each of the levels as described above. Indeed, certain airway issues such as epiglottic trap-dooring (where a floppy epiglottis clamps shut on the laryngeal inlet upon inspiration) or concentric pharyngeal collapse (at the level of the palate or oropharynx) can only be identified during DISE. The information gained from DISE will provide additional information and allow for realistic discussions with patients about the likelihood of success with targeted surgery.

At DISE, the effect of proposed jaw advancement from either a device or orthognathic surgery may also be mimicked by means of a gentle jaw thrust.

A full general anaesthetic after the DISE can be administered to assess suitability for transoral robotic surgery (TORS) or laser microsurgery as well as to perform surgery to fixed structural or obvious targets such as the septum/turbinates or tonsils.

DISE should be a mandatory investigation in patients who have persistent OSA or relapse after initial successful airway surgery; the effect of the previous surgery may unmask other anatomical sites contributing to airway obstruction.

There are, however, problems with universal acceptance of DISE, in part due to the procedure relying on an artificial drug (usually propofol and midazolam) inducing sleep.

The method of induction has to be carefully titrated to prevent apnoea which may be difficult in some patients. The use of bispectral index (BIS) monitoring may help maintain consistency and depth of anaesthesia and should be encouraged—a BIS score of 50–60 is usually an adequate depth of sedation in the authors' experience.

The interpretation of DISE is observer dependent, with a potential bias to over-diagnose surgical targets that are within the surgeon's armamentarium and vice versa. In the earliest classifications of sleep nasal endoscopy, the focus was to identify surgically correctable areas and as transoral laser tongue base techniques were not widely available or restricted to treating neoplastic lesions, tongue base obstruction was given the highest grade and was largely deemed incurable and ignored by the ENT surgical fraternity.

Currently many transoral techniques are available for dealing with the pharynx and tongue base and so the description and assessment of this area during DISE is imperative to guide surgery. Video documentation can allow for re-evaluation after the procedure to ensure consistency of observations as well as counselling and planning the surgery.

Tongue base surgery

Addressing the tongue base appears to be a key factor in surgical success. Positive predictors of successful surgery include a high lymphoid tissue-to-tongue base muscle ratio, a short height of tongue base (indicated by a low hyomandibular plane), and predominant AP collapse at the level of the oropharynx.[4]

A relative exclusion criteria for tongue base surgery is a very high BMI (>35 kg/m^2). The tongue has a higher propensity for fat deposition compared to other muscles. Obese patients have a much higher volume of fat deposition in the pre-epiglottic space, tongue base musculature, and in the sublingual region which acts to counteract any remediable tongue base surgery (**Box 51.1**).

Traditionally, tongue base surgery could only be performed using transcervical techniques, for example, via a lateral pharyngotomy to

Box 51.1 Different surgical techniques to address the tongue base

Tongue base reduction

- Open transcervical (covering tracheostomy).
- Midline partial glossectomy: laser/monopolar/coblation.
- Radiofrequency-mediated ablation.
- Transoral robotic monopolar or laser microsurgery.

Framework expansion

- Mandibulotomy with genioglossal advancement.
- Hyoid advancement:
  - Hitching the hyoid to the mandible.
  - Thyrohyoidpexy.
- Maxillomandibular advancement.
- Hypoglossal nerve stimulation synchronized to inspiration.
- Tracheostomy.

provide direct access. This surgery was invasive, required a tracheostomy to protect the airway, caused a prolonged recovery, and risked long-term pharyngeal dysphagia.

Median glossectomy, with a reduction of the central tongue musculature, has a limited role in macroglossia where the tongue muscle-to-lymphoid tissue ratio is high. This can be performed transorally by monopolar cautery or laser techniques to reduce a central rhomboid of tongue with subsequent suture closure. Safe tissue reduction was restricted to the midline, for fear of injuring large-calibre, laterally lying neurovascular bundles. However, this often failed to address the true anatomical problem at the tongue base.

Safer, less painful modifications subsequently evolved, utilizing radiofrequency and coblation techniques in an attempt to reduce/shrink the tissue at the tongue base. The probes could be inserted across the tongue base, rather than being restricted to the midline. The effect on tissue was not always immediately apparent, and was sometimes met with post-procedural swelling, but a gradual reduction of tissue to variable degrees was achieved. Further repeat procedures could be safely undertaken if initially inadequate.

However, none of these techniques have demonstrated significant long-term success in reversing OSA and, more importantly to the patient, negated the need for CPAP machine use.

In the authors' practice, any patients who have tongue base obstruction, either solely or in part, are offered TORS resection. This technology affords unparalleled three-dimensional visualization and access to provide safe haemostatic tissue resection with minimal collateral injury to surrounding tissue.

Transoral robotic surgery

In the authors' clinical practice, patients are not considered for TORS unless there has been a failure of social, lifestyle, and non-invasive therapeutic interventions.

Patients must have sleep study-proven OSA with symptoms—ideally at least moderate or severe sleep apnoea. Patients with a grossly elevated BMI greater than 35 kg/m^2 will be referred for dietetic advice and bariatric surgery if suitable to help achieve a BMI closer to 30 kg/m^2. This reduces the adiposity of the tongue and pharynx, optimizing the chances of success.

Managing expectations is a key factor in ensuring long-term success. Patients are usually counselled on the potential decline in long-term success of surgical interventions. This may be due to multiple factors including softening of the pharyngeal scar tissue, regrowth of lingual tonsil, and regain of the weight that patients lose in the immediate postoperative period.

Relative contraindications to robotic surgery relate to accessibility and anatomical contraindications. Mouth opening must be sufficient (ideally >2.5 cm) to allow insertion of the endoscope and instruments; a relatively large tongue (correlating to a high Friedman mouth opening score (3 or 4)) can also be a hindrance to instrumentation and obtaining an adequate view. If there is doubt, an examination under anaesthetic while performing DISE can help clarify the situation.

The oropharynx in patients with OSA is usually crowded. This may pose a challenge for safe surgical intervention without excessive collateral injury. If more than one subsite of the oropharynx requires intervention, performing surgeries sequentially can help minimize circumferential pharyngeal stenosis, which is the main concern as it is extremely difficult to remedy.

A final consideration is pharyngeal function, both velopharyngeal closure as well as swallow. Any pre-existing impairment warrants full evaluation which includes instrumental assessments such as videofluoroscopy swallow, flexible endoscopic evaluation of swallow, and/or functional magnetic resonance imaging scanning. A speech and swallow therapist should be involved in the care of such patients and a multidisciplinary team decision made on suitability for surgery and potential rehabilitation thereafter.

Surgery to other levels of obstruction

As mentioned, patients with OSA often have other anatomical contributors/patterns of airway obstruction.

In the authors' experience, the commonest include enlarged palatine tonsils, prominent posterior (palatopharyngeal) palatine pillars, and AP or concentric palatal closure.

The palatal contribution is usually not as important as the tongue base and should be addressed if necessary at a later date once the tongue base, with or without tonsillectomy, sites have healed, in order to prevent circumferential oropharyngeal scarring (pharyngeal stenosis). The latter is extremely difficult to surgically manage and can pose a significant airway/swallow hazard.

The pattern of palatal obstruction can be dichotomized into AP or lateral/concentric and therefore surgery can be conveniently simplified into two techniques: anterior palate reduction (via laser or monopolar cautery) or expansion palatopharyngoplasty,[5] respectively. Indeed, the authors' have discovered in their practice that an adequate expansion pharyngoplasty procedure suffices to resolve most patterns of velopharyngeal collapse, including AP, and has the advantage of retaining most of the soft palate tissue, thus minimizing the risk of velopharyngeal incompetence.

In essence, expansion sphincter pharyngoplasty entails division, rotation, and fixation of the cut palatopharyngeus muscle onto the ipsilateral hook of the hamulus of the bony palate. This elevates and lateralizes the soft palate and posterior pharyngeal wall. The use of a barbed stitch to perform this, which is also passed through the palate, serves to stiffen the soft palate which will also help reduce the vibrations responsible for snoring.

Complications

Most comprehensive series reveal an overall figure for complications of 20–24%.[6] Most are minor and short term, such as protracted postoperative pain, odynophagia, and temporary dysphagia, infection, and taste disturbance.

Nasopharyngeal incompetence or the need for gastrostomy is extremely rare but the short-term risk of aspiration increases with the number of procedures simultaneously undertaken. A formal swallow assessment in a few limited studies indicate a 6% incidence of aspiration in the immediate surgical aftermath.

Bleeding rates of about 5% are similar to other pharyngeal procedures such as tonsillectomy; of those that bleed, over 98% settle with conservative measures. Adequate analgesia is the key to ensuring enteral intake. A readmission rate of up to 10% has been noted in most case series mainly due to pain or bleeding.

Altered taste is perhaps more common than the reported rate of 14.2%.[6] This indeed may be true for all forms of oropharyngeal surgery and its paucity may be a reflection of inadequate postoperative

questioning. In order to reduce the risk, the authors prefer to release the mouth gag every 30 minutes in a prolonged surgical case (for at least 3 minutes).

Long-term sequelae of surgery include stricture and scarring across the pharynx causing pharyngeal stenosis (0.4%), which is more likely with multiple simultaneous procedures. This can be minimized by actively preserving mucosal bridges at the glossotonsillar sulci and performing palate surgery at a later stage.

Addressing surgical failures

If patients still have OSA (proven on a full comprehensive in-patient polysomnograph) after the initial surgical intervention, and CPAP is still not a viable option, then they should have another DISE procedure. The previous procedures may have unmasked other surgical targets, such as a trap-door epiglottic phenomenon, or may have caused new sources of obstruction, such as pharyngeal scar bands. If epiglottic obstruction remains an issue after tongue base surgery with scarification of its lingual surface (the authors' preferred first-line treatment for epiglottic obstruction), then a limited central V-shaped wedge can be excised, with a small attendant risk of aspiration. Aspiration is minimized by retention of the lateral epiglottic cartilage.

A difficult anatomical problem to deal with following the above surgical procedures is lateral pharyngeal wall collapse. This may necessitate an open approach to effectively tighten and plicate the redundant pharynx. In this setting, a less invasive alternative is a hyoid or laryngeal suspension technique. These techniques not only advance the tongue base, but, by virtue of the constrictor insertion into the hyoid and laryngeal framework, also stiffen the pharyngeal constrictors.

Finally, one should also reassess the facial skeletal anatomy in refractory cases, as the patient may have complex orthognathic issues precluding endoluminal approaches. Maxillomandibular advancement in such carefully select cases can be highly successful.

Results

The success of OSA-related surgery is difficult to gauge due to a range of operative strategies, techniques, and indeed outcome measures reported. A number of centres have provided their figures as single case series. Vicini et al. reported a large multicentre study that included 243 patients who underwent TORS as part of multilevel airway surgery. The mean preoperative AHI score fell from 43 to 17.9 post treatment.[7]

Overall success is significantly influenced by the patient's BMI; for example, Garas et al. demonstrated that patients with a high BMI of 30–35 kg/m^2, have only a 50% 'successful' reduction in AHI scores, in comparison to 75% for those with a BMI less than 30 kg/m^2.[2] For carefully selected patients, maxillomandibular advancement surgery appears to be the most successful surgical procedure in reducing AHI scores; but the corresponding TORS figures from a much wider pool of patients mirror such results in patients with a BMI of less than 30 kg/m^2.[8] Hyoid suspension or genioglossus advancement alone are less successful than TORS to the tongue base and should be reserved as second-line treatment, with specific indications as described.

Hypoglossal nerve stimulation

Paced hypoglossal nerve stimulation has met with some success in the US and other European centres, but the cost is currently prohibitive in the UK at the time of writing. Stimulator electrodes are connected to the anterior branches of the hypoglossal nerve. When activated, they stimulate the geniohyoid and medial genioglossus muscles in phase with inspiration to improve tongue muscle tone and resist retrolingual collapse, even if the tongue does not protrude very much. A DISE pattern of concentric palatal collapse is a relative contraindication and success, as indeed is the case for TORS, is best achieved in patients with a BMI of less than 32 kg/m^2.[9]

Conclusion

SRBDs encompass a broad range of conditions from socially disruptive snoring to severe life-threatening OSA. To manage patients effectively, a multidisciplinary team approach is needed to provide comprehensive and holistic care. The severity of the condition should be determined by sleep medicine physicians through detailed sleep studies if required. Anatomical airway issues should be assessed and addressed by an experienced ENT airway specialist in a systematic manner to maximize the chances of CPAP independence.

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52

Acute and subacute airway obstruction

Navin Mani, Amy Teasdale, and Sofia Clegg

Introduction

Management of the obstructed upper airway is a core skill required by the ear, nose, and throat (ENT) surgeon. Initial assessment must be accurate and prompt, and decision-making on timing and intervention needs to be clear and well communicated to the patient and team. Common causes of acute and subacute airway obstruction include trauma, tumours, infection, and post-surgical airway obstruction. ENT surgeons and anaesthetists work collaboratively when managing such patients to ensure safe control of the obstructed airway; decision-making in terms of when, how, or whether to intubate and need for a surgical airway are often made jointly.

Basic science, anatomy, and physiology

Airway obstruction can occur at any level of the airway from the oral cavity, pharynx, larynx, and upper airways including the trachea and main bronchi. The relevant anatomy is described in Chapter 47. Of note is the anatomy of the larynx (**Figure 52.1**), which extends from the epiglottis and aryepiglottic folds to the cricoid cartilage, which communicates with the hypopharynx superiorly and the trachea inferiorly. Knowledge of this anatomy is required for intubation and front-of-neck airway access with either cricothyroidotomy or tracheostomy.

Evaluation and assessment of the airway

Management of the obstructed airway requires initial rapid evaluation, ideally by an experienced ENT surgeon and anaesthetist for early decision-making. This should assess whether immediate intervention is required by way of intubation or front-of-neck access or whether there is time to bring the patient to the more controlled environment of the operating theatre, which is most comfortable and familiar for both surgeon and anaesthetist. Alternatively, if there is no need for immediate intubation or front-of-neck access then medical management and holding measures should be instituted.

As per any initial assessment, this should begin with gaining a pertinent history, giving an idea as to the underlying cause. The speed of symptom onset is important to elicit as it helps to inform urgency of intervention. Any underlying pathology known to the patient may indicate chronic progressive obstruction, such as slow growth of a lesion, and this may alert the assessor to possible physiological

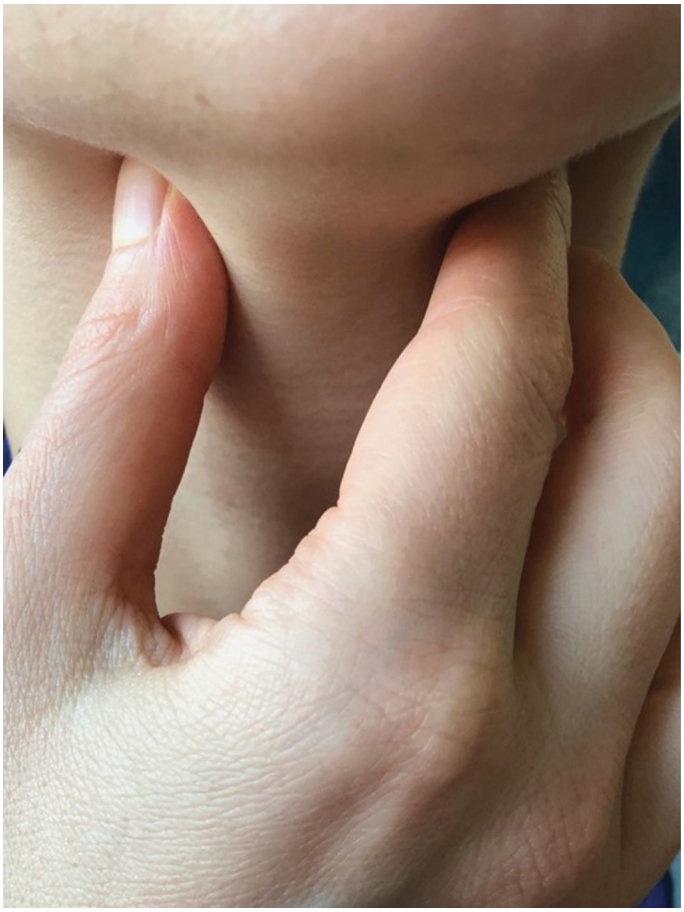

Figure 52.1 Laryngeal handshake: identification of the trachea.

Reproduced from Webber A, Faria J. Rigid bronchoscopy and surgical airway procedures, foreign body aspiration, and gastric content aspiration management. In: Clebone A, Burian BK, eds. *Pediatric Anesthesia Procedures*. Oxford University Press; 2021; online edition, https://doi.org/10.1093/med/9780190685188.003.0005, with permission from Oxford University Press.

compensation through altered breathing patterns or positioning which can mask signs but present the risk of acute decompensation. If time allows, reviewing notes should lead to information on previous surgery to the airway or any past attempts at airway manipulation by interrogating operation notes and anaesthetic charts.

On initial physical examination of the patient both inspection and noise of breathing are important facets of the initial assessment. Examination will reveal signs related to work of breathing, including respiratory rate, use of accessory muscles, ability to talk in full sentences, and presence of any signs of cyanosis. Assessment of the noise of breathing can give valuable clues as to the level of upper airway obstruction, beginning at the level of the oral cavity and oropharynx; obstruction in this area will give rise to stertorous breathing. Obstruction at the level of the glottis leads to an inspiratory stridor sound, obstruction further below in the airway at subglottis and extra-thoracic trachea often gives a biphasic stridor. A purely expiratory stridor can occur with more distal tracheal obstruction at the intrathoracic trachea. The sound and quality of voice can also be important with a 'hot potato' voice typically associated with obstruction at the oropharyngeal level and a muffled voice reflecting supraglottic obstruction, a hoarse voice may indicate obstruction at the vocal cord level from an obstructing lesion, whereas a breathy or weak voice with poor cough may suggest vocal cord paralysis.

Further assessment involves the use of pulse oximetry to assess oxygen saturations, palpation of the neck, and auscultation of the airways and chest.

In a more stable patient, further assessment measures include inspection of the oral cavity and oropharynx and fibreoptic nasopharyngolaryngoscopy, which can often give a definitive assessment of the larynx and pharynx and an idea of the criticality of any impending airway obstruction (**Box 52.1**).

These bedside measures should be enough to make the initial assessment of whether a patient is stable or unstable. In a deteriorating patient, a plan should be made to secure the airway either through endotracheal intubation or front-of-neck access via cricothyroidotomy or tracheostomy and the decision between these options is dependent on the likely underlying nature of the airway obstruction based on the history and examination as detailed above. In a stable patient, further investigation can take place with other treatments and airway temporizing measures put in place.

Investigations

If time allows, investigations can provide important information such as size, position, and movement of lesion, and what may influence the choice of interventions.

Box 52.1 Signs of impending airway obstruction

- Stridor.
- Respiratory distress.
- Inability to speak in a full sentence.
- Cyanosis.
- Oxygen desaturation.

Fibreoptic nasopharyngolaryngoscopy can be a very useful dynamic assessment tool in patients who are able to tolerate this. Where possible this should be performed without local anaesthetic, which may risk precipitating airway obstruction, cough, or laryngospasm. This should ideally be performed on screen as a joint assessment between the ENT surgeon and anaesthetist; useful information can be gleaned not only with regard to the degree and nature of airway obstruction but also as to the difficulty of intubation and can pre-empt any airway adjuncts and method of intubation used.

In stable patients and where there is a new or existing diagnosis of malignancy, cross-sectional imaging with computed tomography or magnetic resonance imaging of the neck can be useful to assess the extent of disease. This is also used often in case of blunt and penetrating neck trauma, and in penetrating neck trauma, computed tomography angiography will inform as to vessel patency. It is imperative that if the decision made is to delay intervention in order to perform these scans, the patient is able to tolerate the duration of the scan and is safe to lie flat without exacerbation of airway obstruction. Mobile lesions may preclude these imaging modalities. In cases of suspected foreign body, plain lateral X-rays of the neck or chest X-rays may be useful.

Ultrasound of front of neck may be used to identify and pre-emptively mark the cricothyroid membrane. In addition to being used as an investigation, it may be used to guide cannula cricothyroidotomy prior to attempting a perceived difficult intubation.

If a blood gas has been performed in the emergency department, elevated carbon dioxide levels may suggest a tiring patient indicating the need for rapid intervention.

Differential diagnoses

Trauma patients will usually be brought into the hospital via the emergency department and in cases of blunt trauma one should consider laryngeal oedema or traumatic perforation. In cases of penetrating trauma, this may be directly into the airway. In cases of anaphylaxis, a history may often be elicited or exposure to common recognized allergens or drugs may be noted in the history. Such patients will often have obstruction at the level of the tongue due to tongue swelling but this may extend into the larynx. Infective causes include supraglottitis, laryngitis, or laryngotracheobronchitis and a history of coryzal symptoms or upper respiratory tract infection and fever may be elicited. In cases of neoplasia, a history of known malignancy may be present or smoking, lifestyle, and alcohol history may be relevant. Postoperative patients may present with airway obstruction from a range of possible pathologies including oedema or haemorrhage depending on the surgery performed. Tracheal obstruction from mediastinal mass should be considered in the differentials for a lower airway obstruction.

Treatment: holding measures

Oxygen

Initial administration of oxygen should be via non-rebreathe mask to treat any potential hypoxia.

High-flow nasal oxygenation

Administering oxygen via a high-flow nasal cannula is invaluable in acute airway obstruction, and it has revolutionized safe airway management over recent years. It is a device that can provide high inspired oxygen concentration at flows up to 70 L/min. Benefits include improved oxygenation, increased apnoea time thus reducing time pressure on team performance, maintaining oxygenation during awake intubation of any technique, reduced work of breathing lessening patient distress, and providing gentle positive pressure to improve airway patency.

Heliox

Another useful adjunct is heliox, which may be of varying gas composition but always a mixture of helium with at least 21% oxygen. The low density of helium compared to the usual nitrogen mix in air allows gas flow of reduced resistance through a narrowed airway, and can also be helpful in relieving patient anxiety. Use of heliox reduces inspired oxygen concentration from other gas delivery techniques therefore preoxygenation with a heliox mixture prior to airway manipulation is suboptimal.

Steroids

Both topical nebulized and systemic steroids when administered in airway obstruction can reduce oedema or inflammation, which may either be the primary cause of the obstruction in cases such as angio-oedema or infection, or can be secondary due to another cause of obstruction such as tumour or foreign body. A common regimen of systemic steroids is intravenous dexamethasone at a dose of 8 mg three times a day. Nebulized topical steroids may also be used, budesonide being a common choice, but onset of action is delayed when compared to nebulized adrenaline. Definitive treatment should not be delayed for administration of steroids in the acute setting, as benefits are not achieved until hours later. Steroids may be more useful in elective or expedited surgery where the drug has time to take effect.

Nebulized adrenaline

Adrenaline acts as a potent vasoconstrictor with a rapid onset of action when given in topical nebulized form. It is usually used at a dose of 1 mL of 1:1000 diluted in 4 mL normal saline and given in nebulized form. This can have an immediate and significant effect on reducing airway oedema and temporizing an unstable airway, which is often used in conjunction with systemic steroids. The effects are, however, short lived and it needs to be given regularly to maintain the efficacy, but nonetheless it can be useful as an interim measure while awaiting definitive treatment (Box 52.2).

Box 52.2 Summary of temporizing measures

- Oxygen.
- High-flow nasal oxygenation.
- Heliox.
- Steroids.
- Nebulized adrenaline.

Treatment: airway interventions

At this point in the patient's management there needs to be a clear strategy including stepwise rescue plans in case of failure at any stage. For example, in a patient presenting with severe facial trauma and perceived difficult facemask ventilation, if primary intubation via the upper airway is plan A but is unsuccessful, plan B might be insertion of a supraglottic airway followed by passage of a fibreoptic scope and intubation, followed by plan C involving rescue techniques such as emergency tracheostomy or rigid bronchoscopy. All team members should know exactly what plans A, B, and C are as well as transition points for moving on to the next step.

Basic airway adjuncts

Airway adjuncts are sometimes used during anaesthesia. Their role in an acute airway obstruction is less useful. Occasionally in a patient with oral/oropharyngeal obstruction, a nasopharyngeal airway may help bypass this, although if the insertion is traumatic it can cause bleeding or precipitate further problems. An oropharyngeal airway is rarely useful in the acute setting and is often not tolerated by a conscious patient, and if incorrectly placed may exacerbate obstruction.

Non-surgical airway management

Communication and coordination between the ENT surgeon and anaesthetist are paramount. Patients with airway obstruction should be identified prior to induction of anaesthesia and an approach to intubation and securing a safe airway should be made between the anaesthetist and the ENT surgeon. Points to consider include initially whether it is possible or sensible to attempt endotracheal intubation, time allowing. The Difficult Airway Society has a set of algorithms for use in unanticipated difficult airway scenarios but not in expected difficult airway situations.

Awake intubation techniques may be required in high-risk situations, provided that the patient is cooperative and not agitated or hypoxic. Options for an awake technique include fibreoptic nasal intubation, video laryngoscopy, and local anaesthetic tracheostomy or cricothyroidotomy. Needle cricothyroidotomy is another technique that may be performed awake, but this technique presents a high risk of barotrauma in airway obstruction. Sedation and local anaesthetic may be helpful in of all the above techniques but need to be used with extreme caution to avoid precipitating complete airway obstruction.

Asleep techniques under general anaesthesia include direct and indirect laryngoscopy, flexible laryngoscopy, or a rigid (Bonfils) stylet (Figure 52.2b). Direct intubation can be performed by the ENT surgeon over a 0° Hopkins rod, or with the use of specialized ENT laryngoscopes. If a general anaesthetic technique is chosen for plan A, then plans B, C and D should be decided upon and discussed with the whole team beforehand. Attempting to wake a patient with reversal of neuromuscular blockade following a failed plan A is high risk and not advisable. A patient with an obstructed airway, despite complete reversal of neuromuscular blockade, is not likely to wake up appropriately. Once a GA has been planned then the airway should be secured through intubation or front of neck access.

Induction of anaesthesia can be either by intravenous or by anaesthetic gas induction. Gas induction has fallen out of favour due to slow onset, difficulty achieving adequate depth of anaesthesia, and requirement for very deep anaesthesia to intubate without a muscle

Figure 52.2 (a) Mellker cricothyroidotomy kit. This kit facilitates cricothyroidotomy using Seldinger technique. (b) The Bonfils retromolar intubation endoscope.

(a) Reproduced from Wang HE. Airway management. In: Yealy DM, Callaway C, eds. *Emergency Department Critical Care*. Oxford University Press; 2013; online edition, https://doi.org/10.1093/med/9780199779123.003.0015, with permission from Oxford University Press.

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relaxant leading to further obstruction and inability to wake the patient in case of failure. It is possible to either allow the patient to maintain spontaneous ventilation or to take over ventilation by invasive positive pressure ventilation, a decision which may be dictated by certain pathologies. Maintaining spontaneous ventilation is the safer technique for mediastinal masses, while laryngotracheal stenosis would require a technique using invasive positive pressure ventilation. Another factor in the decision-making process would be whether the patient is at significant risk of aspiration, and therefore requiring a rapid sequence induction.

Following securement of the airway, the source of the obstruction may be addressed such as by laser debulking of a tumour, or in some cases keeping the patient intubated until, for example, a supraglottic infection settles with the use of intravenous antibiotics and dexamethasone. A plan for safe extubation will also need to be made.

Surgical airway—front-of-neck access

When an immediate airway is required, and an endotracheal intubation is deemed unsafe or not feasible, a prompt surgical airway must be undertaken.

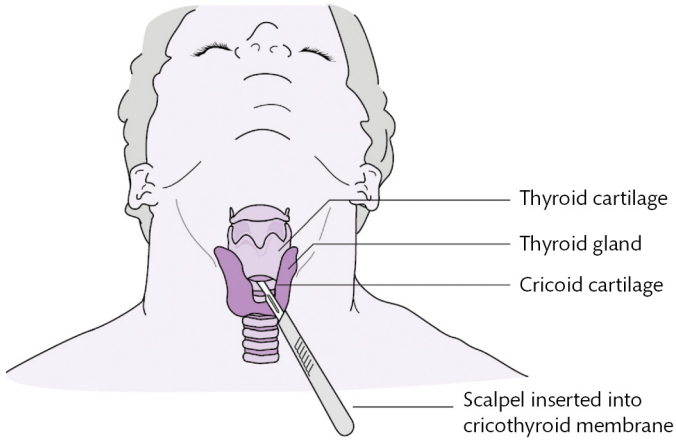

Figure 52.3 Technique of surgical cricothyroidotomy.

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There are two main techniques to obtain an airway in this situation, either cricothyroidotomy or tracheostomy. Scenarios in which this may be required and where intubation is not feasible include:

- major trauma involving facial, oral, laryngeal, or neck structures
- haemorrhage or vomiting that is excluding view or landmarks required for successful intubation
- obstructing tumour or other airway swelling precluding a likely successful intubation
- failed intubation/ventilation emergency scenarios.

Cricothyroidotomy is generally considered to be the quicker of the two methods and simpler to perform (Figures 52.2 and 52.3). A tracheostomy performed in experienced hands can also be quick and efficient and may be the preferred option in certain situations such as tumours of the subglottic region or thyroid.

Tracheostomy

Ideally tracheostomy should be performed in a controlled setting, preferably in the operating room with adequate lighting, instruments, and assistance available. A tracheostomy is most easily performed in an intubated patient under general anaesthesia; however, if that is not possible then a local anaesthetic tracheostomy can safely be performed by an experienced ENT surgeon. This may involve a collaborative approach with the anaesthetist who may titrate sedation to tolerance with extreme caution, where tiny amounts of sedation may lead to complete obstruction or obtund the patient.

The strategy: human factors and non-technical skills in the obstructed airway

Challenging, multidisciplinary shared airway cases require close working relationships between ENT surgeons and anaesthetists, who are expected to both lead, and work as, a team. Shared decision-making reduces pressure, and avoids poor judgement or fixation. Important overlapping non-technical skills expected

of ENT surgeons and anaesthetists are described below, and are arguably as fundamental to success in an emergency situation as clinical skills are.

Situation awareness

- A leader should maintain full awareness of the patient's clinical state throughout the changing situation, as well as the environment and team capabilities, and avoid task-fixation. *Is the patient stable or deteriorating? Is this a district general hospital where complex airways are unfamiliar, and what equipment is available? Is this a junior anaesthetist, and therefore is their consultant available?*
- Understanding the implications of a changing situation and how they affect the strategy. *The patient has become hypoxic and agitated; an awake technique is no longer possible.*
- Proactively anticipate a likely course of events based on situation analysis. *Further deterioration is very likely during induction of anaesthesia.*

Teamworking

- Introductions and allocated team roles create a non-confrontational environment and encourage team cohesion and contribution by all team members.
- Promoting a culture to speak up for patient safety reduces hostility.
- Ensure a shared mental model among the team, with the same mission and goals.

Leadership

In order to maintain situation awareness, the leader may switch from surgeon during anaesthetic interventions to the anaesthetist during a surgical airway.

- Use authority where appropriate.
- Avoid fixation on a failed plan.
- Support the team through the sequential plans.

Communication

- Brief the whole team to communicate urgency and what the lesion/cause is.
- Articulate the strategy and any perceived problems to the team.
- Encourage contribution from all team members.
- Debriefing after a critical event should not be overlooked.

Decision-making

- What is the best current course of action based on resources and clinical situation?
- Following local processes may improve team performance if familiarity ensues.
- Ensure appropriate skill allocation.
- Sequential strategy as per Fourth National Audit Project of the Royal College of Anaesthetists and Difficult Airway Society (NAP4) recommendations.
- Re-evaluation when the situation changes.

Controversies

- Method of intubation.
- When to attempt intubation versus local anaesthetic tracheostomy.

Tips and pearls

- In a patient with an unstable airway, a close working relationship between the ENT surgeon and anaesthetist is paramount to ensure safe outcomes.
- Emergency tracheostomy can be a life-saving procedure and should be performed promptly when needed.

Guidelines

There are no current best practice or consensus guidelines specifically on airway obstruction, therefore teams act on prior knowledge and experience. However, all clinicians involved in airway management should be aware of the Difficult Airway Society guidelines for the unanticipated difficult airway which are a useful tool in any situation involving airway difficulties. These guidelines outline a stepwise approach to tracheal intubation. There are also guidelines from the National Tracheostomy Safety Project for patients of airway obstruction with tracheostomies *in situ* and Advanced Trauma Life Support guidelines for airway management in trauma.

Conclusion

Prompt, rapid airway assessment is required for all patients presenting with acute or subacute airway obstruction. Medical optimization can be undertaken where stability allows, and holding measures should be instituted to delay decompensation. Definitive management may involve intubation or a surgical airway. The overarching goal for managing a patient with airway obstruction in the acute setting is oxygenation. Being decisive, planning for failure, and communicating a clear strategy to the whole team can be achieved through fundamental non-technical skills that will promote team success in any difficult clinical scenario.

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53

Head and neck trauma

Jaiganesh Manickavasagam and Kishore Shekar

Neck trauma

Introduction

The neck is a particularly critical area of assessment and management in the trauma patient. It contains many vital structures. Concern for vascular, neurological, digestive tract, and airway injury are of paramount importance in the evaluation of these patients, as all can be life-threatening. Neck trauma can be split into penetrating injury and blunt injury.

Penetrating injuries of the neck can be classified according to the anatomical level of the injury which becomes important in evaluating and managing these patients (**Figure 53.1**).

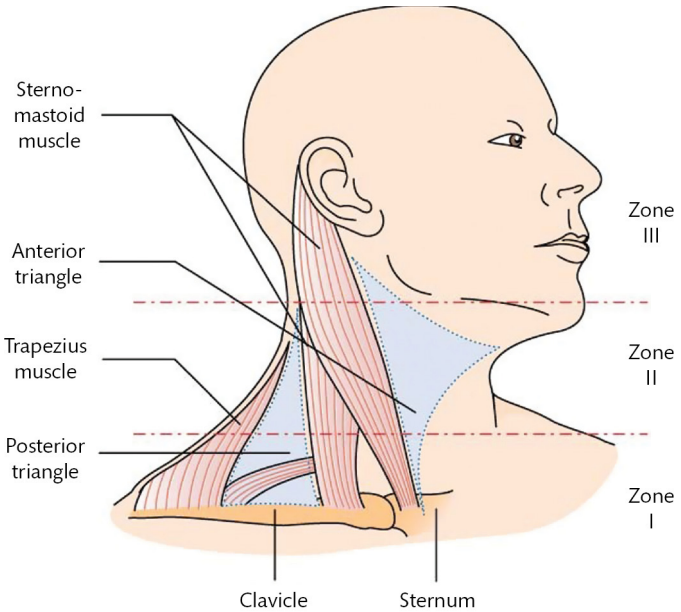

Figure 53.1 Classification of zones of injury for penetrating neck injury. The neck is divided into a posterior and anterior triangle; the anterior triangle is subsequently divided into three horizontal zones.

Reproduced from Van Waes OJ, Cheriex KCAL, Navsaria PH, van Riet PA, Nicol AJ, Vermeulen J. Management of penetrating neck injuries. *Br J Surg*. 2012;99(Suppl 1):149–154, https://doi.org/10.1002/bjs.7733, with permission from Oxford University Press.

- Zone I is defined as the region between the clavicles, sternal notch, and the level of the cricoid cartilage; contains mediastinal structures, thoracic duct, proximal carotid artery, vertebral/subclavian artery, trachea, lung, and the oesophagus.
- Zone II lies between the cricoid and the angle of the mandible; contains carotid/vertebral artery, larynx, trachea, oesophagus, jugular vein, vagus, and recurrent laryngeal nerves.
- Zone III comprises the area between the angle of mandible and the skull base; contains distal carotid artery, vertebral artery, distal jugular vein, salivary glands, and cranial nerves IX–XII.

The assessment and surgical management of injuries in zones I and III are complicated by the proximity of the thoracic cavity, and skull base region, respectively. Most penetrating injuries occur in zone II, where the major structures are relatively more accessible surgically. Zone II injuries also have the best prognosis due to easy access and surgical exposure.

The incidence of penetrating neck trauma is 0.55–5% of all traumatic injuries. Worldwide, the most common cause is a stab wound, followed by gunshot wounds and road traffic accidents. Gunshot wounds can cause tissue injury by three mechanisms: direct tissue injury, temporary cavitation, and transmission of shock waves. Penetrating neck trauma has the potential for serious morbidity and an estimated mortality of up to 6% (**Box 53.1**). Blunt neck trauma is less common than penetrating neck trauma, and is mostly caused by road traffic accidents, assault, and strangulation.

Clinical assessment

Frequently, the neck trauma patient may appear stable, only to have delayed injury found later, causing increased morbidity and mortality. Missed or delayed diagnosis can occur with blunt trauma. **Figure 53.2** provides an overview of the initial management of penetrating neck trauma.

Box 53.1 Causes of death

- Haemorrhage from vascular injury.
- Airway obstruction.
- Cerebral ischaemia.
- Sepsis from oesophageal injuries.

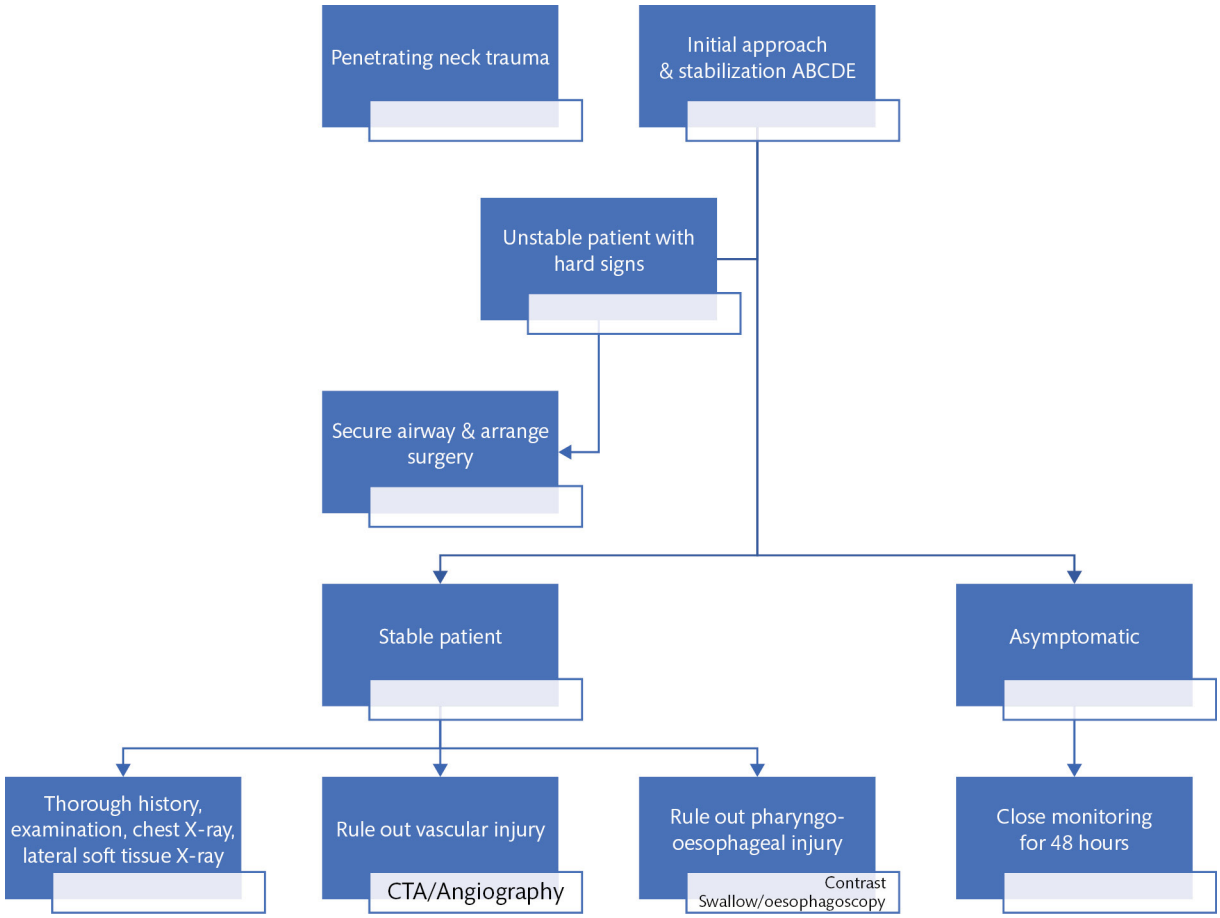

Figure 53.2 Initial management of penetrating neck trauma. CTA, computed tomography angiography.

Stable patients should be evaluated for 'hard' and 'soft' signs. 'Hard' signs indicate the need for operative intervention. 'Soft' signs indicate close observation and monitoring, though not necessarily surgical intervention. As part of the neurological assessment of the patient, all cranial nerves and the sympathetic trunk should be assessed.

Soft signs

- Haemoptysis or haematemesis.
- Dyspnoea.
- Dysphonia or dysphagia.
- Subcutaneous air or mediastinal air.
- Chest tube air leak.
- Non-expanding haematoma.
- Focal neurological deficits.

Hard signs

- Expanding haematoma.
- Severe active bleeding.
- Massive subcutaneous emphysema.
- Air bubbling through a wound.
- Evolving stroke.
- Pneumothorax, severe dyspnoea.
- Oesophageal trauma.
- Shock not responding to fluids.
- Decreased or absent radial pulse, vascular bruit or thrill.
- Cerebral ischaemia.
- Airway obstruction.

Initial approach and stabilization

The need to prioritize the management of significant haemorrhage is especially appropriate in the context of penetrating neck trauma, given that exsanguination accounts for 50% of the mortality from such injuries. Many preventable deaths occur because of massive haemorrhage that may be controlled with pressure.

Given the lack of benefit and indeed possible detriment of cervical spine immobilization in the context of a low-velocity penetrating injury to the neck, cervical spine protection is not advocated as part of an initial stabilization strategy. Exceptions to this guidance include a focal neurological deficit at presentation or a high index of suspicion for spinal injury in an unconscious or heavily intoxicated patient.

If the patient is stable, a portable chest X-ray, as well as antero-posterior/lateral views of the neck should be obtained to look for any bullet fragments, soft tissue swelling, or air outside the trachea. Detailed investigations are discussed in the appropriate later sections.

Airway management

Physical signs that warrant immediate airway management include stridor, respiratory distress, shock, or a rapidly expanding

Table 53.1 Various investigative modalities to assess vascular injury

Modality	Advantage	Disadvantage
Catheter angiography	Simultaneous endovascular interventions feasible	Limited availability outside of office hours, time-consuming, higher risk of neurovascular complications
Computed tomography angiography	Ready availability and minimally invasive	
Colour flow Doppler	Non-invasive	Operator dependent, unlikely to give useful information regarding non-vascular traumatic injuries

haematoma. A variety of means of securing the compromised airway have been described in penetrating neck trauma. This can be achieved by oral endotracheal intubation using rapid sequence induction of anaesthesia and followed by a tracheostomy. Fibreoptic intubation can be considered based on the patient's status and the skillset in theatres. The airway will be more difficult to secure with time, and repeated attempts could be detrimental. Once the airway is secured, other injuries to the larynx or trachea are usually treated surgically. In the presence of significant disruption of laryngotracheal anatomy, surgical control of the airway is advocated. When an injury to the upper aerodigestive tract is suspected or identified, further evaluation with panendoscopy and bronchoscopy under general anaesthesia is often helpful. In hanging and strangulation victims, and where the nature of injury is unclear, early or prophylactic intubation should be considered. In addition, these patients have a propensity to develop pulmonary oedema and acute respiratory distress syndrome.

Management principles

Management of penetrating wounds to the neck remains controversial. The low threshold for operative management in the past has been largely replaced by a selective conservative approach.

The principles of the management of high-velocity injuries are similar to those for low-velocity injuries, but the unpredictable trajectory of firearm projectiles through soft tissue, potential for cavitation or thermal injury, and impairment of cross-sectional imaging by retained metallic fragments mean that high-velocity injuries are much more likely to require operative exploration than low-velocity injuries.

Vascular injury

Investigations for suspected vascular injury

Catheter angiography is the 'gold standard' investigation for suspected vascular injury. But for reasons mentioned in **Table 53.1**, multislice computed tomography (CT) angiography has now supplanted angiography as the test of choice—in all three zones of the neck.

Management of vascular injury

Consultation with a vascular surgeon and interventional radiologist is a good practice to determine the most appropriate management for stable patients (**Figure 53.3**). A positive angiogram usually requires operative or endovascular intervention, while a negative angiogram is taken to signify a venous injury and conservative management is appropriate. Penetrating jugular venous injuries can be safely managed without operative intervention in asymptomatic patients as it involves a low-pressure system.

The leading cause of immediate death is exsanguination. Packing of wounds with no attempt to reconstitute vascular continuity provides the mainstay of initial management in a non-specialized trauma centre. Foley catheter balloon tamponade may obtain temporary control of haemorrhage from a penetrating neck injury if manual compression is unsuccessful. When a common or internal carotid artery injury is identified during a neck exploration, the consensus from the literature is that repair of the artery is generally preferable to ligation, irrespective of whether a preoperative focal neurological deficit was present or not. Any attempt at arterial repair is clearly best performed in conjunction with an experienced vascular team.

Laryngotracheal injury

Assessment

The presence of hoarseness and subcutaneous emphysema in the absence of pneumothorax indicates trauma to the upper aerodigestive

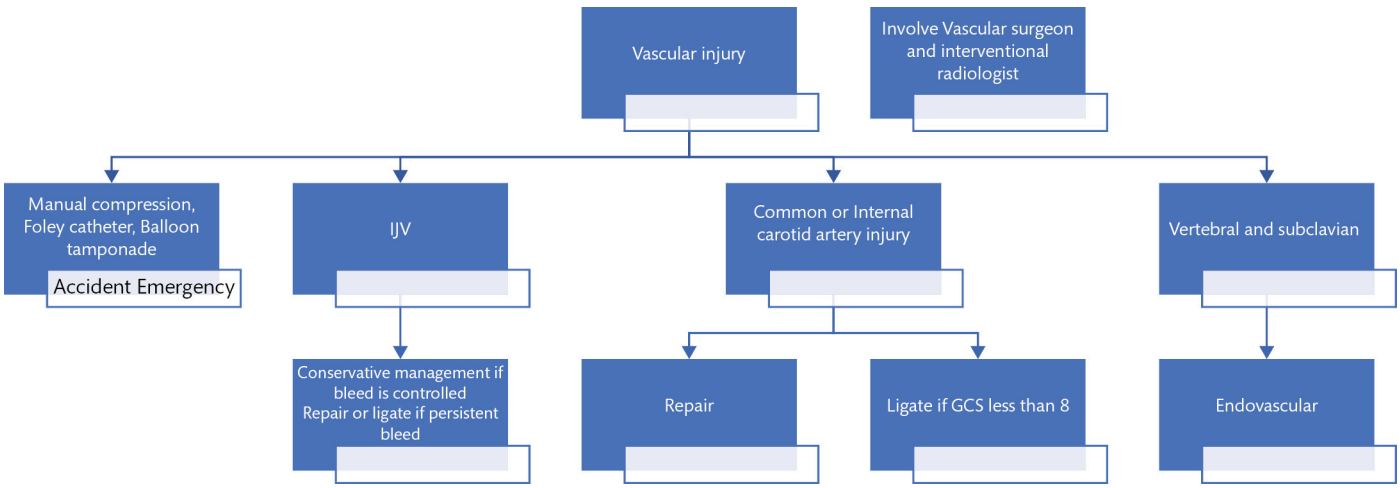

Figure 53.3 Management algorithm for vascular injury. GCS, Glasgow Coma Score; IJV, internal jugular vein.

Table 53.2 Schaefer Classification System for laryngotracheal injury

Group	Severity of injury	Management
1	Minor endolaryngeal haematoma, lacerations without fractures	Flexible endoscopy, steroids, antibiotics, humidification, head-up position, antireflux treatment, and voice rest
2	More severe oedema, haematoma, without exposed cartilage, or non-displaced fractures	Serial flexible endoscopy examinations as injuries may worsen
3	Massive oedema, large mucosal lacerations, exposed cartilage, displaced fractures, or vocal cord immobility	Tracheostomy and surgical repair often required
4	Group 3 and more severe disruption of larynx, unstable fractures	Tracheostomy and surgical repair, often require stent
5	Laryngotracheal separation	Airway usually temporarily established using endotracheal tube inserted through neck. Laryngotracheal repair is carried out

tract. Laryngeal fractures are rare, often present in association with maxillofacial injuries, and may lead to life-threatening airway obstruction, impaired vocal function, dysphagia, chronic aspiration, and death. There is also a high incidence of associated recurrent laryngeal nerve injury in patients with fracture of the cricoid cartilage; concurrent injury to major vessels and the thyroid gland are also commonly seen.

Stable patients, and those in whom the airway is secure, can undergo cross-sectional imaging to further elucidate the nature of the injury.

There have been many proposed classifications for laryngeal trauma that categorize injuries according to sites, tissues injured (cartilage, mucosa, ligaments, nerves, joints), and severity. The Schaefer Classification System appears to be the most useful as it allows the clinician to make treatment decisions based on the severity of the injury (Table 53.2).

Various treatment approaches have been advocated, depending on the extent of the endolaryngeal injury and the amount of cartilaginous displacement (Figure 53.4).

Hyoid bone fractures are managed conservatively.

A calcified laryngeal cartilage in an older patient may fracture in more than one place, whereas a more elastic larynx will usually fracture at a single site in a younger person. Fracture of the thyroid cartilage occurs when the cartilage is forced against the cervical spine, flattens, and springs back into position, resulting in an anterior linear fracture.

In patients with minor injuries and non-displaced fractures with a safe airway, where there is no evidence of internal derangement on endoscopy, tracheostomy is not required and they are candidates for non-operative management with supportive measures.

All displaced fractures of the thyroid cartilage should undergo open reduction via a low cervical thyroidectomy incision and should

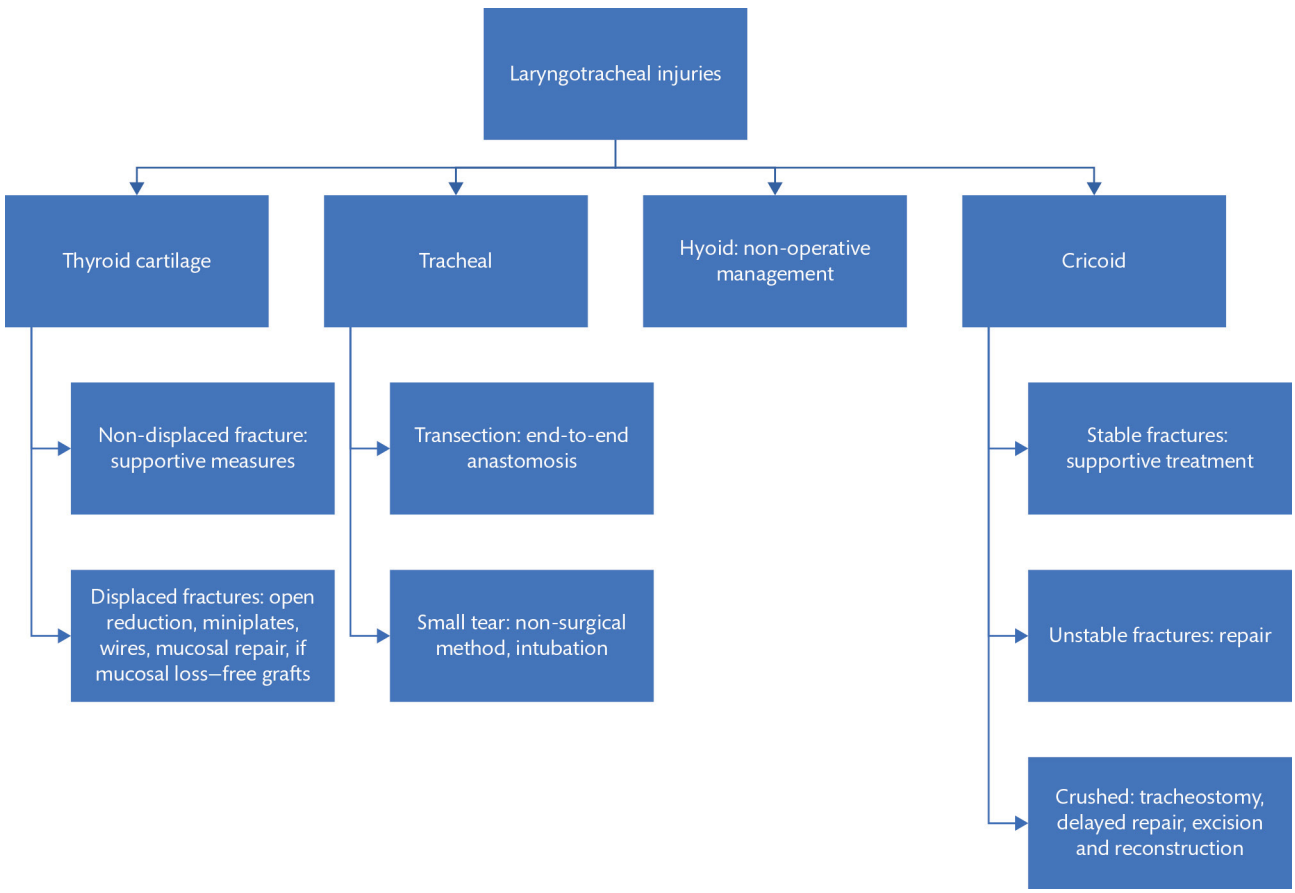

Figure 53.4 Management of suspected laryngotracheal injury.

be realigned with a miniplate, wire, or non-absorbable monofilament suture. Miniplate fixation was shown to be superior to suture or wire fixation because it promotes complete cartilaginous union, whereas the latter two methods promote healing by the fibrous union. The external perichondrium of the cartilage should always be re-approximated. If there is also evidence of internal derangement on preoperative endoscopy, such as an avulsed vocal cord or displaced epiglottis, this must be repaired after the cartilage to ensure that a proper scaffold is obtained before realigning the mucosa. The laryngeal lumen can be accessed either through the fracture itself or via a laryngofissure midline incision through the thyroid cartilage. Mucosal defects are repaired with absorbable suture and buried knots to prevent granuloma formation. If there is extensive mucosal loss, free grafts from the buccal mucosa, skin, or dermis may be used to fix the defect.

Cricoid cartilage Since the cricoid is a complete ring, it usually fractures in two places, anteriorly and posteriorly. Non-displaced, stable fractures can be managed non-operatively. If the fractured segments are unstable, the cartilage should be wired and a soft stent, such as silicone, should be inserted and kept in place for 4–6 weeks. Patients with crushed cricoid rings should have a tracheostomy placed on presentation, and the cricoid should be excised in a delayed fashion once the surrounding oedema has improved. This usually only requires excision of the anterior half of the cartilage, which can be replaced with a hyoid bone or rib graft.

Stents Airway stents are often placed in the setting of massive endolaryngeal injury to prevent mucosal adhesions and laryngeal stenosis. Another indication for use is for laryngeal injuries in which the anterior commissure is disrupted. Stent placement helps to maintain the proper alignment of the commissure and prevent anterior glottic webs. In general, however, if a robust repair of mucosa and full reduction and stabilization of the laryngeal fracture can be achieved, it is preferable to avoid the use of stents given their potential complications. These include infection, granulation tissue and scar formation, and pressure necrosis, which is usually caused by using stents that are too large for the airway. Scarring from stents may lead to stenosis and impaired vocal cord mobility. A large variety of hollow and moulded stents that vary in size, shape, and material are available. Silicone stents are generally preferred. On average, stents are usually left in place for 2 weeks.

Tracheal transection In cases where the cervical trachea is completely transected, a transverse incision is made, and an endotracheal tube is inserted into the distal trachea. The entire anterior chest should be prepared and draped into the operative field so that thoracic surgical help can be sought if there is a significant loss of the trachea. Devitalized tracheal tissue and ragged edges should be debrided. However, care must be taken to preserve as much remaining tracheal tissue as possible. Repair of the trachea is performed in an end-to-end interrupted fashion with Vicryl or Monocryl 4-0 sutures, through all tracheal layers, at an interval distance of 3–4 mm. Care should be taken to preserve the blood supply, which runs along both lateral aspects of the trachea, to prevent ischaemia which would cause poor healing and delayed stricture. Internal stents are not necessary.

Intrathoracic tracheal injuries are approached via a right posterolateral thoracotomy incision. If both the trachea and oesophagus have been injured, the oesophagus must be repaired in two layers. Meticulous repair is needed to prevent a tracheo-oesophageal fistula. Small tracheal wounds where there has been no loss of tissue and where the tracheal edges are well apposed can be treated non-operatively by temporary orotracheal or nasotracheal intubation. The cuff of the endotracheal tube should be inflated below the wound to prevent leakage of air into the subcutaneous and mediastinal spaces, thereby decreasing contamination of these areas. Small tracheal wounds usually seal spontaneously within 48 hours. Cricoid–tracheal separation is treated initially with a tracheostomy. It is frequently associated with bilateral recurrent laryngeal nerve injury and subsequent tracheal stenosis. Recurrent laryngeal nerve injury should be repaired directly where possible but carries a poor chance for functional return.

Airway rupture extending beyond the carina requires placement of an endotracheal tube under bronchoscopic visualization into the mainstem bronchus opposite to the side of the injury to provide single-lung ventilation.

Pharyngo-oesophageal injury

Injury to the oesophagus is uncommon (5%) in neck trauma. However, delayed diagnosis can cause mediastinitis and is the leading cause of delayed death in neck trauma. Oesophageal injuries are often clinically silent, so they ought to be investigated and ruled out. Contrast-enhanced oesophagraphy has a sensitivity of 89%, with rigid endoscopy having a similar sensitivity. Flexible endoscopy has a lower diagnostic yield than rigid endoscopy but has a lower complication rate. When both contrast-enhanced imaging and endoscopy are used, sensitivity approaches 100%.

Signs of oesophageal injury

- Odynophagia.
- Haematemesis.
- Surgical emphysema.
- Prevertebral air on radiology.

Management of pharyngo-oesophageal injury

Figure 53.5 outlines a scheme for the management of pharyngo-oesophageal injuries. All patients with suspected penetrating oesophageal trauma should receive intravenous antibiotics and kept nil by mouth. Nutrition should be managed via either a nasogastric tube or total parenteral nutrition. Many centres now adopt a more selective approach to exploration unless there is evidence of a large defect or deterioration of the patient's condition. The need for close monitoring in the hospital is crucial if the conservative approach is adopted.

Controversies/areas of interest for the future

Some studies have highlighted concerns over the use of rapid sequence induction in trauma patients. These concerns relate to the loss of airway tone induced by rapid sequence induction, which may lead to further airway compromise.

Although controversy continues to surround the management of penetrating neck trauma, the role of selective non-operative management and the utility of CT angiography to investigate potential vascular injuries appears to be increasingly accepted. Mandatory surgical exploration for all injuries that breach the platysma is now no longer widely indicated.

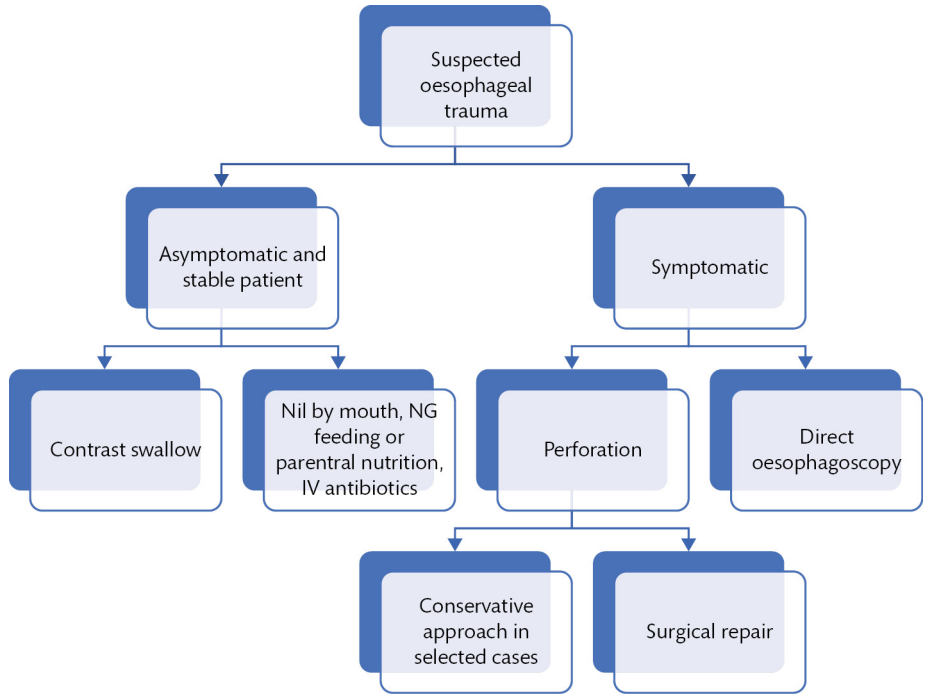

Figure 53.5 Management of pharyngo-oesophageal injury. IV, intravenous; NG, nasogastric.

The 'no zonal approach' to penetrating neck trauma is a selective approach with superior patient outcomes in comparison with traditional management principles.

Mandatory exploration for suspected oesophageal trauma has traditionally been advocated as delays while investigating can lead to increased morbidity, However, small, contained pharyngo-oesophageal leaks can be treated by conservative measures alone.

The use of laryngeal stents remains an area of controversy. The rationale behind stenting is that they serve initially as an internal fixation device and thereafter should prevent endolaryngeal scarring and maintain the internal configuration of the larynx. However, certain studies have shown that stents may increase the incidence of infection and granulation tissue.

Regardless of the type of stent, most centres advocate their removal at the earliest opportunity, usually within 10–14 days to help minimize mucosal damage. But a severely damaged larynx may need stenting for up to several months.

Tips and pearls

- Secure the airway early or prophylactically if necessary.
- CT angiography is the test of choice.
- Involve a multidisciplinary team early and often.
- Oesophageal injury is often missed and is a cause of delayed mortality.
- Monitor patients closely if a conservative approach is adopted.

Facial trauma

Introduction

Complex craniofacial injuries are less common. The cause of these injuries has also changed. This is attributed mainly to changes in road safety regulations. While in the 1980s road traffic accidents were the commonest cause, interpersonal violence and sport injuries are more common in the present day. The changes in the causative factors and incidence have also led to changes in trauma pattern.

In the maxillofacial region, soft tissue injuries are the most common followed by dentoalveolar injury, zygomatic complex fracture, mandible fractures, orbital floor fractures, midface fractures, and complex craniofacial and pan facial fractures. This following sections aim to give an overview of the management strategy for these injuries.

Soft tissue injury

Most soft tissue injuries are facial lacerations with minimal or no soft tissue loss. The excellent blood supply to the facial skin with numerous anastomotic areas makes dealing with these injuries simple and easy.

Management

The surgical principles for the care of soft tissue are:

- clean and debride
- haemostasis
- attention to neural injury
- closure.

Attention to neural injury

Where a transection of the facial nerve is suspected, it is best to consider nerve repair at the time of initial exploration, with or without a nerve graft. Delayed neural repair is more difficult and results vary.

Closure of wounds

There is often enough elasticity of the facial skin to allow for undermining and primary closure of lacerations. However, in areas around

the eyelid, nasal sill, and oral commissure, it may be best to consider a local flap in case of tissue loss. This avoids complications such as ectropion, distortion of the commissure, and widening/narrowing of nasal aperture. Where experience of managing these lacerations may be limited, one may consider placing a skin graft on the wound bed and a more definitive reconstruction carried out as a planned elective procedure.

One of the areas of contention is closure of wounds because of human/animal bites. Thorough debridement and a course of antibiotics is recommended. There are proponents of both primary and secondary reconstruction.

Fractures of the facial skeleton

This section aims to cover the fundamental principles in the management of fracture of the facial bones.

Nasal bone fractures

Circumorbital ecchymosis, deviation of nasal bones, deviation of nasal tip, nasal bleeding, and septal haematoma are the likely clinical features. Isolated nasal bone fractures require no further investigation.

Management

Septal haematomas require immediate attention; drainage can be accomplished by a simple incision along the nasal septal mucosa. Low-volume suction tends to help in accomplishing adequate evacuation. An inflatable nasal pack placed for 24 hours prevents recollection.

Reduction of nasal fractures can be carried out under local or general anaesthesia. The aim is to set the nasal bones and nasal septum back to their preinjury anatomical site. Closed manipulation of the fracture often achieves this. An external splint, left in place for 7–10 days, will protect the realigned segments. Comminuted fractures may require a second revision surgery electively, in the form of an open rhinoplasty.

Fractures of the zygomatic complex

Clinical features include flattening of the face, numbness in the distribution of the inferior orbital nerve (skin over the cheek and lateral wall of the nose), restriction in mouth opening, and subconjunctival ecchymosis with no lateral extent—if the lateral extent is seen, this is pathognomonic of a tripod fracture; associated orbital flow fractures may result in diplopia. Zygomatic complex fractures may also be associated with other midface fractures.

Investigations

Occipitomental radiographs of 15° and 30° will show the fractures; where a comminution of the tripod is clinically suspected, or a fracture of the orbital floor is diagnosed, CT scans are mandatory.

Management

Fractures of the tripod are treated surgically when there are functional or cosmetic issues. Surgery can be done electively as a day case, via a Gille's temporal approach and closed reduction with a lift can be accomplished. When significant displacement with periosteal tear is present, open fixation with mini plates is indicated.

Orbital floor fractures

These are often seen following sport injury and interpersonal violence.

Clinical features

Diplopia is present on upward gaze, indicating entrapment of either muscle or periorbita. The absence of double vision does not mean the absence of a fracture. Large orbital defects do not cause entrapment of the muscle as such; there may be no evidence of double vision. The other features include circum-conjunctival ecchymosis, periorbital ecchymosis, and paraesthesia of the infraorbital nerve.

Investigations

A thorough clinical examination should include visual acuity assessment and further referral to ophthalmology as needed.

White eye blow out

This occurs in children, where the floor of the eye acts as a trap door that opens and shuts, resulting in trapped periorbita and profound diplopia at all gazes including the primary gaze.

Management

A white eye blow out is an emergency as strangulation of the orbital contents results in avascularity and scaring. A CT scan is obtained, and orbital exploration is carried out as an emergency. Other orbital fractures can be treated electively after 7–10 days, allowing orbital swelling to settle down. The surgical principles are outside the scope of this text.

Fractures of the mandible

Contact sport, interpersonal violence, road traffic accidents, and industrial injuries remain the main causes of mandibular fractures.

Clinical features

The mandible has three areas of weakness where most fractures occur. In addition, mandibular fracture sites are often multiple at presentation. The areas of weakness are the neck of condyle, angle of mandible, and parasymphysis. The common combinations of fractures are:

- angle plus parasymphysis
- midline plus bilateral condyle
- bilateral body fractures.

All displaced fractures have a change in occlusion and the patient reports an 'altered bite'. The pathognomonic sign of a bicortical fracture of the mandible is ecchymosis in the floor of mouth. Greenstick fractures may not have these two features. Other clinical features may include numbness of lip, fractured teeth, and fractured dental segments.

Investigations

The minimum radiological investigation is and orthopantomogram and a posteroanterior view of the mandible. Where an orthopantomogram is not available, a lateral oblique X-ray may suffice. CT scans are not routinely needed and are reserved for comminuted fractures, condylar fractures (high condyles), and late presentation resulting in a non-union or malunion.

Management

Appropriate attention to the airway is needed. Bilateral fractures of the body or parasymphysis of the mandible can lead to a haematoma

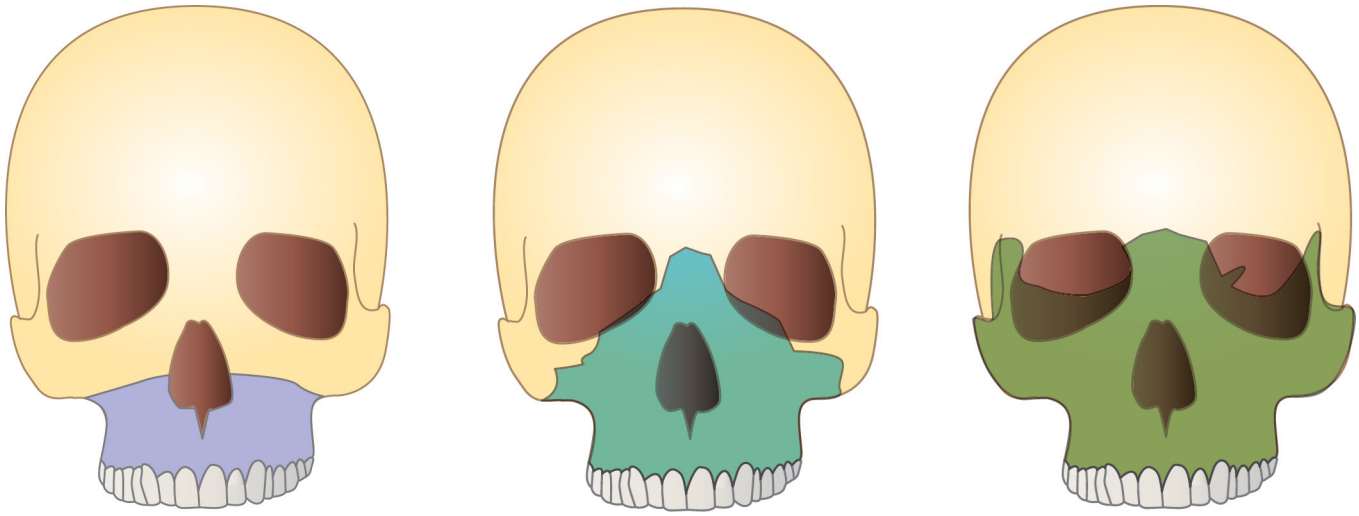

Figure 53.6 Le Fort's classification of midfacial fractures. Le Fort I, horizontal fracture of the maxilla, also known as Guérin fracture. Le Fort II, pyramidal fracture of the maxilla. Le Fort III, craniofacial disjunction.

Source data from Wooley CCH, in Converse JM, ed. *Reconstructive Plastic Surgery: Principles and Procedures in Correction, Reconstruction, and Transplantation*. 2nd ed. Saunders; 1977.

in the floor of mouth, raising the tongue, thus compromising the airway.

A mandible fracture involving the dental segment is a compound fracture due to its communication into the oral cavity, needing pre-operative antibiotics.

Pain secondary to mobility of fractures at the dental segment can be controlled with temporary lasso wiring around the teeth. Stainless steel wires can be placed under local anaesthesia.

The definitive management of fractures of the mandible is open reduction and fixation under general anaesthesia. Occasionally intermaxillary fixation by wiring the jaws together may be indicated in high condyle fractures, infected mandible fractures, and comminuted fractures.

Midface fractures

Midface/maxillary fractures are now relatively rare, often being caused by high-velocity trauma secondary to road traffic accidents.

Clinical features

Maxillary fractures are classified as Le Fort I, II, or III (Figures 53.6 and 53.7). These fractures can occur in isolation or as combination fractures, with the clinical presentation being varied. Mobility of the maxilla is not a necessity as the displacement of maxilla in a posterior and downward direction along the skull base can cause the maxilla to impact. Anterior open bite and palatal ecchymosis indicate a Le Fort I fracture; bilateral circum-orbital ecchymosis, Le Fort II and III; and skull base and dural tear with CSF leak a Le Fort III fracture. A displaced Le Fort II and Le Fort III results in what is classically described as a 'dish face'.

Investigations

Fine-cut CT scans of the midface remain the investigation of choice. Three-dimensional rendering makes planning easier.

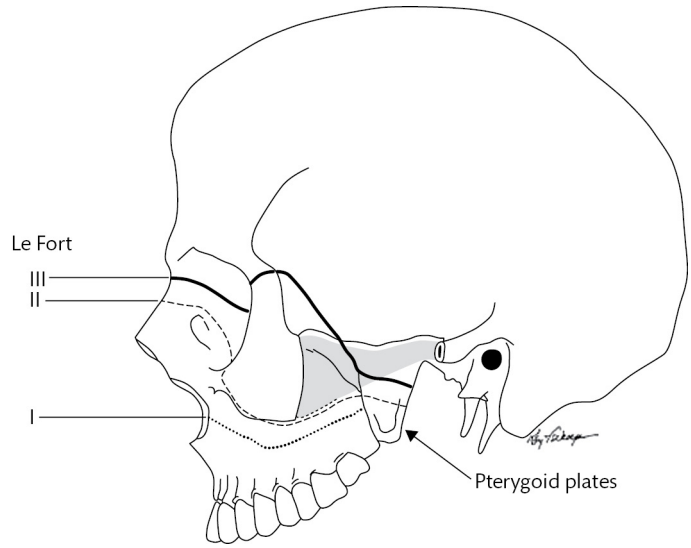

Figure 53.7 Le Fort fractures, lateral views. Note that all the fractures extend posteriorly through the pterygoid plate (arrow).

Source data from Converse JM, ed. *Reconstructive Plastic Surgery: Principles and Procedures in Correction, Reconstruction, and Transplantation*. 2nd ed. Saunders; 1977.

Management

The immediate management involves the principles of ATLS. Concomitant head injuries, if present, take precedence. Patients are often kept for a period of observation. Bleeding from the nose may necessitate nasal packing. Bleeding from the maxilla itself can be reduced by placement of a bite block between the teeth and bringing the jaws together, causing maxillary impaction against the skull base to stem the flow of blood. The definitive management of a midface fracture is open reduction and internal fixation when the swelling has settled.

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54

Oral ulceration

A pragmatic guide to diagnosis and management

Stefano Fedele and Stephen Porter

Introduction

Oral ulceration is a common complaint of patients of all ages attending the Otolaryngology outpatient clinic. Painful symptoms are often associated with oral ulceration and can lead to difficulty with eating, speaking, and swallowing. Occasionally, dehydration and malnourishment may be seen in patients with severe widespread oral ulceration. In immunocompromised patients, the breach in mucosal integrity caused by persistent oral ulceration can lead to septicaemia, which is a significant problem especially in patients undergoing stem cell transplant and other cancer treatment including chemoradiotherapy of the head and neck.

Generally speaking, local trauma is the most common cause of acute transient oral ulceration in adults, whereas younger people are usually affected by recurrent aphthous stomatitis (RAS). However, attention should be paid to consider potential mucocutaneous and systemic disorders, malignancy, and drug reactions presenting as oral ulceration. For example, a single indurated ulcer lasting more than 3 weeks should always raise the suspicion of malignancy or a chronic infection, whereas multiple recurrent or persistent ulcers can be associated with systemic diseases such as blood disorders, infections, gastrointestinal disease, and skin disease. Table 54.1 summarizes the main causes of oral ulceration.

Reviews on the possible causes of oral ulceration are widely available; however, there remains little practical guidance for clinicians regarding how to approach patients with oral ulcers in day-to-day clinical practice.

The present chapter is aimed at providing specialists and trainees in otolaryngology with a pragmatic guide on assessing patients with oral ulceration including medical history and clinical examination, how to navigate through the differential diagnosis of potential causes, when to suspect a malignancy, how to treat some of the common causes of ulceration, and when to refer to other specialists (see diagnostic algorithm in Figure 54.1).

Taking a detailed history

A pragmatic approach to the diagnosis of oral ulceration starts with taking a detailed medical history and history of the presenting complaint, in particular:

- the number of ulcers (single vs multiple)
- their site(s)
- their duration, for example, whether they are acute (lasting only a few days), persistent (lasting weeks or longer), or recurrent (healing spontaneously and occurring again)
- possible triggers (trauma or medication).

A single isolated ulcer lasting more than 3 weeks may represent malignancy or a chronic infection whereas multiple persistent ulcers are more typical of systemic disease.

Multiple, recurring, short-lasting small ulcers affecting multiple intraoral sites in children or young adults are typical of RAS, but when they repeatedly affect the same unilateral intraoral site or lip, a diagnosis of recurrent herpes simplex virus (HSV) infection should be considered.

Sharp or broken teeth, dentures, and other appliances (e.g. orthodontic braces), bites during chewing, and medications applied to or held in the mouth (such as aspirin) are common local causes of traumatic acute ulceration.

The medical history should always consider potentially associated systemic disorders (haematological disease, gastrointestinal and skin diseases, infections, vasculitic and other immunologically mediated disorders, systemic malignancy) or drug use.

Clinical examination

A comprehensive examination of the whole oral mucosa and the presenting ulcer(s) is of critical importance. The authors strongly suggest that clinical examination is performed under appropriate lighting in order to allow a full appreciation of all intraoral mucosal sites and a

Table 54.1 Local and systemic causes of oral ulceration

Local causes/disease affecting primarily the oral mucosa	
Trauma	• Physical/chemical • Burns • Irradiation
Immunological disease	• Recurrent aphthous stomatitis • Eosinophilic ulcer (traumatic ulcerative granuloma with stromal eosinophilia (TUGSE))
Infection	• Acute necrotizing ulcerative gingivitis (ANUG) • Recurrent oral herpes (herpes simplex virus reactivation) • Herpes zoster (shingles) (varicella zoster virus reactivation)
Malignancy	• Oral squamous cell carcinoma • Sarcoma • Extranodal lymphoma (including cutaneous T-cell lymphoma)
Systemic causes/oral ulceration as part of a systemic disease with extraoral symptoms	
Blood diseases	• Anaemia • Neutropenia • Hypereosinophilic syndrome
Infections	• Primary herpetic stomatitis (herpes simplex virus) • Chickenpox (varicella zoster virus) • Infectious mononucleosis • Cytomegalovirus • HIV • Coxsackieviruses and echoviruses (herpangina and hand, foot, and mouth disease) • *Mycobacterium tuberculosis* and atypical mycobacteria • *Treponema pallidum* • Mycoses (candidosis, histoplasmosis) • Leishmania • Others
Gastrointestinal disease	• Coeliac disease • Crohn's disease • Ulcerative colitis
Mucocutaneous disease	• Lichen planus/lichenoid disorders • Pemphigus • Pemphigoid • Erythema multiforme • Dermatitis herpetiform • Epidermolysis bullosa
Vasculitis	• Lupus erythematosus (systemic and discoid) • Behçet's disease • Granulomatosis with polyangiitis • Sweet syndrome • Reiter syndrome
Other immunological disorders	• Autoinflammatory syndromes • Immune-related adverse events of checkpoint inhibitors (cancer immunotherapy)
Drug-induced	• Cytotoxic agents (bone marrow toxicity) • Alendronate • Nicorandil • Non-steroidal anti-inflammatory drugs
Malignancy	• Leukaemias and myelodysplastic syndromes • Lymphomas • Metastatic solid malignancy (e.g. breast cancer, sarcoma)

detailed assessment of the clinical features of the ulceration. Standard dental lights or examination/operation lamps commonly used for minor surgery are preferable to head mirrors/lamps or penlights, which often have insufficient intensity or colour balance. A detailed description of oral cavity examination, in addition to the areas of the upper aerodigestive tract mucosa, can be found in Chapter 46. Visual inspection includes the detection of the major characteristics of oral ulceration, but also the assessment of all other intraoral and perioral areas with a view to detect any associated abnormality. Attention should be paid to assess the number and sites of oral ulceration (single vs multiple), the dimension (small vs large), the appearance of the margins (flat vs elevated) and shape (regular vs irregular), any bleeding, as well as the presence of other oral mucosal lesions (any site) or submandibular/cervical swelling (e.g. lymph nodes).

Removable appliances (e.g. dentures) should be removed prior to clinical examination and a gauze swab is recommended to help in

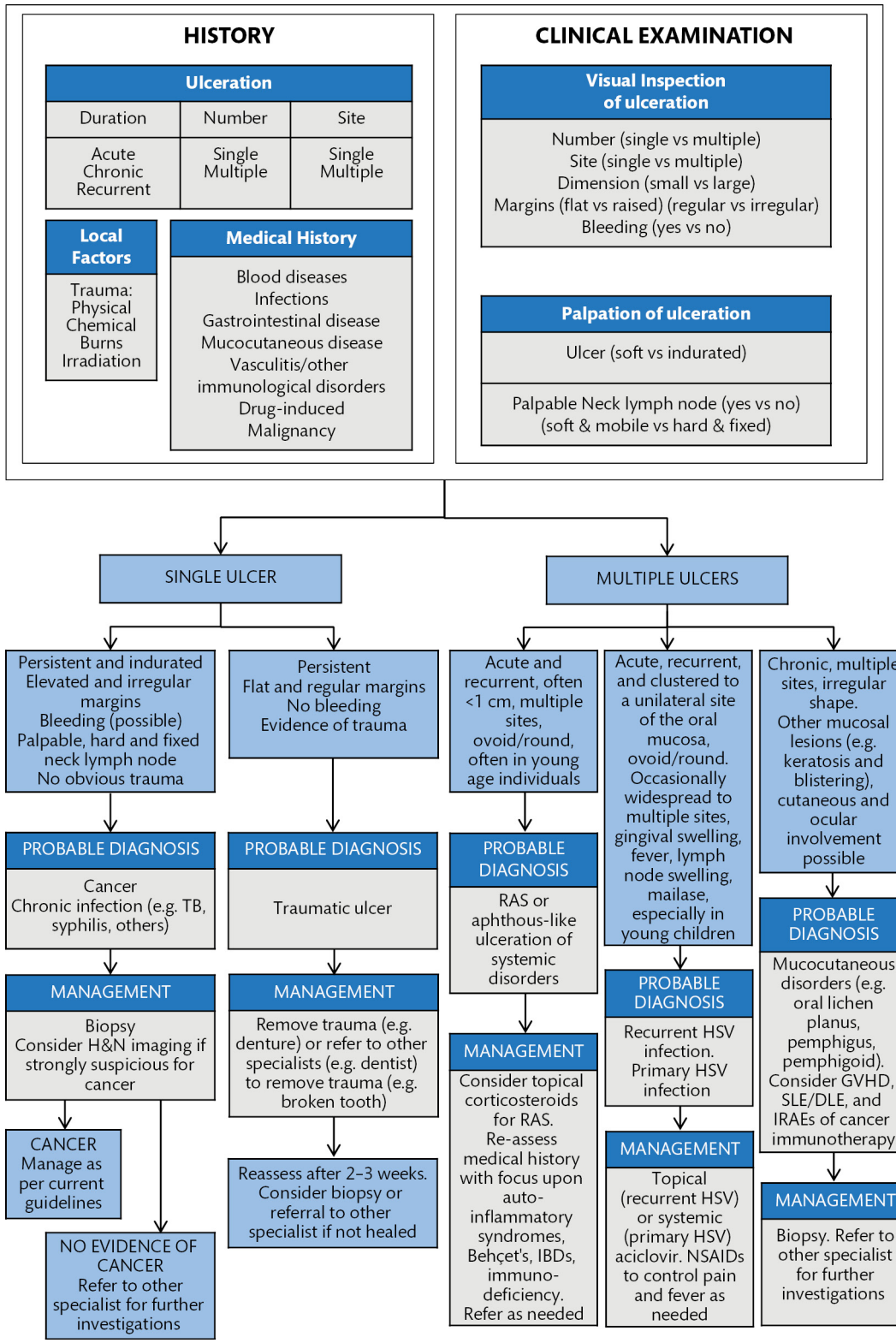

Figure 54.1 Diagnostic and management algorithm of the most common causes of oral ulceration. GVHD, graft-versus-host disease; HSV, herpes simplex virus; IBD, inflammatory bowel disorders; IRAEs, immune-related adverse effects; NSAIDs, non-steroidal anti-inflammatory drugs; SLE/DLE, systemic and discoid lupus erythematosus; RAS, recurrent aphthous stomatitis.

stretching and pulling the oral tissues and to permit careful evaluation of all areas. A dental or laryngeal mirror may also aid adequate visualization of the intraoral areas and is preferable to the use of a wooden tongue depressor which cannot direct or reflect the light and is not rigid enough. Palpation is a critical part of the clinical examination as it allows to capture important characteristics of the oral ulceration including the overall consistency (soft vs indurated) and margins (soft vs firm and infiltrating), as well as the presence and features of palpable submandibular or cervical swelling (mobile vs fixed).

Diagnosing the cause of oral ulceration

Single mouth ulcer

Single persistent ulcer with clinical features suggesting malignancy or chronic infection

A single long-standing ulcer with indurated and elevated margins should always raise the suspect of a malignancy, particularly if it shows an irregular shape with local bleeding, no obvious history of trauma, and associated enlarged neck lymph nodes. Chronic bacterial (e.g. tuberculosis or syphilis), fungal (e.g. histoplasmosis), or other infections (e.g. leishmaniasis) can occasionally give rise to similar clinical features, often as part of the systemic spreading of the disease. The diagnostic process of a patient presenting with a single persistent ulcer and clinical features suggestive of malignancy or chronic infection invariably requires a biopsy. Additional investigations are to be arranged on the basis of the relevant histopathology. For example, in case of oral squamous cell carcinoma, a computed tomography or magnetic resonance imaging scan for disease staging is required, followed by a discussion in a head and neck cancer multidisciplinary team meeting. Further details on oral cancer management can be found in Chapter 57. Patients with oral ulceration due to *Mycobacterium tuberculosis* or *Treponema pallidum* or other infections may need additional serological investigations (e.g. Venereal Disease Research Laboratory (VDRL) or rapid plasma reagin, Quantiferon-TB Gold test, and HIV antibodies), and imaging (e.g. chest radiography), and should be referred to a relevant specialist for further assessment and management.

Single acute or persistent ulcer without obvious clinical features of malignancy but with evidence of trauma

A traumatic cause for a single oral ulceration can be reasonably suspected through careful medical history and careful clinical examination. There is a wide range of possible intraoral traumatic causes, some of which may seem bizarre. Clinicians should enquire whether patients have experienced accidental self-induced bites during mastication, recent dental treatment, direct application of medication to the oral mucosa, and even orogenital sexual practices (e.g. trauma to the lingual frenulum during orogenital intercourse). Clinical examination should include the search for any broken teeth or dental appliances and an assessment of the topographic relationship between the ulcerated area and the possible traumatic factor. These patients can be initially managed by ensuring that the offending agent is removed or avoided (e.g. sharp broken teeth next to an ulcer should be repaired or extracted by a dentist; ill-fitting dentures should be removed) and an antiseptic mouthwash (e.g. chlorhexidine 0.2% aqueous solution) used for 1 or 2 weeks. However, the diagnosis should be reconsidered and further investigations (or a referral) arranged in cases of oral ulceration failing to heal within 2–3 weeks following the removal of the putative traumatic factor.

Multiple mouth ulcers

Multiple acute or recurrent oral ulcers, often of small diameter (<1 cm), and affecting different mucosal sites

Most individuals experiencing episodes of multiple acute or recurrent ulcers with a characteristic ovoid or round shape are otherwise well and likely to have RAS. Ulceration of RAS can be managed symptomatically with the aim of speeding the healing of ulcers and reducing pain. Although topical analgesics (such as benzydamine hydrochloride) can reduce mild painful symptoms, topical corticosteroids (e.g. 0.05% clobetasol propionate ointment, off label) are invariably needed to lessen the duration of ulceration and can be used by the patients as symptoms dictate. Haematinic deficiencies have been associated with RAS and it would be sensible to assess full blood cell count, red cell folate, iron, and vitamin B_{12} levels.

Aphthous-like oral ulceration can arise with a range of systemic disorders including Behçet's disease, gastrointestinal disorders (such as coeliac disease or inflammatory bowel disease), Sweet syndrome, inherited autoinflammatory syndromes (e.g. familial Mediterranean fever, periodic fever, aphthous stomatitis, pharyngitis and adenitis, Muckle–Wells syndrome), and immunodeficiency syndromes (such as HIV infection or neutropenias). It is therefore prudent to take a thorough medical history focusing on gastrointestinal, cutaneous, ocular, genital, and articular symptoms, as well as recurrent febrile episodes, especially in children and young individuals, and refer to the relevant specialist where required.

Multiple acute and recurrent clustered ulcers (<1 cm), often affecting a single unilateral mucosal site

Recurrent HSV lesions typically manifest as multiple clustered ulcers, often localized to a single unilateral mucosal site (e.g. one side of the hard palate or the tongue) or the lip. Ulceration of recurrent HSV is usually self-limiting, but affected people may experience considerable pain and have difficulty eating, speaking, and swallowing. Antiviral therapy (e.g. topical aciclovir for labial lesions or aciclovir tablets 200 mg five times daily for 5 days) can reduce the duration of symptoms and lesions. Of note, primary herpetic gingivostomatitis (primary HSV infection, most commonly seen in children) can be severe and cause fever, malaise, and lymphadenopathy in addition to widespread (not unilateral) oral ulceration and gingival swelling. Reactivation of varicella zoster virus (shingles) causing multiple unilateral clustered oral ulceration is uncommon, notably painful, and requires a higher dosage of antiviral therapy (aciclovir tablets 800 mg five times daily for at least 7 days). Atypical manifestations of recurrent HSV and varicella zoster virus lesions (e.g. bilateral, long-standing, and unresponsive to therapy) can be

seen in immunodeficient patients (e.g. those on corticosteroids, immunosuppressants, or biologics for rheumatological or dermatological conditions).

Multiple recurrent or chronic oral ulcers (often of diameter >1 cm)

Patients presenting with recurrent, chronic, and often progressive multifocal large and irregularly shaped oral ulcers are likely to be affected by a mucocutaneous disease (mainly lichen planus/lichenoid lesions, pemphigoid, erythema multiforme, or pemphigus). This is particularly true if the patient has other coexisting mucosal lesions (such as keratoses or scarring), or cutaneous, genital, or conjunctival abnormalities.

Patients with oral lichen planus typically present with white keratotic reticular, or ray-like striae in addition to areas of erosion or ulceration, and often bilaterally to the buccal mucosae, tongue, or gingivae. A suspected diagnosis of oral lichen planus indicates the need for lifelong surveillance due to the associated increased risk of oral squamous cell carcinoma development (1–3% overall prevalence). It would be prudent to arrange a baseline biopsy in all patients with possible lichen planus in order to assess the potential presence of oral epithelial dysplasia and stratify patients according to risk (e.g. 5% of individuals with mild dysplasia vs 25% of those with severe dysplasia would progress to oral squamous cell carcinoma).

Lichenoid lesions represent a group of well-defined disorders that mimic oral lichen planus clinically and histologically, and require additional diagnostic investigations. Examples include lichenoid lesions of discoid and systemic lupus erythematosus, lichenoid contact lesions, oral graft-versus-host disease, lichen sclerosus, and immune-related adverse effect of immunotherapy, which can be suspected in patients with relevant medical history and confirmed with further investigations (e.g. circulating antinuclear, double-stranded DNA and anti-Smith antibodies), patch tests for dental materials, or direct immunofluorescence.

In addition to oral mucosal ulceration, patients with mucous membrane pemphigoid or other immune-mediated subepithelial blistering disorders may have, or be at risk of, genital, ocular, and laryngeal blistering and ulceration. Conjunctival lesions of mucous membrane pemphigoid can lead to formation of synechiae, entropion, and loss of vision. Laryngeal lesions may cause stenosis requiring surgical intervention. Pemphigus vulgaris often gives rise to blistering/ulceration to the oral mucosa which, if untreated, is followed by progressive widespread blistering to other mucosal surfaces and the skin. If untreated, pemphigus vulgaris may be lethal. Diagnosis of both pemphigus and pemphigoid require demonstration of circulating autoantibodies (via indirect immunofluorescence and/or an enzyme-linked immunosorbent assay).

The clinical appearance of oral ulceration in this group is not diagnostic, with the possible exception of the keratotic striation of lichen planus/lichenoid lesions. Therefore, it would be sensible to refer these patients to an oral medicine specialist in order to establish the definitive diagnosis and start appropriate therapy. Topical and/or systemic corticosteroids and immunosuppressants represent the mainstay of therapy for the patients in this group and are usually provided by the oral medicine specialist.

Other less common inflammatory causes of multiple recurrent or persistent irregularly shaped oral ulcers include oral erythema multiforme, granulomatosis with polyangiitis, and orofacial granulomatosis, which would all benefit from a referral to an oral medicine specialist for further investigations and management.

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55

Laryngeal precancer and cancer

David Hamilton and Anna Thompson

Introduction

Laryngeal cancer affects 2400 new patients each year in the UK, many from deprived communities.[1] Its incidence increases in line with age with an incidence peak between the sixth and seventh decades of life.[1] Tobacco and alcohol use account for around 75% of cases of head and neck cancer (HNC). Their effects are not isolated but multiplicative.[2] Smoking alone has a clear dose–response relationship for the frequency, duration, and number of pack-years of cigarette smoking. When drinkers are compared to never-drinkers, people who take more than three drinks per day have twice the risk of pharyngeal and laryngeal cancer[3]; this effect is found to be more pronounced if drinking beers and spirits than if drinking wine.[4] A family history of HNC in first-degree relatives, especially siblings, also increases the risk of HNC.[5]

Risk factors

- Smoking.
- Alcohol.
- Age.
- Immunosuppression.

Assessment

History

All patients should have a comprehensive ear, nose, and throat (ENT) history. The key points specific to the head and neck to include in the history are:

- speed of onset of symptoms
- duration
- fever/rigors
- weight loss
- anorexia
- hoarse voice
- dysphagia
- odynophagia
- otalgia
- haemoptysis/bleeding.

It is vital that a full and comprehensive history is taken from every patient, including past medical and surgical history, drug history (including allergies), and social history, including details of living arrangements, employment, support networks, and accurate documentation of smoking and alcohol consumption.

Examination

All patients should have a complete ENT examination, which is set out in detail in Chapter 46. This should include neck examination of all nodal levels including the parotid and submandibular glands. In addition, flexible laryngoscopy is an essential part of this examination and the most important points to note are as follows:

- The patient's airway.
- Vocal cord mobility.
- Tumour position and size.
- Anterior commissure involvement.

If there are concerns about the patient's airway, these should be addressed as a priority (see Chapter 52).

If laryngeal cancer is suspected, a speech and language therapy assessment to determine swallow function, risk of aspiration, and voice quality, and to define patient preferences is important when making subsequent treatment decisions.

Investigation

If the patient's airway is safe and laryngeal cancer is suspected, cross-sectional imaging should take place prior to a biopsy as inflammation following biopsy can sometimes lead to inaccurate radiological staging. Most departments use computed tomography scanning of the neck to provide an accurate picture of tumour and nodal stage and a computed tomography scan of the chest and upper abdomen to provide a metastasis stage. An ultrasound scan of the neck to assess for nodes and cartilaginous involvement and extralaryngeal spread may contribute to this staging information and allows sampling of suspicious lymph nodes. Imaging modalities for HNC are set out in detail in Chapter 69. The AJCC staging is separate for supraglottic, glottic, and subglottic sites.

In particular, it is important to ascertain the following from the imaging:

- Involvement of more than one laryngeal subsite.
- Involvement of the anterior commissure.
- Involvement of the thyroid cartilage.
- Extralaryngeal extension.

The next stage of tumour assessment involves a microlaryngoscopy and biopsy of the tumour. Examination under anaesthetic of the entire upper aerodigestive tract with photographic documentation of the tumour should take place. This can be done under local anaesthetic in selected patients if required. This means that assessment of the tumour extent takes place in tandem with assessment for a synchronous primary tumour (present in 1–6% of cases). In a similar way to the flexible laryngoscopic examination, the most important parts of the assessment under anaesthetic are as follows:

- Involvement of the anterior commissure.
- Involvement of the contralateral vocal cord.
- Subglottic involvement.
- Involvement of the pyriform fossa.
- Involvement of other laryngeal subsites.

The assessment under anaesthetic can sometimes provide extra information about how well the patient copes with a general anaesthetic and pre-assessment information, which can help decision-making, especially in the advanced setting.

Management

Management of the airway

This is the primary concern in laryngeal cancer. Any potential airway issues need to be identified before definitive treatment is initiated to avoid the catastrophic situation of 'can't intubate, can't oxygenate' (see Chapter 52).

Airway assessment takes place at each stage of the diagnostic process. In particular, the clinician should watch for the following features when assessing patients:

- Stridor.
- Patient fatigue.
- Use of accessory muscles of respiration.

These findings should prompt an urgent management of the patient's airway. More subtle warning signs to look out for include the following:

- Inability to speak in full sentences.
- Elongation of the inspiratory phase of respiration.
- Limitation of exercise tolerance due to airway issues.

Decision-making around airway management should involve the patient, their supporters, the ENT clinical team, and the anaesthetic team. The clinician should consider the potential behaviour of the airway following induction of anaesthesia: if problems are anticipated, the surgical team should have all airway adjuncts at hand before commencing with anaesthesia. This should include a ventilating bronchoscope and tracheostomy tray, open and ready before induction begins. The surgical and anaesthetic team may consider that a tracheostomy under local anaesthetic is the safest option. Communication between the surgical and anaesthetic teams, together with detailed planning of the steps to be taken in the event of a 'can't incubate, can't oxygenate' situation, is paramount.

Box 55.1 NICE guidelines on treatment of early-stage squamous cell carcinoma of the larynx

- Offer transoral laser microsurgery to people with newly diagnosed T1a squamous cell carcinoma of the glottic larynx.
- Offer a choice of transoral laser microsurgery or radiotherapy to people with newly diagnosed T1b–T2 squamous cell carcinoma of the glottic larynx.
- Offer a choice of transoral surgery or radiotherapy to people with newly diagnosed T1–T2 squamous cell carcinoma of the supraglottic larynx.

Early disease

Early laryngeal cancer (stage 1/2) can be managed either surgically or with radiotherapy (for the UK National Institute for Health and Care Excellence (NICE) guidelines, see Box 55.1).[6] Lymph node involvement in early laryngeal cancer is extremely rare so treatment is usually limited to the larynx only. A Cochrane review found there to be no good-quality randomized controlled trials available comparing mortality, disease-free survival, quality of life, or voice outcomes for laser resection or radiotherapy in early laryngeal cancer.[7] Clinical practice has been informed by a number of case series and other non-randomized research[8–11] which suggest that survival is similar between the two treatments. There is clear uncertainty over the comparative survival and voice outcome between laser surgery and radiotherapy with overall survival ranging between 45% and 77% for T1/T2 tumours.[6] Although some have suggested a survival advantage for those treated surgically, this depends on radiotherapy dose and included patients are selected (observational studies) rather than randomized studies. Transoral laser is, however, considered to be more cost-effective.[12] Treatment pathways between the two options are also very different; radiotherapy typically involves daily outpatient treatment for anything between 3 and 7 weeks whereas laser surgery can be performed in 1 day. This difference in treatment process is one of the main barriers to recruitment to randomized trials and the most recent attempt to conduct a UK randomized controlled trial failed as a result.[13]

Surgical management of early laryngeal cancer

Laser resection is usually the preferred mode of surgical management (Figure 55.1). The patient is placed under general anaesthesia and the tumour is reassessed together with photographic documentation. All laser safety precautions should be observed. Most surgeons use the Steiner technique where the tumour is divided in half to accurately assess the depth and then removed in two pieces. The specimen is mounted for the pathologist and accurately orientated. Most surgeons then take additional margins in order to demonstrate complete resection.

Non-surgical management of early laryngeal cancer

See 'Principles of radiotherapy management of laryngeal cancer'.

Advanced disease

Locally advanced laryngeal cancer (stage T3 or T4) represents around 40% of laryngeal cancer cases.[14] Historically, patients would be treated with a total laryngectomy, but robust evidence has shown

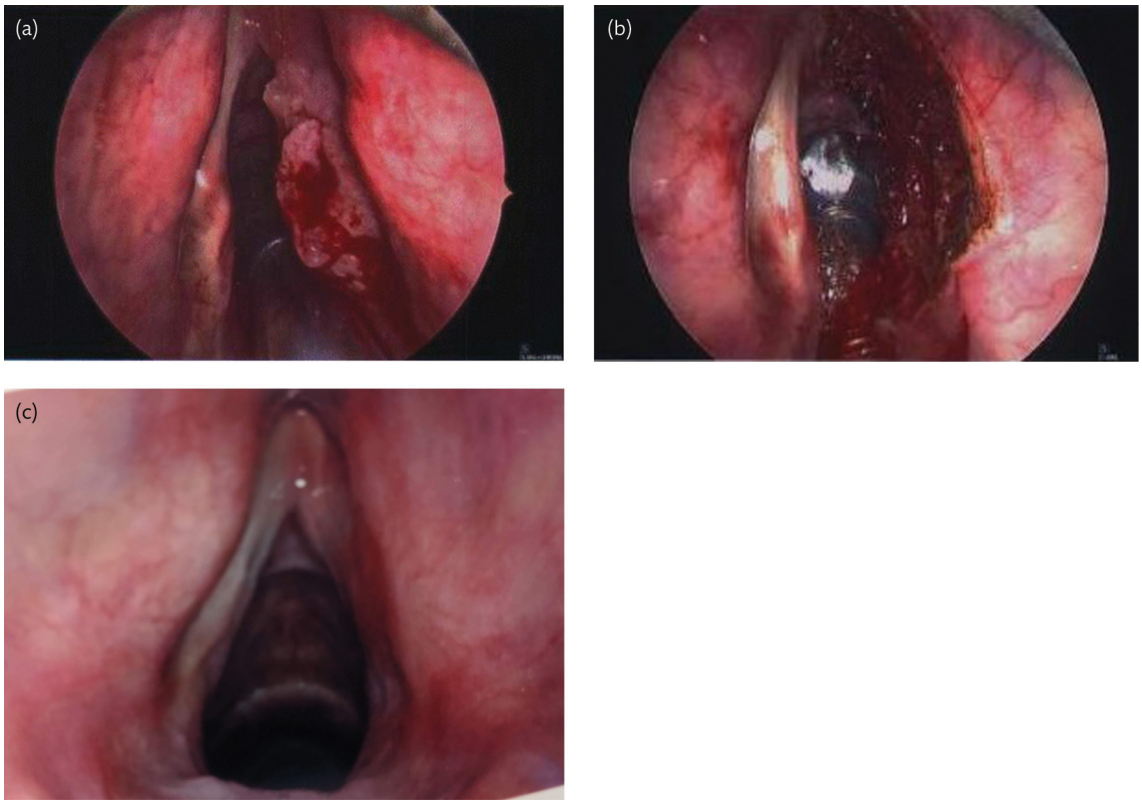

Figure 55.1 An image panel demonstrating transoral laser-assisted surgical resection of a vocal cord tumour and long-term outcome. Panel (a) shows an ulceroproliferative lesion on the right true vocal cord. In (b), a type 3 cordectomy has been performed which includes the thyroarytenoid muscle. Panel (c) demonstrates the cancer-free healed scar tissue in the place of the vocal cord 12 months down the line.

that larynx-preserving therapies such as radiotherapy with or without chemotherapy are valid alternatives. However, if laryngeal function is poor at diagnosis, patients may benefit from primary laryngectomy to gain a safe swallow and better voice quality with reconstruction. These treatment decisions need to be made with full multidisciplinary team input for the clinician and patient to make an informed choice. See the NICE guidelines for management of advanced squamous cell carcinoma of the larynx in **Box 55.2**.[6]

For people with T4a squamous cell carcinoma of the larynx, consider surgery with adjuvant radiotherapy, with or without concomitant chemotherapy.

Box 55.2 NICE guidelines for management of advanced squamous cell carcinoma of the larynx

Offer people with T3 squamous cell carcinoma of the larynx a choice of:

- radiotherapy with concomitant chemotherapy *or*
- surgery with adjuvant radiotherapy, with or without concomitant chemotherapy.

Discuss the following with people with T3 squamous cell carcinoma of the larynx and their carers, to inform their choice of treatment:

- The potential advantages of laryngeal preservation.
- The risk of needing salvage laryngectomy (and its associated complications).
- The benefits of primary surgery in people with existing compromised swallowing and airway function.
- Likely voice and swallowing function after treatment (including the need for a long-term feeding tube).

Decision-making in advanced laryngeal cancer

Over the past 35 years or so, two landmark trials[15,16] demonstrated that chemoradiotherapy achieved enhanced laryngeal preservation rates with equivalent locoregional tumour control to laryngectomy and have changed the management of advanced laryngeal cancer treatment substantially. However, since this change in practice survival has declined[17] and patients who fail to respond to chemoradiotherapy experience severely impaired function and quality of life. These patients face salvage surgery, resulting in a high risk of complications, a poorer overall treatment experience, and compromised survival.[18] The challenge, therefore, is to identify those patients who are likely to respond and for whom chemoradiotherapy is the most appropriate treatment and to avoid tri-modality treatment.

Recent research has now come to focus on quality of life as a treatment outcome[19,20] and even as a predictor of survival.[21] Treatment for advanced HNC has profound effects on a patient's quality of life, especially their voice and swallow function. Despite the obvious difference in treatment regimens, quality of life does not seem to be significantly different for patients treated surgically and non-surgically in early[22] or advanced disease[23,24]; however, these population-based data may obscure wide variations between individual patients. Quality of life is significantly reduced in those patients who have salvage surgery after the failure of non-surgical treatment.[18]

It is recommended that all cases are discussed in the head and neck multidisciplinary team given the importance of allied health professional input to decision-making for both the clinician and patients. However, many of the factors which influence decision-making in

advanced laryngeal cancer are related to the value that the patient places on functions such as swallow or voice. The values that staff members place on these attributes have been shown to vary widely and are often different to the patients that they treat. For this reason the decision process should place the values and preferences of the individual patient at their centre. Putting the values and preferences of the patient at the centre of a multidisciplinary decision-making process is profoundly challenging; there are currently no decision aids to use in the context of the head and neck multidisciplinary team and the decision-making process often makes true patient involvement problematic.

In general, the following features usually push the multidisciplinary team towards surgical management:

- Pre-existing laryngeal dysfunction, especially if leading to weight loss or requirement for tube feeding.
- Requirement for airway adjunct (e.g. tracheostomy).
- Advanced disease (T4).
- Patient preference.

Surgical management of advanced disease

Surgical management of advanced laryngeal cancer commonly involves a total laryngectomy or complete removal of the voice box. This is often combined with neck dissection of one or both sides of the neck. Following this treatment patients usually require postoperative radiotherapy with or without chemotherapy. Total laryngectomy results in a permanent tracheal stoma; this has obvious effects on appearance, but also voice production and swallowing. However, some large retrospective case series suggest a possible survival advantage for surgical patients.[25]

If the laryngeal cancer extends out with the larynx (i.e. into the pyriform fossa, supraglottis, or base of tongue) then reconstruction with a local or free flap is likely to be necessary. Practice varies around the country depending on local expertise. A pectoralis major flap is quick and safe and can be performed by many ENT surgeons; however, the cosmetic outcome is not as good and the flap is quite bulky. Free flaps from the radial forearm or anterolateral thigh are commonly used if pharyngeal mucosa needs to be resected.

Partial laryngectomy can be considered for removal of smaller tumours. This procedure allows the patient to maintain a normal swallow and the patient can continue to produce that voice in a normal way. A tracheostomy is often used as cover during the operation. Only a small number of tumours and patients are suitable for this procedure. The patient should be willing to engage with an often lengthy rehabilitation process.

Radiotherapy management of laryngeal cancer

To deliver external beam radiotherapy a number of factors must be considered: the patient must be able to lie flat for approximately 20 minutes without oxygen while wearing a customized radiotherapy planning mask which ensures accurate daily positioning of the patient. The patient will be alone in the treatment room during treatment delivery so they must be safe and able to follow instructions. They must be able to do this throughout treatment even as the side effects increase and must remain of adequate performance status to complete treatment. Radical treatment is commonly given daily (Monday–Friday) for between 3 and 7 weeks depending on the volume treated. Considerations prior to radiotherapy are detailed in **Box 55.3**.

Box 55.3 Considerations prior to radiotherapy

- Good performance status and capacity, able to follow instructions.
- Able to lie flat without oxygen for duration of daily radiotherapy treatment.
- Airway security.
- Swallow function and feeding tube placement if appropriate.
- Communication (if the patient has poor/no voice).
- Refer to dietician, speech and language therapy team, head and neck clinical nurse specialist, and palliative care team as appropriate.
- Dental assessment pretreatment if radiotherapy to mandible/maxilla is anticipated.
- Referral to smoking and alcohol cessation services as appropriate.
- Consider referral to clinical psychologist.

For early laryngeal cancer the volume treated with radiotherapy is likely to be relatively small; however, in more advanced cancers where the primary tumour is larger and/or lymph node levels are involved, the radiotherapy target volume will be significantly larger to include all sites of potential disease spread in the head and neck. Once a decision to treat has been made and the patient consented appropriately, they must undergo radiotherapy planning.

Radiotherapy planning involves identifying and delineating all areas of macroscopic disease and potential microscopic disease using diagnostic imaging, clinical examination, and operation notes including examination under anaesthetic findings. Organs at risk are also identified such as the spinal cord, brainstem, and salivary glands. A radiotherapy plan is created to deliver the prescribed dose to the target area(s) while minimizing dose to the organ at risk depending on their dose limits and priority (**Figure 55.2**). For example, the dose limit to the spinal cord or brainstem is never exceeded whereas the dose to the parotid gland may have to be compromised in order to treat an adjacent level 2 node, in which case the patient will be consented for xerostomia accordingly. For details, see published guidelines.[26]

Side effects may include increased soft tissue swelling of the target area so airway management is critical and must be considered before radiotherapy planning (e.g. prophylactic tracheostomy or a de-bulking procedure if necessary). Similarly, nutrition should be considered if swallow function is also expected to deteriorate during radiotherapy due to toxicity (e.g. placement of a nasogastric or gastrostomy tube). Voice function again is likely to worsen during radiotherapy so managing communication throughout is important. These side effects and others are listed in **Box 55.4**. The acute side effects build up throughout treatment, usually peaking 1–2 weeks after treatment completion and resolving over the following weeks/months depending on the dose and fractionation used.

Primary radical radiotherapy with or without chemotherapy for advanced disease

Radical radiotherapy is planned and delivered to the primary tumour and involved lymph nodes as previously described. In node-negative disease, lower-dose prophylactic radiotherapy is usually

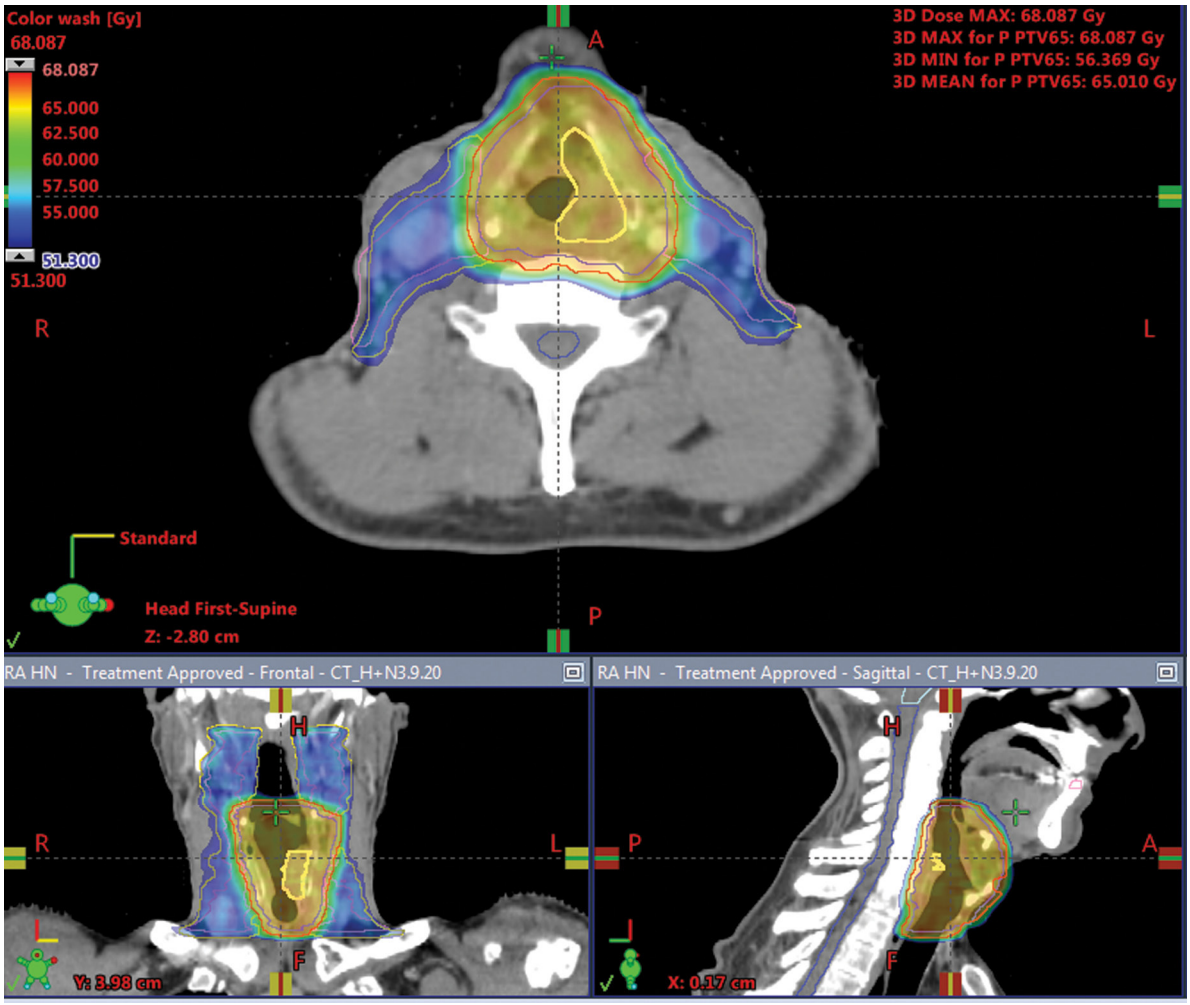

Figure 55.2 Cross-sectional imaging demonstrating intensity-modulated radiation therapy plan for a patient with laryngeal cancer. Note the highdose areas in yellow closer to the tumour receiving a radical dose and those areas in blue indicating a prophylactic dose to the neck.

Box 55.4 Common side effects of radiotherapy (severity and recovery time depend on dose/fractionation used)

Acute toxicity (weeks to months)

- Pain.
- Soft tissue swelling compromising swallow and airway.
- Hoarse voice.
- Skin breakdown.
- Dry mouth.
- Thick secretions.
- Loss of taste and appetite.
- Hair loss in treatment area.
- Tiredness.

Late toxicity (months to years)

- Dysphagia.
- Hoarse voice.
- Skin fibrosis and telangiectasia.
- Hair loss.
- Trismus.

Rare toxicity (years)

- Bone/cartilage necrosis.
- Second malignancy.

planned to the uninvolved draining lymph node levels, that is, levels 2–4 plus retropharyngeal nodes bilaterally. In node-positive disease, level 5 and the retrostyloid nodes are usually additionally included on the involved side(s). Prior to primary radiotherapy ensure airway and nutritional management has been addressed.

The addition of concurrent chemotherapy to radical radiotherapy using a radiosensitizer such as cisplatin is routinely recommended in patients with advanced disease and good performance status. The MACH-NC meta-analysis update[2] demonstrated a 5-year absolute survival benefit of 6.5% with concurrent chemotherapy which decreased in patients over 71 years old or performance status of 2 or greater. Contraindications to cisplatin include glomerular filtration rate less than 60 mL/min, grade 2+ hearing impairment, tinnitus, or peripheral neuropathy. As a radiosensitizer chemotherapy can worsen the radiotherapy toxicity in addition to the direct side effects of the agent. The side effects of cisplatin chemotherapy are shown in Box 55.5.

Postoperative radiotherapy with or without chemotherapy

Postoperative radiotherapy is recommended following primary surgery for locally advanced disease to increase the chance of cure. The

Box 55.5 Side effects of chemotherapy

- Nausea and vomiting.
- Taste changes and loss of appetite.
- Neutropenia and risk of severe infection.
- Anaemia.
- Tiredness.
- Thrombocytopenia.
- Bruising or bleeding.
- Renal impairment.
- Hearing loss and tinnitus.
- Peripheral neuropathy.

indications are listed in Box 55.6.[28,29] In this setting the radiotherapy is used to target the operative bed and adjacent structures with the highest doses planned to areas at risk of residual macroscopic or microscopic disease. Prophylactic radiotherapy can be delivered to areas not included in the surgery such as the retropharyngeal and retrostyloid lymph nodes.

Two randomized controlled trials[30,31] have demonstrated that the addition of concurrent chemotherapy increases local control and survival in high-risk patients with positive resection margins and extracapsular nodal extension. For a complete indication list see Box 55.7.[15]

Induction chemotherapy

A number of trials have looked at using induction chemotherapy as a test of tumour sensitivity in locally advanced laryngeal cancer with responders going on to have radiation treatment rather than laryngectomy with a laryngeal preservation rate of around 60–70%.[32] However, in the RTOG 91-11 trial laryngeal preservation was superior in patients treated with chemoradiation compared to induction chemotherapy and radiotherapy. The MACH-NC meta-analysis[27] found concurrent chemotherapy plus radiotherapy superior to induction chemotherapy plus radiotherapy.[16] In practice, induction chemotherapy is typically used before radical (chemo) radiation in fit patients with rapidly progressing or bulky neck disease to reduce the tumour bulk and so avoid significant volume changes and re-planning during radiotherapy. Pre-chemotherapy imaging is used to delineate the radiotherapy treatment volumes as these tissues are considered involved even if there is a radiological response.

Box 55.6 Indications for postoperative radiotherapy

- Positive resection margins (<1–2 mm).
- Extracapsular spread in lymph node metastases.

Or any two of the following:

- Close margins (<5 mm).
- Invasion of soft tissues such as skeletal muscle.
- At least two lymph nodes positive for metastatic spread.
- One lymph node region positive for metastatic spread.
- Involved node greater than 3 cm in size (≥N2a disease).
- Multicentric primary.
- Perineural invasion.

Less important factors to consider

- Lymphovascular invasion.
- Poorly differentiated (grade 3) disease.
- T3–T4 disease.
- Carcinoma *in situ* or dysplasia at the resection margins where further surgery would be difficult.
- In cases where there are uncertainties regarding the surgical or histopathological findings.

Box 55.7 Indications for postoperative chemoradiotherapy

- Incomplete resections.
- Extracapsular spread in lymph node metastases.

Consider if:

- soft tissue invasion
- advanced stage
- extensive perineural or lymphovascular invasion.

Rehabilitation and follow-up

Close clinical surveillance to monitor side effects and response to treatment is important. As radiotherapy-related oedema and tumour response can take a number of months to complete, cross-sectional imaging is usually done 3 months after treatment completion as a baseline scan. NICE guidelines recommend consideration of fluorodeoxyglucose positron emission tomography–computed tomography 3–6 months after chemoradiation to guide management in those people who have a laryngeal primary site with one or more positive nodes in the neck. NICE does not recommend neck dissection to people with no abnormal fluorodeoxyglucose uptake or residual soft tissue mass on this scan. Regular review of side effects and response to treatment where appropriate is essential and relies heavily on the support of our allied healthcare professionals to monitor airway, swallow, and nutritional status plus symptom control. All multidisciplinary teams should have input from speech and language therapists, dieticians, restorative dentists, clinical psychologists, and clinical nurse specialists who are central to the rehabilitation of the patient after treatment for laryngeal cancer. Regular follow-up is recommended for those patients most at risk of tumour recurrence or side effects from treatment.

Palliation

In some circumstances tumour or patient factors may lead to the clinical team discussing palliative options. Palliative decision-making in laryngeal cancer is fraught with difficulty as the patient is often likely to need a tracheostomy with or without a feeding tube. This clearly has a hugely detrimental effect on their quality of life; however, without a breathing tube the mode of death can be particularly unpleasant. Also, there are very few factors clinicians can use to predict how a patient is going to respond to treatment and therefore choosing which patients to palliate can be extremely challenging. This leads to variation in palliative practice throughout the country. Palliative radiotherapy and/or systemic therapy (chemotherapy, biological agents, or immunotherapy) may be considered to improve symptoms and may prolong life but the side effect profiles of these agents may also affect quality of life so must be discussed fully with the patient. Early involvement of the palliative care team and discussions around end-of-life airway management and feeding are

paramount. Clinicians should make every effort to involve patients and their carers in this decision.

Laryngeal dysplasia

Laryngeal dysplasia is a premalignant condition. Between 11% and 25% of cases will develop into a squamous cell carcinoma[33]: laryngeal dysplasia is divided into low, moderate, and severe.

When managing dysplasia, patients should be encouraged to stop smoking. Management depends on whether the dysplasia is focal or widespread. Focal lesions of any grade should be excised with a laser and the patient kept under follow-up. More widespread mild or moderate grade dysplasia can be kept under regular review. There is variation in follow-up practice for these patients: safety net information about voice changes should be given to patients and follow-up should continue for at least 5 years.

Sometimes radiotherapy is considered for severe dysplasia, especially in those with widespread disease, radiological suspicion of invasive disease, high anaesthetic risk, multiple recurrences, or due to patient preference. However, radiotherapy is usually reserved for lesions classified as carcinoma *in situ* and invasive cancer.

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56

Oropharyngeal cancer

Zaid Awad and Dorothy M. Gujral

Introduction

Squamous cell carcinoma is the commonest type of malignancy in the oropharynx and forms the focus of this chapter. The reader should be aware that several other malignant lesions such as minor salivary gland carcinomas and lymphomas can also occur at this site. Smoking and excessive alcohol intake continue to be important risk factors; however, the rising incidence of human papillomavirus (HPV)-related oropharyngeal squamous cell cancer (OPSCC) has been well documented. Unlike other parts of the upper aerodigestive tract, the incidence of OPSCC continues to increase, with approximately 93,000 new cases worldwide in 2018 reported by the World Health Organization's International Agency for Research on Cancer.

Risk factors and the role of human papillomavirus

HPV infection is linked to OPSCC and is a significant prognostic factor for survival.[1] HPV-16 and/or HPV-18 are detected in the majority of cases of HPV-positive OPSCC. The p16 protein, a surrogate marker for HPV infection, is overexpressed due to the negative feedback loop for p16 expression being abrogated by the HPV E7 oncoprotein binding to hypophosphorylated retinoblastoma protein (pRb).[2] In the presence of p16 overexpression, treatment response rates exceed 80% and are associated with improved survival.[3]

The HPV-positive OPSCC patient is generally younger (between 40 and 60 years), male (8:1 male:female ratio), with no or minimal smoking history, predominantly Caucasian, and more affluent, unlike the traditional head and neck cancer patient (advanced age, history of substance abuse, HPV-negative). Tobacco use has a negative impact on prognosis and is independent of HPV status. While the rate of locoregional failure in HPV-positive OPSCC is lower, the rates of distant metastases, most commonly to the lung, are similar to HPV-negative OPSCC.

HPV-positive OPSCC is now considered a distinct biological and clinical entity. Consequently, the staging of these tumours has been amended in the current eighth edition of the TNM staging system to reflect disease severity and prognosis. However, it must be noted that these stage groupings reflect prognosis, and treatment guidelines do not recommend modifications based on HPV status.

Anatomy

The oropharynx comprises the palatine (faucial) tonsils, the tongue base or posterior third of the tongue, the soft palate including the uvula, the lingual surface of the epiglottis, and the corresponding posterior and lateral pharyngeal walls between the nasopharynx and hypopharynx (**Figure 56.1**).

Arterial supply is from branches of the external carotid artery via terminal branches of the facial, lingual, ascending pharyngeal, and internal maxillary arteries (**Figure 56.1**b).

Venous drainage is through tributaries of the lingual and ranine veins as well as the pharyngeal venous plexus. The main sensory nerve of the oropharynx is the glossopharyngeal nerve. The vagus nerve supplies the lingual surface of the epiglottis, reflecting its different embryological origin (**Figure 56.1**c).

Lymphatic drainage of the oropharynx is mainly through the deep jugular chain, with the echelon nodes located in the ipsilateral upper deep cervical nodes, specifically level IIa. Retropharyngeal, intraparotid, submandibular triangle nodes, and contralateral level II nodes are also occasionally the first stop for metastatic OPSCC. The palatine tonsils have less contralateral drainage than the tongue base and soft palate and this can affect treatment choice and strategy.

Symptoms

Symptoms vary a great deal: while some patients present late with large neck lumps, dysphagia, and aspiration pneumonia due to laryngopharyngeal insufficiency, others present very early with small, indolent neck nodes or may even be asymptomatic, with the disease identified incidentally during a diagnostic tonsillectomy for asymmetrical enlargement. In addition, the wider availability of ultrasound scans, magnetic resonance imaging, and positron emission tomography (PET)/computed tomography (CT) scans has also led to early identification of some asymptomatic, incidental cases of OPSCC. Other symptoms include persistent throat pain, referred

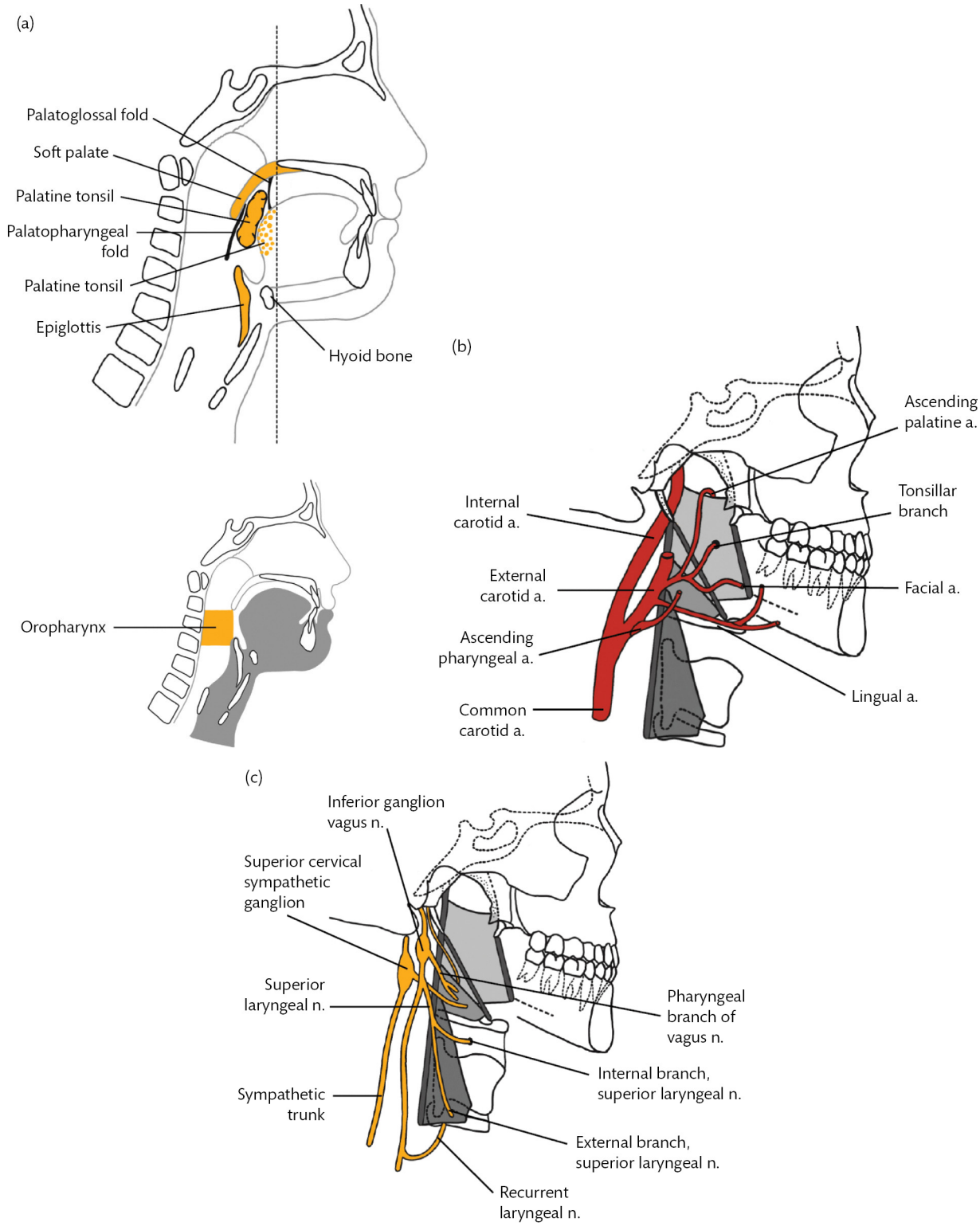

Figure 56.1 (a) The oropharynx. (b) Blood supply to the pharynx. (c) Contributions to the pharyngeal plexus.
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otalgia, dysgeusia, and voice change alongside other general symptoms of malignancy such as unintentional weight loss and lethargy.

For the approach to and assessment of patients presenting with these head and neck symptoms, please refer to the respective chapter within Part 4 of this textbook.

Assessment and investigations

These should start with general blood tests, but ultrasound assessment and guided fine needle aspiration cytology is the first step for patients with neck lumps and suspected OPSCC, as described

in Chapter 50. Aspirates should be tested for the p16 marker. Core biopsy obtained by skilled radiologists can also aid in obtaining histological confirmation of HPV-related disease. A high index of suspicion should guide the investigations of any patient over the age of 40 with cystic cervical nodes, considering as malignant unless proven otherwise.

Cross-sectional imaging allows for radiological staging and the consensus in the UK is to obtain at least a high-resolution contrast CT scan of the head and neck, but many units add magnetic resonance imaging of the neck which can provide better detail of the palatine tonsils and tongue base.[4] PET superimposed on CT scan images is becoming increasingly popular as a tool that allows the detection of small OPSCCs as well as identifying distant metastasis and synchronous tumours.

Early dental assessment using an orthopantomogram and clinical examination by an oral surgeon is needed as part of planning for future radiotherapy treatment.

Necrotic malignant neck nodes can pose a particular challenge in diagnosis as central samples may only show dead cells and cross-sectional imaging findings may mimic those of branchial cleft cysts or chronic infections. PET/CT scanning would only show a rim of enhancement which is not specific to malignant pathology. If an open biopsy is required to achieve a diagnosis, this needs to be carried out using a compartmental approach by experienced head and neck surgeons. A selective neck dissection is preferred to single node sampling. Surgical incisions can be planned in a way that allows them to be extended if further surgery is needed at a later date.

Examination under anaesthesia and structured upper aerodigestive tract endoscopy and biopsies processed as per the Royal College of Pathologists guidelines[5] form the final step in achieving a full assessment of OPSCC. Tumours should be described by site, size, and extension into and involvement of anatomical landmarks, and carefully palpated for mobility before biopsies are taken. Other important findings to document include mouth opening and accessibility should surgery be considered as a treatment option. Finally, this may be the best time to carry out any required dental extractions to reduce the risk of osteoradionecrosis.

Staging

The eighth edition of the American Joint Committee on Cancer (AJCC) staging system separates oropharyngeal cancer staging into two entities based on p16 expression. The new system also introduces a different pathological staging based on resective pathology and the overall group staging reflects the better prognosis of the p16-positive group. Detailed information regarding the latest AJCC staging system is available from the eighth edition of the *AJCC Cancer Staging Manual*.[6]

Management

All cancer patients should be managed in a multidisciplinary setting as discussed in Chapter 46.

Treatment options for OPSCC may be largely divided into radiotherapy, systemic chemotherapy, and surgical resection. Depending on patient factors, stage, and presence and extent of adverse features, definitive treatment may include a combination of modalities.

Principles of treatment

Overriding clinical principles are to maximize disease control while minimizing morbidity.

There is broad consensus that transoral surgery with neck dissection as single modality treatment is appropriate for T1–T2 OPSSC with no nodes or a single metastatic node, where appropriate margins have been achieved and there are no adverse prognostic factors in the resective pathology. If this option is to be considered, it is important to highlight the risks and benefits of this approach to patients, as well as the possibility of potential escalation of treatment should high-risk features exist. In larger tumours, and with more extensive lymph nodal involvement, primary non-surgical treatment is the standard of care. Surgical resection with clear margins and no adverse features can avoid the use of concurrent chemotherapy and reduce radiation dose from radical to adjuvant dose. Such reduction has the potential to reduce treatment toxicity but should be offered within the context of the ongoing trials and multidisciplinary team supervision. There is also broad agreement that triple modality therapy should be avoided, especially in early-stage tumours, thus emphasizing the importance of case selection for surgical treatment.

To date, no high-level evidence exists to support an upfront surgical approach with adjuvant treatment versus a primary non-surgical approach with surgery reserved for salvage. Transoral resection followed by adjuvant treatment is comparable to non-surgical management in terms of local control, regional control, and survival.[7] Therefore, the focus on treatment-related morbidity and functional outcome should drive the treatment approach. Based on current guidelines, patients will require chemotherapy in addition to postoperative radiation if extranodal extension or positive margins are found on the resective pathology. Patients with evidence of gross extranodal extension on pretreatment imaging should be treated with primary chemoradiation unless triple modality treatment is planned from the outset such (e.g. N3b deposits).

Several ongoing studies are investigating the efficacy of reduced dose of radiation, with or without surgery in this low-risk cohort. While some have reported results, there is no consensus on de-escalation as these are primarily phase II studies.[8–10]

Radiation therapy

Intensity-modulated radiation therapy is a highly conformal radiation technique that has been instrumental in preserving excellent control rates while reducing treatment-related toxicity to the salivary glands and pharyngeal structures.[11]

Unilateral neck radiation is utilized for lateralized tonsil cancer (**Figure 56.2**), and bilateral neck radiation for bulky, non-lateralized tonsil cancer and tongue base primaries (**Figures 56.3 and 56.4**). The role of unilateral neck radiation in the context of lateralized tongue base cancer is currently debated and should only be undertaken in the context of a clinical trial.

While primary radiation is commonly utilized concurrently with chemotherapy, radiation monotherapy is an effective modality for early-stage OPSCC (T1–T2, N0).

Radiation doses

The standard radical dose is 65–70 Gy, delivered in 30–35 fractions over 6–7 weeks to the macroscopic disease, referred to as the gross tumour volume. A margin is added to account for microscopic spread,

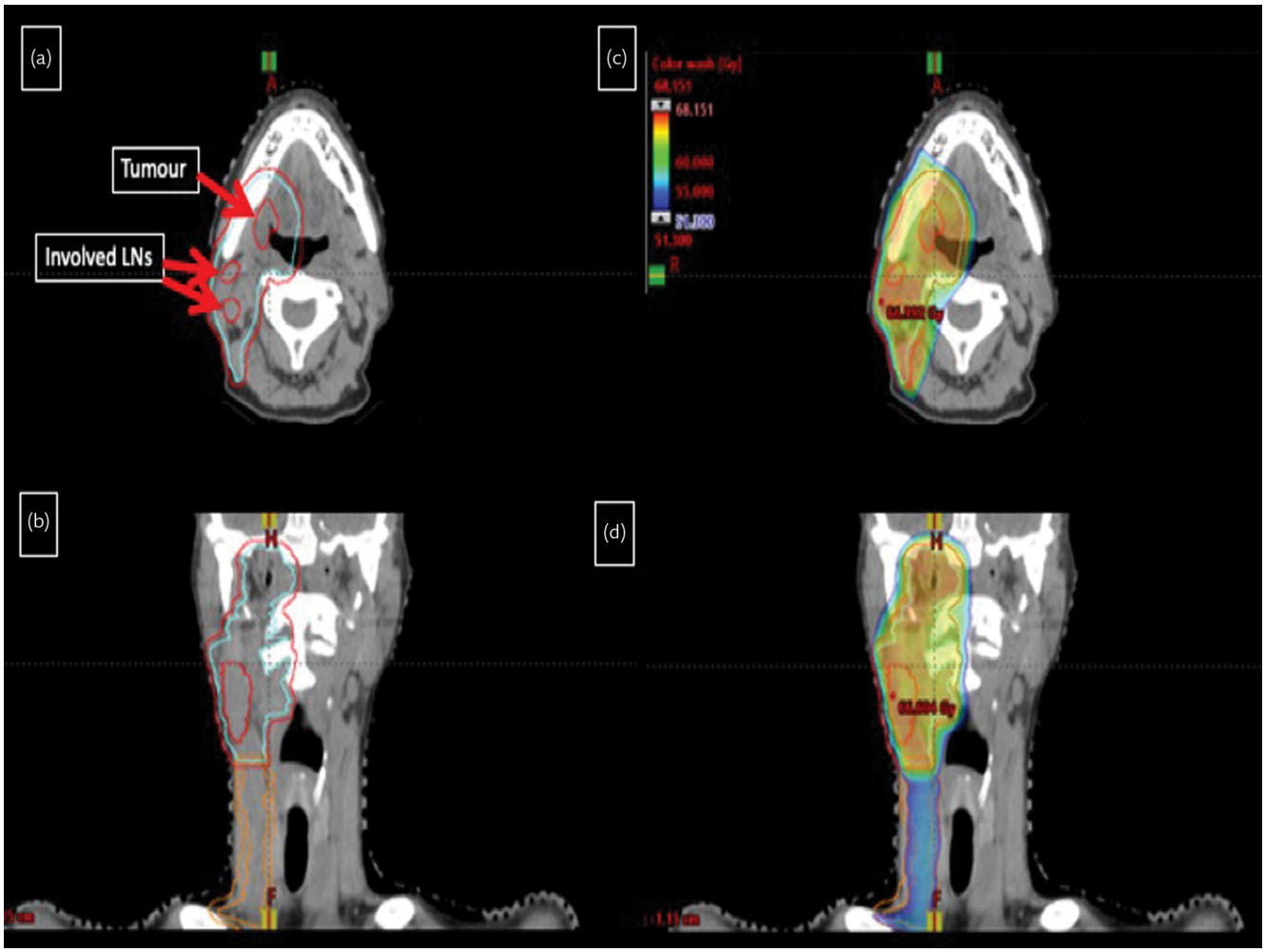

Figure 56.2 (a) Axial and (b) coronal CT slices demonstrating right tonsil cancer and involved level IIb lymph nodes (LNs; outlined in red), with margins to account for microscopic disease and uncertainties in daily set-up. Radiation dose distribution (in dose colour wash) in (c) axial and (d) coronal planes, with dose limited to the ipsilateral oropharynx and neck.

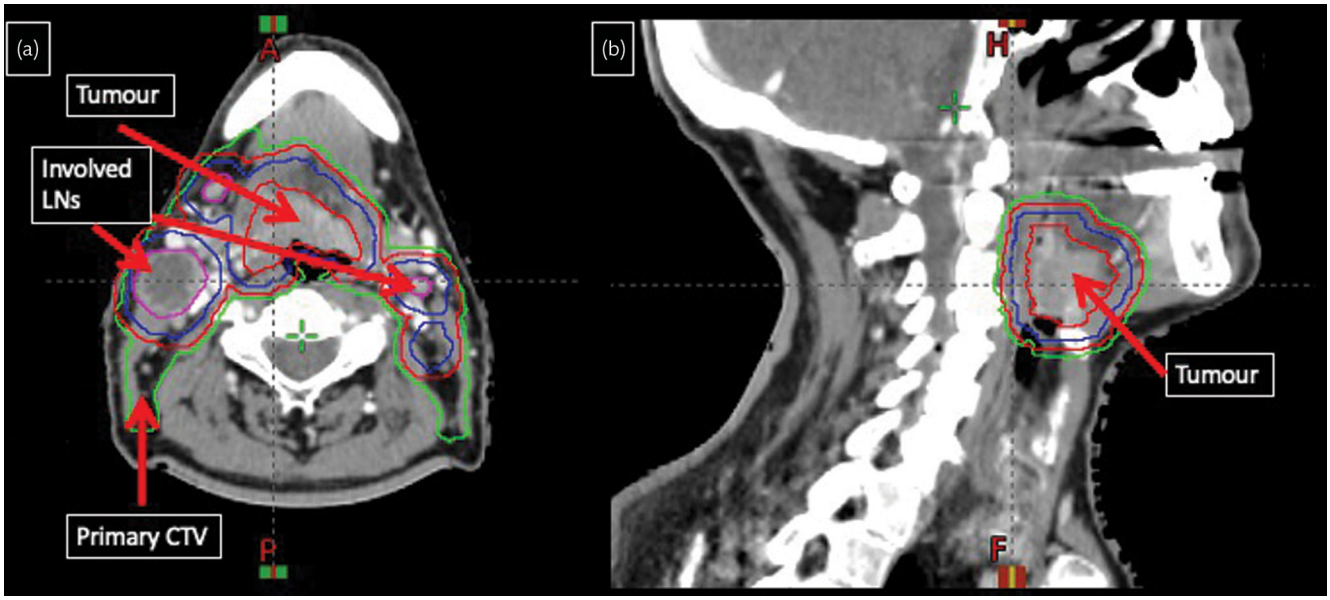

Figure 56.3 (a) Axial and (b) sagittal CT slices demonstrating right tongue base cancer crossing midline and involved bilateral level II lymph nodes (LNs; outlined in red), with margins to account for microscopic disease and uncertainties in daily set-up. CTV, clinical target volume.

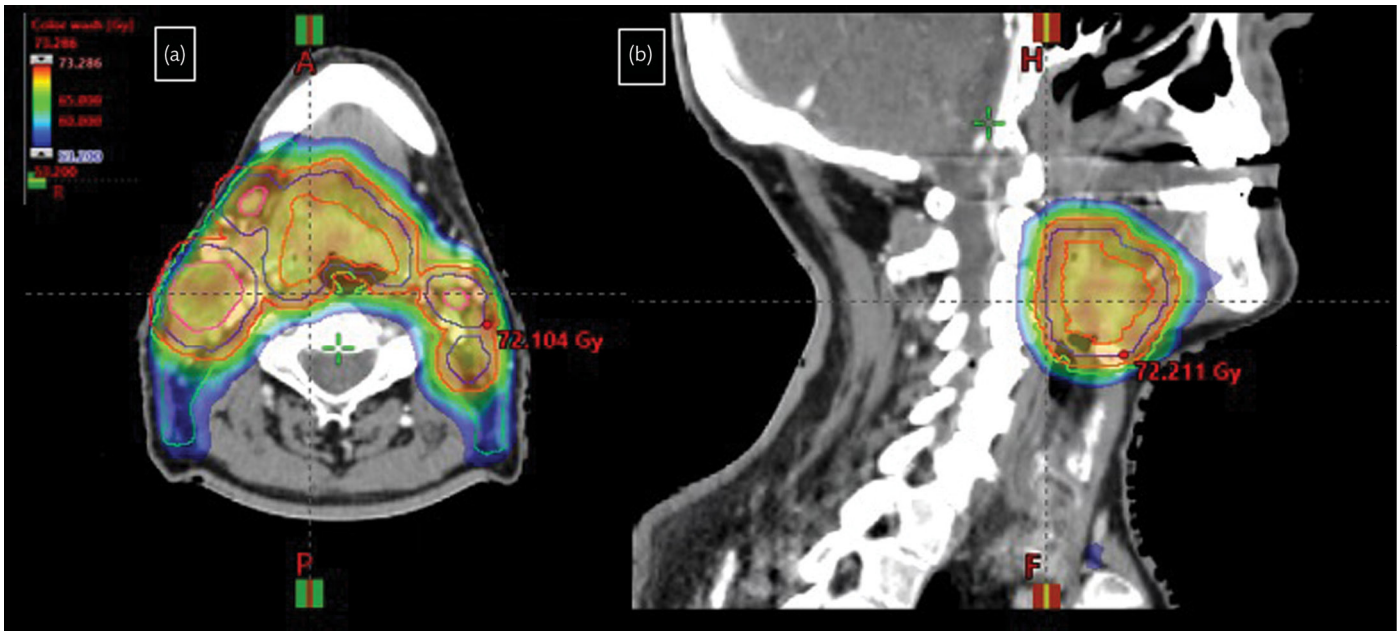

Figure 56.4 Radiation dose distribution (in dose colour wash) in (a) axial and (b) sagittal planes, with dose to the whole oropharynx and bilateral neck.

for daily set-up uncertainties, called the planning target volume (Table 56.1). Areas at risk of microscopic disease, such as postoperative tumour bed and postoperative neck, receive 60 Gy in 30 fractions over 6 weeks; 54 Gy in 30 fractions over 6 weeks is the prescription for areas requiring elective nodal radiation representing uninvolved neck nodal levels with a greater than 10% risk of microscopic disease.

Systemic therapy

High-dose cisplatin is considered the standard agent for concurrent therapy. Concurrent cetuximab (a monoclonal antibody which targets epidermal growth factor) incurs a 10% survival benefit at 3 years in advanced-stage head and neck squamous cell carcinoma compared to radiation alone.[12] However, more recent data comparing cetuximab to cisplatin in combination with radiation as a de-escalation strategy demonstrated similar rates of acute and late toxicity but inferior survival with cetuximab.[13,14]

Concurrent chemoradiation increases acute and late toxicities compared to radiation alone. Chemotherapy is also used with postoperative radiation in patients with high-risk adverse features such as extranodal extension and positive margins, offering improved disease-free survival and locoregional control,[15,16] but is associated with an increased mortality of 3–4%.[17] Potential complications of chemotherapy and radiotherapy are listed in Box 56.1.

Surgery

The role of surgery in the management of OPSCC has evolved significantly in recent years. In general, surgical treatment, when possible, has the benefit of shorter delivery time and less long-term morbidity. Transoral surgical approaches are used for lower T-stage tumours. Open surgical approaches are used for advanced tumours with the transcervical, transmandibular access to the oropharynx being the most common option. As phase III trials have demonstrated efficacy

Table 56.1 Tumour and radiation doses

Tumour variable	Radiation dose and fractions	Radiation volume
Stage I, II	65–70 Gy delivered in 30–35 fractions	Primary and ipsilateral neck[a] for tonsil cancers, bilateral neck for tongue base cancers, and those at or within 1 cm of the midline
Stage III, IVa, IVB	65–70 Gy delivered in 30–35 fractions with concurrent chemotherapy[b]; 54 Gy in 30 fractions	Primary and bilateral neck (except elective nodal regions) For elective nodal regions
Postoperative adjuvant treatment (close margin, multiple positive lymph nodes, perineural invasion, lymphovascular invasion)	60 Gy in 30 fractions over 6 weeks	Based on resective pathology
Postoperative adjuvant treatment (gross residual disease, positive margin, extranodal extension)	60–70 Gy in 30–35 fractions over 6 weeks with concurrent chemotherapy[b]	Based on resective pathology

[a] To include regional nodes (ipsilateral or bilateral levels Ib to V) and retropharyngeal lymph nodes.

[b] Cisplatin 100 mg/m^2 every 3 weeks × 2–3 cycles is the standard of care. Carboplatin or cetuximab can be considered in patients not suitable for cisplatin.

Box 56.1 Potential complications of chemotherapy and radiotherapy

- Skin erythema.
- Mucositis.
- Dry mouth.
- Loss of taste.
- Dysphagia.
- Odynophagia.
- Weight loss.
- Osteoradionecrosis.
- Ototoxicity and otitis media with effusion.
- Nephrotoxicity.
- Secondary cancers.

for a chemoradiation approach, they remain the treatment of choice for advanced OPSCC.[18]

Transoral surgery

Transoral laser microsurgery and transoral robotic surgery (TORS) are two minimally invasive modalities used in OPSCC management. The aim of transoral surgery is to avoid the morbidity associated with open surgery and the toxicity of (chemo)radiotherapy without compromising oncological outcomes, and yet deliver better functional outcomes, especially related to swallowing and dry mouth, which have the most impact on quality of life. The carbon dioxide laser, which is an established modality for laryngeal cancer, is a precision tool for surgery, but has poor haemostatic ability. The main disadvantage of transoral laser microsurgery is the restriction to laser delivery along the line of sight, and the need for mosaic resections that can make margin interpretation difficult. Flexible lasers may abrogate this problem, but long-term outcomes and high-level evidence are lacking.[19]

TORS is now the mainstream option for transoral surgery. The optics and the manoeuvrability of the robotic arms do not require the tumour to be in the 'line of sight', yet they provide a high-definition, close-up view of tumours and an en-bloc resection. While level I evidence comparing TORS to radiation therapy is lacking, many centres have reported excellent treatment outcomes of early OPSCC using TORS. Figure 56.5 shows a standard resection outline in early tonsillar tumours.

Irrespective of the tool used, it is essential to select patients carefully for transoral surgery as well as to inform them of the risks and benefits of this approach. Achieving clear margins is paramount to avoid escalation of treatment in an otherwise good prognostic group. Concurrent or staged neck dissection is also performed for histological neck staging, vessel ligation, and, in some cases, for free flap anastomosis. If bilateral neck dissection is carried out, especially for tumours involving the base of tongue, a prophylactic temporary surgical tracheostomy may be needed. Contralateral diagnostic tonsillectomy may be needed either in select cases, at the time of examination under anaesthesia, or during definitive surgery (Figure 56.6), as well as a thorough examination of the remainder of the upper aerodigestive tract to exclude a second malignancy.[20]

Open surgery

Open surgery for the treatment of primary cancer is not widely espoused, although large database analyses have shown a benefit for primary surgical treatment in larger HPV-negative cancers. Transcervical, transmandibular access to the oropharynx is the most commonly used approach; lingual release and lateral pharyngotomy approaches do not involve median mandibulotomy, but all open

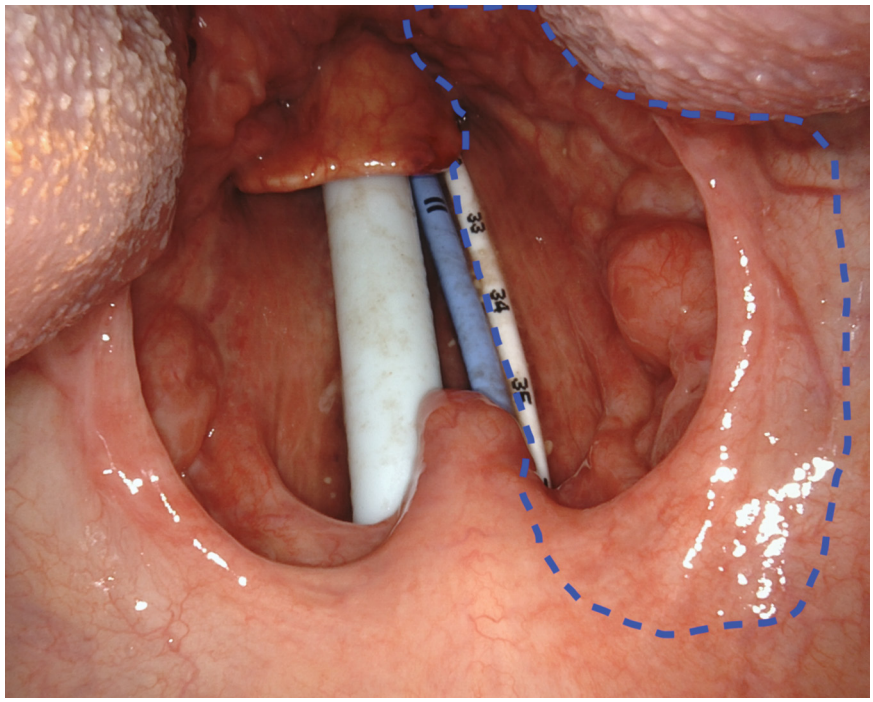

Figure 56.5 Endoscopic view of the oropharynx showing planned resection outline for an early, right tonsil OPSCC. Note the endotracheal tube, nasogastric tube, and nasopharyngeal temperature probe can be moved to access the posterior pharyngeal wall and achieve clear gross margins.

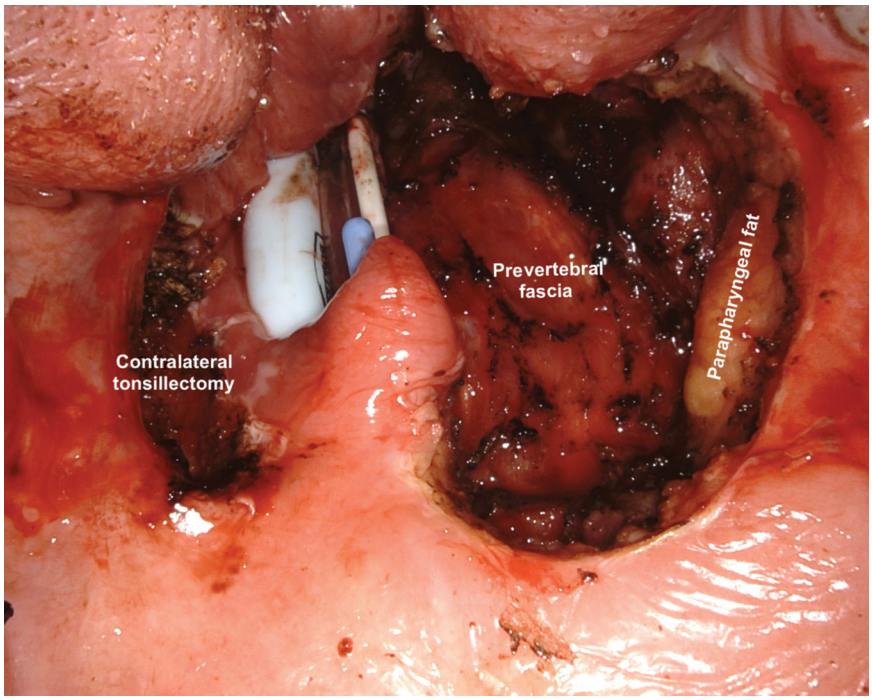

Figure 56.6 After right hemi-oropharyngectomy and diagnostic contralateral tonsillectomy.

approaches are associated with prolonged hospital stay, the need of a flap for reconstruction, pharyngocutaneous fistula, and functional impairment.

Salvage surgery

Tumours that do not respond to or recur after primary treatment may be considered for salvage surgery as a curative option or for locoregional control, irrespective of HPV status. Compared to primary cancers, outcomes are poor, both from an oncological and a functional perspective.

Before undertaking salvage surgery, it is essential to assess the extent of locoregional tumour and exclude distant metastasis (see Chapter 69). If the disease is limited to the neck with no disease at the primary site, a salvage neck dissection is sufficient. The salvage surgical field is usually scarred with poor blood supply, and a vascularized flap will aid in healing and expedite discharge from hospital.[21]

Open surgery is the commonest approach to residual or recurrent OPSCC at the primary site. TORS for recurrent OPSCC, even with free flap reconstruction is feasible in selected cases with good short- and long-term outcomes comparable to open surgery.[22] Potential complications of surgery are listed in Box 56.2.

Box 56.2 Complications of surgery

- Bleeding, haematoma, life-threatening primary, and delayed or secondary haemorrhage.
- Persistent pain, paraesthesia, and altered or loss of taste.
- Dental injury, trismus, and dental misalignment in mandibulotomy approach.
- Persistent dysphagia, dysarthria, and aspiration pneumonia.
- Osteoradionecrosis in salvage cases.
- Neurovascular injury, most commonly to the spinal accessory nerve causing shoulder dysfunction, as well as injury to the hypoglossal, lingual, vagus and phrenic nerves; chyle leak; and internal jugular vein injury, causing swelling and lymphoedema.
- Risks of general anaesthesia such as chest infection, deep venous thrombosis, pulmonary embolism, and cardiovascular events.
- Tracheostomy-related complications if needed: mucous plugging, bleeding, and persistent tracheocutaneous fistulae.

Palliation for advanced and metastatic disease

Platinum-based combination chemotherapy (5-fluorouracil +/– cetuximab) is the standard of care for patients with recurrent or metastatic head and neck squamous cell carcinomas who are platinum-naïve or who received platinum chemotherapy more than 6 months previously.

Immunotherapy agents utilize the immune system to fight cancer cells instead of targeting the cancer cell specifically. These are antibodies that block specific immune checkpoint compounds (e.g. programmed cell death protein 1 (PD-1), and its ligand PD-L1).

For patients who have received platinum chemotherapy within 6 months, immunotherapy treatment with nivolumab (a PD-1 inhibitor) is appropriate. Other agents (e.g. docetaxel) can be considered in those who progress after nivolumab. Recent data support the use of upfront PD-L1 inhibitor (pembrolizumab)[23] either in combination with platinum-based chemotherapy or as monotherapy in the first-line setting, with less toxicity and improved overall survival compared to standard platinum-based chemotherapy.

Palliative surgery is rarely considered for patients with metastatic OPSCC. It is seldom offered to patients with localized fungating neck masses to aid nursing of this area and avoid major bleeding, although risks and complication rates are high. Regional flaps such as the pectorals major muscle flap or deltopectoral flaps are used to cover major vessels. External carotid ligation or embolization can be considered in select cases with high risk of life-threatening bleed.

Segmental liver resections and lung lobectomy for oligometastatic disease are both rarely used as most patients have multiple metastatic disease and poor expected outcomes. For patients with small-volume recurrent disease, palliative re-irradiation can be considered if salvage surgery is not an option, particularly if the patient received radiation to that area more than 5 years previously.

Prognosis

Local control rates for tonsil tumours are dependent on T stage (T1, 85%; T2, 75%; T3, 70%; T4, 45%). Overall, 5-year overall survival for early-stage OPSCC is 84%, 66% for locoregionally advanced OPSCC, and 39% for patients with metastatic disease at presentation. Survival is higher for HPV-positive tumours at each stage.

The de-escalation controversy for human papillomavirus-positive oropharyngeal squamous cell cancer

Considering the good prognosis of HPV-positive disease, novel treatment strategies to minimize toxicity while maintaining survival outcomes have been proposed in these cancers: (1) reduction in radiation, and (2) transoral surgery and deintensification of adjuvant treatment.

Reduction in radiation

Using intensity-modulated radiation therapy, deintensification radiotherapy strategies could result in toxicity reduction without compromising survival outcomes.[24,25] Several studies have reported comparable clinical outcomes in patients with HPV-positive head and neck cancer treated with definitive radiotherapy alone instead of standard chemoradiotherapy.[26–28] Other radiation dose de-escalation strategies are currently under investigation.

Ipsilateral neck irradiation allows better sparing of radiation dose to collateral structures and reduces the risk of radiation-related side effects. Careful case selection is essential—well-lateralized lesion, without extension to soft palate or tongue base, no muscle involvement or any suspicion of deeper penetration, and no contralateral neck lymph node metastasis.

Transoral surgery and deintensification of adjuvant treatment

Adjuvant (chemo)radiation dose reduction following primary transoral surgery is also being proposed as an alternative de-escalation treatment strategy for HPV-positive OPSCC. The main advantage of this approach is the selection of adjuvant treatment based on pathological staging. Careful case selection is critical to avoid the need for triple modality therapy. Phase II data with mature follow-up has demonstrated support for this approach. Several clinical trials are ongoing, and treatment should not be routinely de-escalated until these trials have reported.

The unknown primary

The evolution of imaging strategies has resulted in identification of more occult primary OPSCC and targeted treatment accordingly. Identification of the unknown primary tumour occurs in 60– 90% of cases when the palatine and lingual tonsils are comprehensively sampled.[29–32] What is considered adequate mucosal sampling (unilateral vs bilateral tonsillectomy, base of tongue mucosectomy) remains an area of controversy and varying practice but consensus exists to offer tongue base mucosectomy in addition to traditional methods to enhance yield where expertise and facilities allow.[33] Excision of the tonsil beds and neck dissection may be curative in some cases. P16 status can also guide the planning of radiotherapy in those cases as the majority arise in the oropharynx. For those true T0 patients where a primary is not found after all investigations and endoscopic assessments are complete, especially if p16 negative, there is a need for more reliable evidence to guide management and surveillance. As recurrence rates are low in this population, comprehensive treatment of mucosal sites and the contralateral neck may represent overtreatment for some patients.

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57

Oral precancer and cancer

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Introduction

Oral squamous cell carcinomas (OSCCs) are one of the most common cancers of the head and neck region. The incidence is especially high in Southeast Asia, and in other regions where smokeless tobacco is commonly used. Oral cancers can be preceded by precancerous lesions, also known as potentially malignant disorders (PMDs). Often presenting as a surface lesion that can be easily detected with the naked eye, these lesions are most suitable for cancer screening and prevention programmes.

Anatomy

The oral cavity extends from the lips anteriorly to the anterior surface of the tonsil posteriorly. It is bounded by the hard palate above; the tongue, floor of mouth, and mandible below; and the buccal mucosa laterally. The mucosa of the oral cavity is lined by stratified squamous epithelium. It consists of many structures like muscles, minor salivary glands, blood vessels, nerves, teeth, and bones. These together form specialized structures within a confined anatomical area that serve numerous vital functions (e.g. swallowing and speech). The blood supply of the oral cavity is mainly from the branches of the external carotid artery, namely facial, lingual, and maxillary arteries. The cranial nerves supply the sensory and motor innervation of this region. The lymphatic drainage is primarily to levels I, II, and III neck nodes.

Potentially malignant disorders

Definition

In 2005, the World Health Organization combined the terms precancers and premalignant lesions into 'potentially malignant disorders' as these conditions might not necessarily precede cancer, but may harbour some potential for malignant transformation. OSCC is usually preceded by a visible precancerous lesion and identifying and treating these lesions early may be one of the most effective ways to decrease the mortality of oral cancers, especially in high-risk populations. Among the PMDs, some forms of leukoplakia, oral submucous fibrosis, and erythroplakia are the most frequently encountered varieties with a high malignant transformation rate (Figure 57.1).

Clinical features of potentially malignant disorders

The clinical features and the associated risk of various PMDs are tabulated in Table 57.1.

Aetiology of potentially malignant disorders

Various risk factors are related to the development of PMDs and oral cancers, of which tobacco use is most common. In endemic regions, the smokeless form of tobacco is a major risk factor contributing to PMDs and OSCC through contact carcinogenesis. Tobacco smoking increases the risk of leukoplakia. Other risk factors include areca nut use, which is very common in Southeast Asian regions. Areca nut produces reactive oxygen species and is recognized as an aetiological factor for the development of oral submucous fibrosis. Alcohol consumption also acts independently or in synergy with smoking to enhance the penetration of carcinogens through the mucosa. Certain viruses like human papillomavirus and nutritional, environmental, and genetic factors have also been considered aetiological factors of oral cancers.

Investigations of potentially malignant disorders

It is essential to detect PMDs at an early stage to prevent further progression and transformation. The cumulative malignant transformation rate across various studies with follow-up ranging from 12 months to 20 years (mean 7 years) after diagnosis is given in Table 57.1.

The survival rates of oral cancer when detected at an early stage are about 80–90%. However, when the disease progresses to an advanced stage, the 5-year estimated survival drops to around 30–40%. Hence, it is of paramount importance to diagnose and treat PMDs and oral cancers at an early stage to reduce the associated morbidity and mortality.

Clinical oral examination

The clinical oral examination (see Chapter 46) for any oral pathology begins with the visual assessment of the oral cavity subsites under white light illumination. This is followed by palpation of the

Figure 57.1 Various potentially malignant disorders. (a) leukoplakia of right lateral border tongue. (b) Proliferative verrucous leukoplakia lower alveolus. (c) Erythroplakia left buccal mucosa. (d) Lichen planus.

suspicious sites along with examination of the lymph nodes in the neck. High-risk areas of the oral cavity like the floor of mouth, lateral border tongue, retromolar trigone, and hard palate should be carefully examined for any suspicious lesions. Clinical oral examination is reported to have a sensitivity of 93% and specificity of 31%, as other conditions may mimic oral cancers.

Vital staining

This is a method of staining and identifying suspicious lesions using certain dyes. Staining with toluidine blue, an acidophilic metachromatic dye also known as tolonium chloride, is one of the oldest screening methods for oral cancers. Due to its affinity

Table 57.1 Potentially malignant disorders and their risk of malignant transformation

	Disorders	Clinical features	Site	Malignant transformation rate
1	Leukoplakia (Figure 57.1a):		Cheeks, lips, gingiva	8.6% (5.1–13.0%)
	Homogeneous	Thin, white, uniform, and flat plaque		
	Non-homogeneous	Can be speckled or nodular or verrucous		
2	Proliferative verrucous leukoplakia (PVL) (Figure 57.1b)	Multifocal, non-homogeneous, rough surface projections	Gingiva (most common site), buccal mucosa, tongue	49.5% (26.7–72.4%)
3	Erythroplakia (homogeneous, erythroleukoplakia, or speckled) (Figure 57.1c)	Red velvety plaque	The floor of mouth, tongue, retromolar pad, soft palate	33.1% (13.6–56.2%)
4	Oral submucous fibrosis (OSMF)	Trismus, blanching Bands of fibrosis	Buccal mucosa, retromolar area, tongue, soft palate	5.2% (2.9–8.0%)
5	Oral lichen planus (reticular, papular, plaque-like, erosive, atrophic, and bullous) (Figure 57.1d)	Red membrane, ulcers, blisters, reticular pattern with white streaks	Buccal mucosa, tongue, gingiva, palate	1.4% (0.9–1.9%)
6	Tobacco keratosis	White, non-elevated plaque with intertwining fissures	Buccal or labial vestibule	NA
7	Palatal lesions associated with reverse smoking (reverse smokers' palate)	White patches and plaques	Palate	83.3% dysplasia 12.5% squamous cell carcinoma
8	Nicotine stomatitis (smoker's palate)	Speckled white and red appearance	Palate	NA
9	Discoid lupus erythematosus	White keratinized plaques with elevated borders, radiating white striae and telangiectasia	Lip, cheek, palate, tongue, gingiva	NA
10	Actinic cheilitis	Ill-defined atrophic, erosive or crust forming lesions	Vermilion border of the lower lip	6–10%
11	Chronic hyperplastic candidiasis:		Commissure, buccal mucosa, palate, and tongue	15% progress to dysplasia
	Homogeneous	White plaque		
	Nodular/speckled	Erythematous background with white nodules		

Note: the rates in the last column are cumulative malignant transformation rates across various studies with follow-up ranging from 12 months to 20 years.

for nucleic acids, it helps highlight dysplastic and anaplastic cells which have increased quantities of nucleic acid compared to the normal mucosa. The reported sensitivity ranges between 90% and 100%, while the specificity is from 73.3% to 92.9%. The low specificity is attributed to the false-positive rates due to inflammatory or erosive lesions and granulation tissue and repeat assessment after 2 weeks can reduce the false-positive rates. Positive lesions stain dark blue and this might help in predicting molecular changes like loss of heterozygosity, with or without phenotypic changes in PMDs and OSCC. Vital staining has also been shown to identify PMDs at risk of progression to OSCC, to determine the extent of lesion, to assess margins during excision, and also in the follow-up of oral cancer patients after treatment. However, the low specificity and high false-positive rates outweigh its potential benefits for detecting cancers. Another commonly used dye is Lugol's iodine. It has a high affinity for glycogen-rich cells. The normal mucosa has a high glycogen reserve and has a higher uptake of iodine, producing a chocolate brown stain. Due to variations in proliferation and differentiation, the cancer cells have a low content of glycogen and hence they do not retain as much iodine stain and as a result appear pale. This is assumed to be due to the Warburg effect seen in cancer cells which leads to a higher rate of glycolysis in the cytosol. Specific subsites of the oral cavity like hard palate or alveolar gingiva have a higher degree of keratosis and low glycogen and hence may not retain iodine stains. Epstein et al. have shown that Lugol's iodine can be used as a screening technique with a sensitivity of 87% and a specificity of 84%.

Certain studies recommend using a combination of Lugol's iodine and toluidine blue or methylene blue in a double staining method to enhance the specificity of detecting suspicious lesions. Similar to toluidine blue, Lugol's iodine has also shown promise in delineating surgical margins and targeting the site of biopsy. Other vital stains, such as Rose Bengal stain, methylene blue, and acetic acid, have also been used as an adjunct to clinical examination to enhance the visual features of oral pathologies.

Light-based detection

PMDs and cancers cause alteration in the morphology and concentration of naturally occurring fluorophores like collagen, elastin, tryptophan, keratin, and NADH. It is hypothesized that the altered structural and metabolic changes give rise to varied absorbance and reflectance patterns when exposed to different light or energy

sources. When the reflected illumination is blocked with an absorbing filter, it helps to visualize longer wavelength fluorescence even with the naked eye.

Tissue fluorescence imaging devices like VELscope (Visually Enhanced Lesion Scope; LED Dental Inc., White Rock, BC, Canada) is a handheld device which helps in screening and evaluation of oral pathology. Blue light (400–460 nm) is used to excite green–red fluorescence from fluorophores, resulting in abnormal tissue appearing dark brown to black, in contrast to normal mucosa, which appears pale green. The obtained wavelength is guided into a computer which allows for the mathematical analysis of various spectra. With histology as the gold standard, autofluorescence is reported to have a sensitivity of 98–100% and specificity of 74–100% in discriminating normal mucosa from dysplasia, carcinoma *in situ*, or invasive carcinoma and also helps to target biopsies. However, the results need to be interpreted considering the above diagnostic parameters. Autofluorescence spectroscopy can also identify minor mucosal alterations as molecular changes precede clinical manifestation.

In chemiluminescence techniques like ViziLite (Zila Pharmaceuticals, Phoenix, AZ, USA) and Microlux/DL (AdDent Inc., Danbury, CT, USA), the patient is asked to rinse the oral cavity with 1% acetic acid, which helps to remove the glycoprotein layer and causes dehydration of the nucleic acid, followed by examination with a chemiluminescent light source. ViziLite Plus uses a disposable light packet while Microlux offers a battery-powered reusable light source. ViziLite plus also provides a toluidine blue solution to mark an acetowhite lesion for subsequent biopsy. ViziLite is shown to have a high sensitivity of 100% but low specificity ranging from 0% to 14% and positive predictive value of 18–80%.

Narrowband imaging is an endoscopic technique based on the use of special optical filters that narrow the bandwidth of light to enhance the visualization of the mucosa surface and its microvasculature. The reported sensitivity, specificity, positive predictive value, negative predictive value, and accuracy of narrowband imaging in detecting oral neoplasia is around 95–96%, 97–100%, 91–100%, 93–99%, and 97%, respectively.

While all these technologies aid physicians diagnose lesions better, they have certain limitations. They can only be used to assess mucosal lesions, cannot penetrate deeper tissue, and is not helpful in lesions with extensive keratosis, such as submucous fibrosis.

Cytology

Exfoliative cytology is based on a normal physiological process where cells are shed from epithelial surfaces. Loss of cohesion secondary to malignant transformation also leads to exfoliation. These disaggregated cells can be collected by scraping the mucosa or by rinsing the mouth. The resultant smears can be analysed quantitatively by analysing the nuclear size and shape, nuclear–cytoplasmic ratio, size of a cell, and so on, and qualitatively by cytomorphometry and computer-based image analysis.

Brush cytology is simple and inexpensive with a high specificity and positive predictive value.

False-negative results can be due to non-representative sampling, processing, and subjective error and the use of cytomorphometry can increase its dependability. Liquid-based cytology seems to offer better cytological morphology and improves the thickness of smear and cell distribution. The sensitivity and specificity of liquid cytology are around 88.8% and 100%, respectively, conferring significant advantage over brush cytology.

It can be used to monitor and follow-up PMDs and as an additional diagnostic test for early OSCC. However, cytology has a low negative predictive value and hence a negative report may not always exclude malignancy.

Histopathology

Biopsy is considered as gold standard for the detection of invasive oral cancer and can be incisional or excisional. An excisional biopsy is particularly valuable for dysplastic lesions when oral squamous cell carcinoma is clinically undetectable, as it not only provides a diagnosis but also serves as a treatment. Based on the extent of involvement of the epithelial layers, dysplasia can be classified as mild, moderate, and severe. As the lesion progresses, carcinoma *in situ* is characterized by dysplastic cells seen across all the layers of epithelium, with an intact basement membrane. Eventually, a breach in the basement membrane and involvement of stroma leads to an invasive carcinoma.

Management of potentially malignant disorders

The choice of treatment of PMDs depends on several factors such as the size of the lesion, appearance, location, histology of the lesion, and field cancerization as well as the presence of high-risk factors like use of tobacco and alcohol. The treatment can be divided into the following:

Observation with close follow-up

Patients with small lesions that clinically appear benign and are located at sites accessible to close surveillance like the lip, buccal mucosa, and oral tongue can be kept on close follow-up. Around 5–18% of epithelial dysplasias can transform into malignancies. The risk of malignant transformation is increased in:

- erythroleukoplakia
- proliferative verrucous leukoplakia
- high-risk sites like tongue or floor of mouth
- multiple lesions
- patients with no history of smoking
- females
- long-standing lesions
- presence of epithelial dysplasia
- leison with size greater than 200 mm^3.

Surgical excision

Excision helps in histological examination of the entire lesion but results in scarring and contracture and has not been shown to decrease the risk of malignant transformation.

Laser-assisted excision

Compared with other modalities, this is associated with less pain, minimal bleeding and oedema, and reduced risk of infection than conventional techniques. Re-epithelization over the raw surgical bed occurs in 3–4 weeks. Unfortunately, histopathological confirmation and genetic studies are not possible if ablation techniques are used. Recurrence rates with laser techniques using carbon dioxide, Nd-YAG, and KTP laser are 34.2%, 28.9%, and 17.0%, respectively.

Cryosurgery

This is associated with less bleeding, pain, scarring, and fibrosis than conventional techniques. Unavailability of a histopathological diagnosis, logistic issues around the storage of liquid nitrogen, and the initial cost are the associated disadvantages. Recurrence rates of 20–71.4% have been reported with cryosurgery.

Photodynamic therapy

This is an ablative method for the treatment of oral premalignant lesions. A photosensitizing agent (hematoporphyrin derivatives or 5-ALA) is administered either intravenously or topically and the targeted tissue is exposed to light of a specific wavelength which activates the photosensitizer, thereby generating reactive oxygen species and subsequent tissue damage. The subepithelial collagen and elastin are spared, resulting in better healing, minimal scarring, and better cosmesis. Though it has shown promising results in the management of oral premalignant lesions, its efficacy remains questionable.

Preventive measures for potentially malignant disorders

Primary prevention

The ideal and initial step towards cancer management is avoidance of risk factors known to cause PMDs and OSCC. Attributable risk for development of OSCC is 25% for smoking, 18% for alcohol, and 40% when used in combination. Population-based health campaigns should be promoted among the high-risk groups and adaptation of a healthy lifestyle should be encouraged.

Secondary prevention

This is achieved by early detection of premalignant lesions and/or preventing malignant transformation by removal of the aetiological factors as discussed above. The time to malignant transformation is unpredictable and varies from months to years. In the case of field cancerization, new lesions can present adjacent to the site of excision or other subsites, and therefore these patients need long-term follow-up.

Chemoprevention

This is the use of natural or synthetic substances to prevent initiation and promotion of carcinogenesis to cancer and its progression. Compounds including retinoids, epidermal growth factor receptor inhibitors/antagonists, cyclooxygenase-2 inhibitors, p53 modulators, and topicals, such as bleomycin, vitamin E, Bowman–Birk inhibitor concentrate (derived from soybeans), curcumin, and green tea polyphenol epigallocatechin-3-gallate are among several other agents that have been trialled for chemoprevention. Although antioxidants have shown promising results, the success rate remains uncertain with no long-term results.

Screening

Screening is a healthcare intervention designed to identify disease at an asymptomatic stage when the natural course can be interrupted, if not cured. A good screening test should be acceptable, applicable, effective, have high sensitivity, high positive predictive value, and be cost-effective. Screening of oral cancers has not been successful since early lesions can be asymptomatic and not easily recognized, with clinical symptoms being reported in the advanced stages. Moreover, these lesions are highly varied in their presentation, mimicking benign pathologies. Many case-finding or screening tools have been developed, but none have been shown to improve the sensitivity and specificity beyond clinical examination alone. A prospective randomized trial compared the efficacy of clinical examination as a screening tool. The results demonstrated better three-year survival in the intervention group, which was likely influenced by lead-time bias. However, the long-term follow-up results, spanning 15 years, showed no difference in overall survival among the population. Notably, improved survival was observed in males with high-risk habits.

Oral cancers

Oral carcinogenesis has a multifactorial aetiology: exogenous sources (alcohol, tobacco), physical causes (radiation), and infectious agents (viral and fungal). The carcinogens can lead to alteration in the genome (point mutation, deletion, rearrangement, and insertion of genes) resulting in changes in its structure and function, which is the hallmark of cancer pathogenesis.

Predominantly, around 90% of cancers arising from the oral cavity include OSCC, while the rest is contributed by the salivary gland tumours, sarcomas, lymphomas, and melanoma, etc.

The GLOBOCAN 2020 cancer database estimated 377,713 new cases of lip and oral cavity cancers with 177,757 deaths worldwide. The majority of incidence and mortality was from Southeast Asia and Pacific islands.

Symptomatology of oral cancers

Oral cancers are mostly asymptomatic during their initial stages. The common symptoms that should raise concern include:

- non-healing oral ulcer (most common presentation)
- persisting red/white lesions
- nodular thickening in the mucosa
- pain usually reflects an advanced infiltrative lesion
- recent onset of trismus indicating involvement of the infratemporal fossa, while long-standing trismus can be secondary to submucous fibrosis
- decrease in tongue mobility
- loosening of dentures
- increasing tooth mobility or pain may be secondary to alveolar invasion
- voice changes
- loss of weight/loss of appetite.

Assessment of oral cancers

The clinical examination of patients with oral cancer is of the utmost importance as the mucosal extent of disease needs to be well documented. The points to be noted while examining a patient with oral cancer are listed in Table 57.2.

Investigations of oral cancers

Histopathology

Tissue biopsy is the gold standard for the diagnosis of OSCC. Based on the differentiation, squamous carcinomas can be further

Table 57.2 Checklist for clinical evaluation of oral cancer

General examination	• General health • Nutritional status • Performance status (Karnofsky or ECOG)
Local examination	• Mouth opening • Presence of trismus/submucous fibrosis • Extent of tongue mobility • Oral hygiene and tooth mobility • Dentition • Examination of all subsites, especially hidden areas like the floor of mouth, retromolar trigone, and tonsillo-lingual sulcus • Documentation of the size, extent of the lesion, thickness, submucosal extension/induration in the surrounding area, fixity to underlying structures, and involvement of soft tissues of cheek and neck or skin involvement (Figures 57.2–57.4)
Neck examination	• Laterality of neck nodes • Number of palpable nodes • Size of the largest node • Level of the involved neck node • Mobility of the nodes
Endoscopy	• Nasal endoscopy • Fibreoptic laryngopharyngoscopy

Figure 57.2 Severe dysplasia of left buccal mucosa.

classified as well, moderately, or poorly differentiated. It also helps in identifying non-squamous pathologies. Biopsies can be done as an office procedure under local anaesthesia or may require sedation.

Imaging

This is discussed in detail in Chapter 69.

Computed tomography (CT) scan

This is the initial investigation of choice for evaluating oral cancers. Contrast-enhanced multidetector CT provides sufficiently thin slices

Figure 57.3 Early cancer of the lateral border of oral tongue.

Figure 57.4 Early tongue cancer arising in a background of non-homogeneous leucoplakia.

for reformatting the tumour in various axes for complete evaluation. The puffed cheek technique helps in a comprehensive assessment of mucosal surfaces of the oral cavity, and is especially useful in cancers involving the retromolar trigone and gingivobuccal sulcus. The details to be noted while evaluating a CT scan are given in Table 57.3. In addition, non-contrast CT thorax is advocated in advanced cancers to rule out lung metastases.

Table 57.3 Checklist for evaluation of oral cancer on contrast-enhanced computed tomography

Epicentre	The most probable site of origin of the cancer
Dimensions	To be evaluated in all three planes at the maximum dimension
Extent of disease	All the subsites involved must be enumerated
Soft tissue involvement	Attention must be given to the extent of involvement of skin, floor of mouth, base of the tongue, and the masticator space and infratemporal fossa
Bone involvement (bone windows)	The presence of cortical erosion or gross destruction of bone (marrow involvement) as well as the residual mandibular height
Perineural spread	Gross perineural spread along major nerves like the mandibular, lingual, and hypoglossal nerves must be identified and traced to the skull base
Cervical lymph nodes	Size, number, level, laterality, necrosis, extranodal extension, and relationship with surrounding structures must be reported

Magnetic resonance imaging

This is particularly useful in cancers of the tongue and floor of mouth as it offers better soft tissue definition than a CT scan. Magnetic resonance imaging is more sensitive in the early detection of marrow involvement, and superior to CT images in detection of perineural spread, involvement of the masticator muscles and infratemporal fossa.

Fluorodeoxyglucose (FDG) positron emission tomography (PET)/CT

In the case of locoregionally advanced disease, high-grade tumours, or lower neck node involvement, evaluation of distant metastasis by FDG PET/CT is recommended. FDG PET/CT is also preferred while evaluating recurrent tumours, post-treatment changes, second primaries and distant metastasis.

X-ray

Although chest X-rays are used as annual screening to detect early metastasis, they are less sensitive than CT scans. Dental imaging is required prior to radiation as part of comprehensive dental evaluation/prophylaxis, and after radiotherapy (RT) in case of osteoradionecrosis.

Staging of oral cancers

This can be found in the American Joint Committee on Cancer's eighth edition staging of oral cancer (AJCC Cancer Staging Manual).

Management of oral cancers

Treatment involves a multidisciplinary team approach (see Chapter 46) consisting of surgeons, radiation and medical oncologists, dentists, nutritionists, specialist radiologists and pathologists, speech language therapists, and rehabilitation specialists.

The definitive management for most oral cancers is surgery followed by risk-based adjuvant therapy in the form of RT or concurrent chemoradiotherapy. This approach has been shown to offer the best chance of disease control across all stages of operable oral cancer. In certain situations, such as lesions involving the lip or the hard palate where surgery might severely compromise form and function, brachytherapy or external beam radiotherapy (EBRT) might be considered a possibility. Chemotherapy is mainly used as a radiosensitizer in curative settings as a part of adjuvant or non-surgical definitive treatment protocols. There is an emerging role of induction chemotherapy in borderline unresectable disease.

In addition to the definitive management, patients should also receive speech and swallow therapy, nutritional optimization, and de-addiction and fertility counselling if needed. Prior to initiating treatment, the oral health needs to be evaluated and optimized. If necessary, dental extractions should be performed prior to RT to prevent osteoradionecrosis.

Goals of treatment

The goals of treatment of oral cancer are complete extirpation of the disease, preserving as much normal tissue structure and function within oncological limits. This is done with an aim of achieving maximal locoregional control and improving overall survival. It is important to try and restore as much normal functions of swallowing, mastication, and speech as possible, with the help of appropriate reconstruction techniques. T4b tumours, unresectable nodal disease, and distant metastatic disease with a good performance scale are treated by chemotherapy or RT. Patients with poor general health are directed towards supportive care. Early palliation care should be integrated in the management of oral cancer at advanced stages.

Management of the primary site

The goal of surgery is to achieve complete removal of the cancer with wide mucosal and soft tissue margins in three dimensions. The most widely accepted definition of adequate margins is 5 mm of normal tissue around the tumour. It is established that positive margins after surgery result in reduced overall survival. The details of the various surgical procedures and reconstruction are beyond the scope of this chapter.

Management of the neck in oral cancer

If patients are being treated with surgery, the draining nodal stations in the neck, namely levels I, II, and III, must be surgically treated with an elective neck dissection. Elective neck dissection improves locoregional control and saves lives in patients with node-negative early oral cancer. There is emerging data on the role of sentinel node biopsy in early oral cancer which may be an oncologically safe procedure with reduced morbidity. In node-positive cancers, comprehensive neck dissection of the neck from levels I to V is suggested. There is emerging evidence that in certain cases of ipsilateral level I positivity, level I–IV neck dissection may be adequate, but further studies are warranted. In case of lesions crossing the midline or N3 nodal disease, a contralateral neck dissection might also be performed.

The treatment algorithms for each stage of OSCC are summarized as in Figure 57.5.

Principles of reconstruction

The main goal of reconstruction is to restore function, form, and aesthetics. Emphasis is on restoring swallowing, speech, and dentition in oral cancer. Wherever reconstructing the oral cavity, 'like-for-like tissue replacement' should be followed as far as possible.

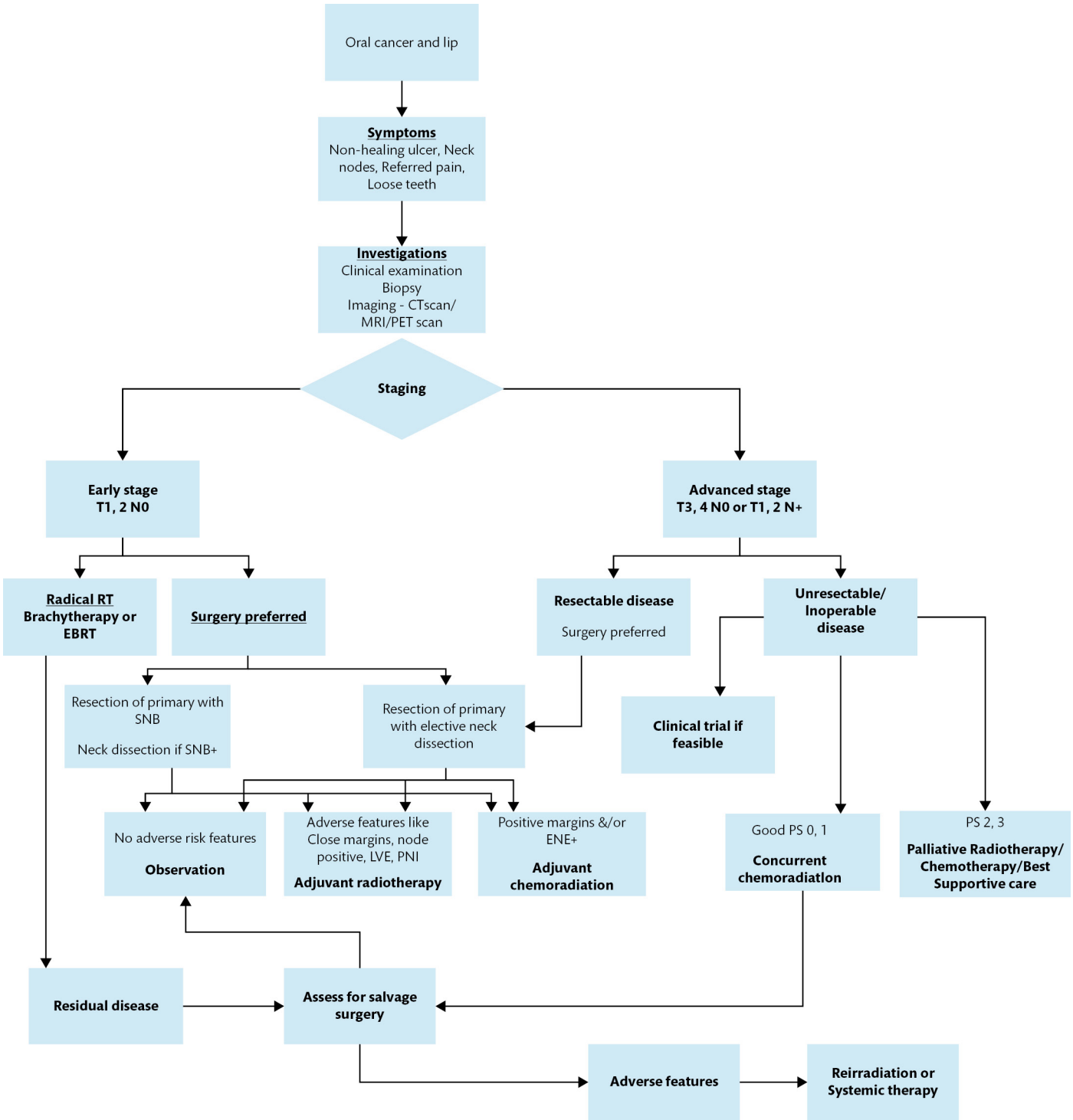

Figure 57.5 Algorithm for the management of oral cancer. ENE, extracapsular nodal extension; LVE, lymphovascular embolism; MRI, magnetic resonance imaging; PNI, perineural invasion; SNB, sentinel node biopsy.

Borderline and unresectable oral cancers

Locally advanced T4b cancers are deemed unresectable when there is frank involvement of skull base, tumour encasing the internal carotid artery, or involvement of the prevertebral fascia.

Some T4b cancers are considered to be borderline resectable as the extent of soft tissue disease makes it difficult to achieve clear margins without causing major functional morbidity. The features that are considered to make a disease borderline resectable include:

- extensive skin involvement (Figure 57.6)
- buccal mucosa cancers with peritumoural oedema reaching above the zygomatic arch
- advanced anterior tongue cancers extending up to or below the hyoid
- anterior tongue cancers reaching the vallecula

Figure 57.6 Advanced buccal cancer with skin fungation.

- infratemporal fossa involvement above the sigmoid notch—there is recent evidence that suggests that infra-notch diseases are amenable for surgery with satisfactory outcomes.

There is a role for using neoadjuvant chemotherapy in these borderline cancers, with reduction in the soft tissue extent facilitating resection of the tumours with clear margins and thereby improved outcomes.

Complications of surgery

Vascular complications

- Haemorrhage is the most common immediate complication. It can be due to the slipping of ligatures or clips. Secondary haemorrhage can occur due to infection.
- Carotid blowout is the most dreaded complication in head and neck cancer surgeries. It is a rare complication in the primary surgical setting (~1%), but more commonly seen in recurrent disease and the radiated neck. Wound infection, exposure to saliva, radical stripping of the carotid sheath, and flap necrosis can predispose to carotid blowout, especially in the post-radiation setting.

Neurological complications

- Marginal mandibular nerve paresis can occur during neck dissection secondary to traction, use of diathermy, and devascularization. The Hayes Martin manoeuvre is a simple technique to safeguard the marginal branch of the facial nerve.
- Injury to the spinal accessory nerve during neck dissection, especially during the clearance of the posterior triangle, can result in trapezius muscle atrophy and shoulder dysfunction.

Wound dehiscence

This is more common in patients who are previously radiated. Placement of incisions should be planned without compromising the blood supply to the skin.

Wound infection

The use of antibiotic prophylaxis has reduced the risk of wound infection in clean-contaminated head and neck surgeries. The nutritional status, history of alcohol consumption, complexity of the surgical procedure, choice of antibiotics, stage of the disease, blood transfusion, use of drains, tracheostomy, and nasogastric tube may impact the incidence of wound infection. It can significantly increase the morbidity and postoperative hospital stay.

Seroma

This occurs due to the accumulation of fluid within the operated site. Small seromas resolve while a larger one may require aspiration. Seromas can get secondarily infected and interfere with wound healing.

Chylous fistula

The presence of milky, greasy fluid in the drains should raise the suspicion of a chyle leak. It is more common during left-side neck dissection (8.3%) and occurs due to thoracic duct injury while clearing level IV lymph nodes and can lead to delayed wound healing and fistula formation. It is usually managed conservatively with pressure dressing, fat-restricted diet, and continuous drain. Any persistent or high output leaks may require surgical intervention.

Salivary fistula

A salivary leak should be suspected in patients with fever of unknown origin, neck collection, or inflammation of the skin flap. Anticholinergics may be prescribed to decrease the salivary flow and the wound should be explored and attempts should be made to direct saliva away from the vital structures in the neck. Injection of botulinum toxin into salivary glands will hasten the resolution of the fistula by reducing salivary flow.

Respiratory complications

The incidence of pulmonary complications varies from 5.1% to 30.6% after oral cancer surgery. This can be attributed to advanced age, male sex, and long duration of surgery, tracheostomy, and delayed mobilization.

Deep vein thrombosis (DVT)

Patients undergoing major head and neck cancer surgery have the risk of developing DVT and pulmonary embolism. Advanced age, obesity, history of alcohol and smoking, heart disease, prolonged duration of surgery, and immobilization in the postoperative period predisposes to the development of DVT. Preoperative pneumatic compression devices and postoperative prophylaxis based on well-rehearsed local or regional protocols should be used to prevent DVTs.

Role of non-surgical management

Early oral cancers

Early oral cancers can be managed by a single modality of treatment, either surgery or RT. There is no randomized trial comparing RT to surgery in early oral cancers. RT is indicated in a specific subset of cancers like lip and oral commissure where surgery may lead to cosmetic and functional defects. Radical RT is generally offered in patients with advanced age, in the presence of major comorbidities that preclude surgery, or a patient's refusal for surgery.

EBRT using advanced techniques like intensity-modulated RT and image-guided RT helps achieve good locoregional control with

reduced adverse effects. Brachytherapy can be offered for lip cancers and certain buccal mucosa cancers away from the bone alone or in combination with EBRT. The decision on radical brachytherapy versus a combination of brachytherapy with EBRT is dictated by the location of the lesion, size, thickness, and probability of lymph node involvement.

Advanced oral cancers

The mainstay of treatment of resectable oral cancers is surgery unless the patient is unfit or unwilling to undergo surgery. RT, with or without chemotherapy, is usually reserved for adjuvant treatment after surgery for locally advanced cancers. The primary indications for adjuvant EBRT are T3 and T4 tumours, nodal positivity, and the presence of pathological adverse features like the presence of lymphovascular or perineural spread and close surgical resection margins. Positive margins and extranodal extension would warrant the use of concurrent chemoradiation. Cisplatin (either weekly or 3-weekly) is the most commonly used agent along with RT.

When surgery is not being considered, concurrent chemoradiotherapy may be used. Biologically, target agents have been found to be promising when used in combination with chemotherapy and RT and are under evaluation in certain subgroups of patients.

Rehabilitation

Rehabilitation of all oral functions is a desirable but challenging task to achieve. Early-stage tumours have less functional alterations post-treatment compared to advanced tumours. Rehabilitation measures include the restoration of facial contour, oral competency, speech, ability to chew and swallow food, and prosthodontic rehabilitation. This function and cosmetic disfigurement can have an impact on the health-related quality of life following treatment; hence all attempts should be made to optimize functional outcomes for these patients. Early involvement of allied health professionals, such as speech and language therapists, physiotherapists, and occupational therapists, help in early post-treatment functional recovery.

Follow-up

The risk of developing second primary tumours is around 3.7% per year and accounts for approximately 25% of deaths in oral cancers. This should be borne in mind while assessing oral pathologies as it mandates close follow-up and regular clinical assessment of patients with a prior history of oral cancer. Follow-ups should include regular clinical evaluation and imaging at appropriate intervals. Special attention should also be taken to ascertain tobacco cessation.

Points to remember

- Oral cancers are common cancers in the Indian subcontinent and Southeast Asia.
- PMDs should always be evaluated and followed up for malignant transformation.
- Screening of high-risk populations (tobacco and alcohol users) may be beneficial in reducing oral cancer mortality.
- Early diagnosis of oral cancer can be done by visual examination and confirmed by biopsy.
- Surgery is the mainstay of treatment in early oral cancers.
- Surgery followed by adjuvant RT/chemoradiation is the treatment of locally advanced oral cancers.

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58

Salivary gland tumours

Khanh K. Nguyen and Nitin A. Pagedar

Introduction

The group of salivary gland neoplasms is diverse and demonstrates a variety of histological types. They account for up to 6% of head and neck tumours with malignant lesion having a population incidence of 1/100,000 cases.[1] The cause of salivary tumours is unclear although some risk factors have been identified for malignancy including radiation exposure, previous radioactive iodine treatment, history of other malignancy, and a pre-existing benign salivary tumour.[2,3]

The most common benign salivary neoplasms include pleomorphic adenoma and papillary cystadenoma lymphomatosum (also known as Warthin's tumour), among many others. Common malignant salivary tumours include mucoepidermoid carcinoma, acinic cell carcinoma, adenoid cystic carcinoma, and adenocarcinoma. Squamous cell carcinoma very rarely presents as a primary malignant tumour of salivary glands and more often represents a metastatic lesion. In addition to the various histological subtypes, salivary malignancies are categorized as low-, intermediate-, or high-grade tumours. The prognosis for patients with high-grade tumours is often worse due to an increased rate of local recurrence and distant metastases. As a result, the management of salivary malignancy should be approached as a multidisciplinary team with adjuvant treatment options.[4] This chapter will discuss a general overview of management of this diverse disease entity. It will highlight specific aspects in the workup, management, and/or follow-up of a particular tumour if it is uniquely different from the group.

Presentation

The majority of salivary neoplasms occur in the parotid gland and up to 80% of tumours identified are benign. The submandibular gland, minor salivary glands, and sublingual gland can also be affected, in order of decreasing incidence. In contrast to the parotid gland, tumours arising from the minor salivary and sublingual glands are malignant in 80% of cases, and in the submandibular gland, 40% of cases (Table 58.1).

Patients are often referred for evaluation due to an asymptomatic mass identified on examination or found incidentally on an imaging test. The history and physical examination are critical first steps to help the clinician determine the risk for malignancy. Key historical features that would suggest malignancy include rapid development of the tumour, pain in the tumour bed or ear, a history of radiation to the head and neck area, a history of other cancers, and previous known tumour at the same location.[5] Rarely, patients may present with acute sialadenitis due to salivary outflow obstruction. In these cases, the clinician should be aware of this possibility and re-evaluate the patient when the acute inflammatory status is improved.

Table 58.1 Incidence of salivary neoplasms and rate of malignancy by site

Site	Incidence of neoplasm (%)	Rate of malignancy (%)
Parotid	70–85	20
Submandibular	8–15	40
Minor salivary	5–8	80
Sublingual	1	80

Often a benign tumour will be well circumscribed and mobile relative to adjacent structures. Physical examination findings that suggest malignancy include overlying skin changes, a firm and fixed mass, and any cranial nerve deficit such as facial weakness, tongue paraesthesia, or atrophy.

Neoplasms are not the only cause of salivary enlargement. Otolaryngologists should be aware of the wide range of pathophysiological conditions which might present with isolated or generalized salivary swelling.[6] Obstruction of a gland's ductal system, from sialolithiasis or stenosis, can affect a portion of or the whole gland, most often causing prandial symptoms. Swelling and pain commonly accompany radioiodine therapy for thyroid cancer.[7] Systemic inflammatory disorders such as Sjögren's syndrome and sarcoidosis can present with salivary gland enlargement, as can viral infections including human immunodeficiency virus (HIV). In experienced hands, ultrasound is effective in characterizing non-specific salivary swellings,[8] with sialendoscopy sometimes offering both diagnostic information and treatment.[9]

Diagnostic testing and workup

The history and physical examination are the beginning of the workup for salivary tumours, but further evaluation is necessary for appropriate management and counselling for the patient. Although not universally recommended, the authors find that tissue diagnosis and radiographic evaluation help to determine the appropriate surgical management.

Because salivary tumours are often accessible, fine needle aspiration biopsy is an effective tool for the clinician.[10] Sensitivity and specificity for this technique approach 90%. As with its utilization for other neoplasms, the results can vary by operator, and the most common cause for false-negative results is due to inadequate sampling. Despite its usefulness, however, fine needle aspiration biopsy can yield indeterminate results in up to 30% of cases. This is largely due to the diverse histological features of salivary tissue.

Salivary gland imaging is discussed in detail in Chapter 69. In conjunction with tissue diagnosis, the imaging modalities are immensely helpful in further evaluation of salivary tumours.

Preoperative counselling

The primary modality of treatment for salivary neoplasm of all types is surgical excision.[11] In preparing for surgery, patients should be appropriately counselled, including the main risks for the surgery as well as the expected course for recovery. In addition to the usual risks for surgery, such as scarring, keloid formation, and infection, the most obvious complication of surgery of the salivary gland is temporary or permanent injury to the facial nerve which depends on the extent of surgery as well as tumour location. The incidence for immediate postsurgical paralysis is reported as being as high as 35% but is much lower at 7% for permanent injury. With the use of continuous facial nerve monitoring, the former result has been demonstrated to be significantly lower at 22% but there is no reported difference for permanent facial nerve weakness.[12]

Other common and unique risks to salivary surgery include auriculotemporal syndrome, also known as Frey syndrome, first bite syndrome,[13–15] and injury to adjacent cranial nerves. A summary of the incidences and description of these is shown in Table 58.2.

Familiarity and early recognition of these complications is part of providing good patient care but taking further steps to manage such complications may be necessary and will be addressed further in the chapter.

The parotid gland contributes to the normal contour of the lateral face, and removal of tissue at that region often results in a notable soft tissue deficiency and malposition of the auricle and ear canal. The resultant impacts on quality of life have been described by several groups.[16,17] Soft tissue augmentation can be performed with free fat grafts, buried adipofascial or muscle flaps, and free flaps.[18–21] Although often ignored, the authors believe the benefits and costs of contour restoration should be considered as the surgeon and patient discuss surgery.

Surgical planning

In cases of known benign salivary tumours, a superficial parotidectomy or submandibular gland removal is often adequate and can be scheduled as an outpatient procedure if the patient does not have any underlying requirement for additional observation after surgery. Where there is confirmed or clinical suspicion for malignancy, the surgeon should be prepared to perform the appropriate extent of lymphadenectomy. Because most salivary tumours primarily involve the submandibular and parotid gland, this section will focus primarily on tumours arising from these areas.

Submandibular gland surgery

Incision planning for removal of the submandibular gland should be oriented along a relaxed skin tension line and can often be hidden in a cervical skin crease. The incision should be planned approximately 3–4 cm below the lower border of the mandible to limit inadvertent injury to the marginal mandibular nerve. The length of the incision can be extended as necessary for adequate subplatysmal flap elevation and exposure. The following are general steps for removal of the gland:

1. Elevate a subplatysmal flap to the level of the lower border of the mandible after the incision is made. The deep surface of the platysma should be visualized at all times to assure that surgical dissection is proceeding in the appropriate surgical plane.

Table 58.2 Most common complications, their description, and likely site of occurrence

Complication	Description	Site	Incidence (%)
Auriculotemporal syndrome (Frey syndrome)	Sympathetic response due to aberrant reinnervation of postganglionic parasympathetic fibres. Can result in flushing, discomfort, or sweating over the parotid region prior to eating	Parotid—superficial	20
First bite syndrome	Hypersensitivity of myoepithelial cells in the parotid region due to loss of sympathetic innervation leads to parasympathetic activation and results in onset of sharp pain in the parotid area in anticipation of oral intake. Pain subsides with subsequent bites and worsens with sialagogues	Parotid—deep lobe	10–38
Sialocele/salivary fistula	Draining tract or collection of saliva in surgical wound bed	Parotid	6–10
Cranial nerve injury: Facial nerve Hypoglossal nerve Lingual nerve	Facial paresis in the distribution of affected branch Uncommon but can result in dysarthria Ipsilateral paraesthesia of the tongue	Parotid Submandibular Submandibular	7 0–7 0–22

2. Identify the lower border of the submandibular gland and expose the inferior extent of the gland. The fascia can then be opened anteriorly and posteriorly being mindful that, posteriorly, one will encounter the common facial vein. The anterior belly of the digastric muscle represents the medial extent of the dissection.
3. Retract the gland inferiorly and elevate the superficial fascial layer superiorly. This step helps to ensure that the marginal mandibular nerve is protected.
   a. Alternatively, careful dissection of the superficial fascia layer overlying the submandibular gland using a scalpel can allow for direct visualization of the marginal mandibular nerve. This should be done in a direction parallel to the lower border of the mandible. Once the nerve is identified, it can be dissected free and retracted superiorly (Figure 58.1).
4. Release the submandibular gland from the lower border of the mandible dividing the distal facial artery and vein.
5. Release the gland off the anterior belly of the digastric and the mylohyoid muscle. Identify the lingual nerve and release the gland from the submandibular ganglion. The submandibular duct should also be divided leaving the gland attached only by the proximal facial artery.
   a. In some instances, the entire facial artery can be easily dissected free from the submandibular gland and preserved (Figure 58.2). However, the procedure can be simplified by division of the facial artery distally as described above and then proximally.
6. Obtain haemostasis and close the incision. A closed suction drain can be placed, depending on the size of the wound bed and the surgeon's preference.

Parotid surgery

A superficial parotidectomy with or without facial nerve dissection is the standard approach for removing tumours of the parotid gland. Various incisions have been described, such as the periauricular incision, the face lift incision, and variations of the cervicomastoid incision—also known as the modified Blair incision.[22,23] Each of these approaches has been demonstrated to provide sufficient exposure with reasonable cosmetic outcomes, and each surgeon should choose what is most familiar and comfortable. However, when there is a high probability that a neck dissection would be included, the modified Blair incision is the best approach as it can be readily extended in a cervical skin crease.

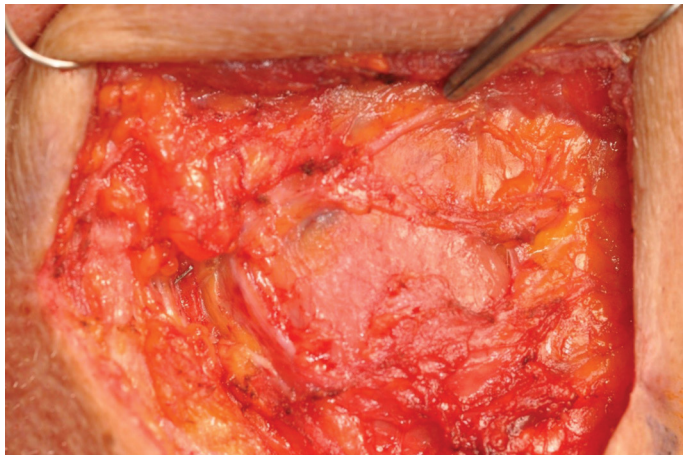

Figure 58.1 Flap elevated and marginal mandibular branch of facial nerve exposed, shown by tip of forceps.

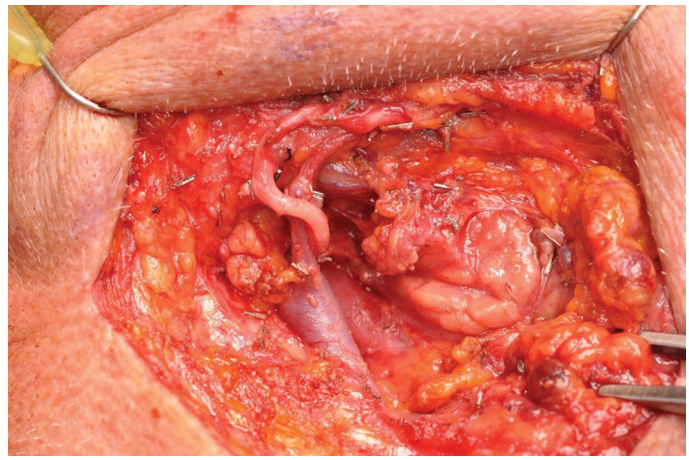

Figure 58.2 Facial artery preserved with submandibular gland retracted medially.

The following are the general steps and approach for a superficial parotidectomy:

1. Make the preauricular incision to the superficial parotid fascia and through the platysma in the neck. Keep in mind that the subcutaneous tissue is often thin in the preauricular area and one will encounter the parotid fascia quite easily.
2. Elevate the facial skin flap over the superficial parotid fascia. In the neck, the flap should be elevated in the subplatysmal plane. When performed correctly, the two flaps elevated will join at the level of the mandible where the platysma transitions into the superficial musculoaponeurotic system. Flap elevation should extend anteriorly to the anterior edge of the tumour. Identification of the superficial parotid fascia is facilitated by injection of dilute epinephrine (1:100,000), with or without lidocaine (Figure 58.3).
3. Identify the facial nerve trunk utilizing the well-established landmarks (Table 58.3 and Figures 58.4 and 58.5).

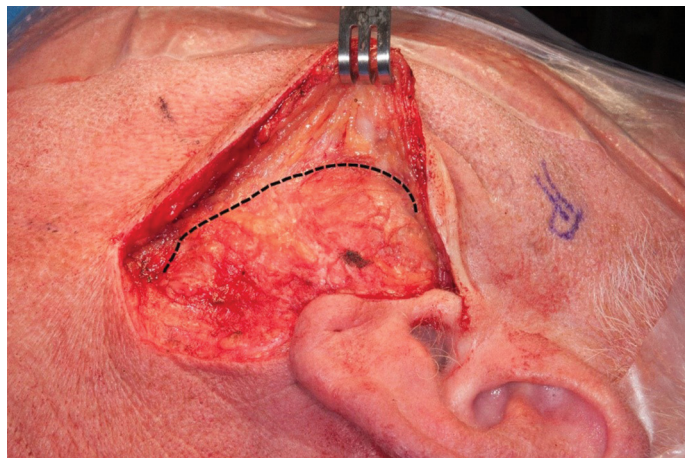

Figure 58.3 Flap elevated in the plane of the superficial parotid fascia to uncover the tumour. The dashed line shows the extent of flap dissection.

Table 58.3 Methods for identifying facial nerve

Method	Description
Posterior belly of digastric and tragal cartilage	Expose the tragal cartilage and follow it medially as the parotid is retracted anteriorly. The facial nerve lies about 10 mm anterior and inferior to the medial aspect of the tragal cartilage. Exposing the posterior belly of digastric and tracing it towards the mastoid tip provides a sense of depth of the point of exit for the facial nerve
Tympanomastoid suture	Palpate for the fissure between the external auditory canal and mastoid bone—this is the tympanomastoid suture line. Release parotid tissue anteriorly while following the suture line medially. The facial nerve lies ~2 mm inferior to the medial most extent of the suture line[24]
Retrograde dissection of peripheral branch	The marginal mandibular nerve is typically the most readily identifiable peripheral branch due to its size and consistent course. Once it is identified over the submandibular gland, follow the nerve proximally until the lower division or main trunk of the facial nerve is identified
Middle ear and mastoid	Perform a cortical mastoidectomy and identify the vertical segment of the facial nerve lateral to the horizontal semicircular canal within the mastoid cavity. This could then be followed inferiorly while removing the mastoid tip to expose the stylomastoid foramen

4. Perform an anterograde dissection of the facial nerve, only tracing out the necessary branches for tumour resection. Divide overlying parotid tissue using bipolar cautery and taking care to avoid cutting through the tumour.
5. Close the incision over a small-calibre closed suction drain:
   a. Free abdominal fat graft can be harvested and utilized for facial contouring and to limit postoperative facial depression. One could anticipate about a 30% atrophy of the graft, therefore overcorrection should be considered.

In the last 20 years, there has been interest in limiting the extent of parotid surgery for benign tumours. Whereas a superficial parotidectomy with facial nerve dissection may have been favoured

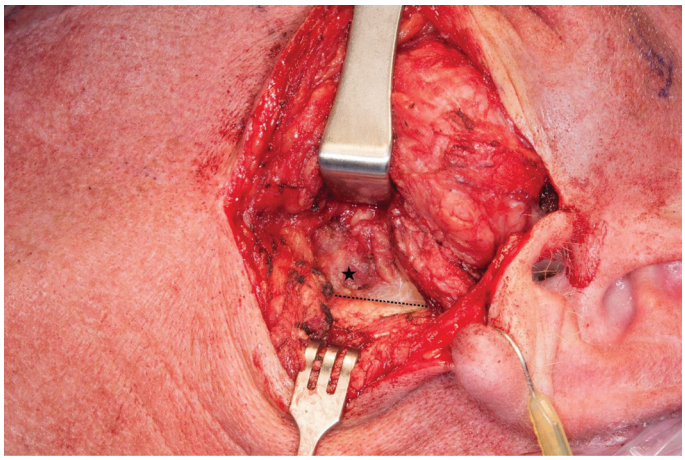

Figure 58.4 Sternocleidomastoid and posterior belly of digastric exposed near the mastoid tip. Dotted line shows sternocleidomastoid and the star shows the posterior belly of digastric.

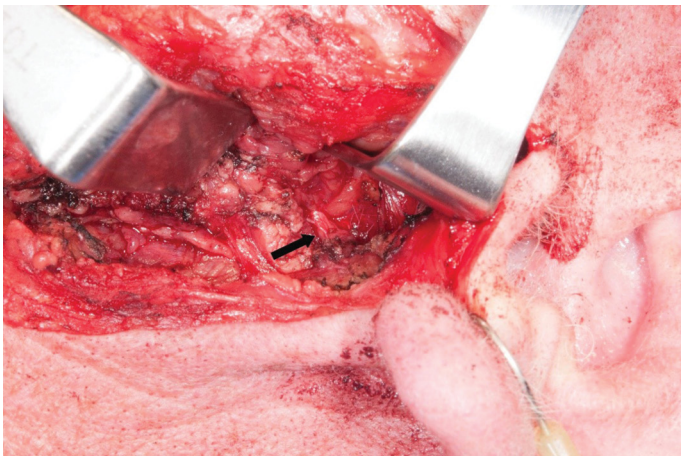

Figure 58.5 Facial nerve exposed at the stylomastoid foramen, shown by the arrow.

for these cases, there is now growing favour for extracapsular dissection of benign lesions without direct visualization of the facial nerve or very limited nerve dissection.[25–27] This technique involves identification of the tumour and its capsule and dissection along its loose areolar plane. Bipolar cautery should be utilized as well as loupe magnification to aid in visualization and minimize the risk of facial nerve injury. Studies have demonstrated only about a 1% occurrence of Frey syndrome, salivary fistula, or facial nerve injury with no difference in tumour recurrence compared to standard parotidectomy in cases for pleomorphic adenoma. By limiting facial nerve dissection, studies have also demonstrated lower rates of early facial nerve paresis and decreased operative time. The recommendation for this technique is for tumours less than 4 cm and to be performed by surgeons experienced in parotid surgery. The surgeon should be ready to perform a standard parotidectomy where there are intraoperative concerns for malignancy. In the authors' view, this technique is most appropriate for benign tumours arising in the tail of the parotid gland that are readily palpable and appear superficial radiographically. In these cases, an incision that extends posterior to the angle of the mandible along a cervical crease often provides adequate exposure and direct access to the tumour.

As discussed previously, the use of continuous facial nerve monitoring does not appear to confer significant additional benefit with regard to decreasing the risk for long-term facial nerve paralysis. However, as this tool is often available and its use is not burdensome, it is the authors' opinion that it should be utilized. It requires experience to understand how best to utilize the information gained from the facial nerve monitor, and surgeons should be vigilant and not solely rely on it in order to preserve the facial nerve and its branches.

Consideration for neck dissection

There is a wide variation in the rate of occult metastasis from malignant parotid tumours ranging from 12% to 49%, with histological type being an important predictor. The main risk factors for cervical involvement include high histological grade and local invasion, factors which are sometimes not known until surgical resection and final histological evaluation. The decision to perform an elective neck dissection requires the surgeon to take individual patient

circumstances into consideration. When elective neck dissection is performed, levels I–III should be addressed for submandibular gland tumours, and levels II–IV and Va for parotid tumours. For the latter, studies have demonstrated only about a 6% rate of occult disease in levels I and V.

The authors suggest a formal level 1b neck dissection when removing tumours of the submandibular gland especially when there is any suspicion for malignancy. Reoperation of this surgical bed at a later time if necessary is much more challenging while there is minimal added morbidity during time of primary surgery.

Adjuvant treatment

Adjuvant radiation treatment should be considered for malignant tumours with high-risk features such as high grade, positive surgical margins, perineural invasion, lymphovascular invasion, skin invasion, deep lobe involvement, and regional node metastasis.[28] Additional consideration for radiation treatment should be given to patients who did not receive surgical treatment of the cervical lymph nodes but are high risk for occult disease. In rare circumstances, radiotherapy might be appropriate for patients with recurrent or unresectable pleomorphic adenoma.[29]

Management of surgical complications

In the immediate postsurgical period, patients can develop a collection of saliva in the surgical bed. These collections should be aspirated, similar to the management of a postoperative seroma.[30,31] Use of a pressure dressing or anticholinergic medication could also be considered. Botulinum toxin injection into the residual parotid gland is useful in recalcitrant cases. Depending on the rate of accumulation, patients may need to have frequent aspiration with weekly clinic visits until resolution.

Management of temporary facial nerve weakness depends on the location of nerve injury. Branches that innervate the orbicularis oculi are of greatest concern and management of paresis of these branches primarily involves providing supportive eye care to prevent corneal drying and injury. The patient should undergo ocular examination and ongoing evaluation until nerve function returns. Intermediate- or long-term impairment in eye closure could be managed with placement of an eyelid gold weight, lateral tarsal strip, or tarsorrhaphy.

While injury to other facial nerve branches can cause dissatisfaction due to facial asymmetry, they do not often cause significant functional deficits. Smile asymmetry from injury to the marginal mandibular nerve can be treated in an algorithmic manner which is beyond the scope of this chapter.

Due to its relative high incidence, the salivary surgeon should be ready to recognize, counsel, and treat patients who may describe symptoms of auriculotemporal syndrome after surgery. In the authors' experience, this does not often cause significant distress, and many are often satisfied with an explanation as to what is taking place. For patients whose quality of life is significantly affected, small aliquot injections of botulinum toxin in the dermis can improve symptoms. While this is a relative safe and effective treatment modality, there is a high rate of recurrence and patients may need to have repeat injections in the future.

While it is uncommon, first bite syndrome can be highly uncomfortable for patients and various treatment options have been described. Some patients seem to respond to non-steroidal anti-inflammatory drugs and use of carbamazepine, pregabalin, gabapentin, or tricyclic antidepressant medications can decrease pain severity. As with auriculotemporal syndrome, botulinum toxin injection to the symptomatic area can also be temporarily effective.

Outcome and prognosis

For benign tumours, most patients do not require ongoing follow-up once healed postoperatively. Some patients with pleomorphic adenoma at the tumour resection margin, often due to proximity of the facial nerve, might benefit from follow-up. Patients who had a malignant tumour should be treated and followed in accordance with the local or national guidelines. Usual recommendations are follow-up every 1–3 months for the first year after treatment, every 2–6 months in the second year, and every 4–8 months until year 5.

Patients who had a high-stage, high-grade tumour, or perineural invasion have been shown to have a worse prognosis compared to their counterparts with an overall survival at 5 years decreasing from 94% to 50% for those with stage 3 or 4 disease.[32] Regular evaluation is critical to detecting early recurrence, but routine radiographic evaluation should not be performed unless there is clinical suspicion.

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59

Thyroid cancer

Hannah Nieto and Neil Sharma

Introduction

Thyroid cancer is the most common endocrine cancer worldwide, affecting women three times more frequently than men. The incidence of thyroid cancer has increased significantly over the last three decades, with the associated mortality also increasing, but not as rapidly. This suggests that a number of the newly identified thyroid cancers have a less aggressive phenotype, possibly due to an increase in radiologically detected thyroid cancers, which would have otherwise have not presented clinically. As well as being detected on scanning, thyroid cancers can present as an anterior neck lump, or less commonly, by causing symptoms such as dysphagia, dysphonia, or a pressure sensation in the neck. Thyroid nodules are very common and distinguishing benign from malignant nodules is key to their appropriate management.

Anatomy and physiology of the thyroid gland

The thyroid is a bilobed gland that sits anterior to the trachea, with an isthmus that joins the lobes over the second to fourth tracheal cartilage rings. A pyramidal lobe, projecting superiorly from the isthmus, is a normal anatomical variant. Embryologically, the thyroid arises from a proliferation of endodermal cells at the base of the tongue, and remains connected to the tongue at the foramen caecum during its caudal descent to its final position. The persistence of this tract can present clinically as a thyroglossal duct cyst.

The blood supply to the thyroid is from the superior thyroid artery and the inferior thyroid artery, derived from the external carotid artery and the thyrocervical trunk, respectively. The venous drainage is by the superior and middle thyroid veins, which drain into the internal jugular vein, and the inferior thyroid vein, which drains into the brachiocephalic vein. The thyroid gland sits posterior to the strap muscles—the sternohyoid anteriorly and sternothyroid—and is encased by the pretracheal fascia. The four parathyroid glands sit on the posterior surface of the thyroid gland, with variable positions, although typically the superior gland is superolateral to the recurrent laryngeal nerve (RLN) and the inferior gland anteromedial to the RLN. The inferior glands are more variable in location than the superior glands due to their embryological origin; they arise from the third pharyngeal pouch and travel inferiorly towards the mediastinum alongside the thymus. In contrast, the superior parathyroids originate from the fourth pharyngeal pouch.

The RLN has a close relationship with the thyroid gland, making identification of its intraoperative location a critical step of thyroid surgery. It originates from the vagus nerve; the left RLN loops around the aorta, whereas the right loops around the right subclavian artery, with the right RLN consequently running more obliquely in the neck. The RLN ascends vertically in the tracheo-oesophageal groove behind the thyroid gland, and enters the larynx posterior to the cricothyroid joint, just under the inferior pharyngeal constrictor. The tubercle of Zuckerkandl, a protuberance from the posterolateral surface of the thyroid gland, overlies the RLN.

Thyroid hormone biosynthesis is controlled by the hypothalamic–pituitary axis; the hypothalamus releases thyrotropin-releasing hormone which stimulates the anterior pituitary to release thyroid-stimulating hormone (TSH) which acts upon the thyroid. TSH functions as a growth factor once bound to the TSH receptor on thyroid follicular cells and activates thyroid hormone synthesis by increasing expression of the genes thyroglobulin, thyroid peroxidase, and the sodium–iodide symporter (NIS). Iodide is required in the synthesis of triiodothyronine (T3) and thyroxine (T4) and is taken up into the cells by NIS, which is the mechanism exploited in radioactive iodine therapy.

Thyroid pathology

Thyroid cancers can be categorized into differentiated thyroid cancer, medullary thyroid cancer, and anaplastic thyroid cancer. The most common thyroid cancers are differentiated, which arise from thyroid follicular cells, and include papillary thyroid cancer (70–80% of all thyroid cancers), follicular thyroid cancer (10–15% of thyroid cancers), and Hurtle cell carcinoma. Several additional categories of thyroid tumour have recently been described, including follicular tumour of uncertain malignant potential (FT-UMP), well-differentiated tumour of uncertain malignant potential (WDT-UMP), and non-invasive follicular thyroid neoplasm with papillary nuclear features (NIFTP). While the first two are 'borderline'

neoplasms and compare similarly to carcinoma *in situ* lesions in other tissues, NIFTP is considered benign.

Anaplastic thyroid cancer was previously known as undifferentiated thyroid cancer and is thought to be a progressive dedifferentiation from differentiated thyroid cancer. It usually affects an older population (>65 years) and is fortunately rare (2% of thyroid cancers) as it is aggressive and invasive, and is the least responsive to treatment with an almost universally poor prognosis.

Medullary thyroid cancer (5–10% of thyroid cancers) arises from the parafollicular (C) cells that produce calcitonin, which regulates blood calcium levels. It can be familial, sporadic, and a manifestation of syndromes such as multiple endocrine neoplasia (MEN) types 2a and 2b. Medullary thyroid cancers are associated with *RET* proto-oncogene mutations.

Finally, lymphoma can also arise in the thyroid gland, and is often a differential for anaplastic disease, and rarely metastases from other organs can occur.

Assessment

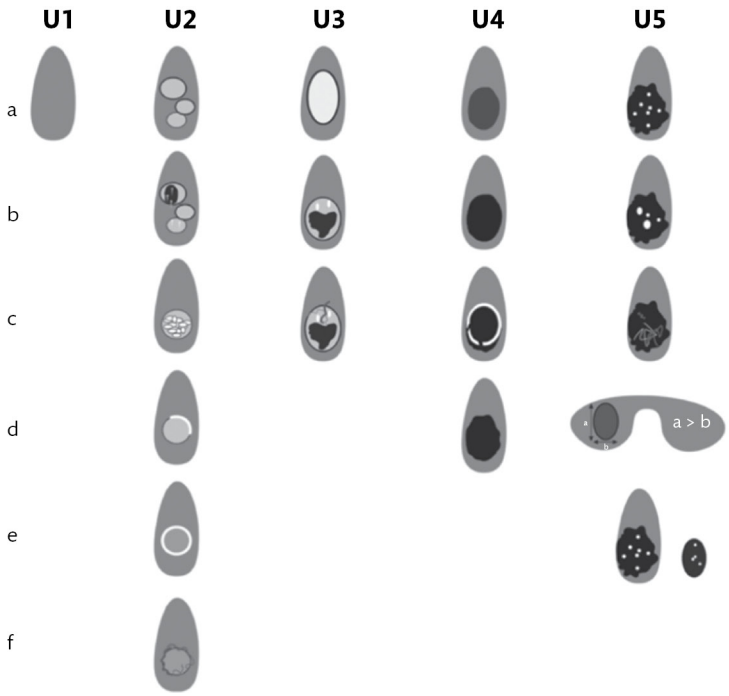
Detailed descriptions of clinical assessment are set out in earlier chapters in Part 4 of this textbook. Usual presenting symptoms include neck swelling, dysphagia, dysphonia, or an incidental finding on imaging. Other features such as sudden growth, pain, haemoptysis, stridor, and other neck lumps should be elucidated. Symptoms of hyper- and hypothyroidism should be addressed. Predisposing factors, such as radiation exposure, iodine deficiency, Gardner syndrome, family history, or known *RET* proto-oncogene mutations, should be noted.

Investigations

Patients should have thyroid function tests and autoantibodies levels assessed. Imaging considerations are discussed in Chapter 69. Ultrasound is key in assessing the thyroid nodule, and sonographers will categorize the nodule on a clinical grading system as per the British Thyroid Association U classification system (Figure 59.1)[1] or the American College of Radiology Thyroid Imaging, Reporting and Data System (TIRADS).[2] Cross-sectional imaging should be arranged for those with suspected retrosternal extension, fixation of the tumour, or haemoptysis.

Fine needle aspiration cytology

Fine needle aspiration cytology (FNAC) grades the thyroid nodule in terms of likelihood of malignancy, and should be performed under ultrasound guidance for all nodules graded as U3, U4, or U5 (Table 59.1).[1] It is an important part of the investigations and helps inform the management of the nodule, from reassurance to diagnostic or therapeutic surgery. All nodules suspicious for cancer (Thy3 and above) or with discordant imaging and cytology should be discussed at the thyroid multidisciplinary team (MDT) meeting. In the presence of hyperthyroidism, FNAC is less accurate and should be deferred until the patient is rendered euthyroid, unless there are other clinical or radiological findings to suggest malignancy.



U1. Normal.

U2. Benign:
(a) Halo, iso-echoic/mildly hyper-echoic
(b) Cystic change +/– ring down sign (colloid)
(c) Micro-cystic/spongiform
(d & e) Peripheral egg shell calcification
(f) Peripheral vascularity.

U3. Indeterminate/Equivocal:
(a) Homogenous, hyper-echoic (markedly), solid, halo (follicular lesion).
(b) ? hypo-echoic, equivocal echogenic foci, cystic change
(c) Mixed/central vascularity.

U4. Suspicious:
(a) Solid, hypo-echoic (cf thyroid)
(b) Solid, very hypo-echoic (cf strap muscle)
(c) Disrupted peripheral calcification, hypo-echoic
(d) Lobulated outline

U5. Malignant:
(a) Solid, hypo-echoic, lobulated/irregular outline, micro-calcification. (? Papillary carcinoma)
(b) Solid, hypo-echoic, lobulated/irregular outline, globular calcification (? Medullary carcinoma)
(c) Intra-nodular vascularity
(d) Shape (taller >wide) (AP>TR)
(e) Characteristic associated lymphadenopathy

Figure 59.1 British Thyroid Association U classification system for ultrasound.

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Table 59.1 British Thyroid Association Thy score for fine needle aspiration

Category	Diagnosis	Risk of malignancy (%)	Management
Thy1	Non-diagnostic	4.5–8.5	Ultrasound assessment and repeat FNAC
Thy2	Non-neoplastic	<3	Correlate with clinical and radiological (ultrasound) findings
Thy2c	Non-neoplastic cystic		
Thy3a	Atypical features not enough to place in another category	10–43	Further investigation, usually ultrasound assessment and repeat FNAC (Thy3a FNAC on repeat sample requires MDT discussion)
Thy3f	Follicular neoplasm		Diagnostic hemithyroidectomy
Thy4	Suspicious of malignancy	68–70	Diagnostic hemithyroidectomy
Thy5	Diagnostic of malignancy	98–100	Therapy appropriate to tumour type, usually surgery for papillary or medullary thyroid carcinomas

Treatment

Thyroid cancer is managed by the MDT, including clinical oncologists, clinical nurse specialists, endocrinologists, nuclear medicine teams, pathologists, radiologists, and surgeons.

Differentiated thyroid cancer

Diagnostic hemithyroidectomy is used in the case of Thy3f or Thy4 lesions, and completion thyroidectomy is indicated in patients with malignancies greater than 4 cm in size, or multifocal disease, bilateral disease, extrathyroidal spread, familial disease, and nodal or distant metastases. Therapeutic surgery will be indicated for Thy5 lesions and may be in the form of hemi- or total thyroidectomy depending on the size of the tumour and the presence of metastases. Consent for total thyroidectomy is outlined in Box 59.1.

Patients with lateral neck disease should have therapeutic lateral neck dissection (levels II–V) and central neck dissection. Central neck dissection in differentiated thyroid cancer is much debated and is explored further in the 'Controversies' section below. It is generally indicated in high-risk disease and can be omitted if the patient is under 45 years of age, with a unifocal tumour less than 4 cm in size, with classical type papillary thyroid cancer, and no extrathyroidal spread.

Recurrent or residual disease should be primarily managed surgically, if amenable to it, otherwise radioactive iodine can be considered.

Papillary thyroid microcarcinomas are differentiated thyroid cancers less than 10 mm in size and if they are solitary lesions with no adverse features, they can be managed with a hemithyroidectomy and no TSH suppression. However, if there is multifocal or bilateral disease then personalized decision-making, via the MDT, needs to be implemented—this can be completion thyroidectomy, monitoring, or follow-up. Similarly, for pT2 tumours without any high-risk features, there is a role for therapeutic hemithyroidectomy, as a less aggressive approach becomes more established, reflected in the American Academy Guidelines update in 2015.[3]

After total or completion thyroidectomy the patient should be started on 2 mcg/kg levothyroxine once a day with the aim of fully suppressing their TSH. This is continued until at least 1 year following radioiodine and treatment response is assessed.

Box 59.1 Consent for total thyroidectomy

Risks, benefits, and alternatives to be discussed, including the risks: infection, bleeding (haematoma/return to theatre), scar, numbness, damage to RLN (voice damage), damage to superior laryngeal nerve (voice change), low calcium/calcium replacement, lifelong T4 replacement, tracheostomy, and general anaesthetic risks.

Radioiodine

Decisions about radioactive iodine treatment are made on an individual basis but those with a large tumour (>4 cm), extrathyroidal extension, or distant metastases should have radioactive iodine (Table 59.2). Historically, patients would be on T3 postoperatively as it has a shorter half-life than T4 and would have this withdrawn before their radioactive iodine therapy, but this has been almost universally replaced by the administration of recombinant human TSH prior to therapy, which allows the patient to continue on levothyroxine up until the point of treatment, and avoids the effects of thyroid hormone withdrawal.

The dose of radioactive iodine therapy was adjusted in the HiLo[4] and the ESTIMABL1[5] trials, both concluding that the lower dose of

Table 59.2 Indications for radioactive iodine in differentiated thyroid cancer

Radioactive iodine	Clinical details
Definite I^{131} ablation (any criteria)	Tumour >4 cm
	Extrathyroidal extension
	Distant metastases
Probable I^{131} ablation	Unfavourable histology (tall cell, columnar, sclerosing, poorly differentiated elements)
	Widely invasive histology
	Multiple lymph nodes
No I^{131} ablation (all criteria)	Tumour <1 cm
	Favourable histology (classical papillary, follicular variant papillary, follicular)
	Minimally invasive (no angioinvasion)
	No extrathyroidal extension

1.1 GBq had a similar post-ablation success rate to the standard high dose of 3.7 GBq.[6] These trials also confirmed the safe use of recombinant TSH in replacement of withdrawal therapy. Further studies have since confirmed that the lower-dose radioactive iodine does not affect thyroid cancer recurrence rates.

After radioactive iodine TSH should be suppressed to less than 0.1 mIU/L, using a supraphysiological dose of T4, until a dynamic risk assessment is undertaken at 9–12 months post-treatment.

Follow-up

Following radioactive iodine, at 9–12 months post-treatment dynamic risk stratification should occur, which incorporates results from suppressed and stimulated thyroglobulin, neck ultrasound scanning, and any cross-sectional or nuclear medicine scans if they are performed (Table 59.3). This categorizes patients as low, intermediate, or high response based on these findings, and the amount of TSH suppression required is calculated from this (Table 59.3). This is important, as previously post-treatment thyroid cancer patients had long-term TSH suppression therapy, using supraphysiological doses of levothyroxine to reduce the risk of recurrence. Subclinical hyperthyroidism does, however, have side effects, including increased cardiovascular risks, osteoporosis, and an association with decreased cognitive function, and dynamic risk stratification allows for the relaxation of this in patients for whom the risk of recurrence is very low.

Medullary thyroid cancer

Preoperatively, all patients with suspected medullary thyroid cancer should be discussed in the thyroid MDT meeting and managed in a thyroid cancer centre. A detailed family history is important, as well as exploring for any history of endocrinopathies. Any patients with suspected medullary disease should have blood tests for calcitonin and carcinoembryonic antigen. Calcitonin levels can be helpful to confirm the diagnosis, and can be used as a reference baseline to compare postoperative levels. All patients should have plasma nor/metanephrines (and/or urinary catecholamines) checked before undergoing surgery due to the syndromic association with phaeochromocytoma, to ensure the general anaesthetic is safe. Postoperatively, *RET* gene sequencing should occur, although this will need to be discussed with the patient as it has implications for them and their family, who may want genetic counselling and screening. Families also need to be counselled about the role of prophylactic surgery.

The management of medullary thyroid cancer is predominantly surgical, and patients should undergo a total thyroidectomy and central neck dissection. If there is suspicion of locally advanced disease, preoperative computed tomography scanning of the neck (or magnetic resonance imaging) is recommended. In the presence of lateral neck disease, bilateral neck dissection (levels II–V plus levels VI–VII) is indicated; central compartment neck disease requires elective ipsilateral neck dissection of levels II–V.[7] Even in the context of distant metastases, the primary management of medullary thyroid cancer is surgical, with control of the neck disease to prevent local progression and airway symptoms, as patients will often have a prolonged survival even with the metastases.

For disease-free family members with germline *RET* gene mutations, prophylactic total thyroidectomy is recommended. For MEN 2B syndrome this is recommended to occur in the first year of life, and MEN 2A (with codon 634 mutation) before the age of 5 years.[1] Those with familial medullary thyroid cancer or other mutations associated with MEN 2A can have surgery delayed until after 5 years old, but the aim should still be to perform this prophylactically (rather than for disease) as this improves patient outcomes/survival. All should have preoperative basal calcitonin checked, and if this is raised preoperatively then additional prophylactic neck dissection will be considered.

Medullary thyroid cancer patients should be followed up with serial calcitonin and carcinoembryonic antigen measurements from 6 months post-treatment onwards. Rises should be investigated with ultrasound scanning and cross-sectional imaging, usually computed tomography scans of the neck and chest and magnetic resonance imaging of the liver. Calcitonin doubling times are useful prognostic indicators. For unresectable disease, palliative radiotherapy can play a role, and targeted therapies including tyrosine kinase inhibitors have been used but not shown any survival benefit.

Anaplastic thyroid cancer

Anaplastic thyroid cancer is thought to be due to dedifferentiation of differentiated thyroid cancer, with an accumulation of mutations in genes such as *TP53*, *RAS*, *BRAF*, *PIK3CA*, and *PTEN*. Patients often present with advanced disease, with tracheal invasion and oesophageal invasion, and carotid artery involvement is frequently seen. Distant metastases are also often present on presentation. Patients should be investigated with an ultrasound FNAC/core biopsy and computed tomography scans of the neck, thorax,

Table 59.3 Post-treatment dynamic risk stratification

Excellent response	Indeterminate response	Incomplete response
All the following:	**Any of the following:**	**Any of the following:**
Suppressed and stimulated Tg <1 mcg/L	Suppressed Tg <1 mcg/L and stimulated Tg ≥1 and <10 mcg/L	Suppressed Tg ≥1 mcg/L or stimulated Tg ≥10 mcg/L
Neck ultrasound without evidence of disease	Neck ultrasound with non-specific changes or stable sub-centimetre lymph nodes	Rising Tg values
Cross-sectional and/or nuclear medicine imaging negative (if performed)	Cross-sectional and/or nuclear medicine imaging with non-specific changes, although not completely normal	Persistent or newly identified disease on cross-sectional and/or nuclear medicine imaging
Low risk: Maintain TSH 0.3–2.0 mIU/L	**Intermediate risk:** Suppress TSH 0.1–0.5 mIU/L for 5–10 years then reassess	**High risk:** Suppress TSH <0.1 mIU/L indefinitely

Tg, thyroglobulin; US, ultrasound.

and abdomen to stage the disease. Appropriate support and interventions may be needed for compromised airway and swallowing (see Chapter 66). Tracheostomy (see Chapter 70) can be technically difficult due to anterior neck disease and will often only prolong life in a palliative setting, so patients and families should be adequately counselled on the risks of conservative and surgical airway management. Thyroidectomy should only be performed if an R0 or R1 resection can be achieved, and patients will need postoperative chemoradiotherapy too. Targeted treatment for patients with BRAFV600E mutations include Dabrafenib and Trametinib, and patients should undergo sequencing (as per national test directory) based on the mutational profile. Further treatments (currently consigned to clinical trials) might be available.

Controversies

Prophylactic central neck dissection

The central neck compartment is bound by the hyoid bone (superiorly), carotid arteries (laterally), and deep layer of cervical fascia (posteriorly) and a central neck dissection clears all nodal disease from this region. There is an associated morbidity with central neck dissection, with an increased risk of transient hypoparathyroidism, and higher risk of transient RLN injury. It is generally accepted that patients with large, differentiated thyroid cancer, extrathyroidal extension, or lymph node metastasis will benefit from a central neck dissection, as there is a documented improvement in disease-free survival. However, in those with smaller (<2 cm) and low-risk disease, the benefit is less clear cut.[8] It also may cause up-staging of low-risk disease (detection of micrometastases), causing patients to be indicated for radioactive iodine where they otherwise would not, the benefit of this remaining unclear. Further controversy arises regarding unilateral or bilateral central neck dissection. For now, for small, low-risk tumours, patient-specific decision-making is recommended dependent on both tumour and patient factors.

Active surveillance

Some thyroid cancers progress very slowly, or not at all; therefore, active surveillance for small, low-risk thyroid cancers can be considered.[9] Active surveillance is a safe option in papillary microcarcinomas (T1a; <1 cm). For low-risk T1b thyroid cancers (1–2 cm), emerging evidence suggests that active surveillance can be a safe option, with no significantly increased risk of cancer-related deaths or recurrence after delayed surgery.[3,10] With this approach, several patients will avoid the morbidity associated with surgery. However, acceptability of this to thyroid cancer patients (and surgeons) remains a significant concern. Long-term outcomes will help build confidence in the appropriateness of active surveillance for T1b tumours.

Alternative management of papillary microcarcinoma

Minimally invasive therapeutic options for thyroid nodules are available in some centres. These are generally ultrasound-guided interventions and include thermal ablation, radiofrequency ablation, and ethanol ablation as alternatives to surgery. Thermal ablation causes coagulative necrosis and has been demonstrated as effective in papillary thyroid microcarcinoma.[11] Data are emerging on the use of the other techniques, such as radiofrequency ablation in a therapeutic cancer setting, albeit solely for papillary microcarcinoma.[12] In a disease such as papillary microcarcinoma, it is difficult to evaluate the comparative effectiveness of active surveillance; minimally invasive therapy and surgery as the outcomes are generally excellent (with a low frequency of events such as recurrence/progression). However, a safe, less invasive, alternative to surgery for patients with papillary microcarcinoma would be a welcome therapeutic option.

Conclusion

Thyroid cancer is a predominantly surgically managed disease but is unique in the availability of targeted radiation in the form of radioactive iodine treatment. Recurrent and residual disease is much harder to treat than primary disease.

FURTHER READING

Guidelines

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60

Hypopharyngeal cancer

Laura Warner and Kee Wong

Introduction and epidemiology

With a global annual incidence of approximately 84,000, hypopharyngeal cancer is considered a rare malignancy and represents 0.4% of all cancer diagnoses. It comprises 2% of head and neck cancers in the UK with an age-standardized incidence rate of 0.63/100,000 in England. Since 2012, the annual worldwide incidence has increased by more than 20,000 cases per year.

Hypopharyngeal cancer is more common in men than in women, although the relative incidence in women has increased in recent years. More than 95% of hypopharyngeal cancers are squamous cell carcinomas. Known aetiological factors include tobacco and alcohol use, which have a synergistic effect, and betel nut and areca nut use are also contributory factors in developing countries.

Hypopharyngeal malignancies carry a poor prognosis with 5-year survival of approximately 30% for all stages (**Box 60.1**). The poor prognosis is related in part to the significant proportion of patients (>85%) who present with advanced disease. In addition to this, hypopharyngeal tumours have a high propensity for extensive nodal involvement (75% of patients have a palpable neck node at presentation) and extranodal extension is frequently encountered. Distant metastases are commoner in hypopharyngeal cancers than in other head and neck malignancies. Furthermore, hypopharyngeal cancers have the highest rates of synchronous and metachronous malignancies. The age at diagnosis typically ranges from 40 to 80 years, with median ages of 55 years for males and 60 years for females. Many patients have significant comorbidities and nutritional deficiencies at presentation which, as a result, negatively impacts treatment choices and survival.

Box 60.1 Reasons for poor prognosis in hypopharyngeal cancer

- Advanced disease at presentation in 85%.
- High propensity for extensive nodal involvement: 75% palpable node at presentation, 5% N2c+.
- Extranodal extension common.
- Frequent distant metastasis.
- High rates of synchronous and metachronous cancers.
- Advanced age and comorbidity status.
- Nutritional deficiencies.

Prognosis for those who present with early-stage disease is much improved at approximately 60%, although radical treatment regimens are often still espoused for early-stage disease, resulting in long-term treatment-associated morbidity, including post chemoradiation laryngopharyngeal dysfunction with tracheostomy and/or enteral feeding tube reliance or natural voice loss due to laryngopharyngectomy. Treatment paradigms for hypopharyngeal disease are typically based upon results from trials of advanced laryngeal cancers, although regrettably this extrapolation of results is unlikely to be entirely appropriate for guidance of treatment regimens due to the differing nature of the two diseases.

While early death is often related to second primary cancers or comorbidities, advances in the treatment of metastatic disease are improving prognosis in selected patients. Furthermore, with recent advances in both surgical and oncological primary treatments for head and neck cancers, improvements in overall prognosis and reduced treatment-related morbidity are becoming evident for this challenging disease.

Aetiology

Tobacco use and alcohol consumption are the main risk factors for hypopharyngeal malignancy (**Box 60.2**). Both factors increase the risk of head and neck cancers in isolation; however, they carry a synergistic effect which is particularly evident in hypopharyngeal disease where a high proportion of patients exhibit these risk factors at presentation compared to other head and neck subsites. Betel quid chewing is another significant independent risk factor

Box 60.2 Risk factors for hypopharyngeal cancer

- Smoking.
- Alcohol.
- Betel nut.
- Diet.
- Paterson–Brown Kelly syndrome.
- Lower socioeconomic status.
- Previous head and neck irradiation.
- Human papillomavirus.

for hypopharyngeal cancer with an effect similar in risk profile to alcohol use and further synergistic effect when combined with tobacco and/or alcohol use.

Dietary factors may also increase the risk of hypopharyngeal malignancy with data from India suggesting that a diet rich in fruit and leafy green vegetables may confer a protective effect in both patients with and without a history of tobacco use. There is a known association between post-cricoid carcinoma and iron deficiency anaemia in patients with Paterson–Brown Kelly (Plummer–Vinson) syndrome, with an increase in risk between 4% and 16%. Correction of iron deficiency anaemia in this condition may reduce the risk of malignancy developing.

Other identified risk factors include previous head and neck irradiation and human papillomavirus, although the significance of human papillomavirus-associated hypopharyngeal disease is significantly less than in oropharyngeal cancer. Socioeconomic status and occupation also influence the risk of developing hypopharyngeal cancer, independent of tobacco and alcohol use.

Hypopharyngeal anatomy

The hypopharynx, also known as the laryngopharynx, is the caudal portion of the pharynx, positioned posterior and lateral to the larynx, between the oropharynx superiorly and the oesophagus inferiorly. It is a triangular space bounded by the aryepiglottic folds superiorly and the upper oesophageal sphincter inferiorly.

The subsites of the hypopharynx are the two piriform fossae (also known as piriform sinuses), the posterior hypopharyngeal wall, and the post-cricoid space. Hypopharyngeal cancers are most commonly located in the piriform fossa (65–80%), with 10–20% affecting the posterior pharyngeal wall and 5–15% in the post-cricoid area.

The hypopharynx is lined by stratified squamous epithelium, surrounded by the inferior constrictor muscles (thyropharyngeus and cricopharyngeus) which are innervated by the pharyngeal plexus with supply from the glossopharyngeal (IX) and vagus (X) cranial nerves. Arnold's nerve, a branch of the vagus nerve, also supplies the external auditory canal, which explains the otalgia frequently experienced by patients with hypopharyngeal malignancy.

Hypopharyngeal physiology

The role of the hypopharynx in swallowing physiology is discussed in detail in Chapter 48. Hypopharyngeal cancers may disrupt any part of the complex swallowing mechanism and therefore dysphagia and/or odynophagia are common presenting symptoms. Vocal fold immobility may be evident, resulting in aspiration, dysphonia, and stridor. Regrettably, swallowing dysfunction frequently occurs as a result of both surgical and oncological treatments for hypopharyngeal cancer.

Assessment and investigations

A comprehensive assessment of the entire upper aerodigestive tract is warranted. The principles of clinical assessment and detailed description of examination of these areas can be found in Chapters 46–50, and appropriate radiological workup for suspected head and neck malignancies is discussed in Chapter 69.

Box 60.3 Key symptoms of hypopharyngeal cancer

- Dysphagia.
- Odynophagia.
- Throat pain—persistent and unilateral.
- Otalgia.
- Unintended weight loss.
- Aspiration.
- Haemoptysis.
- Stridor.

Direct endoscopic assessment under general anaesthetic is the usual means of obtaining a pathological diagnosis. This is also particularly useful if there is a clinical or radiological suspicion of hypopharyngeal malignancy without a visible mass on fibreoptic endoscopy, as areas such as the piriform apex and post-cricoid space may not be readily visualized during fibreoptic endoscopy, and offers thorough tumour assessment that is required for treatment planning. Transnasal oesophagoscopy with endoscopic biopsy or channelled laryngoscopy for biopsy may facilitate biopsies of visible masses in the outpatient setting and is particularly helpful in patients in whom general anaesthesia poses a high risk (**Boxes 60.3** and **60.4**).

Differential diagnoses

Differential diagnoses of hypopharyngeal malignancy include the following:

- *Malignancy*: involvement of other aerodigestive tract sites—oropharynx, larynx, and upper oesophagus.
- *Pharyngeal pouch, pharyngeal or upper oesophageal stenoses*: the presence of throat pain and odynophagia in a patient with dysphagia should be considered suspicious.
- *Infective (viral/bacterial/fungal)*: if symptoms are persistent, progressive, and do not respond to simple treatments, a hypopharyngeal malignancy should be ruled out. Many patients diagnosed with hypopharyngeal cancers have been initially treated with antibiotics or antifungals for suspected *Candida* infection.
- *Globus pharyngeus*: this is discussed in Chapter 48.

Box 60.4 Examination findings in hypopharyngeal cancer

- Neck mass.
- Visible hypopharyngeal mass.
- Mucosal ulceration or oedema.
- Vocal fold immobility.
- Pooling of secretions in piriform fossae (Chevalier Jackson's sign).
- Loss of laryngeal crepitus.
- Loss of laryngeal elevation upon swallowing.

Treatment

After workup is complete, every case should be discussed at a head and neck multidisciplinary team meeting; the role of this team, from diagnosis to follow-up, in managing head and neck cancer patients is set out in Chapter 46. This enables individualized treatment planning, taking into account tumour factors, patient factors including social circumstances, as well as patient preference.

Available treatments include radiotherapy, with or without concurrent chemotherapy, endoscopic or minimally invasive surgery, major head and neck surgery, and palliative measures. Combined treatment with surgery and (chemo)radiotherapy is often employed for advanced disease whereas early-stage disease may be managed with single-modality treatment.

Surgical management

Minimally invasive techniques such as transoral laser microsurgery or transoral robotic surgery offer organ-preservation surgery with equal efficacy to oncological treatments for early-stage disease. One advantage to surgery is the ability to pathologically stage the disease, offering prognostic information such as perineural or lymphovascular invasion and extranodal extension. Careful case selection is paramount to ensure adequate resection as the intention of surgical management in early-stage disease is to avoid adjuvant (chemo)radiotherapy, while minimizing swallowing morbidity. These techniques are ideal for resection of small lesions, limited to one subsite (Tis and T1). Suitability for transoral laser microsurgery or transoral robotic surgery should be carefully assessed by the operating surgeon at diagnostic endoscopy.

While resection of more extensive T2 lesions of the piriform fossa and posterior pharyngeal wall lesions may be feasible, these can cause unacceptable swallowing morbidity due to effacement of the piriform fossa, resulting in aspiration; posterior hypopharyngeal wall resection can cause aspiration through anterior diversion of the bolus and narrowing of the post-cricoid area due to oedema and stricture formation. These risks should be carefully balanced against the sequalae of non-surgical treatments and discussed with the patient.

Endoscopic access to the hypopharynx may be achieved with distending diverticuloscopes (Figure 60.1) and the Feyh–Kastenbauer retractor with hypopharyngeal blades (Figures 60.2 and 60.3) can offer access to the more inferior areas of the hypopharynx.

Figure 60.1 Weerda diverticuloscope.

Figure 60.2 Karl Storz Steiner distending laryngoscope.

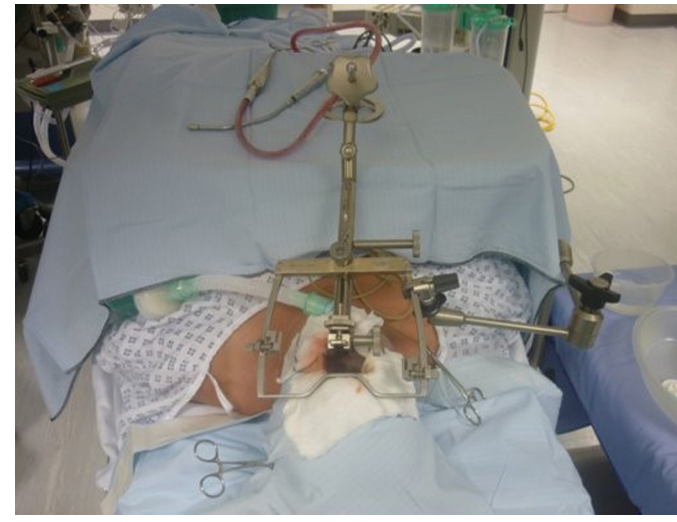

Figure 60.3 Feyh–Kastenbauer retractor in place.

When endoscopic access is problematic, open techniques via a lateral pharyngotomy approach or partial laryngectomy, with or without reconstruction, may also be considered for early-stage disease. However, in such cases non-surgical treatments are usually favoured, when possible, as the swallowing outcomes from open surgery for hypopharyngeal cancers remain unpredictable.

Advanced disease usually requires multimodality treatment, often with surgery and adjuvant (chemo)radiotherapy. Total laryngectomy with partial or total pharyngectomy is the mainstay of surgical treatment for advanced hypopharyngeal cancer.

Organ preservation with radiotherapy and concomitant chemotherapy is the preferred approach for T3 lesions. However, in cases where pretreatment laryngeal compromise is evident with aspiration, poor swallow, stridor, or dyspnoea, primary surgery should be considered. In patients with pre-existing laryngopharyngeal dysfunction, it is widely believed that functional outcomes are better with a primary surgical approach and adjuvant treatment. In such cases, the loss of organ function with loss of the natural voice is balanced against retaining some swallowing ability and the stability of a laryngectomy airway versus a long-term tracheostomy. These are difficult choices to make, and patients should be supported in this decision-making by the whole multidisciplinary team. For patients with T4a disease, complete response from oncological treatments is unlikely and pharyngolaryngectomy is frequently required.

High rates of submucosal extension in hypopharyngeal cancers necessitate wide resection (with recommended macroscopic clinical margins of 1.5 cm superiorly, 2 cm laterally, and 3 cm inferiorly) to ensure adequate clearance. Particular care must be taken to ensure deep soft tissue margins are not compromised when the thyroid lamina is breached by disease.

The dimensions of the pharyngeal remnant after resection dictate the method of reconstruction. The amount of remaining pharynx is rarely sufficient for primary closure, meaning pharyngeal reconstruction is often necessary to avoid tight neo-pharyngeal closure, risking pharyngocutaneous fistula and neo-pharyngeal stricture.

Reconstructive principles are discussed in detail in Chapter 64. Issues relevant to hypopharyngeal cancer alone are set out in brief here. Pedicled flaps such as the pectoralis major myocutaneous or myofascial flaps and the supraclavicular flap offer options for lesions where a small patch is required to augment the neo-pharynx. Free flaps such as the radial forearm, the anterolateral thigh, or medial sural artery perforator free flaps are useful for larger patch reconstruction for partial pharyngectomy defects. Where total pharyngectomy with circumferential resection is required, a tubed cutaneous free flap (usually anterolateral thigh due to the size of skin paddle required) or jejunal flap are the preferred choices. Tubed cutaneous flaps should be designed to minimize stricture formation; however, significant dysphagia due to neo-pharyngeal stricture is commonplace following a tubed cutaneous pharyngeal reconstruction. Gastro-omental free flaps offer robustly vascularized tissue that is useful in a previously irradiated field, although the procedure has added morbidity from abdominal surgery. For inferior hypopharyngeal disease involving the upper oesophagus, where reconstruction may extend inferior to the clavicles, pharyngo-laryngo-oesophagectomy with gastric pull-up may be required.

Surgical management of the neck with neck dissection is mandated for N+ cases and for N0 cases in anything other than Tis lesions treated with primary surgery due to the high propensity for nodal disease in hypopharyngeal cancer. Levels II–IV should be included as a minimum, with consideration of central compartment clearance (level VI) in cases where there is extralaryngeal spread into the anterior soft tissues or significant subglottic involvement. Ipsilateral neck dissection may suffice for N0/N1 cases where the disease is well lateralized (a minority), whereas bilateral neck dissection is required for N2+ or primary disease approaching the midline.

Oncological management

A detailed discussion of the principles of non-surgical treatment can be found in Chapter 65. Radiotherapy has become the preferred radical treatment for early-stage hypopharyngeal (T1/T2) cancer due to organ and function preservation. It offers high local control rate of around 80% for T1–T2 tumours with surgical salvage being reserved for treatment failure. As hypopharyngeal tumours are characterized by high risk of lymph node metastases, elective irradiation of regional lymph nodes is required, even in radiologically N0 patients. Lymphatic drainage of hypopharynx is to bilateral levels II–IV and retropharyngeal nodes, so these levels are always included in the radiation field.

For locally advanced disease, which is often the case at presentation for hypopharyngeal cancer (i.e. T3/T4 and/or N+), treatment choice depends upon extent of disease and patient preference, acknowledging the balance between possible cure, treatment morbidity, and likely functional outcomes. Phase III data exist to support the use of induction chemotherapy to stratify patients to either surgical intervention or chemoradiation based on response. For locally advanced disease, concurrent platinum-based chemotherapy is indicated in those aged 70 or under as it confers additional overall survival advantage. For patients who are treated with primary surgery, the role of adjuvant (chemo)radiotherapy is determined by postoperative histopathological findings. Indications include T3/T4 tumour, close or positive resection margin, pN2/N3, and those with extranodal extension (**Figure 60.4**).

Salvage surgery

Recurrence after radiotherapy or incomplete response to oncological treatment may be surgically salvaged if the patient's performance status allows and the patient is willing to accept the high risk of complications following salvage surgery. Resection must be radical with wide mucosal margins, as close or involved margins are likely to result in early further recurrence, negating any benefit from surgery. This may not apply posteriorly, where the prevertebral fascia is robust and rarely invaded by disease. Reconstruction is almost always needed, and options such as the gastro-omental flap may help to revascularize the soft tissue of the neck in these instances.

Best supportive care

Approximately 25% of patients with hypopharyngeal malignancy are not suitable for curative treatment at presentation, due to either age and comorbidities or the extent of disease. For patients unsuitable for radical treatment and those with recurrent or metastatic disease, palliative treatment can be offered to improve symptoms and prolong survival. Systemic options include platinum-based doublet chemotherapy and immunotherapy, that is, anti-PD1 checkpoint inhibitors. Pembrolizumab and nivolumab are current immunotherapy options available in the UK, in the first- and second-line setting respectively. Short-course palliative radiotherapy can be offered

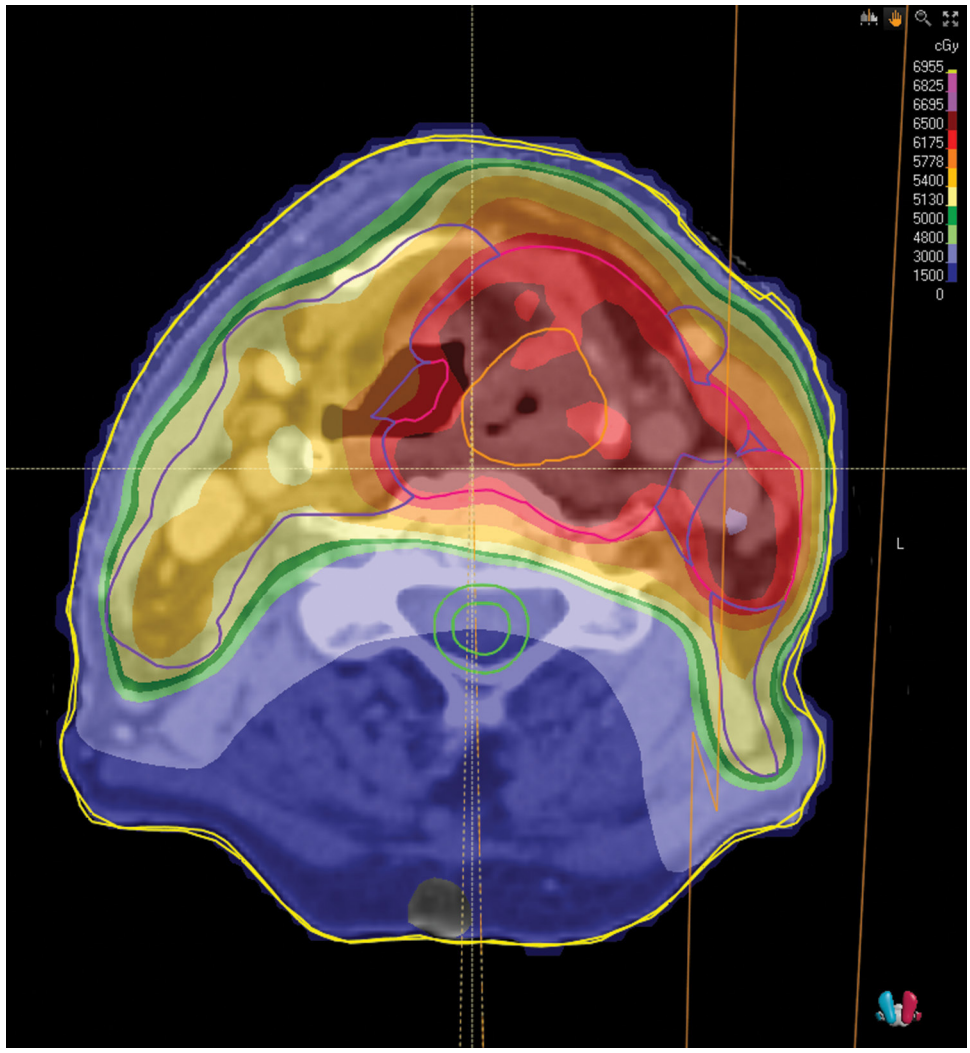

Figure 60.4 Example of volumetric arc radiotherapy plan for a patient with T3N2bM0 left piriform fossa squamous cell carcinoma. The macroscopic tumour plus margin received 65 Gray in 30 fractions whereas uninvolved lymph nodal levels received 54 Gray.

to help with pain and bleeding. The role of surgery in palliation is controversial as pharyngolaryngectomy surgery is a major undertaking, particularly when reconstruction is required as healing is likely to be significantly compromised in the presence of metastatic disease. Palliative neck surgery may be considered for painful or fungating neck disease (Box 60.5).

Guidelines

The National Institute for Health and Care Excellence published a guideline entitled 'Cancer of the upper aerodigestive tract: assessment and management in people aged 16 and over' in 2016, and the United Kingdom National Multidisciplinary Guidelines on hypopharyngeal cancer were also published in 2016.

Box 60.5 Prognosis for hypopharyngeal cancer

- Overall 5-year survival 30%.
- Stage I and II 5-year survival 60%.
- 50–60% of patients experience recurrence within 12 months of treatment.

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61

Nasopharyngeal cancer

Shreerang A. Bhide and Pablo Nenclares

Introduction

Nasopharyngeal cancer (NPC) is an epithelial malignant neoplasm arising from the nasopharyngeal mucosal lining. Despite originating in the head and neck region, NPC is distinct from other epithelial head and neck tumours in biology, epidemiology, histology, natural history, and response to treatment. These differences justify a different approach for management of these neoplasms.

With an age-standardized incidence rate around 0.4/100,000 population in the UK, NPC is a relatively uncommon tumour. However, its geographical global distribution is extremely unbalanced, with more than 80% of the new cases diagnosed in Eastern and South-Eastern Asia where the incidence rate is as high as 8/100,000 people. The incidence of NPC is two- to threefold higher in males compared to females.[1] The distinct racial and geographical distribution is reflective of its multifactorial aetiology.

Pathology and risk factors

According to the World Health Organization (WHO), NPC compromises three histological types[2]: keratinizing squamous, non-keratinizing (divided into differentiated and undifferentiated subtypes), and basaloid squamous carcinomas (**Box 61.1**). Non-keratinizing carcinoma is by far the most common type in both endemic and non-endemic areas, and it is predominantly associated with Epstein–Barr virus (EBV) infection.[3]

EBV is the most common causal agent of NPC although other risk factors including host genetics and environmental factors have been shown to contribute to the development of NPC. High consumption of salted fish in southern Asia has been implicated as a possible aetiological factor in these areas. Other potential environmental factors include alcohol consumption, tobacco smoking, and poor oral hygiene.[4] Genetic factors have also recently been identified. Variations in the human leucocyte antigen region on chromosome 6p21 have been recognized as a major risk locus conferring NPC risk.[5] However, it remains unclear whether this impacts development of NPC directly or by affecting the immune response to EBV. In addition, loss-of-function mutations in the NF-κB negative regulators as well as genetic lesions including the loss of the *CDKN2A/CDKN2B* locus, *CCND1* amplification, P53 and PI3K/MAPK mutations, and DNA damage repair alterations, have been associated with the development and progression of disease.[6,7]

The association of EBV and NPC has led to the hypothesis that serological testing or plasma EBV DNA detection may be an attractive screening strategy to identify early-stage disease in endemic areas. Real-time polymerase chain reaction of circulating EBV DNA in plasma has shown to be a promising screening tool for high-risk populations, potentially aiding diagnosis at an earlier stage.[8]

Box 61.1 Histological classification (WHO 2005)

- Keratinizing squamous cell carcinoma (WHO type I).
- Non-keratinizing carcinoma—this is subdivided into:
  - differentiated type (WHO type II).
  - undifferentiated type (WHO type III).
- Basaloid squamous cell carcinoma.

Patterns of spread

NPC typically arises in the lateral pharyngeal recess (fossa of Rosenmüller) and can spread extensively to adjacent structures (**Figure 61.1**). It tends to spread submucosally with early infiltration of the palatal muscles within the parapharyngeal space. The tumour spreads through areas of lesser resistance of the pharyngobasilar fascia and infiltrates along neural pathways and base of skull foramina. Anteriorly, NPC may easily extend to the nasal fossa and from there to the pterygopalatine fossa via the sphenopalatine foramen, and subsequently into the foramen rotundum or through the orbit, both eventually resulting in intracranial invasion. In addition, the disease can also spread through the foramen lacerum and reach the cavernous sinus. Laterally, spread to the parapharyngeal space can occur very frequently. From there, it can invade the infratemporal fossa or extend perineurally along the mandibular nerve into the foramen ovale and upwards into the cavernous sinus. Posterolateral extension to the retropharyngeal space and prevertebral muscles, destruction of basisphenoid and clivus, and involvement of the

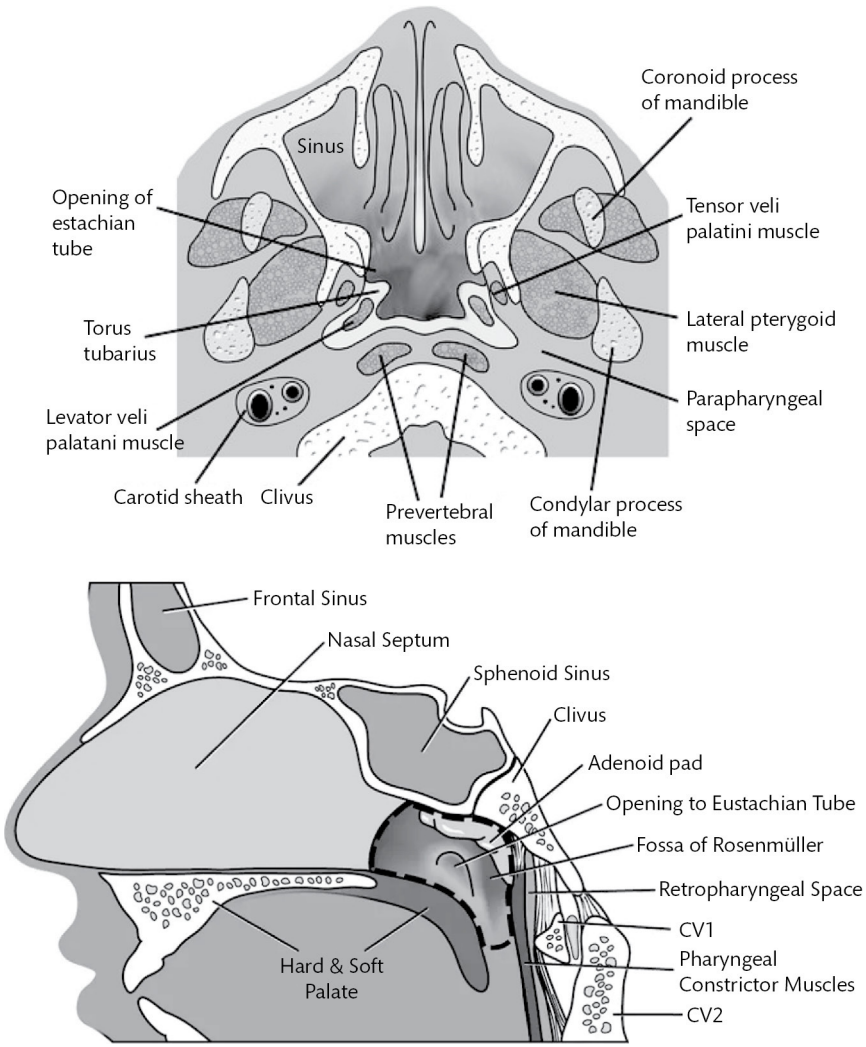


Figure 61.1 Anatomical relations of the nasopharynx important for local patterns of spread of NPC.

jugular foramen and hypoglossal canal, can be seen in very advanced tumours. Inferiorly, NPC extend can into the oropharynx via submucosal spread.[9]

Neck lymphadenopathies are present in up to 90% of patients. The retropharyngeal is the most commonly involved nodal chain, with the majority of cases involving the lateral retropharyngeal group. The frequency of nodal metastases in the jugular lymph node chains decrease in the cranio-caudal direction (from level II to IV). Posterior cervical nodes (level V) are commonly affected and usually accompanied with multinodal involvement in other levels.[10]

Haematogenous spread is not uncommon, with distant metastases occurring at initial diagnosis in around 4% and in around a quarter of patients after definitive treatment in the era of intensity-modulated radiotherapy (IMRT). Bone is the most common site of metastases followed by lung and liver.[11]

Symptoms

The clinical signs and symptoms of NPC are correlated with the involved anatomical regions and lymphatic spread. Unilateral or bilateral neck lumps is one of the commonest presenting symptoms. Local growth can cause epistaxis, nasal obstruction, and discharge. Eustachian tube dysfunction due to obstruction of its orifice can lead to conductive hearing loss and tinnitus. When tumour invades the base of skull or extends intracranially, it can cause cranial nerve neuropathies (the Vth and VIth nerves are most commonly affected). Lower cranial nerves IX, X, XI, and XII could also be involved as they emerge from the skull base to the parapharyngeal space. Headaches or pain in the temporal or occipital regions can occur secondary to skull base destruction.[12]

Paraneoplastic syndromes are very rare, but dermatomyositis has been described in patients from endemic areas. Systemic symptoms including fatigue, loss of appetite, and weight loss are late symptoms which may suggest distant metastases.[13]

Initial assessment and investigations

Clinical assessment

The initial clinical evaluation of patients with head and neck diseases is discussed in Chapter 46. If a neck lump is present, this can

often be used to conform the diagnosis, with evaluation described in Chapter 50. Nutritional and dental status assessments are indicated, especially prior to the start of radiotherapy (RT).[14] Imaging considerations are discussed in Chapter 69 and further reading in the references.[17–19]

Pathology

Histological examination is required for the definitive diagnosis and is discussed in Chapter 68. All NPC subtypes share morphological and immunohistochemical features of squamous differentiation to varying degrees. The histological features of WHO type I are similar to well-differentiated squamous carcinoma. By contrast, WHO types II and III often exhibit a dense lymphoid component, from which the synonym lymphoepithelial carcinoma is derived. Immunohistochemistry is used to identify the production of keratin intermediate filaments and the use of antibodies AE1/AE3 and MNF116 helps to detect a broad of keratin molecules that support the diagnosis of carcinoma. In addition, cytoplasmic expression of cytokeratins 5/6 and nuclear expression of p63 can be used as evidence of squamous differentiation.[3] The presence of EBV in the biopsy sample is detected using hybridization for EBV-encoded early RNA (EBER). Serological markers of EBV infection have limited diagnostic utility. However, plasma EBV DNA quantification using polymerase chain reaction techniques represents a promising strategy that could help in the diagnostic risk stratification, treatment adaption, and disease surveillance.[15,16] Assay standardization and validation, alongside with technical optimization, are warranted before routine use of this biomarker in the clinic.

Staging and prognosis

NPC is classified according to the International Union Against Cancer/American Joint Committee on Cancer (UICC/AJCC) tumour–node–metastasis (TNM) staging system. Its last update to the eighth edition in 2016 incorporated some changes in the primary and nodal disease classification.

In the era of modern RT techniques, early disease (stages I–II) has an excellent chance of cure (5-year local failure-free and overall survival (OS) rates being 94–100% and 91–100%, respectively).[17,18] For locally advanced stages (III–IVA), the most recent studies with combined and intensive chemoradiation treatments show a 3-year local failure-free and OS of around 80% and 90%, respectively.[19] Metastatic patients (stage IVB) present a median OS of approximately 20 months, which can be greatly increased if the patient is suitable for more ablative treatments.[20]

Management

Primary treatment

RT is the mainstay of treatment and is an essential component of curative intent in non-metastatic disease. Surgery has no role in the initial treatment given its particularities of the anatomy of the area, the propensity for locoregional spread, and the proximity of critical structures.

The proximity of numerous critical structures within the nasopharynx makes it challenging to provide an adequate dose coverage to an irregularity shaped tumour while keeping the dose to surrounding tissues within acceptable limits, as discussed in Chapter 65. IMRT provides highly conformal and precise coverage with sharp dose gradients and its adoption for treatment of NPC has shown an improvement in locoregional control and survival and a reduction in toxicity.[21,22] The use of IMRT has reduced the 5-year locoregional failure rate for newly diagnosed non-metastatic NPC to less than 10%[23] and has demonstrated a significant reduction in radiation-induced toxicities such as temporal lobe neuropathy, xerostomia, and trismus.[21] Successful RT of NPC requires precise delineation and exact dose delivery to the gross tumour volume (GTV), clinical target volume (CTV), and organs at risk (OARs). Moreover, IMRT permits the delineation of several CTVs: high-risk CTV for gross primary and nodal disease which receives full therapeutic dose and intermediate-risk CTV for subclinical disease which receives a prophylactic dose. This practice is known as simultaneous integrated boost or dose painting (**Figure 61.2**). Contrast-enhanced magnetic resonance imaging (MRI) and positron emission tomography (PET)/computed tomography (CT) images performed closely before RT, ideally acquired in the treatment position, and fused with the planning CT scan allow an accurate delineation of the primary tumour and nodal GTVs. International consensus guidelines have been published for delineating the CTV and OARs to guarantee appropriate target coverage and lessen adverse effects.[24] The general principles of CTV contouring are based on the natural behaviour of tumour invasion and the complex anatomical relationship between the nasopharynx and adjacent soft tissues. In summary, high-risk CTV delineates volumes to receive a dose of 70 Gy equivalent, which includes the primary tumour and involved nodes with 0.5–1 cm margins. The entire nasopharynx is usually encompassed unless the tumour is well lateralized. Intermediate-risk CTV delineates volumes to receive a dose of 50–60 Gy equivalent (**Table 61.1**). Some centres may choose to implement a third low-dose level, making a total of 60 Gy equivalent, given to intermediate-risk primary and nodal CTV, while a lower dose of 50 Gy equivalent is given to the lower neck. Despite these set of consensus guidelines, the high interobserver variability has prompted efforts to improve countering accuracy, consistency, and efficiency using deep learning algorithms. In addition, current studies are also looking at the possible reduction of the CTVs and prophylactic neck irradiation based on an individualized, patient risk-based approach.

Proton beam therapy and carbon ion RT are newly emerging technologies which may further improve the therapeutic ratio even further given its dosimetry advantages.[25] However, evidence to support a difference in outcomes to those achieved with IMRT is lacking and large-scale studies with long-term follow-up data are warranted.

While early disease (stage I) NPC is treated with RT alone, intermediate (stage II) and locoregionally advanced disease (stages III–IVA) requires the combination of chemotherapy. A number of trials and a recent meta-analysis have demonstrated the survival benefit of concurrent chemoradiotherapy (CRT) with or without adjuvant chemotherapy versus RT alone in advanced NPC.[26–29] However, whether adjuvant chemotherapy after concurrent CRT confers further survival benefits remains controversial. Indeed, results of a phase III trial failed to show significant improvements when adjuvant

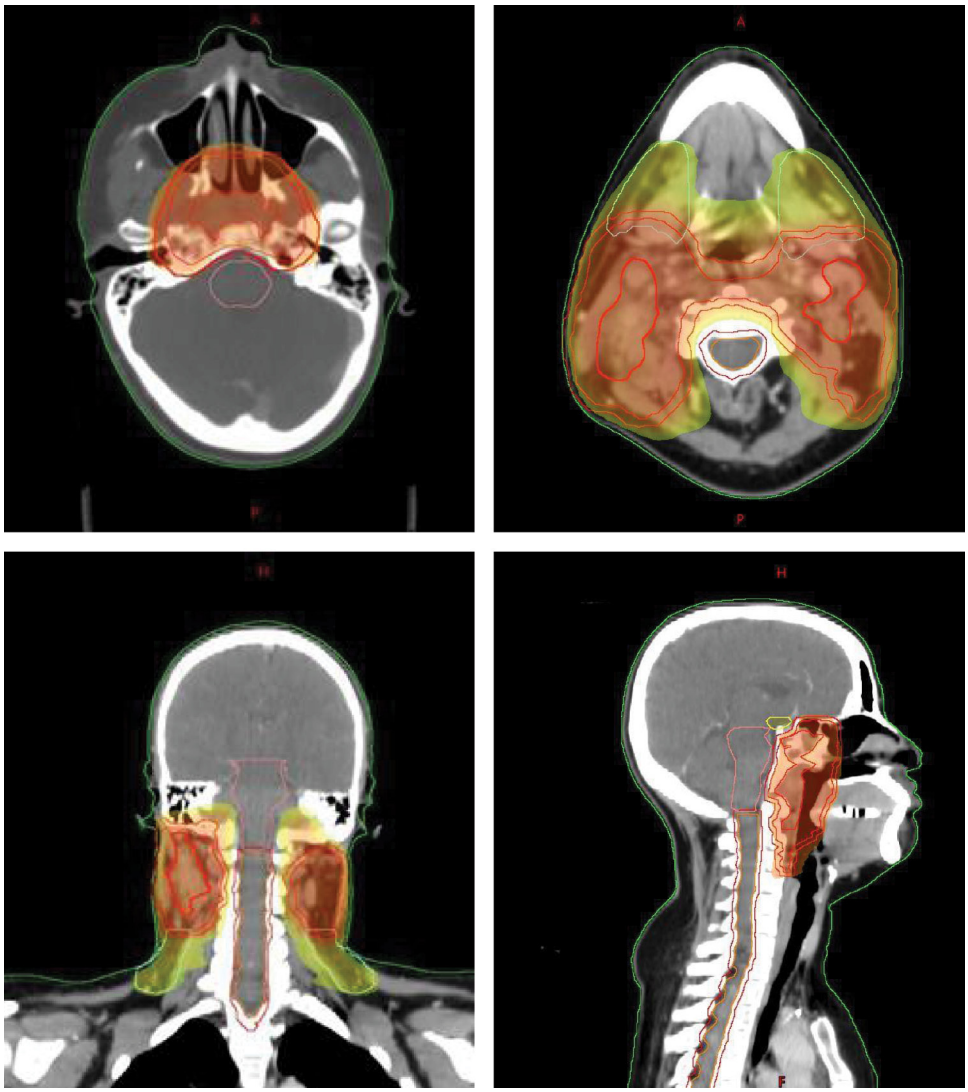


Figure 61.2 Dose colourwash illustrating IMRT planning for a case of a cT4N2M0 NPC.

chemotherapy was added to concurrent CRT.[30] The most commonly used chemotherapy schedule is 3-weekly cisplatin 100 mg/m^2 based on the United States Intergroup Study 0099.[27] Weekly cisplatin 40 mg/m^2 is acceptable in clinical practice too. Nonetheless, rather than the regimen used, the dose intensity acquires more importance, with cumulative 200 mg/m^2 as the threshold for optimal efficacy,[31] and 160 mg/m^2 when receiving induction chemotherapy.[32] The role of adjuvant chemotherapy with cisplatin and gemcitabine on detectable serum EBV DNA patients after completion of RT or CRT was studied in a phase III trial. However, despite using this biomarker to

Table 61.1 IMRT target delineation for nasopharyngeal carcinoma

High-risk CTV (70 Gy equivalent)	CTV primary[1]: primary GTV + 5 mm (± whole nasopharynx) (Minimal margin if tumour in close proximity to critical OARs is 1 mm)
	CTV nodal[1]: all positive lymph nodes (GTV nodal) + 5 mm (consider 10 mm if extracapsular spread)
Intermediate-risk CTV (50–60 Gy equivalent)	CTV primary[2]: primary GTV + 10 mm, whole nasopharynx, posterior part of the nasal cavity and maxillary sinuses, skull base covering foramina ovale, rotundum, lacerum and petrous tip, cavernous sinus (if T3–T4), pterygoid fossae and parapharyngeal spaces, sphenoid sinus (inferior half if T1–T2 and whole if T3–T4), clivus (inferior 1/3 if no invasion and whole if invasion)
	CTV nodal[2]: • Bilateral retropharyngeal lymph nodes, levels II, III, Va, and VIIb, and at least ipsilateral one level below the involved levels • Consider level Ib if there is disease involvement of the submandibular gland, or involvement of structures that drain to level Ib as the first echelon site or involvement of level II with extracapsular extension or a maximum nodal axial diameter >2 cm • Cover cervical lymph node levels IV and Vb ipsilaterally if there are any involved lymph nodes on the same side of the neck (excluding retropharyngeal lymph nodes)

identify patients with higher risk of relapse, the study did not show survival benefit.[33] On the other hand, induction chemotherapy followed by concurrent CRT represents a promising treatment strategy. Recently, two large-scale, multicentre phase III trials have shown that induction cisplatin and fluorouracil significantly improved 3-year disease-free survival[34]; and induction gemcitabine and cisplatin added to CRT significantly improved recurrence-free survival and OS, as compared with CRT alone.[19] In summary, CRT is the mainstay of treatment for patients with locoregionally advanced NPC with induction chemotherapy followed by CRT playing an increasingly important role by improving distant control and subsequently survival. However, the ideal intensity and regimen for induction has not been established. In this regard, a network meta-analysis involving seven randomized clinical trials showed that the combination regimen of taxane, platinum, and 5-fluorouracil (TPF) was the most effective for improving the OS of locoregionally advanced NPC at the cost of high rate of severe adverse events.[35] Despite induction or concurrent chemotherapy, patients with high-risk locoregionally advanced NPC present high rates of disease relapse. Two recent phase III randomized controlled trials showed that the addition of oral metronomic or adjuvant capecitabine improved failure-free survival compared to observation, with a manageable safety profile. Therefore, this may be considered in high-risk locoregional disease.[36,37] In the near future, effective biomarkers such as pre-treatment EBV DNA or relevant genomic signatures may help to identify patients most likely to benefit from induction or adjuvant therapies.

Assessment of treatment response and follow-up

Despite combined aggressive treatment, local, regional, and/or distant failure may occur. Therefore, assessment of treatment response and follow-up is imperative. In addition, late toxicity induced by RT or CRT highlights the importance of an appropriate follow-up. Twelve weeks after completion of RT or CRT is widely considered the appropriate timepoint for initial assessment of residual disease as, by then, the treatment-related inflammation would have largely resolved and most tumours would have regressed. A comprehensive response assessment includes clinical history with endoscopic examination and neck palpation and radiological imaging. ^{18}F-FDG-PET/CT is more accurate in distinguishing residual disease from post-RT fibrosis, and combined with MRI constitutes the most accurate assessment modality.[38] Plasma EBV DNA may have a role as a complementary surveillance method to conventional imaging for monitoring failure, particularly distant metastases.[39] The presence of radiological abnormalities alongside with morphologically viable malignant cells with evidence of EBER by *in situ* hybridization is strongly suggestive of residual disease.

Follow-up includes periodic examination of the upper aerodigestive tract, using fibreoptic nasoendoscopy, and neck, and evaluation of other systemic complaints suggestive of distant relapse. Most guidelines recommend clinical examination every 3–4 months for the first 2 years, every 6 months until 5 years, and yearly afterwards; and radiological imaging (CT or MRI) on a 12-month basis for the first 5 years after treatment.[14]

Locally recurrent or persistent disease

Salvage neck dissection is the primary choice for patients with isolated resectable regional nodal failure. The high incidence of extracapsular spread demonstrated in persistent nodal disease following CRT may require excision of involved skin and repair with pedicled or microvascular flaps.[40] Due to this, postoperative RT in the form of external beam RT or interstitial brachytherapy placed in the dissected neck are sometimes offered as adjuvant treatment.

Given the deep location and complex adjacency of the nasopharynx, salvage surgery of local failure at the primary site can be technically challenging with potentially severe morbidity. Small and centrally located residual disease (rT1) can be removed using an endoscopic or transoral robotic-assisted approach. Endoscopically guided microwave coagulation has been described as having low morbidity and good outcomes.[41,42] For tumours with parapharyngeal extension (rT2), the maxillary swing (facial translocation) approach provides a wide exposure, allowing tumours to be resected with adequate margins. A lateral approach with radical mastoidectomy and exposure of the infratemporal fossa after mobilization of the internal carotid, trigeminal nerve. and floor of the middle cranial fossa has been described.[43] Surgery for rT3–T4 tumours is rarely indicated.

Non-surgical options include re-irradiation and palliative systemic treatment. The scope for re-irradiation depends on the tumour volume, location, dose received by adjuvant critical OARs, and disease-free interval since primary irradiation. In general, a dose of greater than 60 Gy needs to be deliverable for re-irradiation to be worthwhile.[44] This is more achievable using IMRT, intracavitary brachytherapy. and stereotactic radiosurgery. Achieving a high dose safely is usually limited by the vital surrounding normal structures. As previously mentioned, proton and carbon ion therapies could be a promising salvage treatment considering its physical advantages to IMRT.[45] OS rates of 41% at 5 years have been reported in a recently published meta-analysis of re-irradiation with IMRT. However, a pooled event rate of grade 5 events of 33% stresses the fact that re-irradiation should be approached with caution.[46] Recently, a prognostic scoring system for locally recurrent NPC, which takes into account age, disease volume, prior RT-induced toxicities, rT category, and repeat IMRT dose, has been developed,[47] where low-risk patients may be the ideal candidates for curative repeat IMRT. Despite the lack of randomized data, induction and concurrent chemotherapy is being used increasingly for re-irradiation.

For residual/recurrent disease that is neither resectable nor suitable for re-irradiation and for high-risk patients with poor prognosis, palliative systemic anticancer treatment is the treatment of choice.

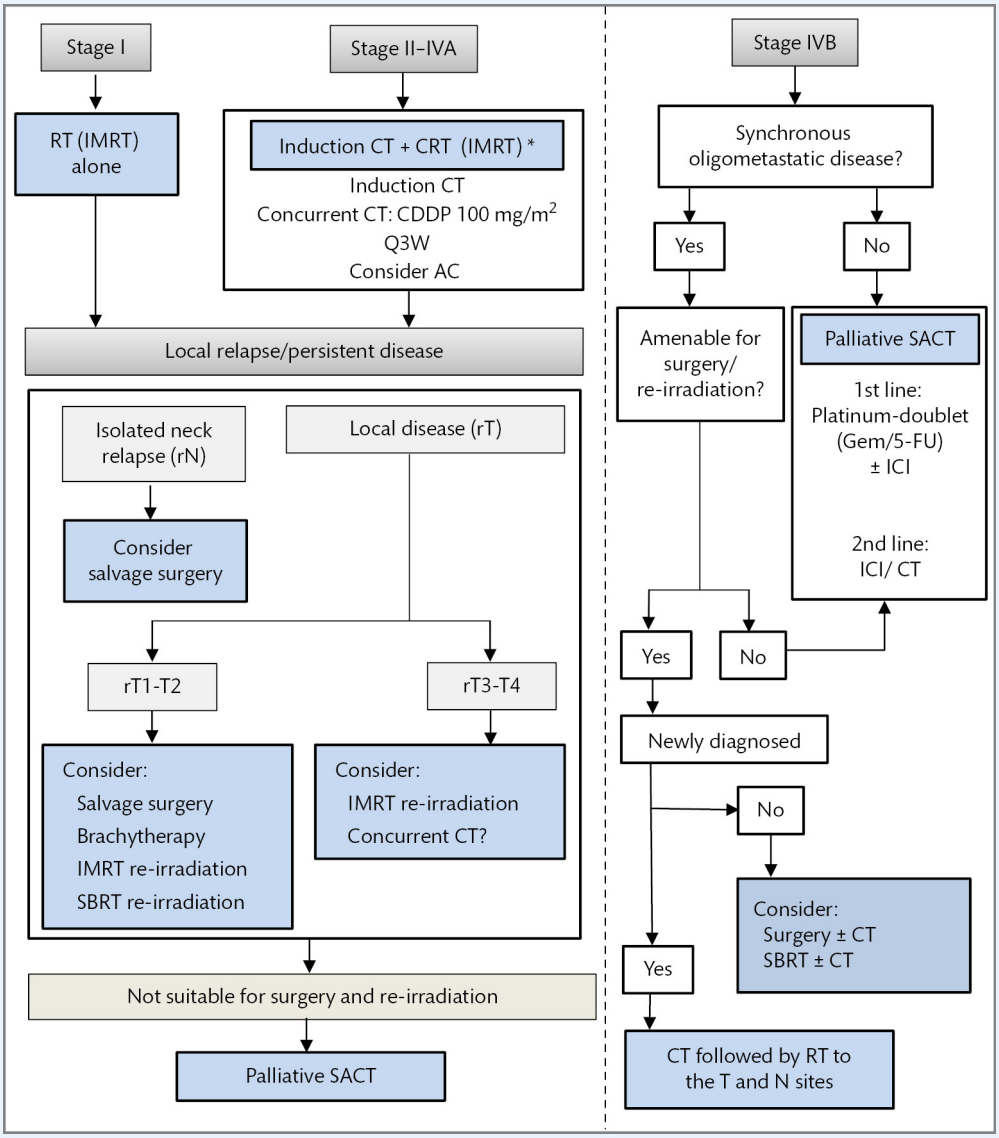
Metastatic disease

Patients with metastatic NPC represent a heterogeneous group who display a wide range of survival outcomes. In the event of *de novo* metastatic disease, the prognosis is closely related to the site and extent of metastatic lesions. A recently published phase III study in this setting randomized patients who achieved partial or complete response after three cycles of cisplatin and fluorouracil to three further cycles of chemotherapy or CRT to the primary disease with radical RT doses. The study demonstrated a statistically significant benefit in OS (hazard ratio 0.42) and progression-free survival (PFS) (hazard ratio 0.36) in the CRT arm with high treatment compliance (96.7%).[48] The survival benefit might be as a result of reduction in tumour load and the decrease in the likelihood of death from fatal complications resulting from local invasion, such as intracranial disease or intractable bleeding. In addition, local therapy of metastatic lesions with ablative RT and/or surgery with primary CRT

for locoregional disease, may be strategies to consider in prognostic groups with favourable features.[49]

Platinum-containing doublet of cisplatin and fluorouracil was the first-line systemic therapy for relapsed and metastatic disease for decades, with response rates of 40–65%.[50] A large phase III randomized trial comparing gemcitabine plus cisplatin versus fluorouracil plus cisplatin in patients with recurrent or metastatic NPC showed that the gemcitabine group outperformed the fluorouracil group in terms of objective response rate (64% vs 42%), median PFS (7 vs 5.6 months), and median OS (29.1 vs 20.9 months), at the expense of increased haematological toxicities but less mucositis.[20] Up to 83% of the recruited patients in this trial were type III NPC and improvement in PFS was not consistent for the non-type III histology. Nevertheless, this study established gemcitabine plus cisplatin as a new standard first-line treatment option for recurrent/metastatic non-keratinizing undifferentiated NPC. Recently, two randomized phase III clinical trials of East Asian populations showed an increase in PFS when immunotherapy in the form of anti-PD-1 checkpoint inhibition (camrelizumab or toripalimab) was added to first-line treatment with cisplatin and gemcitabine, followed by maintenance immunotherapy. The addition of immunotherapy should therefore be considered, pending long-term results in OS. However, at the time this chapter was written, camrelizumab and toripalimab have not been approved by the European Medicines Agency, US Food and Drug Administration, and National Institute for Health and Care Excellence UK for NPC. Nevertheless, immunotherapy alone or its combination with targeted therapies has attracted great interest as a promising approach towards improving clinical outcomes. In this

Box 61.2 Treatment algorithm according to AJCC/UICC, eighth edition



* For T2N0: consider omission of induction chemotherapy.

5-FU, 5-fluorouracil; AC, adjuvant capecitabine; CDDP, cisplatin; CRT, chemoradiation; CT, chemotherapy; Gem, gemcitabine; ICI, immune checkpoint inhibitors; IMRT, intensity-modulated radiotherapy; RT, radiotherapy; SACT, systemic anticancer treatment; SBRT, stereotactic body radiotherapy; T, tumour; N, regional lymph nodes.

regard, a recent phase II clinical trial that tested the combination of camrelizumab with an anti-angiogenic agent (apatinib) has shown promising efficacy with a measurable safety profile.

Upon progression after first-line therapy, the efficacy of non-platinum monotherapy (such as capecitabine, docetaxel, and gemcitabine) or doublets (gemcitabine–vinorelbine, ifosfamide–fluorouracil, or ifosfamide–doxorubicin) has been tested in small phase II studies, with reported response rates that vary from 30% to 68%,[51–55] with poly-chemotherapy showing higher anticancer activity compared to monotherapy.[56] The lack of comparator arm makes it impossible to recommend the most suitable therapy in the second-line setting. The estimated PFS and OS with second-line therapy area are around 5 and 12 months, respectively. In this context, treatment choice should be based on previous lines of treatment, the patient's symptoms, performance status, expected toxicities, and preferences. In addition, immunotherapy represents a promising strategy, with some phase II studies with anti-PD-1 checkpoint inhibitors (nivolumab, pembrolizumab, and camrelizumab) shown to be safe and active as monotherapy, with objective response rates of 20%, 25%, and 34%, respectively, and with most of the best responses occurring at first radiological evaluation.[57–59] Cytotoxic T-cell lymphocyte adoptive immunotherapy has demonstrated activity in highly pretreated patients.[60] In the setting of prolonged PFS following first-line therapy (>12 months), reintroduction of platinum-based chemotherapy can also be considered. Moreover, consideration of inclusion in clinical trials should be encouraged.

A practical treatment algorithm is proposed in **Box 61.2**.

Conclusion and areas of future interest

NPC is a significant healthcare burden in endemic areas where EBV infection and other hereditary and environmental factors play a crucial role in the pathogenesis. The advances in RT techniques and systemic treatments have substantially improved outcomes and transformed prognosis. However, several questions on clinical management and biological mechanisms remain unanswered. In addition, prognosis in the metastatic setting remains poor. Future research points are addressed in **Box 61.3**.

Box 61.3 Research questions on nasopharyngeal cancer

- What is the biological mechanism underlying the EBV infection and development of NPC?
- Which is the target population to screen for NPC and how should this be done?
- How should plasma EBV DNA be incorporated into the current staging system?
- How can plasma EBV DNA during surveillance be applied to individual risk-stratified treatment?
- Is there a role in proton therapy and carbon ion radiotherapy?
- Should patients with T2N0 disease receive induction and/or concomitant chemotherapy?
- How can the RT volumes and dose be adapted based on individual and anatomical risk stratification?
- How can artificial intelligence be applied in the RT delineation and planning?
- Which patients with residual or recurrent NPC should undergo surgery or RT?
- Which patients in synchronous metastatic disease should receive definitive local CRT?
- What is the role of ablative therapies in the metastatic setting?
- How can immune-checkpoint inhibitors be incorporated in the treatment of metastatic NPC?

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62

Adult deep neck space infections

Zi-Wei Liu and Abdul Ahmed

Introduction

Deep neck space infections present at a rate of 2/100,000 each year in the UK to both ear, nose, and throat and maxillofacial surgery as emergencies.[1] These can broadly be divided into bacterial, mycobacterial, and other causes. Of these, bacterial deep neck space infections are by far the commonest and of most interest to a surgeon. Bacterial infections are subdivided by location of the abscess into parapharyngeal, anterior neck, and retropharyngeal. The management of these abscesses varies according to surgical preference and availability of interventional radiology, but whichever strategy is adopted, regular daily review is essential to ensure abscess resolution and repeat interventions are often required.

Anatomy of deep neck space infections

Parapharyngeal space infection

The commonest type of deep neck space abscess, parapharyngeal space infections, generally arise from a tonsil (80%) or dental cause (20%). This space is external to the pharyngeal constrictors, lying in an inverted triangle bordered by the skull base superiorly, hyoid bone inferiorly, and mandible laterally. For surgical purposes it can be considered as the fatty connective tissue deep to the muscles of the tonsillar bed and wrapped around the internal carotid artery. It is more common for the abscess to develop medial to the internal carotid artery in which case it is best drained via an intraoral approach; however, sometimes an external approach is required.

Anterior neck infection

This includes infections affecting level I–V lymph nodes in the neck. These infections most commonly arise from suppurative lymphadenitis; however, submandibular and submental abscesses can have a dental origin and a maxillofacial review would be prudent. Mycobacterial infections and, rarely, lymphoma with necrotic nodes can present in this area. Bacterial abscess in lymph nodes levels II to IV may cause Lemierre's syndrome, characterized by systemic septic emboli from thrombosis of the internal jugular vein.

Retropharyngeal infection

This is a space that contains fatty tissue and a few lymph nodes, which lies posterior to the pharynx and oesophagus in the neck and the mediastinum. It is bounded anteriorly by the buccopharyngeal fascia that envelops the constrictor muscles of the pharynx and posteriorly by the prevertebral fascia and extends down to the tracheal bifurcation.

Behind the retropharyngeal space and posterior to it is the danger space, which extends further down to the posterior mediastinum and infected collections here may cause fatal complications. Both clinically and radiologically it is extremely difficult to determine whether an abscess has extended to the danger space, but radiologists will generally report whether there is evidence of mediastinitis.

Retropharyngeal abscesses are due to suppurative lymphadenitis from an upper respiratory tract infection in children, as the nodes drain the nasopharynx and sinuses. In adults, penetrating trauma such as a retained fishbone is more likely. A rare cause of retropharyngeal or prevertebral abscess is mycobacterial osteomyelitis of the spine spilling over into this space, but changes to the adjacent vertebra should give a clue.

Submandibular space infection

This is a triangular-shaped space, bounded superiorly by the mylohyoid muscle, medially by the anterior belly of the digastric muscle, and the mandible laterally. It contains lymph nodes, the superficial lobe of the submandibular glands, and blood vessels. It communicates with the sublingual space above, and the submental space anteriorly.

Submental space infection

This is contained by the two anterior bellies of the digastric muscles. Above is the mylohyoid muscle and below the deep cervical fascia covered by platysma and skin. It contains submental lymph nodes and communicates posteriorly with the submandibular space. Surgical access is obtained behind the mental prominence in the anterior neck.

Clinical presentation and acute management

Deep neck space abscesses present with a wide spectrum of severity depending on size and location. The mildest cases present with throat pain or neck swelling over days to weeks, often without skin changes due to the abscess being contained by the deep investing fascia. A concurrent history of tonsillitis, dental infection, or preceding upper respiratory tract infection may be present. Often, patients have received antibiotics in primary care, leading to symptoms being masked for a few days. With further progression, local effects such as voice change, swallow difficulty, and stridor may occur. The most severe cases will have systemic sepsis.

Acute management therefore focuses on the above problems. An ABCDE approach as per Advanced Life Support can be followed if the patient has airway difficulty or sepsis. Maximal medical airway treatment (with adrenaline nebulizers, dexamethasone/prednisolone) and an anaesthetic review should be the first treatment if stridor is present. Intravenous access should be established with routine bloods and inflammatory markers sent, as well as blood cultures. An antibiotic with broad-spectrum cover should be started in accordance with local microbiology guidelines.

Cross-sectional imaging should be obtained in the first instance (see Chapter 69). If available, abscesses can be aspirated under ultrasound (US) guidance to give relief to symptoms and obtain a sample for microbiology. A decision then needs to be made as to whether to pursue a conservative management plan or to arrange urgent surgical drainage.

Parapharyngeal abscess

Parapharyngeal abscesses typically present as an intraoral swelling displacing the tonsil medially and often extend from an existing peritonsillar abscess.[2] Symptoms in the early stages include pain, swelling, and 'hot potato' voice. As the abscess enlarges, masseter and lateral pterygoid muscles are irritated, causing trismus. On further extension into the neck, lateral rotation of the head becomes painful. There is rarely external swelling or skin change. Examination will elicit neck tenderness and sternocleidomastoid (SCM) spasm.

Acute treatment needs expert anaesthetic input at an early stage as trismus makes oral intubation difficult and the abscess may obscure the view for a nasal intubation.

Anterior neck abscess

These present with a painful swelling of the neck with or without overlying skin change. Large abscesses can limit cervical rotation from pain. If there is voice change, this tends to be hoarse rather than 'hot potato'. Flexible nasendoscopy will either be normal or show some oedema of the larynx. A special case is a thyroid abscess, which can occur spontaneously or be iatrogenic (e.g. from an US fine needle aspiration), which does cause laryngeal oedema and airway compromise early and requires urgent drainage.

Retropharyngeal abscess

These have a subtle clinical presentation and may be missed on casual examination. A sore throat, odynophagia, and difficulty rotating the head are key symptoms. Sepsis is possible. On intraoral examination and flexible nasendoscopy, a bulge of the posterior pharyngeal wall may or may not be present. The rest of the pharynx and larynx may be unremarkable. A high index of suspicion is required to diagnose a retropharyngeal abscess. Drooling, unwell children with otherwise minimal clinical findings may have a retropharyngeal abscess (see Chapter 76). Adults will usually have a history of a retained foreign body or trauma. Transoral aspiration or incision and drainage under local anaesthesia is possible if there is an obvious bulge in the posterior pharyngeal wall in a compliant patient, otherwise early intubation and surgical exploration under general anaesthesia is recommended.

Patients with retropharyngeal abscesses often have significant difficulty in swallowing despite adequate drainage and may require nasogastric feeding for a short time.

Submental/submandibular abscess

Patients usually present with trismus, swelling, and usually a history of dental work or tooth pain (Figure 62.1). Investigations usually would include bloods and an orthopantomogram. The trismus is caused by oedema and spread of infection into the buccal and submasseteric spaces.

When examining a patient, if the lower border of the mandible is not palpable the submandibular space is invariably involved and must be drained. Any infection involving the lower posterior teeth (notably the wisdom teeth) can spread into the submandibular space directly through the bone, or spread to the submandibular lymph nodes.

Ludwig's angina

This is a special case[3] of submental/submandibular abscesses as discussed above, and the clinical features include firm, brawny, indurated cellulitis involving the submandibular, sublingual, and submental spaces bilaterally. Patients are usually septic.

There is a massive swelling involving both the neck and the floor of the mouth. Ludwig's angina is a potential airway emergency and

Figure 62.1 A patient with infection of the wisdom tooth presenting with submandibular swelling and trismus. Note blunting of the lower border of mandible.

Figure 62.2 Bilateral submental, and submandibular gland swelling—with floor of mouth oedema.

patients can quickly become septic or develop other complications. Historically this condition has had a mortality rate exceeding 50%. With aggressive surgical intervention, good airway control, and antibiotics this has now dropped to around 8%. Usually, the initial cause is a submandibular space infection which is typically dental in origin.

From its initial focus in the submandibular space, the infection then spreads into the sublingual space (following the tissue space around the deep lobe of the submandibular gland) and from there to the other side, via the floor of the mouth (Figure 62.2). The tissues here are very loose and are easily distensible. Very often, rather than an abscess collection, the infection is usually a cellulitis, with diffuse inflammation of soft tissues spaces.

Ludwig's angina is a life-threatening emergency. It is best treated by early recognition, prompt resuscitation, and aggressive surgical drainage (Figure 62.3). This includes high-dose antibiotics, steroids, and removal of the underlying cause (often extraction of the offending tooth).

Drains should be left in the sublingual, submandibular, and submental spaces. Often repeat return to theatres with further washouts is indicated. Most patients will need high dependency care. Complications include airway obstruction, septicaemia, mediastinitis, and even death.

Mycobacterial abscess

These are 'cold' abscesses[4] with neck swelling presenting over weeks or months rather than days. Presentation is delayed because there is little pain. The overlying skin will typically have a purplish discoloration with minimal erythema. The key is not to rush into incision and drainage but rather arrange for US-guided biopsy with tuberculosis cultures and histology (will show caseating granulomas and histiocytes). Since the laboratory investigations take weeks to return a result, the diagnosis and management plan is based on history and clinical findings. If a cold abscess is accidentally incised, this may lead to chronic discharge but will heal as the patient starts on antimycobacterial medication.

Figure 62.3 Submandibular drain *in situ*.

Investigations

Flexible nasendoscopy

Important in assessment of deep neck space abscesses to ensure the airway is stable. Any laryngeal oedema or pharyngeal swelling should be noted, and an airway plan discussed with the anaesthetist in case surgery is required (see Chapter 52). Repeat endoscopy should be done regularly for inpatients to document response to treatment.

Routine blood tests

Full blood count, urea and electrolytes, clotting and group and save (in case of surgery), and blood cultures.

Imaging

Plain lateral soft tissue X-ray of the neck can be helpful in retropharyngeal abscess as it shows expansion of the retropharyngeal space and possible foreign bodies in adults. The retropharyngeal soft tissue (measured at the level of the oesophagus) should be no more than 50% of the width of the adjacent vertebral body. Loss of lordosis can also be present.

Plain X-rays are not indicated in the investigation of other deep neck space abscesses.

Computed tomography of the neck with or without chest with contrast

Computed tomography (CT) of the neck and chest is the best imaging modality for deep neck space abscesses. It shows the location of the abscess, relation to key structures, and the degree of suppuration (important if surgical drainage is planned). Complications arising from the abscess such as thrombosis of the internal jugular vein, mediastinitis, and septic emboli in the thorax can be seen. It can also suggest alternative diagnoses such as mycobacterial infection or lymphoma.

US of the neck with or without aspiration

The availability of this investigation is dependent on availability of local head and neck radiology expertise. It can be therapeutic as well as diagnostic in visualizing the abscess and providing a sample for microbiology and cultures. In children and in cases of suspected mycobacterial infection, US with or without aspiration is the first-line intervention ahead of surgical drainage. However, in adults with large, thick walled, and loculated abscesses, US drainage is unlikely to result in cure and surgery is preferred. In addition, US visualization and access to the parapharyngeal and retropharyngeal spaces is quite limited and is not recommended in these sites.

Orthopantomography or CT of the mandible

This is indicated for deep neck space infections of suspected dental origin. Orthopantomography is the first-line investigation, with CT of the mandible a useful adjunct where the diagnosis is in doubt or cross-sectional imaging is required to guide surgery.

Surgical approaches to deep neck space infections

Surgical drainage is indicated for patients presenting with severe symptoms such as airway swelling, swallow difficulty, sepsis, and those who have failed conservative measures. Surgical drainage is not helpful where there is only cellulitis and no suppuration, and generally an abscess with a liquid component smaller than 1 cm is best managed conservatively. Intubation can be challenging, and a skilled airway anaesthetist should be involved in these cases (see Chapter 52).

Parapharyngeal abscess

Intraoral approach

This approach is not advised for the inexperienced surgeon. The imaging findings should be very carefully studied, with an accurate idea of the great vessels to avoid inadvertent injury, which can be difficult to control from a transoral access.

Intubation may be difficult due to severe trismus and transnasal intubation with fibreoptic scope guidance may be required. The larynx is usually normal once the flexible scope is navigated past the tonsil area. Trismus may also limit mouth opening for intraoral drainage, although adequate muscle relaxation under general anaesthesia usually improves the view. If there is no access for surgical instruments, a wide-bore cannula can be used to aspirate the abscess.

The surgical set-up is as for a standard tonsillectomy. These are incised lateral to the tonsil, entering the space through the anterior pillar and through the superior constrictor muscle that forms the tonsillar bed. A simple method would be to start with a standard tonsillectomy and then pass a pair of blunt artery forceps through the tonsillar bed and open along the line of the muscle fibres. The scans should be examined to ensure that the internal carotid is displaced laterally by the abscess for this approach. A dental review may be required if the source of the abscess is due to the lower second/third molars rather than the tonsil and extraction on table can be done at the same sitting.

External approach

It is rare for parapharyngeal abscesses to be lateral to the carotid sheath, in which case intraoral drainage may be difficult. The parapharyngeal space can be accessed through the neck via a transverse skin crease incision at the level of the hyoid bone. Following retraction of the SCM, find the posterior belly of the digastric muscle and trace the internal jugular vein and carotid artery running under it into the parapharyngeal space. Blunt dissection anterior to the carotid artery and under the digastric should be performed to access the abscess cavity. The posterior belly of the digastric can be divided to improve access and visualization of the great vessels.

Profuse irrigation of the cavity is recommended, and a corrugated drain should be placed before closing the wound lightly; samples should be set for microbiology and culture.

Anterior neck abscess

These abscesses can be amenable to US-guided drainage if small and abscess contents are not too thick. Surgically, they are usually drained via an external approach. The neck should be extended and lightly rotated away from the abscess. A horizontal skin crease incision is marked out, usually anterior to the SCM, but not directly over the muscle as this will limit access. The surgeon should aim for the area of maximum fluctuance or towards the lower end of the abscess to allow gravitational drainage. If the planned incision is posterior to SCM, the course of the accessory nerve, which emerges one-third of the way down the posterior border of the muscle, should be borne in mind.

Once the incision is made, raise a subplatysmal flap and identify the border of the SCM. The SCM is retracted and the deep cervical fascia overlying the great vessels is incised. This will usually drain the abscess as the lymph nodes affected lie over the internal jugular vein and carotids.

If the abscess is loculated and has multiple compartments, all the loculations are broken down. Rarely, multiple stepladder incisions may be required. As before, irrigation of the wound and drain is recommended, with samples for microbiology.

Retropharyngeal abscess

Intraoral approach

If there is a clinically obvious bulge, retropharyngeal abscesses can be approached intraorally. With the patient under general anaesthesia and intubated, an incision is made over the area of maximum fluctuance in the posterior pharyngeal wall. The incision needs to be deep enough to go through the constrictor muscles of the pharynx and the fascia. Pus should be encountered at this point. Care should

be taken to ensure the thick prevertebral fascia is not breached as this may introduce infection into the cervical spine. Aspirating with a needle first may help to localize the abscess cavity if the location is not immediately apparent.

External approach

This is rarely required but may be helpful in cases where there is inferior extension of the retropharyngeal abscess as a mediastinal drain can be placed through the neck.

Through a lateral skin crease incision at the level of the abscess anterior to the SCM muscle, the internal carotid artery and internal jugular vein are identified and retracted laterally away from the pharynx/larynx and oesophagus. Sometimes the superior thyroid vessels may need to be ligated to allow lateral retraction of the internal carotid artery. Blunt dissection down the posterior aspect of the pharynx and oesophagus is performed to enter the abscess cavity. The wound is closed over a corrugated drain.

Submental and submandibular abscess

Surgical access can be made via an incision 2–3 cm below the lower border of the mandible (to avoid injury to the mandibular branch of the facial nerve). Skin and subcutaneous tissues are incised. Using blunt forceps, the deep cervical fascia is penetrated, aiming towards the mandibular border. The forceps is opened and slowly pulled out without closing the jaws. This is repeated several times and the wound copiously irrigated. From the same incision the submental space can be also drained by directing forceps anteriorly, and the sublingual space by pushing the forceps underneath the mandible and into the floor of the mouth. It should be possible to palpate the instrument in the sublingual space via the oral cavity. Once drained, a corrugated drain is left in each of the drained spaces for at least 24 hours.

Adjunct procedures and controversies

Tracheostomy

A decision to proceed to tracheostomy depends on patient and surgical preference, and also on whether postoperative high dependency unit care with one-to-one nursing is available. Clinically, a tracheostomy is unlikely to be required in patients who did not have airway symptoms preoperatively. As well as potential complications, tracheostomies almost always prolong a patient's stay in hospital. Alternatives include intubation for 24–48 hours post surgery with steroid cover to reduce swelling.

Reasons for inserting a tracheostomy may include (1) airway compromise at presentation, (2) patient likely to need multiple trips to theatre for washouts, (3) shortage of out-of-hours airway expertise for emergency reintubation, and (4) patients who have already been intubated for 5–7 days (see Chapter 70).

Enteral feeding

Deep neck space abscesses can compromise swallow due to pain, oedema, or mass effect. Incision and drainage of the abscess, especially intraoral approaches, have been used. Again, if the patient's swallow is unaffected preintervention, it is likely to remain intact after surgery. However, some patients, particularly in the elderly group, may require nasogastric tube feeding. Patients who will remain intubated postoperatively should have a nasogastric tube inserted.

If a decision is made to feed via a nasogastric tube, a speech and language therapy assessment should be done as soon as possible to rehabilitate swallowing function. Most people will recover normal swallow function in less than 2 weeks and should not remain nil by mouth without reassessment.

Tonsillectomy

A tonsillectomy is usually done as a part of intraoral drainage of parapharyngeal abscesses if the tonsils are thought to be the source of infection. Bilateral tonsillectomies were routinely done for this reason. However, some surgeons will choose to do unilateral tonsillectomy only to reduce the risk of bleeding in an inflamed surgical field and it can be argued that paratonsillar or parapharyngeal abscesses are almost always unilateral.

Conclusion

Deep neck space abscesses are surgical emergencies that require urgent assessment and intervention. Left untreated, speech, swallow, airway complications, and sepsis may ensue.

Drainage of neck abscesses can be done either surgically or with US guidance. Cross-sectional imaging is crucial to guide surgical approach.

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63

Parathyroid disorders

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Introduction

Parathyroid disease consists of benign and malignant pathology. For the purposes of this chapter, we will focus on benign parathyroid diseases relevant to the ear, nose, and throat surgeon: primary, secondary, and tertiary hyperparathyroidism (HPTH). Parathyroid cancer is a very rare entity and will not be discussed.

Primary hyperparathyroidism (PHPTH) is a condition characterized by hypercalcaemia driven by excess parathyroid hormone (PTH) secretion. Secondary HPTH occurs when there is a deficiency of vitamin D in the blood causing the parathyroid glands to produce more than normal amounts of PTH to rectify this. Tertiary HPTH occurs following prolonged secondary HPTH, when the parathyroid glands autonomously overproduce PTH in the absence of ongoing hypocalcaemia.

Parathyroid diseases are best managed in a multidisciplinary setting, involving endocrinologists, surgeons, geneticists, pathologists, radiologists, and clinical nurse specialists.

Aetiology of hyperparathyroidism

- The most common cause of PHPTH is a single hyperfunctioning parathyroid adenoma which accounts for 85–90% of cases. The remaining cases are caused by multiple parathyroid gland hyperplasia or multiple parathyroid adenomas.
- Parathyroid carcinoma is a rare cause of PHPTH, which accounts for 1% of diagnoses.
- Ectopic parathyroid glands can arise as a result of abnormal embryological migration of the inferior parathyroid glands and are typically located near the thymus in the anterior mediastinum but can also be found in the thyroid gland, in the para-oesophageal position, or near the tracheal bifurcation.[1] Ectopic parathyroid glands can be difficult to localize preoperatively, making surgical cure challenging on the first attempt.
- Sporadic causes: HPTH is predominately a sporadic disease.
- Familial and hereditary causes: approximately 10% of cases are familial and a hereditary predisposition should be considered in patients presenting with HPTH at a young age (<50 years), patients with clinical features of an endocrine neoplasia syndrome (e.g. pancreatic neuroendocrine tumours, pituitary adenoma), or patients with a family history of HPTH or endocrine tumours.[2] Hereditary causes include multiple endocrine neoplasia (MEN) type 1, MEN2A, MEN4, and HPTH and jaw tumour syndrome (Table 63.1). Patients with familial HPTH present most often with parathyroid hyperplasia or multiple parathyroid adenomas. Table 63.1 expands on the hereditary HPTH syndromes including the responsible genes and expressed phenotypes.
- Exogenous calcium and thiazide diuretics can cause elevated calcium levels. Drugs such as frusemide, bisphosphonates, and anticonvulsants can cause elevated PTH.
- Lithium therapy and radiotherapy to the head and neck region can cause parathyroid hyperplasia and PHPTH.
- Chronic vitamin D deficiency, which can stimulate parathyroid hyperplasia and PTH secretion (secondary HPTH) or adenomatous growth and autonomous parathyroid secretion (tertiary HPTH).[3]

Secondary HPTH is most commonly seen in renal failure but can be seen in severe vitamin D deficiency and malabsorption.

Tertiary HPTH usually occurs when a patient with secondary HPTH due to renal failure receives a transplant. It can also develop after any period of prolonged hypocalcaemia.

Table 63.1 Hereditary hyperparathyroidism syndromes

Hereditary disorder	Gene	Clinical phenotype
MEN1	*MEN1* *CDKN2B* *CDKN2C* *CDKN1A*	PHPTH, pancreatic neuroendocrine, extra-pancreatic neuroendocrine tumours, pituitary tumours, lipomas, adrenal tumours
MEN2A	*RET*	Medullary thyroid carcinoma, phaeochromocytoma, PHPTH
MEN4	*CDKN1B*	PHPTH, pituitary tumours, pancreatic neuroendocrine tumours, thyroid, gonadal, and renal tumours
HPTH–jaw tumour syndrome	*CDC73*	PHPTH, parathyroid carcinoma, ossifying tumours of jaw, uterine and kidney tumours

Primary hyperparathyroidism

Assessment

The assessment should focus on (1) clinical symptoms and patient comorbidities, (2) confirming a biochemical diagnosis of PHPTH, (3) the end-organ effects of HPTH (on bones and kidneys), and (4) radiological localization of parathyroid disease.

Clinical findings

PHPTH affects female patients preponderantly and typically presents in the fifth to sixth decade of life. PHPTH has evolved as a symptomatic disease presenting with renal stones, nephrocalcinosis, and fractures in the past to a disease that is often asymptomatic and picked up incidentally on routine blood tests or suggested by reduced bone density on dual-energy X-ray absorptiometry (DXA) testing. PHPTH is a common endocrine condition with a prevalence of one to seven cases per 1000 adults.[4–7]

Referrals are often made by primary care physician to secondary care. Common symptoms and signs with which patients present include the following:

- Generalized body aches with muscle weakness and fatigue.
- Excessive thirst.
- Low mood with or without depressive symptoms.
- Frequent micturition with dehydration.
- Altered bowel habit with a tendency towards constipation.
- Calcinosis leading to renal stones.
- Fractures—unexplained (usually secondary to osteoporosis).

Biochemical diagnosis

For the majority of patients with PHPTH, investigations can be safely performed in the outpatient setting. PHPTH is defined biochemically as an elevated calcium level in the presence of an elevated or inappropriately normal PTH level. Diagnosing PHPTH is sometimes incidental from an unexpectedly raised serum calcium level.

Diagnostic serum tests must include serum corrected calcium, intact PTH (measured using an immunoradiometric or immunochemical assay), a 25-hydroxyvitamin D level, serum alkaline phosphatase, serum urea and creatinine levels, and serum phosphorus. Urinary calcium measurements are required to differentiate PHPTH from familial hypocalciuric hypercalcaemia (FHH) and to determine the risk of renal stone disease. FHH is a rare, benign, autosomal dominant condition caused by mutations in the calcium-sensing receptor gene and very rarely requires medical or surgical management. The biochemical features of FHH can mimic those of PHPTH including hypercalcaemia and inappropriately elevated PTH. The main diagnostic difference between FHH and PHPTH is relative hypocalciuria and can be detected by measuring 24-hour urinary calcium and calculating a calcium creatinine clearance ratio (CCCR). A CCCR value of less than 0.02 is suggestive of FHH and should prompt referral for confirmatory genetic testing, whereas a value greater than 0.02 confirms a diagnosis of PHPTH.

Assessment of end-organ damage

In addition to measuring 24-hour urinary calcium excretion, a renal ultrasound should be considered to screen for silent renal stone disease or nephrocalcinosis. Finally, a DXA scan of the lumbar spine, distal radius, and hip should be considered to screen for osteopenia or osteoporosis in patients with HPTH.

Preoperative localization studies

Superior parathyroid glands: these are classically located near the posterolateral aspect of the superior pole of the thyroid, 1 cm superior to the junction of the recurrent laryngeal nerve, and the inferior thyroid artery. They classically lie deep to the plane of the recurrent laryngeal nerve.

Inferior parathyroid glands: these glands are classically located within 1 cm of the inferior poles of the thyroid glands, inferiorly, laterally, or posteriorly. They classically lie superficial to the plane of the recurrent laryngeal nerve.

Up to 16% of parathyroid glands can be ectopic with the superior ones being more likely to be in their usual position. Ectopic superior glands can be found most often in the retropharyngeal region or para-oesophageal anywhere down to the mediastinum. Ectopic inferior glands may be found in the anterior mediastinum in association with the thymus. Intrathyroidal adenomas are more likely to be inferior glands and rarely glands can be found in the carotid sheath (Table 63.2).

Preoperative localization of hyperfunctioning parathyroid tissue is a prerequisite for minimally invasive parathyroidectomy (MIP) and therefore preoperative imaging should be offered to those patients being considered for surgical management of HPTH. The two most commonly used imaging modalities include one or a combination of:

- ultrasound
- technetium (Tc)-99m sestamibi (MIBI) parathyroid scintigraphy.

Ultrasound has the benefit of being a cost-effective, non-invasive, and non-irradiating modality but it is operator dependent and will miss deep or ectopic parathyroid adenomas. Tc99m MIBI parathyroid scintigraphy detects an increase in the number and activity of mitochondria in a hyperfunctioning parathyroid gland (Figure 63.1). In modern practice, scintigraphy typically includes a single-photon emission computed tomography (SPECT)/computed tomography (CT) acquisition, which improves resolution. This modality

Table 63.2 Anatomical locations of parathyroid glands

	Normal	Ectopic
Superior parathyroid	1 cm superior to inferior thyroid artery/recurrent laryngeal nerve crossing	Posterolateral surface of upper half of thyroid lobe Retro-oesophageal/retrolarynx Above superior pole Intrathyroidal Carotid sheath Para- or retro-oesophageal down to posterior mediastinum
Inferior parathyroid	Within 1 cm of thyroid inferior pole	Thyrothymic ligament >1 cm lateral to inferior pole Medial on trachea Anterior mediastinum/ lower thymus Undescended, from angle of mandible to thoracic inlet Carotid bifurcation Intrathyroidal

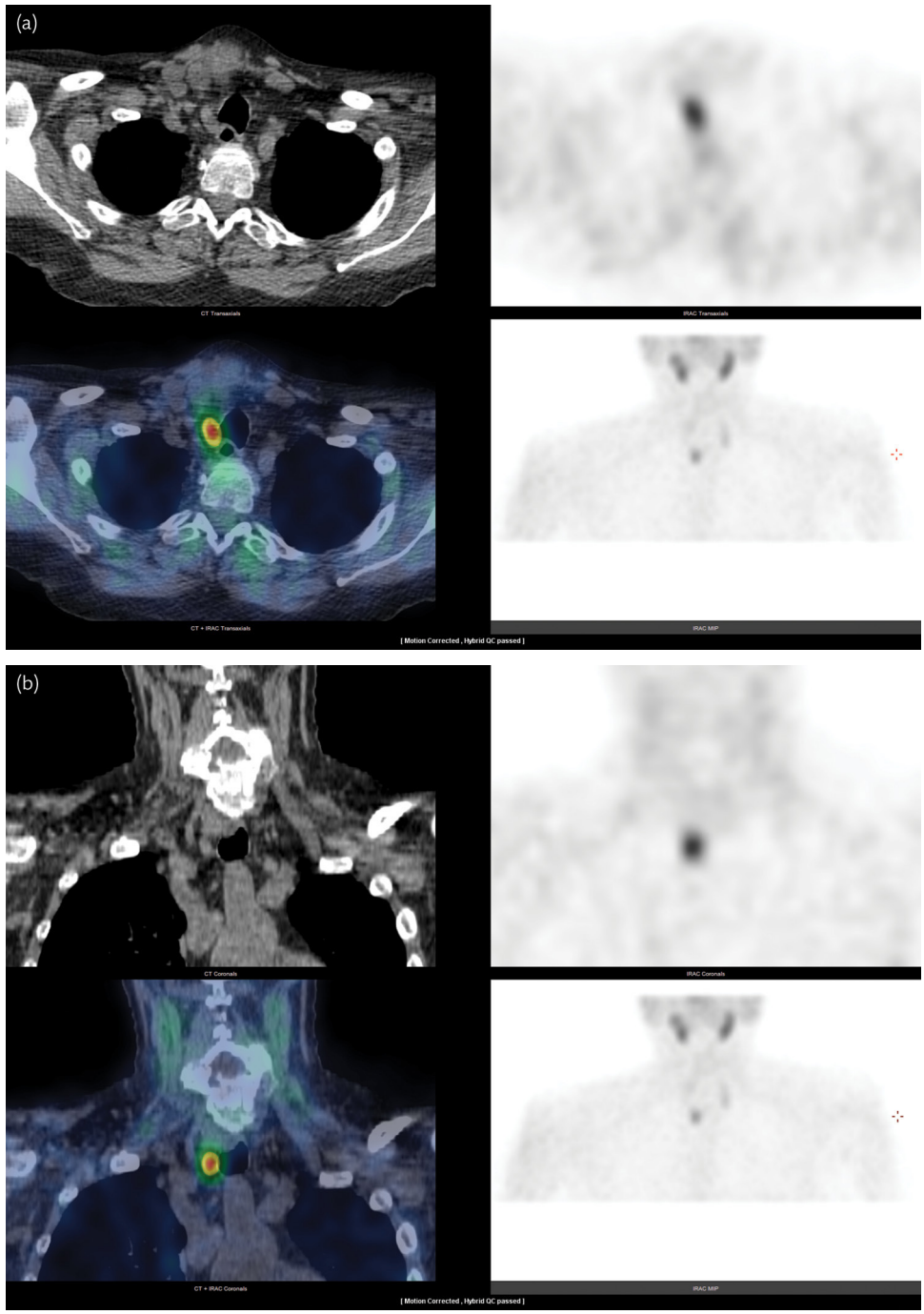

Figure 63.1 Positive Tc99m MIBI parathyroid scintigraphy. (a) Coronal views of right lower parathyroid adenoma. (b) Axial views of right lower parathyroid adenoma.

does involve a low radiation dose and is quite time-consuming for patients but can detect adenomas not seen on ultrasound.[8]

In parathyroid imaging, sensitivity can be defined as the percentage of people who have a single adenoma, who are correctly picked up by imaging tests or the percentage of people in whom imaging guides curative surgery. The pooled sensitivity of ultrasound versus sestamibi SPECT for parathyroid adenoma localization is 76.1% versus 78.9% respectively.[9]

Potential explanations for failed preoperative localization are multiple parathyroid gland hyperplasia, ectopic parathyroid adenoma, and early or mild parathyroid disease. In such instances, four-dimensional (4D) CT[10] is useful, with the 'fourth' dimension referring to changes in perfusion of hyperfunctioning glands observed over time. Parathyroid adenomas typically show a rapid initial uptake of contrast, followed by a washout over 1–2 minutes. The sensitivity of 4D CT is 79.8%[11] and it is a sensitive imaging modality to detect ectopic parathyroid adenomas.[12] There is growing interest in the utility of 4D CT as an adjunct to ultrasound and sestamibi SPECT or as an alternative imaging modality in clinical practice.[11,13]

Genetic testing

Genetic testing should be offered to patients in accordance with the local and national guidelines. In the UK, the NHS England National Genomic Directory testing criteria (see 'Further reading') recommend that genetic testing is offered to those patients with HPTH presenting at a young age (<50 years) or patients at any age with

(a) a confirmed or relevant family history, OR
(b) multi-glandular disease or hyperplasia in the presence of relevant family history, OR
(c) parathyroid carcinoma or atypical or cystic adenoma, OR
(d) ossifying fibroma(s) of the maxilla and/or mandible.

Treatment

Patients should be offered the option of surgery if they have symptomatic PHPTH, evidence of end-organ damage (e.g. renal stones or osteoporosis), or significantly elevated calcium levels.

Medical management

PHTH is commonly encountered in the elderly population who may have significant comorbidities which increase the risks associated with surgery. For patients with significant comorbidities who do not meet the criteria for surgery, a conservative or medical management approach should be adopted.

Patients can be offered treatment with a calcimimetic such as cinacalcet or evocalcet if they are considered unsuitable for surgery and have a calcium level greater than 2.85 mmol/L with symptomatic hypercalcaemia, or a calcium level greater than 3.0 mmol/L without symptoms. A bisphosphonate should also be considered to reduce fracture risk for patients with HPTH and for those in whom surgery is contraindicated or has failed.

Surgical management

Primary parathyroid surgery

All surgical intervention for parathyroid surgery must be preceded by a full preoperative assessment, including clinical evaluation of the vocal cord mobility.

The conditions for surgical management by way of parathyroidectomy are listed in **Box 63.1**. Surgery may take the form of:

- minimally invasive parathyroidectomy: focused/minimal access surgery, video-assisted surgery, and remote access surgery.
- open bilateral neck exploration: transcervical surgery also known as four-gland exploration.

Box 63.1 Indications for considering surgery in primary hyperparathyroidism

- Clinical symptoms (e.g. polyuria, polydipsia, musculoskeletal symptoms, cognitive or mood disturbance).
- Evidence of end-organ damage (e.g. renal stones, fragility fractures, or evidence of osteoporosis on DXA scanning).
- Significantly elevated serum corrected calcium (>2.85 mmol/L).

Adapted from National Institute for Health and Clinical Excellence. Hyperparathyroidism (primary): diagnosis, assessment and initial management. NICE guideline [NG132]. 2019. https://www.nice.org.uk/guidance/ng132/documents/evidence-review-11

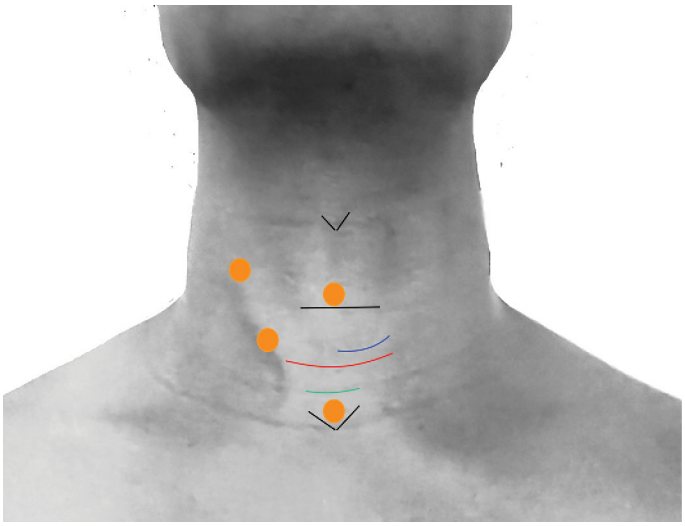

Figure 63.2 Landmarks for surgical access to parathyroidectomy. Key: orange, total endoscopic parathyroidectomy: port access; red, bilateral neck exploration; blue, MIP; green, MIVAP. The black lines indicate the neck landmarks: from superior to inferior, these are the thyroid notch, the cricoid cartilage, and the suprasternal notch.

The aetiology of up to 90% of cases of HPTH being secondary to an adenoma makes surgery a rewarding treatment option. The primary outcome of surgical intervention is to achieve permanent resolution of the biochemical abnormality. **Figure 63.2** shows the various landmarks for the differing surgical approaches discussed.

Minimally invasive parathyroidectomy The MIP approach uses a 2.5 cm neck incision, approaching via the midline or laterally, allowing access to both ipsilateral parathyroid glands. Surgery can be offered as a day case and even under regional anaesthesia. Excision of the localized lesion can be accompanied by intraoperative serum analysis of PTH which shows a real-time reduction upon excision of the responsible gland. In select centres, this can be combined with gamma probe-guided (radio-guided or scan-directed) parathyroidectomy, where Tc99m MIBI is administered intravenously 1–2 hours before surgery and an intraoperative gamma probe used to identify the adenoma.

Minimally invasive video-assisted parathyroidectomy Using a 1.5–2 cm incision over the suprasternal notch, access is obtained without gas insufflation; angled telescopes, custom retractors, and special endoscopic instruments are used to remove the adenoma. The use of endoscopic video-assisted parathyroidectomy allows single adenoma and four-gland exploration. Minimally invasive video-assisted parathyroidectomy (MIVAP) results in a HPTH cure rate of up to 97%, which is comparable to standard MIP of up to 96–98%. However, up to 25% of cases of MIVAP lead to conversion to an open procedure.[14] **Figures 63.3** and **63.4** show minimally invasive parathyroid surgery and the resulting resected adenoma.

Total endoscopic parathyroidectomy This technique involves up to four ports being placed in the neck and is performed using carbon dioxide insufflation. A similar conversion rate as for MIVAP has been reported.

Remote access parathyroidectomy Aimed at avoiding a scar in the neck, robot-assisted parathyroidectomy has been performed

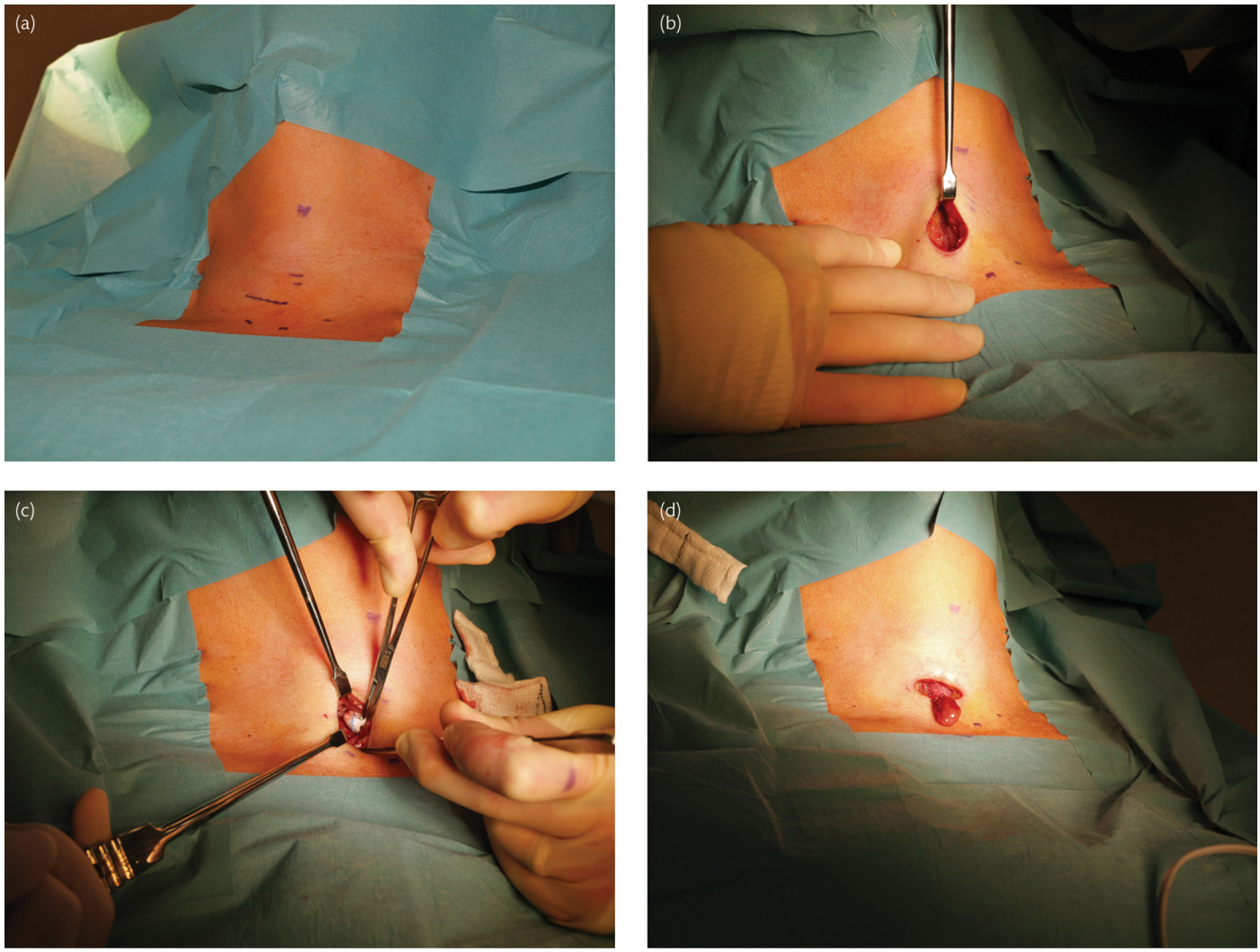

Figure 63.3 Minimally invasive parathyroid surgery. (a) Skin markings minimally invasive surgery of right lower parathyroid adenoma. (b) Open incision. (c) Retraction. (d) Excised parathyroid adenoma.

via axillary, breast, infraclavicular, retroauricular, and transoral approaches. These do not fall under the MIP umbrella.

Bilateral neck exploration In patients where gland localization fails or when the imaging findings are discordant, a four-gland exploration is needed. This approach is also used for more complex pathology such as familial and hereditary causes of HPTH where subtotal or radical total parathyroidectomy is required. In these cases, the principles of surgery are based on achieving normocalcaemia for as long as possible, avoiding hypoparathyroidism and planning for inevitable recurrence. Three or three and a half parathyroid glands are removed, or all parathyroid glands are removed with heterotopic autotransplantation of a single gland is performed to avoid hypoparathyroidism.

Intra-operative localisation of parathyroid glands

Parathyroid glands possess endogenous fluorophores which result in their ability to exhibit near infrared autofluorescence (NIRAF) in comparison to surrounding tissues. In recent times, technologies such as probe-based detection systems and image based detection systems have been developed. They leverage the NIRAF capabilities of the gland to facilitate real time intra-operative localisation of parathyroid glands.

Postoperative follow-up

Patients are reviewed in the outpatient department afterwards for wound inspection, assessment of vocal cord function, and discussion of pathology. Cosmetic outcomes of scars are comparable between the minimally invasive techniques but superior in the remote surgical approaches as the latter avoid an incision in the neck. Postoperatively, if serum calcium and PTH levels have normalized and histology confirms a parathyroid adenoma, further clinical follow-up is rarely required. If histology suggests parathyroid hyperplasia, endocrine follow-up is generally recommended because of the risk of recurrent PHPTH and in those patients in whom serum calcium or PTH levels do not normalize postoperatively, further investigations and consideration for revision surgery may be required.

Revision parathyroid surgery

The indications for revision surgery are the same as for primary surgery. However, the added morbidity of surgery means further

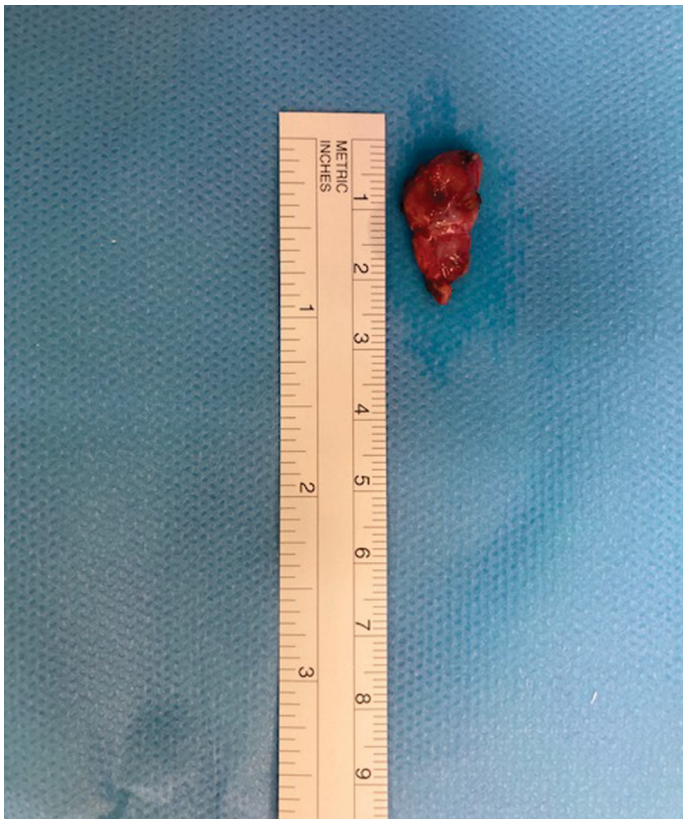

Figure 63.4 Image of a resected parathyroid adenoma.

intervention ought to be carefully considered with the input of a multidisciplinary team. The endocrinologist and surgeon should weigh up the risks and benefits of surgery and the option of non-surgical management should also be considered and discussed with the patient. A missed hyperfunctioning gland is a more common cause for PHPTH refractory to treatment than a supernumerary gland. The incidence of the latter is only about 1%.[15]

Preoperative workup is more rigorous and involves laryngoscopy and localization studies. Ultrasound, MIBI scan, SPECT, magnetic resonance imaging, and 4D-CT are complementary, and careful assessment of the mediastinum should be performed. In complex cases where cross-sectional imaging is not successful, parathyroid venous sampling and parathyroid arterial angiography, performed with appropriate expertise, is useful.

Positron emission tomography radiotracers have been evaluated for the localization of parathyroid disease and the most commonly used tracers include carbon-11 (^{11}C)-methionine[16] and fluorine-18 (^{18}F)-fluorocholine.[17] The sensitivity of ^{11}C-methionine has been reported in the range of 79–90%.[16,18] ^{18}F-fluorocholine is a marker of cellular proliferation and its longer half-life provides more flexibility in clinical practice.[18] At present, the utility of these tracers in combination with positron emission tomography/CT is limited to a few specialist centres and should be reserved for complex cases including those patients who have undergone at least one previous unsuccessful surgery.

The surgical approach will depend on preoperative localization and the previous surgical intervention. Indeed, the success of surgery including the avoidance of hypoparathyroidism depends very much on preoperative localization and the experience of the operating surgeon. An intraoperative PTH assay also contributes to improved surgical outcomes.

Parathyroid autotransplantation is used to avoid hypoparathyroidism and is often offered when there is certainty that no other parathyroid tissue is present in the neck. It is most commonly offered during revision surgery for MEN.

Controversies

Intraoperative nerve monitoring

Monitoring of the recurrent laryngeal nerve during surgery is advocated by some as a gold standard and should particularly be considered for revision parathyroid surgery.

Cryopreservation of parathyroid glands

This technique involved cryopreservation at the time of initial surgery followed by later transplantation when the need arises. There are few indications, and the technique has not shown reproducible success. Robust options for medical management of hypocalcaemia and concerns regarding graft failure has resulted in few centres offering this intervention.

Secondary and tertiary hyperparathyroidism

Many patients with these conditions are asymptomatic. In patients with secondary HPTH, calcium levels will be low or normal. They may also have extremely elevated intact PTH levels and decreased vitamin D levels. Patients with tertiary HPTH will have normal or elevated serum calcium concentrations in combination with elevated PTH levels. They may also have decreased vitamin D and phosphate levels and elevated alkaline phosphatase.

When symptomatic, untreated secondary HPTH leads to progressive bone disease. Patients with tertiary HPTH may experience bone pain or fractures, nephrolithiasis, pancreatitis, soft tissue or vascular calcifications, pruritus, and mental status changes. DXA scans may show changes consistent with osteopenia or osteoporosis.

Treatment of secondary HPTH is primarily medical with surgery rarely having a place except in cases of medical failure or calciphylaxis.

Treatment of tertiary HPTH is primarily surgical and is usually by way of subtotal parathyroidectomy or total parathyroidectomy with autotransplantation.

Conclusion

The management of parathyroid disease occupies a range of complexities and is best managed in a multidisciplinary setting. Outcomes of investigations and treatment are tied to the expertise within these multidisciplinary teams who, over time, have developed a responsive and capable service.

FURTHER READING

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64

Principles of head and neck reconstruction

Vijay V. Haribhakti

Introduction and basic principles

Reconstruction and restoration of function and quality of life are the fundamental pillars that support all major head and neck surgical endeavours. Over the past four decades, the greatest advances in the entire domain of head and neck surgery have been in the areas of reconstruction and rehabilitation, permitting routine performance of procedures hitherto regarded as mutilating and life-changing.

No longer is it acceptable for any patient to be left functionally impaired or an 'aesthetic recluse', even after a massive surgical resection.

Indeed, it is crucial to establish a completely holistic plan of treatment at the very start, such that the primary head and neck surgeon performs an *'ideal' resection* to create an *'ideal' defect* for the reconstructive surgeon, who in turn chooses the best alternative to provide an *'ideal' outcome*, which enables both adjuvant treatment in order to maximize survival and disease control, and a return to normal life.

The ultimate goal of the entire treating team is to effectively save life and restore function to achieve good quality of life.

Basic principles of sound reconstruction are included in **Box 64.1**.

Every patient undergoing a major resection must be routinely assessed by both teams to completely understand the functional and aesthetic impacts of the tissues that need resection, and to make a realistic plan of reconstruction based upon the patient's unique needs and expectations. In this exercise, the importance of detailed radiological assessment to establish the true extent of disease is obvious.

There should routinely be a dialogue between both teams to visualize the extent and complexity of the defect that will result, and to choose the most ideal reconstructive alternatives for a given case, based upon individual needs and physical characteristics such as quality of tissues, obesity, comorbidities, fitness levels, expectations, and, importantly, comfort zones of the reconstructive team (**Box 64.2**).

Box 64.1 Basic principles of sound reconstruction

- Detailed clinical and radiological assessment.
- Anticipation of the defect in consult with the resection team.
- Estimating individual patient needs and expectations.
- Assessing reconstructive alternatives and choosing the most ideal one.

Box 64.2 Assessment and preparation

- On-table assessment of defect details and requirements.
- Preparing the 'right' flap/tissue needed for reconstruction.
- Executing the best inset possible.

Frequently, the size and demands of a given defect differ significantly from those anticipated. This may happen due to inadequate preoperative clinical assessment owing to factors such as trismus and submucous fibrosis, which are 'endemic' in certain populations, or to unexpected disease extensions found on frozen section assessment of margins. Hence, it is optimal to plan the *final flap dimensions by means of a pattern* only after the defect has been created, even when simultaneous harvest is undertaken.

Having raised an optimal flap, a very careful *primary inset* is absolutely crucial in order to ensure best, complication-free outcomes.

In those patients where free flaps are necessary, microvascular anastomosis has to be a very meticulous exercise with absolute attention to smallest details. Once the flap is revascularized and perfusing satisfactorily, it is important to turn attention to the *lie* of the *pedicle*, ensuring a free flow of blood without turbulence and external compressions. In all pedicled flaps, suitable safety measures need to be taken to ensure correct lie and to *avoid pedicle compression* (**Box 64.3**).

The immediate and early postoperative periods are vital, and every flap must be carefully monitored to detect interruptions in blood flow, which demand immediate intervention for correction. The position of the head, the planning of dressings and drain fixations, and planned mobilization are all extremely important.

Box 64.3 Safety measures

- Microvascular anastomosis with free flaps.
- Pedicle protection following haemostasis and closure.

Box 64.4 Roles of the rehabilitative team

- Vigilant postoperative monitoring.
- Rehabilitation as needed.

The rehabilitative team, who have ideally seen the patient ahead of surgery, come into action in the early postoperative period, and ensure efficient implementation of all measures to restore mobility and function (Box 64.4).

Fundamentals of decision-making

When encountering any defect, a number of questions need to be answered, as listed in Box 64.5.

Box 64.5 Questions when encountering any defect

- Is primary tension-free closure possible?
- Is it possible to achieve a satisfactory outcome with a skin graft/equivalent?
- Is primary reconstruction necessary?
- Is a flap needed? If yes, which one would be ideal, depending upon size and complexity of the anticipated defect as well as donor site qualities:
  - Local?
  - Regional?
  - Distant?
- In the event of flap loss, what would be a possible lifeboat/saviour?

The 'reconstructive ladder'

We often speak of a 'reconstructive ladder', based upon the inherent requirements of each defect, keeping in mind the ultimate recovery goals mentioned above.

Accordingly, there are certain *defects that are best left alone* or simply covered with a biological material such as 'Kollagen' which provides temporary cover while the natural process of epithelialization is underway.

Superficial carbon dioxide laser excisions over the cheek mucosa, palate, and tongue, without loss of major substance, with healthy and vascularized tissue at the floor of the defect, are very adequately treated in this manner. In numerous centres, even moderate-sized tongue defects are treated using this approach, mainly to prevent tethering and loss of function. Defects of the palatal mucosa, with intact bone and sound oronasal separation, are also treated this way with consistent success.

A very important factor that determines such choices is the quality of the native mucosa. In the presence of submucous fibrosis, healing occurs with dense fibrosis, giving rise to thick, unyielding scar formation which detracts from good functional recovery.

Figure 64.1 demonstrates a 29-year-old lady with no habits or addictions, who developed a variegated lesion on the lateral aspect of the tongue, covering a considerable area of the junctional tongue. A superficial excision was required to address this lesion (which was ultimately reported as a carcinoma *in situ*). Primary closure would clearly hamper function, and so we chose to go ahead with a carbon dioxide laser excision and covered the substantial raw area with Kollagen. The patient had uneventful healing and returned to normal function.

Figure 64.1 (a) Variegated primary lesion. (b) Peroperative view. (c) Large post-excisional defect. (d) Kollagen used to cover raw area.

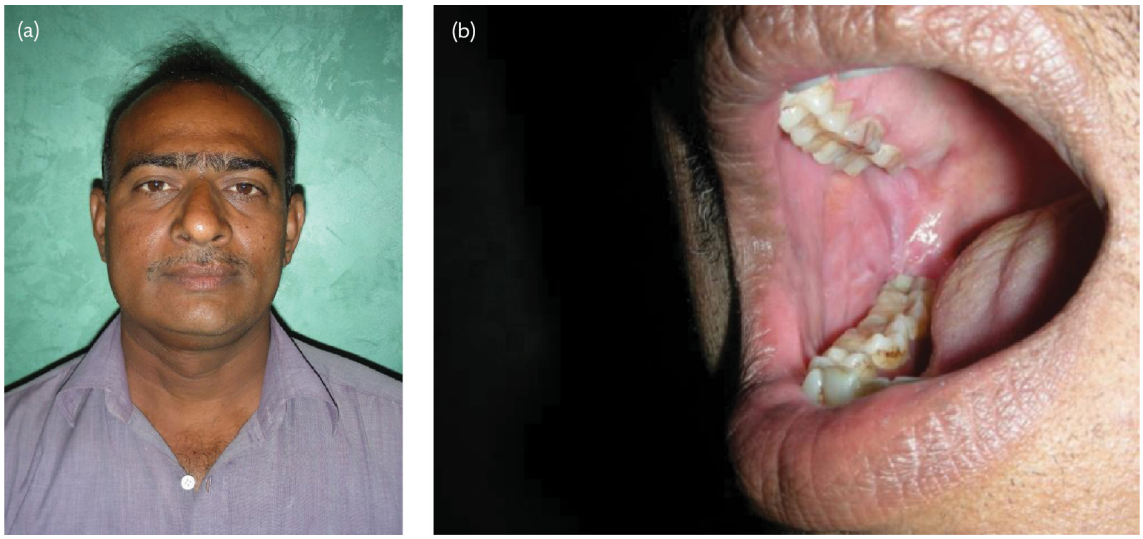

Figure 64.2 (a) Final appearance, front. (b) Adequate mouth opening, scar inconspicuous.

Wherever possible without tension, *primary closure* is an excellent option. With defects of the cheek mucosa, it is crucial to ensure that adequate mouth opening remains possible after primary closure. Failing this, trismus is likely and must be avoided. If sutureline dehiscence is anticipated with immediate, excessive mouth opening, a nasogastric feeding tube is often recommended and the patient counselled in detail to give sufficient rest before resuming mastication.

Figure 64.2 demonstrates the end results in a 35-year-old old male with a T2N1 lesion of the right cheek, which was managed with primary closure. The final outcomes post radiotherapy are seen.

For laterally based tongue defects, carefully planned primary closure of small and selected medium-sized defects works very well and preserves excellent function of the remnant, albeit with occasional foreshortening.

Figure 64.3 demonstrates a 69-year-old male who presented with a cT2 N0 lesion of the right lateral border tongue. A wide-local excision with selective neck dissection (I–IV) was performed. As all margins were clear and nodes negative, with a DOI of 6 mm, patient was on surveillance only. The 1-year result is seen:

With defects of the cheek mucosa that are large in surface area but have an intact layer of buccinator muscle beneath, *split-thickness skin grafts* work well. Being a mobile area though, it is essential to quilt the graft and use a good bolus tie-over in order to secure stable fixation. In all cases, a temporary feeding tube is inserted to maintain oral hygiene and provide rest and relative immobilization to the grafted area. Figure 64.4 demonstrates management of a middle-aged male with a large, verrucous/well-differentiated ca involving the left commissure and a substantial portion of the cheek, without deep invasion. A split skin graft was carefully quilted into place and held down with a tie-over. The 4-week result is seen.

For superficial cheek defects that are significant in size and liable to heal with graft contracture and trismus, we frequently employ a full-thickness skin graft (FTSG) harvested from the supraclavicular area. This is an excellent donor site that heals extremely well with a barely visible scar, and the graft retains elasticity and pliability, so necessary for restoring mouth opening and averting trismus. We have often used the FTSG even over a well-vascularized suctorial fat pad, with good outcomes. In all cases, it is essential to quilt the graft and employ effective fixation in order to achieve a good take.

Figure 64.5 demonstrates a young male who presented with a T2 squamous cell carcinoma in the right posterior cheek. Wide local

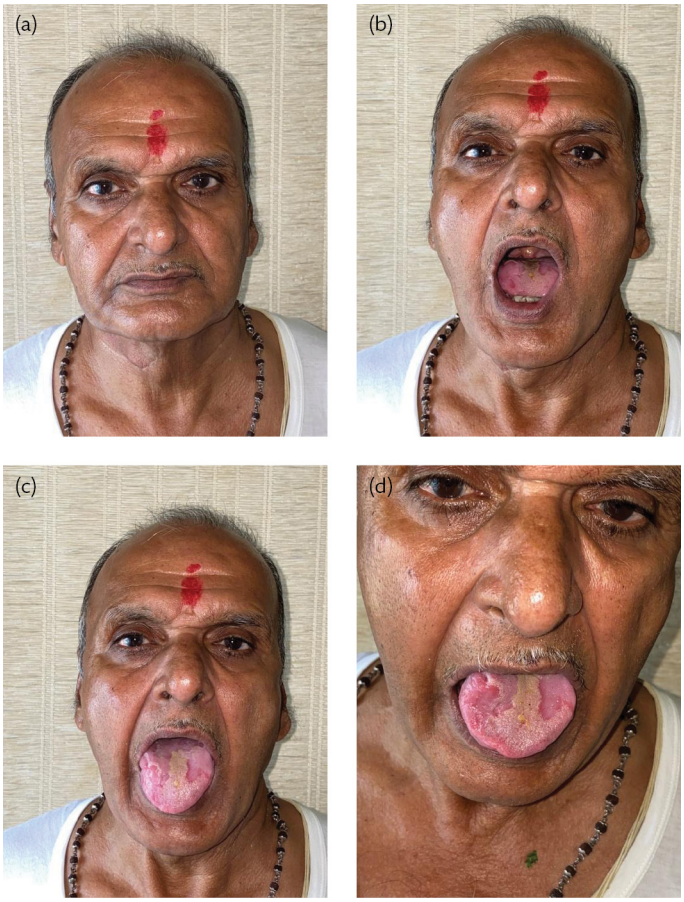

Figure 64.3 (a) One-year post-surgery. (b) View of tongue, within mouth. (c) The 13-month outcome. (d) Close-up view, restored tongue.

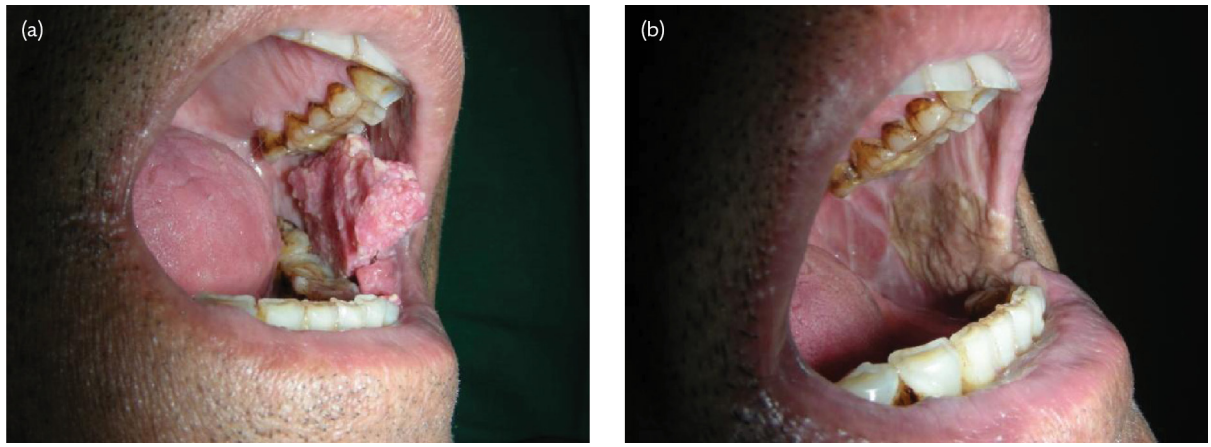

Figure 64.4 (a) Proliferative left cheek lesion: well-differentiated squamous cell carcinoma. (b) Final outcome, post split-thickness skin graft repair.

Figure 64.5 (a) Final functional outcome: FTSG. (b) Open mouth view demonstrating FTSG. (c) Close-up view: well-established graft. (d) Left supraclavicular donor area.

excision incorporated the buccinator muscle. The suctorial pad was brought down and covered with a FTSG. Excellent long-term outcomes are seen.

For relatively small and superficial external defects over the nose and forehead, which have an adequately vascularized base, FTSGs work very well too. Colour match is crucial, and supraclavicular skin most often achieves it. If not, postauricular skin may be harvested.

Figure 64.6 shows a lady in her sixties, who presented with a basal cell carcinoma over the left nasal ala, just behind the tip. The perichondrium-deep defect was resurfaced with a FTSG. Final outcomes were satisfactory.

Once we are confronted with a *defect* of such *size and complexity* that all the above-described techniques are likely to prove inadequate, we need to plan *flap reconstruction*. A very large variety of flaps are available, and must be appropriately selected following a detailed consideration of defect requirements, donor site availability, and suitability in a given individual. This applies to virtually all defects anywhere in the head and neck area, whether of the mucosal lining, the external skin, or bone, or complex composite defects.

The major priorities for *oral reconstruction* are as follows:

- To ensure adequate *mouth opening.*
- To achieve sufficient *commissure diameter.*
- To assure *oral competence.*
- To restore normal *occlusive relationships.*
- To preserve *lingual function* for efficient mastication, speech, and deglutition.
- To provide for *dental rehabilitation* whenever possible.

For *external defects* over the face/neck, it is essential to provide tissue of matching size, colour, texture, and properties in order to provide optimal aesthetics.

For medium-sized defects that require vascularized tissue replacement, a variety of local flaps are extremely useful and require perfect excision design in order to permit a suitable fit, as demonstrated.

'Workhorse techniques'

Local skin flaps

A large number of designs of local skin flaps are described in the face and neck area, the commonest among which for small to moderate-sized defects is the *Limberg* flap, a *transposition flap* rhomboidal in design, which has to be accurately planned in order to plot scars in the most ideal planes.

Figure 64.7 demonstrates a substantial 2 cm defect of the lateral forehead following rhombic excision of a basal cell carcinoma, repaired with a Limberg flap, some scars being hidden within the hair-line.

Another interesting and highly useful flap is the *island nasolabial flap*, based on the profuse, random vascularity of the face in addition to the *facial artery* that nourishes the *nasolabial* flap. In general, the author does not prefer to use nasolabial flaps for intraoral defects except in individuals with lax, hairless skin wherein no prominent scars would result, and problematic intraoral hair growth is avoided.

Figure 64.6 (a) Patient with basal cell carcinoma of the nasal ala. (b) FTSG repair, front face appearance. (c) FTSG repair: profile. (d) Donor site: inconspicuous scarring.

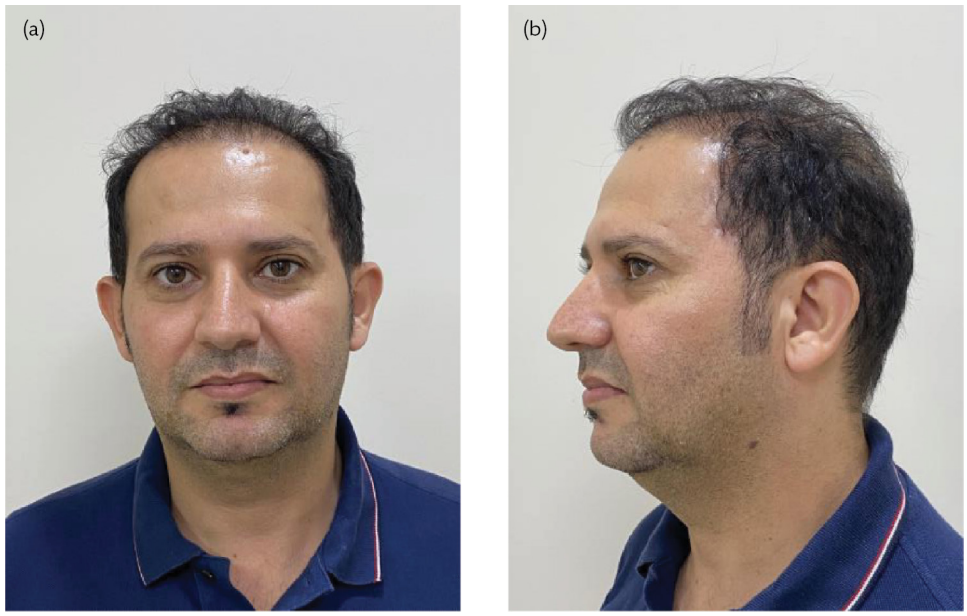

Figure 64.7 (a) Lateral forehead defect: Limberg flap repair. (b) Profile view: demonstrates scar placement.

Figure 64.8 demonstrates use of a nasolabial flap for a large defect over the nasal bridge, down to cartilage, following excision of a localized sarcoma.

Figure 64.8 (a) Deep nasal defect and planning. (b) Nasolabial flap raised and transposed into defect. (c) Close-up view, immediately postoperative. (d) Early result post surgery and radiotherapy.

We also have *rotation–transposition* flaps such as the *bi-lobed* flap as demonstrated in Figure 64.9.

Intraoral: facial artery myomucosal flap

Figure 64.10 demonstrates management of a thick, plaque-like lesion (squamous cell carcinoma) in the lower gingivobuccal sulcus in a young patient in his thirties. Wide local excision with clear margins left a substantial defect which was repaired as shown.

Figure 64.11 demonstrates use of the facial artery myomucosal flap for reconstruction of a small but highly significant defect in the anterior and lateral floor mouth, following resection of a 6 cm adenoid cystic carcinoma originating in the left sublingual gland in a 35-year-old male. This patient returned to normal appearance and function.

Lip and commissures: the Karapandžić technique

This highly versatile technique enables excellent outcomes following resection of significant amounts of lower lip (Figure 64.12). In essence, this technique enables neurovascular islands of lip tissue to be moved, preserving sensation, aesthetics, and function. It is important to achieve sufficient commissure diameter and mouth opening following resection and reconstruction in such cases.

Regional flaps

A large number of regional skin flaps have been described, of which few have retained relevance in the 'free flap' era.

The *forehead flap*, based on the anterior division of the superficial temporal artery, was among the first and provided reliable tissue of large surface area. Currently, it is of historical interest only.

The *deltopectoral flap*, based on perforators of the internal mammary artery, described by Bakamjian, remained in use for many years, but has fallen out of favour over the last quarter century as it entails a second division procedure.

The *submental flap* based on the submental branch of the facial artery, is a pliable flap with good vascularity. The author, however,

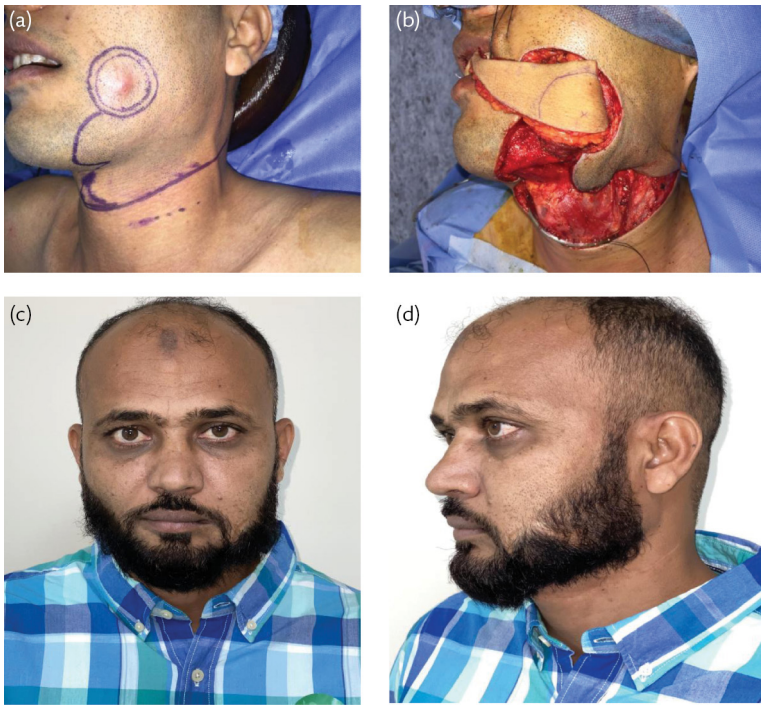

Figure 64.9 (a) Marking for a T4a cheek cancer in a young male: the bilobed flap for outer cover. (b) Large mucosal and soft tissue defect repaired with a free anterolateral thigh flap. (c) Final outcome demonstrating contour. (d) The bilobed flap provided a 'natural' result over the face.

Figure 64.10 (a) Thick lesion, lower central gingivobuccal sulcus. (b) Significant defect, post excision. (c) Primary inset: facial artery myomucosal flap. (d) Final aesthetic outcome.

Figure 64.11 (a) Small but significant anterior floor of mouth defect facial artery myomucosal flap elevated. (b) Primary flap inset as shown. (c) Front face view post treatment (surgery + radiotherapy). (d) Intraoral view of facial artery myomucosal flap.

Figure 64.12 (a) Proliferative, bulky lateralized lower lip lesion. (b) Post-excisional defect. (c) Final, on-table result. (d) Appearance at 8 days.

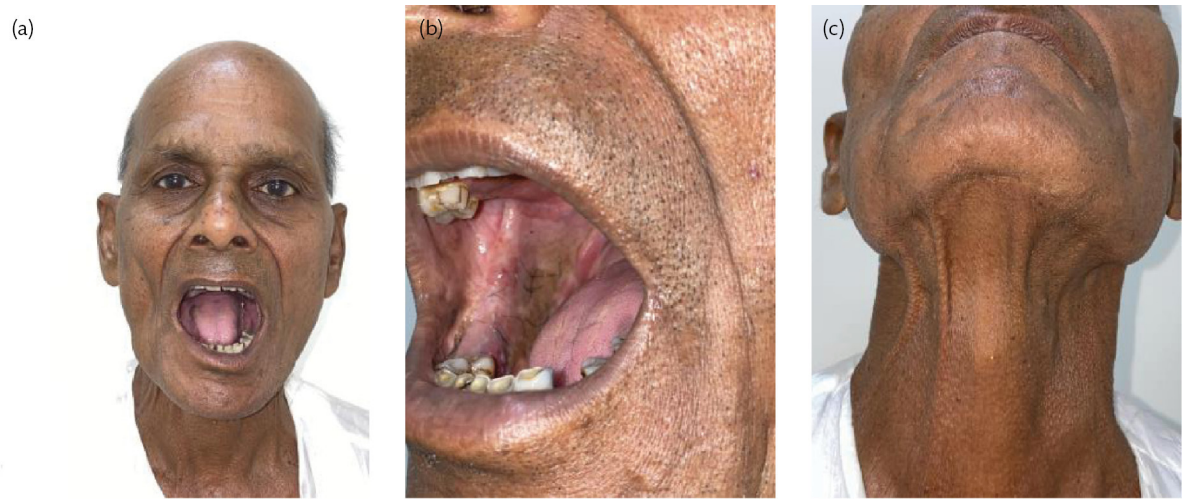

Figure 64.13 (a) Normal mouth opening. (b) Well-settled flap. (c) Donor defect: completely hidden.

considers its use inappropriate in all cases where a neck dissection is considered necessary, and in male patients with dense hair follicles, owing to the profuse hair growth that results intraorally. Figure 64.13 demonstrates outcomes after a whole right cheek defect, repaired with a submental flap.

The *supraclavicular flap*, based on branches of the transverse cervical artery, is useful in theory but seldom finds application in most hands.

The regional flap that has truly stood the test of time and continues to be in use all over the world is the *pectoralis major myocutaneous* (PMMC) flap. In our hands, it is still a preferred flap for pharyngeal augmentation in suitable individuals and as a source of well-vascularized muscle for a variety of indications including primary pharyngeal closure post radiotherapy/chemoradiotherapy, risk of carotid exposure in hostile necks, and repair of large-volume, recalcitrant lymphatic leaks following thoracic duct injury, etc. Most importantly, it is an amazing lifeboat following failed free flaps, and is also extremely useful for providing external cover in large neck defects impossible to close primarily without tension.

Figure 64.14 illustrates utility of the PMMC flap even in a large-volume composite cheek–mandible defect involving a segmental mandibulectomy.

Free flaps

Over the past three decades, the availability of free tissue transfer has completely revolutionized the field of head and neck reconstruction. It is now possible to tailor-make a reconstruction responding to the unique needs of any defect and the preferences of the individual. Although a large number and variety of free flaps are now available, there are a few workhorse flaps that are integral to any reconstructive armamentarium.

Free radial forearm flap

The free radial forearm flap (FRAFF) is an amazingly reliable and versatile flap that can be brought in for a variety of defects, with predictably good outcomes.

Figure 64.15 illustrates a 45-year-old gentleman who presented with a massive lesion (verrucous and well-differentiated squamous carcinoma) over the tongue, floor mouth, alveolus, cheek and oropharynx. A very complex, contoured defect resulted (Figure 64.15c), which was repaired with a FRAFF, with satisfactory outcomes.

Anterolateral thigh flap

The anterolateral thigh (ALT) flap is perhaps the most widely used free flap today, and is highly dependable and versatile for all varieties of significant soft tissue defects. It is customary to harvest simultaneously while the excision is ongoing, permitting ease and efficiency, and finalizing the posterior incision only after the defect is created. A variety of designs are possible, including chimeric flaps for complex defects.

The bulk provided by this flap frequently makes it a good solution in those patients where bony reconstruction cannot be offered.

Figure 64.16 demonstrates ALT flap reconstruction in an 18-year-old male presenting with a sarcomatous lesion involving the cheek, retromaxillary space, and infratemporal fossa.

Fibular osteocutaneous flap

Without doubt, the fibula has become the versatile workhorse for all cases needing bony reconstruction, whether of the mandible or the maxilla.

In addition, a good understanding of perforator anatomy has now made it possible to transfer large areas of skin and soft tissue with complete ease, often as chimera, as three-dimensionally independent skin paddles, in addition to bone.

Figure 64.17 demonstrates reconstruction of a large subtotal mandibulectomy defect for a T4a lesion of the lower alveolus, involving the central arch.

The fibular osteocutaneous flap can be employed for highly complex and difficult defects as demonstrated in Figure 64.18. This lady, aged 55 years with no addictive habits but with oral lichen planus, had multiple osseous and soft tissue lesions within the oral space, which needed a subtotal mandibulectomy (angle to angle) as well as a left infrastructure maxillectomy. She had a one-stage reconstruction of this very complex defect with a fibular osteocutaneous flap.

Figure 64.14 (a) Segmental mandibulectomy plus whole cheek defect. (b) Flap planning with major anatomical landmarks, illustrating potential for contour restoration. (c) Early postoperative outcome. (d) Close-up view face and neck. (e) Adequate mouth opening. (f) Close-up view demonstrating inset.

Figure 64.15 (a) Extensive lesion over numerous anatomical areas. (b) Large excision specimen with mandible rim. (c) Large, complex, contoured defect. (d) Flap elevated, with large adipofascial component. (e) Final postoperative outcome. (f) Adequate functional restoration.

Figure 64.16
normal mouth opening.

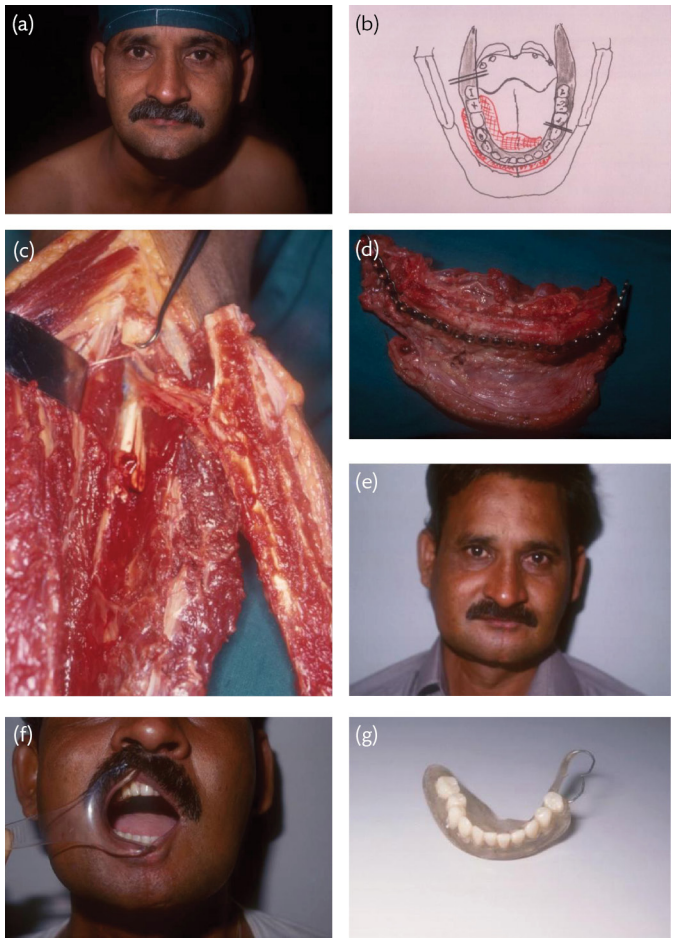

Figure 64.17 Preoperative picture of a young, 38-year-old male with a large T4a lesion of the lower alveolus. (b) Illustration depicting the extent of tumour and mandible resection needed. (c) Fibular osteocutaneous flap elevated on peroneal pedicle. (d) Osteotomies complete, flap ready for transfer. (e) Final aesthetic outcome. (f) Functional outcome. (g) Partial denture fabricated.

Figure 64.18 (a) Final aesthetic outcome. (b) Functional outcome. (c) Soft tissue repair, alveolar defect. (d) Palatal repair, with oronasal separation.

Pharyngeal reconstruction

The major priorities of pharyngeal reconstruction include the following:

1. Prompt restoration of effective swallowing.
2. Ability to withstand adjuvant radiotherapy/chemoradiotherapy.
3. Minimize incidence of pharyngocutaneous fistula.
4. Minimize risk of strictures, early and delayed.
5. Permit effective voice rehabilitation.

Following laryngectomy alone, primary pharyngeal repair is often possible. The simple measures for mucosal sufficiency are either a measurable width of 3 cm of unstretched mucosa at the narrowest portion or an ability to tube the mucosa over a 34–36 French tube without tension.

If it is a salvage laryngectomy, especially after radiation therapy with concomitant chemotherapy, we prefer to cover the suture line with a muscle flap (pectoralis major) for additional vascularity.

Following laryngectomy with subtotal pharyngectomy for pyriform lesions, the above conditions for mucosal sufficiency can seldom be met, and some form of pharyngeal augmentation is needed.

The PMMC flap is readily available in the near vicinity and can be used in male patients without an excess of subcutaneous fat. In obese males and in females, we prefer a fasciocutaneous free flap, either the ALT or the FRAFF.

Figure 64.19 demonstrates the case of a short-statured, obese lady with a T4N3b hypopharyngeal cancer, who was left with just a narrow strip of posterior pharyngeal wall mucosa. We used a FRAFF for reconstruction, along with a primary tracheo-oesophageal puncture.

She went on to develop excellent swallowing as well as TEP speech, despite the need for adjuvant chemoradiotherapy.

For circumferential pharyngeal defects, the critical question to be asked is, is the defect above the thoracic inlet? If not, gastric transposition is the answer. If yes, it can either be a free jejunal graft or a tubed fasciocutaneous flap.

Figure 64.20 demonstrates use of a jejunal graft, a highly efficient conduit, albeit with a few drawbacks.

Although the jejunum restores excellent swallowing, it secretes excessively, rendering speech rehab poor. These patients are forced to use an electrolarynx.

In recent times, we have tended to move to tubed ALT flaps, which allow better voice rehabilitation with a tracheo-oesophageal puncture prosthesis.

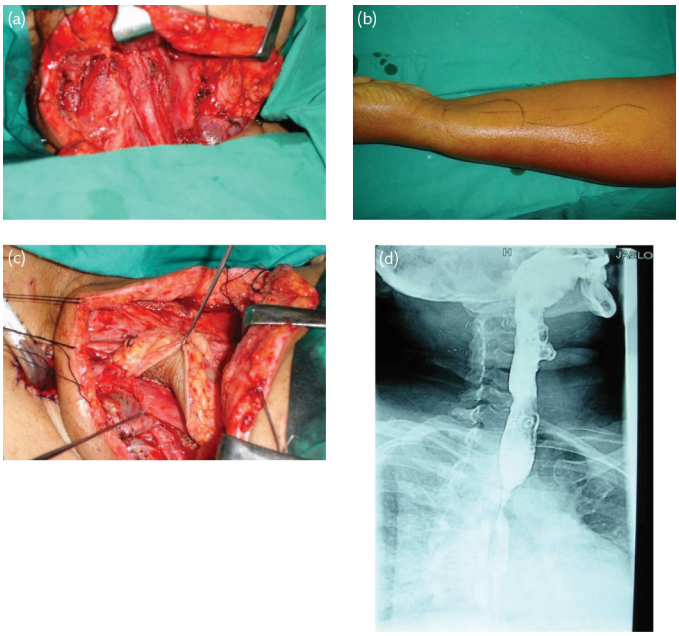

Figure 64.19 (a) Near-total pharyngeal defect. (b) FRAFF marking. (c) Flap inset. (d) Postoperative barium swallow.

Individual techniques

A very large number and variety of techniques are available for head and neck reconstruction, and details of each need to be mastered by review of the excellent sources available. Technical nuances of each, although extremely important, are beyond the scope of this chapter.

Apart from the 'workhorse flaps' cited above, there are numerous other alternatives available, such as the lateral arm flap, the latissimus dorsi flap, the rectus flap, and the deep inferior epigastric perforator flap, all of which are well described in the literature cited.

Controversies

Most controversies pertain to choices we need to make in every case and are, quite frankly, impossible to avoid completely.

Participative decision-making, patient and family education, detailed counselling, and rehabilitation efforts all go a very long way in building patient confidence and trust.

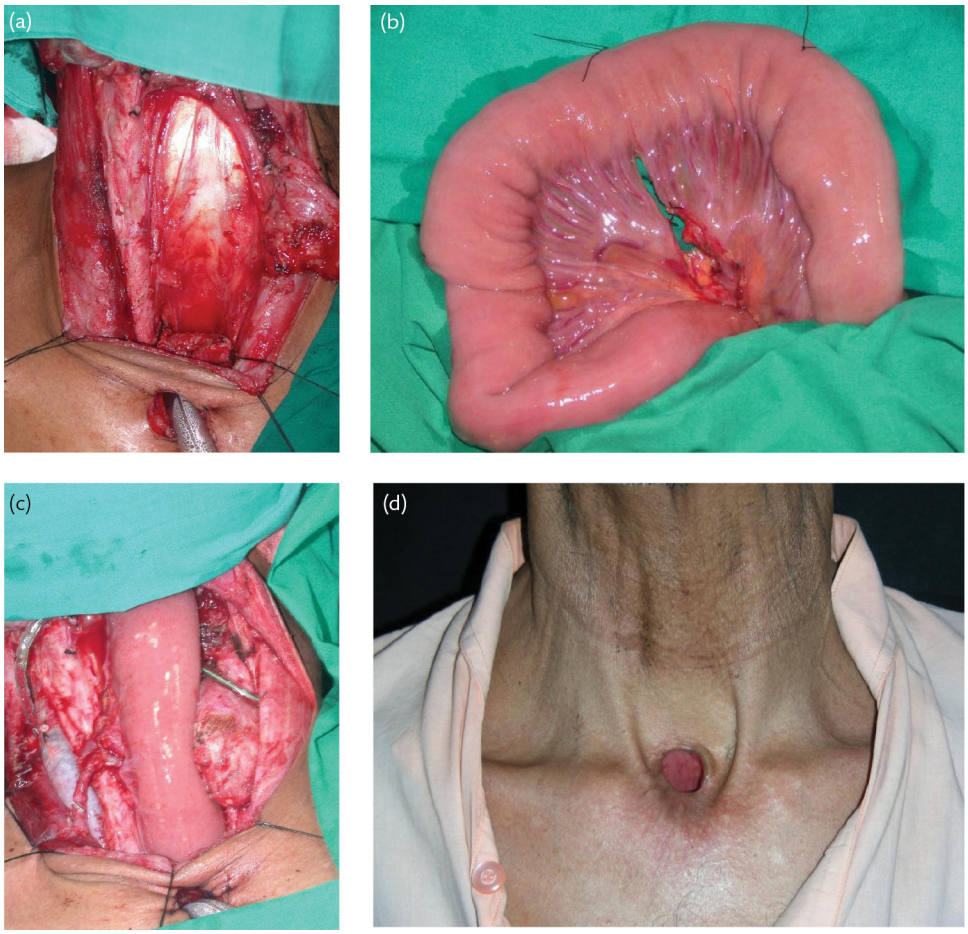

Figure 64.20 (a) Circumferential defect. (b) Jejunal segment, to be transferred iso-peristaltic. (c) Graft with microvascular anastomosis completed. (d) Final outcome at 2 weeks.

Tips and pearls

'Begin with the end in mind'

This is perhaps the most important pearl of reconstructive surgery. Both the excision as well as the reconstructive teams must come together a priori to 'imagine' the defect completely, set forth a detailed plan, and conceptualize the end result, keeping in mind all the hurdles.

'Assess the recipient and donor sites at great length'

This is absolutely crucial in every case. It allows a more complete 'preparedness' for the actual procedure, and optimal selection of donor tissue based on anticipated defect dimensions and complexity. We cannot follow a 'cookbook' approach. There needs to be careful individualization in each case.

'Reassess, remeasure thoroughly post excision, and prepare a pattern'

Oftentimes, defect dimensions and complexity are at variance with initial estimations. Thus, each defect has to be measured and treated individually to ensure that tissue of the correct dimensions and quality is prepared for transfer. Whether in terms of shape and size/need for de-epithelialization/preparing chimera, and so on, an accurate pattern should be created to ensure accurate reconstruction.

'Remember: the primary inset represents the best opportunity'

The primary inset is extremely important. Later changes are frequently difficult or impossible.

'Be obsessive about anastomotic technique and pedicle lie'

The majority of microvascular occlusions are technical failures and must be minimized by accurate and complete attention to detail, and once adequate perfusion sets in, the pedicle lie must be accurately arranged to avoid torsion, kinks, and external compression.

'Supreme vigilance, prompt decisions postoperatively'

The initial 24–48 hours are critical, and complete vigilance is essential. In centres where it is available, audible monitoring of pulsations is performed. Alternatively, the surgeon should at the conclusion of the operation, mark a point over the skin where pedicle pulsations are best detected with a surface Doppler. The nursing staff are taught how to monitor frequently and inform the surgeon immediately if any changes are observed.

'Rehabilitation is the true icing on the cake'

Every patient undergoing major reconstruction, and most certainly all patients needing further adjuvant treatment such as radiotherapy or chemoradiotherapy, should routinely be assessed by the rehabilitation team and measures decided to ameliorate symptoms and ensure prompt functional restoration.

'Review, review review . . . and honestly critique every outcome'

It is critical to note all complications, problems, and failures and carefully document in every case. Though inherently subjective, such quality-of-life evaluations are vital and enable us to attend to subtle needs which are important for each patient.

Common complications and their effective prevention

Complications and sequelae are an immutable part of the work we do as reconstructive surgeons, and remind us of our inherent fallibility. Nonetheless, every attempt must be made to minimize them.

The commonest complication in reconstructive surgery is *flap necrosis*.

In local and regional flaps, *partial necrosis* is frequently found and one has to decide if conservative measures will suffice.

With free flaps, *total necrosis* is typically encountered and most often needs re-exploration, debridement, and replacement with a new flap.

In the head and neck area, we find the *PMMC* flap to be one of the *best lifeboats* available.

At all times, it is crucial to act before sepsis sets in, with significant attendant morbidity.

In order to avoid major issues, appropriate preoperative counselling and consent are necessary for all reconstructive surgery.

Adverse functional and aesthetic outcomes are to be expected in any busy reconstructive practice, and need detailed analysis, complete counselling, and rehabilitation efforts to encourage patients to adapt to life with clear adaptive strategies. In carefully selected instances, patients may be offered major corrective procedures, following exhaustive planning and counselling.

Future directions

We are now in an era where success rates in excess of 90–95% should be the norm. Hence, future directions clearly take us to the exciting field of rehabilitation, where truly no limits exist. Dental and masticatory rehabilitation is a clear case in point. Most centres practise osseointegration, and ongoing efforts are being made to incorporate implants even before flap transfer to optimize functional outcomes.

Although we have a large armamentarium of flaps available today, the search continues, to seek donor sites with optimal reliability and minimum morbidity.

For patients with adverse outcomes, revision procedures and strategies will become a norm, all finally aimed at enhancing quality of life.

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65

Non-surgical treatments for head and neck cancer

Muhammad Shahid Iqbal and Charles Kelly

Introduction

The management of head and neck cancer (HNC) is complex and often involves a combination of surgery, radiotherapy, and chemotherapy. Radiation therapy is an important modality for managing patients with locoregional HNC. A multidisciplinary approach is required for optimal shared decision-making with each patient on an individual case-to-case basis.

Principles of radiotherapy

The tumouricidal effects of ionizing radiation work through producing ionized atoms which disrupt molecular bonds, which in turn cause wide-ranging molecular damage throughout the cells. This process leads to the production of free radicals and irreversible DNA damage by generating double-stranded breaks in DNA. In tumours, loss of proliferative ability due to this DNA damage leads to tumour cell death. Radiation not only damages tumour cells but also normal cells. The reason for radiotherapy being a successful cancer treatment lies in the fact that normal cells are, in general, better able to repair the damage and subsequently resume normal function. However, normal tissues have certain limits on the radiation dose which they can safely tolerate. These limits are crucial in helping to determine the maximum dose which can be safely administered to the tumour during the course of treatment.

Practical considerations for head and neck radiotherapy

The practice of precise radiotherapy delivery requires specific radiotherapy volumetric planning skills and input from the dosimetry (radiotherapy planning) and radiation physics teams.

The practical steps for state-of-the-art precise radiotherapy delivery for patients with HNC are as follows:

1. *Dental assessment*: this is an essential part of the pre-radiotherapy management and any teeth with a poor long-term prognosis should be removed prior to commencement of radiotherapy. Dental extraction is not recommended within 1 year of radiotherapy, if the tooth is in the radiotherapy volume. One of the potential long-term side effects from radiotherapy on mandible and teeth is osteoradionecrosis (ORN) due to radiation damage to the blood vessels supplying the bone and the osteoblasts needed for bone repair, causing devitalized bone to present as a non-healing ulcer. ORN typically affects the mandible, presenting 2–4 years after the radiation is complete. Although the incidence of ORN is not high, the risk can be reduced by pretreatment dental assessment and intervention as appropriate, better dental care during and after radiotherapy, and sparing the salivary glands; xerostomia, secondary to radiation damage to the salivary glands, contributes towards the development of ORN. With modern radiotherapy techniques such as intensity-modulated radiotherapy (IMRT), the dose delivered to the parotid and submandibular glands can be reduced, unless the cancer or involved nodes are immediately adjacent to these structures. However, it is not possible to spare the minor salivary glands spread throughout the oral cavity. Radiotherapy-induced xerostomia can also have a detrimental effect on the teeth, causing radiation caries which are characterized by appearing around the neck of the tooth.
2. *Patient immobilization*: this is a basic first principle of radiotherapy delivery. The patient must be in a position which is comfortable and reproducible throughout the treatment. In HNC radiotherapy, immobilization is generally obtained by making a thermoplastic shell that is a precise fit around the patient's head and neck. A planning computed tomography (CT) scan at this stage permits 'voluming' the radiation targets.

3. *Target volume definition*: the International Commission on Radiation Units published recommendation reports on the criteria for delineating tumours for radiation. Gross tumour volume is the primary tumour as judged by clinical examination, examination under anaesthesia, and imaging. It encompasses the primary tumour and any lymphadenopathy. Gross tumour volume is then grown with a margin to give a clinical target volume in order to encompass any microscopic disease around the gross tumour volume. Historically, a margin of 1 cm, excluding bone, air cavities, or other anatomical barriers has been considered the standard margin in HNC; however, there is growing consensus towards using less than a 1 cm margin, as patient immobilization and radiotherapy planning techniques allow greater accuracy and reduce the risk of 'geographical miss'. This reduction in clinical target volume is aimed at reducing the incidence and severity of treatment-related toxicity. Taking into account the possible intra- and interfractional internal organ movement and variation in daily set-up, a further final margin is applied circumferentially to make the planning target volume, a further 3–5 mm depending on the technology and protocol at the treating centre.
4. *Organs at risk (OARs)*: these are healthy tissues/organs near the clinical target volume whose irradiation could cause damage that would make changes to the radiotherapy treatment plan. They can be considered as any tissue that a radiation beam passes through but in practical terms they vary in importance. Within HNC practice, the brain, spinal cord, eyes, lens, and optic nerves are considered among the most important to be blocked to reduce dose, and, if possible, the parotid glands. For most HNC radiotherapy set-ups there will be mandatory dose constraints for these OARs. Rarely, doses to OARs may constrain the dose to the tumour or nodal radiotherapy volume, but this may be necessary, for example, to avoid radiation-induced quadriplegia because of spinal cord dose exceeding tolerance. In even fewer cases, potential doses to critical OARs may contraindicate radiation therapy. All OARs are delineated to avoid unnecessary radiotherapy dose. With the increasing ability to reduce the dose to specific delineated areas, new OARs are emerging and being given greater consideration. Examples include pharyngeal constrictor muscle, mandible, the oral cavity mucosa, the auditory apparatus, and submandibular glands.
5. *Radiotherapy planning*: modern software tools enable improved localization and segmentation of target volumes and regions of interests. With the help of automatic and beam's eye view field designs, improved beam trajectories are used to achieve more accurate three-dimensional dose calculations to the target volumes excluding OARs.
6. *Radiotherapy delivery*: with the evolution of radiotherapy techniques, IMRT is employed to treat HNC in most of the developed world. With this technique, multileaf collimators (which are strips of a high atomic number and, therefore, dense material) that are programmed to move independently in and out of the path of a radiotherapy beam, perpendicular to the beam direction, will shape the beam and vary its intensity. This will produce the beamlets that deliver the desired uniform and/or non-uniform dose distribution in the target volume. IMRT can be delivered using the older step-and-shoot method or the relatively newer techniques using volumated arc therapy or tomotherapy.
7. *Image-guided radiotherapy (IGRT)*: this is an important advancement in precision radiotherapy delivery. Various techniques including cross-sectional daily cone beam CT scans or X-rays are used during treatment in order to check that the actual treatment delivered matches that which was planned originally. Obtaining cone beam CT scans during the course of treatment varies from centre to centre, from once a week to daily imaging. If the radiotherapy machine head could be considered as a television screen with each pixel representing a radiotherapy beamlet, which can change in intensity separately from other beamlets, and the patient made up of individual tiny volumes similar to children's building blocks, then consider the radiotherapy head moving around the patient in either an arc or a spiral, with the beamlets changing energy as the machine head moves. Complex three-dimensional shapes of photon energy deposits within the patient's 'building blocks' can be built up, increasing dose to the tumour target and minimizing dose to OARs; for example, dose sparing the parotid while creating a high-dose region within a very close tumour volume.
8. *Adaptive radiotherapy*: following the principles of the state-of-the-art radiotherapy delivery, if there is significant change during treatment, either due to a patient's weight loss or change in disease characteristics (significant regression or progression of primary disease), it is pertinent to obtain a new planning scan and to re-plan the remaining treatment which helps make sure that the targeted area receives a full dose with a minimal dose to OARs, despite any alteration in anatomy. In some centres, resources exist for daily re-planning for even the smallest change in volume and this is likely to become much more common in the future.
9. *Quality assurance*: all radiotherapy departments must keep robust procedures for quality assurance to ensure the best possible care is delivered to the patient. Routine checks of machine calibrations, dosimetry protocols, planning checks, and patient documentation is carried out on a defined time intervals.

Radical radiotherapy for head and neck cancer

Radical radiotherapy is delivered as primary treatment for a disease process, or adjuvant treatment, usually in the postoperative phase. Adjuvant treatment can be considered as a type of 'insurance policy' treatment when the primary tumour has undergone macroscopic surgical resection, but the patient is still at risk of local recurrence from potential microscopic disease. Adjuvant radiotherapy with or without chemotherapy can be given to reduce this local risk of recurrence.

As an organ-preserving approach, radical radiotherapy provides an alternative to surgical treatment with equivalent locoregional cancer control rates, with the advantage of better long-term function from the preserved organ. When used as adjuvant treatment, postoperative radiotherapy is delivered for locally advanced tumours (T3–T4 disease), close resection margins, the presence of perineural or vascular invasion, or in the presence of multiple nodal spread.

Box 65.1 Most commonly used dose fractionation schedules for radical radiotherapy in head and neck cancer

Primary radiotherapy dose fractionation schedules

- 70 Gy in 35 fractions over 7 weeks.
- 65–66 Gy in 30 fractions over 6 weeks.
- 55 Gy in 20 fractions over 4 weeks (mainly for T1–T2N0 carcinoma of the glottis).

Elective lymph node radiation doses

- 50 Gy in 25 fractions.
- 54 Gy in 30 fractions.

Adjuvant radiotherapy dose fractionation schedules

- 60–66 Gy in 30–33 fractions over 6–6.5 weeks.

Adjuvant radiation is used with concomitant chemotherapy when resection margins are positive or when extranodal extension is seen on the resective pathology.

However, the indication for primary radiotherapy as opposed to primary surgery varies from subsite to subsite. For example, in primary nasopharyngeal carcinoma, primary radiotherapy is the treatment of choice, whereas for oral cavity and paranasal sinus tumours, primary surgery is performed followed by adjuvant radiation as needed. Therefore, multidisciplinary management, centred around patient preferences and values, as discussed in Chapter 46 is the standard of care for HNC.

Elective lymph node irradiation

In a radiologically node-negative neck, if the risk of microscopic nodal disease is more than 15–20%, elective lymph node irradiation is offered. A biological equivalent dose of 50 Gy in 25 fractions is a standard dose to effectively treat the elective nodal basin. With altered dose fractionation, 54 Gy in 30 daily fractions is an equivalent option for a 6-week radiation regimen. This elective lymph node irradiation is commonly delivered, simultaneously within a single IMRT plan in conjunction with the high-dose areas.

The most commonly used dose fractionation schedules for radical radiotherapy in HNC are summarized in Box 65.1.

Figures 65.1–65.3 show a diagnostic image for a patient with a base of tongue tumour and bilateral level II lymphadenopathy, a customized thermoplastic shell for patient immobilization during the course of radiotherapy, and an IMRT plan for the same patient, respectively.

Addition of concomitant chemotherapy to definitive or adjuvant radiotherapy

In the non-surgical management of HNC, concomitant chemotherapy with radiotherapy is a frequently used management strategy. Addition of chemotherapeutic agents yields not only a therapeutic effect by radiosensitization, making the radiotherapy more effective, but also provides the benefit of an additive cytotoxic effect. There have been number of trials which consistently showed superiority of concurrent chemoradiotherapy as compared to radiotherapy alone.

In a large meta-analysis (Meta-analysis of Chemotherapy on Head and Neck Cancer (MACH-NC)) of 93 trials involving more than 17,000 patients, concomitant cisplatin chemotherapy along with radiotherapy showed that there was a 19% reduction in the risk of death and an absolute 6.5% increase in 5-year overall survival as compared to radiotherapy alone ($p < 0.0001$). This survival benefit was predominantly because of a 13.5% improvement in local or regional disease control. It also showed a modest 2.9% reduction in the risk of distant metastases, but this was not statistically significant. Cisplatin 100 mg/m^2 given three-weekly is the

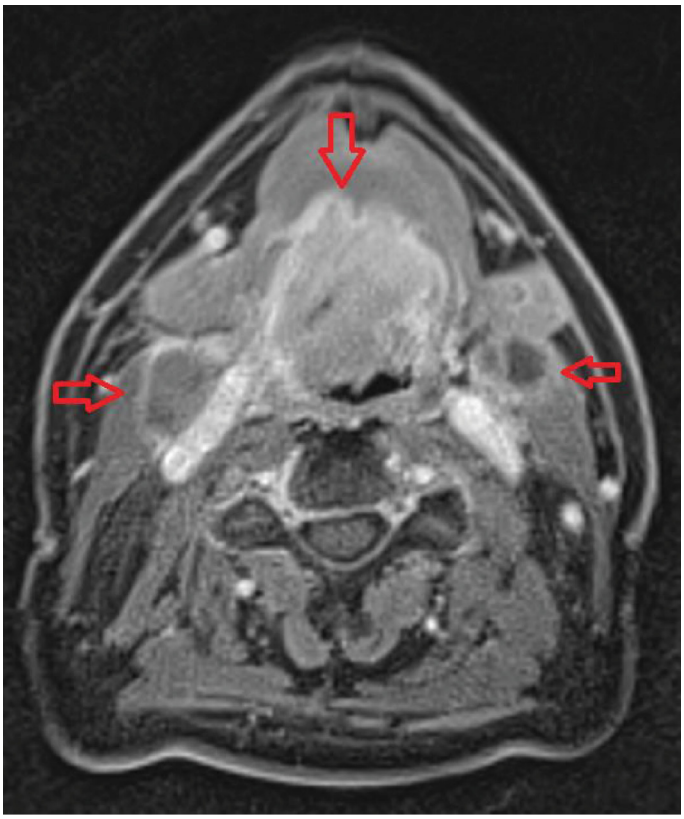

Figure 65.1 An axial image of magnetic resonance imaging scan showing base of tongue tumour with bilateral level II lymphadenopathy—red arrows.

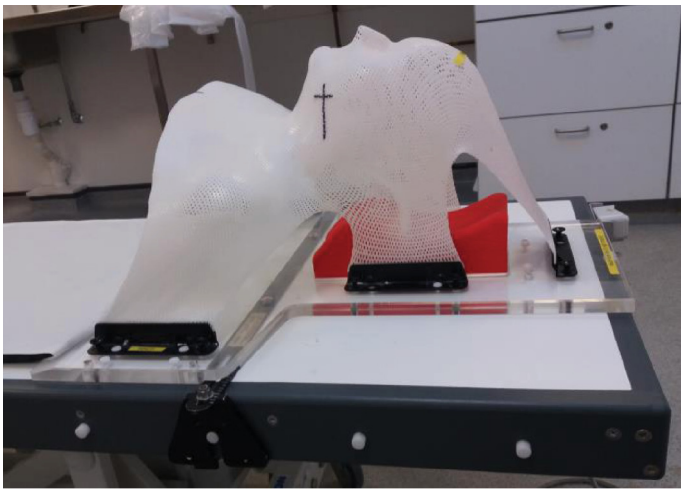

Figure 65.2 A customized thermoplastic shell for patient immobilization during the course of radiotherapy.

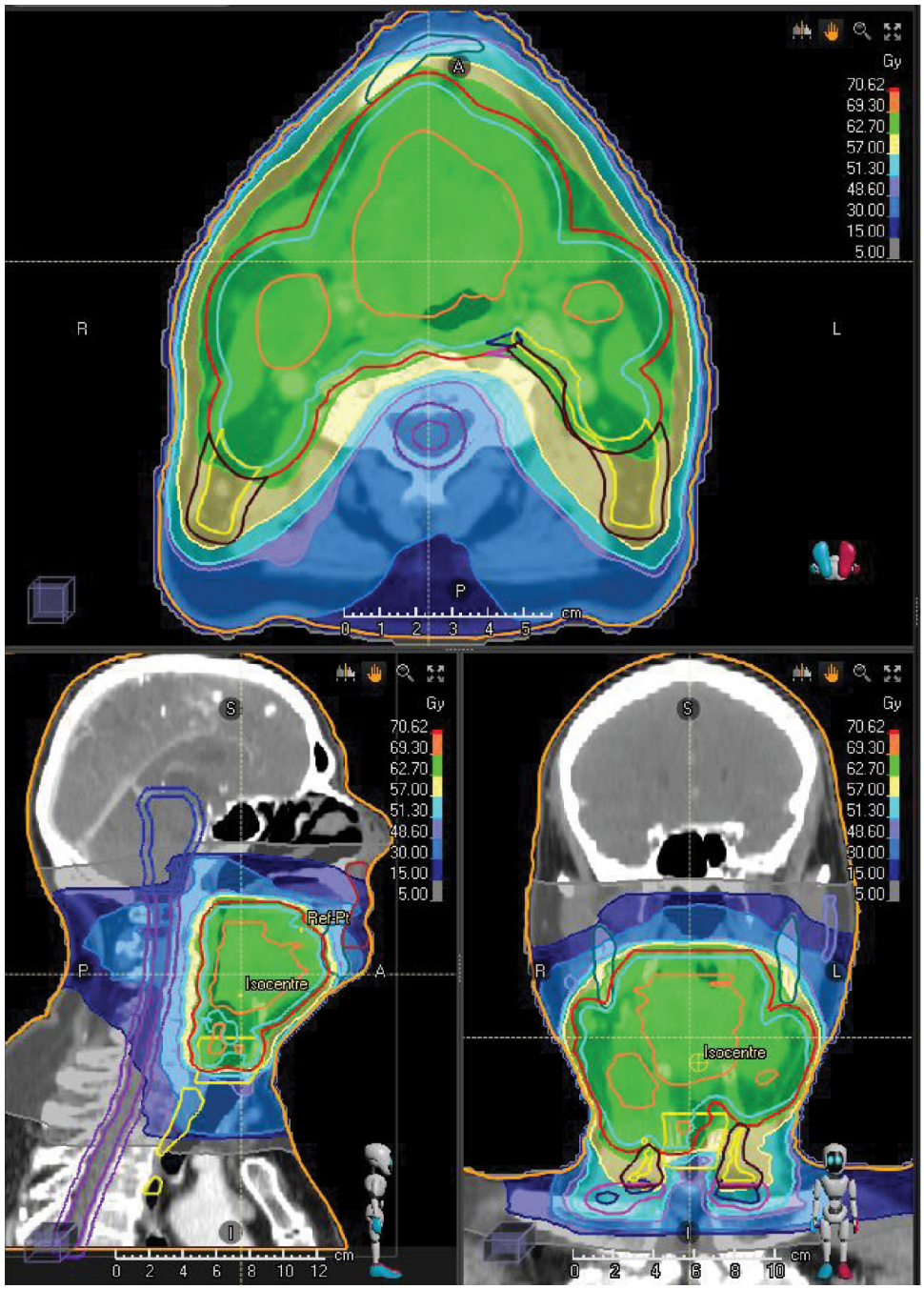

Figure 65.3 A volumetric modulated arc therapy plan for the same patient as in Figure 65.2 (T4N2c base of tongue tumour). Various colour scheme denotes different radiotherapy dose levels.

most commonly concomitant chemotherapy regimen. However, a lower dose of cisplatin, for example, 40 mg/m^2 given weekly for 6–7 weeks, with radiation is also being increasingly used, as it can have logistical advantages and add flexibility. There has been a comparative randomized controlled trial between these two cisplatin regimens, and a comparative meta-analysis and systematic review which showed that three-weekly high-dose cisplatin chemotherapy was more toxic, including more chemotherapy-induced nephrotoxicity, when compared to the weekly cisplatin chemotherapy, with similar efficacy.

In patients who are ineligible for cisplatin chemotherapy (mainly due to suboptimal kidney function, hearing loss or tinnitus, or significant ischaemic or peripheral vascular disease), the use of cetuximab (an immunoglobulin G monoclonal antibody against epidermal growth factor receptor) to radiotherapy has shown benefit. In a multinational trial involving 424 patients with locoregionally advanced cancers of the oropharynx, hypopharynx, or larynx (oral cavity cancers were not included), addition of cetuximab at a dose of 400 mg/m^2 as a loading dose 1 week prior to radiotherapy, followed by 250 mg/m^2 weekly during radiotherapy was evaluated. Three-year

overall survival was 55% with cetuximab and radiotherapy versus 45% with radiotherapy alone, with this difference being significant. The benefit was more marked in patients under the age of 65, good performance status of 0–1, and those with an oropharyngeal primary. More recently, there has been reduced interest in using cetuximab-based bio-radiotherapy in HNC following the publication of two trials where the addition of cetuximab led to inferior outcomes when compared to cisplatin-based chemoradiotherapy; these trials were performed in human papillomavirus (HPV)-related oropharyngeal cancer. The de-ESCALATE HPV trial with 334 patients demonstrated lower overall survival for the cetuximab arm (89% vs 98% at 2 years) and increased disease recurrence (16% vs 6% at 2 years). The RTOG 1016 trial with 987 patients showed lower estimated 5-year overall survival (77.9% vs 84.6%) and progression-free survival (67.3% vs 78.4%) for the cetuximab arm. The cetuximab arm did not show reduction in toxicity either.

For patients ineligible for cisplatin, carboplatin is another option but the level-one evidence for its use is lacking. Carboplatin is associated with more acute myelosuppressive toxicity than cisplatin but causes fewer other side effects in terms of neurotoxicity, nephrotoxicity, or gastrointestinal upset. There is also published evidence emerging for the use of taxanes in the form of docetaxel as a substitute for those patients ineligible for cisplatin, and further work on this may well appear in the near future.

Although age alone should not be used as a criterion in patient selection for addition of concomitant chemotherapy, in the MACH-NC meta-analysis the addition of chemotherapy did not show benefit in those patients over the age of 70 years, although the number of patients over 70 years receiving chemotherapy was small. Overall, appropriate patient selection should occur based on taking patient and disease factors into account, with patients being individually considered for the addition of concomitant systemic therapy.

Role of neoadjuvant/induction chemotherapy

Induction chemotherapy prior to definitive radiotherapy may reduce the disease volume, treat subclinical distant metastatic disease, and has been shown to improve local regional control, organ preservation, and prolong survival in early-phase studies. This approach is also often associated with increased toxicity, which may delay or even prevent definitive local treatment. The toxicity of induction chemotherapy may also limit patients' compliance with definitive radiotherapy. There is also a major concern that if induction chemotherapy does not work, potentially curable disease may become incurable by delaying definitive chemoradiotherapy. Analysis of six trials involving 861 patients comparing concurrent chemoradiotherapy versus induction chemotherapy followed by definitive locoregional therapy showed no significant improvement in overall survival with induction therapy, although there is a greater survival benefit from use of induction chemotherapy, specifically in locally advanced nasopharyngeal cancer.

There has been long debate for the use of neoadjuvant/induction chemotherapy prior to definitive treatment in locally advanced HNC. The evidence is contrary and there is a large variation in clinical practice. A combination induction chemotherapy, either cisplatin and 5-fluorouracil (5FU), or docetaxel and cisplatin with or without 5FU, has been frequently used, but much less so now, for locally advanced unresectable tumours. Appropriate patient selection is absolutely key. Induction chemotherapy may still be considered for a very small number of patients with bulky primary or nodal disease and who are otherwise healthy, with a performance status of 0 or 1, only. This induction chemotherapy should be offered in cancer centres with a relatively high volume of HNC patients with optimal supportive care.

Acute toxicities and supportive care during treatment

Radiotherapy, with or without concomitant chemotherapy, is a toxic treatment and a multidisciplinary team approach is required during treatment for toxicity management and supportive care. The principles of multidisciplinary care are set out in Chapter 46. Standard of care involves patient assessment in a multidisciplinary, on-treatment clinic, on a weekly basis, with ad hoc daily interventions as required.

Treatment assessment and post treatment surveillance

Patients with advanced disease treated with primary (chemo)radiation routinely undergo a positron emission tomography (PET) scan 12 weeks after treatment. The role of PET scanning in these settings was established in the PET-Neck trial, where 564 patients with advanced disease (N2 or N3 nodal disease) were randomized to either planned neck dissection or PET-based surveillance at 12 weeks post treatment; of the 270 patients randomized to PET scan-based surveillance, 69% had complete response, 6% had complete response in the neck but not at the primary site, and 24% did not have complete response in the neck. Medically fit patients with a residual disease in the neck underwent a neck dissection. Of the patients randomized to planned neck surgery, 78% actually underwent went neck dissection. The 2-year overall survival rates were 81.5% and 84.9% in the planned surgery and surveillance groups, which met the predefined objective of non-inferiority of the two treatment strategies. The results were similar in the HPV-positive and HPV-negative patient populations.

For the first 2–4 years post treatment, follow-up surveillance should be robust as approximately 80–90% of all recurrences occur within this time period. The purpose of robust surveillance is to detect locoregional recurrence early, which may still be amenable to curative treatment, detection of second primary cancers, and the management of late side effects. On each visit, patients should have a detailed history of symptoms taken, especially any new symptoms, assessment of treatment-related side effects, clinical and endoscopic examination, and cross-sectional imaging if there is any suspicion of a cancer recurrence.

Long-term side effects include swallowing dysfunction, xerostomia, taste alteration, neck stiffness, loss of hair in the radiated field, risk of hypothyroidism (~50%), ORN (~5–10%), radiation caries, and a low risk of increased carotid artery atherosclerosis. There is also a small risk of developing second cancers in

the radiotherapy field; radiation-induced cancers usually present 15–20 years later.

Radical re-irradiation

Despite optimal management of locoregional disease, locoregional recurrence is sadly a common occurrence; data from RTOG 0129 and 0522 trials suggested that 55% of patients recurred locoregionally only, and 40% showed distant metastasis. In the remaining 5% of patients, recurrence was both locoregional and distant.

Management of locoregional recurrent disease and second primary cancers in the previously irradiated field is a unique challenge. In general, surgery is considered the gold standard salvage option, if feasible. However, a considerable number of patients present with inoperable recurrences. Historically, re-irradiation with a radical dose in the head and neck region has been thought to be a high-risk strategy with a significant cumulative incidence of grade 3 or 4 toxicities. Nevertheless, with evolving IMRT technologies and more precise delivery of radiation, radical re-irradiation in the field of previously irradiated head and neck region is gaining popularity. However, the evidence base is weak, and this should be undertaken only at specialist centres, preferably within the envelope of a trial.

Palliative radiotherapy in head and neck cancer

Palliative radiotherapy is often used for disease control, symptom control, and to delay further symptoms. As far as dose fractionation is concerned, there is a large heterogeneity in practice, and it varies from giving a single fraction to several weeks of radiotherapy. Given the relatively short life expectancy of this patient population, it is important to keep overall treatment time as short as possible with minimal toxicity and minimal reduction in quality of life, because of treatment. In the era of IMRT, a short course of hypofractionated radiotherapy (high dose per fraction over a few days, e.g. 25 Gy in five fractions over a week) has shown promise with less hospital visits and good clinical outcome.

Palliative systemic therapies

Initiating systemic therapy for advanced or metastatic HNC has to be a considered decision. Survival for this cohort of patients is often approximately 6–12 months and many patients have limited prognosis even without their HNC because of other comorbidities. Choice of which systemic therapy to use depends on performance status, comorbidities, physiological reserve, renal function, potential treatment morbidity, which previous systemic therapies the patient has had, and patient choice. In the elderly, as many of these patients are, geriatric assessment is useful before starting any treatment.

There is little up-to-date evidence in the literature on palliative use of chemotherapy, and few comparative trials. Carboplatin or cisplatin is the standard chemotherapy used, usually combined with 5FU, or if the patient is fit enough with a taxane, as in the carboplatin/docetaxel combination. Carboplatin may be the preferred platinum agent in treating metastatic disease. The combination of cetuximab, cisplatin, and 5FU (EXTREME regimen) has been used in better performance status patients with inoperable advanced or metastatic intraoral primary cancer.

Immunotherapy is a robust therapeutic option for metastatic disease with a burgeoning evidence base. While nivolumab and pembrolizumab, both programmed cell death protein 1 (PD1) inhibitors, are in use, other new agents are being developed. Immunotherapies are associated with novel and at times serious and even life-threatening toxicities such as colitis, pneumonitis, hepatitis, nephritis, carditis, and pituitary ablation, so patients require close monitoring.

Interest is increasing in those patients with only a few metastases, oligometastatic disease, most commonly lung metastases, with the use of either surgical metastectomy or stereotactic body radiation therapy to a small number of well-defined metastases. This may delay the use of systemic treatment as well as offering patients another treatment option.

Best supportive care without systemic treatment may be the optimal management strategy for those patients with a poor performance status, significant comorbidities, especially the presence of other advanced cancers, marked weight loss, or hypercalcaemia, all of which indicate a worse prognosis. The aim of best supportive care is to maintain as good a quality of life as possible with control of any symptoms including avoiding iatrogenic chemotherapy morbidity. The decision to offer best supportive care should be taken in conjunction with the patient and family and not taken on grounds of age alone. Older HNC patients with good performance status and few comorbidities may tolerate systemic therapies as well as younger patients but may show increased toxicity.

Future developments

Both surgical and non-surgical treatment technologies are evolving with time. From a non-surgical point of view, radiotherapy with IMRT is now considered the standard of care. The following are potential future avenues for non-surgical treatment in HNC:

- Proton therapy for primary treatment.
- Stereotactic ablative body radiotherapy for re-irradiation.
- Immunotherapy, in combination with IMRT and other radiation modalities.
- Local injectables with small molecules and oncolytic viruses.

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66

Rehabilitation of the head and neck cancer patient

Sarah Stephen and Joanne Patterson

Introduction

The rising prevalence of head and neck cancer (HNC) and improved survival has led to an increasing focus on the numerous and complex needs of HNC follow-up. Broadly, this encompasses the functional, psychosocial, and quality-of-life sequelae of HNC and its treatment, surveillance for recurrent or new primary disease, and the promotion of a healthy lifestyle. This chapter covers the rehabilitation of function in HNC, focusing on voice, speech, and swallowing. These are critical functions and subsequent impairments are strongly related to poor quality-of-life outcomes.[1,2]

Rehabilitation of function needs to begin *early* in the care pathway, involving comprehensive assessment and tailored interventions. Although speech and language therapists (SLTs) hold a key role, multidisciplinary team (MDT) working is essential to attain optimal patient outcomes.[3,4] This chapter follows the clinical pathway, with a section dedicated to laryngectomy issues.

Assessment

Clinical assessment

A clinical assessment typically begins by gathering background information on the presenting disease, treatment/planned treatment, and patient characteristics. For swallowing, particular attention is given to risk factors that have the potential to increase medical complications, for example, older adults have less efficient swallowing mechanism, chronic lung disease reduces tolerance to aspiration, and pre-existing neurological conditions that can delay recovery. Patient report of the problem provides insights into self-awareness, priority, perception of severity, changes over time, and impact on daily living. This information is supplemented by validated patient-reported outcomes, summarized in Table 66.1.

A clinical examination then follows, comprising observations of oral sensory-motor function (rate, range, strength, and sensation), oral secretions and hygiene, voice, and speech quality. This is accompanied by volitional tasks of function such as pitch glides, sustained maximum phonation time, and voluntary cough. A swallowing assessment also includes an observation of eating/drinking, evaluating safety and efficiency. Where significant dysphagia is suspected, the assessment may begin with small, measured amounts of water, progressing to larger aliquots and observing for an airway response (coughing/throat clearing, 'wet' voice quality). Information is then integrated to generate

Table 66.1 Examples of functional outcome measures used in head and neck cancer

Target function	Description
Swallow	
Patient reported	MD Anderson Dysphagia Inventory[5] Swallowing-Specific Quality of Life Questionnaire (SWAL-QOL)[6] Sydney Swallow Questionnaire[7] Eating Assessment Tool (EAT-10): assessment of symptom severity[8]
Clinician rated	Performance status scales[9]: subscales; Normalcy of Diet and Eating in Public Functional Oral Intake Scale[10] Functional Outcome Swallowing Scale (FOSS)[11] 100 mL Timed Water Swallow Test[12] Penetration Aspiration Scale (PAS)[13]
Voice	
Patient reported	Voice Handicap Index (VHI)-10[14] Voice-related quality of life (V-RQOL)[15] Voice Symptom Scale (VoiSS)[16] Vocal Performance Questionnaire (VPQ)[17]
Clinician rated	GRBAS[18] (Grade, Roughness, Breathiness, Asthenia, Strain scale) Consensus Auditory-Perceptual Evaluation of Voice (CAPE V)[19] Dynamic Imaging Grade of Swallow Toxicity (DIGEST-VF)[20]
Speech	
Patient reported	Speech Handicap Index[21]
Clinician rated	Performance status scales[9]: subscale; Understandability of Speech

a management plan. Clinician-rated scales (Table 66.1) are also recorded for benchmarking and outcome measurement.

Instrumental assessment

Swallow

Instrumental swallow assessments provide detailed information on dysphagia aetiology, safety (aspiration risk), efficiency, anatomical and pathophysiology, and severity. The two main assessments are videofluoroscopy and flexible endoscopic evaluation of swallowing (FEES).

Videofluoroscopy

Videofluoroscopy is a dynamic swallow assessment under X-ray, tracking radio-opaque boluses from the oral cavity to the upper oesophagus, enabling a view of all phases of oropharyngeal swallowing. The assessment is conducted by an SLT and radiologist or radiographer, under relatively controlled conditions. Test boluses are usually given in measured amounts, increasing in volume and texture, depending on the focus of the assessment.

Flexible endoscopic evaluation of swallowing

FEES provides direct visualization of the pharynx and larynx and their non-deglutitive functioning (e.g. base of tongue retraction). The assessment includes observations of secretion management (especially pertinent for tracheostomy weaning) and food and drink trials. Test items may be dyed to increase visibility. Although not all components of oropharyngeal swallowing are observed (e.g. the oral stage), the benefits of FEES include portability of equipment, repeatability, and avoidance of radiation exposure.

Voice and speech

There are no agreed gold standard instrumental voice and speech assessments for HNC. Examples of acoustic and aerodynamic voice measures that may be employed are summarized in Table 66.2.[5–21]

Protocols for acoustic speech measures have been published for isolated sounds but there is limited work on spontaneous speech.[22] The Iowa Oral Performance Instrument (an air-filled tongue bulb connected to a peak pressure measurement device) assesses lip, tongue strength, and endurance and may be used for speech and swallowing assessment.[23]

Interventions

Rehabilitation for speech, voice, and swallowing begins before treatment and continues after treatment, until maximal potential has been attained.

Table 66.2 Voice instrumentation measures

Acoustic and aerodynamic measures	Fundamental frequency—vocal fold vibration frequency
	Perturbation—jitter: irregularities in vocal fold frequency; shimmer: amplitude irregularities
	Harmonic-to-noise ratio—ratio of sound frequencies to noise energy in the voice
	Maximum phonation time—glottic efficiency

Pretreatment

HNC patients whose primary treatment will disrupt the ability to speak, eat, or swallow should be seen by a specialist SLT. This consultation serves many purposes including (1) a thorough assessment with tailored advice/intervention; (2) information provision about impending functional impacts; (3) facilitation of decision-making; and (4) completion of baseline measures to plot progress, predict outcome,[24,25] and offer pre-habilitation advice where indicated.[26]

Information provision

Providing information on functional outcomes is central to preparing the patient and their family, setting expectations for recovery and survivorship as well as giving an outline of the rehabilitation plan. Patients prefer to receive individually tailored information, delivered by a knowledgeable person, including the impact on short- and long-term functioning with an estimate of recovery time.[27–29] Patient experience videos and peer-to-peer support (particularly well established for the laryngectomy community) supplement this service.

Decision-making

Patient involvement in decision-making is crucial, with high importance being attributed to functional outcomes when weighing up HNC treatment options.[30] Eliciting patient priorities and values forms part of this process, supported by giving realistic information about alterations to function and the impact on daily living. In addition to this, there are subsequent decisions (e.g. placement of a prophylactic gastrostomy before radiotherapy) where there may be a choice of routes and the impact on function is unclear. SLTs and the MDT need to be equipped with functional prognosis for options, presented in an easily understood format.

Pre-habilitation

Performing swallowing exercises throughout radiotherapy may have a positive influence on maintaining normal long-term swallowing function,[31–34] although the impact on physiology is debatable.[35,36] Adherence to exercises is variable, given the significant symptom burden during radiotherapy such as pain and fatigue.[37] Preoperative swallowing techniques may also be beneficial for certain procedures, such as laryngeal conservation surgery, to enhance skill development in performing swallowing manoeuvres accurately.[38]

On-treatment

The impact of HNC treatment on speech, voice, and swallow function will depend on the size and location of the disease, the extent and type of treatment, and the nature of any reconstruction. The functional impact following multimodal treatment will be far greater than after single modality treatment,[39] with subsequent implications for the amount and intensity of rehabilitation. The rest of this section will give an overview of acute rehabilitation requirements during HNC treatment.

Surgery

Integrated working is essential in the early postoperative period with clear communication and multidisciplinary ward round representation, to facilitate decision-making processes and optimize patient management. Commencement of oral intake and rehabilitation programmes will be patient specific and dependent on oedema, pain,

secretion management, and adequate mucosal healing. Resections requiring reconstruction or in compromised patients (previous HNC treatment, malnutrition, diabetes) require a longer healing period prior to swallow assessment and initiation of therapy programmes. Site and size of tumour resection will influence assessment aims, patient progress with swallow rehabilitation, and feeding tube requirements. Disorders of the swallow mechanism are vast and variable, see Table 66.3 for a summary of typical swallow impairments related to site of resection.

For patients who receive an elective tracheostomy, early intervention targets holistic communication needs with careful selection of tubes, fenestrations, and cuff management. Assessment of voice production and communication strategies can facilitate speech and have a positive impact on patient anxiety levels.[40] A team approach to early identification of suitable timing for cuff deflation and introduction of speaking valves facilitates vocalization and promotes laryngeal function by desensitizing the laryngopharyngeal mucosa and reflexes, supporting the decannulation process.[41,42] Assessment of secretion management and airway safety when swallowing is needed and may be augmented by FEES to determine risk of silent aspiration.[43]

Voice

Surgery for hypopharyngeal, supraglottic, and laryngeal cancers can have a deleterious effect on critical structures and movements necessary for voice production. Damage to the superior and recurrent laryngeal nerves can result in hoarseness and reduced vocal range.

Speech

Speech impairments following surgical resection of oral and oropharyngeal cancers are common. Speech is characterized by articulation errors, with reduced intelligibility, particularly in the early postoperative period where oedema is severe. Unilateral resections, avoiding anterior structures, have better functional speech outcomes.[44] Microvascular free flap reconstruction for advanced oral cancers yields superior speech function than primary closure.[45] Hypernasal resonance is common following resections of the soft palate and a palatal obturator should be considered. Speech strategies, communication devices, and exercises optimize communication and patient experience.

Radiotherapy

Acute radiotherapy side effects, such as mucositis, pain, dysgeusia, fatigue, oedema, and nausea, have significant impact on the ability to eat and drink. These side effects are dose dependent, increase when radiotherapy is combined with chemotherapy, and persist for several weeks or months after completion of treatment.[46] Regular assessment by the SLT team is recommended, ideally within an MDT environment.[47] Nutritional support is required during treatment, with rates of tube feeding between 60% and 100%.[48] Periods of 'nil by mouth' should be avoided to prevent deconditioning and atrophy of swallowing musculature.[31,33,49] Neuropathic pain control may aid the patient's ability to maintain oral intake during treatment.[50]

Table 66.3 Swallow impairment characteristics dependent on site and location of resection

Site of resection	Location	Dysphagia Impairment												Functional impact
		Lip closure/sensation	Tongue control/ sensation	Bolus preparation/transit	Oral residue	Initiation of swallow	Soft palate elevation	Pharyngeal / BOT contraction	Pharyngeal residue	Hyoid excursion	Laryngeal elevation	Laryngeal closure	Cricopharyngeal opening	
Oral	Lip													• Anterior spillage of saliva, food and drink • Modified textures • Extended mastication • Oral and pharyngeal residue
	Tongue													
	Floor of mouth													
	Mandible													
Oropharyngeal	Soft palate													• Bolus uncontrolled • Pre-swallow aspiration • Post-swallow aspiration from residue overspill • Modified textures
	BOT													
	Tonsil/PPW													
Pharyno-laryngeal	Hypopharyngeal													• Reduced airway protection • High risk of aspiration of secretions, food and drink • Intensive and complex rehabilitation needs • Modified textures
	Supraglottic													
	Laryngeal													

Rehabilitation

This section focuses on SLT interventions for the remediation of function.

Swallowing interventions

Swallowing interventions largely fall into compensatory strategies, direct therapy, and indirect therapy techniques. They are tailored according to pathophysiology and usually used in combination to improve the safety and efficiency of swallowing.

Compensatory strategies include (1) changes to head posture, (2) texture/food preparation modification, and (3) alteration of swallow/bolus volume. Direct therapy aims to improve swallow physiology, such as the introduction of swallow manoeuvres. For example, an extended airway protection technique can be effective at reducing/eliminating aspiration.[51] These manoeuvres can be complex to learn. Providing visual feedback via endoscopy has been shown to expediate skill development.[52] Indirect therapy techniques address discrete components of the swallowing mechanism—examples of which are provided in Table 66.4.

Voice and speech therapy

Early introduction of voice therapy improves patient-reported outcomes and discrete vocal performance parameters post radiotherapy for laryngeal cancer.[63,64] Preliminary evidence suggests that patients with non-laryngeal HNC may also benefit from voice therapy. Voice rehabilitation may include breathing techniques, voice exercises, postural advice, and indirect voice care.[65,66]

Trismus can adversely affect speech intelligibility. Jaw stretching and strengthening exercises may prevent trismus developing in the short and long term.[67] Strategies such as slowing speech rate, compensatory strategies, and environmental considerations are helpful for increasing intelligibility.

Table 66.4 Examples of indirect therapy techniques to facilitate post-treatment swallowing function

Intervention	
Exercises	Tongue resistance exercises may improve strength but limited evidence of translation to swallowing function or patient-reported outcomes[22,53] Exercises to improve hyolaryngeal elevation are helpful for postradiation dysphagia[54]
'Pharyngocise' programme	Exercise-based intensive programme with preliminary evidence to support improvement in functional swallowing outcomes[55]
Swallow–respiratory cycle training	Preliminary evidence showing trend for improvement in swallow function[56]
Devices	
Expiratory muscle strength training	Patients forcefully expire through device containing a one-way spring-loaded valve. Small-scale studies suggest an improvement in cough function[57,58]
Neuro-muscular electrical stimulation	May prevent decline of swallow function post radiotherapy, but limited evidence to support gains[59] and may lead to further deterioration[60]
Jaw stretch and strengthening devices	Devices targeting trismus can help improve mobility for eating[61,62]

Psychosocial

Psychosocial adjustment is intrinsically linked to HNC rehabilitation. Studies have shown that SLTs and dietitians can integrate talking therapies such as cognitive behavioural therapy and motivational interviewing alongside their clinical practice to improve psychosocial adjustment and behaviour change.[68,69] Peer-to-peer support also facilitates adjustment to loss of function, provides an environment to practise new skills, and reduces isolation.[70]

Late effects

There is an emerging body of evidence identifying the progressive deteriorative nature of late-radiation-induced dysphagia.[71–75] Pathophysiology may include cranial neuropathy, severe fibrosis, and atrophy of vital speech, voice, and swallowing musculature.[76] Several factors have been associated with the incidence of late radiation complications: treatment for nasopharyngeal and oropharyngeal cancer, increased total radiation dose and mean pharyngeal constrictor dose, extended periods of 'nil by mouth', acute inflammation resulting in lymphoedema, the addition of surgery and scarring, and patient's age.[33,77–81]

Presentation

Time to onset of late-radiation-induced dysphagia is variable, with some patients experiencing progressive deterioration, whereas others may present with a sudden severe onset of dysphagia.[75,82] Late effects can be under-reported, with patients developing malnutrition, dehydration, and recurrent chest infections or pneumonias, but not recognizing changes due to poor sensation.[82,83] Attribution to previous HNC treatment may be overlooked, particularly if the onset of symptoms is after the 5-year cancer surveillance period. Improved awareness and referral pathways to ear, nose, and throat (ENT) services are required for specialist MDT assessment and management.

Assessment

Regular evaluation of function should be incorporated into HNC surveillance, including onset of problems, symptoms, and psychosocial burden[85] (see 'Clinical assessment'). Late-effect dysphagia is often characterized by impaired hyolaryngeal excursion, pharyngeal constriction, and tongue base retraction causing pharyngeal residue and high rates of aspiration.[71,77,86] Instrumental assessment is necessary to provide pathophysiology assessment of anatomical changes, potential strictures, internal lymphoedema, muscle atrophy, and cranial nerve palsies.[82]

Management

Optimum management is not based solely on clinical judgement but includes wider determinants of health and eliciting patient values to inform shared decision-making. For example, patients may decide to continue with oral intake for quality of life, despite aspiration risk. Chronic management, support, and education are needed to guide patients through adjustments of their long-term changes to function.

Swallow interventions described in the 'Rehabilitation' section are also suitable for late-effect dysphagia.[86] Collaboration with ENT is

essential to determine appropriateness of surgical interventions (e.g. oesophageal dilatations). Elective salvage laryngectomy surgery is a treatment option for dysfunctional larynx. Surgery will prevent aspiration, but careful patient selection and preparation is required as laryngectomy surgery will not resolve impaired oral and pharyngeal contraction and bolus propulsion needed for effective swallowing. Surgical complications need to be considered as they can be significant in this population.[87,88]

Laryngectomy

Total laryngectomy is a treatment option for advanced primary laryngeal and hypopharyngeal cancer. Salvage laryngectomy may be offered for residual or recurrent disease and functional laryngectomy may be considered for a dysfunctional larynx resulting in chronic and severe aspiration, airway compromise, and/or radionecrosis.

Anatomical changes

Surgery includes complete removal of the laryngeal cartilage, epiglottis, hyoid bone, sections of the base of tongue, and strap muscles of the neck (see Chapter 55). Surgery requires the creation of a permanent tracheostoma to maintain respiratory airflow. For extensive tumours, insufficient pharyngeal mucosa prohibits primary closure. Reconstruction with free or pedicle flap repairs, jejunal free graft, or gastric pull-up reconstruction will have further impacts on voice and swallow function.[88] Anatomical problems following surgery can include narrowing of the reconstructed pharynx, fistula formation, stricture or stenosis, or formation of a diverticulum or pseudo-diverticulum.[89] Multiprofessional management and treatment may include surgical interventions including myotomy, Botox injection, and dilatation. Salvage surgery following chemoradiotherapy has a higher rate of postsurgical complications.[87,88]

Pretreatment assessment

Pretreatment assessment is essential to determine suitability for surgery and patient preparation. Patient capacity and capability to maintain a permanent breathing stoma needs to be verified. Characteristics of alaryngeal communication options need to be carefully considered and discussed with the patient and their family to elicit communication values. Preparation for postoperative changes to communication, breathing, swallowing, taste, and smell as well as the potential impact on a patient's psychological well-being need to be discussed in detail (see 'Pretreatment').

Postsurgical management

Rehabilitation starts early postoperatively with support and advice to facilitate communication, discuss changes to breathing, and emphasize the importance of tracheostoma care and pulmonary rehabilitation. Placement of a nasogastric feeding tube provides nutritional input in the early postoperative period, to allow for tissue healing. Early oral feeding after primary total laryngectomy may reduce hospital stay.[90] Time to oral intake usually follows institutional protocol, but evidence suggests no increased risk of pharyngocutaneous fistula in patients who receive early oral feeding (<5 days) when compared to later oral feeding (>7 days).[91] Feeding may be delayed to 7–14 days in extensive resections or in patients with compromised healing following previous HNC treatment, increased frailty, or poor nutrition.[88] Institutional protocols may specify blue dye swallow assessment of subcutaneous fistula or radiological examination of internal and external fistula prior to commencement of oral intake in complex patients, or if there are early signs of fistula formation.

Communication

There are three post-laryngectomy communication techniques: oesophageal speech, surgical voice restoration (SVR), and alaryngeal voice. For oesophageal speech and SVR techniques, the source of voice is a reconstructed segment created during surgery, known as the pharyngo-oesophageal segment. See Table 66.5 for a summary of communication techniques post total laryngectomy surgery.

Table 66.5 Comparison of surgical voice restoration, oesophageal speech, and artificial larynx in terms of production, requirements, and characteristics

	Surgical voice restoration	Oesophageal speech	Artificial larynx
Vibratory source for voice	Pharyngo-oeosophageal segment	Pharyngo-oeosophageal segment	Vibrating membrane on device
Energy source of voice	Pulmonary air	Oesophageal air reservoir	Battery
Requires equipment	Yes	No	Yes
Requires visual acuity	Yes; prosthesis care	No	No
Requires hand/arm dexterity	Yes; finger occlusion of stoma for voice production and prosthesis care. If patient unable to perform, will require carer assistance	No	Yes; holding the device in the correct position and manipulating the controls
Regular hospital visits	Yes; prosthesis changes, depending on valve and patient training	No	No
Requires maintenance	Yes; daily cleaning, regular prothesis changes	No	Yes; care for the device and batteries/charging
Voice quality	Hoarse, rough, wet, breathy; generally preferred over oesophageal speech and artificial larynx	Hoarse, rough, wet, breathy; less preferred than SVR	Mechanical, unnatural; generally less preferred than oesophageal voice and surgical voice restoration
Overall 'success'	Generally a high success rate, comparable to artificial larynx	Generally lowest of the three options due to long therapeutic process	Generally a high success rate

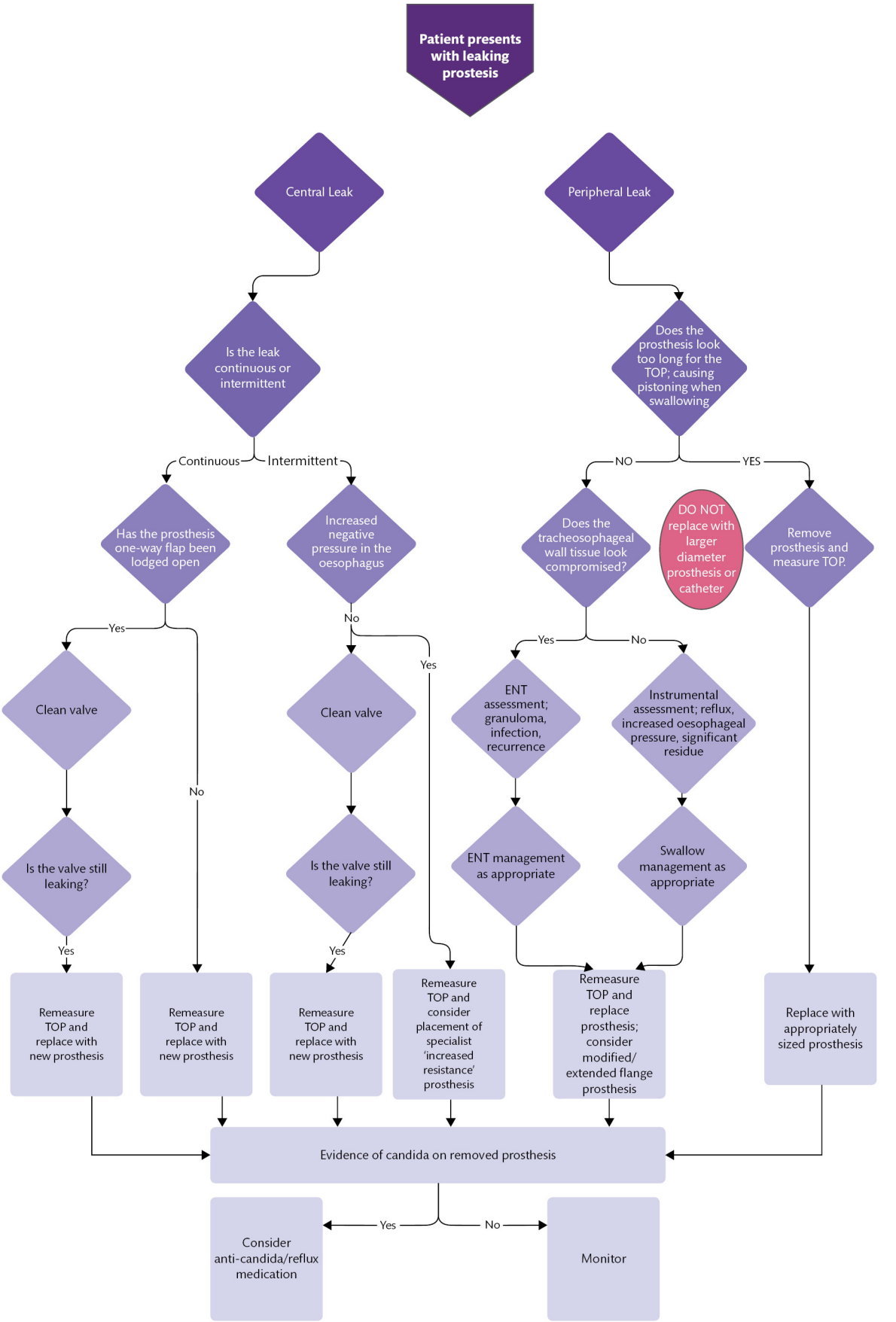

Figure 66.1 Flowchart demonstrating basic troubleshooting for surgical voice restoration prosthesis leaks; TOP = tracheo-oesophageal puncture.

Surgical voice restoration

SVR is considered the 'gold standard' for achievement of voice post laryngectomy.[92,93] Despite this, careful consideration for patient selection is required prior to surgery and in an MDT environment. SVR involves placing a silicone voice prosthesis into a surgically created tract within the tracheo-oesophageal party wall, known as a tracheo-oesophageal puncture (TOP). This allows for redirection of pulmonary airstream into the neopharynx when the tracheostoma is covered. Surgical technique is a factor for successful development of SVR voice. The TOP can be placed either during primary surgery for select patients or as a secondary procedure following wound healing. In the latter situation, a detailed assessment including an oesophageal insufflation test prior to TOP formation is often required.

Institutional protocols will determine if a voice prosthesis is placed in the TOP intraoperatively or once the tract has stabilized. For the latter, the TOP is kept patent using a catheter or feeding tube, with frequent rotation of the stenting device to ensure stable healing. Superior clinical and patient benefits are associated with intraoperative voice prosthesis placement during primary TOP.[94]

A range of voice prostheses are now available, with Blom Singer and Provox being the commonly used devices within the UK. There are two types of prostheses: indwelling (removed and refit by professionals) and ex-dwelling (removed and by the patient, if competent), which come in a range of sizes and diameters to fit each individual TOP. The valve prosthesis lifetime can vary from a number of weeks to months. When a prosthesis leaks or is dislodged, it needs to be changed as soon as possible to prevent aspiration of saliva, food, and drink and prevent the TOP closing. Despite SVR being the gold standard, TOP complications can be problematic, require MDT management, and have a significant impact on a patient's quality of life. See **Figure 66.1** for TOP and valve prosthesis troubleshooting.

Oesophageal speech

The oesophagus acts as an air reservoir to drive airflow for voice production. Insufflation of the oesophagus can be achieved by two techniques: the inhalation method and the injection method. As the air returns from the oesophagus, approximation of the pharyngo-oesophageal segment provides a vibratory source.

Artificial larynx

An external vibratory source is used by placing an electrolarynx on the neck or by using an intraoral tube to divert the vibratory source into the oral cavity.

Swallow

Separation of the trachea from the oesophagus prevents aspiration; however, significant changes to swallow efficiency are common.[95] Altered anatomy and pressure changes due to reconstruction of the pharyngo-oesophageal segment result in altered bolus transit and propulsion through the neopharynx.[89] Dysphagia prevalence rates can be up to 90%.[96,97] Patient symptoms include significant residue, regurgitation, extended mealtimes, globus sensation, modification of oral intake, and reduced quality of life.[95,98] There are multiple contributing factors including extent of surgical resection, use of jejunum flap, and postsurgical adjuvant treatment.[87,88,99]

Conclusion

Changes to speech, voice, and swallow function can be a debilitating consequence of HNC and its treatment and can result in compromised nutrition, aspiration, and impaired quality of life. The nature and severity of functional changes are complex and have the potential to change over time depending on the treatment received. Surgical interventions can result in acute oedema, pain, and desensitization of functional musculature. Over time this can develop into scaring and atrophy. Radiotherapy treatment has short-term side effects which can progress into long-term effects, such as dysphagia, taste changes, and xerostomia. There is an emerging body of evidence identifying late-stage changes to functional outcomes following radiotherapy, with patients experiencing significant changes to function and quality of life. Laryngectomy results in significant changes to communication, breathing, and swallowing. Patient preparation and education is essential, alongside support and management of communication and pulmonary and swallow rehabilitation.

Input from SLT is required throughout the patient's diagnosis and treatment of HNC, as supported by national guidance. Pretreatment input is essential to establish patient function and to prepare patients for the potential acute and long-term functional changes following treatment.

Speech, voice, and swallow interventions are varied and complex, depending on tumour, treatment, and patient factors. Multidisciplinary working is essential to ensure optimum and efficient patient management.

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67

Benign neck lumps

Samantha Hassid, Georges Lawson, and Sébastien Van Der Vorst

Introduction

Benign neck lumps can be challenging diagnostic entities, and surgical management is nuanced and complex, needing multiprofessional input in some cases. The approach and assessment of neck masses is covered in Chapter 50. This chapter describes some of the most common benign neck lumps and their management.

Branchial cleft cyst

Branchial arch anomalies represent 20% of cervical masses, almost all are diagnosed during childhood. They are the second most common head and neck congenital lesions after thyroglossal duct remnants.

Anatomy and embryological considerations

Around the fourth week of gestation, neural crest cells migrate into the future head and neck region and develop *six pairs of pharyngeal arches* (Figure 67.1). The fifth arch does not contribute to anatomical structures in humans. These arches are made by mesenchymal tissues and are covered externally by an ectoderm recess, referred to as a *branchial cleft*, and internally by a layer of endoderm, referred to as a *branchial pouch* (Figure 67.2).

Each arch contains its own central cartilaginous/skeletal supporting element, an aortic arch artery, and an arch-associated cranial nerve (Table 67.1).

At around 7 weeks of gestation, the second arch enlarges caudally and thickens, combining with the third and fourth arches to form a new cavity called the *cervical sinus* of His. This sinus rapidly involutes during embryogenesis (Figure 67.3).

Branchial cleft anomalies can occur due to incomplete involution of the branchial cleft structure:

- *A fistula* is a communication between a persistent pouch and cleft.
- *A sinus* is a blind-ending tract, which may connect with either the skin (branchial cleft sinus) or with the pharynx (branchial pouch sinus).
- *A cyst* is a trapped branchial arch remnant and occurs when there is no communication with the inner mucosa or outer skin.

Assessment

Fistulas are typically present in infants/young children during the first decade of life. They are usually diagnosed by the opening to the skin or the drainage of secretions or purulent material.

Cystic lesions are more common than fistulas, but they occur later, often in the second decade of life or in the young adult. They are often asymptomatic, but can be diagnosed as a painless, compressible lateral neck mass. They can become tender, enlarged, or inflamed in cases of infection or abscess formation, triggered by episodes of upper respiratory tract infection (in 25% of cases). A purulent drainage of the sinus can occur to the skin or the pharynx. Occasionally, dysphagia, dyspnoea, and stridor in cases of upper airway compression can be a presenting symptom. They can be bilateral in 2–3% of cases (often with familial history).

First branchial arc anomalies: these represent 5–10% of branchial arch anomalies. They are more common in females and are often misdiagnosed. They extend from a cutaneous opening in the submandibular triangle, superolateral to the hyoid bone, ascending to the region of the parotid salivary gland to terminate at the external auditory canal. So, an otologic examination is crucial in these patients. The tract may pass above or below the facial nerve. If infected, there may be associated adenitis in the submandibular area. They are subclassified into two types by Work. Type I are purely ectodermal and type II are ectodermal and mesodermal in origin and can contain skin adnexa or cartilage.

Second branchial arch anomalies (Figure 67.4): these are the most common branchial arch anomalies, comprising 40–95% of cases. They are typically located in the skin at the lower anterior border of the sternocleidomastoid (SCM), course between the internal and the external carotids, and enter the pharynx at the level of the tonsillar fossa. They can be associated with branchio-oto-renal syndrome. They are classified into four types, depending on their extension.

- Type I: most superficial and lies along the anterior surface of the SCM deep to the platysma without contact with the carotids.
- Type II: most common, lies along the anterior surface of the SCM, lateral to the carotid space and posterior to the submandibular gland.
- Type III: extends medially between the bifurcation of the internal and external carotids, lateral to the pharyngeal wall.
- Type IV: lies deep to the carotid space and opens into the pharynx.

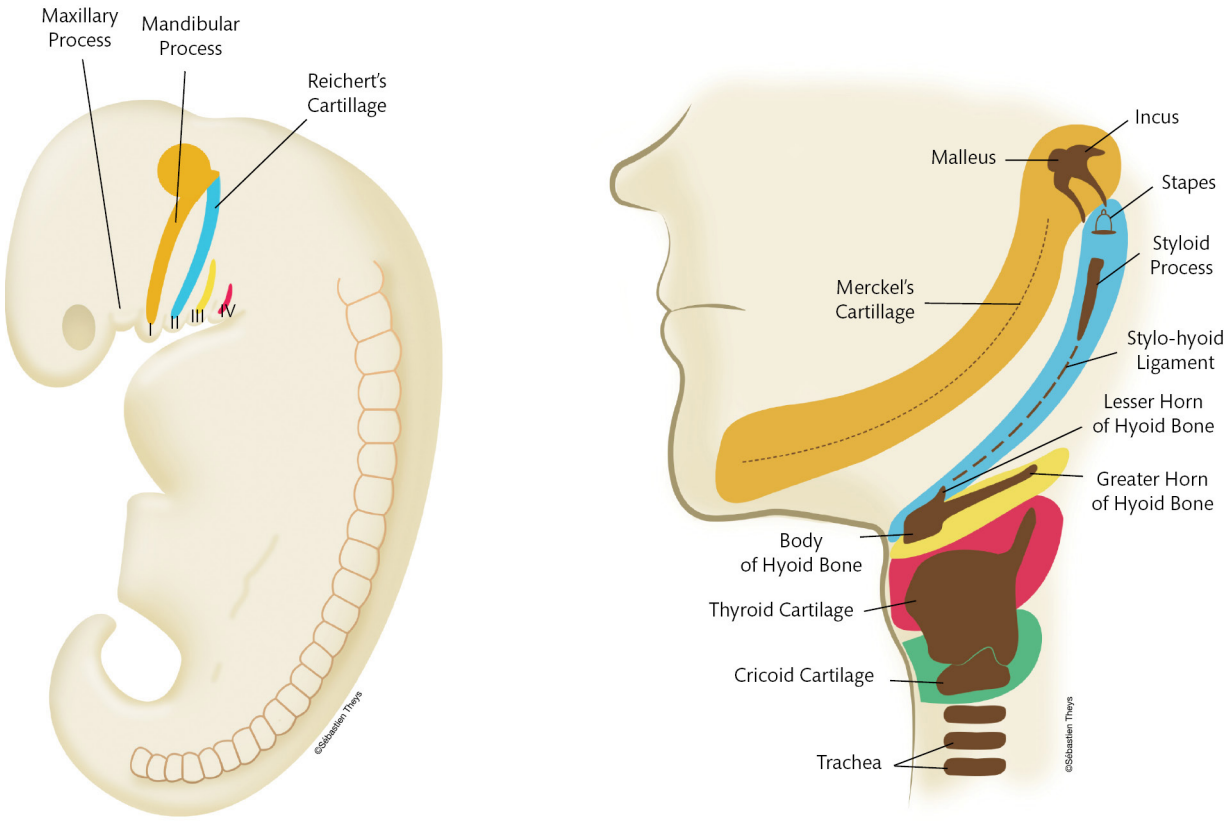

Figure 67.1 Pairs of pharyngeal arches.
Courtesy of Sebastien Theys.

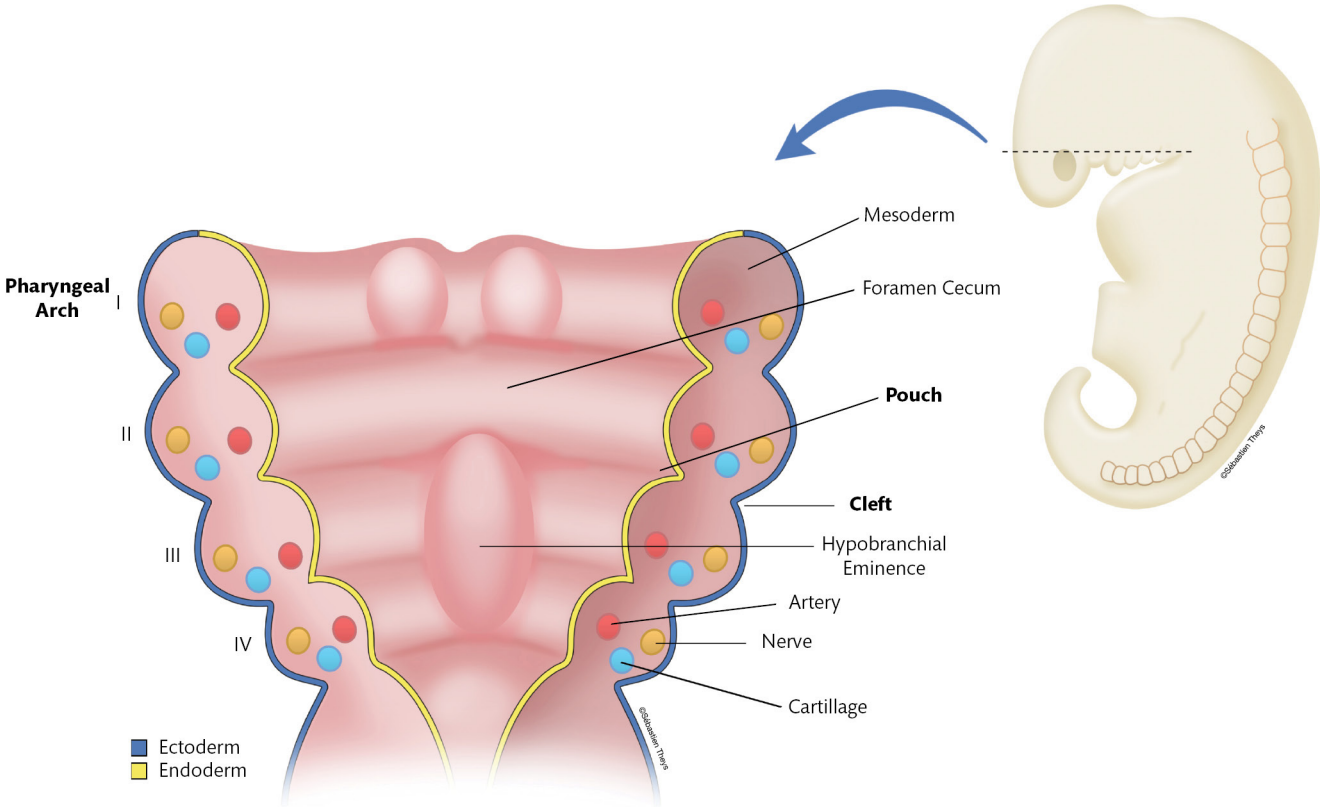

Figure 67.2 Pharyngeal arch composition.
Courtesy of Sebastien Theys.

Table 67.1 Each branchial arch contains its own central cartilaginous/skeletal supporting element, an aortic arch artery, and an arch-associated cranial nerve

Arch	Skeleton	Muscle	Nerve	Artery
First	*Quadrate cartilage:* • Incus • Malleus *Merckel's cartilage:* • Sphenomandibular ligament • Anterior ligament of malleus • Spine of sphenoid • Genial tubercle of mandible	• Muscle of mastication • Anterior belly of digastric • Mylohyoid • Tensor tympani • Tensor veli palatini	Trigeminal (V)	*First aortic arch:* Maxillary artery
Second	*Reichert's cartilage:* • Stapes • Styloid process of temporal bone • Stylohyoid ligament • Lesser horn of hyoid bone	• Facial muscles • Posterior belly of digastric • Buccinator • Stapedius	Facial (VII)	*Second aortic arch:* External carotid artery
Third	Body and greater horn of hyoid bone	Stylopharyngeus muscle	Glossopharyngeal (IX)	*Third aortic arch:* • Common carotid artery • First part of internal carotid artery
Fourth	Thyroid cartilage Corniculate cartilage Cuneiform cartilage	Pharyngeal and extrinsic laryngeal muscle	Vagal (X) Superior laryngeal nerve	Subclavian artery (right) Arch of aorta
Sixth	Arytenoid cartilage	Intrinsic laryngeal muscle	Vagal (X) Recurrent laryngeal nerve	Pulmonary trunk dorsal aorta ductus arteriosus (left)

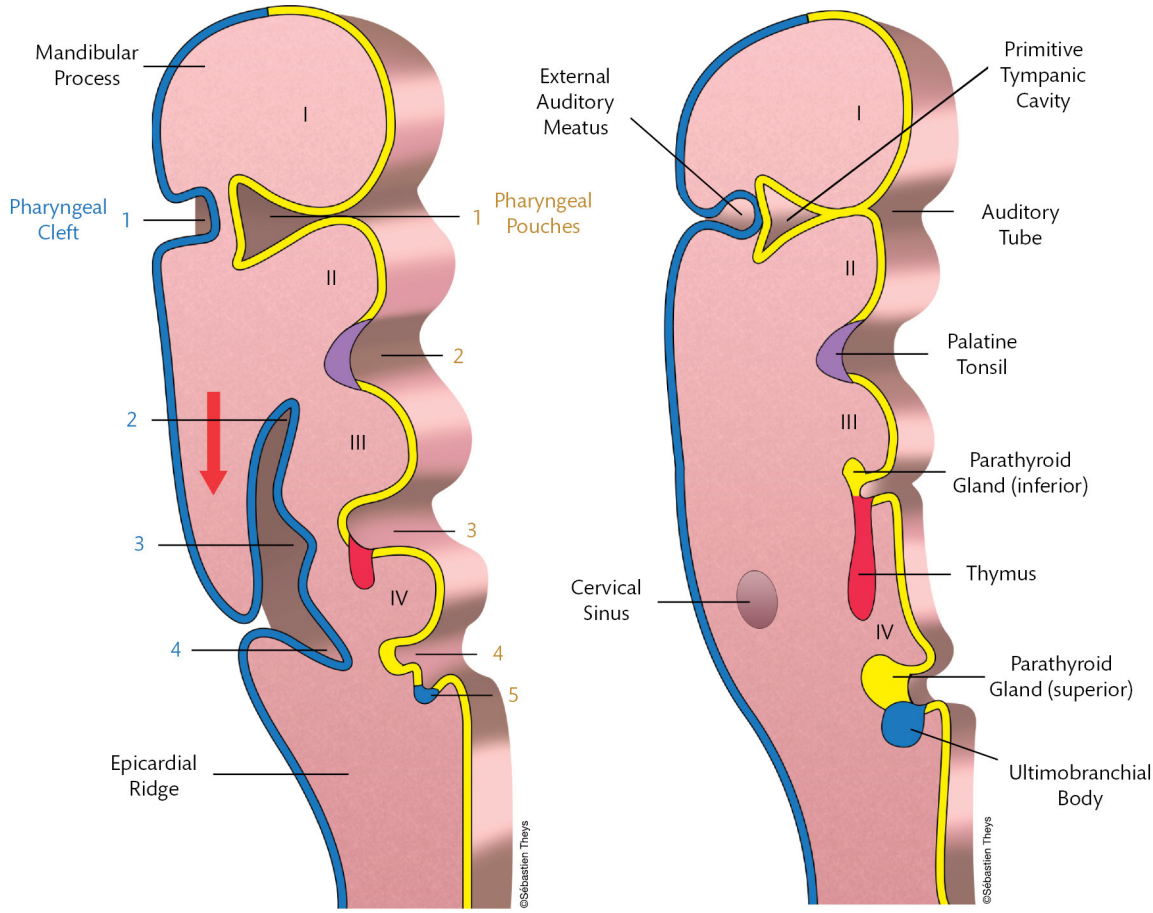

Figure 67.3 Cervical sinus formation.
Courtesy of Sebastien Theys.

Figure 67.4 Second branchial cyst.

Third and fourth branchial arch anomalies: these are very rare and represent 2–8% of branchial arch anomalies. Clinically, they appear similar to the second branchial arch anomalies, with a cutaneous opening in the supraclavicular area, but internally they enter the pharynx through the pyriform sinus. They are separated anatomically by their relationship to the superior laryngeal nerve (the third is above and the fourth below the nerve) and their origin from the pyriform sinus (the third is from the lateral wall and the fourth is from the apex). The third cleft anomalies are often present in the posterior cervical space, posterior to the SCM. The fourth cleft anomalies are more common on the left side, inferior to the mediastinum, following the path of the tracheo-oesophageal groove, and can occur with recurrent neck abscesses or suppurative thyroiditis.

Differential diagnoses

- *First branchial cleft anomalies*: parotitis with abscess formation, lymphatic malformation, parotid sialocele, and benign lymphoepithelial cyst.
- *Second branchial cleft anomalies:* lymphatic malformation, cystic nodal metastasis (human papillomavirus cancer, papillary thyroid cancer), lymphadenopathy/abscess, schwannoma, and paraganglioma.
- *Third branchial cleft anomalies:* lymphatic malformation, abscess, cervical thymic cyst, infrahyoid thyroglossal duct cyst, cystic nodal metastases, and laryngocele.
- *Fourth branchial cleft anomalies:* cervical thymic cyst, lymphatic malformation, thyroglossal duct cyst, thyroid colloid cyst, and abscess.

Investigations

The clinical history and the physical examination are the most important factors for the evaluation of a congenital neck mass. There are no pathognomonic imaging features. The correct diagnosis depends also on the understanding of anatomy and a high index of suspicion. Some evaluation can help to confirm the diagnosis or to evaluate the anatomy preoperatively, which are dealt with in detail in Chapter 69:

- *Ultrasonography*: they are well-circumscribed, thin-walled, and anechoic with evidence of compressibility and posterior acoustic enhancement.
- *Computed tomography (CT) scan*: they are well-circumscribed, low-density cystic masses (**Figure 67.5**). Cyst wall thickness and enhancement are variable and increase with recurrent infections. The 'beak sign' is pathognomonic of a second branchial cyst and represents a curved rim of tissue or 'beak' pointing medially between the internal and external carotid arteries.
- *Magnetic resonance imaging (MRI)*: on T1-weighted imaging, they may turn from low to high signal depending on the proteinaceous content of the cyst, but are typically hyperintense on T2-weighted imaging. This can be relevant in cases of extension into the mediastinum or deep spaces of the neck.

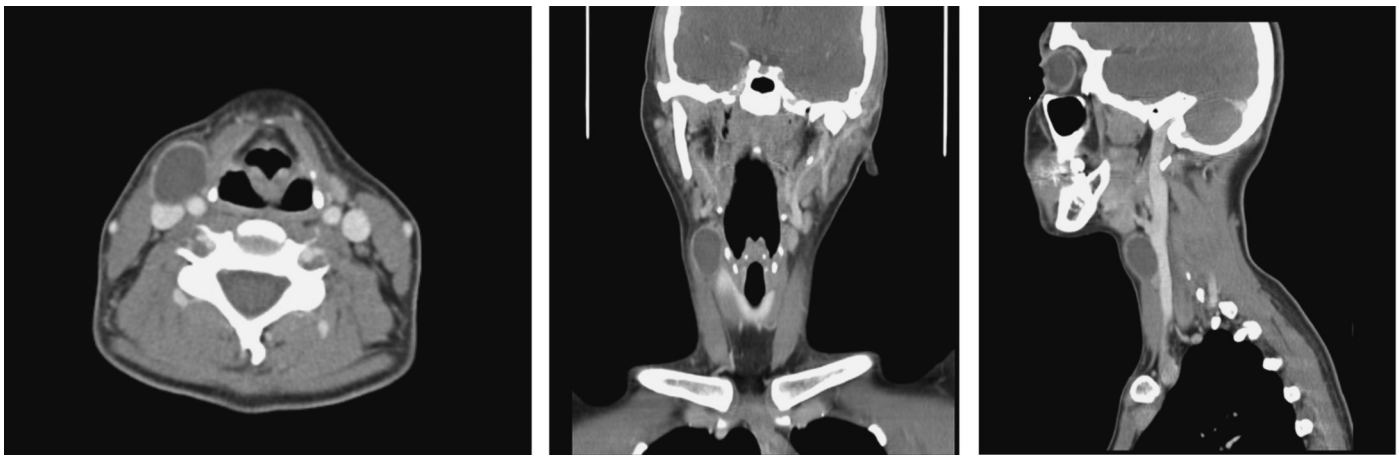

Figure 67.5 CT scans of second branchial cysts.

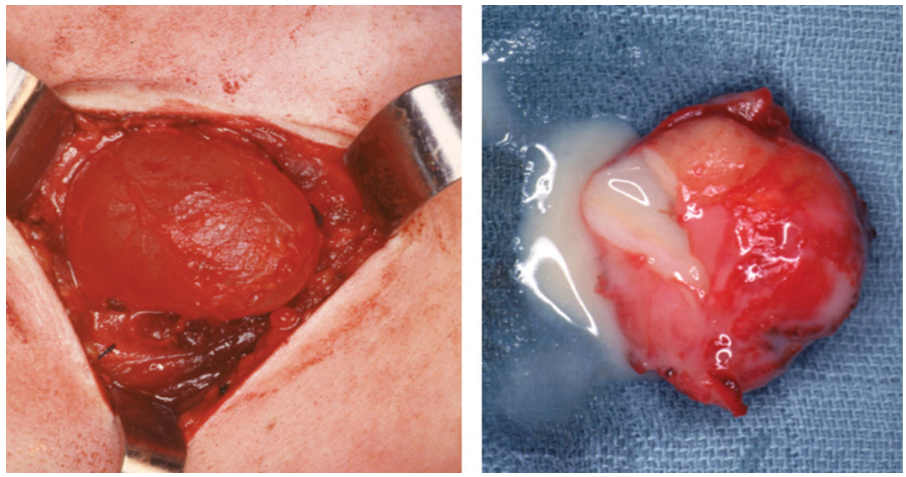

Figure 67.6 Second branchial cyst: (a) surgical view; (b) specimen with its typical fluid.

- *Barium swallow test/fistulography*: injecting radiopaque dye can help to delineate the path of the tract and determine the size, especially when the tract communicates with the pharynx.
- *Panendoscopic evaluation*: to evaluate a pharynx communication.
- *Fine needle aspiration*: is helpful to distinguish a branchial cleft cyst from malignant neoplasm, especially when it appears the first time in an adult older than 40 years.

Histologically, they are filled with a turbid yellow fluid containing cholesterol crystals and are limed by stratified squamous epithelium.

Treatment

The treatment is surgical with a *complete excision* of the lesion, encompassing the external sinus opening with the dissection of the sinus tract (Figure 67.6). The complete excision is essential for good outcomes and to avoid recurrences (<3%). In case of acute infection, systemic antibiotics are required, with abscess drainage if needed. The latter might cause more scarring, but definitive management should be postponed by 3–6 months. If previous surgery or recurrent infection has occurred, the recurrence can be as high as 20%.

Some teams recommend specimen analysis for the adult over 40 years by fresh frozen section during the surgery to exclude malignant pathology, as this can influence further treatment on table.

Depending on the lesion location, the dissection will be made carefully to avoid nerve or vessel damage. For example, the first branchial cleft cyst resection needs a good knowledge of the facial nerve and sometimes a superficial parotidectomy with nerve monitoring. Third and fourth branchial clefts cysts require recurrent laryngeal nerve monitoring with some needing a thyroid lobectomy.

Tips and pearls

- In the case of a fistula, the cutaneous tract can be identified and followed by the *insertion of a lacrimal probe or a catheter and the injection of methylene blue* (Figure 67.7). This helps to ensure complete excision and decrease the chance of recurrence.

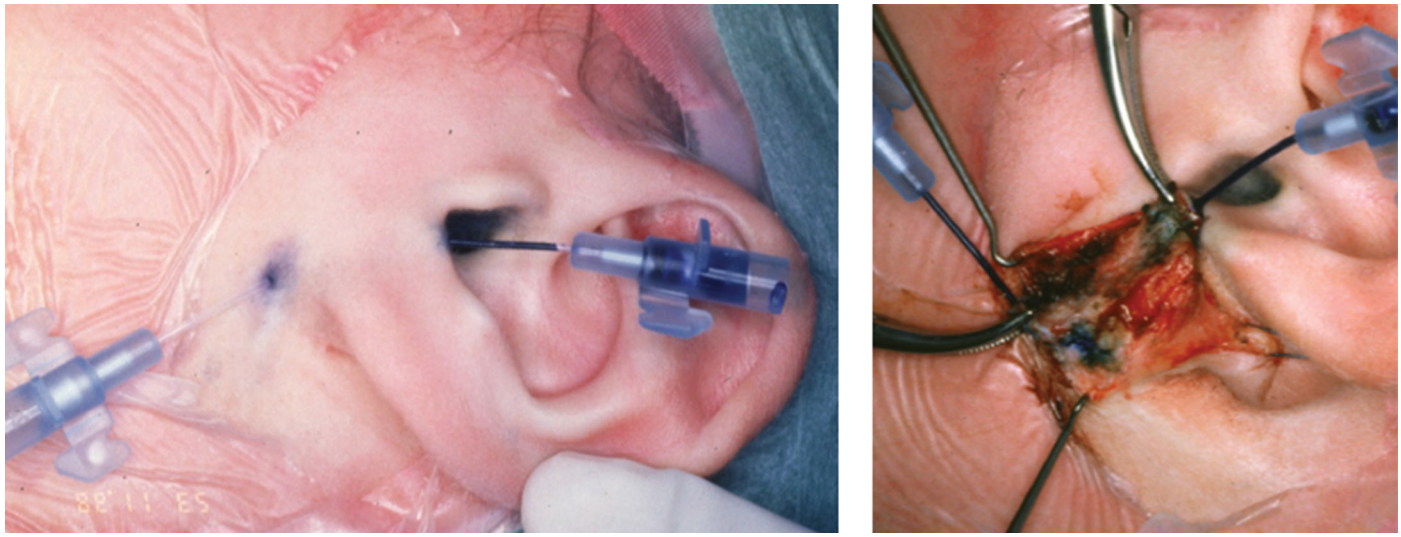

Figure 67.7 First branchial cyst: methylene blue injection.

Figure 67.8 Second branchial cyst: second stepladder incision.

- If the tract is long, a *second 'stepladder' incision* is sometimes needed to obtain better exposure (Figure 67.8).

Controversies/areas of interest for the future

- *Endoscopic cauterization approach*: when there is a pharyngeal communication, an alternative to the classical external surgery is to cauterize endoscopically the origin of the fistula. Options include electrocautery, carbon dioxide laser, or chemocautery by silver nitrate, especially for fourth branchial cleft lesions. Some teams recommend this approach for the children younger than 8 years, to minimize complications.
- *Ethanol ablation*: this technique can be used for patient who refused or are ineligible for surgery. It involves an ultrasound-guided aspiration of the cystic fluid, followed by injection of absolute ethanol into the lesion. The ethanol provides a sclerosis of the cyst without severe adverse effect. Mild pain and discomfort related to the needle puncture can occur. Some patients need multiple sessions (especially in cases of larger cysts).
- *Retro-auricular approach*: this is an alternative to the classical transcervical approach. The surgery takes more time with a bigger subplatysmal skin flap but the cosmetic outcomes are better. No other complications have been described.
- *New endoscopic/robotic approach*: these technics can be used by a transcervical or a retro-auricular approach without negative outcomes. This reduces the length of incision with a magnification of the surgical field compared to the classical open approach.

Thyroglossal duct cyst

Anatomy and embryological considerations:

Around the third week of gestation, the primordium thyroid, originating at the foramen caecum in the tongue, starts to descend to the neck. At the seventh week, it reaches its final position in front of the second and third cartilage of the trachea. The involution of this pathway, named the *thyroglossal duct*, starts at the fifth week of gestation, and is completely closed at the fourth month. If any portion of the duct persists, it can result in *thyroglossal duct cyst* formation (Figure 67.9).

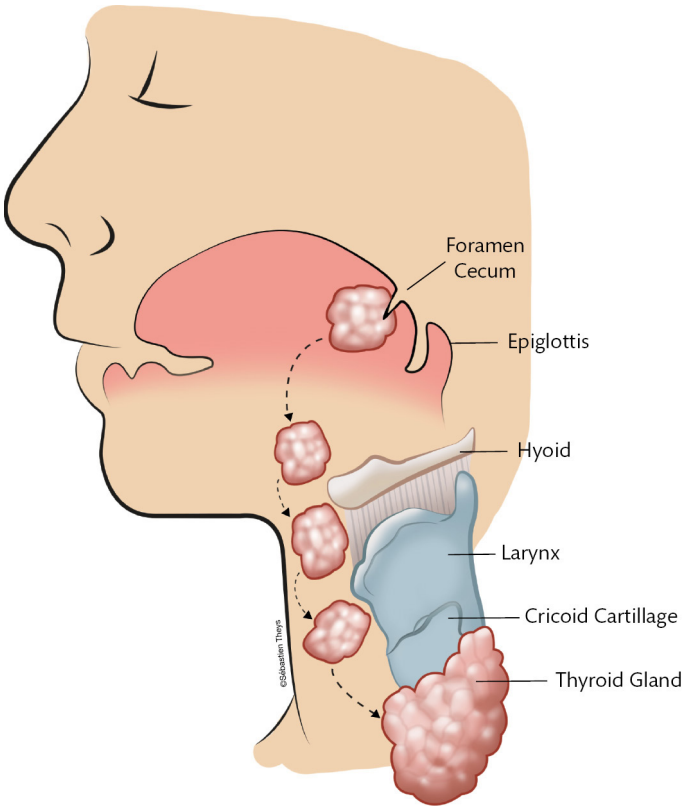

Figure 67.9 Thyroglossal duct cyst formation.
Courtesy of Sebastien Theys.

Assessment

Thyroglossal duct cysts are the most frequent congenital cervical anomalies, with a 7% population prevalence worldwide, and are mostly diagnosed during the first decade of life. Clinical examination shows a mobile midline mass, near the hyoid bone, which elevates with tongue protrusion or swallowing. Most cystic masses arise at the infrahyoid level (65% of the cases). They can become tender, enlarged, or inflamed in cases of superinfection or abscess formation (Figure 67.10). Fistula can occur in cases of infection (spontaneous suppuration) or surgical postoperative fistulization (simple incision or incomplete excision).

Differential diagnoses

They include cystic metastatic lymph nodes (papillary thyroid carcinoma or squamous cell carcinoma), dermoid or epidermoid cysts, lipoma, laryngocele, thymic cyst, bronchogenic cyst, and second branchial cleft cysts.

Investigations

Ultrasound comprises the first line of preoperative examination to diagnose the cyst and search for the presence of normal thyroid tissue. *CT scanning or MRI* are not necessary but can be used in cases of recurrences. Some teams recommend *thyroid function testing*. *Fistulography* should be avoided, as it can be responsible for fistulization of the cyst to the skin. But it can be useful in cases of recurrences to evaluate the course of deep fistula tract.

Figure 67.10 Thyroglossal duct cyst.

Fine needle aspiration can be used for adults over 40 years to exclude neoplasia.

Histologically, they are lined by a respiratory epithelium, a squamous epithelium, or both. Some ectopic thyroid gland tissue can also be found.

Treatment

The treatment is a complete surgical excision of the cyst with the *Sistrunk* technique (Figure 67.11). This consists of the resection of the body of the hyoid bone en bloc with the cyst. Indeed, the resection of the cyst alone has a risk of recurrence around 45–55% compared to less than 5% with the Sistrunk procedure. The risk factors for recurrence are surgery during the inflammatory phase, cyst rupture during dissection, multiple thyroglossal ducts, or technical surgical errors. Surgical complications can be infection, laryngotracheal defect, haematoma, and hypoglossal nerve injuries.

In case of acute infection, systemic antibiotics and needle aspiration are recommended, to facilitate identification of the microorganisms and guide appropriate antibiotics. Drainage by incision should be avoided, as it increases the risk of fistula or recurrences. The surgical approach should be postponed until 3 months after infection. After surgery, thyroid function testes are recommended.

Surgical tips

- Avoid rupture of the cyst during dissection.
- Resection of the body of the hyoid bone en bloc with the cyst.
- Avoid excessive lateral dissection of the hyoid bone (to avoid risk of hypoglossal nerve or lingual artery injuries).
- Perform a ligation of all possible thyroglossal ducts.
- Put a finger in the mouth over the area of the foramen caecum to avoid entry in the oropharynx.

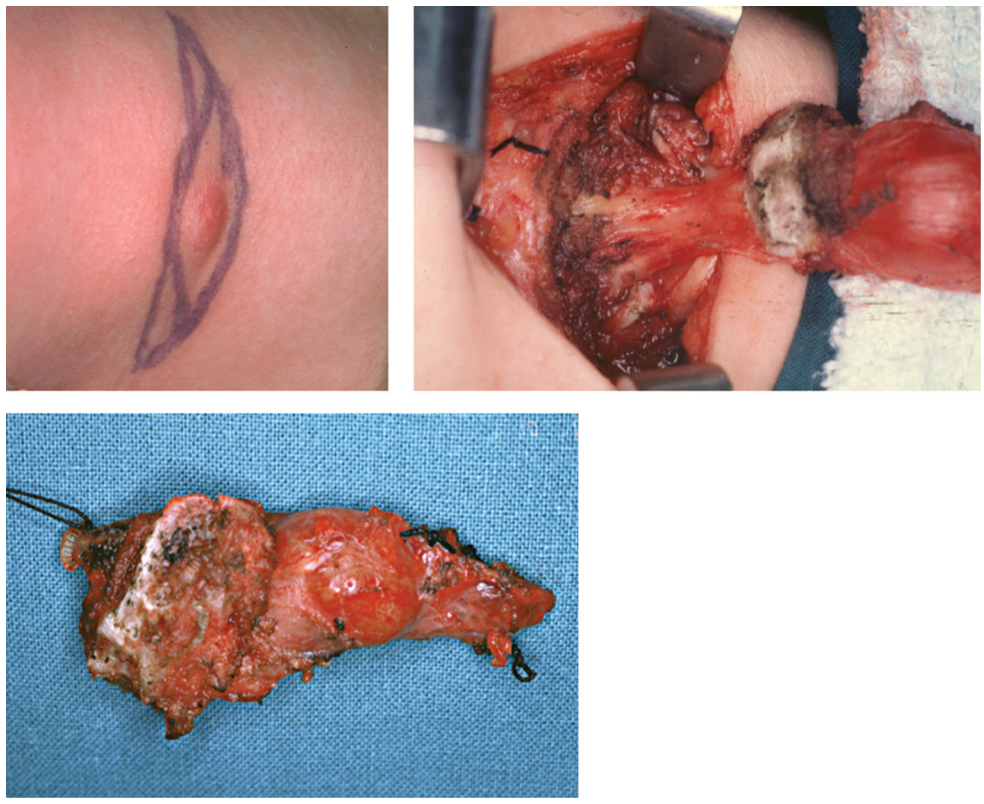

Figure 67.11 Thyroglossal duct cyst: Sistrunk surgery.

- Remove the cutaneous fistula (if present) with a wedge-shaped incision.
- Re-approximate the two ends of the hyoid bone at the end of the surgery.

Controversies/areas of interest for the future

- *Thyroglossal duct cyst carcinoma*: can occur in less than 1%, the most common finding histologically is papillary carcinoma followed by squamous cell carcinoma. The surgical management remains controversial. Some authors systematically add a total thyroidectomy to the Sistrunk procedure. The rationale for this is the possible presence of concomitant carcinoma in the main thyroid, the possible utilization of radioactive iodine as adjuvant therapy, and the role of serum thyroglobulin as a follow-up marker of cancer. Other authors propose a staging strategy based on low-risk patients (wait and see), moderate-risk patients (total thyroidectomy +/− radioactive iodine), and high-risk patients (and add cervical lymph node dissection). Criteria for low risk are age under 45 years, no history of radiation, tumour size less than 4 cm, no soft tissue invasion, no signs of distant lymphatic metastases, and no aggressive tumour histology. Overall prognosis is excellent.
- *Transoral surgical approach*: this has been developed by some teams. It consists of making a transoral anterior incision above the submandibular gland opening, to separate the genioglosses muscles and to identify the cyst and the thyroglossal duct. The surgery is guided by an endoscope and the hyoid bone is also cut and pulled upward. The advantages of this procedure are the magnification of the view, which helps in the identification of the thyroglossal duct, and the cosmetic satisfaction without cervical incision. No other complications have been descripted.

Schwannoma

Anatomy and embryological considerations

Schwannomas are also called neuroma, neurinoma, or neurilemmoma. They represent a benign tumour composed of Schwann cells which come from the neural crest. Approximately 75% are intradural, 10% intra- and extradural, and 15% extradural.

They can occur along cranial nerves (except the optic and olfactory nerves), spinal nerves, and the autonomic nervous system. They are found throughout the body, but a head and neck location accounts for 25–40% of all schwannomas. Cervical vagal schwannoma constitutes about 2–5% of neurogenic tumours. Some rare locations are also described, such as the base of tongue.

These tumours are mostly solitary but can occur in multiple locations and may be associated with syndromic conditions such as neurofibromatosis type 2 and schwannomatosis. The *NF2* gene on chromosome 22 plays a role in sporadic and syndromic development.

Assessment

Schwannomas are slowly growing tumours and remain asymptomatic for quite long periods. These are generally well encapsulated and can incidentally be found on imaging for other pathologies. The disease is more prevalent in the fifth decade of life.

Some symptoms can appear with time, such as tenderness of the cervical mass and neurological symptoms in larger lesions (mostly hoarseness by vocal fold paralysis). A paradoxical cough during palpation of the cervical mass is another sign of vagal schwannoma (due to vagal stimulation). In cases of vestibulo-cochlear schwannomas, some other neurological symptoms can occur, such as hearing loss, tinnitus, and imbalance. There is often a 5-year delay between symptoms and diagnosis.

Differential diagnoses

The differential diagnoses are neurogenic tumours (neurofibroma, chordoma), meningioma, giant cell tumours, lipoma, metastatic lymph node, haemangioma, lymphangioma, and benign salivary gland tumours.

Investigations

- *ENT examination*: oral examination can show a tonsillar bulging (in case of parapharyngeal mass effect) and laryngoscopy is recommended to assess vocal fold motion.
- *CT scanning and MRI*: show the well-defined mass and its anatomical rapport with the nerve. MRI is preferred. The tumour appears isointense or hypointense on T1-weighted and hyperintense on T2-weighted images. The lesion enhances uniformly with gadolinium contrast.
- *Histology*: confirms the diagnosis. They are characterized in microscopic examination by Antoni A and Antoni B pattern. Immunohistochemistry is often positive for S-100 protein.

Treatment

Different treatments can be proposed:

- *Surgical management*: this is the treatment of choice. Schwannomas do not infiltrate the nerve of origin; therefore, they can usually be separated from it. Intracapsular excision with gentle dissection between the tumour capsule and normal fascicles minimizes the risk of nerve damage. The use of a microscope during surgery is helpful and reduces postoperative complications due to nerve damage.
  - In case of a large tumour, the nerve will need to be sacrificed. Reconstruction by anastomosis or sural nerve graft can be used.
  - Some postoperative complications can appear, such as dysphagia, vocal cord palsy or paralysis, eye ptosis, or marginal mandibular nerve paresis. If the nerve is complete and non-damaged, patients have a total recovery in 1-year follow-up. Speech therapy can be useful.
  - Finally, the malignancy transformation is rare (8% of all head and neck schwannoma). In case of malignancy suspicion, the surgical approach must be considered, and the specimen must be sent for anatomopathological examination.
- *Radiotherapy and stereotactic radiosurgery*: can be used when surgery is contraindicated (elderly patients, comorbidities with large or symptomatic tumours, or non-resectable lesions). However, the rate of radiotherapy resistance is high.
- *Observational management*: due to their slow-growing characteristic and in cases of elderly patients or patients with multiple comorbidities, observational management can also be proposed. Yearly brain MRI follow-up assesses the rate of tumour growth.

The prognosis is excellent and recurrence after total resection is uncommon. Long-term surveillance is not recommended.

Paraganglioma

Anatomy and embryological considerations

Paraganglioma are rare neuroendocrine tumours which originate in the *neural crest* from where they migrate beyond the sympathetic chain. Approximately 85% occur in the abdomen (largely represented by pheochromocytoma), 12% in the thorax, and 3% in the head and neck. In the head and neck, the carotid body is the most common location, followed by jugular foramen, vagal nerve, and laryngeal locations. Some other names used for these tumours are chemodectomas and glomus tumours. Two types are described:

- *Parasympathetic*: usually asymptomatic and inactive (non-secreting tumours), mostly located in head and neck.
- *Sympathetic*: highly active with catecholamine (norepinephrine) secretion and symptomatic. They are mainly located in the abdomen and pelvic regions but can sometimes occur in the head and neck.

Assessment

Paragangliomas are hypervascular and slow-growing tumours. They affect women more often (3:1) over 45 years. Paragangliomas are mostly sporadic, but 30–40% are familial, and can be associated with genetic syndromes including mutations in the succinate dehydrogenase (*SDH*) gene, neurofibromatosis, von Hippel–Lindau disease, and multiple endocrine neoplasia type 2. In these cases, they can be bilateral or in multiple locations. Familial paragangliomas often present a decade earlier and can be multifocal.

They can be asymptomatic (incidental finding on radiological imaging) or present with a painless and pulsatile lateral neck mass. Some symptoms due to vascular or nervous compression by mass effect can occur (cranial nerve paralysis, dysphagia, hoarseness, bradycardia, syncope, and pulsatile tinnitus). In cases of catecholamine secretors, symptoms can be tachycardia, hypertension, dry month, flushing of the face, fatigue, headaches, profuse sweating, tremors, and general weakness.

These are usually benign tumours, but less than 5% can turn malignant; the only indicator for malignancy is the cervical lymph nodes or distant metastases (local invasion is *not* a criterion).

Differential diagnoses

Differential diagnosis includes cervical lymph nodes, schwannomas, vascular tumours, branchial cysts, and salivary tumours (deep lobe of parotid).

Investigations

- *ENT examination*: will show a tonsillar bulge (in cases of parapharyngeal masses) and laryngoscopy is recommended to assess vocal fold motion.
- *Blood and urine tests*: will identify plasma or urinary catecholamine or its metabolites (metanephrines), indicating secreting tumours. False-positive results from concurrent medications (tricyclic antidepressants, phenoxybenzamine) and consumption of catecholamine-rich foods (fruit, nuts, cheese, and alcohol) should be considered.
- *CT scans*: show a hypervascularized lesion and can be helpful in identifying its anatomical relationship with the carotid artery and the skull base. The 'lyre sign' is pathognomonic (splaying of the internal and the external carotid system).
- *MRI*: this is the standard imaging and shows the typical 'salt and pepper' lesion (that reflect flow voids and blood supply). It can also evaluate the relationship with the neural structures and the surrounding soft tissue involvement (**Figure 67.12**).
- *Functional imaging*: can be performed to identify multiple lesions, for example, ^{111}In Octreoscan or ^{18}F-DOPA positron emission tomography/CT.
- *Genetic evaluation* for *SDH* mutation is usually performed; centres may choose to additionally look for *RET*, neurofibromatosis type 1, and multiple endocrine neoplasia mutations.
- *Fine needle aspiration biopsy*: should be avoid because of the bleeding risk.

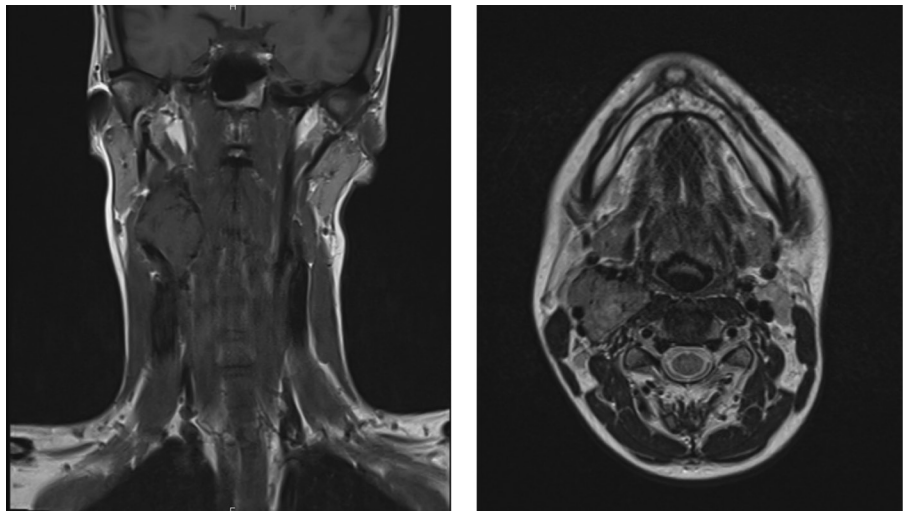

Figure 67.12 MRIs of paraganglioma.

- *Angiography and occlusion imaging*: may be needed in selected patients to evaluate the loop of Willis and cerebral perfusion and to minimize risk of stroke prior to tumour surgery.

Treatment

No guidelines exist about the management in head and neck paraganglioma. Different approaches are possible and are in constant evolution such as surgical excision, observational management, radiotherapy, and stereotactic radiosurgery. Multidisciplinary planning and counselling are recommended.

- *Surgical management*: is indicated for smaller to medium tumours and for younger, relatively healthy patients (with a local control between 94% and 100%). Complications can be significant and are based on the lesion size and vessel/nerve involvement. Cranial nerve deficit rates are around 14% for a tumour smaller than 5 cm compared to 67% if it is larger than 5 cm. In particular, complete resection of vagal nerve location requires removal of the associated vagal nerve and creates univocal fold paralysis and pharyngeal plexus deficit. So surgical management for this location is less commonly recommended.
  - Additional factors for which surgery should be considered include secreting tumours, concern for malignancy, pain, rapid growth, or contraindications for radiation.
  - *Preoperative embolization* can reduce blood loss and operating time and facilitate the dissection (Figure 67.13). It should be performed 24–48 hours prior to surgery with a carotid angiography visualization. The main risk of embolization is product migration. Some teams reserve this procedure for larger lesions only.
- *Observational management*: because of the slow-growing characteristic of paragangliomas (approximately 1 mm/year), observation should be considered for elderly patients, those with significant comorbidities, and those who are asymptomatic or minimally symptomatic. New or progressive cranial nerve deficits can appear in time with a risk around 30%. There are no widely recommended observation protocols, with 6–12-monthly MRI (depending symptoms) being recommended. Earlier imaging is performed in cases of progression of symptoms, pain, tumours growing in size, or concern for malignancy.
- *Stereotactic radiosurgery and radiotherapy*: should be considered for large tumours, particularly in situations with a skull base invasion, vagal nerve location, or risk to vessel or nerve sacrifice with the surgery (with permanent long-term morbidity). Other indications are elderly patients or patients with comorbidities and contraindications for surgery, tumours recurring after surgery, or when a progression occurs during an observational approach. Stereotactic radiosurgery allows high-dose gradients on the tumours and limited toxicity to healthy tissues and is therefore suitable for paraganglioma smaller than 3 cm. Radiotherapy can be used for lesions larger than 5 cm (45 Gy in 25 fractions over 5 weeks). The goal of radiotherapy is not a complete eradication but a lesion size stabilization or partial regression.

Figure 67.13 Preoperative embolization.

Surgical tips

- Adequate exposure.
- Vascular control:
  - Preoperative embolization especially for large tumours.
  - Carotid stent can be uses as an alternative to embolization to exclude the tumour blood supply (without interrupting the arterial blood flow and to facilitate the tumour resection).
  - Careful dissection of the carotid bulb which is the zone with greatest risk of rupture.
  - When a resection of carotid artery wall is necessary, a vascular surgeon should repair it with a patch of an interposition vein graft. A temporary shunt is necessary when more than 25 minutes is required.

Controversies/areas of interest for the future

Management of bilateral paraganglioma: the rationale for surgery and especially the side to do first is matter of debate, and there is no conclusive literature to guide clinicians. The goal is to minimize the risk of bilateral cranial neuropathies, damage to the baroreceptor system, and/or impact on the cerebral circulation. The authors recommend that the smaller side be operated first. If complications appear, observation or radiotherapy should be considered for the other side. In cases of bilateral small lesions, the larger side is usually excised first. In case of a vagal nerve paraganglioma on one side and a carotid bulb on other side, the carotid bulb should be managed first.

Dermoid cyst

Anatomy, embryological considerations, and assessment

A dermoid cyst is a congenital, benign cutaneous tumour that comes from the entrapment of ectodermal elements along the lines

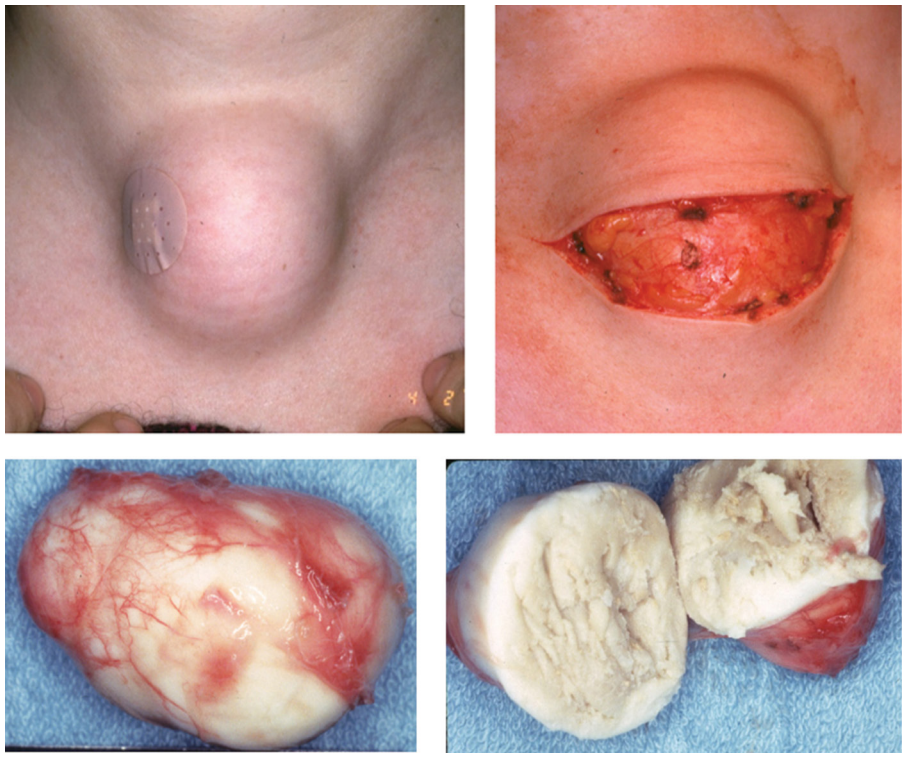

Figure 67.14 Dermoid cyst.

of embryonic closure (mostly on the midline). The histology shows a well-defined cyst lined by a stratified squamous epithelium. It can contain mature adnexal structures of mesodermal origin such as hair follicles, sebaceous glands, and eccrine glands. These cysts may be present at birth, grow slowly, and are often diagnosed before 5 years. They are mostly present in facial region locations (periorbital, nasal, occipital, and frontal) and can occur also in the neck (often in the midline, but some lateral locations are described) or in the floor of the mouth. They are usually asymptomatic and present as subcutaneous nodules. Hair protruding from a dermoid cyst punctum is pathognomonic (Figure 67.14).

Differential diagnoses

These include epidermoid cyst, lipoma, pilar cyst, teratoma, subcutaneous abscess, neurofibroma, lymphatic malformation, and thyroglossal duct cyst.

Investigations

The diagnosis is made clinically; in cases of uncertainty, ultrasound can be used and shows a well-defined heterogeneous and hypoechoic cyst. For tumours located more cranially, CT scanning or MRI are often used to exclude intracranial or intraspinal invasion.

Treatment

The treatment is the complete surgical excision without disruption of the cyst. If the cyst wall ruptures, the remnant tissue needs to be removed with curettage and abundant irrigation. The specimen needs pathological examination to confirm the diagnosis and exclude malignancy.

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68

Head and neck pathology

Max Robinson and Selvam Thavaraj

Introduction

This chapter offers a perspective into the broad range of pathological diagnoses seen within the various subsites of the head and neck, focusing on the more common entities. With greater understanding of the molecular profile of tumours and the availability of drugs against actionable mutations, this is a rapidly evolving field.

Sample acquisition

Accurate diagnosis requires an adequate sample. Head and neck anatomy is complex and access to many sites is challenging; sampling is often compounded by the friable nature of diseased tissue. Adequate access and optimal visualization of the biopsy site are prerequisites to providing a satisfactory sample. Blind biopsies typically produce superficial disrupted samples, which are difficult for the laboratory to process and for the pathologist to interpret, potentially delaying diagnosis and treatment. Endoscopy is essential for guiding upper aerodigestive tract biopsies and ultrasound is routinely used for needle and core biopsies of salivary gland pathology and neck lumps.

The pathology request form

Please remember that completing the pathology request form, although tiresome, is an important step in the diagnostic process. Ensuring that the correct sample is sent with the correct form is essential and should be carried out contemporaneously, prelabelling multiple forms and specimen pots ahead of a biopsy list is bad practice and risks mixing samples up. Patient details, hospital location, consultant name, clinical history, specimen type, and date and time of biopsy are key fields that should be completed. A clinical differential diagnosis and indication of the level of clinical suspicion of malignancy are helpful. All samples must pass strict quality checks in the laboratory before they are accepted and assigned a unique accession number and barcode for tracking purposes.

Sample pathway

Cytology samples

The contents of fine needle aspiration biopsies can be used to prepare direct smears on glass slides, but this requires careful technique and is a difficult skill to master. The smears can be air-dried or sprayed with an alcohol-based fixative. A safer option for the novice is to use a liquid-based cytology technique, where the needle contents are deposited into a proprietary preservation fluid (e.g. SurePath, BD; ThinPrep, Hologic). The needle can also be 'washed' several times in the fluid to maximize the yield of cells. In fact, it is recommended that even when direct smears have been prepared 'needle washings' in alcohol preservative should be collected as these can be centrifuged to prepare a cell block for further analysis. The direct smears or liquid-based cytology specimen are sent to the laboratory for processing. Cytology samples are usually stained with either Papanicolaou (PAP) or Giemsa. Liquid-based cytology has the advantage that numerous slides can be prepared for ancillary techniques such as immunocytochemistry.

Tissue samples

Tissue biopsies are routinely collected and fixed in 10% neutral buffered formal saline (formalin). The volume of fixative should be at least ten times that of the tissue to allow adequate preservation. For routine diagnostic samples specimen pots containing 60 mL of formalin are used. In some circumstances it is necessary to send fresh tissue to the laboratory, for example, if a vesiculobullous (blistering) disorder is suspected or if intraoperative frozen sections are required to assess excision margins. Routine fixed tissue samples are processed by dehydration and infusing with paraffin wax prior to embedding in a wax block from which 5 μm sections are cut using a microtome and mounted on glass slides prior to staining with haematoxylin and eosin (H&E). Ancillary histochemical stains and tests such as immunohistochemistry and *in situ* hybridization can be performed on additional sections from the block. Furthermore, routine molecular tests such as fluorescence *in situ* hybridization and standard polymerase chain reaction may also be performed on formalin-fixed paraffin-embedded tissue. Currently, pathologists

assess the glass slides using a microscope; however, there is a drive to a digital workflow, whereby slides are scanned at high resolution to generate a navigable digital image. Such technology will enable the development of artificial intelligence algorithms and the pave the way for automated diagnosis and quantitative analysis of key biomarkers.

Excision specimens

Careful preparation of the *ex vivo* surgical specimen in the theatre optimizes the yield of information the pathologists can provide the oncology team. An indication of anatomical landmarks, critical margins, and the relationship of margin biopsies to the main specimen are all helpful. For example, transoral robotic surgical specimens can be pinned to a board or stitched to a sponge sheet, and laser vocal cordectomy specimens can be glued to a cucumber biomount (Figure 68.1). Pathologists' work is enriched by communication with the surgical team and the ability to provide accurate information that guides patient treatment options. In the laboratory the specimens are described and photographed, prior to the application of ink to margins, which maintains specimen orientation during block dissection (Figure 68.2). Sampling of the surgical specimen, also known as 'cut-up' or 'grossing', is carried out using standard operating procedures to ensure that important anatomical features and excision margins are selected for processing into wax blocks.

Interpreting the pathology report

The pathology report includes the patient demographics, hospital location, responsible clinician, and reporting pathologist. The macroscopic report documents the gross specimen features and the block selection. The microscopic report typically includes a narrative description of the histological features but should come to a conclusion or diagnosis. For resection specimens, proforma-style reports that document items from evidence-based datasets are recommended. For malignancies, staging manuals from the Union for International Cancer Control (UICC) or American Joint Committee on Cancer (AJCC) are used to estimate the burden of disease, which can then be linked to prognosis. In future, as molecular profiling of tumours becomes more commonplace, these details will become integrated into pathology reports documenting prognostic factors and actionable mutations that inform personalized patient care.

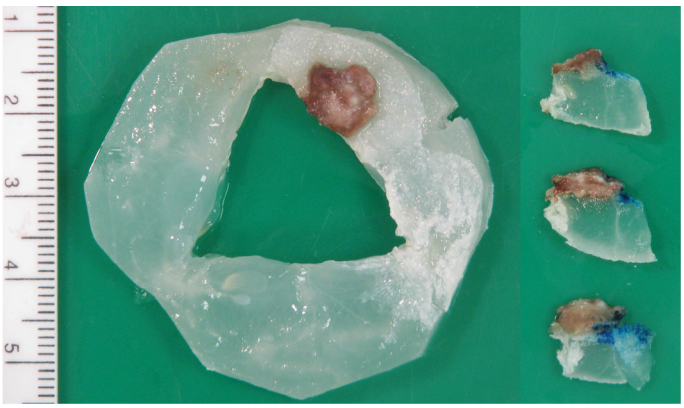

Figure 68.1 A cucumber biomount with a cordectomy specimen glued to the surface. Slices through the specimen demonstrate how the cucumber supports the orientation of the mucosal slices.

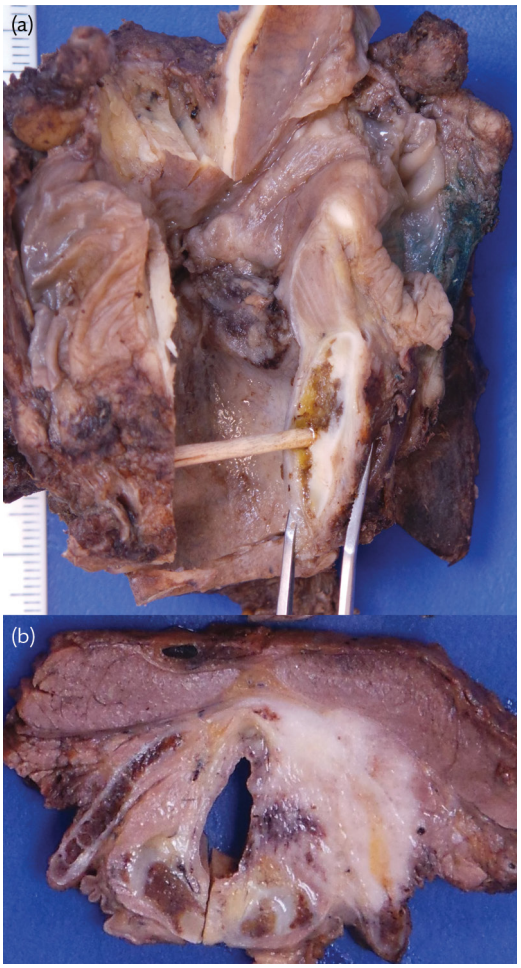

Figure 68.2 (a) A formalin-fixed laryngectomy specimen opened posteriorly to demonstrate an ulcerated transglottic tumour. (b) An axial slice demonstrates the extent of the tumour and in this case extralaryngeal spread into the anterior strap muscles.

Squamous cell carcinoma

The majority (90%) of head and neck malignancies are squamous cell carcinomas (SCCs) arising from the squamous epithelium lining the upper aerodigestive tract. A progression model from normal squamous epithelium to epithelial dysplasia and then SCC has been proposed and is underpinned by accumulating genotoxic damage caused by environmental carcinogens. The most common risk factors are tobacco and alcohol abuse, although oncogenic viruses such as human papillomavirus (HPV) and Epstein–Barr virus (EBV) are aetiological agents implicated in the development of oropharyngeal and nasopharyngeal carcinoma, respectively. It is becoming increasingly common for oncologists to request PD-L1 testing prior to using targeted immune-oncology drugs (e.g. nivolumab, pembrolizumab), particularly in the setting of recurrent and metastatic head and neck SCC that is refractory to standard chemoradiotherapy.

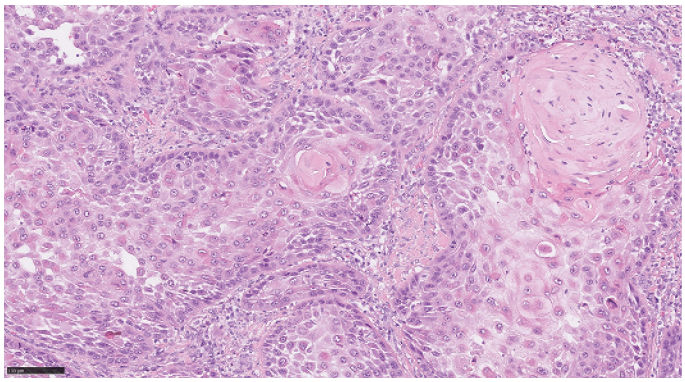

Figure 68.3 A moderately differentiated SCC showing areas of keratinization.

Laryngeal squamous cell carcinoma

Laryngeal SCC is strongly associated with tobacco smoking. Countries that have shown a reduction in the prevalence of smoking in the last few decades now have fewer laryngeal cancer cases, although the disease still represents a significant workload for most ear, nose, and throat (ENT) surgeons and oncologists around the world. Early symptoms, such as altered voice or cough, herald the changes on the vocal cord (covered by squamous epithelium). Initially there may be a white patch (leukoplakia) on the cord characterized by hyperkeratosis and hyperplasia or a red patch (erythroplakia) caused by epithelial atrophy and inflammation. Changes in the organization of the squamous epithelium, such as architectural disturbances and cytological abnormalities, reflect altered proliferation and differentiation and are termed epithelial dysplasia. Pathologists assign a grade based on the degree of disturbance; different grading systems have been recommended over the years, but currently high-grade and low-grade epithelial dysplasia are the preferred categories in the larynx. High-grade lesions are most likely to develop into cancer. High-grade dysplasia at the extreme end of the spectrum may also be referred to as carcinoma *in situ* by some pathologists. One of the problems of assessing laryngeal dysplasia is the small size and disrupted nature of the biopsy material, which is often tangentially cut in histological preparations. The pathologist may diagnose 'high-grade epithelial dysplasia' and indicate that an invasive component cannot be excluded; in these circumstances careful clinicopathological correlation is required. By contrast, sampling of a tumour mass is likely to provide a definitive diagnosis of SCC. The pathologist will then grade the tumour as well-, moderate-, or poorly differentiated SCC, based on the degree of keratinization and cellular anaplasia (Figure 68.3). For poorly differentiated SCC, immunohistochemistry may be required to render a diagnosis. Typically, the report will indicate that the malignant cells express cytokeratin intermediate filaments (AE1/AE or MNF116 positive), including cytokeratins 5, 6, and 14, and there is also nuclear expression of the transcription factors p40 and p63.

There are a few uncommon variants of SCC, but one worth mentioning is spindle cell carcinoma, which typically presents as a polypoid mass arising from the vocal cord. The tumour can be mistaken for a benign reactive lesion, but in reality is a polypoid mass of malignant cells undergoing epithelial–mesenchymal transition (Figure 68.4). Immunohistochemistry is usually not helpful as the epithelial markers are completely lost; however, focal areas of epithelial dysplasia and/or tiny islands of conventional SCC are clues to the diagnosis.

In other circumstances, when the pathologist encounters a poorly differentiated carcinoma, it is important to consider the possibility of a poorly differentiated adenocarcinoma or a neuroendocrine carcinoma, including small cell carcinoma. Head and neck primary adenocarcinomas typically express cytokeratin 7; occasionally the panel of immunohistochemical markers may need to be expanded to investigate the possibility of metastatic adenocarcinoma from another site. For neuroendocrine carcinoma the use of CD56, synaptophysin, and chromogranin is required to secure a diagnosis. Primary neuroendocrine carcinomas of the head and neck region are uncommon and consequently metastasis from a distant site should also be considered—small cell carcinoma of the lung would be top of the list.

Oropharyngeal squamous cell carcinoma

Oropharyngeal SCC is now classified into HPV-associated and HPV-independent disease. HPV-associated SCC has become more prevalent in the last decade or so and is now one of the most commonly diagnosed cancers by ENT surgeons. The significance of HPV-associated oropharyngeal SCC is that patients have a better prognosis by comparison to those with HPV-negative disease. Good prognosis seems to be irrespective of the treatment modality and similar outcomes are achieved using chemoradiotherapy or primary surgery with or without adjuvant treatment. The detection of oncogenic HPV is carried out using a combination of p16 immunohistochemistry and an HPV-specific test, such as HPV DNA or RNA by *in situ* hybridization

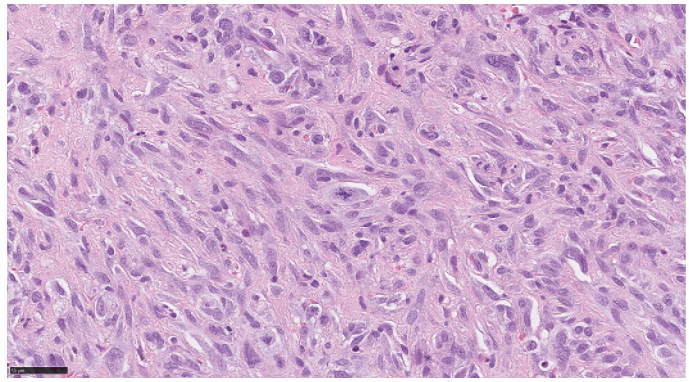

Figure 68.4 A laryngeal spindle cell carcinoma composed of highly atypical malignant cells. This particular tumour was completely negative on cytokeratin immunohistochemistry.

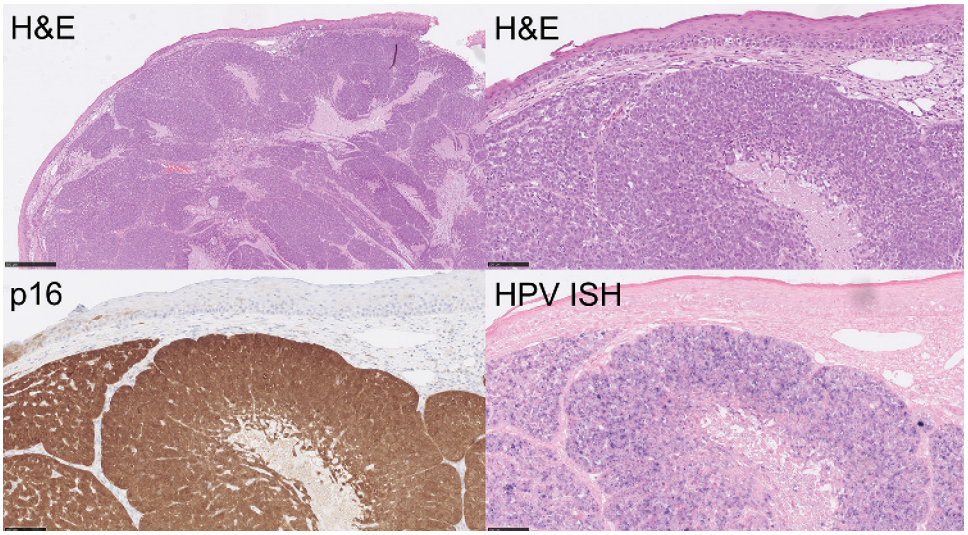

Figure 68.5 An HPV-associated oropharyngeal SCC. The carcinoma is non-keratinizing, p16 positive, and shows evidence of high-risk HPV DNA by *in situ* hybridization (ISH).

or by using a polymerase chain reaction to detect HPV nucleic acid sequences (Figure 68.5). The majority (90%) of infections are caused by HPV-16, although other high-risk HPV genotypes can be detected, such as HPV-18 and -33. Surgical treatment of oropharyngeal SCC has been revolutionized using transoral approaches including robotic surgery. The specimens require careful annotation and communication with the pathologist to ensure that the margins can be accurately assessed and the disease appropriately staged.

Nasopharyngeal carcinoma

Nasopharyngeal carcinoma is a type of SCC. Typically, it is non-keratinizing and blends in with the resident lymphoid tissue at the site and is well recognized by pathologists as a potential 'pitfall' in diagnosis. Small, superficial biopsies may fail to adequately sample the tumour. In geographical areas where nasopharyngeal carcinoma is endemic (e.g. South East Asia and China), the main aetiological agent is EBV. EBV can be detected in tissue biopsies using *in situ* hybridization against EBV early encoding RNA. Although the presence of EBV does not have prognostic implications, its use in diagnosis is invaluable. The other types of nasopharyngeal cancer and their impact on prognosis and treatment selection in the event of recurrence are discussed in Chapter 61.

Sinonasal tumours

The majority of sinonasal carcinomas can be classified as conventional non-keratinizing or keratinizing SCC. Oncogenic HPV is occasionally identified, but the prognostic significance of this is unclear. A small number of benign sinonasal papillomas are known to transform to cancer; judicious sampling of these specimens is required to exclude malignancy. Sinonasal undifferentiated carcinoma is an entity, but effectively a diagnosis by exclusion. The use of molecular pathology is starting to diminish this group and new entities such as NUT carcinoma, SWI/SNF complex-deficient carcinomas, and *DEK::AFF2* fusion carcinomas are beginning to be more widely recognized.

Adenocarcinomas of the sinonasal complex are uncommon, but the intestinal-type is rather distinct and has morphological features that resemble colorectal carcinoma. Non-intestinal-type adenocarcinomas are also encountered and may include the spectrum of salivary gland-type adenocarcinomas. HPV-related multiphenotypic sinonasal carcinoma is a recently recognized entity which may have overlapping features with sinonasal adenoid cystic carcinoma.

Olfactory neuroblastoma is one of the so-called small round blue cell tumours, which are diagnostically challenging for even the most experienced pathologist, based on the lack of morphological clues, the broad differential diagnosis, and the interpretation of the numerous ancillary tests required to derive a definitive diagnosis. Olfactory neuroblastoma has a distinct clinical and radiological presentation and typically presents as a dumbbell-shaped mass straddling the cribriform plate and producing a tumour mass in the vault of the nasal cavity; providing the pathologist with this clinical information will facilitate a timely and accurate diagnosis.

Salivary gland neoplasms

Salivary gland neoplasms are diagnostically challenging as they are uncommon, few pathologists have the experience to confidently diagnose the full range encountered as there are more than 20 malignant entities to choose from! Furthermore, there is morphological diversity within the named entities and the classification is constantly evolving as molecular profiling defines new categories. Many of the entities have overlapping morphological features and therefore it is often not possible to provide a definitive diagnosis on cytology or core biopsy alone. A pragmatic approach for the beginner is to focus on the most common epithelial neoplasms (Table 68.1). For malignant salivary gland tumours, pathologists may provide a grade (low, intermediate, high), based on cytology and invasive characteristics. Mucoepidermoid carcinoma has a clinically validated grading system that includes assessment of the proportion of cystic change, neural invasion, necrosis, mitoses, and anaplasia. Stage is considered a better indicator of prognosis than grade. It is not unusual to diagnose a lymphoma, particularly marginal zone

Table 68.1 Common salivary gland neoplasms

	Features	Molecular profile
Malignant		
Mucoepidermoid carcinoma	Most common salivary gland malignancy. Solid/multicystic neoplasm composed of mucous cells, squamous (epidermoid) cells, and intermediate cells (Figure 68.6)	*MAML2* fusions
Acinic cell carcinoma	Wide spectrum of histological features but characterized by the presence of serous acinar cells and non-specific glandular cells. Solid, macrocytic and microcytic areas and papillary arrangements (Figure 68.7)	*NR4A3* rearrangements
Adenoid cystic carcinoma	Composed of basaloid cells, arranged in cribriform (Swiss cheese), tubular, and solid patterns (Figure 68.8). Neural invasion is a characteristic feature (Figure 68.9)	*MYB/MYBL1* rearrangements
Benign		
Pleomorphic adenoma	The most common salivary gland tumour. Bosselated mass composed of glandular and mesenchymal elements (so-called mixed tumour). There is often a characteristic chondro-myxoid stroma (Figure 68.10). Malignant change, carcinoma ex pleomorphic adenoma, is associated with a long-standing tumour that shows recent increase in size	*PLAG1, HMGA2* fusions
Basal cell adenoma	The tumour is encapsulated and composed of isomorphic basaloid cells arranged in tubular and trabeculae configurations	*CYLD, CTNNB1* mutations
Warthin tumour	Papillary cystic tumour. Papillae are covered in a double layer of columnar oncocytic (pink) cells. There is a dense lymphoid stroma with organization into follicles (Figures 68.11)	Not known

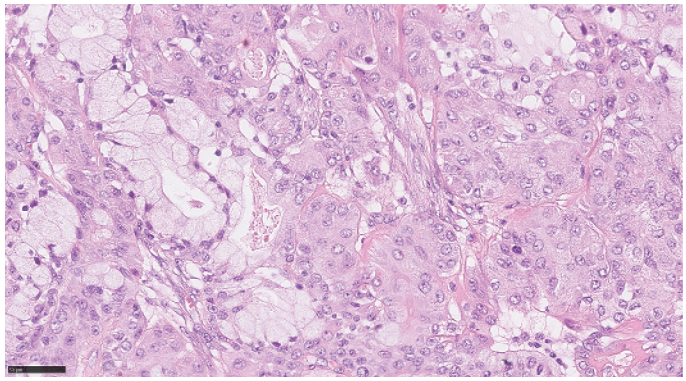

Figure 68.6 A mucoepidermoid carcinoma showing mucocytes (pale cells) and epidermoid cells (pink cells).

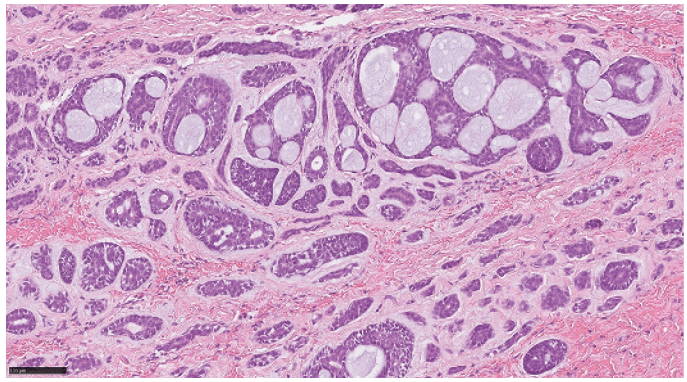

Figure 68.8 An adenoid cystic carcinoma with characteristic cribriform islands and tubular structures.

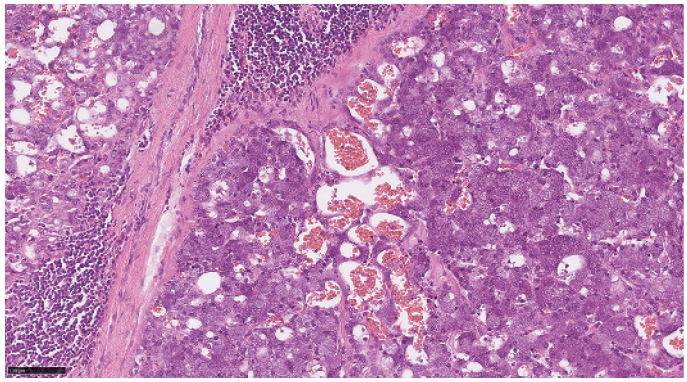

Figure 68.7 An acinic cell carcinoma composed of serous acinar cells (purple granular cells).

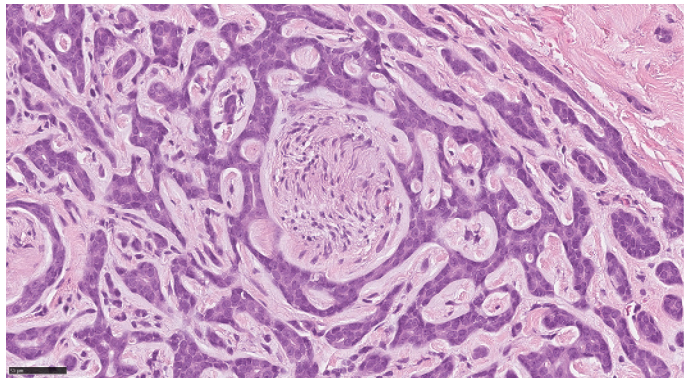

Figure 68.9 An adenoid cystic carcinoma showing neural invasion.

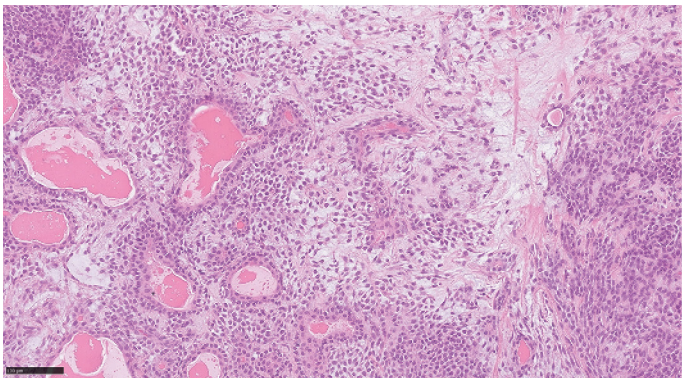

Figure 68.10 A pleomorphic adenoma composed of ductal structures and chrondro-myxoid matrix.

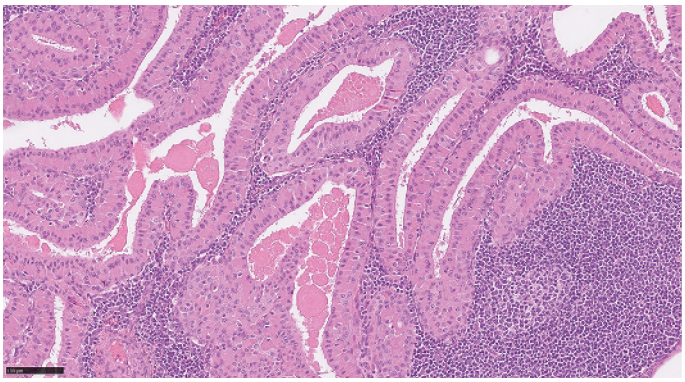

Figure 68.11 A Warthin tumour showing a papillary cystic architecture, oncocytic epithelium (pink cells), and lymphoid tissue (purple areas).

lymphoma, on a background of lymphocytic sialadenitis/Sjögren syndrome. Soft tissue tumours are rarely encountered in the major salivary glands. It is important to note that SCC or malignant melanoma in the parotid gland is likely to represent metastasis from a head and neck skin primary to an intra/para-parotid lymph node.

Tumours of the ear

The most common pathology specimens derived from the ear are usually a consequence of acute and chronic inflammation and pathologists tend to default to an inflammatory process at this site. If the ENT surgeon is suspicious of SCC, this should be clearly stated. Superficial disrupted fragments of atypical squamous epithelium are insufficient for a definitive diagnosis of SCC, but careful clinicopathological correlation may be sufficient to institute appropriate management. Problematic diagnoses can usually be resolved by harvesting additional tissue or by conservative excision. Other malignancies at the site are uncommon but include ceruminous adenocarcinoma and salivary gland-type neoplasms such as adenoid cystic carcinoma. Langerhans cell histiocytosis may present with temporal bone involvement and be part of multifocal eosinophilic granuloma.

It is important to make sure your pathologist knows that the clinical term 'malignant otitis externa' pertains to an inflammatory condition caused by *Pseudomonas aeruginosa*. The acute inflammation can cause a pseudo-epitheliomatous hyperplasia that mimics SCC. *Pseudomonas* infection requires microbiological testing to establish the diagnosis. By contrast, cholesteatoma is characterized by well-organized thin strips of hyperkeratotic epithelium producing abundant keratin debris and is rarely confused with SCC. Inflamed granulation tissue and the breakdown products of blood—haemosiderin deposits and cholesterol clefts—complete the histopathological features. Benign tumours unique to the ear include ceruminous adenoma and middle ear adenoma, the latter shows neuroendocrine differentiation. Paragangliomas also arise in the middle ear (glomus jugulare, glomus tympanicus), but are uncommon.

Metastatic carcinoma of unknown primary

Diagnosis of metastatic carcinoma of unknown primary location, most commonly presenting as cervical lymphadenopathy, initiates the search for the primary tumour. The pathologist uses panels of immunohistochemical markers to direct the search, relying on tumour markers that are specific for particular anatomical sites. For example, viral testing of metastatic SCC can be used to localize the search for the primary to the oropharynx (HPV) and nasopharynx (EBV). Metastatic adenocarcinoma may be from a salivary gland malignancy, but distant sites should also be considered particularly the common cancers: lung, colorectal, breast, and prostate.

Lymphomas

Lymphomas commonly present with cervical lymphadenopathy and may also affect extranodal sites such as Waldeyer's ring of lymphoid tissue. Hodgkin lymphoma is a nodal disease and typically affects young adults. Non-Hodgkin lymphomas (all the rest) are broadly classified as B- or T-cell lymphomas. B-cell lymphomas are most common and include small lymphocytic leukaemia/chronic lymphocytic leukaemia, follicular lymphoma, mantle cell lymphoma, diffuse large B-cell lymphoma, Burkitt lymphoma, and plasmablastic lymphoma, to name a few. The lymphocytic component of Sjögren syndrome may transform to marginal zone lymphoma, and low-grade tumours may transform to high-grade lymphomas. T-cell lymphomas are uncommon in the head and neck region, but nasal NK/T-cell lymphoma is an aggressive disease that typically causes mid-face destruction. The disease can be difficult to diagnose as the malignant cells are often masked by a dense inflammatory infiltrate. Clues to the diagnosis include accumulations of atypical lymphocytes around blood vessels (angiocentric) with endarteritis obliterans and necrosis. Persistent destructive nasal disease that can mimic lymphoma include granulomatosis polyangiitis and chronic cocaine abuse. The diagnosis of lymphoma requires consultation with a specialist lymphoreticular pathologist. The current lymphoma classification has a bewildering number of entities, many of which have specific molecular profiles. While ENT surgeons are the point of initial diagnosis, haematologists ultimately manage these patients and a timely referral is essential.

Mucosal malignant melanoma

Mucosal malignant melanoma is a rare tumour, the most common site in the head and neck region is the sinonasal tract. They typically

present with nasal obstruction and on clinical examination a brown/black polypoid mass. The malignant melanocytes are usually polygonal cells with large nuclei and prominent cherry red nucleoli. Mitotic figures are easily identified, and ulceration and necrosis are common. It is not always possible to demonstrate an *in situ* component or pre-existing nevus to pronounce a definite mucosal primary, in which case metastasis should also be considered but is unlikely. While the melanoma cells express similar tumour markers as cutaneous melanoma, immunohistochemistry for S100 and MelanA are positive; by contrast, nasal melanomas do not usually harbour BRAF V600 mutations and consequently are not eligible for vemurafenib treatment.

Malignant mesenchymal tumours

Malignant mesenchymal tumours, tagged with the suffix sarcoma, are uncommon in the head and neck region, but include the following: rhabdomyosarcoma (nasal cavity), chondrosarcoma (larynx), malignant peripheral nerve sheath tumour (neck), and fibrosarcoma, among others.

Conclusion

The ENT surgeon has the challenge of providing care for patients with functional disorders, an array of inflammatory conditions, both infective and non-infective, and a broad range of benign and malignant neoplastic lesions. While a working knowledge of the pathogenesis of common malignancies is essential, it is recognized that increasingly complex tumour classifications, the emergence of molecular diagnosis, and changing treatment paradigms require specialist head and neck surgical training, ultimately for the benefit of patients.

FURTHER READING

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69

Head and neck radiology

Derfel ap Dafydd

Introduction

As with most systems in modern medicine, advanced imaging plays a crucial role in the diagnosis and treatment planning of head and neck (H&N) pathology. The anatomical complexity of the neck, diversity of disease processes, and tumour types in this region and the need for multimodality analysis can make imaging interpretation of this region intimidating even to radiologists. Complete evaluation of certain processes, such as tumours of the upper aerodigestive tract, rely on a combination of both clinical assessment of the mucosal extent (e.g. with nasendoscopy) together with radiological assessment of the deep extent. A close working relationship between surgeon and radiologist is therefore vital. It is good practice for a surgeon to always review the cross-sectional imaging of their patients. A significant proportion of time in the H&N cancer multidisciplinary team (MDT) meeting is dedicated to discussion of cross-sectional imaging, which is pivotal to treatment decisions. The surgical members of the multidisciplinary team will need to make prompt assessments of resectability. The ability to read cross-sectional imaging (especially computed tomography (CT)) relating to their area of interest is therefore an essential skill for modern surgeons. The full spectrum of H&N imaging is not addressable in a single chapter; however, this chapter provides a general approach to the principal imaging modalities, outlines the imaging considerations of common and important neck pathologies (particularly malignancies), illustrates classic examples, and provides lists of important differential diagnoses.

Anatomy

The H&N is among the most complex anatomical regions and an understanding of cross-sectional anatomy is the most important element of imaging interpretation. Neck anatomy is conventionally divided into the suprahyoid and infrahyoid neck, and further divided by fascial planes into neck spaces (**Figure 69.1**). Localizing a pathological process to a particular neck space can significantly narrow the differential diagnosis, based on the known contents of the corresponding space and the common disease processes those structures are most prone to (**Table 69.1**). Displacement of adjacent neck spaces (e.g. the parapharyngeal space) by a mass may indicate the space of origin (**Figure 69.2**).

Modalities

H&N imaging is a prime example of a multimodality discipline, often requiring a combination of the main modalities: CT, magnetic resonance imaging (MRI), ultrasound (US), nuclear medicine (especially positron emission tomography (PET)/CT), and image-guided biopsy. Other supplementary modalities include plain radiography (largely superseded), orthopantomography for dental evaluation, fluoroscopy for ductal calculi (sialography), and water-soluble swallow for suspected pharyngeal pouch, fistulae, and for assessment to plan rehabilitation strategies.

The imaging modality of first choice will depend on many factors, including clinical urgency, lesion location, biology, patient factors (e.g. renal function, contrast allergy, metalwork, or claustrophobia), as well as local availability and expertise.

When reading any modality, a systematic approach is helpful; for example, beginning with the aerodigestive tract, followed by the cervical nodal stations, major salivary glands, thyroid, and bones. 'Review areas' (where pathology may be overlooked) include the imaged parts of the brain, globes, pterygopalatine fossae, fossae of Rosenmüller, retropharyngeal nodes, tongue, and lung apices. When assessing an abnormality, such as a mass, features to note include size, texture (solid versus cystic), enhancement, contour (smooth versus infiltrative), and local invasion. In general, it is preferable to image a potentially malignant process prior to biopsy. Analysis of post-biopsy imaging can be significantly hindered by artefact (e.g. inflammatory enhancement and haemorrhage).

Computed tomography

CT has become the dominant modality in emergency and oncological imaging. It is readily available, fast, high resolution, standardized, and familiar to both radiologists and clinicians. In the

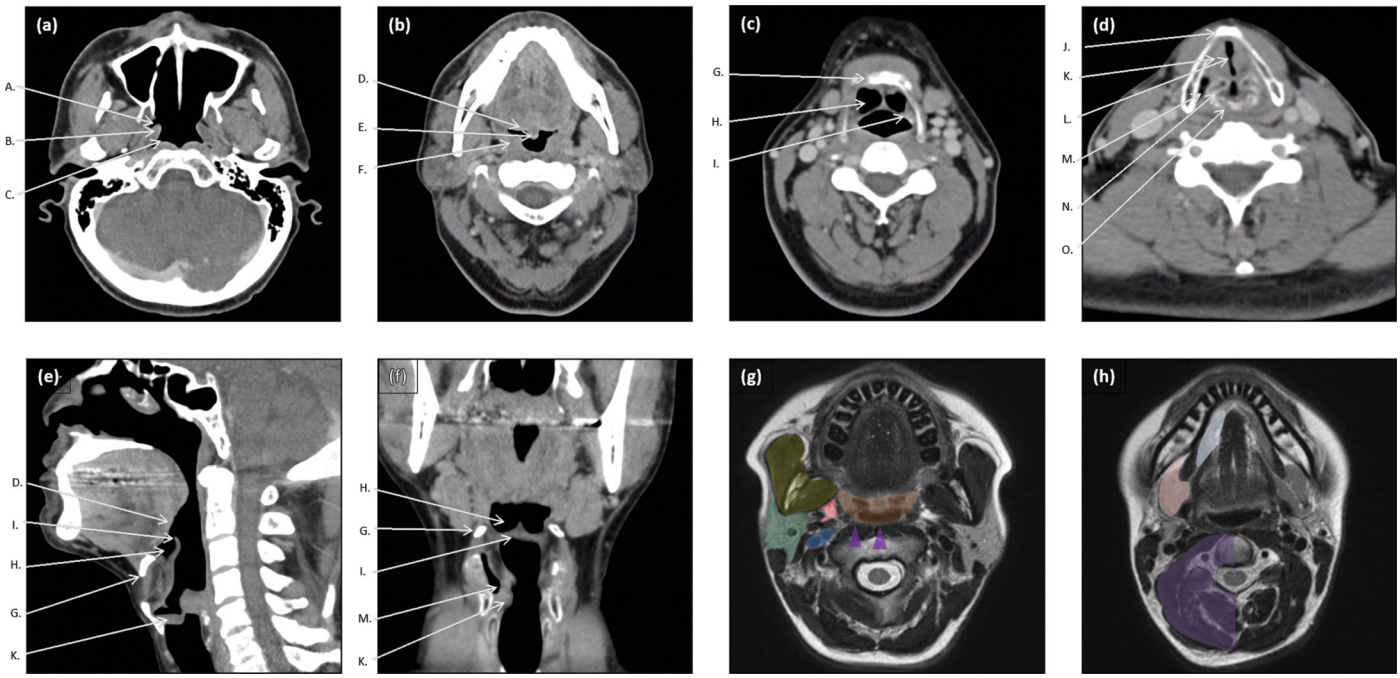

Figure 69.1 Cross-sectional anatomy of the upper aerodigestive tract and suprahyoid neck spaces. Axial contrast CT images at the level of the (a) nasopharynx, (b) oropharynx, (c) hyoid, and (d) glottis, and contrast CT images in the (e) sagittal and (f) coronal planes (A, eustachian tube orifice; B, torus tubarius; C, fossa of Rosenmüller; D, tongue base; E, uvula; F, palatine tonsil; G, hyoid bone; H, vallecula; I, epiglottis; J, thyroid cartilage; K, vocal cord; L, glottic airway; M, pyriform fossa). (g, h) Axial T2-weighted MRIs, depicting the suprahyoid neck spaces: the masticator space (yellow), parotid space (green), parapharyngeal space (red), carotid space (dark blue), pharyngeal mucosal space (orange), retropharyngeal space (purple arrowheads), sublingual space (light blue), submandibular space (pink), perivertebral space (purple fill).

Table 69.1 Suprahyoid neck spaces: differential diagnosis

Space	Malignant neoplasm	Benign neoplasm	Inflammatory	Congenital	Other
Parapharyngeal space	SCC (spread) Adenoid cystic carcinoma	Pleomorphic adenoma	Pharyngitis (spread)	Branchial cleft cyst	Carotid artery dissection/aneurysm
Pharyngeal mucosal space	SCC Lymphoma Minor salivary gland tumour (malignant)	Minor salivary gland tumour (benign)	Inflammatory masses	Tornwaldt's cyst	Asymmetric mucosa/ pseudomass
Masticator space	Metastasis SCC (spread) Sarcoma Lymphoma	Schwannoma	Odontogenic abscess/ osteomyelitis	Fibrous dysplasia	Accessory parotid tissue Denervation
Carotid space	Malignant lymph node	Schwannoma Paraganglioma	Thrombophlebitis	Branchial cleft cyst	
Parotid space	Mucoepidermoid carcinoma Adenoid cystic carcinoma Acinic cell carcinoma Lymphoma	Pleomorphic adenoma Warthin tumour Oncocytoma Facial schwannoma	Sialadenitis Abscess Intraparotid lymph node Benign lymphoepithelial cyst	First branchial cleft cyst Lymphangioma/ haemangioma	
Retropharyngeal space	Malignant lymph node SCC (spread) Lymphoma	Lymphangioma/ haemangioma Lipoma	Cellulitis/abscess Infected lymph node	Tortuous carotid artery	
Submandibular and sublingual spaces	Mucoepidermoid carcinoma Adenoid cystic carcinoma Acinic cell carcinoma Lymphoma	Pleomorphic adenoma Nerve sheath tumour	Sialadenitis/sialolithiasis Ludwig's angina Abscess	Ranula (+/– plunging)	
Perivertebral space	Bone metastasis Malignant lymph node Sarcoma Lymphoma	Schwannoma Chordoma Benign bone tumours	Osteomyelitis-discitis/ abscess	Lymphangioma	Vertebral artery dissection/aneurysm

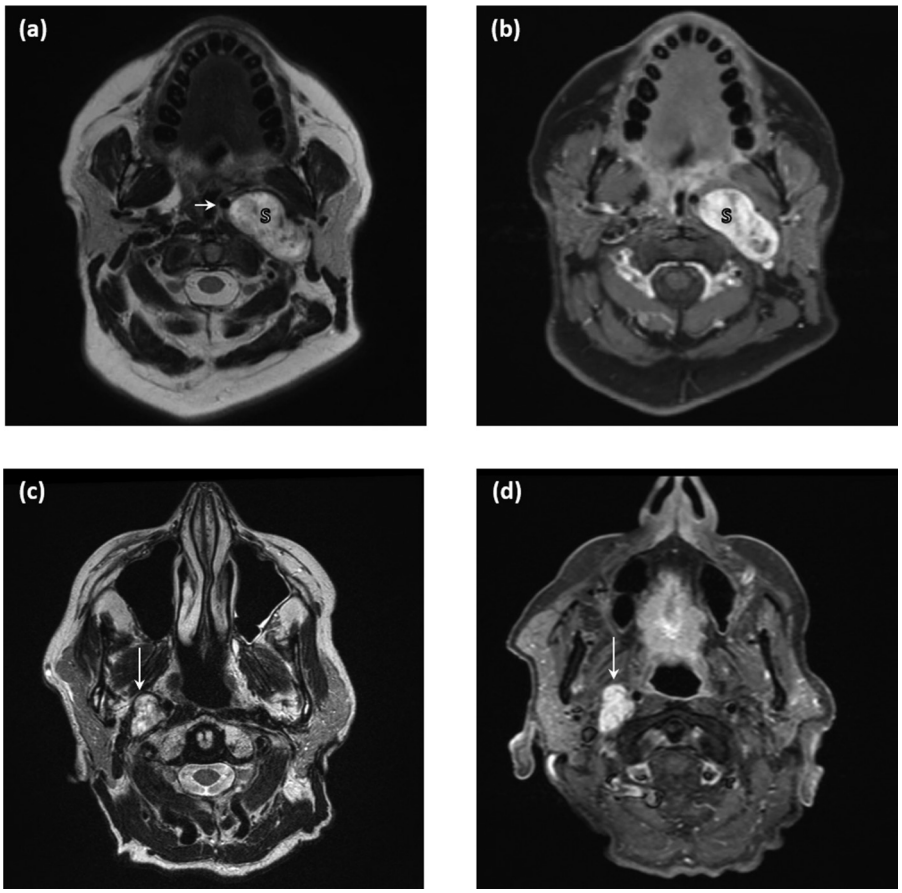

Figure 69.2 Two different patients with deep neck space masses. Patient 1: (a, b) axial T2-weighted and post-contrast fat-saturated T1-weighted images of a left vagal schwannoma (S) in the left carotid space. It exhibits heterogeneous, but predominantly hyperintense T2 signal and marked enhancement. The mass displaces the left internal carotid artery medially (small arrow), the parapharyngeal space anteriorly and parotid space laterally. Patient 2: (c, d) axial T2-weighted and post-contrast, fat-saturated T1-weighted images showing a right glomus vagale in the right carotid space (long arrow). It is predominantly T2 hyperintense, with linear, isointense internal flow voids and exhibits homogeneous enhancement. The mass displaces the internal carotid artery medially and parapharyngeal space anteriorly.

H&N, CT is well suited to imaging the temporal bones, infrahyoid neck, vascular complications, and cortical bone erosion and for completion of staging, response evaluation, and post-treatment surveillance. CT is reasonably good at imaging oropharyngeal and nasopharyngeal masses. However, oral cavity processes are often poorly depicted and often obscured by metal artefact from dental amalgam. Other disadvantages include the use of ionizing radiation and the risks of iodinate contrast in those with allergy and renal impairment. Iodinated contrast is also contraindicated in hyperthyroidism and prior to radioactive iodine therapy.

Magnetic resonance imaging

MRI exploits the varying responses of different tissues (e.g. fat and muscle) to changes in magnetic fields. Although spatial resolution is lower than CT, MRI produces superior soft tissue contrast. The oral cavity is much better assessed with MRI than CT. The oropharynx and rest of the suprahyoid neck are better assessed with MRI than CT, but either will suffice, depending on local practices. Aggressive sinonasal and orbitofacial tumours are best assessed with MRI, as orbital invasion, marrow infiltration, and intracranial invasion are better appreciated. Perineural spread is much better depicted on MRI than any other modality. MRI is, however, potentially hazardous due to the magnetic field, is time-consuming, less available, less standardized, prone to metal- and gas-related artefacts, and to patient-related factors (e.g. claustrophobia).

A standard H&N MRI protocol would include at least T1-weighted and T2-weighted (anatomical) sequences usually followed by diffusion-weighted imaging (an assessment of cellularity) and post-contrast (gadolinium enhanced) sequences. On T1-weighted imaging, fluid (water) is isointense (grey), fat is hyperintense (white), and muscle is isointense. On T2-weighted imaging, fluid (water) is hyperintense (bright), fat is hyperintense, and muscle is isointense. Most pathological processes are either T2 isointense or hyperintense and T1 isointense. Certain processes, however, are T1 hyperintense, such as blood, fat, and melanin. The axial T2 and axial, fat-saturated, post-contrast T1 are good sequences to obtain an initial overview of the neck.

Ultrasound

This technique relies on high-frequency sound waves which are reflected to varying degrees by different tissue types. It is safe, quick, cheap, and readily available. It offers high-resolution imaging of superficial structures and is therefore well suited to neck imaging. It is excellent in assessing palpable neck masses, vessels, thyroid,

salivary glands (e.g. for stones, inflammation, abscesses, or tumours), nodes, and (in some centres) intraoral US of the oral cavity.

On US, fluid (water) is anechoic (black), muscle is isoechoic (grey), fat is hyperechoic (white), and tumour is variable, but typically isoechoic (grey). Doppler can be used to assess the vascularity/solidity of a mass. Abnormalities found on US can then be subjected to US-guided sampling (fine needle aspiration or biopsy) as appropriate. If a biopsy is anticipated, the referring clinician should arrange a clotting screen beforehand. Air, bone, and metal obliterate the US beam and produce US artefacts. Assessment of deep structures is limited, and US is an operator-dependent technique.

Nuclear medicine

Nuclear medicine is a functional imaging specialty, involving the injection of a radioactive tracer into the patient. The tracer accumulates at regions of interest and the radiation emitted by the patient is detected and displayed typically as a heat map.

PET/CT has a key role in oncological imaging. A radiotracer (commonly ^{18}F-fluorodeoxyglucose (FDG)) is injected intravenously and its accumulation at sites of high metabolic activity (normal and pathological) are measured in 'standard uptake values' on a low-resolution colour map. Its principal applications in the H&N are investigation of carcinoma unknown primary (e.g. malignant neck nodes with no obvious primary), improved nodal staging, and in completion of staging and post-treatment surveillance in selected cases (see Chapter 56). Its disadvantages are that it is time-consuming, a high radiation dose, expensive, less available, and offers low anatomical resolution. It is also prone to false positives from brown fat, reactive nodes, asymmetrical uptake in benign tonsils and the larynx, benign parotid, and thyroid nodules, and post-treatment inflammatory uptake.

Other nuclear medicine techniques in the H&N include radioactive iodine in the treatment of thyroid cancer and ^{99m}Tc sestamibi for parathyroid adenoma localization.

Skull base and sinonasal disease

Surgical planning for functional endoscopic sinus surgery is best achieved with CT. The presence of bony sclerosis suggests chronicity. Fungal sinus disease tends to produce dense secretions.

Assessment of suspected sinonasal tumours is optimized with MRI. Invasion of local anatomy suggests malignancy. Important management considerations are orbital invasion and intracranial extension (**Figure 69.3**). Sinonasal tumours invading the

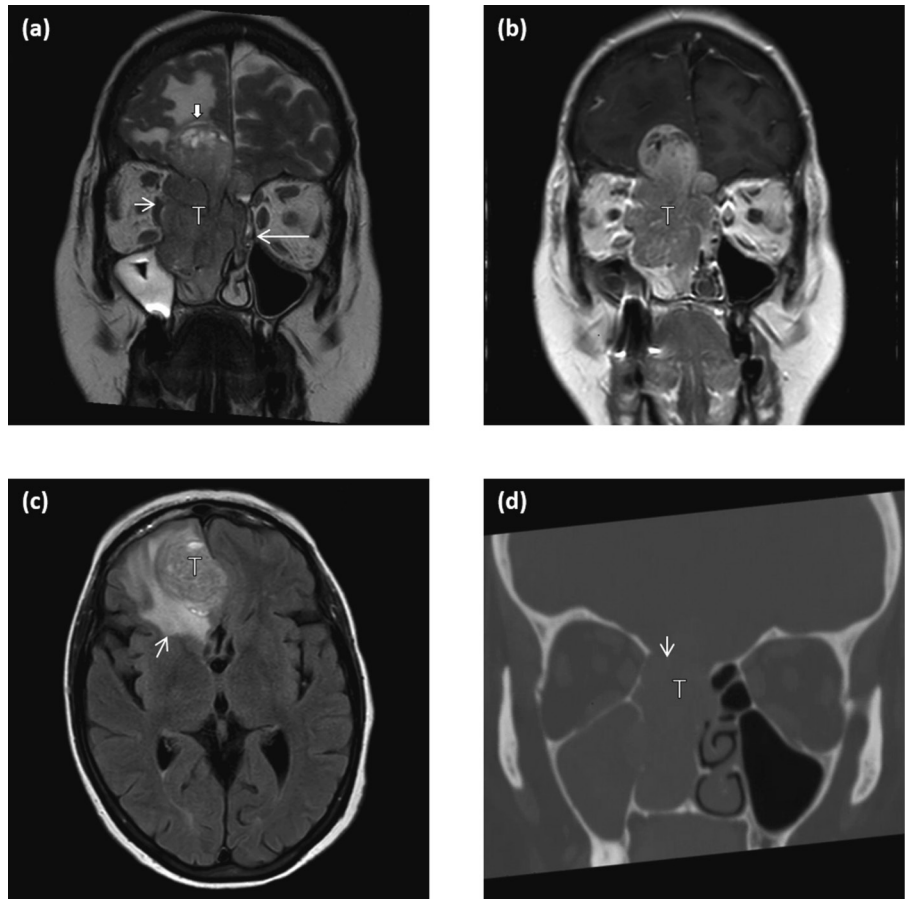

Figure 69.3 A 78-year-old woman with an olfactory neuroblastoma (esthesioneuroblastoma). (a) Coronal T2-weighted image, showing a large, aggressive, heterogeneous anterior skull base tumour (T), filling the right nasal cavity, invading into the anterior cranial fossa and parenchyma of the right frontal lobe (thick arrow) and with gross invasion of the right orbit. Note the loss of the T2 hypointense line of the right periorbita (short arrow), in contrast to the normal left periorbita (long arrow). (b) Corresponding post-contrast T1-weighted image, showing marked, heterogeneous enhancement. (c) Axial FLAIR image through the brain, showing parenchymal invasion (T) and surrounding vasogenic oedema (short arrow). (d) Coronal CT image on bone window, demonstrating more obvious cortical destruction of the cribriform plate (short arrow).

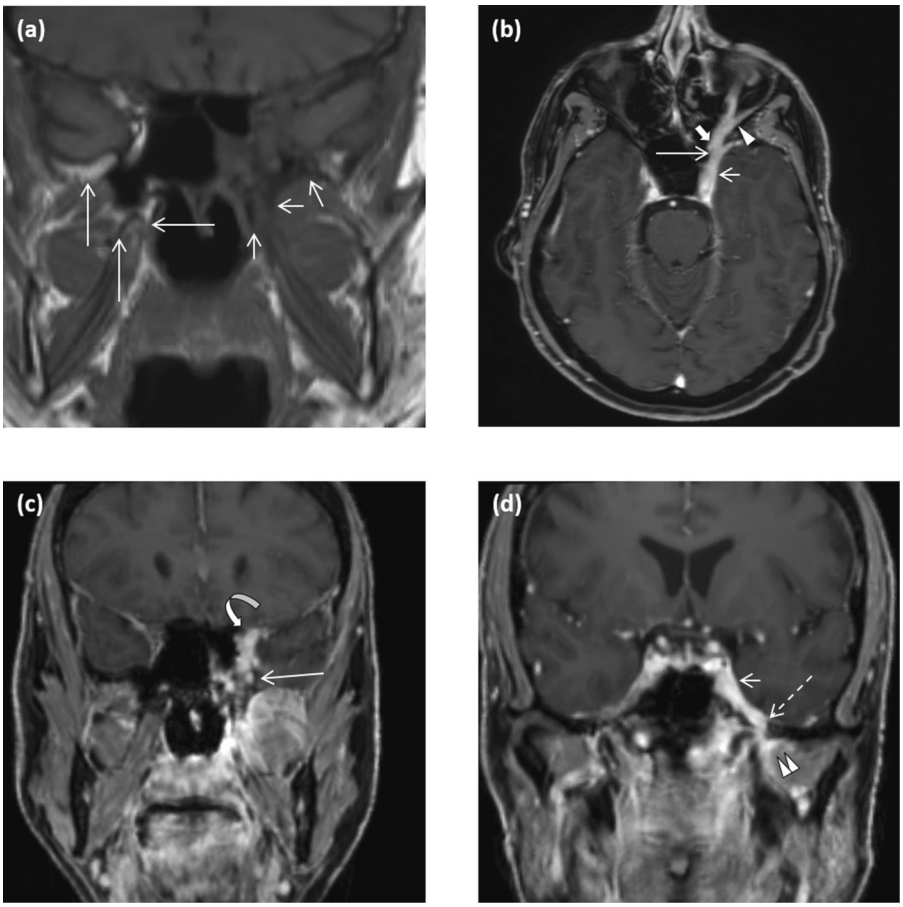

Figure 69.4 A 54-year-old man with an adenoid cystic carcinoma of the left maxilla and intracranial perineural spread. (a) Coronal T1-weighted image showing abnormal, low T1 signal in the left pterygoid plates and greater wing of sphenoid, in contrast with the normal high T1 marrow signal (fat) on the right (long arrows). (b) Axial and (c, d) coronal post-contrast, fat-saturated images, showing abnormal perineural thickening and enhancement (perineural spread) in the left cavernous sinus (short arrow), foramen rotundum (long arrow), pterygopalatine fossa (thick arrow), infraorbital foramen (single arrowhead), orbital apex (curved arrow), foramen ovale (dashed arrow), and infratemporal fossa (double arrowhead).

medial orbital wall may necessitate orbital exenteration if the periorbital (periosteum) is involved. MRI is most reliable in assessing the periorbita, which appears as a thin black line on the coronal T2. Any threat to the orbital apex should be noted, as this may lead to loss of vision. Intracranial invasion may preclude resection, particularly if there is dural invasion or parenchymal brain invasion. Perineural spread (e.g. in adenoid cystic or mucoepidermoid carcinomas) is best appreciated as thickened, enhancing nerves on post-contrast, fat saturated, T1-weighted sequences (Figure 69.4). Lists of differential diagnoses are provided in Tables 69.2 and 69.3.

Pharynx

Oral cavity and oropharynx

Cancers of the oral cavity and particularly the oropharynx are among the commoner malignancies encountered at the H&N multidisciplinary team meeting. The vast majority are squamous cell carcinomas (SCCs), with a minority being other cell types, such as minor salivary gland tumours.

Tumours of the oral tongue and buccal mucosa can be occult on conventional imaging. Local staging often relies principally on clinical assessment, with imaging used for regional and distant staging. Radiological interpretation should also allow for potential superimposed biopsy-related inflammation, which may have been performed in outpatients prior to imaging (Figure 69.5). If assessable on imaging (e.g. MRI or intraoral US), an oral tongue primary should be described in terms of anterior, middle, and posterior thirds of the oral tongue, depth of invasion, and local extent.

Retromolar trigone tumours may spread in several directions: laterally to the buccal mucosa; medially to the glossotonsillar sulcus/tonsillar pillars; posteriorly into the mandible; and upwards or downwards into the buccogingival sulci.

Oropharyngeal SCC is often discovered following the investigation of a malignant neck node (Figure 69.6). Incidental tonsillar enlargement (symmetrical or asymmetrical) either on anatomical imaging or PET/CT should be interpreted with caution, as the majority of these will be false positives. The prognosis of oropharyngeal SCC depends on the p16 status (found in human papillomavirus-associated SCC). Although not reliably distinguishable with current

Table 69.2 Skull base and temporal bone: differential diagnosis

Site	Malignant neoplasm	Benign neoplasm	Inflammatory	Congenital	Other
Anterior skull base	Esthesioneuroblastoma Sinonasal tumour	Meningioma*	Mucocele	Anterior neuropore anomalies (neonates)	
Central skull base	Craniopharyngioma PNS from nasopharyngeal SCC Metastasis (osseous/meningeal)* Lymphoma/leukaemia* Plasmacytoma* Chondrosarcoma* Haemangiopericytoma* Histiocytosis*	Pituitary macroadenoma Meningioma* Chordoma Nerve sheath tumour*	Cavernous sinus thrombosis Osteomyelitis* Sarcoid*	Fibrous dysplasia* Ecchordosis physaliphora	Carotid aneurysm Carotid-cavernous sinus fistula
Posterior skull base and cerebellopontine angle	Metastasis*	Paraganglioma (glomus jugulare) Vestibular schwannoma/neurofibroma Meningioma*	Osteomyelitis* Sarcoid*	Arachnoid cyst Epidermoid Lipoma	Posterior circulation aneurysm
Temporal bone	SCC (external auditory canal or middle ear) PNS from parotid tumour Endolymphatic sac tumour	Paraganglioma (glomus tympanicum) Osteoma	Cholesteatoma Necrotizing otitis externa Acute mastoiditis AOM/COM Exostosis Otosclerosis Cholesterol granuloma Labyrinthitis Apical petrositis Keratosis obturans	Congenital anomalies of the cochlea and labyrinth External auditory canal atresia Haemangioma/vascular malformations	Temporal bone fracture Ossicular dislocation Vascular anomalies (e.g. persistent stapedial artery, aberrant internal carotid artery)

*Can affect any part of the skull base. AOM/COM, acute otomastoiditis/chronic otomastoiditis; PNS, perineural spread.

techniques, p16-positive disease tends to produce large volume, cystic nodes. Assessment of the primary should focus on features that would influence staging and management, such as involvement of the epiglottis, extrinsic tongue muscles, bone, and carotid artery (Figure 69.7).

Nasopharynx

Nasopharyngeal SCCs may underlie a presentation of conductive hearing loss and middle ear effusion in an adult. An incidentally detected middle ear effusion on cross-sectional imaging (e.g. an MRI brain) should therefore prompt radiological examination of the nasopharynx/fossa of Rosenmüller for an associated tumour (Figure 69.8). It is typically a locally aggressive cancer, with a propensity to invade the skull base and extend intracranially through foramen lacerum causing cranial nerve palsies. Nodal disease at presentation is detectable in 75% of cases. Nasopharyngeal carcinoma is therefore generally a non-surgical disease with its unique staging system.

Hypopharynx and larynx

The supraglottis, hypopharynx, and larynx are generally best assessed with CT (Figure 69.9). Tumours in this region can be very subtle/occult on conventional imaging, for example, early vocal cord lesions, particularly when imaged with the cords adducted. Again, evaluation is best concentrated on features that upstage the tumour. Certain staging and management-dependent features can be challenging to assess. Paraglottic fat and thyroid cartilage involvement can be overestimated, particularly after recent biopsy, where superimposed inflammatory change can exaggerate tumour extent. Visible erosion of the cartilage is generally reliably predictive. However, this can be complicated by reactive sclerosis on CT scanning or reactive oedema on MRI and by variable or asymmetric ossification of the cartilage. Visible soft tumour beyond the outer surface of the cartilage should reliably predict extralaryngeal invasion. The presence of subglottic extension can be determined if the distance from the laryngeal ventricle to the lower extent of the tumour exceeds 10 mm. The UK 2016 National Institute for Health and Care

Table 69.3 Sinonasal processes: differential diagnosis

Malignant neoplasm	Benign neoplasm	Inflammatory	Congenital
SCC Adenocarcinoma Esthesioneuroblastoma Sinonasal undifferentiated carcinoma Melanoma Lymphoma Osteosarcoma	Inverted papilloma Juvenile angiofibroma osteoma	Sinusitis (acute/chronic/fungal) Sinonasal polyposis Antrochoanal polyp Mucocoele Connective tissue disease	Nasal glioma Nasal encephalocele Fibrous dysplasia Choanal atresia Pyriform aperture stenosis

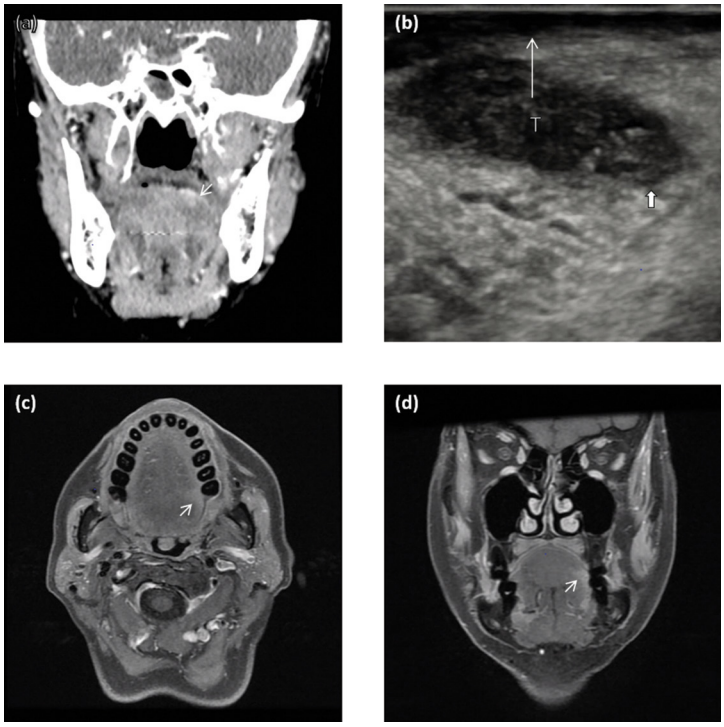

Figure 69.5 Two different patients with oral cavity lesions. Patient 1: (a, b) coronal contrast CT and intraoral US showing a small SCC in the posterior third of the left lateral oral tongue. It is subtle on CT (short arrow). It is well depicted in intraoral US (T), where the mucosal surface (long arrow) and deepest margin of the tumour (thick arrow) are appreciable. Patient 2: (c, d) axial and coronal post-contrast, fat-saturated T1-weighted images of a different patient with carcinoma *in situ* in the mid-third of the left lateral oral tongue (small arrow). The small focus of abnormal enhancement is easily mistaken for an invasive SCC. However, this was a post-biopsy MRI, leading to potential overestimation of the lesion due to super-imposed inflammation. Biopsy revealed carcinoma *in situ* only.

Figure 69.6 A 67-year-old man with left tonsillar SCC and left-sided nodal disease, presenting with a left neck mass. (a) US image, showing an overtly malignant left level II node (short arrow), with central necrosis (long arrow). (b) Axial contrast CT, showing the malignant node (short arrow), but no obvious primary tumour. (c) PET/CT showing the FDG-avid node (short arrow) and primary tonsillar cancer (long arrow).

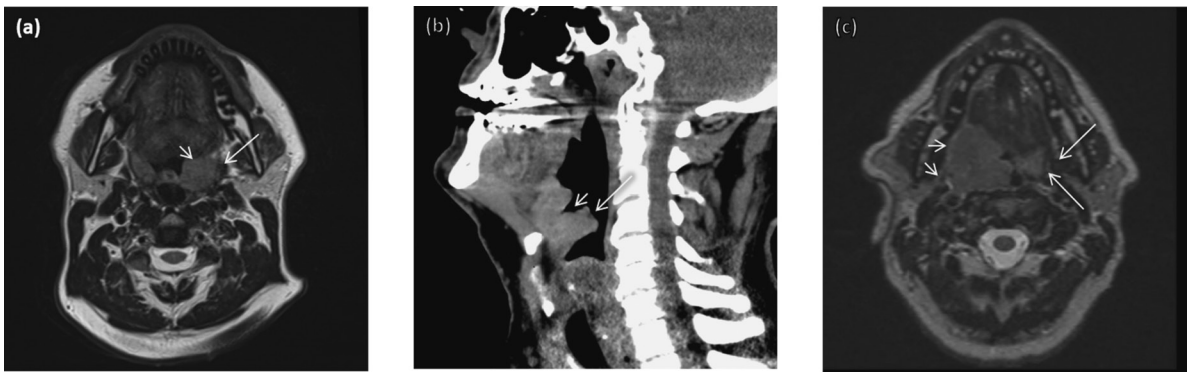

Figure 69.7 Three different patients with oropharyngeal SCC. Patient 1: (a) axial T2-weighted image, showing a (TNM) T2 tumour of the left palatine tonsil (small arrow). The darker fibres of the pharyngeal constrictors and left styloglossus muscle are intact (long arrow). Patient 2: (b) a different patient with a (TNM) T3 tumour of the tongue base, involving the vallecula (small arrow) and lingual surface of the epiglottis (long arrow). Patient 3: (c) another patient with a (TNM) T4 tumour of the right palatine tonsil; the low-

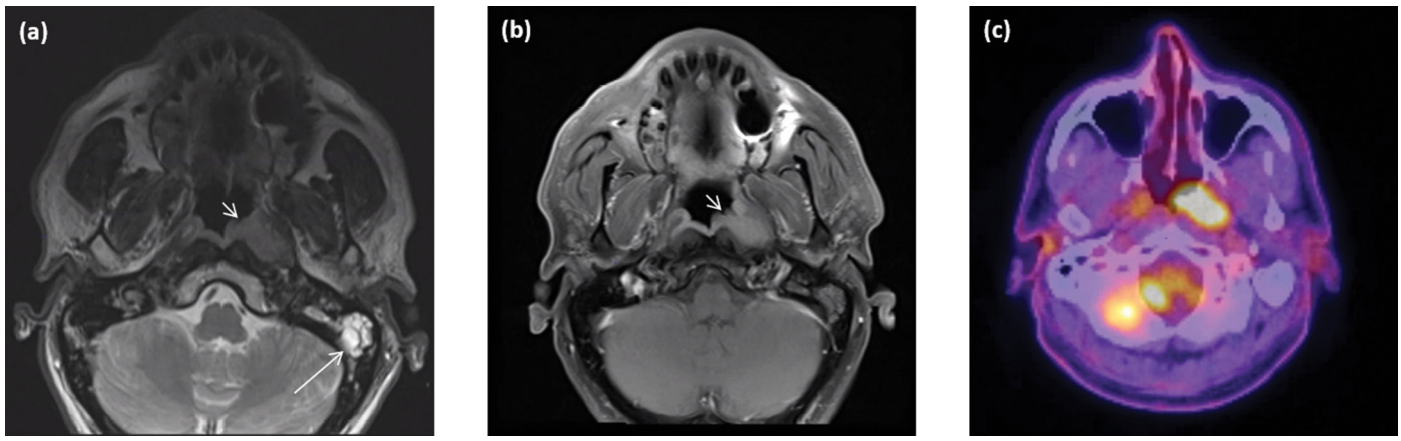

Figure 69.8 A 57-year-old man with a left nasopharyngeal SCC. (a) Axial T2-weighted and (b) post-contrast, fat-saturated T1-weighted images, showing a small, T2 isointense, enhancing mass filling the left fossa of Rosenmüller (short arrow), with secretions in the left mastoid air cells (long arrow) due to occlusion of the eustachian tube orifice. (c) Corresponding PET/CT scan, showing intense FDG accumulation in the tumour.

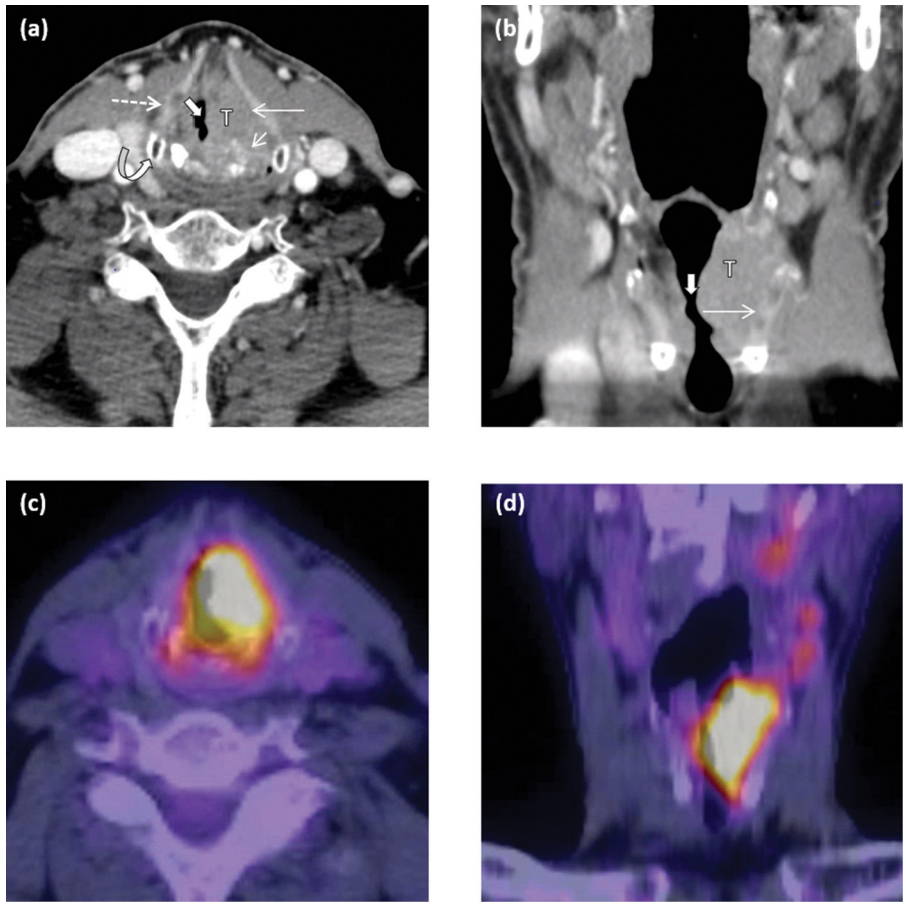

Figure 69.9 A 53-year-old man with laryngeal SCC. (a, b) Axial and coronal contrast CT images, showing a left glottic and supraglottic tumour (T), invading the left arytenoid cartilage (short arrow) and paraglottic fat up to the inner surface of the thyroid cartilage (long arrow) and narrowing the glottic airway (thick arrow). The thyroid cartilage is variably ossified, with an ossified superior cornu (curved arrow) and non-ossified lamina, making assessment of invasion difficult, but in this case the cartilage is spared. (c, d) Corresponding PET/CT images showing intense FDG accumulation.

Excellence guidelines for management of H&N malignancies recommend PET/CT for T4 hypopharyngeal cancer given the high propensity for metastatic disease and second primaries.

Major salivary glands

Considerable overlap exists between the imaging features of benign and malignant solid masses of the major salivary glands. US and US-guided sampling are the mainstay. Other imaging modalities may be additive where malignancy is suspected or confirmed (Figure 69.10). Although certain clinical or imaging features may suggest either a benign or malignant major salivary gland mass, not infrequently, definitive characterization may not be possible on imaging. Unfavourable features on any modality include an infiltrative front, invasion of local anatomy (including skin or bone), perineural spread, or associated pathological nodes or metastases. In general, a cytopathological diagnosis is advisable. PET/CT is not reliable at distinguishing benign and malignant salivary gland mases, as pleomorphic adenomas and Warthin tumours can both be FDG avid. The 80% rule of major salivary gland masses is helpful (see Chapter 58): 80% of salivary gland tumours are in the parotid; 80% of parotid gland tumours are benign; 80% of benign tumours in the parotid are pleomorphic adenomas; and 80% of pleomorphic adenomas remain benign. On imaging, the retromandibular vein can be used as a surrogate marker of the facial nerve and arbitrary boundary between the deep and superficial lobes, with obvious implications on surgical planning.

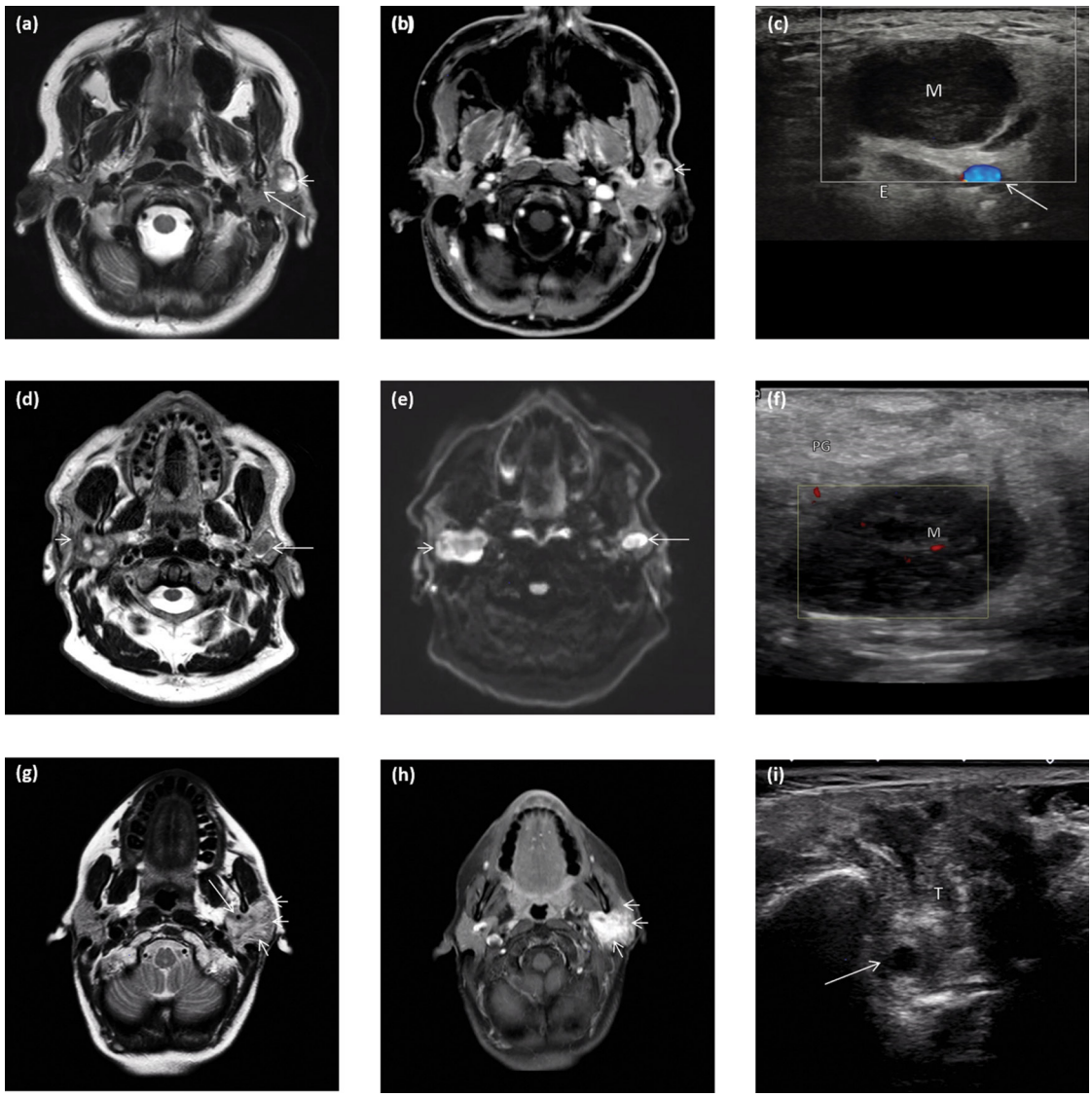

Figure 69.10 Three different patients with parotid lesions. Patient 1: (a, b, c) axial T2-weighted and post-contrast, fat-saturated MRIs and an US scan of a left parotid pleomorphic adenoma. It shows a mostly T2 hyperintense and enhancing mass in the superficial left parotid gland (short arrows), and on US an isoechoic, hypovascular mass (M) with posterior acoustic enhancement (E). Note the retromandibular vein (long arrows). Patient 2: (e, f, g) axial T2-weighted and diffusion-weighted MRIs and an US scan of a different patient with bilateral Warthin tumours (short and long arrows). The masses are solid cystic, with T2 hypointense elements, restricted diffusion (cellular) and areas of vascularity on Doppler (M). Patient 3: (h, i, j) axial T2-weighted and post-contrast, fat-saturated T1-weighted MRIs and an US scan of a left parotid adenoid cystic carcinoma (T). It exhibits an infiltrative front (short arrows), avid enhancement and extends beyond the retromandibular vein (long arrows) into the deep lobe.

Classically, pleomorphic adenomas are well-defined, lobulated masses, mostly found in the superficial parotid gland. They appear hypoechoic on US (with posterior acoustic enhancement), and enhancing, variably necrotic, and variably calcified on CT. On MRI they appear T1 hypointense and T2 hyperintense, occasionally with a thin T2 hypointense pseudocapsule and pathological enhancement.

Warthin tumours present as well-defined masses, most commonly in older men. They may be multiple or bilateral in 20% of cases and are the commonest cause of multiple parotid masses. They are typically found in the tail of the parotid gland and classically exhibit heterogeneous, solid-cystic texture and moderate enhancement.

Perineural spread may be seen with mucoepidermoid and adenoid cystic tumours, necessitating careful examination of the course of the facial nerve (e.g. the stylomastoid foramen).

Thyroid

In most UK centres, sonographically detectable thyroid nodules are reported according to the British Thyroid Association Ultrasound 'U' classification system (Table 69.4). In general, U1 and U2 nodules can be discharged and U3, U4, and U5 nodules require fine needle aspiration (Figure 69.11). An alternative US grading system is the American College of Radiology Thyroid Imaging, Reporting and Data System (TIRADS), where a score (0–3) is assigned to five sonographic features (composition, echogenicity, shape, margin, and echogenic foci) and the total score determines the risk level from

Table 69.4 British Thyroid Association: ultrasound 'U' classification of thyroid nodules

U1 (normal) • No nodule	**U4 (suspicious)** • Solid hypoechoic/very hypoechoic • Hypoechoic with disrupted peripheral calcification • Lobulated outline
U2 (benign) • Hyperechoic or isoechoic with halo • Cystic spaces + ring-down artefact (colloid) • Microcystic/spongiform • Peripheral egg-shell calcification • Peripheral vascularity	**U5 (malignant)** • Solid hypoechoic with lobulated/irregular outline and microcalcification (papillary thyroid cancer) • Solid hypoechoic with lobulated/irregular outline and globular calcification (medullary thyroid cancer) • Intranodular vascularity • Taller than wide axially • Associated lymphadenopathy
U3 (indeterminate) • Markedly hyperechoic nodule with halo (follicular lesion) • Hypoechoic + equivocal echogenic foci or cystic change • Mixed/central vascularity	

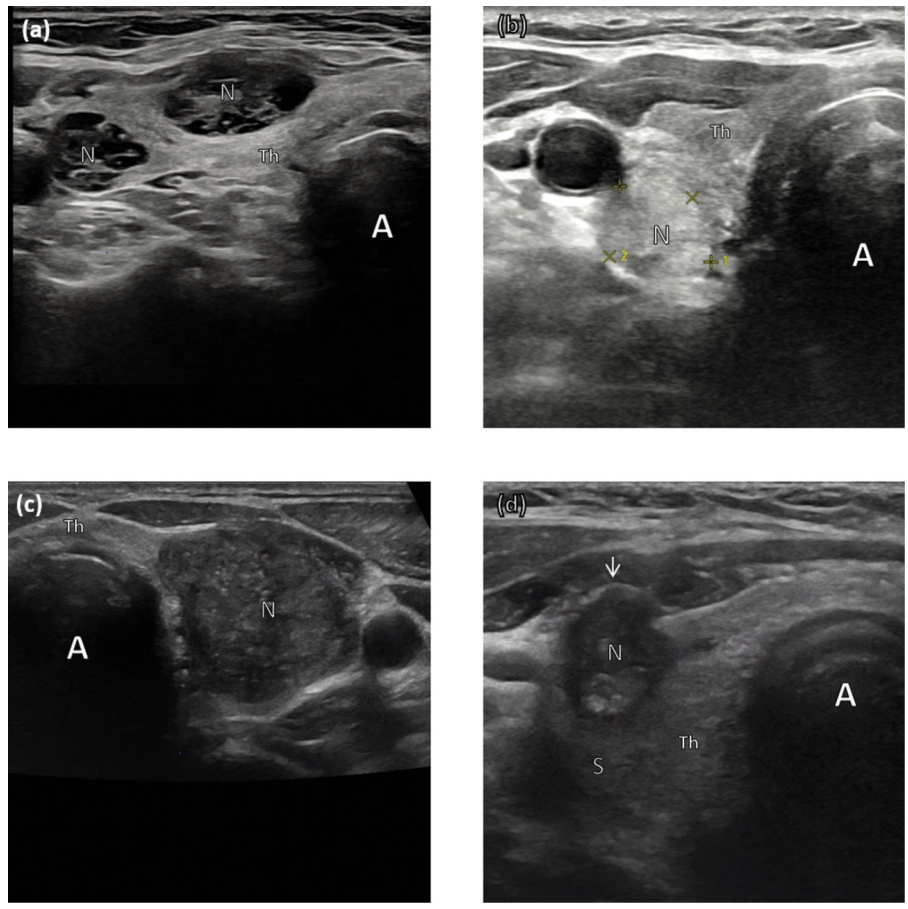

Figure 69.11 Ultrasound images of four different patients with nodules (N) in the thyroid (Th), which can be assigned a U-grading according to the British Thyroid Association. (a) Two benign 'U2' right thyroid nodules, with small cystic (spongiform) spaces. Note the relative position of the airway (A). (b) An indeterminate 'U3' right thyroid nodule, which is hyperechoic and potentially a follicular lesion. (c) A likely malignant 'U4' left thyroid nodule, which is relatively hypoechoic and slightly irregular. (d) An overtly malignant 'U5' right thyroid nodule, which is irregular, with coarse calcification, posterior acoustic shadowing (S) and anterior extrathyroidal extension (small arrow).

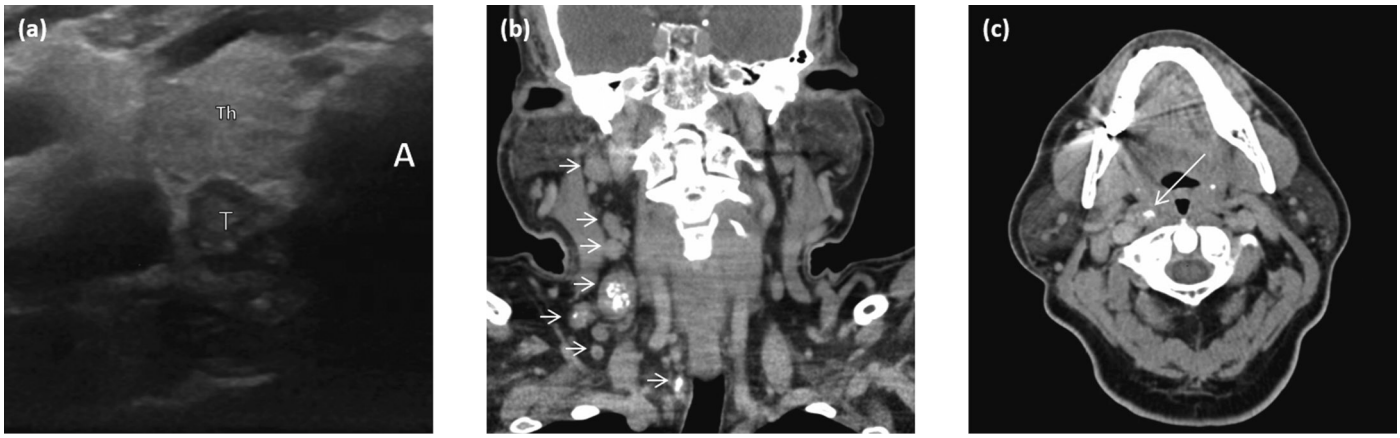

Figure 69.12 A 57-year-old man with papillary thyroid cancer and cervical nodal disease. (a) US image of the primary tumour (T) in the right lobe of thyroid (Th). (b) Coronal and (c) axial contrast CT images, showing multiple, partially calcified malignant nodes in the right lateral neck (short arrows) and a malignant right retropharyngeal node (long arrow). Identification of the involved retropharyngeal node necessitated a more extensive nodal dissection to avoid residual macroscopic disease.

TR1 (benign) or TR5 (highly suspicious). Also assessable on US are extrathyroidal extension, retrosternal extension, and tracheal deviation. A well-defined, solid, echogenic nodule may be a follicular lesion. It is not possible on US, fine needle aspiration, or core biopsy to distinguish a follicular adenoma from a well-differentiated follicular carcinoma. Hemithyroidectomy is usually indicated for definitive characterization.

Intravenous iodinated contrast (for CT) is contraindicated in hyperthyroidism and should be avoided in staging a confirmed thyroid cancer, as it may take weeks for the iodine absorbed by the thyroid to clear and this can compromise the effectiveness of radioactive I^{131} treatment (radioiodine should be delayed 6–8 weeks after intravenous iodinated contrast). MRI neck is preferred in this context. A non-contrast CT chest is recommended for excluding lung metastases where appropriate.

It should be borne in mind when staging thyroid cancer that papillary thyroid cancer has a propensity to spread to locoregional nodes and follicular thyroid cancer tends to spread to the lungs and bones. Medullary thyroid cancer may be sporadic or part of a multiple endocrine neoplasia syndrome, therefore other genetic manifestations should be noted (e.g. pituitary mass, parathyroid mass, pancreatic mass, or phaeochromocytoma). They can produce arterially enhancing metastases; hence, an arterial phase CT of the liver is advisable. A large, locally advanced thyroid tumour in an older patient suggests a possible anaplastic thyroid cancer. Localized disease may be treatable surgically. Radioactive iodine has a role in advanced or metastatic disease.

Lymph nodes and cystic neck masses

Abnormal nodes may be reactive (e.g. due to viral illness, poor dentition, or pharyngeal inflammation) or involved by malignancy (e.g. SCC or lymphoproliferative disease), tuberculosis, glandular fever, HIV, or sarcoid.

Normal lymph nodes vary in size but are typically less than 10 mm in short axis and are smooth, ovoid, symmetrical, with a fatty hilum. Although a 10 mm size threshold is often quoted in nodal staging, it should be noted that 67–71% of malignant SCC nodes are smaller than this and reactive nodes (even in patients with confirmed cancer) may be as large as 20 mm. Other adverse nodal features include roundness, low-echotexture on US, irregularity, asymmetry, loss of fatty hilum on US, and abnormal vascularity on Doppler. A node exhibiting complex cystic/necrotic areas or features of extracapsular extension may be regarded as overtly malignant in the context of a known primary. In the setting of a histologically proven neck primary, it may be appropriate to sample indeterminate nodes. During nodal staging, particular review areas include the central compartment, periauricular nodes, suboccipital nodes, retropharyngeal nodes, and the upper mediastinum (**Figure 69.12**).

An infected branchial cleft cyst may be indistinguishable from a cystic SCC node on imaging and may require further investigation (**Figure 69.13**). **Box 69.1** lists other causes of cystic neck masses.

Conclusion

Additional detail on specific areas can be obtained from the list of further reading; however, this chapter outlines general principles, key aspects, together with pearls and pitfalls in H&N imaging for the general ear, nose, and throat surgeon.

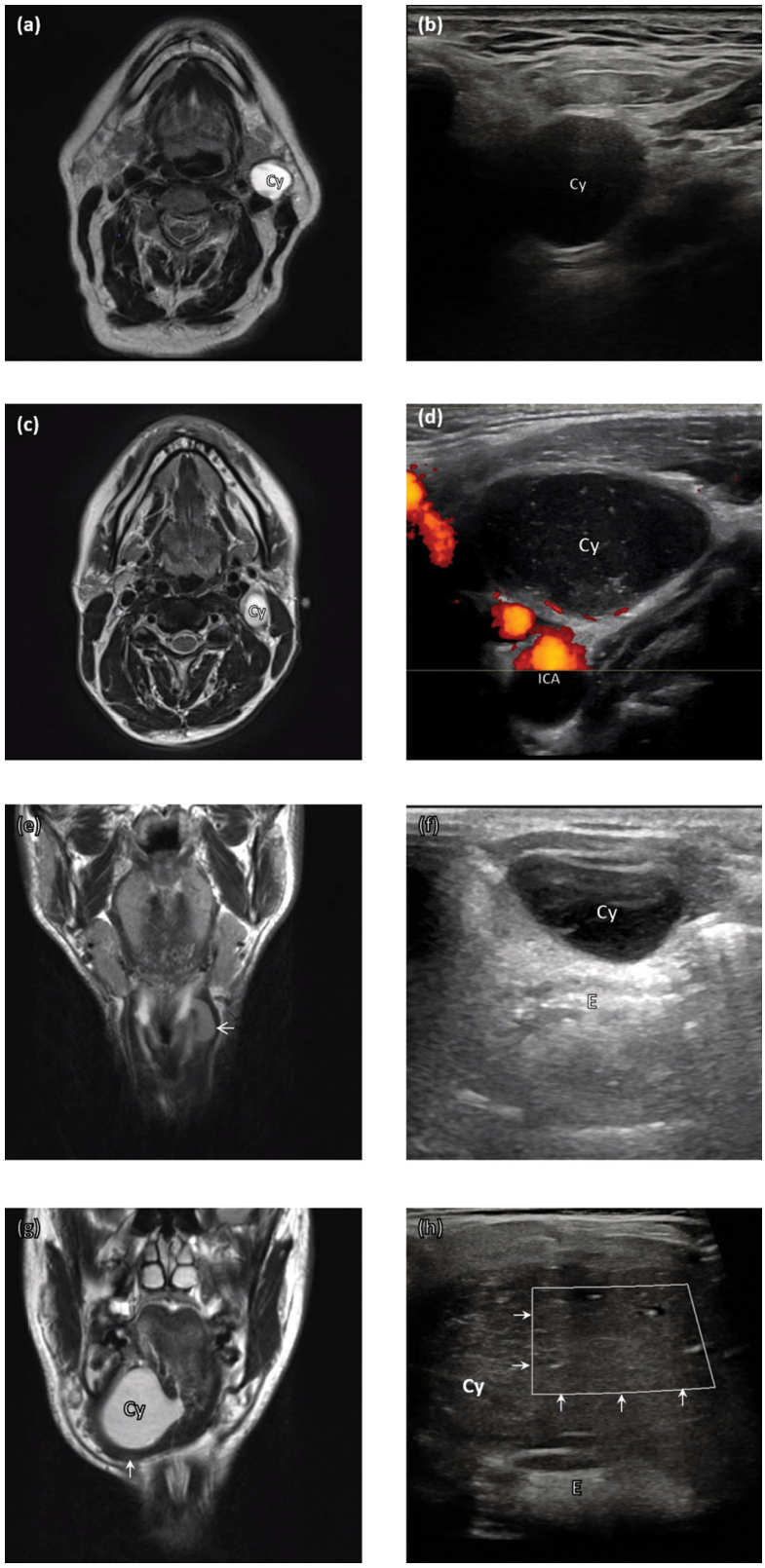

Figure 69.13 Four different patients with a cystic neck mass. Patient 1: (a, b) Axial T2-weighted and US images of a left level II cystic SCC node (Cy) in a patient with carcinoma unknown primary. Patient 2: (c, d) axial T2-weighted and US (with Doppler) images of a different patient with an infected, left branchial cleft cyst (Cy), with avascular echogenic content (debris). Patient 3: (e, f) Coronal T2-weighted and US images of another patient with a left paramedial thyroglossal duct cyst (Cy) overlying the larynx, containing debris and exhibiting posterior acoustic enhancement (E). Patient 4: (g, h) coronal T2-weighted and US (with Doppler) image of a large ranula. It is a smooth, markedly T2 hyperintense (cystic) structure (Cy), confined within the right sublingual space by the mylohyoid sling (short arrow). On US it exhibits avascular, isoechoic content (debris) and posterior acoustic enhancement (E).

Box 69.1 Cystic neck lesions

- Lymphangioma.
- Thyroglossal duct cyst.
- Branchial cleft cyst.
- Ranula.
- Dermoid.
- Epidermoid.
- Salivary gland cystic lesion (abscess/Warthin tumour/dilated ducts).
- Necrotic lymph node (malignant/infected).
- Necrotic infected lymph node.
- Cystic schwannoma.
- Tornwald's cyst.
- Adenoid retention cyst.
- Peritosillar abscess.
- Cystic thyroid nodule.

FURTHER READING

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70

Tracheostomy

Jack Faulkner, Aleix Rovira, and Asit Arora

Introduction

Tracheostomy is one of the oldest known surgical procedures. It involves the creation of a communicating tract between the anterior tracheal wall and the skin, bypassing the upper aerodigestive tract and aiding respiration. Records of tracheostomy procedures have been found as early as 3500 BCE in ancient Egypt and it is now an essential surgical procedure for the management of diseases of not only the head and neck, but also for other systemic indications.

In modern medicine, tracheostomies are performed for three principal indications: upper airway obstruction, airway protection from aspiration, and, most commonly, to aid in long-term mechanical ventilation and weaning.

Tracheostomy can be performed through open surgical techniques or via percutaneous dilatational methods. This chapter focuses primarily on surgical tracheostomy and the management of patients with a tracheostoma.

Patient considerations

Tracheostomy provides safe, long-term airway support for patients requiring airway protection, mechanical ventilation, or enhanced airway toileting. It does, however, carry significant quality-of-life limitations, including speech and oral nutrition, and they require high levels of care. These issues should be fully discussed with patients prior to the procedure where possible.

Surgical tracheostomy technique

Preparation

Where possible, patients should be fully assessed prior to tracheostomy. Cardiopulmonary function should be optimized, and any coagulopathy addressed.

A full head and neck evaluation should be performed to assess the level of any obstruction and ensure that a tracheostomy will sufficiently bypass it. The patient's anatomy should be noted. A short thick neck, enlarged thyroid, poor neck extension, and tracheal deviation may make the procedure more challenging.

The patient should be positioned supine with the neck moderately extended with a shoulder roll. Overextension of the neck can result in a lower tracheostomy and consequent narrowing of the airway.

Typically, the patient is anaesthetized and intubated; however, where endotracheal intubation is not possible, awake local anaesthetic tracheostomy can be performed.

Procedure

Anatomical landmarks of the thyroid notch, the inferior border of the cricoid cartilage, and the sternal notch are marked. The surgeon should palpate the sternal notch to assess the possibility of a high-riding brachiocephalic artery.

A 2–3 cm horizontal skin incision is made halfway between the inferior boarder of the cricoid cartilage and the sternal notch.

The subcutaneous tissue and platysma are divided to expose the strap muscles. The median raphe is identified. If an anterior thyroid vein is encountered, it can be ligated and divided, or retracted laterally.

The avascular raphe is divided vertically in the midline and the strap muscles retracted to expose the thyroid isthmus and trachea.

Normally, the thyroid gland can be retracted superiorly to expose the trachea without the need for ligation and division; if the isthmus is bulky, it can be divided in full, or partly using bipolar electrocautery, taking care not to injure the trachea. The thyroid gland is very vascular and careful haemostasis should be ensured if the isthmus is divided.

A cricoid hook is placed under the cricoid cartilage and utilized to elevate the larynx and with blunt dissection and palpation expose the second and third tracheal rings.

The tracheostomy tube cuff should be tested at this point and the anaesthetic team informed that the surgeon is ready to make a window in the trachea.

Classically, the anaesthetic team deflates the endotracheal tube cuff prior to incising the trachea to prevent damage to the cuff. An alternative technique introduced in the COVID-19 pandemic is to introduce the tube further down the trachea to allow ongoing ventilation and minimize aerosolization risks.

The trachea is opened at the level of the second and third rings.

Electrocautery should be avoided when opening the trachea to minimize the risk of oxygen-related burns.

Once the trachea is opened, the anaesthetist is asked to withdraw the endotracheal tube to above the opening and the tracheostomy tube is inserted.

The inner cannula is introduced, and the cuff inflated without removal of retractors in case of misplacement.

The tracheostomy is connected to the anaesthetic circuit and correct placement confirmed via capnography. Once confirmed, the cricoid hook and retractors are removed and the tracheostomy tube secured with four sutures and soft ties around the neck. The sutures should remain in place until the first tube change between 5 and 7 postoperative days.

Troubleshooting

1. *Management of the thyroid isthmus*: the thyroid isthmus can either be retracted superiorly and preserved, or it might be divided to gain further exposure of the tracheal anterior wall. One of the advantages of preserving the thyroid isthmus is a decrease of the surgical time without an increase of surgical and postoperative complications.[1] Therefore, unless a large thyroid isthmus is encountered and when the patient's anatomy allows, thyroid isthmus preservation should be preferred. When it needs to be divided, classic ligation can be used. However, electrocautery and other vessel sealing devices have proved to be equally effective.
2. *High versus low tracheostomy*: the ideal location of the tracheostomy is between the second and third tracheal ring. Higher entry points into the trachea can lead to subglottic stenosis while a lower tracheostoma is more challenging due to the posterior inferior deflection of the trachea and increased risk of brachiocephalic vessel injury. This issue is controversial when performing a tracheostomy on patients with laryngeal malignancy and airway obstruction. A high tracheostomy allows simpler resection of the stoma at time of laryngectomy and should be the preferred option; the hypothesized greater risk of seeding disease with a high tracheostomy is not clinically significant, as these patients often undergo wide local resection and reconstruction of the high tracheostomy site, and adjuvant radiation reduces chances local recurrence. Low tracheostomy has less risk of tumour seeding, but the resection will need to include a longer tracheal segment to ensure clear margins, making it surgically more challenging.
3. *Tracheal opening*: the trachea can be opened via multiple methods—vertical, horizontal, or cruciate incisions can be performed. A square tracheal window can be resected, or three sides of the square incised to create a Bjö rk flap. Li et al. identified the absence of Bjork flap as a risk factor for tracheal stenosis.[2]
4. *Cricoid hook*: the cricoid hook is very useful instrument that increases the procedure safety in securing and approximating the trachea to the skin when tracheostomy tube insertion is performed.
5. *Prophylactic antibiotics*: tracheostomy is a clean-contaminated surgical procedure. As such, prophylactic antibiotic can reduce the rate of infection as demonstrated in a preliminary randomized control trial with surgical tracheostomies and a large series of percutaneous tracheostomy.[3,4]
6. *Securing tracheostomy tube*: different studies have demonstrated that median or lateral sutures from the tracheostomy flange to the skin exhibit greater resistance to displacement of the tube than the tracheostomy strap alone.[5] A recent study with animal tissue simulation has shown that medially placed sutures are the most secure method in order to avoid tube displacement.[6]

Tracheostomy complications

Intraoperative

Haemorrhage

Bleeding is the most common intraoperative complication and can vary in severity. Fatal intraoperative haemorrhage is a significant risk in patients with a high-riding innominate artery if injured during inferior dissection. If dissection is inadvertently performed lateral to the trachea due to distorted patient anatomy or morbid obesity, attention must be paid to avoid the carotid sheath. Up to 5% of patients may have a thyroid ima artery (an anomalous, embryonic remnant) lying anterior to the trachea and care must be taken to appropriately ligate it when present.[7]

Pneumomediastinum/pneumothorax

A false tract anterior to trachea may be created intraoperatively, secondary to positive pressure ventilation or forced coughing against a tightly sutured or packed wound. This may lead to pneumomediastinum or pneumothorax, and if suspected, a chest X-ray should be performed. Pneumomediastinum is often self-limiting; however, a pneumothorax may need chest drain placement.

Tracheal injury/oesophageal perforation

Most tracheal wall injuries are small and heal uneventfully. Larger tears, perforations, and oesophageal damage might lead to bleeding and air leak and are potentially life-threatening. If perforation occurs, surgical repair is the preferred treatment option for the fistula; tracheal stenting can be performed for those patients not suitable for surgical management.

Early complications

Tube obstruction

Blockage of the tracheostomy tube is common in the early postoperative period due to blood or mucus. Appropriate management with humidified oxygen, regular inner cannula cleaning, and flexible tracheal suctioning can minimize risks of complete obstruction. The UK National Tracheostomy Safety Project has developed an emergency management algorithm for tube obstruction and displacement. It is advised that emergency algorithms are readily available at the patient bedside for all patients with tracheostomy (**Figure 70.1**).

Tube dislodgement

During the first days after tracheostomy insertion, tube displacement represents a medical emergency due to the absence of an established tract that may make tube reinsertion challenging. Stay sutures may be placed at the time of tracheostomy in the lateral trachea will aid in tube replacement (see algorithm in **Figure 70.1**).

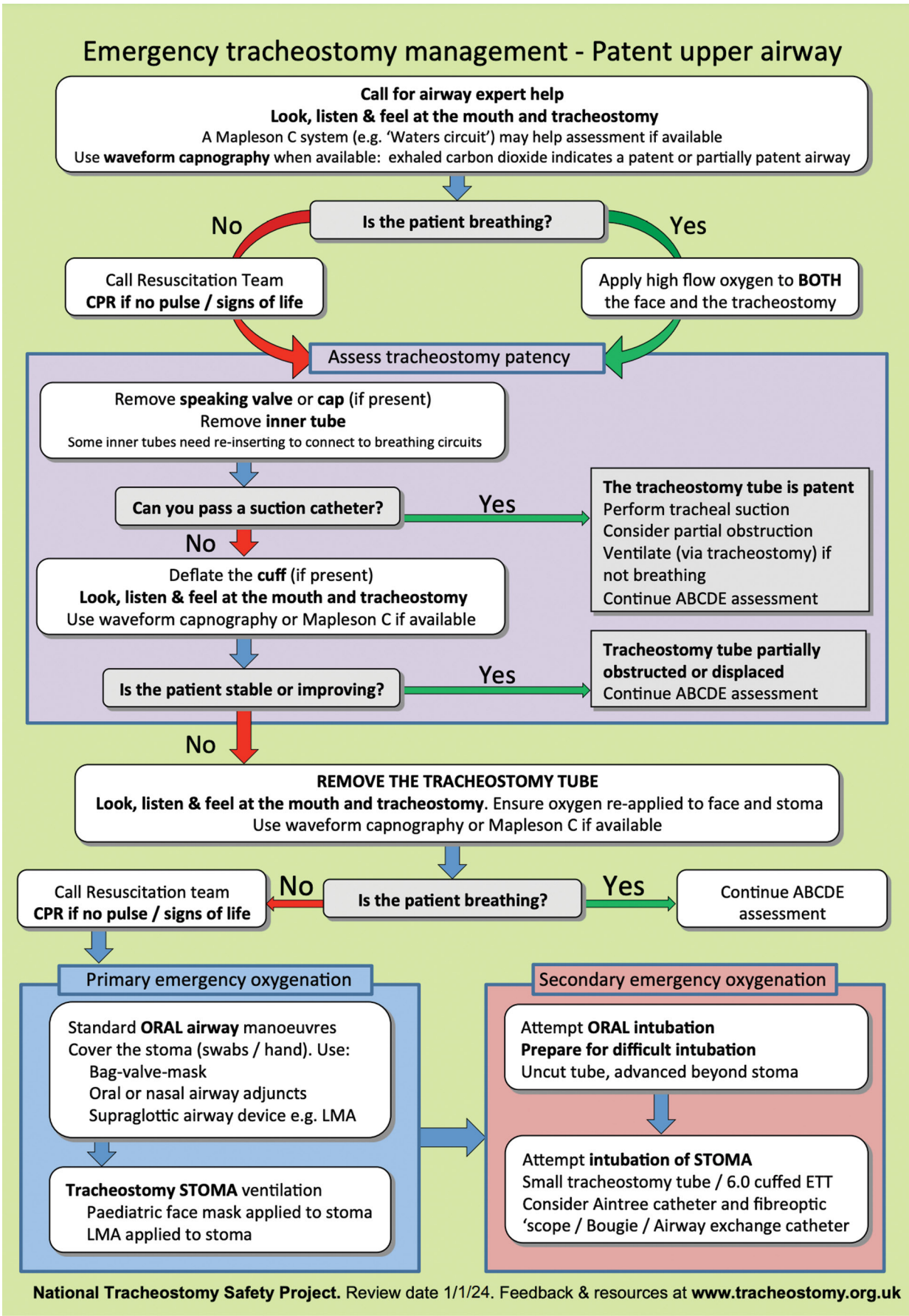

Figure 70.1 National Tracheostomy Safety Project emergency management algorithm.
Reproduced with permission from www.tracheostomy.org.uk

Infection

Local infection and tracheitis is common following tracheostomy; severe infections are rare. Most wound infections can be managed with antiseptic stomal wound care and systemic antibiotics.

Late complications

Tracheal stenosis

Tracheostomy-related tracheal stenosis is challenging to treat and may require treatments ranging from balloon dilatation to tracheal resection. Risk factors include tracheostomy cuff pressures greater than 30 cmH_2O, endotracheal intubation exceeding 10 days, reflux, obesity, tracheostomy size greater than 6 (internal diameter), percutaneous insertion, and absence of Björk flap.[2]

Tracheoinnominate fistula

Fistulation between the innominate (brachiocephalic) artery and tracheostoma is rare with an incidence of less than 1% but it carries a very high mortality rate (>80%).[8] Low tracheostomy, cuff overinflation, and a high-riding brachiocephalic artery are risk factors. Bleeding identified more than 48 hours after tracheostomy insertion may originate from the erosion of a major vessel. This may be due to a tracheoinnominate fistula but also, although less frequent, from erosion of the common carotid artery, inferior thyroid artery, innominate vein, or the aortic arch. Management of possible sentinel bleeding should take place in the operating room using a rigid bronchoscope to examine the stoma and anterior wall with the tracheostomy tube removed. Equipment for prompt surgical intervention should be readily available. In case of a massive bleed, the rigid bronchoscope can be used to compress the innominate artery against the sternum, while providing adequate suction, oxygenation, and ventilation, as measures are put in place for definitive management. This may include vessel repair and/or replacement, with vascularized flaps to protect the repaired vessel.

Tracheo-oesophageal fistula

This is a rare late complication after tracheostomy insertion and is related to pressure necrosis and erosion of the posterior tracheal wall. When suspected, a longer cuffed tube must be inserted to prevent pulmonary infection. The fistula will often need surgical repair with thoracic surgical input.

Persistent tracheocutaneous fistula

Following decannulation, the tracheostoma usually closes within 24–48 hours. This can be inhibited by granulation tissue at the stoma site that may be treated with topical silver nitrate cauterization; mature epithelialized fistulas will need surgical excision and closure of the tract in layers.

Care of tracheostomy patients

As the indications for tracheostomy cover a wide range, care for these patients occurs at multiple settings from the acute hospital to the community. Care should take a patient-centred approach with paramount importance placed on optimizing safety.

Quality improvement

Tracheostomy complications are commonplace and can be catastrophic, despite the fact that tracheostomy-related morbidity and mortality is often preventable. It has been reported that tracheostomy-related complications may account for 50% of all airway-related hypoxic brain injuries and deaths.[9] The majority of tracheostomy complications can be safely and effectively managed by appropriately trained individuals.

The Global Tracheostomy Collaborative is an international quality improvement collaboration centred on reducing tracheostomy-related morbidity and optimizing care. Established in 2012, it aims to improve patient safety through five key principles:

1. *Team-based care*: a multidisciplinary team of surgeons, nursing staff, speech pathologists, intensive care, and other appropriate members should meet weekly to review inpatients and make joint management decisions.
2. *Standard protocols*: established protocols should be utilized to ensure that every tracheostomy patient receives the same preoperative, perioperative, and postoperative care, barring patient-specific needs or complications.
3. *Staff education and assignment*: hospital staff should receive the appropriate training and tracheostomy-trained staff be available to attend an emergency at all times.
4. *Patient and caregiver involvement*: patients and their families should be involved in all aspects of tracheostomy care and patients and family representatives should sit on the institutional tracheostomy committee.
5. *Data collection*: the Global Tracheostomy Collaborative maintains a worldwide database containing key data from member hospitals on each admission for patients with tracheostomies. Regular reports are issued to member hospitals that allow them to track their progress and compare outcomes.

The Global Tracheostomy Collaborative theorize that most tracheostomy complications are preventable through appropriate multidisciplinary care and ongoing quality improvement efforts.[10]

Multidisciplinary care

Bedside information: following tracheostomy all patients should have vital information and emergency protocols displayed at their bedside. These include highlighted date of surgery, tube type and size, and upper airway assessments of the patient as well as emergency algorithms. The National Tracheostomy Safety Project has developed bedhead information sheets and emergency algorithms for this purpose (**Figure 70.2**).[11]

Cuff management: tracheostomy tube cuffs allow positive pressure ventilation and provide airway protection. Cuff pressures should be maintained between 20 and 30 cmH_2O to facilitate ventilation while minimizing tissue ischaemia. When positive pressure ventilation is not necessary, and the patient is able to safely manage their secretions, the cuff may be deflated.

Humidification: humidification of inspired air/gases reduces secretion viscosity and minimizes obstruction risks. Patients receiving mechanical ventilation or supplementary oxygen should do so through humidified circuits and heat moisture exchangers (e.g. Swedish nose or bib) should be utilized to maintain humidification. Regular saline nebulizers can further enhance humidification efforts.

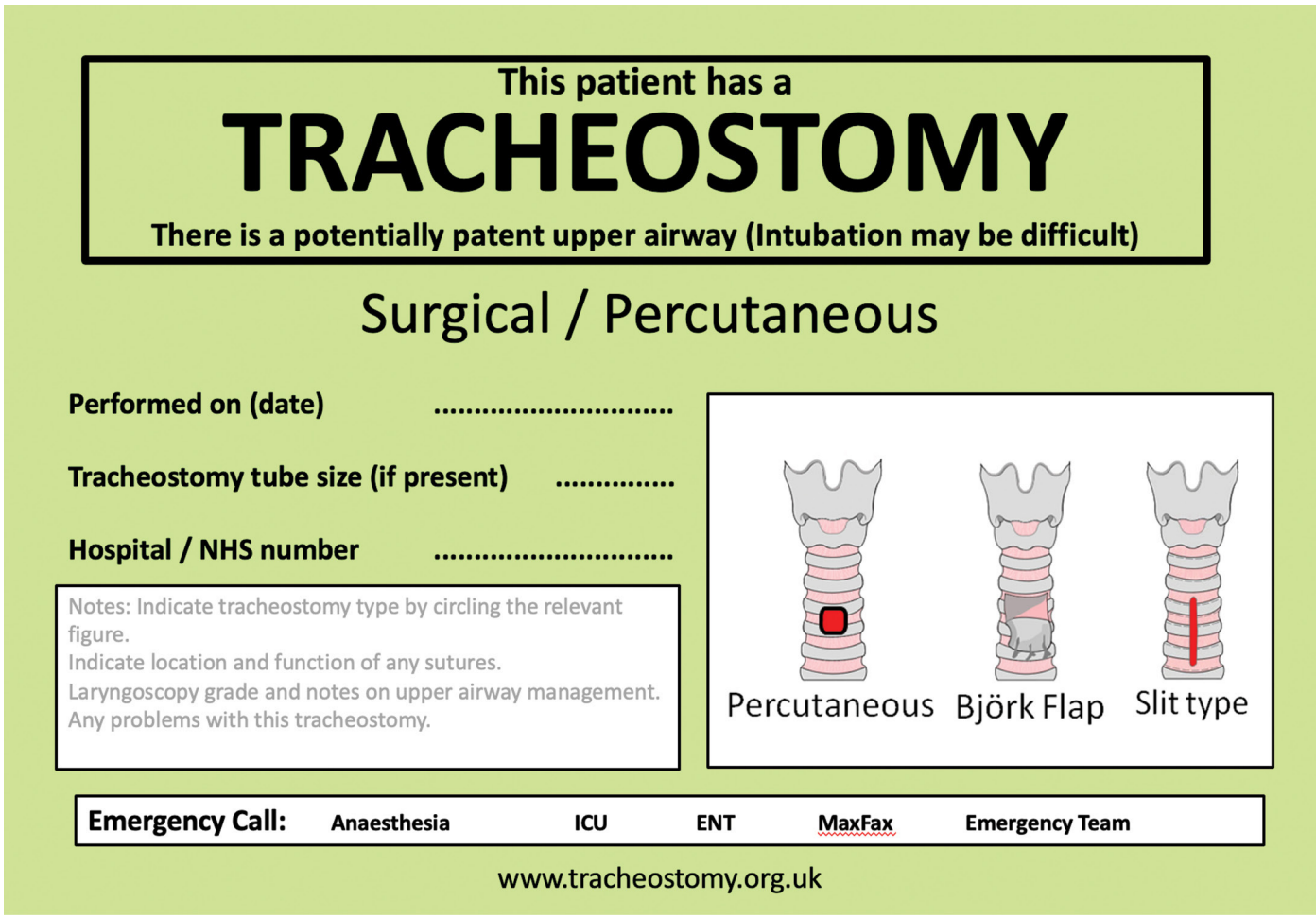

Figure 70.2 Bedhead information from the National Tracheostomy Safety Project.
Reproduced with permission from www.tracheostomy.org.uk

Secretion management: following tracheostomy, regular suctioning of the tracheostomy should be performed. This should be performed hourly for the first 24 hours and can then be tailored to the secretion load.

Swallowing: tracheostomy patients should be assessed by speech and language therapists prior to commencing oral intake.

First tube change: sufficient time should be allowed for the tract to form prior to the first tube change. This should occur 5–7 days following tracheostomy placement and the sutures should also be removed at this point. If earlier changes are required, this should be performed in an appropriate setting with emergency equipment and expertise available to rapidly re-establish a lost airway, with robust planning for all eventualities.

Emergency management

Clinical assessment of patient with tracheostomy should follow the same basic life support (ABCDE) principles used for the evaluation of any patient. However, some differences must be considered.

The airway is not physiological and comprehensive knowledge of the airway pathway as well as management of the potential complications is crucial.

Tracheostomy patients usually have two airways, the native upper airway and the tracheostomy. In assessing airway and breathing, both should be assessed. Supplementary oxygen if required should be applied to the face and to the tracheostomy. Simple and easily reversible scenarios can cause significant issues in tracheostomy patients. Any external devices attached to the tracheostomy should be removed in compromised or distressed patients. Speaking valves and humidification devices, such as Swedish noses, can become blocked, causing airway obstruction.

Tracheostomy inner tubes should be removed and cleaned. Inner tubes allow the lumen to be cleaned without removal of the tracheostomy and reduce risk of obstruction.

Management of a blocked tracheostomy tube

Blocked tracheostomy tubes should be managed in accordance with the National Tracheostomy Safety Project algorithm in Figure 70.1.[11]

The UK National Tracheostomy Safety Project aims to improve the management of tracheostomy critical incidents. The project has standardized resources for education of both healthcare providers and patients. Three self-directed learning modules include basic knowledge (anatomy, physiology, indications, procedures), emergency management (tracheostomy and laryngectomy), and nursing and general care, including associated devices. The emergency algorithms were developed by consensus, and the final algorithms describe a universal approach to management of tracheostomy

emergencies. The full emergency algorithms can be found at www.tracheostomy.org.uk.

Tracheostomy and COVID-19

The coronavirus disease 2019 (COVID-19) pandemic placed an unprecedented demand on healthcare systems around the world and put a premium on rapid evaluation and distribution of data related to COVID-19 management. COVIDTrach is a UK multicentre collaborative that was established to rapidly assess and disseminate data and recommendations on a national scale. Tracheostomy in the context of COVID-19 requires several practice-changing recommendations that differ from pre-pandemic practice.[12]

During tracheostomy there is the potential for aerosolization of pathogens and appropriate precautions must be taken to minimize risks of disseminating the disease to patient and staff. All aerosol-generating procedures should be performed with appropriate personal protective equipment including FFP3 mask, eye protection, fluid-repellent disposable surgical gown, and gloves and only necessary personnel should be present.

It is recommended that all COVID-related tracheostomies are performed in negative-pressure ventilation where possible.

Timing of tracheostomy has been controversial in COVID-19. Early in the pandemic there was significant concern over risks of transmission during the aerosol-generating tracheostomy procedure. This resulted in several organizations advocating a cautious and delayed approach. A review of 26 international guidelines demonstrated recommended timings from 3 to more than 21 days post intubation.[13,14]

As understanding of the natural cycle of COVID-19 developed, most recommendations balanced risk to staff with risk to patients and advocate a minimum of 14 days of mechanical ventilation prior to tracheostomy. As confidence in personal protective equipment improved and outcomes of patients with severe COVID-19 improved, reports have emerged advocating early tracheostomy with potential mortality benefits. Case selection and timings in the context of COVID will continue to evolve as more data become available.[13]

Tracheostomy technique

Location of tracheostomy in COVID-19 patients provides logistical hurdles. Most operating theatres lack negative-pressure facilities and moving critically ill patients from intensive care is not without risk.

Insertion techniques of open surgical versus percutaneous are debated. Perceived benefits to the percutaneous technique are familiarity of technique to critical care staff, reduced air leak from the smaller stoma, and reduced bleeding. However, percutaneous tracheostomy involves more airway manipulation, with withdrawal of the endotracheal tube risking aerosolization. Open surgical techniques allow for insertion of the tracheostomy tube under direct vision and modification of techniques including inserting the endotracheal tube into the trachea at the time of window creation to minimize risks of aerosolization. Large multi-institution data suggest there is no clear advantage of one technique over the other.[15,16]

Most institutions advocate suspending ventilation at times of high aerosolization to minimize risks. This is when the trachea is opened, and dilatation of the stoma and during insertion of the tracheostomy tube. This apnoeic period poses a serious risk to patients and may be mitigated with preoxygenation.[17]

Basic principles of tracheostomy management in COVID-19

The same basic principles of tracheostomy care should be applied to patients with COVID-19 infection as for any other patient with tracheostomy while minimizing aerosolization risks. Strategies to reduce inner cannula changes and suctioning, using 'closed suction' systems, and using heat and moisture exchange filters in ventilator circuits have been proposed.[16]

During weaning prior to COVID-19, regular periods of cuff deflation, use of vocalization strategies through one-way valves, swallowing rehabilitation, and promotion of coughing to the mouth was recommended. However, all these steps have the potential of both droplet and aerosol generation when on a ventilator, hence the decannulation process should be modified to only move through these stages once the patient can tolerate self-ventilation. The early use of one-way valves is a useful tool. They aid in respiratory rehabilitation and additionally may allow patient speech which is critical in communication when visitors may be limited, and telephone communication may be the only means of contact.[16]

As the pandemic evolves, more information regarding tracheostomy management has become available and collaborative research efforts are paramount in expeditious data collection and dissemination.

Conclusion

Tracheostomy is an airway management technique that is safe and well established. It can be performed via open surgical or percutaneous techniques and is integral to the management of long-term mechanical ventilated patients and patients with airway obstruction.

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71

Phonosurgery

Kate Heathcote

Background

As with all aspects of ear, nose, and throat surgery, phonosurgery has been revolutionized by the rapid advancement of endoscopic equipment and in particular video endoscopes.

The subspecialty of laryngology has emerged through the increased ability to assess and treat disorders of the airway, voice, and swallow.

Poor laryngeal function will affect all its roles and thus a patient with a voice complaint may also have airway and swallow issues that must be considered and addressed (see Chapter 48). Therefore, it is imperative that laryngeal assessment should be global, taking into consideration other aspects, and not just 'the voice'.

Introduction

By definition, 'phonosurgery' is the surgical treatment of voice disorders.

By breaking down the larynx into its structural components and function, it is possible to understand the pathophysiology of laryngeal disorders. To identify the cause of a voice disorder, it is essential to examine the larynx with equipment that shows the intricate details and movement of the vocal folds. The gold standard for this is high-definition videolaryngoscopy, with stroboscopy and image enhancement.

Clinical assessment of dysphonia is covered elsewhere (see Chapter 47) and is critical to the decision on whether the patient needs surgical intervention. Several instances exist where it is not possible to determine the diagnosis in an outpatient setting, and assessment under general anaesthetic (GA) will be necessary. Such entities include scars, sulci, and mucosal bridges. Differentiation between the various causes of vocal fold palsy (e.g. neurogenic versus cricoarytenoid fixation) can be difficult without palpating the vocal folds and will need a day-case microlaryngoscopic assessment. All phonosurgical interventions need comprehensive voice assessment before and after surgery to monitor outcomes for patient feedback and personal audit (see Chapter 47).

Lesions of the vocal folds

Vocal fold nodules are commonly used to describe calluses that appear superficially on the edge of the vocal folds due to traumatic voice use. These are best treated by voice rest and input from speech therapy. However, there are cases where this fails or a faster recovery is needed, and microlaryngoscopy for injection of a long-acting steroid into the nodule should be considered. Occasionally, when the nodules become fibrosed, they will never resolve with conservative measures and removal is indicated. Another consequence of voice misuse is microvascular trauma causing bleeding into superficial layers of the vocal folds. Again, this may resolve with voice rest but a *haemorrhagic polyp* may occur, requiring laser-assisted (**Figure 71.1**) or cold steel resection. Some lesions of the vocal folds will not settle spontaneously and require microlaryngoscopy and phonosurgery at first diagnosis. The most common are *intracordal cysts* which should be excised in a timely manner before they rupture and leave an untreatable scar, with a permanent impact on the voice (**Figure 71.2**). These can be confused with *bamboo nodules* or *rheumatoid nodules* which are discrete areas of inflammation related to autoimmune conditions. These cannot be removed but can settle with treatment of the underlying condition and steroid injection into the lesion.

Some conditions specifically affect the vibratory edge of the vocal fold. Areas of cystic change on the vibratory edge and *laryngeal papillomatosis* can persist, needing cold steel or laser-assisted resection. In the latter condition, surgery is not curative, but it controls the disease thus improving voice and maintaining the airway.

Finally, although not strictly a lesion of the vocal folds, *ventricular cysts* are common lesions that protrude out of the laryngeal ventricles and sit on the superior surface of the vocal folds. They cause dysphonia by dampening down the vibration of the vocal folds and thus require endoscopic resection.

Thus, owing to the range of pathology, a firm diagnosis cannot be established until the patient is on the operating table, and thus requiring patients to be listed for microlaryngoscopy with or without phonosurgery and with or without steroid injection.

Vocal fold paralysis

Unilateral vocal fold paralysis is a common diagnosis as the vagus and recurrent laryngeal nerves are vulnerable to many insults (see Chapter 72). Due to an increased understanding of the impact of this condition and improvements in treatment options, most patients benefit from phonosurgical intervention in addition to speech

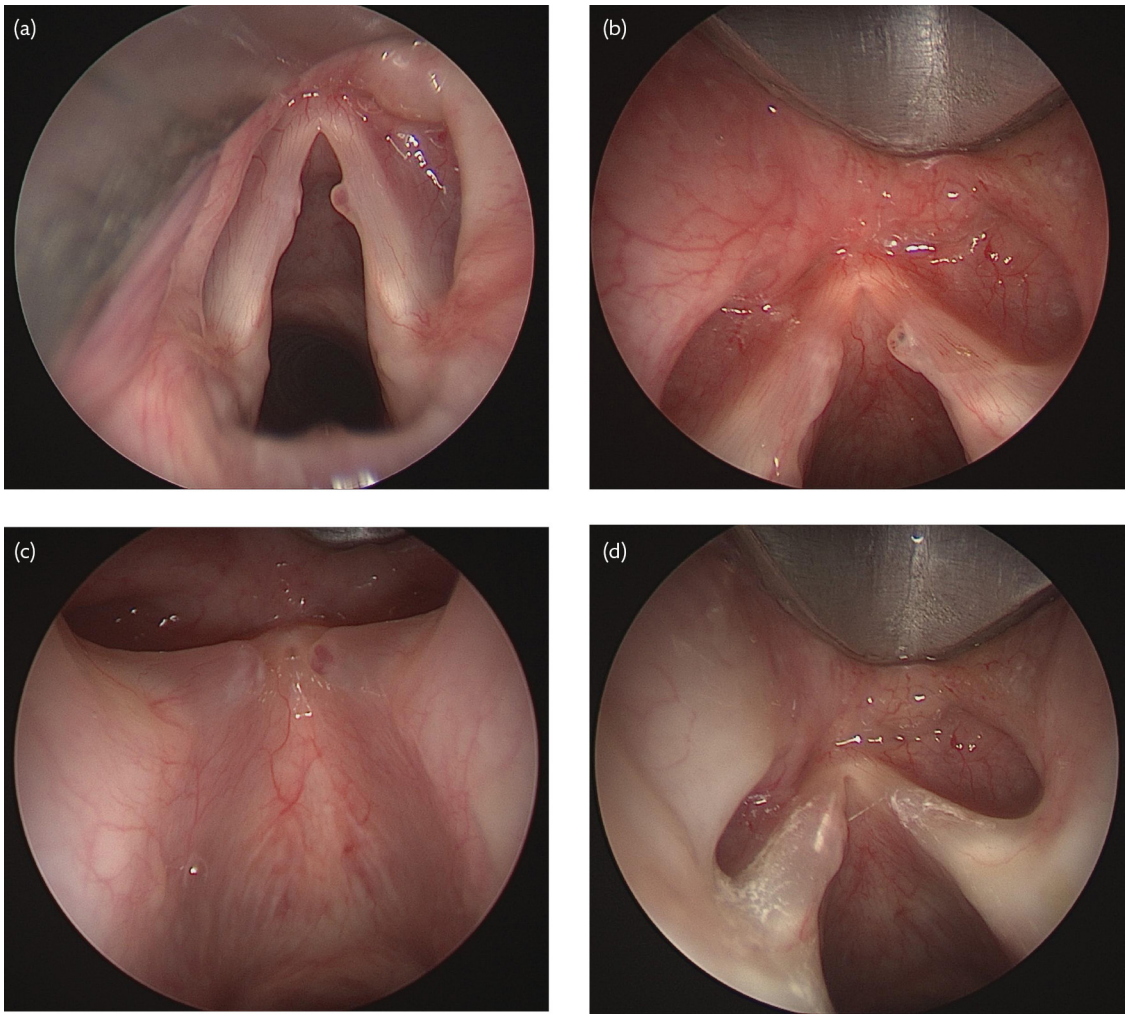

Figure 71.1 (a) Haemorrhagic polyp and contrecoup inflammation. (b) KTP laser to polyp. (c) View with 70° scope. (d) Intralesional steroids bilaterally.

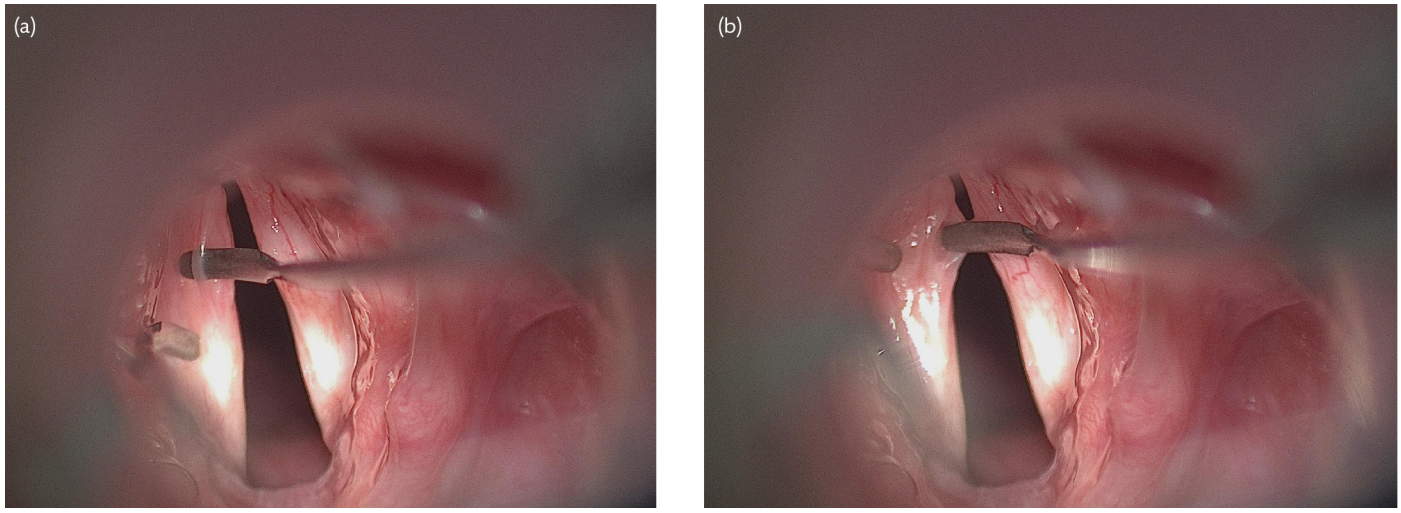

Figure 71.2 Examining a scarred vocal fold from a ruptured cyst under the microscope with palpators. (a) Mucosal bridge. (b) Deep sulcus.

therapy. The aim of phonosurgery is to maintain laryngeal competence. An incompetent larynx leaves the patient at risk of aspiration, with an ineffective cough and poor voice. Due to the wider availability of in-clinic local anaesthetic (LA) injection and augmentation of the vocal folds, it is possible to treat many more patients including those in whom the paralysis may be temporary. Expectant recovery, with intervention reserved only for symptomatic patients with residual deficits, is no longer acceptable. Vocal fold paralysis needs active management from the outset, initially with injection augmentation, followed by repeating this or progressing to a permanent option. This includes laryngeal framework surgery or non-selective reinnervation.

Bilateral vocal fold paralysis is a different matter, as in this condition treatments are focused on improving the critical airway instead of the voice. Rather, treatment is usually at the cost of the voice, and hence these interventions do not fall under the heading of phonosurgery.

Surgical principles

The surgical approach taken must consider patient factors as well as local equipment and expertise. A number of conditions can now be treated under LA in the clinic instead of under GA in theatre and the choice should be in the context of the individual case. For example, a fit and healthy professional singer with vocal fold nodules requiring steroid injection should be listed for microlaryngoscopy and steroid injection under GA. The risk of the GA for this patient is negligible and accurate placement of the steroid for optimum outcome is the priority. In contrast, an older patient with significant comorbidities, who relies on their voice for work but is not a singer, may still warrant a steroid injection but this may be best done via a channelled nasendoscope or transcutaneously under LA in the clinic. The precision is less but the risks of GA are avoided in this higher-risk patient where the goal is voice improvement rather than voice perfection.

As well as consideration of anaesthetic risks, other factors that may determine whether a LA or GA technique is performed include patient preference and poor access with rigid endoscopy.

Endolaryngeal phonosurgical techniques

Anaesthetic considerations

With all phonosurgery techniques under GA, careful consideration of the anaesthetic technique is required. Preoperative planning with the anaesthetist is essential. The ideal conditions are a tubeless surgical field without any movement of the vocal folds. This is possible in many cases with the use of non-invasive ventilation techniques. Transnasal humidified rapid insufflation ventilatory exchange ('THRIVE') allows the patient to be fully anaesthetized without an endotracheal tube and without significant airflow past the vocal folds (Figure 71.3). An alternative non-invasive ventilation technique is jet ventilation. Total intravenous anaesthesia is also used but a jet needle is attached to the side of the rigid laryngoscope. This is connected to the jet ventilator which regulates the pressure and regularity of the jetted oxygen/air. There is some movement of the vocal folds with this technique but it is regular, predictable, and can be stopped and restarted as required, titrated to patient saturations.

Figure 71.3 Suspension laryngoscopy with total intravenous anaesthesia and high-flow humidified nasal oxygen.

However, compared to humidified oxygenation, jet ventilation dries out the vocal folds, requiring the use of saline nebulizers in recovery. Subglottic jetting is an alternative jetting technique where a small catheter is inserted into the trachea and connected to the jet ventilator. This avoids the drying effect and movement of the vocal folds. The catheter is much smaller than an endotracheal tube but caution must be taken with concurrent use of the laser.

It must be said that a significant percentage of patients will not tolerate tubeless anaesthesia due to comorbidities or body habitus. In this setting, phonosurgery can often be achieved by working around a microlaryngoscopy tube but if the lesion is in the posterior aspect of the glottis, the tube will be a significant hindrance. Further, if a laser is required, a laser-proof tube is necessary, and given their bulk, much of the larynx can be obscured.

For these reasons, careful joint planning with the anaesthesia team is essential, and developing a laryngology team with specialist anaesthetic provision is the key to a successful service.

Surgical principles

Microlaryngoscopy and phonosurgery under GA is performed with the patient positioned supine with no shoulder bolster but a head ring. A rigid laryngoscope is inserted with a gum shield to protect the teeth. It is placed in the laryngeal inlet (except in the occasional case when a Lindholm scope is used and sits in the vallecula), close to but not touching the folds. It can be difficult to obtain a full view of the vocal folds and there are some techniques that can be employed to improve this. Slightly raising the head section of the operating table, gradually increasing the pressure on the suspension over several minutes as well as applying some gentle external pressure either manually or with a strap/tape can all improve access. It is almost impossible to operate through an anterior commissure scope, thus negating its routine use for interventions. However, an angled rigid endoscope is very useful to visualize the anterior commissure and essential to assess the ventricles and undersurface of the vocal folds. Rarely, a flexible channelled endoscope can be passed through a suspended laryngoscope to access and treat difficult locations.

Surgery may be performed holding a rigid endoscope in one hand for visualization with the instruments in the other. This technique can be useful if the view is poor and treatment is required to the anterior commissure, for example, an angled rigid scope in one hand and an angled microdebrider in the other to resect anterior ventricular cysts or papillomas (**Figure 71.4**). However, stability is better with a fixed microscope and this allows the use of both hands. A good example of this is using heart-shaped forceps in one hand to hold open a microincision while a cyst is shelled out with crocodile forceps held in the other. Lesions on the vibratory edge such as nodules and areas of cystic change can be removed with heart-shaped forceps in one hand and curved micro-scissors in the other (**Figure 71.5**). Great care should be taken with biopsies to minimize tissue removal and the most precise technique is using micro-biopsy forceps in one hand and curved scissors in the other. Micro-sutures can also be inserted to repair flaps, for example, after the evacuation of Reinke's oedema of the vocal folds.

Phonosurgery instruments are highly specialized and designed to minimize trauma to the delicate tissues, thus avoiding disastrous scarring. All lesions are approached with a view to minimizing tissue disruption. This means micro-incisions overlying the lesion and minimal dissection of the surrounding tissues. The essential instruments required on every phonosurgery set are palpators, heart-shaped forceps right and left, straight scissors, curved scissors right and left, fine sucker, and crocodile forceps right and left (**Figure 71.6**).

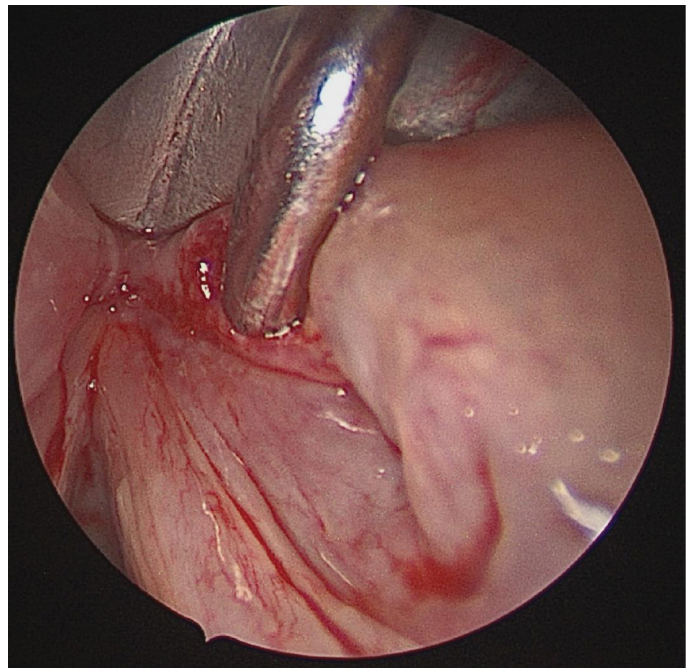

Figure 71.4 Anterior ventricular cyst visualized with 70° scope and debulked with angled microdebrider.

An adjunct used for epithelial preservation is hydrodissection, injecting a small volume of normal saline into Reinke's space. This can be used to exaggerate the plane between the epithelium and an underlying lesion such as a cyst.

Mounted on the microscope and controlled with a micromanipulator, the carbon dioxide (CO_2) laser is an excellent tool for phonosurgery. This set-up is commonly used to ablate papillomas if the microdebrider is not used. It may also be used to perform *arytenoid reduction* to facilitate glottic closure by removing the upper part of a paralysed arytenoid that is prolapsing and blocking closure. Myoneurectomy (see below) also employs the CO_2 laser. The KTP laser is another effective tool against vascular lesions, as its chromophore is haemoglobin, hence targeting blood vessels (**Figure 71.1**b). Unlike the CO_2 laser, it is typically used via a fibre held in a handpiece which may be of various angulations depending on requirements.

The critical thing to always keep in mind when performing phonosurgery is that no salvage options exist for a scarred vocal fold. If there was ever a time to remember the old adage 'first do no harm', it is here.

In-clinic transnasal phonosurgery

In some patients, LA may be the preferred option, and most laryngeal interventions are performed in the outpatient setting. As mentioned previously, steroid injections can be performed through a channelled endoscope to expedite the resolution of vocal fold nodules. Other inflammatory lesions such as vocal process granulomas, rheumatoid and bamboo nodules, and areas of inflammation resistant to voice rest, can be injected in-clinic.

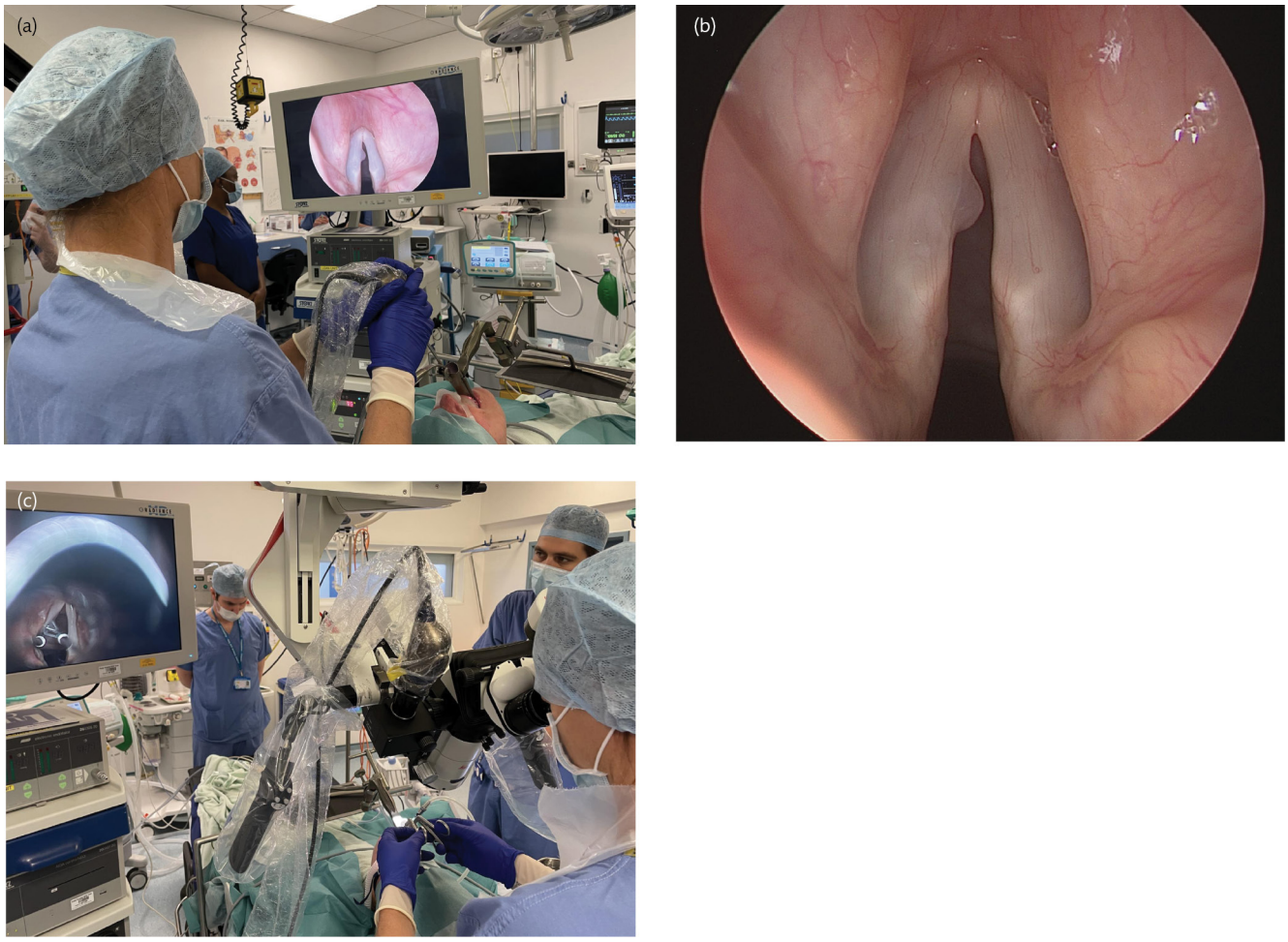

Figure 71.5 (a, b) Examining a superficial cyst with a rigid scope. (c) Excision of the cyst with heart-shaped forceps to the left and curved scissors to the right.

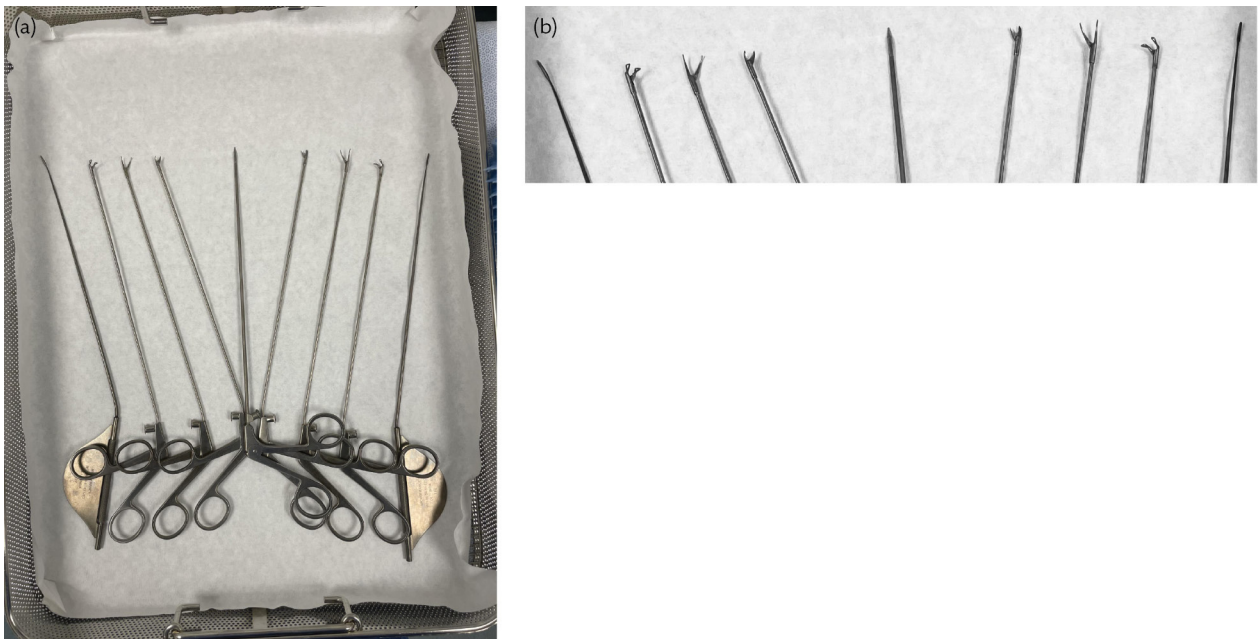

Figure 71.6 (a, b) Basic phonosurgery set with close-up. From centre outwards: straight scissors, curved scissors (left and right), crocodile forceps (left and right), heart-shaped forceps (left and right), palpators (left and right).

Figure 71.7 Trans-thyrohyoid injection augmentation of the vocal folds.

Vocal fold injection augmentation is a very common procedure in a laryngology treatment clinic. The most common indication is for unilateral vocal fold paralysis but it can be a very useful adjunct to speech therapy in cases of vocal cord atrophy and presbyphonia. Here, glottic closure is compromised by wasting of the vocal fold muscles due to prolonged steroid inhaler use and ageing, respectively.

It is also one of the only procedures shown to improve voice in the presence of scar (including sulci, vergeture, and mucosal bridge). Although this does not improve the area of scar, it does facilitate stronger closure of the vocal folds and improved voice volume.

Transcutaneous injection augmentation is performed with a nasoendoscopic view of the larynx to facilitate injection of LA and filler through the front of the neck. The transcutaneous route may be through thyrohyoid or cricothyroid membranes, aiming to augment the lateral aspect of the vocal fold in the thyroarytenoid muscle, adjacent to the internal aspect of the thyroid cartilage (Figure 71.7). Medializing the vibratory edge facilitates glottic closure. Substances may be short acting, such as hyaluronic acid (~3 months) or longer acting such as calcium hydroxyapatite (6–9 months). A short-acting substance is suitable when there may be recovery of the nerve, that is, iatrogenic or idiopathic paralysis/paresis. Other substances such as autologous fat or Bioplastique should be injected under GA as they require high pressures for injection. The advantage of an LA technique is that the patient can phonate to ascertain when glottic closure has been achieved. LA injection augmentation is a really useful technique when the status of the nerve is uncertain, in patients receiving palliative care (recurrent laryngeal nerve paralysis is common in lung and metastatic cancer), and in those who do not want to undergo more invasive external techniques.

In-clinic techniques are also used to manage vascular laryngeal lesions. Both KTP and blue lasers are very effective on vascular lesions (Figure 71.8). A laser-proof clinic room is required and laser safety precautions must be followed. The patient and all others in the room wear protective goggles. The nose is decongested and anaesthetized and then LA is injected through the channel of the endoscope to anaesthetize the larynx. Laser fibres are inserted through the channelled scope and the laser fired at the lesions. This technique is also used for the treatment of low-volume laryngeal papillomatosis.

External laryngeal phonosurgical techniques

Laryngeal framework surgery includes a variety of phonosurgical techniques via the external approach. This often requires entering the larynx through the thyroid cartilage and hence the term 'thyroplasty'. Isshiki described four thyroplasty techniques in the 1970s (Box 71.1). Since then, other laryngeal framework surgery techniques and modifications have emerged. *Type I thyroplasty* is in common use for the treatment of unilateral vocal fold paralysis. It is usually performed under LA (+/– sedation) and involves a skin crease incision at the level of thyroid cartilage. A window is cut in the cartilage and an implant is inserted alongside the vocal fold to medialize it. The position and the effectiveness are verified intraoperatively by asking the patient to phonate and by direct observation of the larynx with a nasendoscope. Various implants may be used including firm silicon, Gore-Tex ribbon, and titanium. Additional techniques may be performed to improve the position of the arytenoid such as *arytenoid adduction* which can be performed through the same incision.

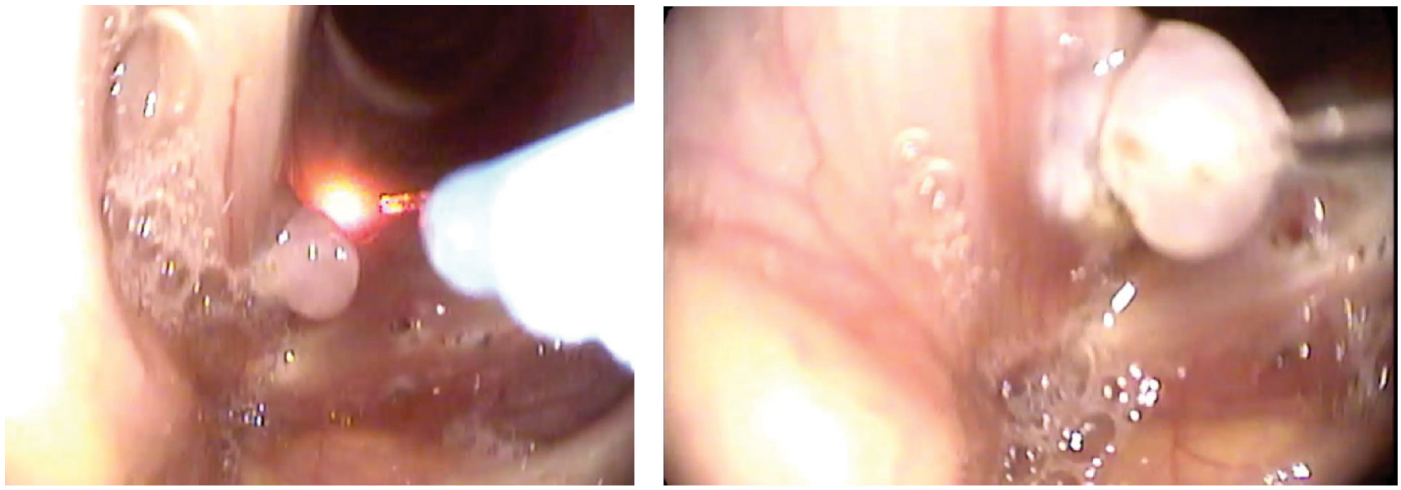

Figure 71.8 Transnasal approach with KTP laser fibre to treat a polyp in a patient not fit for GA.

Box 71.1 Isshiki classification of thyroplasty techniques

- Type I: vocal fold medialization (in unilateral vocal fold paralysis).
- Type II: to separate the folds (in adductor spasmodic dysphonia).
- Type III: to lower voice pitch (in pubophonia or female-to-male gender reassignment).
- Type IV: to raise voice pitch (in male-to-female gender reassignment).

It involves repositioning the arytenoid with a suture placed through the muscular process of the arytenoid.

Non-selective laryngeal reinnervation is an alternative to type I thyroplasty. This is performed under GA through a skin crease incision at the level of the cricoid cartilage. A branch of the ansa cervicalis (motor supply to the infrahyoid ('strap') muscles of the neck) is diverted to anastomose with the recurrent laryngeal nerve. While it does not restore movement of the vocal fold, the procedure provides tone and position to the paralysed hemilarynx. The advantage of this technique over type I thyroplasty is that it can be performed even when there is still filler present after a previous injection augmentation so that glottic competence can be maintained at all times. It is also suitable for children in whom injection augmentations must be done under GA and implants are not effective as the child out-grows them and laryngeal framework surgery can interrupt the growth of the larynx. It may be less effective in the elderly and is not possible in those unfit for GA.

Type II thyroplasty is not currently widely practised outside Japan as adductor spasmodic dysphonia is most commonly controlled with in-clinic botulinum toxin injections. Type II thyroplasty is a technique to widen the anterior commissure and involves a vertical incision through the apex of the thyroid cartilage which is then spread apart and held with titanium bridges. Other surgical techniques for the treatment of adductor spasmodic dysphonia are *selective laryngeal adductor denervation-reinnervation* which is also an open technique or *laser myoneurectomy* where the thyroarytenoid muscles and their nerves are resected via microlaryngoscopy and CO_2 laser. These techniques are also not widely practised due to the efficacy of botulinum toxin injection, but may be considered in patients who develop a resistance to toxin or where funding injections every 3–4 months is problematic.

Type III thyroplasty or 'relaxation' thyroplasty is a technique to lower the pitch of the voice. This is used in pubophonia where the pitch has failed to lower during puberty in the male leaving them with a voice in the female range. It is also used in female-to-male gender reassignment. A midline skin crease incision is made and the thyroid cartilage is made smaller by removing vertical strips of cartilage a few millimetres wide on one or both sides. This reduces the tension in the vocal folds.

Type IV thyroplasty is used to raise the pitch of the voice in male-to-female gender reassignment. It is a tensioning procedure also known as a cricothyroid approximation and is usually performed in conjunction with a cosmetic resection of the laryngeal prominence. Sutures are placed between the thyroid and cricoid cartilages across the cricothyroid membrane. Tightening these sutures tilts the thyroid cartilage forwards on the axis of the cricothyroid joints. This tensions the vocal folds and increases pitch, much as the cricothyroid muscle does but in a permanent way by removing the ability for the vocal folds to shorten. The alternative technique is microlaryngoscopy and suturing of the anterior one-third of the vocal folds together to make them shorter, known as a glottoplasty. This endoscopic technique avoids an external scar, although does not address the laryngeal prominence.

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72

Movement disorders of the larynx

Julia Selby

Introduction

Voice, breathing, swallow, and throat symptoms can exist in the absence of structural pathology and may instead be attributable to disrupted laryngeal function. Changes in laryngeal function may be caused by upper airway sensitization, neurological conditions, and functional voice disorders. The most common presentations within each category are presented below.

Upper airway disorders

Upper airway disorders appear to arise from a process of laryngeal sensitization that triggers an engrained cycle of hypersensitivity and hyperreactivity (Figure 72.1). The larynx may have become sensitized through a precipitating event, such as an upper respiratory tract infection, with further aggravation from comorbid irritants, such as nasal disease or acid/non-acid reflux. Previously innocuous triggers then elicit an exaggerated response, usually in the form of a persistent cough or a tendency towards unwarranted laryngeal closure. These disorders often display significant clinical overlap, with varying, additional impact on voice, swallowing, and throat comfort.

Patients whose upper airway symptoms are centred primarily on breathing will likely be referred directly to respiratory medicine, while those with voice, swallow, and an upper airway cough may commonly be seen in a general ear, nose, and throat (ENT) clinic.

Clinical presentation

Chronic refractory cough

Chronic refractory cough (CRC), typically defined as lasting 8 weeks or more, is a disabling condition that affects between 5% and 10% of the adult population. When severe, it is associated with comorbidities such as incontinence and cough syncope, that may lead in turn to social withdrawal, depression, and reduced quality of life.

Patients typically report a dry, tickly cough and enhanced sensitivity to environmental irritants, such as perfume and temperature changes. Exertion, talking, and laughing are also common triggers.

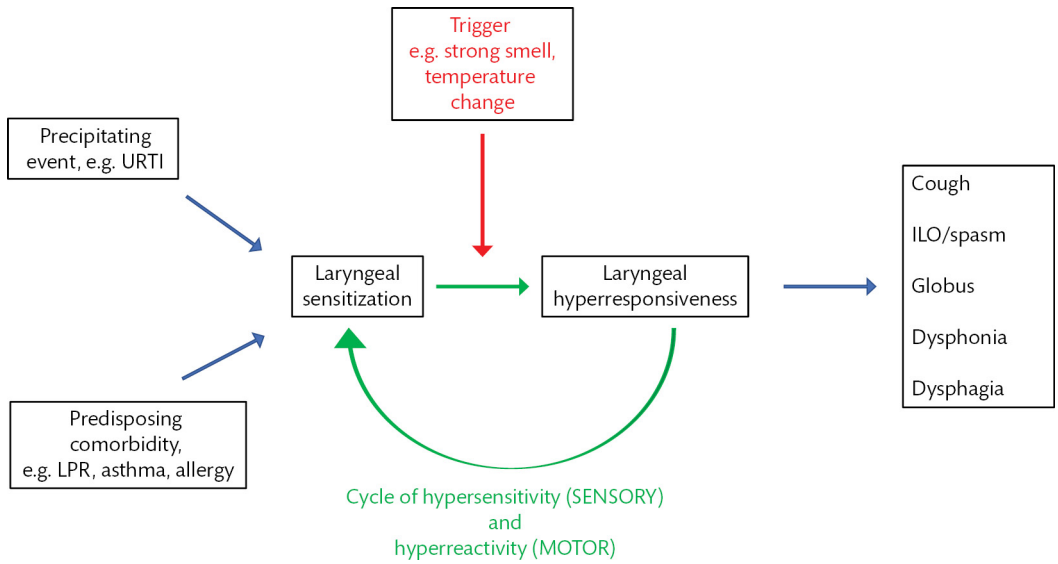

Figure 72.1 Laryngeal sensitization. ILO, inducible laryngeal obstruction; LPR, laryngopharyngeal reflux; URTI, upper respiratory tract infection.

These observations have led to the concept of cough hypersensitivity syndrome as a recognized diagnosis, where the cough reflex becomes hyperactive in response to low thresholds of thermal, chemical, or mechanical stimulation. Cough frequency may increase during periods of emotional stress, often mediated by an increase in shallow, mouth-dominant breathing that perpetuates the cycle of hypersensitivity. CRC is more common in females and typically presents in the fifties or sixties.

Inducible laryngeal obstruction

Inducible laryngeal obstruction (ILO), previously named paradoxical vocal fold movement or vocal cord dysfunction, is characterized by episodes of paradoxical vocal fold adduction on inspiration that cause breathing difficulties. Triggers tend to be external environmental stimuli, as with CRC, and indeed many ILO attacks are initiated by coughing. Patients often attend the accident and emergency department, where they are typically treated for asthma or a panic attack. In severe cases, patients avoid situations where they have been symptomatic previously and, at worst, can become housebound, with obvious socioeconomic consequences.

Laryngeal constriction may occur at either the glottic or supraglottic level, or both simultaneously. Airflow through the larynx is compromised as a result, creating audible turbulence, effortful breathing patterns, and sensations of throat tightness and discomfort. Changes in voice quality are also common due to increased laryngeal muscle tension and disrupted respiratory–phonatory coordination.

Psychological factors may be relevant in ILO. Although controversial, ILO has been proposed by some authors as a physical manifestation of underlying psychological distress, akin to psychogenic voice disorders described later in this chapter.

Exercise-induced laryngeal obstruction is a separate clinical condition, resulting from aerodynamic and pressure phenomena at increased ventilatory demand.

Laryngospasm

Laryngospasm refers to rapid, transient, complete closure of the larynx, which often leads to pre- or full syncope. It serves to protect the airway against entry of a foreign body or substance and can occur during both intubation and extubation. When laryngospasm becomes ingrained and unwarranted, treatment with botulinum toxin may be indicated.

Assessment and differential diagnosis

In some respects, upper airway disorders are easily identifiable due to their primary presenting symptoms: a dry, tickly cough, or episodes of throat tightness and breathing difficulties. The diagnostic challenge, however, is that patients may be asymptomatic in clinic, although that at least distinguishes them from most neurological and functional conditions where symptoms are present more consistently.

In addition to standard case history and examination to exclude malignancy, infection, foreign body inhalation, or, in cases of CRC, use of an angiotensin-converting enzyme inhibitor, information should be gathered on triggering agents, localization of tightness/wheeze, whether inspiratory or expiratory, time course of attacks, including recovery, and additional upper airway symptoms, such as globus sensation, voice or swallowing difficulties, and impact on quality of life.

Laryngoscopy may be unhelpful as a diagnostic tool, due to the transient nature of symptoms. Its use is warranted, however, to identify signs of acid reflux or nasal disease, which may be relevant laryngeal irritants, and to investigate the cause of any concomitant dysphonia or abnormal sensations in the neck or throat.

Laryngoscopy in patients with suspected ILO is likely to be normal unless they are symptomatic. Specialist provocation laryngoscopy clinics have been established, therefore, in many upper airway services, to visualize the larynx during generation of typical symptoms. Many cases of suspected ILO fail to show glottal narrowing on provocation; instead, symptoms are often due to intense sensations of upper airway tightness, breathing pattern changes, or a hypersensitive response in the form of throat clearing or coughing.

Management

Early management of common laryngeal irritants (e.g. postnasal drip, allergic rhinitis, and acid reflux) is highly beneficial. Beyond that, it is recommended that patients with CRC and ILO are referred to a specialist upper airway clinic, usually within respiratory medicine, where they can be treated in a multidisciplinary team setting. Upper airway patients often require expertise from respiratory medicine, otolaryngology, speech and language therapy (SLT), physiotherapy, and clinical psychology; the benefits to patients of receiving centralized, coordinated care should not be underestimated. Furthermore, they may require comprehensive airway assessment, including lung function tests and specialist investigations for asthma, and will likely require highly specialist therapy input to desensitize the larynx and restore normal function.

Onward referral to gastroenterology to establish the presence of reflux episodes may also be warranted.

Neuromuscular conditions

Neuromuscular voice and swallowing difficulties result from disruption to the control, coordination, and strength of laryngeal muscle movement, from underlying peripheral or central neurological changes. In some cases, a change in voice and swallow function may be an early indicator of neurological disease; in others, such changes occur as part of a broader, established disease process.

Clinical presentation

Vocal fold paralysis

Vocal fold paralysis may be unilateral or bilateral. The majority of cases are caused by pathology to the recurrent laryngeal nerve, but conditions affecting the cricoarytenoid joint, such as rheumatoid arthritis or trauma, or changes to the intrinsic laryngeal muscles from myopathy or tumour infiltration are also possible causes.

Unilateral paralysis is most problematic when the affected vocal cord is immobile in a widely abducted position. Voice quality is weak and breathy, with an ineffective cough, and patients are vulnerable to penetration and aspiration on swallowing. They may also

report breathlessness due to air wastage through the glottis, in the absence of respiratory disease.

If the vocal cord is paralysed in a medial position, the voice tends to be relatively unchanged, but patients may exhibit restricted breathing patterns due to the reduction in glottal area. Inspiration may be turbulent, and patients often breathe apically and irregularly, in an attempt to achieve a satisfying breath.

Bilateral vocal fold paralysis poses a trade-off between voice and airway patency. If the vocal folds are paralysed in a lateral position, the airway is preserved, but voice and swallow are compromised. Conversely, if the vocal folds are fixed in a paramedian position, intervention will be needed to improve the airway, although the voice may be strong.

Paralysis from recurrent laryngeal nerve injury is typically visible on laryngeal endoscopy and poses few diagnostic dilemmas. Damage to the external branch of the superior laryngeal nerve is less obvious but may be reflected in ipsilateral deviation of the epiglottic petiole. The resulting cricothyroid muscle impairment results in difficulty raising pitch and reduced vocal strength, with accompanying altered sensation in the laryngopharynx.

Laryngeal dystonia

Laryngeal dystonia (LD) is the term proposed in a recent multidisciplinary update to replace 'spasmodic dysphonia'. It is a focal dystonia characterized by task specificity and selective impairment of volitional phonation, but not whispering or innate vocal behaviours, such as laughing or crying. Adductor LD is the most common form, with intermittent voice stoppages on voiced sounds, leading to a strained-strangled voice quality. Patients subjectively report excessive effort and fatigue on speaking, difficulty modulating pitch, and an inability to project. Abductor LD is much rarer, characterized predominantly by voice breaks on voiceless consonants, while mixed LD exhibits features of both adductor LD and abductor LD. Recognition is now given to singer's LD, characterized by selective impairment during singing, and to adductor respiratory LD, in which adductor laryngeal spasms occur during inspiration, causing stridor and dyspnoea. LD is more common in females than males, with an average onset around 40 years of age, which can be gradual or sudden.

Vocal tremor

Vocal tremor may occur in isolation or as part of a benign essential tremor affecting the head and hands. Over-frequent muscle contraction creates a periodic 'wobble' in the voice and patients often report that they are mistaken for sounding emotional or on the verge of tears. Management is usually led by a neurologist, where beta blockers are often prescribed initially.

Parkinson's disease

The hypofunctional voice associated with Parkinson's disease is quiet, weak, and breathy, with reduced pitch inflection, and is usually accompanied by festinant bursts of speech and decreased articulatory precision. Voice changes tend to be predated by hand tremor, a shuffling gait, and other disruption to movement. In addition to medication (e.g. levodopa), voice therapy is central to treatment, in particular the Lee Silverman Voice Treatment programme.

Motor neuron disease

Voice, speech, and swallow changes are often the first signs of motor neuron disease and ENT specialists play a vital role, therefore, in ensuring prompt referral to neurology for early diagnosis. As the disease progresses, speech becomes slurred, slow, and hypernasal, with increased harshness and strain in the voice. The ability to swallow food and fluids safely becomes increasingly impaired over time, often resulting in enteral feeding.

Myasthenia gravis

Myasthenia gravis is an autoimmune disorder in which symptoms appear with muscle use and fatigue and abate with rest. Laryngeal signs and symptoms include hoarseness, vocal fatigue, loss of pitch control and variability, with increased hypernasality and swallowing difficulties.

Multiple sclerosis

Patients with multiple sclerosis exhibit poor control of pitch and volume, along with a harsh, strained voice quality. Their speech is often described as 'scanning', due to hesitancy and a loss of rhythm.

Assessment and differential diagnosis

The standard case history and laryngoscopic evaluation should be sufficient in identifying vocal cord paralysis and vocal tremor, and in placing other voice, swallow, and breathing symptoms in the context of broader neurological disease. Patients may already have a confirmed neurological diagnosis and the role of ENT assessment (see Chapter 47), therefore, is to provide an opinion on surgical options to optimize function or make an onward referral to SLT. Clearly, if patients have not been assessed by a neurologist, a referral should be made.

Differentiating between LD and muscle tension dysphonia (MTD; see below) can be challenging and often requires specialist voice SLT assessment. However, to elicit the task specificity associated with LD, patients can be asked to count from 60 to 70 and from 80 to 90. Symptoms of adductor LD worsen on voiced phonemes during 80–90, while patients with abductor LD show more difficulty on voiceless sounds from 60 to 70. MTD affects all speech sounds equally.

Signs of airway compromise or severe dysphagia must obviously be managed urgently.

Management

Patients with unilateral vocal fold paralysis should be referred to SLT prior to consideration of surgical intervention, in order to achieve the best possible vocal fold adduction, minimize the risk of developing compensatory strain, and manage accompanying changes to breathing and swallowing. Where therapy is unsuccessful, medialization or reinnervation procedures may be indicated.

Botulinum toxin injection continues to be the most common treatment for LD, but patients may benefit from voice therapy to reduce compensatory strain and trial other facilitative strategies to minimize the impact of muscle spasm (see Chapter 71).

Patients with a known neurological diagnosis may be referred to ENT for consideration of surgical options to maintain or optimize laryngeal function. Communication difficulties may extend far beyond changes in the larynx, however, and the benefits of intervention may be modest at best. Such decisions are best discussed in a

joint voice clinic with SLT, or with the neurological multidisciplinary team.

Functional voice disorders

Functional voice disorders constitute a large, heterogeneous group of non-organic voice disorders that may present as MTD or a psychogenic voice disorder. They can co-occur with an upper airway or neuromuscular condition, but they exhibit unique and distinctive features that differentiate them from each other and their neurological and upper airway counterparts.

Clinical presentation

Muscle tension dysphonia

MTD is generally characterized by hyperfunction of the true and/or false vocal cords, with narrowing of the aryepiglottic sphincter in some cases. It tends to result from excessive vocal demands or from compensation for a weakness at the level of the true vocal folds (e.g. with a vocal fold paralysis). Onset is common following an upper respiratory tract infection, where hyperadduction from coughing and altered phonatory patterns fail to resolve once the infection has cleared.

Patients tend to report a change in the pitch and quality of their voice, which usually sounds deeper and rougher, with accompanying symptoms of vocal fatigue, increased effort on speaking, and varying degrees of discomfort. Onset is usually gradual, except for acute cases of phonotrauma, and symptoms worsen consistently until treatment is initiated. Laryngoscopy will typically reveal the constriction associated with MTD, most commonly visible in an anterior–posterior dimension of the aryepiglottic folds, with overinvolvement of the false vocal folds and/or marked medial compression of the true vocal folds (**Figure 72.2**).

Psychogenic voice disorders

Psychogenic voice disorders result from disrupted psychological processes that lead to a loss of volitional control over phonation, where laryngeal anatomy and physiology are normal. The exact mechanism is unclear, but a proposed mediating factor is the conversion of unresolved psychological distress into laryngeal muscle tension, supported by the fact that onset often occurs following or during periods of emotional stress. Involuntary manoeuvres, such as coughing, laughing, and breath holding, usually elicit full glottal closure, while the vocal cords fail to adduct fully on voicing tasks. Patients are usually unaware of this discrepancy. There may be a total loss of voice (aphonia), or a range of dysphonic qualities that are incongruent with normal laryngoscopic findings, the most common of which is weak, breathy, and falsetto. Many patients report globus sensation; in rare cases, there may be a habitual cough.

Symptom incongruity is the main feature that distinguishes psychogenic voice disorders from MTD and other voice disorders. Yawning, coughing, and throat clearing can be elicited to confirm the potential for full vocal cord adduction, with sensitivity to the fact that the incongruity may not be apparent to the patient.

Puberphonia

Enlargement of the male larynx during puberty results in a drop in vocal pitch. Although rare, some adolescent males fail to utilize this lower register, however, and continue to speak in a high-pitched, breathy, falsetto voice, with difficulties projecting over noise or distance. Relevant causative factors may be difficulty adjusting to pubertal changes, subconscious resistance to voice change in successful soprano choristers, and psychological conversion reactions for cases that occur later in adult life. There are usually no other puberty-related health problems and puberphonia generally responds well to voice therapy, sometimes in conjunction with psychological intervention. Other treatment modalities include laryngeal framework surgery, cricothyroid denervation, and injection laryngoplasty (see Chapter 71).

Assessment and differential diagnosis

Ideally, patients with a functional voice disorder should be triaged into a joint voice clinic, where the combined expertise of a voice SLT and laryngologist will yield an accurate diagnosis, through careful, comprehensive questioning and use of specifically selected voicing tasks. Unfortunately, initial referral information on the nature of the voice difficulty may be brief and patients may be assessed first in a general ENT clinic.

It should be clear from the referral and medical history whether the voice symptoms are self-contained or part of a broader neurological picture, thereby obviating potential diagnostic confusion (e.g. between the breathy voice quality of a psychogenic voice disorder and Parkinson's disease). The exception, as mentioned above, is with motor neuron disease, where voice and swallow changes may indicate the presence of underlying disease that is as-yet undiagnosed. Other organic pathology should also be ruled out; the clinical and objective assessment of dysphonia is set out in Chapter 47.

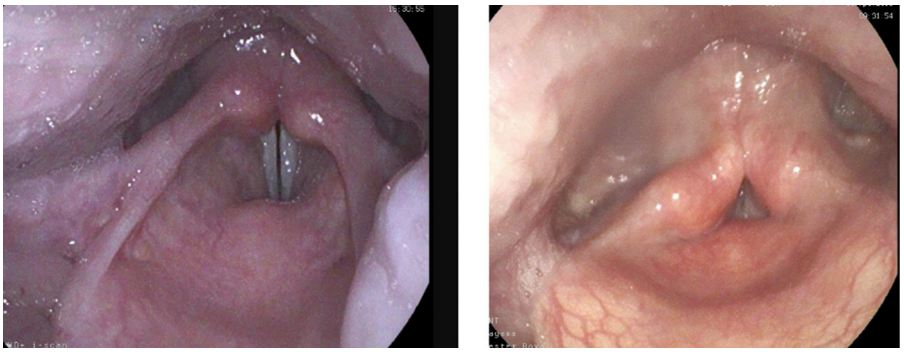

Figure 72.2 Transnasal fibreoptic images of muscle tension dysphonia depicting laryngeal constriction in an anterior–posterior dimension.

Laryngoscopic examination should reveal phonatory patterns to explain the voice disorder (see Chapter 47). Nasendoscopy is preferable to rigid stroboscopy, to facilitate a greater variety of voicing tasks.

Management

Patients should be offered referral for specialist voice therapy and reassured that their symptoms should resolve with a programme of re-establishing normal voice patterns. Patients in vocal crisis, with severe voice and throat symptoms, may need to be signed off work for a short period of time to enforce a period of voice rest.

Where a psychogenic voice disorder is suspected, patients should be referred urgently to a voice specialist/SLT with expertise in this area. Identification of causative psychological factors is no longer a requirement for the diagnosis of conversion symptoms, but emotional stress and trauma are common precipitating factors that must be carefully and sensitively explored by an experienced voice clinician. Treatment will typically involve a course of combined cognitive behavioural therapy and voice therapy, or parallel work with a voice therapist and cognitive behavioural therapy practitioner.

Conclusion

Changes in voice, breathing, swallowing, and throat sensations may be caused by a disruption to normal laryngeal function. Disruption may result from a cycle of laryngeal hypersensitivity and hyperreactivity, neurological changes affecting laryngeal movement, or from the muscle tension and imbalance seen in functional voice disorders. ENT specialists play a vital role in identification of these conditions, to achieve an accurate diagnosis and early intervention, making prompt onward referrals as necessary.

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SECTION 5
Paediatrics

73. **Assessment of the neonate, infant, child, and adolescent** *561*
Hasnaa Ismail-Koch and Andrea Burgess

74. **Acute airway management and paediatric stridor** *570*
Sean E. Evans and Sanjay R. Parikh

75. **Snoring and obstructive sleep apnoea** *580*
Kate A. Stephenson and Michael Kuo

76. **Sore throat in paediatric patients** *588*
Alex Millington and Nico Jonas

77. **Rhinorrhoea and epistaxis** *596*
Sujata De and Ravi Sharma

78. **Blocked nose** *608*
Neil K. Chadha

79. **Paediatric dysphonia** *613*
Eaodaoin O'Cathain and Scott Rickert

80. **Paediatric dysphagia** *624*
Michelle Wyatt and Vicky Thorpe

81. **Paediatric neck masses, cysts and sinuses** *630*
Haytham Kubba and Tash Kunanandam

82. **Paediatric drooling and salivary glands** *638*
Anna Harrison and Neil Bateman

83. **Paediatric hearing loss** *643*
Jaya Nichani, Adam Walker, and Iain A. Bruce

84. **The painful and discharging ear in children** *650*
Ricky Pal and Mark Felton

85. **Dizziness in children** *659*
Alok Sharma and Gundula Thiel

86. **Acquired paediatric airway stenoses and congenital laryngotracheal disorders** *665*
Ben Hartley and Claire Frauenfelder

87. **Tonsillectomy and adenoidectomy** *675*
Keith G. Trimble

88. **Otitis media with effusion** *681*
Mat Daniel

89. **Cleft and craniofacial abnormalities** *685*
Louisa Ferguson

90. **The syndromal child** *691*
Ravi Thevasagayam and Sanjeev Gupta

73

Assessment of the neonate, infant, child and adolescent

Hasnaa Ismail-Koch and Andrea Burgess

Introduction

The key to successful paediatric ear, nose and throat (ENT) assessment lies in understanding that children are not just small adults. Their communication abilities, developmental stage, family dynamics, and unique physiological considerations all influence how we approach their care. This holistic perspective ensures we don't miss important factors that could impact assessment and subsequent management of ENT problems.

Optimizing the clinical environment

Creating the right environment is fundamental to successful paediatric ENT assessment. Consider telephone and virtual consultations when physical examination isn't immediately necessary and adequate visual documentation can be provided remotely (**Figure 73.1**). This approach not only accommodates busy families but proved essential during the COVID-19 pandemic and will remain valuable for future healthcare disruptions.

The clinic room should:

- be large enough for the child and carers
- be friendly and non-threatening with instruments covered where possible so as not to frighten or distract children or accompanying siblings
- have a small collection of wipe-down toys and a play mat for babies
- have a waiting play area that is large and playful for easy distraction during prolonged waits
- include an assistant who is comfortable engaging with children and can help with examination or distract the child or siblings, allowing the parent to focus on the consultation
- a system to ensure long waits are kept to a minimum.

Tips and pearls

Everyone will develop their own style of communicating with children and carers but basic principles are as follows:

- Start the conversation with some casual conversation to establish rapport with parents and, where possible, the child.
- Establish who is with the child and what relationship they have with them. Be aware that only parents and legal guardians have the ability to give consent for any surgical procedure.
- Although parents will generally speak for younger children, talking to the child will improve engagement and can give you information regarding their level of communication.
- Adolescents may require a consultation without parents to encourage disclosure and avoid parents misrepresenting them. *If so, they may also require a chaperone.*

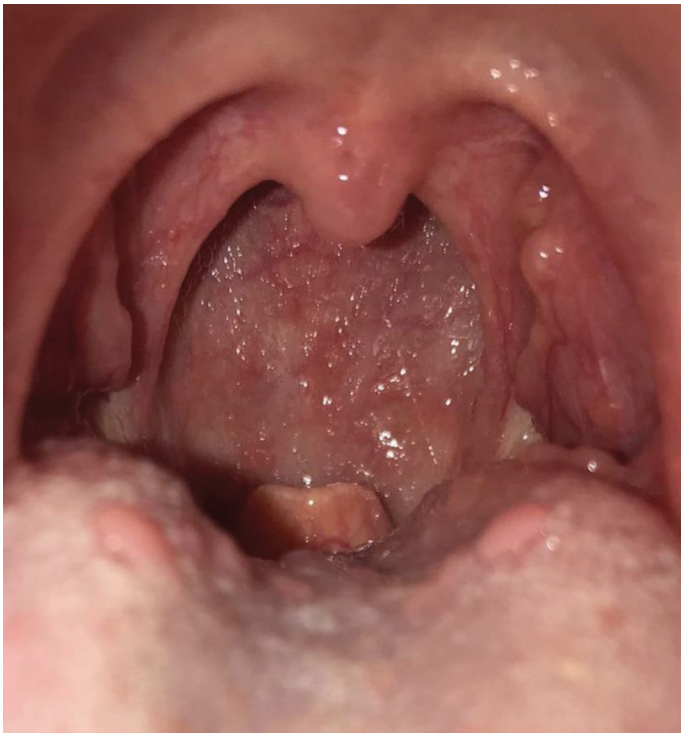

Figure 73.1 Photograph of a child's tonsils obtained and uploaded by

The history

- Main problems presenting to the ENT clinician will be about breathing, feeding, hearing, speech, communications, and infections.
- When asking about the ENT problems related to breathing, feeding, hearing, speech, communication, and infections, focus on how these are impacting their growth, well-being, and development.
- It is important to consider birth history, particularly in those under 3 years old. This includes prenatal problems, scanning abnormalities, polyhydramnios, low birth weight, birth trauma resulting in hypoxia, neurodevelopmental issues, periods of intubation, special care baby unit and neonatal intensive care unit stays, poor feeding, enteral feeding (nasogastric tube, nasojejunal tube), gastro-oesophageal reflux disease, and prematurity predominantly impacting airway and development.
- Ask about developmental milestones and any communication or behavioural issues contributing to whether they have reached an age-appropriate developmental level (Table 73.1 details developmental milestones). This is particularly important in younger children.
- Ask about immunization history. It is important to appreciate that, typically, normal healthy children have an immature immune system and can have up to 11 upper respiratory tract infections per year depending on age, particularly if attending daycare or surrounded by siblings. Box 73.1 suggests those paediatric patients who should be considered for further investigation.
- Ask about newborn hearing screening.
- Ask about current medications and document any drug or other allergies (e.g. environmental, food).
- Ask about social history including whom they live with. Ask if they like school. What do they like doing at school? Ask if older children have any hobbies or even career ambitions.
- Ask about family history of congenital/hereditary illness. Family history of the problem the child is presenting with may also influence the reason they are here or their expectation of the consultation.

Table 73.1 Language, communication, social, and motor milestones in children

Age	Language and communication	Social	Motor
1 month		• Endogenous smile	• Reacts to pain
4 months		• Preferential social smile	• Rolls over supine to prone • Sits with support • Follows objects to past midline
6 months	• Babbles	• Stranger anxiety	• Rolls prone to supine • Sits well unsupported • Transfers objects between hands
1 year	• Uses 10 words	• Parental dependence • Separation anxiety	• Walks • Ascends stairs • Kicks/throws a ball • Stacks 3–4 cubes
2 years	• Two-word sentences • Uses 250 words • More easily understood by parents	• Self-centred • Imitates others • May exhibit aggression • Favourite word is 'no'	• Walks backwards • Descends stairs • Runs • Can turn doorknobs/unscrew lids • Able to aim thrown ball • Stacks 6 cubes • Scribbles with crayons
3 years	• Talks in complete sentences • Uses 900 words • Understood by strangers	• Group play • Gender identity • Understands sharing	• Rides tricycle • Catches ball with arms • Stacks 9 cubes • Unbuttons buttons
4 years	• Can tell a story • Uses plurals • Speaks in compound sentences	• Role play • Nightmares (fear of monsters) • Imaginary friends	• Hops on one foot • Throws overhand • Brushes teeth • Copies rectangle
5 years	• Asks the meanings of words	• Conformity to peers • Feelings towards others	• Complete sphincter control • Catches ball with 2 hands • Partially dresses self • Copies a square
6–12 years	• Vocabulary expands • Able to differentiate between right and left	• Joins in teams • Follows rules	• Rides bicycle • Gains athletic skill • Skips with alternating feet • Ties shoes • Draws recognizable images
12+ years	• Communication becomes focus of relationships • Develops personal speech patterns	• Identity important • Organized sports may diminish • Cross-gender relationships	• Adolescent growth spurt (girls before boys) • Onset of sexual maturity (10+ years)

Box 73.1 Children who should be considered for immunological investigations

- ≥4 or more new severe ear infections within 1 year.
- ≥2 or more serious sinus infections within 1 year.
- ≥2 months of antibiotics with minimal effect.
- ≥2 pneumonias within 1 year.
- Failure to grow and gain weight as expected.
- Recurrent deep skin and organ infections causing collections.
- Persistent oral thrush or skin fungal infections.
- Intravenous antibiotics required to clear infections.
- ≥2 deep-seated infections including septicaemia.
- A family history of primary immunodeficiency.

The observation

It is well worth spending time observing the child and parents and how they engage with each other and those around them. You also can gauge how successful the examination will be and how helpful parents will be in supporting this. Observation of the carer–child interaction and doctor–child interaction may give insight into any communication or social difficulties. If a child is restless and refuses eye contact, it may be that they are tired, hungry, or just frightened of being in a strange place. It may, however, be suggestive of a significant communication disorder such as autistic spectrum disorder or attention deficit disorder (Table 73.2).

The child may have a physical sign aiding diagnosis such as stridor or tracheal tug suggesting work of breathing. Breathing, feeding, hearing, speech, and communication can be assessed by observing the child in clinic. It is also worth encouraging the parents to bring video clips of the child during episodes that concern them such as possible obstructive sleep apnoea during sleep or choking while feeding. It is also important to be vigilant regarding signs of maltreatment/neglect (Table 73.3) and beware of local safeguarding and child protection responsibilities and procedures if this is suspected.

Table 73.2 Behavioural traits suggesting issues/autism (in ages 12–24 months)

Behaviours	Hearing loss	Developmental delay	Autistic spectrum disorder
Eye gaze not appropriate			*
Inappropriate expression with gaze			*
Rarely shares joy or interest			*
May not respond to name	*		*
Does not coordinate expression, gestures, and sounds			*
Lack of engagement			*
Unusual vocal/verbal responses	*		*
Repetitive/recurrent movements and posturing			*
Repetitive movements with objects			*
Rarely responds to cues		*	*
Does not point		*	*
Rarely voicing consonants	*	*	*
Paucity of playing with variety of toys in typical way		*	*

Table 73.3 Signs that may suggest maltreatment

Children at increased risk of maltreatment	• Children not brought to medical appointments • Younger and disabled children (higher risk) • Risk factors present among caregivers, particularly domestic violence, alcohol and/or drug or mental health conditions
The explanation for the bruising does not marry with the clinical presentation	• Bruising in a child who is not mobile unaided • Multiple bruises or clusters of bruises • Bruises that are similar in dimensions • Bruises on non-bony parts of the body or face including the eyes, ears, and buttocks • Bruises on the neck that look like an attempted strangulation • Bruises on the ankles and wrists that look like ligature marks
Non-accidental injury (NAI) in the head and neck	• Left-sided bruising (most perpetrators right-handed) • Fractures • Burns • Abrasions, lacerations
NAI affecting the ear and hearing	• Laceration to the pinna, laceration to the anterior meatal wall • Recurrent perforations of the tympanic membrane • Recurrent bleeding from the ear (no cause found) • Bruising of the ear • Rarely hearing loss (trauma)
NAI affecting the oropharynx and nasopharynx	• Mucosal pharyngeal injury/oropharyngeal bleeding • Palatal petechiae/lacerations (may be a sign of oral sexual abuse) • Extensive dental caries (may be a sign of neglect) • Unexplained epistaxis in an infant (asphyxia should be included in the differential diagnosis)

The examination

Younger children with concerns regarding failure to thrive should have their weight and height assessed and plotted on growth charts (Figure 73.2). Parents may also bring their previous growth records (e.g. in the UK this is usually documented in what is referred to as the 'red book') for comparison. Evidence of failure to thrive aids decision-making in many paediatric conditions. Obesity is also a growing concern and impacts decision-making particularly in airway surgery.

Explain the examination first in a soft, warm, reassuring voice. Some children will also need to see and even feel the examination instruments. Small children may prefer to sit on a carer's lap for both comfort and to aid examination. Using the auroscope examine the ears; the view of the canal and drum can be optimized by pulling the pinna backwards and down (Figure 73.3a,b). Wax can be gently swept to the side using a wax hook guided by a headlight. This is often the best way of clearing wax in younger children as they struggle to tolerate microsuction on a bed. Nasal airflow can be evaluated using a metal spatula (Figure 73.3c,d), mirror, or cotton wool test. Younger children will prefer anterior rhinoscopy by gently pushing

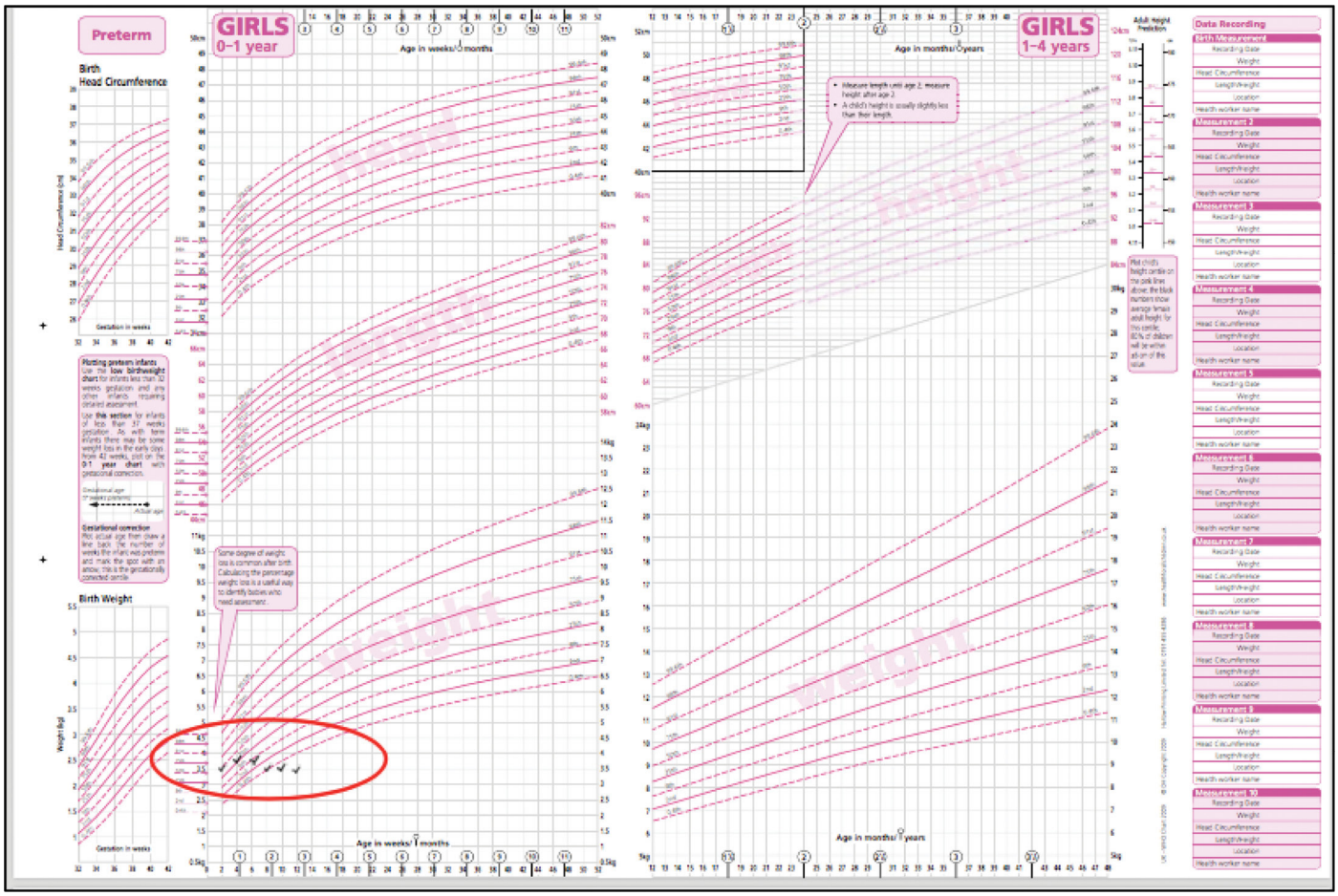

Figure 73.2 Growth chart for a female infant who had laryngomalacia with failure to thrive (showing decreasing weight and falling centile).

the nasal tip upwards and illuminating the nose with the otoscope or headlight. Older children may tolerate a Thudichum speculum and even nasoendoscopy. Oral examination may be satisfactory with or without a tongue depressor. Neck examination in younger children is sometimes easier with the child lying supine across a carer's lap with the head just falling off the lap into extension.

Invasive examinations

With parental support and persuasion, babies and young children will agree to undertake more invasive procedures such as flexible laryngoscopy and nasal cautery but it is important to monitor the situation and stop if the child becomes too distressed and reward positively for trying. They may require gentle restraining for nasolaryngoscopy and functional evaluation of swallowing, this must be done with parents' consent and preferably with their physical support to reassure the child. Try and video record the findings to avoid repeating where possible. Children below 12–18 months old can easily be examined while being swaddled. After this age it is challenging to examine until they reach an age where they will willingly participate which varies from 3 years to adulthood. Some countries use papooses for restraining different age groups but they have not gained popularity in the UK.

It is also important to give praise after examination (consider stickers) to reinforce consultation as a positive experience.

Complex children

Some children with special needs and multiple comorbidities are more challenging in their history and examination. It is important to be sensitive to their specific needs. The involvement of a paediatrician coordinating their overarching care and allied healthcare specialists (i.e. specialist nurses, speech and language therapists, physiotherapists, psychologists, etc.) and the parents making decisions together in a multidisciplinary setting can be invaluable.

The decision-making and consent

The child and carer should be informed that the examination is now finished so that they can relax and focus can now be given to an explanation of the findings and decision-making. This decision is made by the carers for younger children, but in older children it is important to ensure that their voice is also being heard. One has to decide what capacity the child has to make up their own mind. Young adults have the ability to agree or refuse treatments without parental consent if they can show they have the capacity to understand the treatment, the options and the consequences of such treatment (known as 'Gillick' competence). In younger children if parents disagree with a management plan then it is better to involve

Figure 73.3 (a, b) Examination of the ear (facilitated by gently pulling the pinna backwards and downwards). Gaining rapport (c) and using a metal spatula to evaluate nasal airflow (d) (detected by misting).

the wider team. Mediation may prove helpful and should be offered in contentious cases, as this may empower carers. Hospital ethics committees can help in providing balanced opinions—weighing up the benefits and negative effects of treatment options. This is particularly important with complex or premature babies where a multidisciplinary team should decide the appropriate level care for the child with careful consideration given to the carers' opinion.

If shared decision-making is not possible and no agreement can be reached between the clinicians and the parents then it is for the courts to decide what is in the child's best interests. The best interest test currently forms the foundation and is determinative in such contentious cases.

Neglect and withholding treatment can be considered a form of abuse, and all team members should have undergone safeguarding training and understand how to escalate concerns to the appropriate safeguarding officer.

If a child does not attend an appointment or was not brought to clinic it is also important to inform the wider healthcare team and the general practitioner about this, and to inform the social worker/health visitor to ensure there is not a pattern of this across specialties or services.

Ensure adequate information is given to the carer appropriate to their level of understanding and also adequate time. It may be that they do not feel confident to make the decision immediately or cannot as they are not primary carers, such as grandparents helping out by bringing the child to the appointment (**Box 73.2**).

Assessment of the seriously ill 'sick' child

Sick children should have combined care with a paediatrician and other specialties as appropriate. Rapid and efficient clinical assessment is required when faced with a seriously ill child, with a structured approach (**Box 73.3**). Multidisciplinary multiprofessional input is vital and the wider multidisciplinary team should be involved in treatment decisions, together with the carers. If intervention is required, depending on the severity, specialist senior anaesthetic opinion and paediatric intensive care opinion should be

Box 73.2 Key points in the UK law about paediatric consent

- A mother will always have parental responsibility for her child (unless adopted).
- A father has parental responsibility if:
  - married to the mother at the time of the child's birth
  - parental responsibility agreement, court order, or residence order is in place
  - appointed as the child's legal guardian.
- Since 1 December 2003, if named on the birth certificate a father has parental responsibility (regardless if married to the mother). Others may acquire responsibility by:
  - residence/adoption order
  - appointment as guardian.
- The local authority can acquire consent by:
  - emergency care order
  - court order.
- Some responsibilities can be transferred by the mother to another individual under Section 2(9) of the Children's act 1989.
- 'Gillick Competence' applies to a child under 16 years who is deemed to have sufficient understanding of what an intervention involves and as such can consent for themselves.
- A person with 'parental responsibility' can consent if a child aged 16–18 years is not felt to be competent.
- If there is conflict between the clinicians' and parents' view, it is for the courts to decide in the best interests of the child.

sought early and is often required as is careful pre-, intra-, and post-operative planning.

Assessment of the child with upper airway obstruction

The thorough evaluation of a child's airway is dependent on the degree of urgency and whether the child is able to maintain their own airway. Rapid and efficient multidisciplinary assessment of a child in extremis may be warranted in the emergency situation to avoid death. Examination of a child in this situation may be limited to pertinent history and with no or a very minimal examination.

The nature of the airway noise is important in establishing level of airway obstruction, although this may in some cases be present at several levels. *Stertor* is a low-pitched inspiratory noise that most closely resembles the sound of nasal congestion experienced with a cold or a snoring sound; it results from vibration of the pharyngeal tissues due to upper airway obstruction at the level of the nasopharynx, oropharynx, and/or palate. *Stridor* is a high-pitched sound that results from turbulent airflow through an airway that is partially obstructed or narrowed. Inspiratory stridor is usually as a result of airway obstruction at the supraglottic or glottic level. However, biphasic stridor (occurring on inspiration and expiration) denotes a narrowing of the airway at the glottic, subglottic, and/or cervical trachea. This may occur at one level or multiple levels. *Wheezing* is an expiratory noise often described as 'musical' caused by bronchial narrowing.

Work of breathing is a useful indicator of the severity of airway compromise. Stridor—in particular, biphasic stridor—suggesting a subglottic narrowing raises the level of concern. Head bobbing, nasal flaring, opisthotonic posturing, tracheal tug, subcostal recession, sternal recession, the use of accessory muscles, and latterly cyanosis

Box 73.3 A structured approach to evaluate the seriously ill child

Rapid clinical assessment

Airway (A) and Breathing (B)

- Effort and efficacy of breathing.
- Respiratory rate.
- Use of accessory muscles.
- Chest movements and symmetry.
- Additional sounds: stridor, wheeze, grunt.
- Breath sounds.
- SpO_2 in air.

Circulation (C)

- Heart rate.
- Pulse volume; peripheral, central (carotid/femoral).
- Blood pressure.
- Capillary refill time.
- Skin colour and temperature.

Disability (D)

- Conscious level.
- Posture.
- Pupils.

Exposure (E)

- Fever.
- Skin rashes, bruising.

'Don't ever forget glucose' (DEFG)

- Blood glucose measurement.

Actions

- Assessment should take less than 1 minute.
- Treat and reassess.
- Give clear explanations to parents and child.
- Allow parents to remain with child as much as is safely possible.

Adapted from the Advanced Paediatric Life Support Guidelines (2016).

indicate significant levels of airway obstruction. It is of the utmost importance to recognize the tiring child and differentiate the child in whom the respiratory rate is slowing down due to tiring from the child who may be improving following intervention and treatment.

Key points required from the history include:

- a history of previous intubations/airway interventions
- a history of recurrent croup
- a history of rapid stridor
- a history of new-onset drooling
- abnormal voice and cry
- failure to thrive/feeding difficulties
- cough, choke, cyanosis
- birth trauma
- a history of foreign body inhalation/ingestion
- a history of other comorbidities including respiratory, cardiovascular, neuromuscular, neurological, endocrine/metabolic, and genetic or syndrome related.

Key features when examining the child include:

- dysmorphic/syndromic features
- working hard (little reserve)
- biphasic stridor

- neck swelling
- cutaneous haemangiomas
- congenital head and neck malformations.

Assessment of the hypovolaemic child

Three stages of shock are described: compensated (early), uncompensated (late), and irreversible.

Assessment of shock

During compensation, the sympathetic reflexes divert blood away from non-essential tissues and the venous reservoir is mobilized with increased peripheral arterial resistance. Children are very good at compensating and, as such, vital signs in compensated/early shock may be subtle. In the early stages there may be mild tachypnoea and tachycardia. The blood pressure may be normal in early compensated shock, and notably a low blood pressure does not occur until late shock in children. Tachycardia itself may represent a non-specific sign of distress.

Examination findings in compensated (early) shock include:

- dry mucous membranes
- warm or cool skin
- palpable central pulses
- decreased or bounding peripheral pulses
- slightly delayed/poor capillary refill (3 seconds)
- irritable/unconsolable
- not reacting with parent/carer
- 'staring into space'
- poor response to pain
- decreased urine output.

Once uncompensated shock ensues, vital signs reflect the inability to maintain perfusion and comprise tachycardia, tachypnoea, hypotension, agitation, confusion, decreased level of consciousness, poor tone, tacky mucous membranes, cool and mottled extremities, decreased pulses, and delayed capillary refill (>4 seconds). Hypotension is a late sign in children with hypovolaemia and indicates greater than 30–40% volume loss (Table 73.4).

Hypovolaemic shock

Hypovolaemic shock is the commonest form of shock that occurs in children and is caused by a reduction in the intravascular fluid volume. Reduction of the intravascular fluid volume causes a decrease in stroke volume due to the resulting decrease in preload. The decrease in preload impairs cardiac output, leading to inadequate delivery of oxygen and nutrients to the tissues and organs (shock).

The loss of intravascular fluid volume which results in hypovolaemic shock can have a number of causes, such as dehydration from vomiting and diarrhoea, haemorrhage, decreased intake of fluids, and so on. When managing the child with hypovolaemic shock, examination necessitates the need for an estimated blood volume calculation. According to the Advanced Paediatric Life Support (APLS) guidelines, the weight of children between the ages of 1 and 5 years can be safely estimated using the formula: weight = (age × 2) + 8, and for children older than 5 years, weight = (age × 3) + 7.

Estimated normative blood volumes by age group are as follows:

Neonates	85–90 mL/kg
Infants	80 mL/kg
Children	75 mL/kg
Adult	70 mL/kg

Table 73.4 classifies the vital signs of shock by blood volume loss in children.

Currently there are several apps readily available that can be downloaded and websites to help provide this information, normative values, and parameters for children in an emergency.

Continual monitoring and reassessment of the child, especially once treatment is commenced, is vital in order to ensure an improving overall picture.

Assessment of the septic child

There is a rising incidence of severe sepsis in children and ENT infections may result in sepsis. Early recognition and prompt treatment of sepsis leads to decreased morbidity. It is important to have a low threshold for suspecting sepsis as the clinical presentation may be non-specific, especially in younger children. Given the time-critical nature and rapid progression of sepsis it is usually best to initiate investigations and start treatment promptly. The Paediatric Sepsis Six provides a guide to recognizing the signs and management of sepsis (Figure 73.4).

Table 73.4 Clinical signs of haemorrhagic shock in children with varying degrees of blood loss

Clinical signs	% Blood volume loss			
	<15	15–25	25–40	>40
Heart rate	Normal/minimal increase	Slight increase	Increased	Increased
Blood pressure	Normal/increased	May be decreased	Decreased	Decreased
Respiratory rate	Normal	Mild tachypnoea	Moderate tachypnoea	Severe tachypnoea
Mental status	Anxious	Anxious/agitated	Anxious/confused	Confused/lethargic/unresponsive
Capillary refill	Normal	Prolonged	Prolonged	Prolonged
Urine output	Normal	Normal/decreased	Decreased	Absent

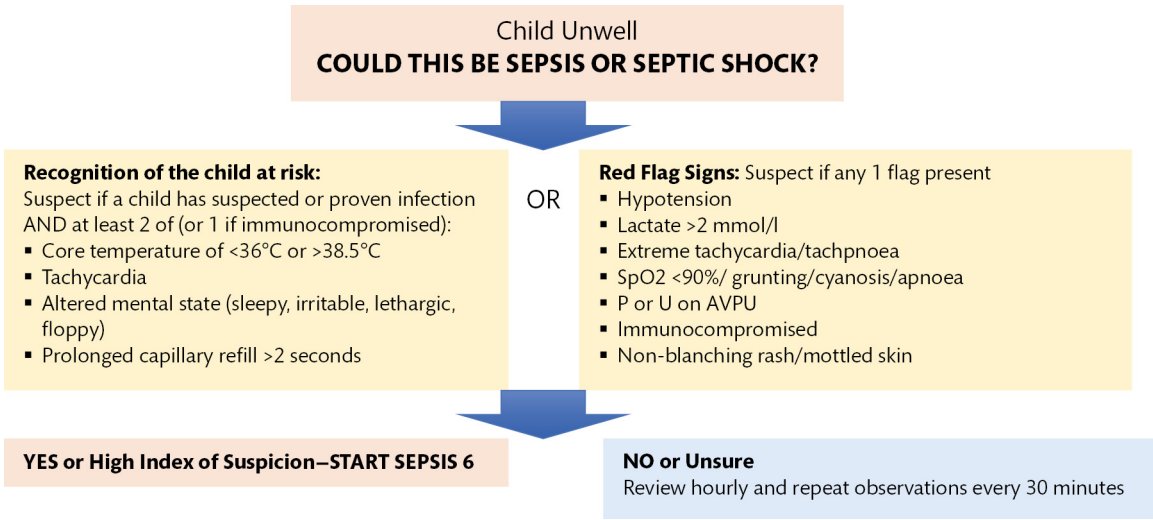

Figure 73.4 Paediatric Sepsis Six recognition and signs.
Source data from the Great Ormond Street Hospital for Children Sepsis 6 form & Paediatric Sepsis 6 version 11.1 August 2015 in collaboration with the UK Sepsis Trust Paediatric Group.

Assessment of the preoperative child

Good perioperative care is fundamental in the pathway towards better surgical care. This care can be significantly optimized by consultant-led paediatric pre-assessment, which can be cost-effective, reduce cancellations, and improve both parent and child satisfaction. Although the majority of children are ASA I and II using the American Society of Anesthesiologists (ASA) physical status classification system, with increased survival rates for neonates, more children are presenting for surgery with complex comorbidities. Pre-assessment allows shared decision-making, thorough anaesthetic and specialist assessment, planning for the complex child, consenting, providing reassurance, improving compliance, and managing anxieties prior to the day of treatment.

Perioperative support

It is not uncommon, especially when an investigation, treatment, or surgery is required, for a child to become uncooperative, especially at the induction of anaesthesia. Stress of hospitalization in children has been ascribed to five general fears: separation from main caregivers, the environment, painful procedures, the operation, and/or the anaesthetic. As such, it is important to be aware of strategies that may help manage these fears and facilitate treatment.

Communication is vital at an appropriate level of understanding for each child and it is important to consider developmental differences when managing children.

Psychological or pharmacological interventions such as sedation may reduce anxiety and stress, improving compliance and the treatment experience.

Frequent attenders and children with psychological, developmental, or behavioural disorders are more likely to require more support and preoperative planning from the outset with active engagement of the carers and staff in formulating a pre-emptive plan.

Perioperative setting for frequent flyer children—Positive Outcome and Experience Management Strategies (POEMS)

Anxiety in children frequently attending can be a challenging problem and there is a link between preoperative anxieties with adverse clinical outcomes (e.g. emergence delirium, increased analgesic requirements, negative behavioural changes, and bed wetting). POEMS focuses broadly on three pre-induction categories to help better manage children preoperatively:

- Sedative premedication.
- Parental presence at induction of anaesthesia.
- Behavioural intervention.

Pre-hospital programmes

Pre-hospital programmes including hospital visits, videos, books, apps, and so on may reduce anxiety on admission. These strategies based on behavioural modelling provide the child with knowledge, which aids coping mechanisms. Research has shown that visits are more effective if undertaken 5–7 days prior to surgery in children older than 4 years. Interventions are less effective in the children of anxious parents or in children with developmental or behavioural issues.

Play therapy

Play therapy with an experienced therapist tailored to the individual child's needs can prove extremely beneficial and experienced therapists may additionally be able to identify children in whom premedication will likely be needed. Specialist psychology support is available in some centres for children and families adjusting to a diagnosis, managing treatments, preparing for surgery (in particular, children who develop major psychological problems around

surgery), managing uncertain outcomes, managing feeling different to others, and living as well as possible alongside a medical difficulty, and often involves working together with parents to ensure that they feel as confident as possible managing their child's difficulties.

Conclusion

Consultation requires special understanding of the child's needs for engagement and the carer's expectations:

- Awareness of age-appropriate developmental stages particularly around language and communication.
- Recognition of special needs and how they impact the success of the consultation and the underlying health issues.
- Recognition of abnormal dynamics and safeguarding concerns.
- Understanding of consent.
- Assessment of the sick child and an understanding of appropriate pathways to escalate their care.

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74

Acute airway management and paediatric stridor

Sean E. Evans and Sanjay R. Parikh

Introduction

Acute paediatric airway compromise is characterized by active or impending airway obstruction that requires immediate attention to prevent significant morbidity or loss of life. Impending airway collapse is a true medical and surgical emergency. Focused history, physical examination, and situational assessment may be paramount to saving a patient's life. This chapter provides an overview of the causes, diagnostics, interventions, and pitfalls to avoid in the management of a patient with acute airway compromise.

An important note should be made regarding the stress of decision-making in critical environments such as airway emergencies. The situation is often hectic, with the presence of multiple providers, ancillary staff, and family members, in addition to complex equipment in a confined space. The most critical steps to ensuring a safe patient outcome include the need for open and clear communication with all team members, preparedness for alternative measures, and collaborative decision-making (**Figure 74.1**).

Anatomy and physiology

Paediatric patients have a number of unique anatomical and physiological properties that put them at greater susceptibility to morbidity and mortality as a result of even mild airway compromise. Greater metabolic rates and immature respiratory control make this population particularly vulnerable. Infants have unique anatomical airway characteristics that contribute to physiological differences. The infant larynx is higher in the neck than in adults, typically at the level of the second cervical vertebrae, and the hyoid bone is also more anterior. The elevated position of the larynx places the epiglottis in contact with the uvula and contributes to the obligate nasal breathing seen until approximately 6 months of age. Additionally, the lack of intravertebral joints above the larynx causes the infantile larynx to not be amenable to the classic flexion–extension position for laryngoscopy, and a greater reliance is necessary on anterior cricoid pressure for visualization. The glottis is the narrowest portion of the infant airway at approximately 4 mm for a term neonate and therefore even minor obstruction can cause significant distress. The overall small size of the upper airway may make rapid diagnoses and interventions challenging.

The most common symptom and sign of paediatric airway obstruction is stridor: the audible consequence of turbulent airflow through restricted anatomy. It is classically characterized by its association with the phase of respiration and may be inspiratory, expiratory, or both. Supraglottic pathology is classically associated with inspiratory stridor, while glottic level obstruction may result in either inspiratory or a biphasic stridor as dictated by Poiseuille's law (see Chapter 70). Isolated expiratory stridor is classically caused by intrathoracic pathology.

Assessment

All patients should be hooked to cardiopulmonary monitoring with frequent (i.e. every 2 minutes) blood pressure cycles, pulse oximetry, and capnography when available. The patient should be succinctly evaluated for their overall work of breathing. In the infant population, it is important to note the presence, degree, and extent of accessory muscle use (e.g. subcostal retractions, tracheal tugging, lateral neck flaring); audible airway sounds including stridor and stertor (remembering that progressively worsening stridor resulting in quiet is impending airway collapse until proven otherwise); the presence, pitch, and volume of cry (lack of cry in an agitated paediatric patient may suggested impending respiratory failure); colour changes of the skin and lips (due to limited lung volumes and respiratory mechanics, paediatric patients may fall quickly off of the oxygen dissociation curve resulting in pallor and cyanosis); and lastly a general look of fear, posturing, and restlessness that may suggest impending doom. It is good to know the history of onset, progression, associated symptoms, potential ingestions, infections symptoms, or situational details regarding trauma when time allows.

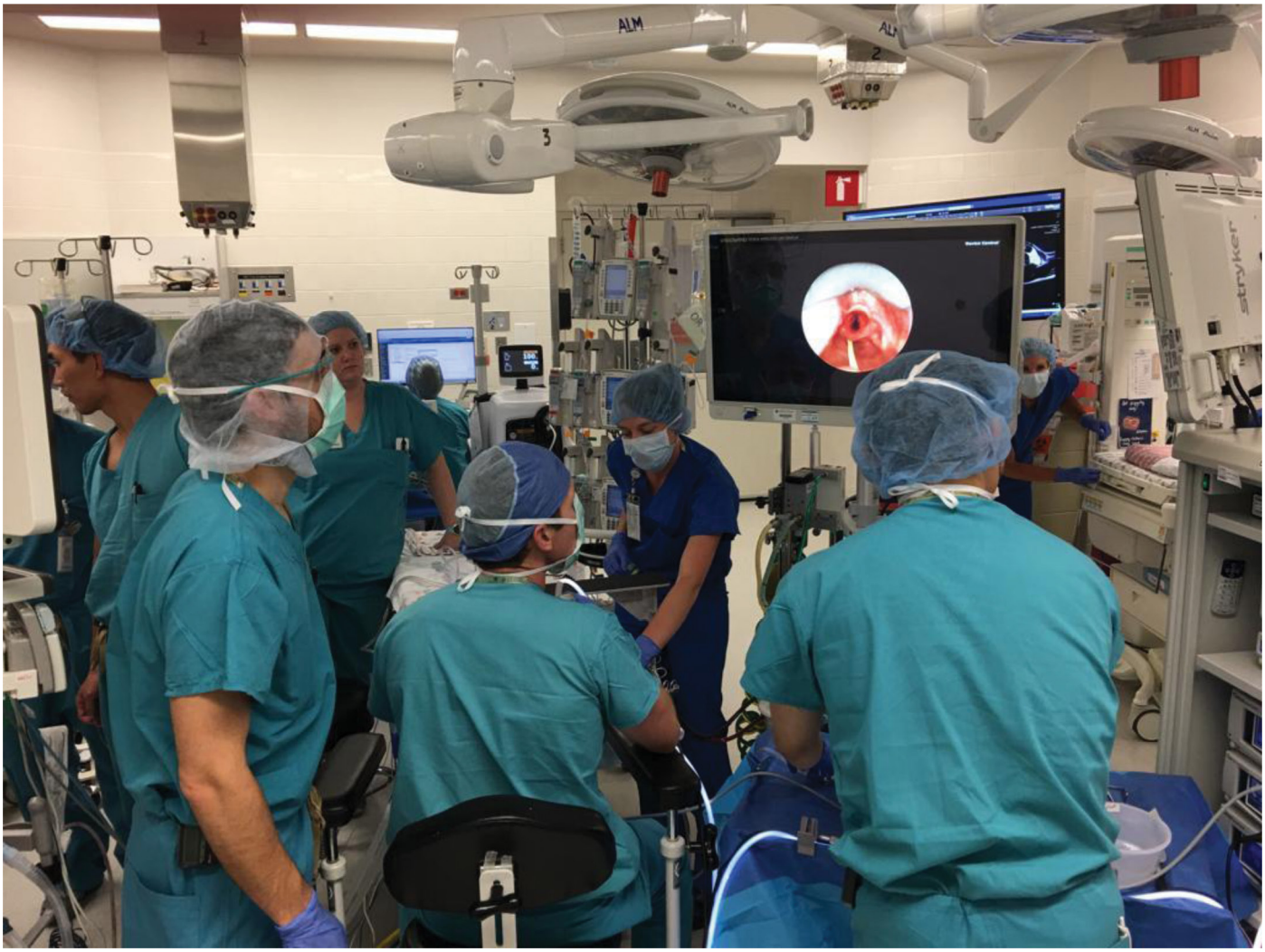

Figure 74.1 Intraoperative photograph of an urgent bronchoscopy, highlighting the hectic nature of these environments.

Investigations

If the patient is deemed to be stable enough to pursue, imaging can be extremely helpful in identifying the cause of the airway obstruction. Standard anterior–posterior and lateral X-rays can typically be done expeditiously and give valuable information about foreign bodies, soft tissue/air interfaces, and bony details. An anterior–posterior chest radiograph can provide information regarding the status of the lungs and or more distal foreign bodies that may be clinically relevant. Computed tomography scans are also readily available in most emergency departments and radiology suites and provide superior detail of soft tissue/air interfaces, particularly for trauma. The addition of contrast can dramatically improve the ability to identify bleeding and infectious aetiologies; however, in an emergent situation, intervention or airway securement should never be delayed for further diagnostic imaging. Blood work is typically reserved for presumed infectious causes of airway obstruction (i.e. complete blood count, C-reactive protein, erythrocyte sedimentation rate) in order to assess response to therapy or in the setting of trauma (i.e. type and screen) when blood replacement may become necessary. Regardless of investigational studies chosen, someone qualified and prepared to manage the airway should be with the patient until they are deemed stable.

Differential diagnosis/associated conditions

Early consideration of differential diagnoses stratified by age group may be helpful when assessing a child in distress. A handheld table of differential diagnosis may be useful, such as Table 74.1.

Neonate

Although laryngomalacia is the most common cause for airway distress in an infant, it is rarely the source for acute airway compromise. Laryngomalacia, originally coined congenital laryngeal stridor, is a condition whereby the supraglottic cartilaginous structures are

Table 74.1 Differential diagnosis of acute airway compromise by age group

Neonate	Laryngotracheal stenosis/CHAOS, vocal cysts, vocal cord paralysis, tongue base obstruction, laryngomalacia
Infant	Laryngomalacia, subglottic stenosis, haemangiomas, vascular compression, tracheobronchomalacia
Toddler	Infectious (croup, epiglottitis, retropharyngeal abscess), foreign body aspiration, acquired subglottic stenosis, caustic ingestion
Child	Infectious (mononucleosis, tonsillitis, tracheitis), foreign body aspiration, trauma, asthma
Adolescent	Infectious (peritonsillar abscess), paradoxical vocal fold motion, trauma, asthma, epistaxis

immature or malformed, leading to glottic hooding and a characteristic inspiratory staccato stridor. It is best diagnosed via awake transnasal endoscopy and demonstrates some combination of the classic triad description of an omega-shaped, furled epiglottis, tight and foreshortened aryepiglottic folds, and posterior arytenoid redundancy causing passive ball-valve hooding and obstruction of the rima glottidis with inspiration. It is a mostly self-limiting conditions, though surgery is occasionally indicated when it presents with failure to thrive, cardiopulmonary stress, or recurrent desaturations or cyanotic episodes. Surgical management is discussed at the end of the chapter. In the newborn, congenital bilateral vocal fold immobility is the most common cause of acute compromise and is associated with the signs of biphasic stridor, respiratory effort, dysphagia, and loud cry. The inability to abduct the vocal folds results in a narrowed glottic aperture. Diagnosis is typically made by flexible fibreoptic examination, and the severity of obstruction is variable based on the amount of glottic gap. The passive folds may be paradoxically drawn into the larynx by negative pressure air flow, further obstructing the rima glottidis, as dictated by Bernoulli's law. Cranial imaging is typically required to assess for associated anomalies such as Chiari malformation. Approximately 90% of cases are idiopathic, emphasizing the lack of clarity of the aetiology of this disorder.

Laryngeal webs represent a spectrum of adhesion along the medial surface of the vocal cord, starting at the commissure and extending posteriorly. Webs limit the lateral excursion of the vocal cord thereby narrowing the glottic inlet and occur more commonly in patients with midline abnormalities (i.e. velocardiofacial syndrome).

The most severe form of laryngotracheal stenosis is known as congenital high airway obstruction syndrome (CHAOS), whereby a patient is born with a laryngotracheal separation or without a cannulated glottis. The diagnosis of CHAOS is typically made a prenatally via ultrasound and confirmed with magnetic resonance imaging. Obstruction at the level of the glottis inlet causes accumulation of fetal lung fluid in the lower airways. Lung expansion and subsequent oesophageal compression may then contribute to significant dysphagia. This sequence of events leads to the findings of diaphragmatic flattening with occasional inversion, polyhydramnios, and ultimately hydrops that help distinguish and diagnose this condition. With CHAOS, there may be associated tracheo-oesophageal anomalies such as distal tracheo-oesophageal fistula that may ultimately require attention as well.

Choanal atresia resulting in partial or complete nasal obstruction is caused by embryonic failure of resorption of the nasobuccal membrane (Figures 74.2 and 74.3). Bilateral choanal atresia will present at birth with cyclical cyanosis, where the patient ventilates when crying but does not in the resting state due to nasal obstruction. Unilateral disease on the other hand may go unnoticed for years and ultimately present as unilateral nasal obstruction or rhinorrhoea (see Chapter 78).

Congenital subglottic stenosis is a condition whereby the normal shape and calibre of the infantile cricoid, the narrowest part of the infant larynx, is malformed, resulting in airway narrowing. Congenital long-segment tracheal stenosis can be caused by the development of complete tracheal rings resulting in newborn acute airway compromise (see Chapter 86).

Benign laryngeal cysts may occasionally become symptomatic enough to create a barrier to airflow obstruction and should also be considered. Obstruction above the level of the vocal cords is

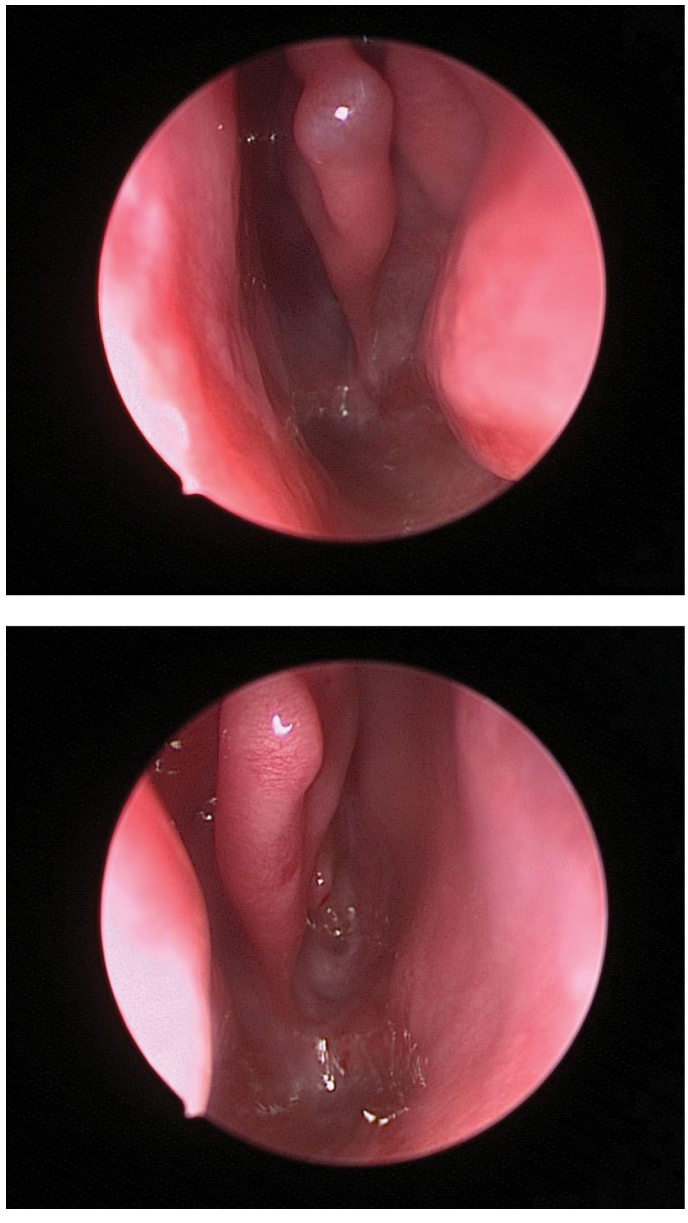

Figure 74.2 Bilateral choanal atresia in a patient with CHARGE syndrome demonstrating absence of a choanae with the nasal cavities ending in a cul-de-sac.

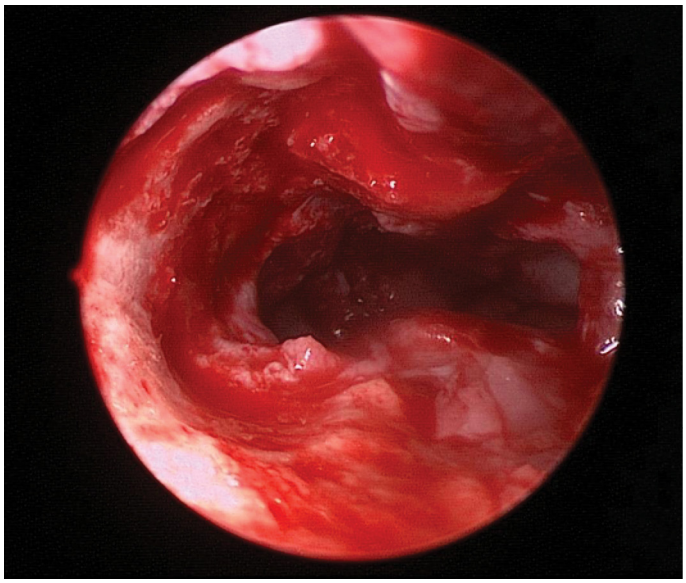

Figure 74.3 Postoperative appearance of the neo-choanae following choanal atresia repair in the same patient as Figure 74.2, demonstrating widely patent bilateral nasal airway.

often seen in children suffering from craniofacial abnormalities or micrognathia as the tongue base may fall back and occlude the oropharynx. Lastly, while laryngomalacia does not typically present with acute airway obstruction, it may be a contributing factor when multilevel pathology is present.

Infants

Infants, defined as being 1 month to 1 year of age, represent the next unique subset of patients to keep in mind when called about impending airway obstruction. While it is intuitive to believe that congenital lesions present at birth should presumably have been recognized by now, this thinking is not always accurate. As the patient grows and their metabolic needs change, previously adequate compensation around fixed lesions may no longer be adequate. Congenital laryngomalacia and subglottic stenosis must therefore also be considered in this population.

Infantile haemangiomas are typically absent at birth and then undergo rapid expansion in the early months of infancy. Patients presenting with large or segmental head and neck haemangiomas are at greater risk of having a synchronous airway lesion, with the classic location being at the left posterolateral subglottic region (Figure 74.4). This obstructive lesion may lead to acute airway compromise necessitating diagnostic or therapeutic interventions such as intubation, steroid injection, and/or systemic propranolol administration which has become first-line therapy in recent years (see Chapter 81 for a protocol).

Tracheobronchomalacia is characterized by immaturity of the lower airway cartilages causing susceptibility to collapse with inspiratory effort. Vascular compression may also result in extrinsic tracheobronchomalacia and should be a consideration in this age group. As the infant airway grows, compression secondary to fixed vascular rings or extrinsic compression may cause weakening of the walls of the airway. As the narrowed airway causes reduced airflow, inspiratory effort is increased through diaphragmatic excursion and accessory respiratory muscle use. The result is increasing

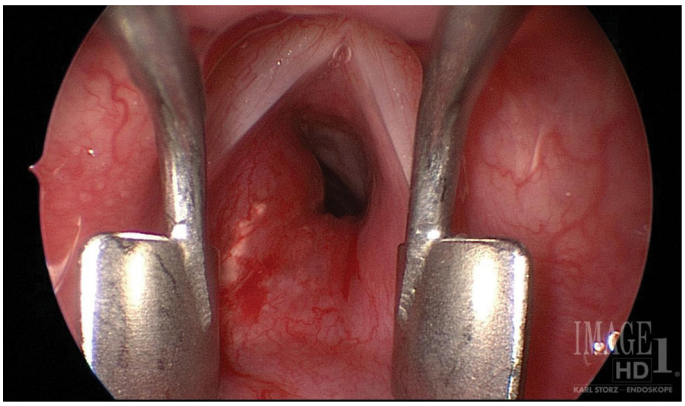

Figure 74.4 Propranolol-resistant large left posterolateral subglottic haemangioma.

negative intrathoracic pressure. The combination of a narrowed lumen and these dynamic thoracic changes may cause the anterior and posterior tracheobronchial walls to touch, resulting in a poorly understood reflex apnoea, bradycardia, and potentially cardiopulmonary collapse. These so-called dying spells are pathognomonic of tracheobronchomalacia (see Chapter 86).

Toddlers

The differential diagnosis of a toddler diverges significantly from that of an infant. One reason for this is that toddlers have typically gained mobility and may no longer be within the parent's reach at all times. Toddlers also tend to explore their world through their mouths, predisposing them to the possibility of caustic ingestion as well as foreign body aspiration. Both caustic ingestion, and foreign body aspiration are associated with significant potential morbidity and mortality, and therefore must be aggressively pursued and managed. The 'classic' triad includes a witnessed aspiration choking event, radiological evidence of air trapping and mediastinal shift, and auscultatory findings of diminished air movement unilaterally, with the right main stem bronchi being more commonly affected due to its acute angulation. In practice, few patients present with such a clear-cut picture as described in the 'classic' triad and the presence of one to two of these may merit operative evaluation.

Toddlers also tend to be at an age where social play and daycare become more common; this in turn exposes them to pathogens that may cause laryngotracheobronchitis, or croup, retropharyngeal phlegmons and abscesses, and the ever-concerning epiglottitis. While the *Haemophilus influenza* type B vaccine has dramatically reduced the number and severity of epiglottitis cases, it should still be considered, particularly in a patient with unknown immunization status. Lastly, a history of prior intubation may lead the examiner to consider the possibility of acquired subglottic stenosis from pressure necrosis, fibrosis, and scarring.

Children

Children aged 3–13 years may suffer from many of the same pathologies as toddlers, especially in their younger years. An additional consideration as the child grows, though, is the possibility of trauma related to sports, recreation, or motor vehicle accidents. These can range from subtle soft tissue trauma to laryngeal fracture and laryngotracheal separation. Severe asthma attacks may manifest as referred upper airway obstruction and may rarely or occasionally

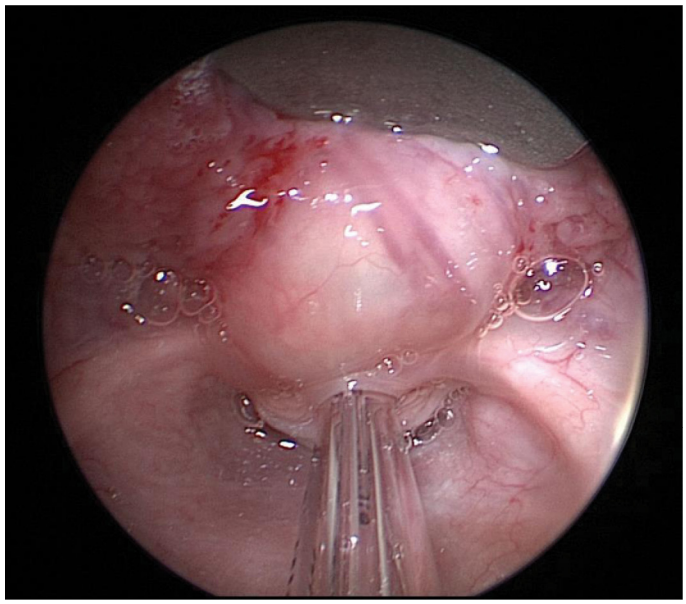

Figure 74.5 Preoperative appearance of a large vallecular cyst. Note the level of laryngeal obstruction from the craniocaudal compression of the supraglottic larynx.

prompt otolaryngological involvement. Additional infectious considerations in this age group may include tonsillitis and mononucleosis which can cause significant oropharyngeal narrowing and airway obstruction.

Cystic masses involving the vallecula may cause retropulsion and posterior displacement of the tongue resulting in upper airway obstruction. This group of lesions includes benign vallecular cysts, lingual thyroids, and vallecular thyroglossal duct cysts (Figures 74.5 and 74.6). These masses have a variable age at presentation but should be considered, especially in the presence of concomitant dysphagia.

While reported in all paediatric age groups, a recent review demonstrated bacterial tracheitis most commonly affected children at approximately 6 years of age. This uncommon infection is characterized by a pseudomembranous exudative infection of the trachea that may progress to complete airway obstruction and therefore should be considered.

Adolescents

Adolescent patients represent the last subgroup for consideration in the paediatric population. As with the toddler-to-child transition, the child-to-adolescent transition lends many of the same pathologies to the latter age group, including trauma and asthma. One unique phenomenon seen in the male adolescent population is juvenile nasopharyngeal angiofibroma. While these lesions may not cause acute airway obstruction themselves due to insidious growth patterns, they may cause life-threatening posterior epistaxis that requires emergent management (see Chapter 77).

Trauma to the larynx may also occur in the adolescent population, resulting in acute airway compromise. Typical causes of laryngeal fractures include motor vehicle accidents and sports injuries either from direct human collisions or from high-velocity projectiles (e.g. baseball, cricket, and lacrosse). Trauma to the larynx, including traumatic intubations, may also result in post-traumatic granulomas which may present with varying degrees of respiratory distress (Figures 74.7 and 74.8).

Paradoxical vocal fold motion (also termed inducible laryngeal obstruction) is a condition whereby the patient has paradoxical adduction of the vocal folds with inspiration. It is usually benign and self-limiting but can present with severe stridor. The classic presentation of inducible laryngeal obstruction is in a high-achieving adolescent with a history of reactive airway disease undergoing significant home/school/athletic stressors (see Chapter 79).

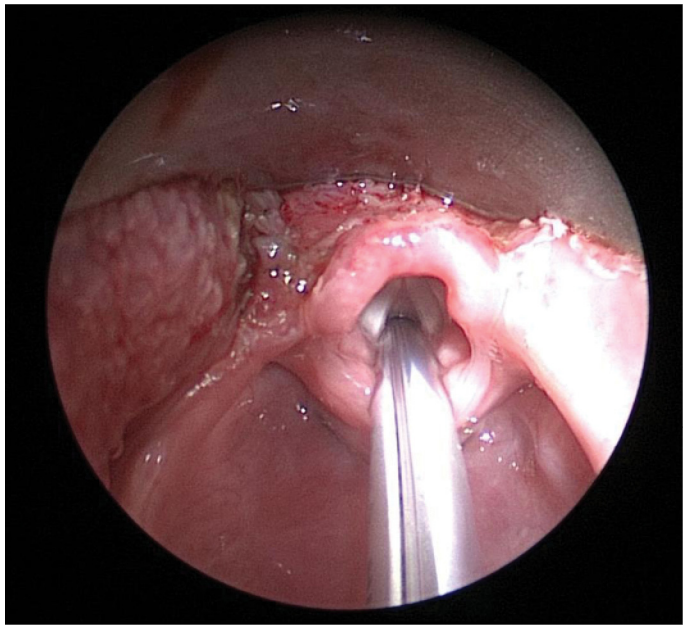

Figure 74.6 Postoperative appearance of the same patient as Figure 74.5 following microscopic dissection of the vallecular cyst. Note the dramatic improvement in laryngeal exposure.

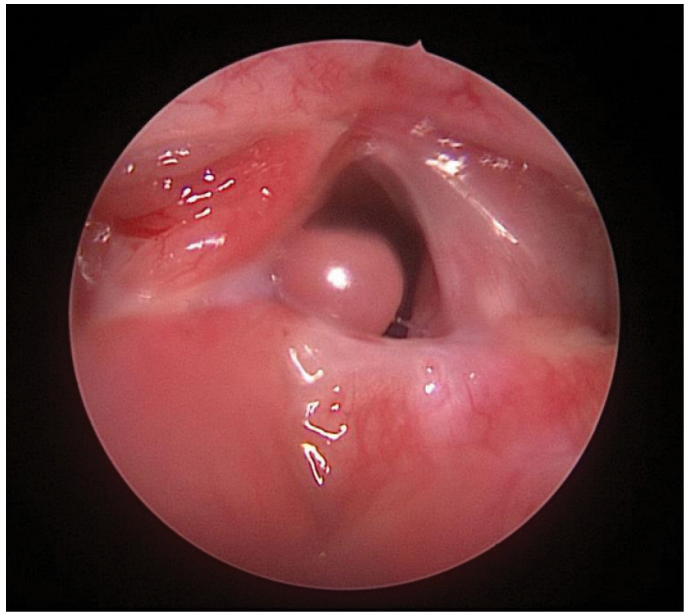

Figure 74.7 Post-traumatic laryngeal granuloma in a patient who suffered a blunt clothesline injury.

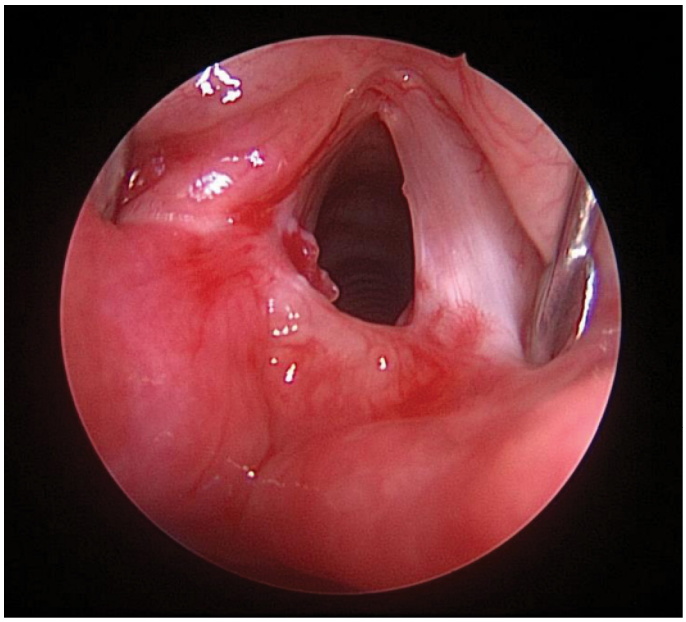

Figure 74.8 Postoperative appearance of the lesion in Figure 74.7 after removal of the granuloma. Note a small posterior glottic scar band that was also incised.

A last consideration in adolescents is the peritonsillar abscesses or deep neck space infection causing airway compromise.

The most important part of the assessment of a patient with acute airway compromise is a determination of how much time is available for diagnostics, evaluation, and intervention. Respiratory therapy, intensivists, emergency medicine physicians, bedside nurses, and anaesthesiologists may all play a vital role in securing a safe airway, managing a critical paediatric patient, and getting that patient to a safe environment.

Medical treatments

Awareness of state-of-the-art medical treatments for acute airway compromise is essential for the rapid implementation of therapy. Familiarity with the different forms of oxygen administration and non-invasive ventilation, adjuncts to ventilator support, and secretion management (e.g. suction set-up) are necessary. In addition to non-invasive ventilation, the otolaryngologist should be familiar with all potentially available intubation tools including a variety of laryngoscope blades (e.g. Miller, MAC, Wisc, Phillips), video-assisted laryngoscope devices, bronchoscopic intubation technique, and a basic understanding of the pharmaceuticals involved in achieving a safe environment for airway manipulation. In the acute setting, the most common medical agents should include high-dose intravenous steroids and nebulized racemic epinephrine (adrenaline).

Once the decision is made to secure the patient's airway, the next immediate decision will be to choose the environment in which to do so. The operating room is the preferred location to secure a tenuous airway. In the event the patient is too unstable to transfer, every effort should be made to obtain the potential tools needed to secure the airway at that location.

Surgical management

Airway securement requires a systematic plan with multiple contingencies and concise, clear communication (e.g. who will intubate, secure the tube, confirm end-tidal carbon dioxide, and assess breath sounds). Essential items to disclose on all airway consents include the potential for therapeutic interventions, typically including augmentation with rigid or balloon dilation, adjunctive injection, destruction of obstructive lesions, and the potential for surgical airway. In addition to the standard risks of bleeding and infection, the otolaryngologist should describe potential associated complications including injury to the lips, gums, teeth, tongue, or pharynx, obstruction recurrence, scarring and stenosis following intervention, and possible death.

When preparing to secure an airway in the operating room, proper planning, anticipation, and communication will optimize success. All equipment should be personally inspected by the surgeon to insure proper assembly and functionality prior to the beginning of the procedure. A full complement of operative laryngoscopes in varying sizes (e.g. Parsons, Lindholm, Benjamin, etc.) and endotracheal tubes should be readily available. Familiarity with the components of the rigid ventilating bronchoscope is a prerequisite for performing bronchoscopy on children (**Figure 74.9**).

During bronchoscopy, the child is kept spontaneously breathing and assisted with mask ventilation. The head of the bed is turned out 90° away from anaesthesia and the otolaryngologist assumes control of the airway. An age-appropriate laryngoscope is used to expose the larynx and topical lidocaine applied. A rigid telescope is then passed through the vocal cords and down the trachea to the level of the mainstem bronchi, typically obtaining pictures at the level of the supraglottis, vocal cords, subglottis, carina, and takeoff for each of the mainstem bronchi by gently rotating the head left and right to allow direct visualization (**Figure 74.10**). If there is a question regarding the calibre of the subglottic airway, a 'leak test' can be performed by inserting an uncuffed endotracheal tube and having the anaesthesiologist administer positive pressure ventilation while directly visualizing the glottis with the endoscope. A leak is identified by air or secretions moving around the tube at less than 25 cmH_2O pressure. Sequentially larger tubes are used until no leak occurs, helping to determine the calibre of the airway, as originally described by Cotton and Myers. If any airway interventions are planned, the patient can be suspended from a Mayo stand with a laryngoscope in place to allow for a two-hand technique or the use of an operating microscope.

In the case of a supraglottoplasty for laryngomalacia, the operating microscope and typically Lindholm laryngoscope are used to expose and visualize the supraglottic larynx. A microlaryngeal triangle grasper or haemostat is used to grab the ipsilateral redundant arytenoid cartilage and place the aryepiglottic fold on tension. Microlaryngeal scissors are then used to release the shortened band of tissue until the epiglottis is released forward. The redundant arytenoid tissue and even corniculate cartilage can then be sharply excised on the ipsilateral side, making sure to preserve the inter-arytenoid mucosa and maintain a small amount of mucosa between the aryepiglottic fold release and the excised arytenoid tissue to prevent potential postoperative scarring and supraglottic stenosis. The procedure is then repeated on the contralateral side and

Figure 74.9 Airway table set up with Parson's laryngoscope, 2.7 mm Hopkins rod telescope, 3.0 and 3.5 ventilating bronchoscopes, anti-fog device, suction tubing, and rubber and glass bronchoscopic diaphragms.

Afrin (oxymetazoline) pledgets are used to assist with hemostasias (Figures 74.11 and 74.12).

Suspension laryngoscopy techniques are also the standard of care for addressing obstructive subglottic pathology, such as stenosis (see Chapter 86) or haemangiomas (particularly when unresponsive to propranolol). This is often assisted by the use of a carbon dioxide laser. In the case of the airway haemangioma, the patient is suspended using the above techniques. The subglottic visualization can be improved through the use of the Lindholm laryngeal distending forceps (i.e. 'vocal cord spreader') which is secured to the suspension arm of the laryngoscope using rubber bands. This will provide an unobstructed view of the airway pathology. The carbon dioxide laser

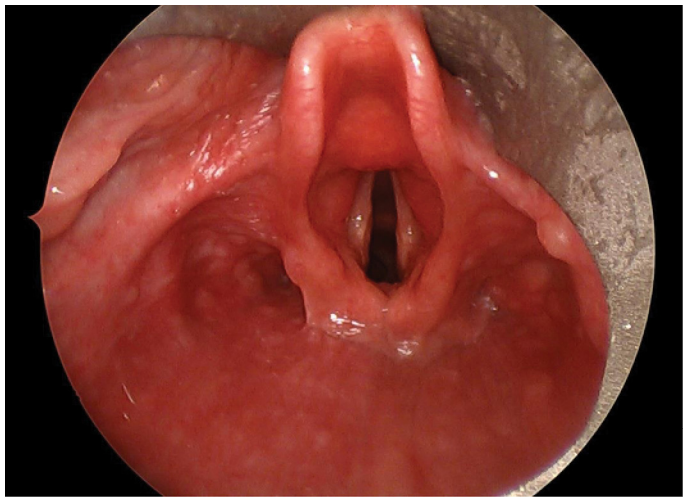

Figure 74.10 Telescopic view on direct laryngoscopy of a normal infantile larynx.

Figure 74.11 Preoperative appearance of a patient with laryngomalacia. Note the foreshortened aryepiglottic folds, omega-shaped epiglottis, and redundant posterior arytenoid cartilage.

Figure 74.12 Postoperative appearance of the patient in Figure 74.11 following supraglottoplasty demonstrating dramatically improved airway calibre with significant aryepiglottic fold release and no further hooding.

fibre can be used to directly ablate the lesion until adequate airway calibre is achieved. Additionally, steroids may be injected into the base of the lesion using the orotracheal injector to reduce recurrence (Figure 74.13).

The use of a pneumatic balloon catheter (e.g. balloon dilation) can be an additional adjunct technique to assist in establishing a capacious airway in airway stenosis (see Chapter 86). Using an endoscopic technique, the uninflated balloon catheter is placed down to the level of the airway narrowing. It is then rapidly inflated and maintained for a period of seconds to a minute depending on the patient's stability. Communication with the anaesthesiologist during this period is critical as significant inspiratory effort against an obstructed glottis could result in post-obstructive pulmonary oedema. The balloon catheter is deflated, and the airway reinspected for response. It can be repeated as necessary. In the event there is concern

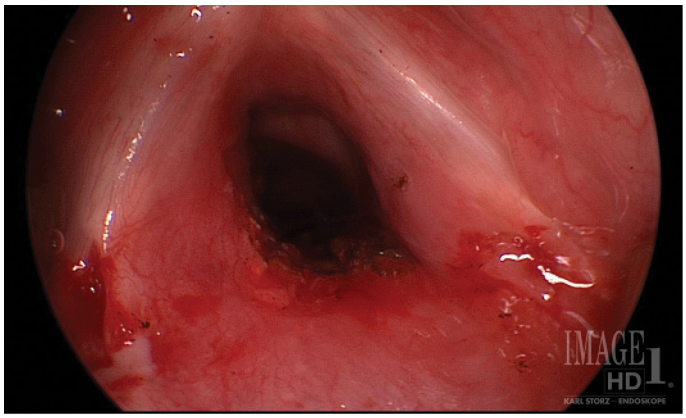

Figure 74.13 Postoperative appearance of the subglottic haemangioma from Figure 74.4 following carbon dioxide ablation. Note the dramatic improvement in airway calibre.

about the patient's ability to safely maintain their own airway, the patient can remain intubated at the end of the case and extubated in an intensive care unit setting when more stable.

If there is concern for long-term stability, consideration should be given to performing a tracheostomy. Patient positioning with a shoulder roll and significant chin extension often assisted with soak tape will help elevate and expose the cartilaginous structures of the neck for surgical planning. The increased subcutaneous fat content and short neck in infants can mislead the surgeon and awareness and identification of the anatomy is paramount to a safe procedure. The midline, cricoid, and sternal notch are identified and marked on the skin. A small horizontal incision is planned at approximately the second or third tracheal rings at the site of the anticipated tracheostomy, generally about 0.5–1 cm below the inferior border of the cricoid. Haemostasis is assisted with the injection of topical lidocaine with epinephrine. Non-flammable preparation is then used to cleanse the skin and the patient is towelled out. After incising through the epidermis and dermis, debulking of the subcutaneous fat is typically performed with needle tip cautery and can assist in visualization of the midline raphe and the strap muscles. These are next identified, divided in the midline and lateralized. The use of Ragnel retractors and blunt dissection with a haemostat can help carry the dissection down onto the trachea without further dividing any tissue, reducing the risk of bleeding. Small blood vessels can be cauterized as needed with bipolar cautery until the pretracheal fascia is identified. The pretracheal facia is then incised and divided from superior to inferior exposing the now-free tracheal surface. If the thyroid isthmus is in the way, it may be divided slowly using monopolar cautery. Alternatively, it can be retracted inferiorly without being divided. The inferior extent of the dissection should be minimally manipulated and palpated multiple times to ensure there is no high-riding innominate artery that could result in devastating vascular injury. Once the tracheal surface has been cleared adequately, typically two or three rings, 3.0 or 4.0 Prolene stay sutures are placed parasagittally immediately parallel to the anticipated vertical tracheotomy site. These are utilized in the postoperative event of an accidental decannulation to help open and expose the tracheotomy site for re-cannulation. At this point, the stoma may also be formalized through a series of half-mattress sutures approximating the skin to the tracheal surface. This reduces the potential for 'false tracking' the tracheostomy tube whereby the tube is placed into the soft tissues around the trachea as opposed to into the lumen of the airway. Prior to incising the trachea, communication with the anaesthetist is critical. They should have access to the patient's head, and ability to un-tape the endotracheal tube while keeping it securely in place. A vertical midline tracheotomy is made between the stay sutures until the tracheal tube is visualized, typically the height of two tracheal rings. A small haemostat can be used to gently spread horizontally in the incision, ensuring adequate airway calibre. The anaesthesiologist then backs the tube out to the superior level of the tracheotomy without removing it, allowing the distal cannulation with a pre-tested, age-appropriate tracheostomy tube. After placement of the tube and removal of the obturator, a ventilating circuit is passed on to the sterile field and connected to the tracheostomy tube. Tidal volumes and carbon dioxide return should be immediately confirmed by the anaesthesiologist. A flexible fibreoptic laryngoscope should be readily available to perform tracheoscopy and determine the height above the carina or help troubleshoot if there is any concern for placement. As always, there should be multiple

back-up options including an endotracheal tube on the field, as well as the indwelling endotracheal tube above the tracheotomy site that can be re-advanced if ventilation is not adequate. Only after confirmation of adequate ventilation through the tracheostomy tube should the endotracheal tube be removed. The tracheostomy tube is then secured around the neck leaving one to two finger breaths to prevent skin necrosis and breakdown. Dressings may be applied underneath the flanges of the tracheostomy tube or the securing ties and/or Velcro. Stay sutures are labelled with their laterality and secured to the chest using Tegaderm. The patient is transported to the intensive care unit where they should remain sedated for 3–7 days prior to performance of the first tracheostomy tube change, typically by the surgical team.

Paediatric airway foreign bodies merit special consideration as they have the potential to be life-threatening; therefore, timely evaluation and a high degree of suspicion are necessary. Tracheobronchial foreign bodies are best dressed using the standard bronchoscopy techniques described with a spontaneously ventilating patient. An age-appropriate bronchoscope should be set up and ready to use, as well as one a size smaller in the event of an unexpectedly smaller airway calibre. After exposure of the larynx and insertion of the bronchoscope through the vocal cords, the ventilating circuit can be hooked up directly to the bronchoscope, permitting ventilatory support while the foreign body is identified and extracted. After identification, the telescope is removed, leaving the bronchoscope sheath in place, and it is replaced with optical forceps chosen based on the identified foreign body. The optical forceps are advanced through the bronchoscopic sheath. The forceps are opened upon exiting the sheath and the foreign body is grasped and gently delivered into the distal opening of the bronchoscope, thereby securing the foreign body. The bronchoscope and foreign body are then removed in concert, being mindful not to drop the foreign body at the level of the glottis. Repeat bronchoscopy is then performed ensuring there are no additional foreign bodies before turning the patient over to anaesthesia for emergence.

One last procedure that merits attention is the *ex utero* intrapartum treatment (EXIT) procedure. EXIT is a rare procedure whereby an infant is partially delivered via caesarean section while being maintained on placental circulation in order to secure the unborn child's airway. These procedures, often taking weeks or even months of planning, are typically reserved for large obstructive head and neck lesions that place the neonate at risk for respiratory compromise without immediate high-level intervention. These procedures are typically attended by multiple teams including anaesthesiologists, neonatologists, obstetricians, maternal fetal medicine, otolaryngologists, and general surgeons. During an EXIT procedure, the full armamentarium of all described techniques in this chapter should be readily available including potential partial or complete resection of any large cervical facial masses that are impinging on the airway, or even cannulation and initiation of extracorporeal membrane oxygenation in the event an airway is not able to be established. After opening the uterus, the head and a single shoulder and arm are delivered. The surgical team then secures the airway by whatever means necessary, while the remainder of the patient remains within the uterus. Only after securing an airway is the patient fully delivered and placental circulation disrupted. The patient is immediately handed over to the neonatology team for critical care.

Conclusion

The successful management of a paediatric patient suffering from acute airway compromise requires a succinct and thoughtful approach, timely decision-making, clear communication, and technical expertise. Failure to escalate and call for support can be catastrophic, and every effort should be made to engage the appropriate team members when called to an urgent situation. The most effective tool in emergent situations is clear and concise communication with teammates to create a rapid collective plan of intervention. Saving a patient's life in the face of impending airway distress, however, is perhaps one of the most rewarding aspects of the field, and while one hopes to rarely, if ever, be called to this situation, the well-trained otolaryngologist must be prepared to act. The authors hope this chapter has provided a guideline for assessing and managing the paediatric patient with impending airway compromise.

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75

Snoring and obstructive sleep apnoea

Kate A. Stephenson and Michael Kuo

Introduction

Snoring and concerns regarding obstructive sleep apnoea (OSA) are among the commonest reasons for referral of a child to a paediatric ear, nose, and throat (ENT) clinic. Differentiation of 'simple' snoring from potentially severe OSA, with its associated significant morbidity, is both a challenge and of great clinical importance.

Simple snoring—also known as habitual or primary snoring—is common and occurs in approximately 10–15% of children, whereas the prevalence of OSA is likely to range between 1% and 4%; this is around two- to fourfold higher in some subgroups, such as obese or black children.

The wide range of adverse health outcomes of OSA include cognitive and behavioural problems, inflammation, and cardiovascular consequences. This important disease burden is associated with significant healthcare use and cost. OSA is also linked to quality-of-life impairment in both the child and the caregivers.

Terminology

It is important to use the correct terminology when referring to 'a snoring child'. Snoring is the lay term for breathing in a noisy way, often with a rough or snorting quality. The correlating clinical term of stertor defines an abnormal breathing noise with a low-pitched inspiratory quality that originates from the upper airway. Simple snoring implies a mild form of upper airway resistance without sleep fragmentation or gas exchange abnormalities.

The term obstructive sleep-disordered breathing (oSDB) is best applied when a clinical diagnosis of relative upper airway obstruction that may impair ventilation and/or oxygenation is suspected; this encompasses a spectrum of disease including OSA. oSDB needs to be differentiated from sleep-disordered breathing as this may describe central apnoeas and hypoventilation. Upper airway resistance syndrome has also been used to describe a less severe form of upper airway obstruction than OSA and tends to be applied primarily in the adult setting (see Chapter 51). It is characterized by increased work of breathing and sleep fragmentation, without significant desaturation episodes or gas exchange abnormalities.

A label of OSA should strictly only be applied when the diagnosis is clinically suspected and then confirmed by appropriate investigation. OSA implies the presence of increased upper airway resistance with resultant periodic airway obstruction (either hypopnoea or apnoea) and sleep disruption, intermittent hypoxaemia, and/or hypercapnia.

Basic science: anatomy and physiology

Anatomy

The upper airway obstruction resulting in oSDB can occur at a single level; however, in most cases several anatomical factors are involved in a picture of multilevel obstruction. Although the nasopharynx and oropharynx are the principal culprits, the entire upper airway is relevant, including the nasal cavities, hypopharynx, and larynx.

The commonest risk factor for childhood oSDB is adenotonsillar hypertrophy; the combined volume of the tonsils and adenoids correlates better with oSDB severity than tonsillar size alone. It is likely that the severity of oSDB is related to a combination of tonsillar and adenoidal relative size, craniofacial anatomy, and neuromuscular tone; the age of the child can influence all of these factors. Onset of OSA usually occurs between the age of 2 and 6 years, coinciding with adenotonsillar growth and peak in relative size. Tonsillar hypertrophy can broadly be defined as grade 3 or above on a standardized scale of 0–4 (Brodsky and Friedman grading systems) by oropharyngeal inspection (**Figures 75.1 and 75.2**). Adenoid size estimation is more challenging and can be gauged endoscopically (**Figure 75.3**), by indirect visualization, palpation, or radiologically. Grading can also be made on a standardized scale or by percentage of choanal/nasopharyngeal obstruction.

Examples of craniofacial risk factors for oSDB include midfacial hypoplasia, micro/retrognathia, and macroglossia.

Physiology

Neuromuscular tone physiologically varies according to phase of sleep. Episodes of airway collapse are more likely to occur during the rapid eye movement sleep phase due to comparatively reduced airway muscle tone. Neuromuscular diseases may result in pathological hypotonia while hypertonia (e.g. in patients with cerebral palsy), is also linked to oSDB. Sedatives and stimulants also affect neuromuscular tone.

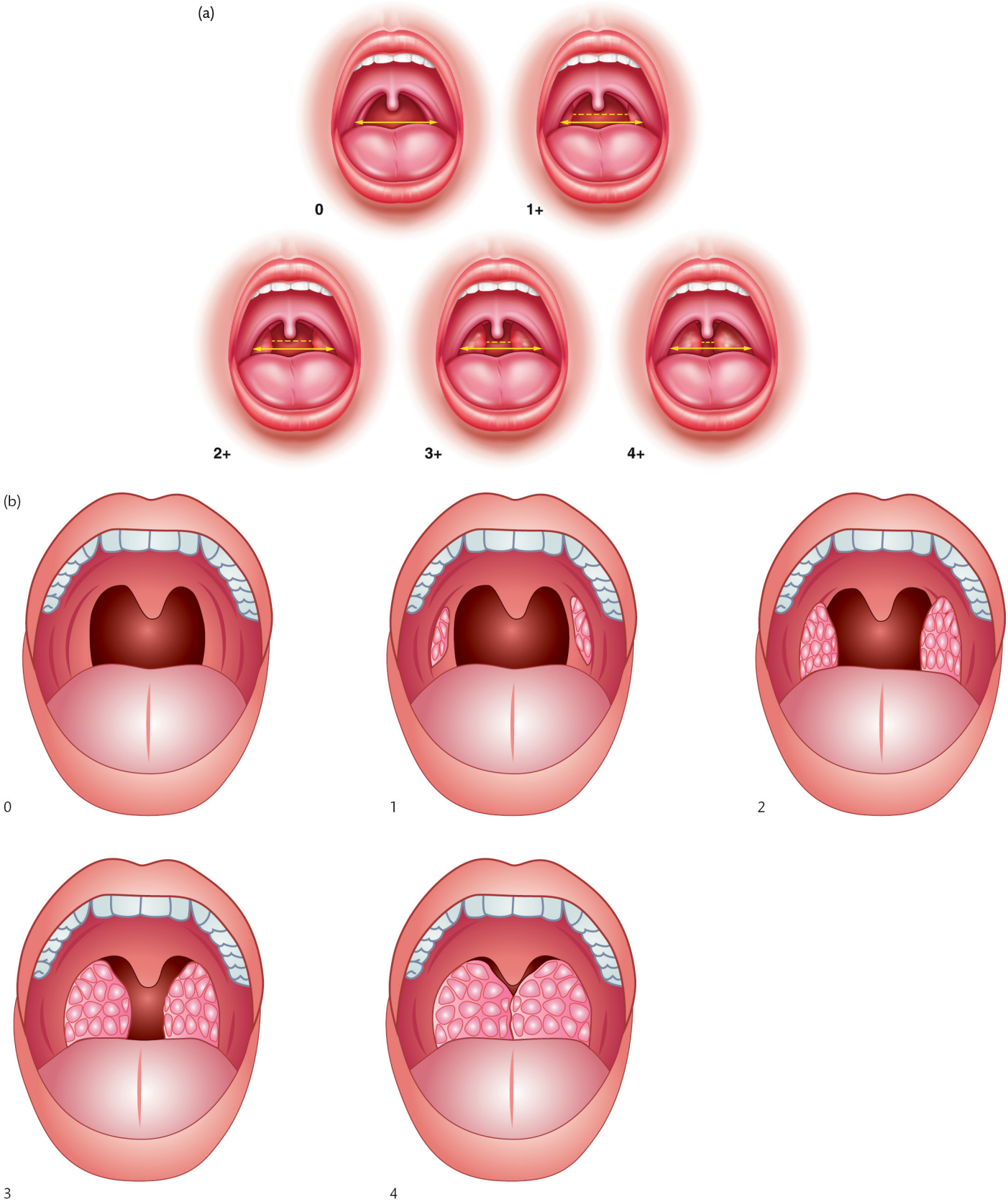

Figure 75.1 Tonsil grading systems. (a) Brodsky scale. 0: tonsils are entirely within the tonsillar pillars. 1+: tonsils occupy less than 25% of the oropharyngeal width (as measured between the anterior tonsillar pillars). 2+: tonsils occupy from 25% to 50% of the oropharyngeal width. 3+: tonsils occupy from 50% to 75% of the oropharyngeal width. 4+: tonsils occupy more than 75% of the oropharyngeal width. (b) Friedman scale. 0: previous tonsillectomy. 1: tonsils are in the tonsillar fossae, barely seen behind the anterior pillars 2: tonsils visible behind the anterior pillars. 3: tonsils extend beyond the pillars/three-quarters of the way to the midline. 4: tonsils extend to the midline.

(a) From Quimby A, Salman SO. Surgical evaluation and airway assessment of patients with OSA. In: Salman SO, ed. *Modern Management of Obstructive Sleep Apnea*. Springer; 2019:25–37. (b) Reproduced from Friedman M, Tanyeri H, La Rosa M, et al. Clinical predictors of obstructive sleep apnea. *Laryngoscope* 10.1097/00005537-199912000-00002, with permission from John Wiley and Sons.

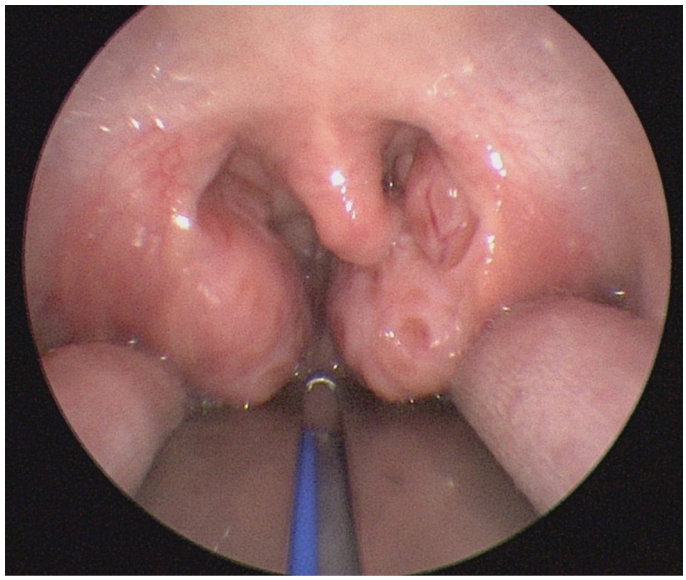

Figure 75.2 Clinical photograph of significant tonsillar hypertrophy.

In OSA, the ventilatory changes (oxygen desaturation, carbon dioxide retention, and intrathoracic pressure swings) cause rapid changes in heart rate and blood pressure due to catecholamine release. These, in turn, result in autonomic nervous system dysfunction, precipitating the cardiovascular sequelae associated with OSA, such as systemic hypertension. Elevated pulmonary vascular resistance leads to pulmonary hypertension and, at worst, right heart failure. There is evidence that OSA also impacts metabolic pathways, contributing to obesity.

Differences between sleep physiology in children and in adults mean that the same measurement parameters cannot be applied. For example, partial airway obstruction is more common and oxygen desaturation less common in children with OSA than in adults (see Chapter 51). Further differences are seen between older children and infants. Table 75.1 defines common sleep physiology terms, as measured by polysomnography (PSG). Application of cut-off values is particularly difficult in children due to lack of a standardization in parameters measured and variation in definitions.

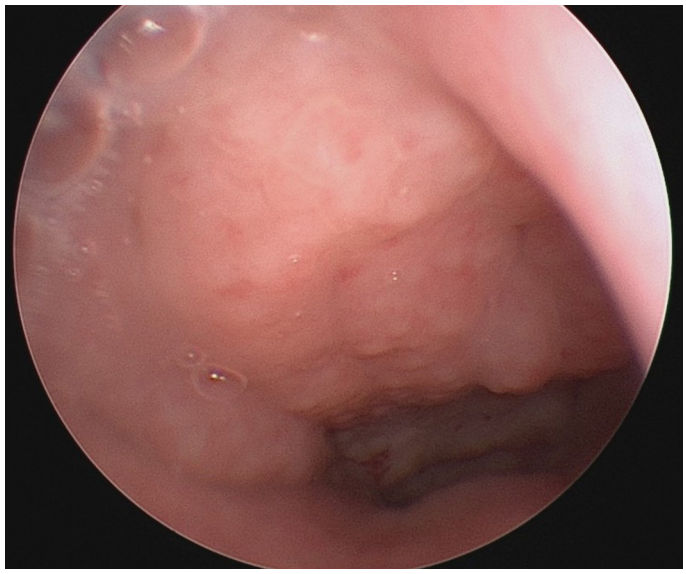

Figure 75.3 A transnasal endoscopic photograph of the adenoids.

Table 75.1 Commonly used paediatric sleep physiology definitions

Apnoea	Cessation of >90% air flow for at least 2 breath cycles
Hypopnoea	Decrease in air flow of >30% for at least 2 breath cycles, associated with cortical arousal or ≥3% O_2 desaturation
Apnoea–hypopnoea index (AHI)	Number of mixed, obstructive, and central apnoeas and hypopnoeas per hour of total sleep time
Obstructive apnoea–hypopnoea index (oAHI)	Number of obstructive apnoeas and hypopnoeas per hour of total sleep time
Respiratory effort-related arousal (RERA)	Increasing respiratory effort, flattening of the inspiratory portion of the nasal pressure waveform, snoring, or an elevation in the end-tidal $PaCO_2$ that leads to arousal and does not qualify as an apnoea or hypopnoea
Respiratory disturbance index (RDI)	Number of obstructive apnoeas, hypopnoeas, and RERAs per hour of total sleep time
OSA*	Obstructive AHI ≥1 *Or* A pattern of obstructive hypoventilation defined as at least 25% of total sleep time with hypercapnia ($PaCO_2$ >50 mmHg) in association with snoring, flattening of the nasal pressure waveform, or paradoxical respiratory effort
Mild OSA	AHI 1–5
Moderate OSA	AHI 5–10
Severe OSA	AHI >10

*A definitive diagnosis of OSA requires both clinical and PSG evidence.

Assessment

History

A detailed history is obtained from the caregivers. There can be great variation in interpretation of the child's breathing and in the terms used to report symptoms; the use of open-ended questions is recommended. Snoring is not always described, while words and phrases such as 'heavy breathing', 'gasping', 'pausing', and 'struggling for air' are often used. Partial or full arousals from sleep ('waking him/herself up') are a typical feature. The child may adopt unusual sleeping positions in an attempt to improve their upper airway, such as neck extension. Evidence of increased work or paradoxical breathing may also be reported, evidenced by flaring of the nasal ala, tracheal tug, sternal recession, and 'see-saw' respiration. Nocturnal sweating is a further common but rather non-specific feature of oSDB.

The level of concern on the part of the caregivers can be a useful, if imprecise, clinical hint of disease severity; often in the worst of cases, families describe extreme anxiety and fatigue due to constant observation of their sleeping child with frequent repositioning and stimulation of the child during apnoeic episodes. A history of nocturnal enuresis, particularly secondary enuresis (occurrence of bedwetting after a period of nighttime control), can be an indicator of

Box 75.1 Key comorbidities with an increased risk of paediatric obstructive sleep-disordered breathing

- Obesity.
- Trisomy 21.
- Neuromuscular diseases, including cerebral palsy.
- Mucopolysaccharidoses.
- Craniofacial abnormalities.
- Achondroplasia.
- Prematurity.

(See also Chapter 90.)

oSDB. Snoring and oSDB symptoms are also typically transiently worsen at the time of upper respiratory tract infections.

In severe cases there can also be a history of stertor when awake and/or mouth breathing. Children that have severe nasal obstruction are obliged to keep an open-mouth posture which lends itself to drooling, particularly in the younger child. Children who are not getting restful sleep at night are frequently paradoxically hyperactive in the daytime rather than more tired; daytime somnolence is a much stronger characteristic of adult OSA. Irritability, poor concentration, and impaired mental performance are key daytime features.

Previous medical and surgical history is important. Key comorbidities of note are detailed in **Box 75.1**. There is high-quality evidence linking a history of prematurity to OSA while a history of OSA or childhood adenotonsillectomy in a parent or sibling is also associated, albeit weakly. A history of asthma, especially severe asthma, influences surgical care setting choice while adenotonsillectomy is linked to an improvement in asthma disease markers. oSDB can also be associated with failure to thrive. The child's growth should be evaluated and consideration given as to whether they are significantly underweight or obese.

The Epworth Sleepiness Questionnaire is an adult sleep medicine tool but is not applicable to children. It has been modified (the Epworth Sleepiness Scale for Children and Adolescents (ESS-CHAD)) and can be used to assess the level of excessive daytime sleepiness in children and adolescents. Other paediatric questionnaires for completion by a parental proxy include the Paediatric Sleep Questionnaire (PSQ), which includes a sleep-relating breathing disorder scale aiming to identify children at risk of oSDB, and the OSA-18 tool, which assesses sleep-disordered breathing-related quality of life. These scales predict OSA to an extent; however, there is poor consensus regarding their exact role in clinical practice.

Examination

Observation of the child—whether awake or asleep—from the very start of the consultation and during history taking is recommended. Is the child dysmorphic? Is stertor or mouth breathing present? 'Adenoid facies' describes the long, thin, and open-mouthed facial appearance of children with chronic severe nasal obstruction.

Simple measures of nasal airflow include a misting test on a metal spatula and use of a wisp of cotton wool close to the nares. Anterior rhinoscopy is often best achieved by gently lifting the tip of the nose with a thumb or forefinger rather than use of a Thudichum speculum. The otoscope is also a useful anterior rhinoscope in the older cooperative or younger restrained child. Check for anterior nasal obstruction causes such as septal deviation, rhinitis, or inferior turbinate hypertrophy.

Tonsillar size should be evaluated. This can be facilitated by tongue protrusion or saying 'ah', with the use of a tongue depressor reserved for when these measures fail or are not possible. During oral examination, it is useful to note any abnormalities of the palate or tongue.

A complete basic ENT examination should be performed in all cases; OSA is often accompanied by otitis media with effusion. Pectus excavatum as a result of chronic severe obstruction occurs more readily in the younger child but is now rarely seen, occurring only in the most severe of cases.

Video or audio recordings of the sleeping child can often be provided by the caregivers. A video recording may be particularly useful if it captures respiratory effort without evidence of air flow suggesting transient complete obstruction.

Investigations

In the majority of children, a detailed history and examination may be sufficient to recommend treatment. An individualized approach to investigation is recommended. Investigation is likely to be particularly useful in certain circumstances, such as younger children (e.g. <2 years), and those with obesity and comorbidities. It is also valuable where there is a poor correlation between symptoms and clinical findings.

The role of investigation in the child suspected to have oSDB is fourfold: firstly, it can be used to confirm the diagnosis and attempt to define the severity of disease, guiding the time frame and clinical setting for any intervention. Secondly, it can also localize the site of the obstruction and therefore guide best intervention. Thirdly, investigation can check for complications of oSDB and further inform anaesthetic practice/perioperative care. Lastly, it also guides expectations of outcomes from interventions and provides a baseline for comparison. Significant controversy exists as to the best investigative strategy and practices may vary widely according to local resources and policy.

Does the child have obstructive sleep apnoea?

A diagnosis of OSA can be strongly suspected on clinical grounds but can only be confirmed by appropriate investigation. A once-off assessment of baseline saturations in the outpatient clinic is likely to be of little or no clinical value in the child without other cardiorespiratory concerns. The presence of night-to-night variation in sleep physiology also presents a challenge when attempting to define and quantify disease.

The umbrella term of 'sleep study' may describe several different investigations, the most basic being overnight oximetry. This records oxygen saturations and heart rate sequentially, typically over a period of one or two nights. This is relatively low-cost, can easily be performed in a home setting, and does not require continuous input from a clinician. Information gained includes oxygen saturation nadir, number of dips, and percentage of total sleep time spent below specified saturation percentage levels. **Figure 75.4** is an example of an oximetry tracing. Overnight oximetry is widely available and often performed. Importantly, the frequency of measurements can vary from machine to machine. It has a high positive predictive value for oSDB but a low negative predictive value (not all

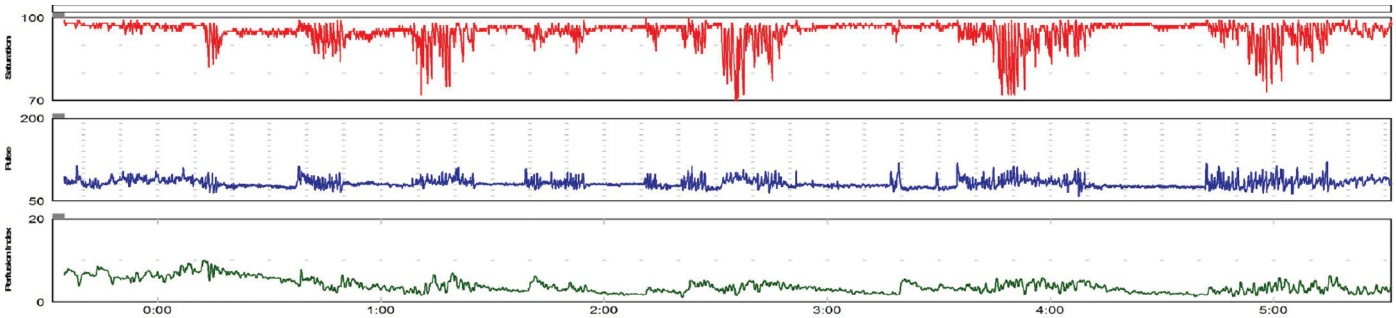

Figure 75.4 An overnight oximetry trace demonstrating episodic desaturations. The upper red line describes oxygen saturations on the y-axis and time on the x-axis.

apnoeas and hypopnoeas result in a desaturation); a normal overnight oximetry therefore does not rule out OSA. Sleep oximetry is not currently recommended as a routine screening tool.

A 'full cardiorespiratory study', PSG, is regarded as the gold standard in the diagnosis of OSA. It is expensive and not widely available. PSG comprises simultaneous multichannel monitoring and typically includes measurement of oxygen saturation, end-tidal carbon dioxide, nasal airflow, and respiratory effort. It often includes electromyography, electroencephalography, electrocardiography, and electro-oculography. In detailed studies, differentiation can be made between obstructive and central apnoeas and hypopnoeas. Adult PSG parameters cannot be applied to children and there is poor consensus as to stratification between grades of severity and parameters reported. Classification of disease severity and treatment decision-making should not be made solely on the basis of certain PSG parameters (e.g. respiratory disturbance index, apnoea–hypopnoea index (AHI), and minimum oxygen saturation); additional useful variables include arousals and symptoms, which may be poorly correlated. Current UK guidelines suggest that PSG should be used selectively in cases with comorbidity or diagnostic uncertainty. In severe or complex OSA cases, post-intervention repeat PSG is recommended to assess the degree of improvement. Critically, PSG does not localize the level of obstruction (Figure 75.5).

Where is the obstruction?

It the vast majority of children, obstruction will be principally due to relative adenotonsillar hypertrophy. While tonsillar bulk is easy to assess to direct visualization, assessment of adenoidal size presents a greater challenge. Flexible nasendoscopy is a rapid, outpatient clinic-based investigation that assesses not only the degree of adenoidal obstruction but also the nasal cavities. It is, however, invasive and not likely to be well tolerated in the child between the age of 1 and 5 years. Lateral neck radiography (a 'postnasal space X-ray') is employed by some clinicians to give greater information regarding the postnasal airway and adenoidal bulk (Figure 75.6). The tonsillar/pharyngeal ratio and the adenoid/nasopharyngeal ratio can be estimated. This is a non-invasive, low-cost, and quick investigation but does require ionizing radiation. One could argue that if severe bilateral nasal obstruction is present and anterior rhinoscopy is normal then the site of nasal obstruction could be assumed to be posterior and related to relative adenoidal hypertrophy in the vast majority of children.

In children without adenotonsillar hypertrophy, or those with persistent OSA despite adenotonsillectomy, there is a need to localize the persistent site(s) of obstruction and inform next management steps. Drug-induced sleep endoscopy (DISE) is used in some centres and aims to create the airway conditions of natural

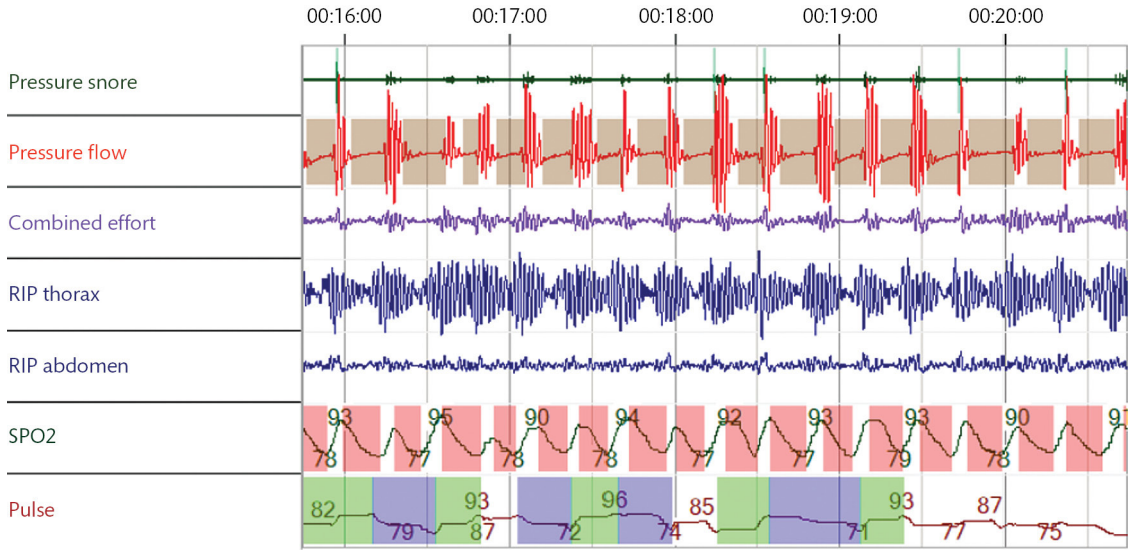

Figure 75.5 A 5-minute sample of a PSG recording, demonstrating obstruction and desaturations. RIP, respiratory inductance plethysmography.

Figure 75.6 A postnasal space X-ray demonstrating significant adenoidal bulk and narrowing of the nasopharyngeal airway.

sleep. DISE requires careful sedation or anaesthesia while a flexible nasendoscopy is performed. It can demonstrate obstruction at several levels, namely the nasopharynx/velopharynx, tongue base, oropharynx and hypopharynx, and the larynx. The practice of DISE is not widespread. If there are greater laryngotracheal concerns, a full laryngotracheobronchoscopy is also advisable.

Magnetic resonance imaging (MRI) of the head and neck describes the soft tissue anatomy related to the upper airway while dynamic or 'cine'-MRI can also show changes in airway patency throughout the breathing cycle. Again, this requires skilled anaesthetic input to achieve conditions that best mimic the natural sleep state. Like DISE, MRI is rarely indicated and reserved for severe, refractory, and complex cases, in specialist centres or research settings. Computed tomography scanning of the head and neck, with its accompanying radiation dose, is rarely applicable; it might be employed in patients with maxillofacial and craniofacial concerns to evaluate bony anatomy.

Is there evidence of obstructive sleep apnoea complications?

Thankfully, significant cardiovascular complications of OSA occur only in the most severe and prolonged cases. A careful preoperative anaesthetic assessment should be made. Some clinicians elect to perform an electrocardiogram and/or chest X-ray, searching for cardiomegaly and evidence of pulmonary hypertension. Echocardiology and cardiac catheterization studies may be indicated if these concerns are present; however, for the vast majority of oSDB cases, these investigations are not required.

Differential diagnoses

A basic knowledge of childhood sleep is important to differentiate potential pathology from normal behaviour. A normal child can be observed to have a periodic breathing pattern with spells of faster and slower respiration in addition to brief physiological apnoeas. Parents may notice these patterns and become falsely concerned.

While OSA affects approximately 1–4% of children, up to 50% can have a 'sleep problem' at some point. Non-breathing-related sleep disorders can also give rise to significant parental worry. They include disrupted sleep patterns (e.g. behavioural insomnia, including difficulty initiating and maintaining sleep) and parasomnias (such as night terrors and sleepwalking). A careful history can largely differentiate between breathing and non-breathing-related sleep-disorders. Importantly, overlap can occur and oSDB is a common trigger for parasomnias.

Central causes of sleep-related hypoventilation are a rare but important differential. Examples include congenital central hypoventilation syndrome ('Ondine's curse') and disorders of the brain and spinal cord. Clinical features may include early presentation with cyanotic episodes, apparent life-threatening events, and apnoeas in the absence of an obstructive picture. Clinical suspicion is important and PSG aids diagnosis. Early specialist paediatric and respiratory input is advisable.

Treatment

The treatment of paediatric OSA is markedly different to that of adult OSA; it is largely a straightforward and very rewarding condition to treat. Given that most cases are due to relative adenotonsillar hypertrophy, surgery (tonsillectomy and/or adenoidectomy) results in a significant improvement in the majority of cases; however, this is by no means universal. The clinician should be cautious not to apply a 'one-size-fits-all' approach. Validated paediatric sleep questionnaires can be used for research purposes and to refine the indication for PSG. Detailed history from the parents and video footage of the child's breathing pattern captured on the parent's smartphone during sleep can be equally informative.

Important factors that may influence treatment decision-making include the age of child, comorbidities, and previous ENT surgical history. In obese children, weight loss interventions improve OSA severity and should be recommended alongside other tailored treatments.

In children suspected to have primary snoring or potential mild OSA, a period of active observation ('watchful waiting') may be appropriate. Evidence has suggested that PSG findings can normalize in around 45% of children with non-severe OSA after a watchful-waiting period of around 6 months; however, this is less likely to occur in children who are obese, black, and in those who have more severe disease. Importantly, this does not necessarily correlate with symptomatic improvement and the long-term natural history of OSA remains relatively poorly understood.

Medical

The principal medical therapies considered in children with oSDB can be broadly classed as anti-inflammatory medications. Raised inflammatory mediators such as prostaglandins and leukotrienes have been found in the upper airways of children with OSA while leukotriene antagonists have been found to reduce adenotonsillar cell proliferation *in vitro*. Anti-inflammatory medications are particularly useful in children with evidence of significant rhinitis and are most frequently considered in cases of likely mild disease or those with residual symptoms post adenotonsillectomy.

Steroids

Topical nasal steroids can be applied in either a spray or drop form; drops may be better tolerated by the younger child. This is frequently an 'off-licence' use according to age and this should be discussed with the caregivers. Serious side effects are rare; however, compliance may be poor and epistaxis can result from either nasal trauma or drying of the nasal mucosa. Some might consider a 1-week short course of a topical decongestant at initiation of topical nasal steroid treatment in order to improve the access of the medication to the nasal cavities.

Systemic steroids are not recommended in the routine management of oSDB. A short course may act as a 'rescue' for selected severe cases and result in short-term relative symptomatic improvement. It is, however, not a definitive or long-term treatment. There are significant adverse effects associated with systemic steroids, especially when used in high doses and for prolonged periods; benefits of prescription must be considered and discussed on an individualized basis.

Leukotriene antagonists

The leukotriene antagonist montelukast is an oral medication that has been found to have potential to reduce OSA severity in the short term, as evidenced by significantly reduced AHI scores. Montelukast tends to be well tolerated and serious adverse effects are rare; however, there is a lack of evidence to support long-term improvement.

There is no high-quality evidence to support the use of antihistamines in the general management of oSDB. Antihistamines may, however, be useful if allergic rhinitis is also present.

Surgical

Adenotonsillectomy

Adenotonsillectomy is the mainstay of treatment in the vast majority of children with oSDB, resulting in prompt subjective symptomatic improvement. Meta-analysis has found that resolution of OSA occurs in around 80% of children following adenotonsillectomy, with PSG-demonstrated improvement in saturations and apnoea parameters. Tonsillar size has not been found to be clearly correlated with improvement in OSA following adenotonsillectomy, although a larger proportion of patients with tonsillar hypertrophy (Brodsky grade 3+) have complete resolution of OSA.

The 2013 Childhood Adenotonsillectomy Trial (CHAT) compared early adenotonsillectomy to watchful waiting in 5- to 9-year-old children with non-severe OSA. This was a major evidence advance; surgery was associated with significantly greater improvement in oSDB symptoms, PSG findings, and also in behavioural and quality-of-life markers.

If surgical management is planned, the best care setting for this is an important consideration. UK national guidance from a multidisciplinary working party was published in 2019 and recommends that children with OSA symptoms should be managed as close to home as possible within good regional clinical networks. It advises that secondary-level centres (district general hospitals without paediatric intensive care facilities) are able to treat children aged 2 years and over (corresponding to a weight of >12 kg) without extremes of body mass index or other high-risk factors (e.g. severe cerebral palsy, achondroplasia, moderate or severe neuromuscular disorders, significant craniofacial abnormalities, mucopolysaccharidosis, or other significant cardiorespiratory concerns) (see Chapter 87).

Other surgical procedures

Surgery should be tailored to the individual anatomy and suspected sites of relative upper airway obstruction. In cases of anterior or mid-nasal obstruction, septoplasty and turbinate reduction surgery (in those that have not responded to medical treatment) can be indicated. Palatal surgery such as uvulopalatopharyngoplasty features much more heavily in the surgical management of adult OSA (see Chapter 51). Tongue base and supraglottic surgery have a role in highly selected cases. Lastly, the treatment of OSA in children with craniofacial abnormalities is highly specialized and can include surgeries such as mid-facial and mandibular advancement procedures.

A tracheostomy is the final rung on the OSA surgical treatment ladder. Although highly effective in bypassing upper airway obstruction, it is a 'last resort', associated with significant care and morbidity concerns that include but are not limited to voice and swallowing.

Non-pharmacological treatments

Continuous positive airway pressure

Application of continuous positive airway pressure during sleep improves the patency of the upper airway. A variety of tight-fitting masks are available for paediatric use; nasal masks or full-face masks can be tailored to the age of the patient and individual tolerance. This requires specialist expertise, typically including a respiratory paediatrician and a sleep laboratory for titration of therapy and compliance monitoring. Continuous positive airway pressure is a long-term therapy and principally has a role when adenotonsillectomy is either not feasible or not beneficial (i.e. OSA persists); this is more frequently the case in obese and syndromic children.

Nasopharyngeal airway

Insertion of a unilateral nasopharyngeal airway (NPA) can alter the tone and dynamics of the upper airway, improving patency. An NPA is of particular value in the neonate with craniofacial abnormalities, such Pierre Robin sequence. Tolerance in the toddler and older children frequently limits its use. 'Sleep studies' should guide and tailor NPA placement. With appropriate caregiver training, children may be managed at home with an NPA. An NPA can also be a useful adjunct in the immediate postoperative airway of children with severe OSA and those with syndromes such as trisomy 21.

Oral appliances such as mandibular advancement splints aim to enlarge the upper airway. They are not routinely employed in the management of paediatric OSA, and their use is reserved for special circumstances, such as children with craniofacial abnormalities. Their use is limited by tolerance and there is little high-quality efficacy data.

Controversies

Who should have polysomnography/drug-induced sleep endoscopy?

The American Academy of Otolaryngology–Head and Neck Surgery clinical practice guideline recommends PSG in children under the age of 2 years or with significant comorbidity. Its utility, in addition to confirming the diagnosis of OSA, lies also in prediction of

opiate sensitivity and likelihood of requirement for postoperative ventilatory support. PSG remains a limited resource in most institutions and prioritization should be based on individual patient risk assessment.

Currently, DISE is largely reserved for children for whom the clinical history suggests a multifactorial and multilevel obstruction, in whom the level of obstruction is unclear, and in those whose symptoms of OSA have not been relieved by adenotonsillectomy.

Is there any benefit of surgery in children with mild obstructive sleep apnoea ?

Four-fifths of children with OSA improve after adenotonsillectomy. The multi-centre Pediatric Adenotonsillectomy Trial for Snoring (PATS) included 459 children aged 3–12.9 with mild OSA (AHI <3). Behavioural problems, sleepiness symptoms and quality of life all improved with adenotonsillectomy over watchful waiting. Furthermore, it showed that early adenotonsillectomy was associated with a 32% reduction in all-cause health care encounters and a 48% reduction in prescription medications.

What should be the infrastructure required to support safe adenotonsillectomy for obstructive sleep apnoea?

Careful preoperative assessment, use of the NPA, and availability of postoperative oxygen have reduced unplanned demand for postoperative intensive care to a very low level. Adenotonsillectomy for OSA in children aged 2 years and weighing 12 kg without high-risk comorbidities can be safely delivered in secondary centres with appropriate paediatric anaesthetic support. Where there is high-dependency unit provision, children aged 1 year and weighing 10 kg can be treated in the absence of high-risk comorbidities.

Guidelines

Guidelines have been published by a combined working party of the British Association for Paediatric Otolaryngology, ENT UK, The Royal College of Anaesthetists, and the Association of Paediatric Anaesthetists of Great Britain and Ireland ('Safe Delivery of Paediatric ENT Surgery in the UK: A National Strategy', 2019); the European Respiratory Society ('Obstructive sleep disordered breathing in 2- to 18-year-old children: diagnosis and management', 2016); the American Academy of Otolaryngology–Head and Neck Surgery Foundation ('Clinical practice guideline: tonsillectomy in children (update)—executive summary', 2019); and the American Academy of Otolaryngology–Head and Neck Surgery (Position statement: tonsillectomy and OSAs, 2020). See 'Further reading' for more details.

Conclusion

OSA in children is common. In the majority of patients, the obstruction is attributable to adenotonsillar hypertrophy although the aetiology may be multifactorial. Although PSG is the 'gold standard' investigation, there is good correlation between parental history and PSG findings. Single-channel overnight oximetry has a good positive predictive value but poor sensitivity. In the absence of comorbidities, the incidence of unexpected need for postoperative intensive care support is very low.

FURTHER READING

American Academy of Otolaryngology—Head and Neck Surgery. Position statement: tonsillectomy and OSAs. 2020. https://www.entnet.org/resource/position-statement-tonsillectomy-and-osas/

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76

Sore throat in paediatric patients

Alex Millington and Nico Jonas

Introduction

Sore throat is one of the most common symptoms experienced by paediatric patients, with a variety of simple and more complex causes. Viral, bacterial, fungal, and parasitic infections; neoplastic processes; and gastrointestinal, neurological, and nutritional problems can all present with sore throat.

The majority of sore throat episodes are simple and self-limiting, with 40% resolving within 3 days and 85% within 1 week, but 10% of patients with sore throat may still see a general practitioner, with approximately half of these receiving antibiotics. Presentation to secondary care is usually associated with more severe or chronic symptoms, and in recent years there has been an upward trend in hospital admissions with severe tonsillitis and its complications.

Estimates put the economic burden of acute tonsillitis at £400–500 million per year in the UK, which, if extrapolated for all sore throats resulting in general practitioner attendance and/or time off school or work, could be many times more costly.

Basic science: anatomy and physiology

The mouth and nose are the primary natural entry points into the human body. As such, *Homo sapiens* have evolved to have a primary focus of immune protection in the head and neck with a proliferation of lymphoid tissue distributed in the lymph node plexuses and deposited around the oro- and nasopharynx. The quartet of adenoid, palatine tonsil, lingual tonsil, and tubal tonsil is known as Waldeyer's ring (**Figure 76.1**). The tubal tonsils, also known as Gerlach's tonsils, are situated just posterior to the Eustachian tube openings.

The palatine tonsils are covered with non-keratinizing stratified squamous epithelium and lie in a capsule formed by a condensation of the pharyngobasilar fascia, bounded anteriorly by the palatoglossus, posteriorly by the palatopharyngeus, and medially by the superior constrictors (**Figure 76.2**). The adenoids are situated on the posterior and superior wall of the nasopharynx and are covered with pseudostratified ciliated columnar epithelium.

The tonsils are well supplied with blood from five arteries, all branches of the external carotid artery (**Figure 76.3**), and the adenoids from six arteries, five from the external carotid and one from the internal carotid artery (**Table 76.1**).

Tonsils tend to be cryptic in nature with a network of primary and secondary crypts (**Figure 76.4**) and adenoids consist of tubular folds, both of which increase surface area for antigen identification. The tissues act as antigen-presenting units to B cells and T cells in the lymph tissue, stimulating an immune response in the form of secretory immunoglobulin A and production and formation of B memory cells. The associated cytokine cascade, increased blood supply, and inflammatory response is responsible for the symptom of sore throat in this context.

The glossopharyngeal nerve, which has a tympanic branch, lies deep to the tonsil and superior constrictor muscles and can be irritated in severe tonsillitis or quinsy abscess formation, causing referred otalgia.

The lymphatic drainage of the head and neck follows predictable patterns, with the adenoids draining to the retropharyngeal and pharyngomaxillary lymph nodes, and palatine tonsils draining to the jugulodigastric and upper deep cervical nodes.

Adenoid tissue continues to grow and reaches a maximum size at approximately 2–6 years before involuting. Tonsil tissue grows until puberty when it ceases before also atrophying. These proliferative periods can be associated with sleep-disordered breathing and obstructive sleep apnoea due to adenotonsillar hypertrophy (see Chapter 75).

Assessment

History

Timing, frequency, and severity of symptoms and the acute, chronic, or recurrent nature of sore throats are important, as are the effects of these on daily life. Symptoms may include:

- odynophagia
- dysphagia
- dysphonia
- pyrexia
- anorexia
- malaise and fatigue
- otalgia
- halitosis
- neck pain
- weight loss
- night sweats.

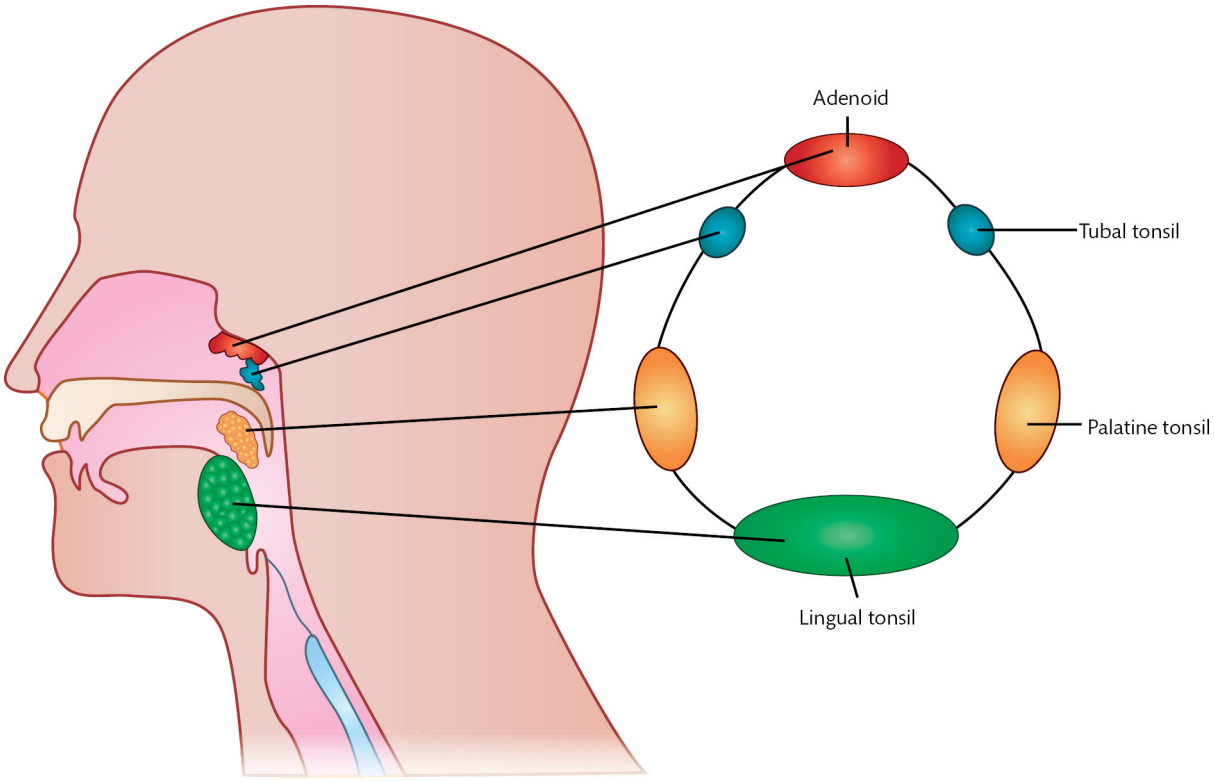

Figure 76.1 Waldeyer's ring.

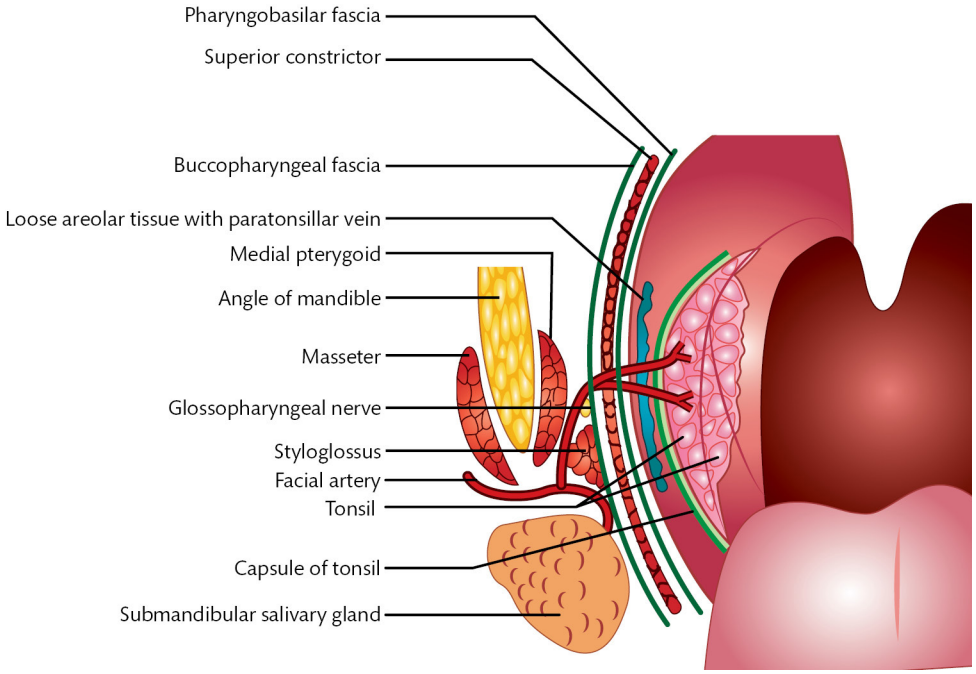

Figure 76.2 Cross-section of the right tonsil.

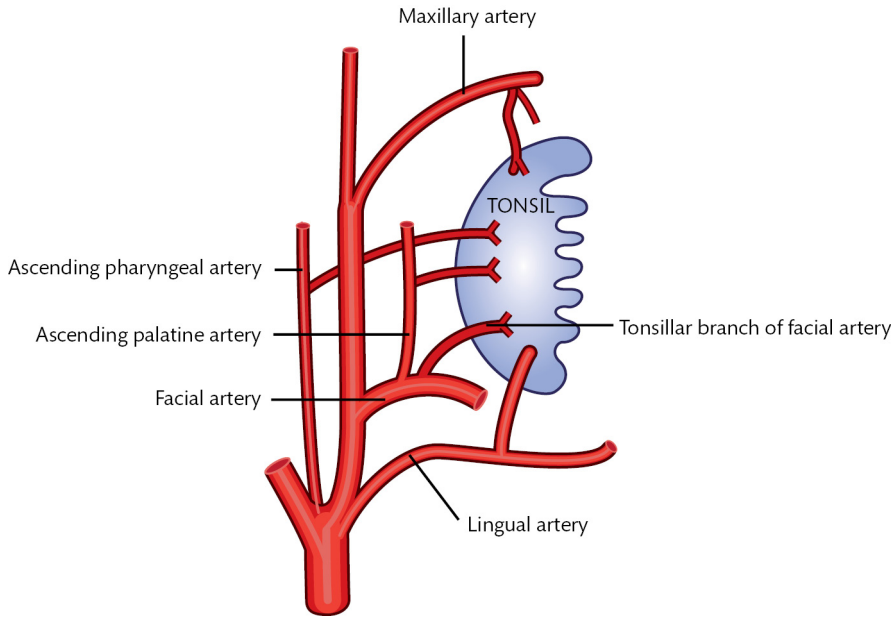

Figure 76.3 Arterial blood supply of the tonsil.

Table 76.1 Adenoid blood supply

External carotid supply	Internal carotid supply
Ascending pharyngeal artery	Basisphenoid artery
Ascending palatine	
Pharyngeal branch of internal maxillary	
Artery of pterygoid canal	
Contributions from tonsillar branch of facial artery	

Clinical examination

Clinical examination should include a general ear, nose, and throat examination and a focused oral and oropharyngeal examination.

Tonsil size can be graded using the Brodsky or Friedman tonsil classification (see Chapter 75).

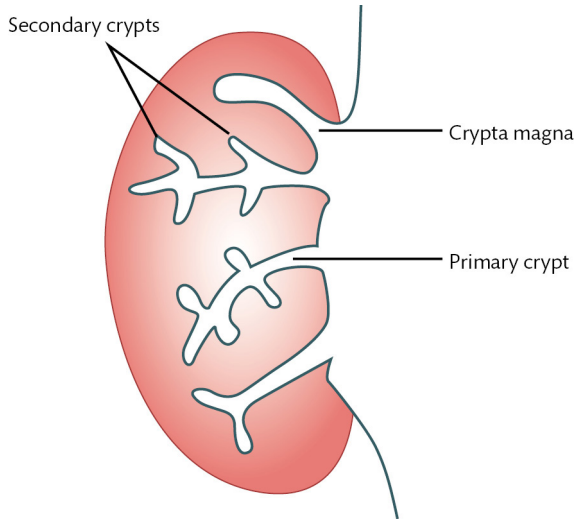

Figure 76.4 Anatomy of the tonsil crypts.

Clinical examination should further include basic dental examination including palpation and percussion. Neck examination is an essential part of the clinical examination and should include palpation for lymphadenopathy and assessment of neck movement, which can be impacted by deep neck space infections.

Investigations

Investigation for a simple sore throat is usually unnecessary in a primary care setting. Rapid antigen swabs for group A streptococcal infections are not recommended by the National Institute for Health and Care Excellence because of their effect on antimicrobial prescribing and lack of cost-effectiveness (2019). Routine throat swabs have a negative yield in 40% of patients. In addition, 20% of the population carry group A beta-haemolytic *Streptococcus* (GABHS) as a commensal infection and there are no criteria to distinguish infection and carriage.

If a patient is acutely unwell and requires secondary care, investigations often become necessary. These include full blood count, renal function, inflammatory markers (C-reactive protein/erythrocyte sedimentation rate), blood cultures (if febrile) and Paul Bunnell or Monospot tests. Taking an arterial or venous blood gas is good practice in severely acutely unwell patients to assess acid/base status.

Should there be a question or clinical suspicion of a deep neck space infection (see below), then a contrast computed tomography scan of the neck with or without chest should be ordered.

Differentials diagnoses/associated conditions

Tonsillitis (acute and chronic)

Acute tonsillitis presents with signs and symptoms of acute infection (pyrexia, pain, inflammation, etc.) and resolves within 2 weeks. Chronic tonsillitis can be defined as recurrent acute tonsillitis or infectious symptoms lasting more than 2 weeks (Table 76.2 and Figures 76.5–76.7).

Table 76.2 Acute and chronic tonsillitis

Classification	Features
Acute tonsillitis	
Acute superficial	Part of generalized pharyngitis
Acute follicular (Figure 76.5)	Infection into crypts with purulent material in crypts
Acute parenchymatous	Tonsil uniformly enlarged and congested
Acute membranous (Figure 76.6)	Follicular exudates coalesce to form surface membrane
Chronic tonsillitis	
Chronic follicular (Figure 76.7)	Tonsil stones in crypts
Chronic parenchymatous	Enlarged tonsils
Chronic fibroid	Fibrotic/small/micro-abscesses

Tonsillitis (viral versus bacterial)

Viral pathogens include adenovirus, respiratory syncytial virus, rhinovirus, influenza, parainfluenza, cytomegalovirus, and Epstein–Barr virus. Bacterial tonsillitis is usually due to GABHS, but other pathogens include *Staphylococcus*, non-haemolytic *Streptococcus*, *Haemophilus influenza*, *Bacteroides*, and *Actinomyces*.

Distinguishing between bacterial and viral infections is difficult because the signs and symptoms are similar whatever the cause. As the majority of infections are viral and episodes self-limiting, unnecessary antibiotic prescribing may cause avoidable harm and may contribute to the global issue of antimicrobial resistance.

Figure 76.5 Acute follicular tonsillitis.

Figure 76.6 Acute membranous tonsillitis (glandular fever tonsillitis).
From James Heilman, MD https://creativecommons.org/licenses/by-sa/3.0/

Examination may be able to determine whether the patient has an acute parenchymatous tonsillitis—the whole tonsil infected but no exudate/pus, or acute follicular tonsillitis—crypts filled with infected fibrin, although this is somewhat academic as they are treated similarly.

Glandular fever/infectious mononucleosis

Usually caused by the Epstein–Barr virus (90%) with 10% caused by cytomegalovirus. Glandular fever often presents as tonsillitis with marked tonsil hypertrophy with profuse exudate, diffuse cervical lymphadenopathy, and generalized malaise (Figure 76.6). Viral inflammation of the liver and spleen can lead to hepatosplenomegaly

Figure 76.7 Chronic follicular tonsillitis (tonsil stones).
Copyright Alex Millington.

and dysfunction. Rarely, the adenotonsillar hypertrophy can be so severe as to cause airway obstruction, particularly in a child with a background of large tonsils, which may require systemic steroid therapy, airway interventions, or even an emergency 'hot' tonsillectomy.

Blood tests will often show a lymphocytosis and elevated liver enzymes, changes which are more evident in older children. Diagnosis can be confirmed with the Monospot or Paul Bunnell tests, although these tests can be unreliable in early infection and in younger children, therefore Epstein–Barr virus serology is recommended if there is strong clinical suspicion but a negative infectious mononucleosis screen.

Patient education is mandatory regarding the associated risk of hepatic or splenic rupture with minor trauma in the weeks following infection. In the majority of cases the spleen will return to normal size within 4 weeks post infection. Abdominal ultrasound can be performed to assess splenic size before allowing patients to return to contact sports.

Pharyngitis

Infections of the pharynx are commonly viral in aetiology, and have many pathogens in common with viral tonsillitis. Other viral pathogens include coxsackie, enterovirus (both of which can cause hand, foot, and mouth disease), coronaviruses, and herpes simplex (HSV-1).

GABHS is the most common bacterial cause, but other bacterial aetiologies include group B and C streptococci, *Chlamydia pneumoniae*, *Mycoplasma pneumoniae*, *Haemophilus influenzae*, *Neisseria meningitidis*, *Neisseria gonorrhoeae*, *Arcanobacterium haemolyticum*, *Fusobacterium necrophorum*, and *Corynebacterium diphtheriae*.

Viral and bacterial infections directly invade the pharyngeal mucosa, leading to an inflammatory response and subsequent symptoms. *Candida* infections can occur commonly in infants, in older children with immunocompromise, or those using steroid therapy (e.g. steroid inhalers for asthma). Topical treatment (e.g. nystatin), mouth care, and patient education is often sufficient.

Epiglottitis/supraglottitis

Inflammation of the epiglottis or supraglottis can occur with viral and bacterial throat infections. In children the relative lack of lymph drainage in the epiglottis results in rapid swelling of the epiglottis that can result in acute airway obstruction. Historically caused by *Haemophilus influenzae B*, the success of the HiB vaccine has resulted in a drastic fall in epiglottis in the last 20–30 years, and is more commonly now caused by GABHS, parainfluenza, and staphylococcal infection. The classic triad of rapid onset dysphagia, dysphonia and drooling on the background of URTI should stimulate a high index of suspicion and be treated as a medical emergency. Patients should be initially assessed in a resuscitation room scenario and any formal examination should be performed in the operating theatre with equipment to secure an emergency airway immediately available.

Quinsy

A quinsy is a peritonsillar abscess, caused by lateral spread of the bacterial tonsil infection through the pharyngobasilar fascia into the space between the fascia and the surrounding superior constrictor muscles. This is often associated with trismus due to irritation of the nearby medial pterygoid muscles, and otalgia (as discussed above). Clinical examination can be challenging due to trismus, but fullness of the soft palate, with medialization of the ipsilateral tonsil and deviation of the uvula away to the contralateral side, will be seen.

Quinsy can be treated with aspiration or incision and drainage (Figure 76.8) under local anaesthetic in the emergency department. In young paediatric patients a general anaesthetic will be required to aspirate or incise the abscess and consideration should be given to performing a 'hot' tonsillectomy. If left untreated, a quinsy abscess can progress to a deep neck space infection.

Deep neck space infections

Deep neck space infections are further suppurative complications of bacterial tonsillitis, among other originating pathologies. The interconnections between fascial planes in the neck mean that pus from the peritonsillar space can migrate to the parapharyngeal, retropharyngeal, and danger spaces, ending up in the mediastinum.

Presentation may be similar to a quinsy, but also with associated neck swelling, reduction in neck movement, and airway compromise in severe cases. Contrast computed tomography imaging is important to assess location/spread. Management is commonly surgical with either a transoral or external transcervical approach and identification of a fluctuant swelling will help direct the surgical approach. This can often only be decided on when the patient is examined under anaesthetic, therefore consent for both the external and

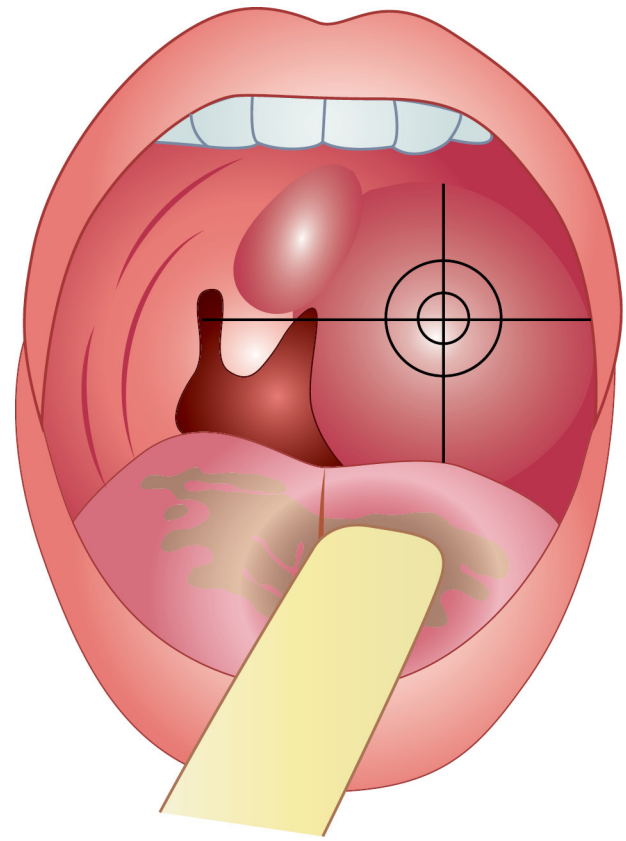

Figure 76.8 Quinsy abscess left tonsil. Targeted drainage and incision is where two imaginary lines meet. The horizontal line is drawn from the base of the uvula and the vertical line along the area of maximum swelling (https://www.i-gp.uk/wellness/wellness-body/health-conditions/quinsy).

the intraoral approach should be sought and the decision on the best approach decided on intraoperatively.

The location of puss can be confirmed by aspiration with a large-bore needle prior to making an incision. The styloid process can be used as a crude landmark on a computed tomography scan, with collections lateral to the styloid more likely to require an external incision via a submandibular approach. Insertion of a large-bore drain to facilitate free drainage is essential. It is a good principle to send puss for microscopy and sensitivity and also a tissue biopsy to exclude rare pathologies.

Lemierre syndrome

This syndrome is characterized by internal jugular vein thrombophlebitis spread via the tonsillar vein secondary to pharyngeal infection most commonly with *Fusobacterium necrophorum*. The sequelae of ensuing bacteraemia and septic emboli include involvement of the chest, bones, and joints. Rarely, intracranial, renal, or haematological complications can ensue.

Diphtheria

Caused by *Corynebacterium diphtheria*, the immunization programmes in the UK and other developed nations have all but eradicated diphtheria. Symptoms of sore throat, lymphadenopathy, skin ulceration, and findings of thick eschar or 'pseudomembrane' covering the tonsils, pharynx, or even larynx would be suggestive. Diphtheria must be considered in the presentation of sore throat in the native unimmunized or migrant population. The treatment is as for suspected bacterial tonsillitis.

Eagle syndrome

Calcification of the stylohyoid ligament, or elongation of the styloid process, can lead to symptoms of chronic sore throat, exacerbated by head movement, which often radiates to the ear (Figure 76.9). Although often a diagnosis of exclusion, cross-sectional imaging can be helpful in determining this as a cause, and it can be managed surgically with a styloidectomy. The styloid can be located via a tonsillectomy approach and separating the superior constrictor muscle fibres in the tonsil fossa by using a haemostat. Patients with persistent symptoms in the absence of styloid or stylohyoid pathology on cross-sectional imaging should be considered for referral to neurology to explore other causes of glossopharyngeal neuralgia.

Oral and dental health

Conditions of the oral cavity may present with sore throat, including dental infection; infective, inflammatory, and ulcerative conditions of the mucosa; and neoplastic processes.

Vitamin deficiency (e.g. vitamin C, B_2, or B_{12}/folate), although unusual, might be considered in the context of persisting or recurrent oral ulceration.

Any concerns or diagnosis of sexually transmitted oral infection must be escalated immediately to the local safeguarding team/unit/paediatric department.

Neoplastic

Lymphoma can present as asymmetrical tonsils causing discomfort, mimicking tonsillitis. True asymmetry is rare and, if associated with cervical or systemic lymphadenopathy or B symptoms (weight loss, night sweats, pyrexia), biopsy of the node or tonsil is mandatory (Figure 75.10). Other malignancies are rare in children but any abnormal mass or non-healing ulceration should be biopsied.

Laryngopharyngeal reflux

Acid reflux is a rare and controversial cause of sore throat in children. It is usually associated with coughing and voice change and in the absence of a definitive diagnostic test is usually a diagnosis of exclusion. A trial course of proton pump inhibitor for 4 weeks can be considered with close monitoring for symptom improvement. Dietary and lifestyle advice can also be discussed with parents.

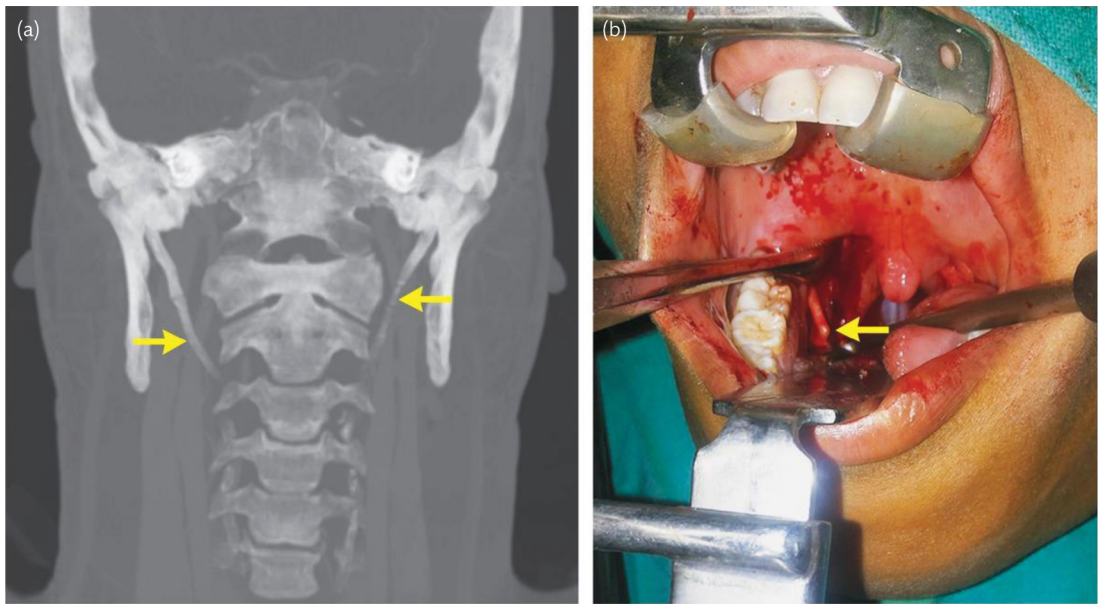

Figure 76.9 Eagle syndrome on coronal CT imaging (a) and intraoperatively (b).
Reproduced from Elmas F, Shrestha BL. Eagle's syndrome. *N Engl J Med*. 2017;377(13):e18.

Figure 76.10 Clinical image of lymphoma of the left tonsil.
Reproduced from Zapater E, Bagán JV, Carbonell F, Basterra J. Malignant lymphoma of the head and neck. *Oral Dis*. 2010;16(2):119–128, with permission from John Wiley and Sons.

Treatment

For the majority of patients with sore throat, simple supportive symptom relief measures such as paracetamol or ibuprofen, fluids, and rest are appropriate. The chief clinical concern for primary care physicians regarding sore throats/tonsillitis and its sequelae is when to consider antibiotic treatment. Two clinical criteria scoring systems (Centor and FeverPAIN) **Boxes 76.1 and 76.2** exist to help determine whether a patient may benefit from antibiotics.

Each of the *FeverPAIN* criteria scores 1 point (maximum score of 5). Higher scores suggest more severe symptoms and likely bacterial (streptococcal) cause. A score of 0 or 1 is thought to be associated with a 13–18% likelihood of isolating streptococci. A score of 2 or 3 is thought to be associated with a 34–40% likelihood of isolating streptococci. A score of 4 or 5 is thought to be associated with a 62–65% likelihood of isolating streptococci.

Each of the *Centor* criteria scores 1 point (maximum score of 4). A score of 0, 1, or 2 is thought to be associated with a 3–17% likelihood of isolating streptococci. A score of 3 or 4 is thought to be associated with a 32–56% likelihood of isolating streptococci.

These tools do not confirm or refute the presence of GABHS, but aim to help make the diagnosis more likely. Antibiotic prescribing should not be used for chronic sore throat, or as 'prevention' for glomerulonephritis or scarlet fever. Trials have suggested that antibiotic prescription for sore throat makes re-attendance more likely, without any reduction in complications and a 'no antibiotic' or 'delayed script' policy may be more appropriate.

If clinically indicated, first-choice antibiotics would be phenoxymethylpenicillin (or erythromycin in penicillin allergy), and consideration of a delayed script may be suitable.

If acutely unwell and particularly in the presence of stridor, progressive difficulty with swallowing, increasing pain, or severe systemic symptoms, acute secondary care referral is mandatory. Patients may present with sepsis and should be treated with intravenous antibiotics (use local microbiological guidelines, but commonly first-line benzylpenicillin), fluids, and steroids if severe swelling or breathing difficulty.

If tonsillitis is recurrent, then consideration for tonsillectomy is warranted. Current UK guidelines based on the 2010 Scottish Intercollegiate Guidelines Network (SIGN) guidance suggest patients experiencing a frequency of seven acute episodes in 1 year, five in 2 consecutive years, or three in 3 consecutive years could benefit from tonsillectomy. Two or more episodes of quinsy, two admissions for severe tonsillitis, or those patients in whom episodes of tonsillitis exacerbate underlying medical conditions (such as psoriasis, nephritis, or periodic fever, aphthous stomatitis, pharyngitis, and adenitis (PFAPA) syndrome), but who do not meet the SIGN criteria, should also be considered for tonsillectomy.

SIGN guidelines

Box 76.3 lists the recommended indications for consideration of tonsillectomy for recurrent acute sore throat in both children and adults.

Various techniques for tonsillectomy exist including cold steel, bipolar, monopolar, and coblation. The intracapsular technique is increasingly being used in children due to its low peri- and postoperative bleeding risk, and quicker and more painless recovery. Adenoidectomy is usually performed in conjunction with tonsillectomy especially if there is an associated history of snoring, nasal obstruction, otitis media, glue ear, or adenoiditis (see Chapter 87).

Box 76.1 FeverPAIN criteria

- **Fever** (during previous 24 hours).
- **P**urulence (pus on tonsils).
- **A**ttend rapidly (within 3 days after onset of symptoms).
- Severely **I**nflamed tonsils.
- **N**o cough or coryza (inflammation of mucus membranes in the nose).

Box 76.2 Centor criteria

- Tonsillar exudate.
- Tender anterior cervical lymphadenopathy or lymphadenitis.
- History of fever (>38°C).
- Absence of cough.

Box 76.3 Indications for consideration of tonsillectomy for recurrent acute sore throat in both children and adults

- Sore throats are due to acute tonsillitis.
- The episodes of sore throat are disabling and prevent normal functioning.
- Seven or more well-documented, clinically significant, adequately treated sore throats in the preceding year ***or***
- Five or more such episodes in each of the preceding 2 years *or*
- Three or more such episodes in each of the preceding 3 years.

Controversies/areas of interest for the future

The use of prophylactic antibiotics is controversial. Patients troubled by recurrent tonsillitis might benefit from an extended course of prophylactic antibiotics leading up to an important occasion such as exams, although high-level evidence for this is lacking.

The recent fall in immunization rates in the UK, associated with the antivaccination movement, socioeconomic deprivation, ethnicity, and a highly mobile population in large cities, may mean that previously rare conditions such as diphtheria and *Haemophilus influenza* type B epiglottitis become more common.

Tonsillectomy has been targeted as a low-priority procedure and the numbers of tonsillectomies performed have fallen to approximately 35,000/year from a height of 80,000 in the 1960s. Many attribute the rising admission rates with severe tonsillitis and its complications with this reduction.

The coronavirus disease 2019 (COVID-19) pandemic and its consequences on healthcare provision may further affect access to tonsillectomy. It will be interesting to see if tonsillitis related hospital admissions increase in the aftermath of the COVID-19 pandemic.

Coblation intracapsular tonsillectomy is fairly well established as the preferred surgical technique in the paediatric patient with sleep-disordered breathing requiring adenotonsillectomy. A large proportion of paediatric ear, nose, and throat surgeons also prefer this technique for patients with recurrent infections as an indication for surgery. The low peri- and postoperative complications make this an attractive option in this patient group.

Tips and pearls

Glandular fever can be reasonably diagnosed in a patient with tonsillitis and a blood picture of lymphocytosis and with a transaminitis or hyperbilirubinaemia, even in the absence of a positive Monospot or Paul Bunnel test (or if these tests are not available). If suspected, but not formally diagnosed, it is still medico-legally indefensible not to educate patients regarding the sequelae (as documented above) of infection. Patients who have any dysfunction of their liver function associated with sore throat should have follow up blood tests arranged to ensure their normalisation. Continued raised LFTs results should prompt further investigation or referral to exclude other causes such as hepatitis.

An acute sore throat in a pre-primary or primary school-age child with pharyngeal ulceration, malaise, and cervical lymphadenopathy is likely to represent primary HSV-1 infection, and will usually resolve within 5–7 days.

Trismus is caused by muscular spasm of the medial pterygoids which are irritated with quinsy and parapharyngeal collections. This spasm resolves with induction of anaesthesia and is therefore not an intubation concern in itself.

Infiltration with local anaesthetic, using a 1 or 2 mL syringe, into the area of maximum swelling of a quinsy can help reduce trismus and improve access when attempting incision and drainage or aspiration.

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77

Rhinorrhoea and epistaxis

Sujata De and Ravi Sharma

Anatomy

In small children, the external nose is soft and has a squared form. The nostrils are angled superiorly relative to the philtrum. This, together with the relative absence of hairs in the nasal vestibule, makes anterior rhinoscopy relatively easy in a child, using a finger to push the tip of the nose upward and an auroscope rather than Thudichum forceps.

The vascular supply of the nose is from branches of both the internal and external carotid artery. Little's area or Kiesselbach's plexus is formed by branches of the ethmoidal and sphenopalatine arteries along with the terminal branches of the facial artery along the anterior cartilaginous septum. This is the most common site of epistaxis in children.

The nose contains relatively more mucosa and lymphoid tissue in a child.

The paranasal sinuses develop as outlined in Table 77.1. The presence of ethmoid and maxillary sinuses at birth explains the propensity of orbital involvement as a result of complicated rhinosinusitis in young children, while frontal and sphenoid sinus complications are likely to occur in an older child.

Rhinosinusitis

Paediatric rhinosinusitis is defined as:

- the presence of two or more symptoms, one of which should be either nasal congestion/obstruction/blockage or nasal discharge (anterior or posterior nasal drip); with or without facial pain/pressure; with or without cough; and *either*
- endoscopic signs of nasal polyps and/or mucopurulent discharge/oedema/mucosal obstruction in middle meatus, *or*
- computed tomography (CT) changes (mucosal changes in the ostiomeatal complex or sinuses).

School-aged children are thought to have seven to ten upper respiratory tract infections of viral origin per year. Of these, 0.5–2% may progress to develop bacterial acute rhinosinusitis (ARS) infections.

Acute rhinosinusitis

ARS is defined as the sudden onset of three or more symptoms/signs of nasal congestion/obstruction/blockage; discoloured nasal discharge; cough; facial or dental pain; fever greater than 38°C; or 'double sickening', for less than 12 weeks. Double sickening is a term describing worsening of symptoms after about 5 days of upper respiratory tract infection, following a perceived initial improvement. Recurrent ARS is, by definition, four or more episodes of ARS with symptom-free intervals.

Factors predisposing to acute bacterial sinusitis are described in Box 77.1.

Table 77.1 Development of paranasal sinuses

Paranasal sinus	Status at birth	Growth
Maxillary	Present	Rapid growth from 3 years; adult size at 15 years
Ethmoid	Present	Adult size by 12 years
Frontal	Not present	Reaches adult size by late teenage years
Sphenoid	Not present	Reaches sella turcica by 7 years of age and dorsum sella by late teens; adult size by 15 years

Box 77.1 Predisposing factors for acute bacterial rhinosinusitis

- *Environmental*: active and passive smoking.
- *Odontogenic infections*: a projecting tooth root into the maxillary sinus or a frank oral antral fistula can be identified on the CT scan.
- *Allergy*: there is little support for the presence of allergic rhinitis as a risk factor for developing acute rhinosinusitis.
- *Ciliary impairment*: impaired mucociliary clearance in AR patient predisposes the patient to ARS.
- *Iatrogenic*: sinus surgery, nasogastric tubes, mechanical ventilation.
- *Immunodeficiency*: HIV infection, immunoglobulin deficiency, cystic fibrosis, Kartagener's syndrome.
- *Mechanical obstruction*: deviated nasal septum, nasal polyps, foreign body, tumour, granulomatosis with polyangiitis; presence of concha bullosa and sphenoethmoidal cell (Onodi cell) are likely to be of causal significance.
- *Mucosal oedema*: preceding viral infection, allergic rhinitis, vasomotor rhinitis.

Source data from Fokkens WJ, Lund VJ, Hopkins C, et al. European position paper on rhinosinusitis and nasal polyps 2020. *Rhinology*. 2020;58(Suppl S29):1–464.

Pathophysiology

ARS can be divided into viral rhinosinusitis (common cold), post-viral rhinosinusitis, and acute bacterial rhinosinusitis.

Viral rhinosinusitis is an acute rhinitis induced by the respiratory viruses including rhinovirus, respiratory syncytial virus, influenza and parainfluenza viruses, coronavirus, adenovirus, and enterovirus. The nasal epithelium is the primary port of entry for the viruses (Figure 77.1) by binding to surface receptors followed by viral replication in the airway. The epithelial cells release antimicrobial surfactants and mucus to delay pathogen transmission in the airway. They also express and secrete various cytokines and chemokines to drive the immune responses against the invading pathogens which result in mucosal oedema and impairment of the mucociliary clearance function. The resultant effects result in obstruction of the sinus ostia and poor clearance.

Acute bacterial rhinosinusitis is indicated by the presence of three or more of the following five presentations: discoloured discharge, severe pain, fever (>38°C), an elevated erythrocyte sedimentation rate/C-reactive protein, or deterioration after an initial improvement. Common pathogens include *Streptococcus pneumoniae*, *Haemophilus influenza*, and *Moraxella catarrhalis*.

Treatment

Treatment of patients with ARS should be symptomatic, with possibly local corticosteroids. Antibiotics are not recommended unless the clinical signs of severe disease are present—high fever, double sickening, severe pain, and elevated erythrocyte sedimentation rate.

Procalcitonin is a potential blood biomarker indicating severe bacterial infection and is being investigated as a tool for guiding antibiotic prescribing in the community.

Chronic rhinosinusitis

Paediatric chronic rhinosinusitis (CRS) is defined as the presence of two or more symptoms, one of which should be either nasal blockage/obstruction/congestion or nasal discharge (anterior/posterior nasal drip); with or without facial pain/pressure and/or cough for at least 12 weeks, with validation by telephone or interview.

Predisposing factors for CRS in children are listed in Box 77.2.

Assessment

- *CT scans*: routine use is not recommended unless suspecting complications or contemplating surgery. It has been suggested that a Lund–Mackay score of greater than 5 should be used as a cut-off to signify disease.
- *Quality-of-life questionnaire*: the only validated disease-specific quality-of-life questionnaire for paediatric CRS is the Sinus and Nasal Quality of Life Survey (SN-5) questionnaire.
- *Nasal endoscopy*: is essential in the diagnosis of CRS.
- *Allergy testing*: skin prick test or blood tests (radioallergosorbent test). In very young children, specific immunoglobulin E (IgE)

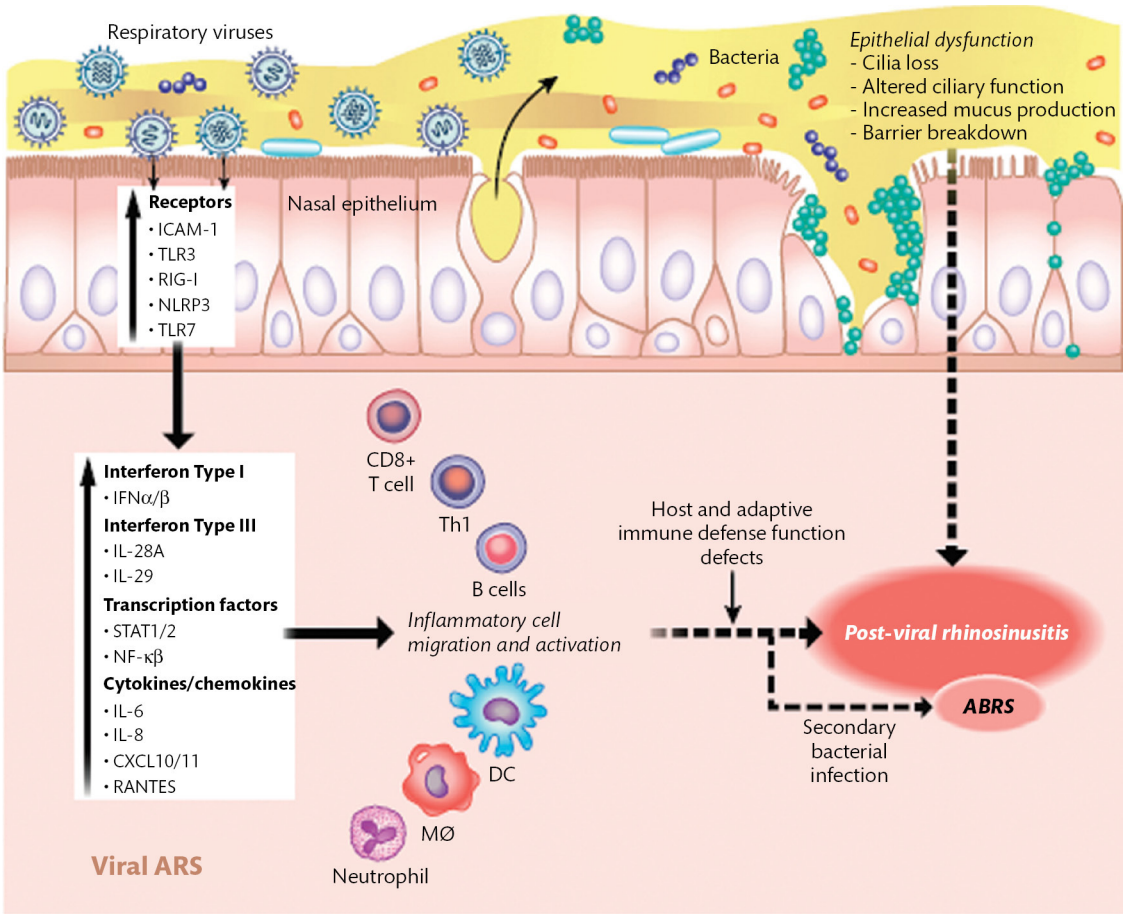

Figure 77.1 Pathophysiology of acute rhinosinusitis.
Reproduced from Fokkens WJ, Lund VJ, Hopkins C, et al. European position paper on rhinosinusitis and nasal polyps 2020. *Rhinology*. 2020;58(Suppl S29):1–464.

can be elevated without clinical significance and return to normal levels during development.

- *Smell test*: it is feasible to use 'sniffin sticks' in children but one should be aware that cultural differences can influence the performance of subjects on the test.
- *Ciliary morphology and function tests*: if required, are best done by paediatric respiratory physicians.
- *Genetic testing*: routine genetic testing is not advised unless cystic fibrosis or primary ciliary dyskinesia are suspected.

Box 77.2 Predisposing factors for paediatric chronic rhinosinusitis

- *Anatomical variations*: although more common in older children, they do not seem to be related to the presence of CRS. Similarly, viral upper respiratory tract infection and environmental factors are not related.
- *Environmental factors*: exposure to tobacco smoke inhibits mucociliary clearance and epithelial regeneration. Both active and passive smoking have been identified as significant risk factors for development of CRS.
- *Allergic rhinitis* (AR): the evidence base for allergy to predispose to CRS is poor. Recent studies have suggested that allergic fungal rhinosinusitis and central compartment atopic disease may have some association with chronic AR.
- *Asthma*: asthma is present in 25–30% of patients with CRS.
- *Adenoids*: adenoids have a significant impact on the development of CRS in younger children. They may contribute to the development of CRS by mechanical obstruction but also by serving as a bacterial reservoir. Symptoms of CRS improve after adenoidectomy in children.
- *Gastro-oesophageal reflux disease* (GORD): although GORD is frequently found in children with CRS, based on the current evidence routine treatment of GORD in children with CRS is not recommended.
- *Immunological factors*: there is good evidence that the immune deficiencies are common in patients with CRS. The most frequent immune deficiency found in these cases is common variable immunodeficiency in 10% of cases and selective IgA deficiency (6%).
- *Bacteria and biofilms*: a biofilm comprises of any syntrophic consortium of microorganisms in which cells stick to each other and often also to a surface. They become embedded within a slimy extracellular matrix composed of extracellular polymeric substances. Microbes form a biofilm in response to various different factors which include cellular recognition of specific attachment sites on the surface, nutritional cues, or by exposure of planktonic cells to subinhibitory concentration of antibiotics. The organisms implicated in forming a biofilm include *Staphylococcus aureus* (associated with severe recurrent or recalcitrant cases), *Pseudomonas aeruginosa*, *Haemophilus influenza*, and *Moraxella catarrhalis*.
- *Cystic fibrosis*: cystic fibrosis is an autosomal recessive disease caused by mutations in the *CFTR* gene which results in formation of thick viscous secretions leading on to infections of the respiratory tract. *Pseudomonas aeruginosa* and *Staphylococcus aureus* are the most frequent culprits. It is the most common disorder encountered in children with nasal polyps.
- *Ciliary impairment*: primary ciliary dyskinesia is an autosomal recessive disorder with the incidence of 1 in 15,000–20,000 births. It is associated with frequent ear and sinus infections. Specimen diagnosis requires examination cilia by light and transmission electron microscopy. The most common described abnormality consists of a lack of the outer dynein arms. Kartagener's syndrome comprises the triad of rhinosinusitis, chronic bronchitis, and bronchiectasis, and situs invertus occurs in 50% of patients with primary ciliary dyskinesia.

Source data from Fokkens WJ, Lund VJ, Hopkins C, et al. European position paper on rhinosinusitis and nasal polyps 2020. *Rhinology*. 2020;58(Suppl S29):1–464.

Treatment

Treatment includes a combination of nasal saline irrigation and intranasal steroids, in addition to antihistamines when the symptoms are related to environmental factors. Antibiotics are indicated for infectious cases (purulent nasal discharge, foetid breath, and/or maxillary sinus fluid level).

Intranasal steroids are recommended for use in children with CRS despite the absence of good evidence, based on safety in children and favourable efficacy data in adults. Surgery is indicated for failure of medical treatment. Initial surgery might consist of an adenoidectomy and an antral washout.

Functional endoscopic sinus surgery, in the form of nasal polypectomy, middle meatal anstrostomy, and anterior ethmoidectomy, is indicated after failure of adenoidectomy in older children.

Complications of rhinosinusitis

In children, orbital complications arise, in order of decreasing frequency, from the ethmoid, maxillary, frontal, and, rarely, the sphenoid sinus.

According to Chandler's classification, orbital complications may be divided into five stages based on the clinical and radiological findings (Table 77.2).

Infection spreads into the orbit from the ethmoid sinus either directly across the lamina papyracea (Figure 77.2) or by vascular means.

Indications for surgical intervention are listed in Box 77.3.

Treatment of complications

The patient should be admitted under the joint care of the paediatrician, and ear, nose, and throat and ophthalmic surgeons.

- Stage 1—oral antibiotics.
- Stage 2—intravenous antibiotics (ceftriaxone)/nasal decongestants/ophthalmic review to test visual acuity, eye movements, and colour vision.
- Stage 3 and 4: intravenous antibiotics and drainage of abscess (either endoscopically or externally) and paranasal sinus washouts.
- Stage 5: high-dose intravenous antibiotics; anticoagulation therapy and endoscopic sinus surgery.

The most common osseous complication is osteomyelitis of the maxillary (in infancy) and frontal bones. Frontal sinus infection can cause osteitis and osteomyelitis of frontal sinus wall and may develop subperiosteal abscess with soft issue swelling and doughy pitting oedema of the anterior wall (Pott's puffy tumour—Figure 77.3).

Similar concomitant infection of the posterior wall may result in meningitis, epidural or subdural empyema, and brain abscess (Figure 77.4).

Intracranial extension is suspected in the presence of superior eyelid oedema, high fever, severe headache, meningeal irritation, photophobia, papillary oedema, coma, and focal neurological signs. Treatment consists of long-term antibiotics with endoscopic or minimal external drainage. Surgical debridement of sequestered bone is no longer considered necessary in uncomplicated Pott's puffy tumour.

Table 77.2 Chandler's classification of orbital complications, features, and radiological findings

Stage	Type	Features	Radiology
Stage 1	Preseptal cellulitis	Inflammation anterior to orbital septum—pain eyelid oedema and erythema	CT—soft tissue swelling better seen on MRI T2-weighted images
Stage 2	Orbital cellulitis	Proptosis; limited movement; chemosis	CT—diffuse swelling within the orbit
Stage 3	Subperiosteal abscess	Above + ophthalmoplegia; raised immature leucocyte count	Oedema of medial rectus; lateralization of periorbital (rim enhancement)
Stage 4	Orbital abscess		Obliteration of extraocular muscles and optic nerve by a confluence mass; air in orbit
Stage 5	Cavernous sinus thrombosis	Intracranial complications—throbbing headaches; ophthalmic neuralgia; papilloedema; spiking temperature; meningeal irritation	Magnetic resonance venogram/high-resolution CT with contrast– filling defect

Source data from Fokkens WJ, Lund VJ, Hopkins C, et al. European position paper on rhinosinusitis and nasal polyps 2020. *Rhinology*. 2020;58(Suppl S29):1–464.

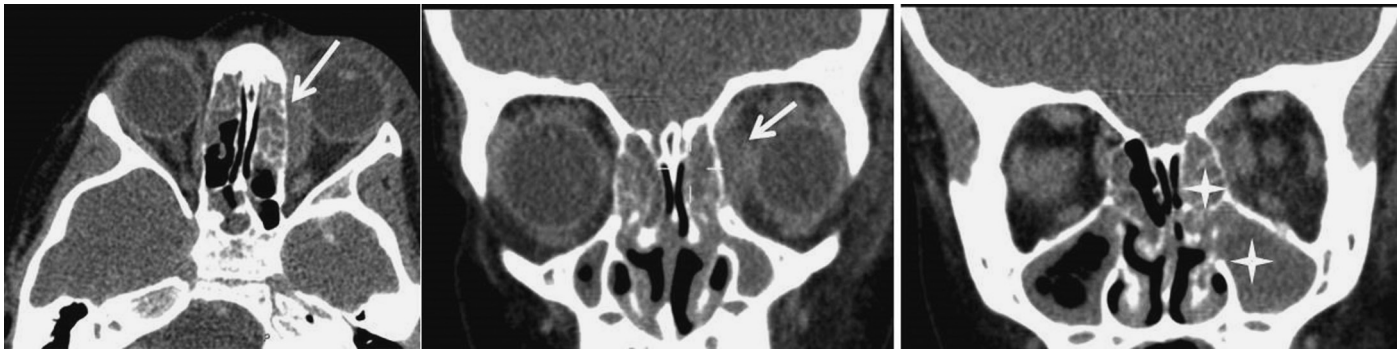

Figure 77.2 Axial and coronal CT scans showing inflammatory mass/early abscess (arrows) formation. Ethmoid and left maxillary arum are opacified (star).

Allergic rhinitis

Allergic rhinitis (AR) is an IgE-mediated chronic inflammatory disease of the nasal mucosae triggered by environmental aeroallergens.

Assessment

Symptoms included nasal congestion, itching, sneezing, watery rhinorrhoea, and possibly loss of smell in severe cases. Ocular symptoms of tearing, itching, and redness increase the likelihood. Patients or their carers may be able to identify triggers, such as pet exposure, or a specific time of year when symptoms worsen, and it can be helpful to elicit these history points to guide avoidance measures (discussed later in this chapter). Typical examination findings in AR patients are described in Box 77.4.

Box 77.3 Indications for surgical intervention

- Subperiosteal abscess or intraorbital abscess.
- Decreased red colour vision/ophthalmoplegia/reduced visual acuity.
- Progression or no improvement in orbital signs (diplopia, proptosis, swelling, chemosis) despite intravenous antibiotics for 48 hours.
- No improvement in the general condition after 48 hours of intravenous antibiotics.

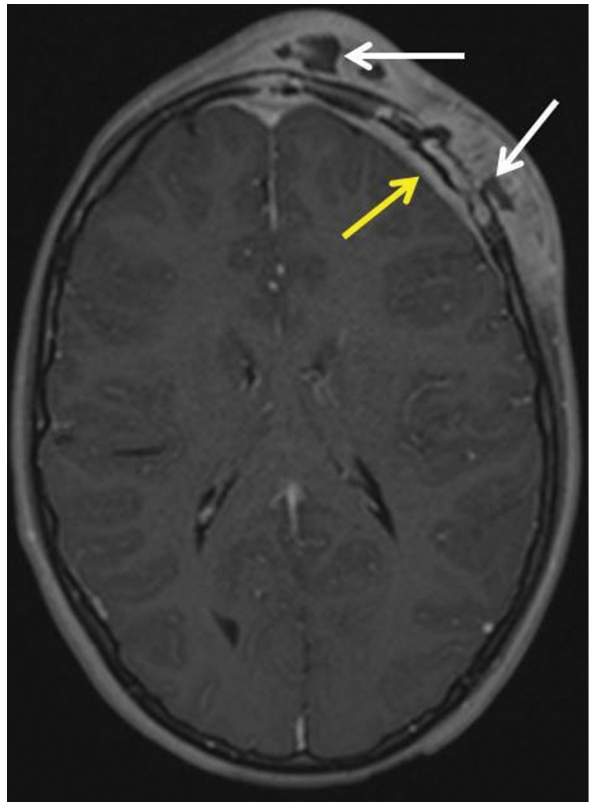

Figure 77.3 Pott's puffy tumour: T1-weighted axial MRI scan showing soft tissue thickening and filling defect of frontal bone resulting from

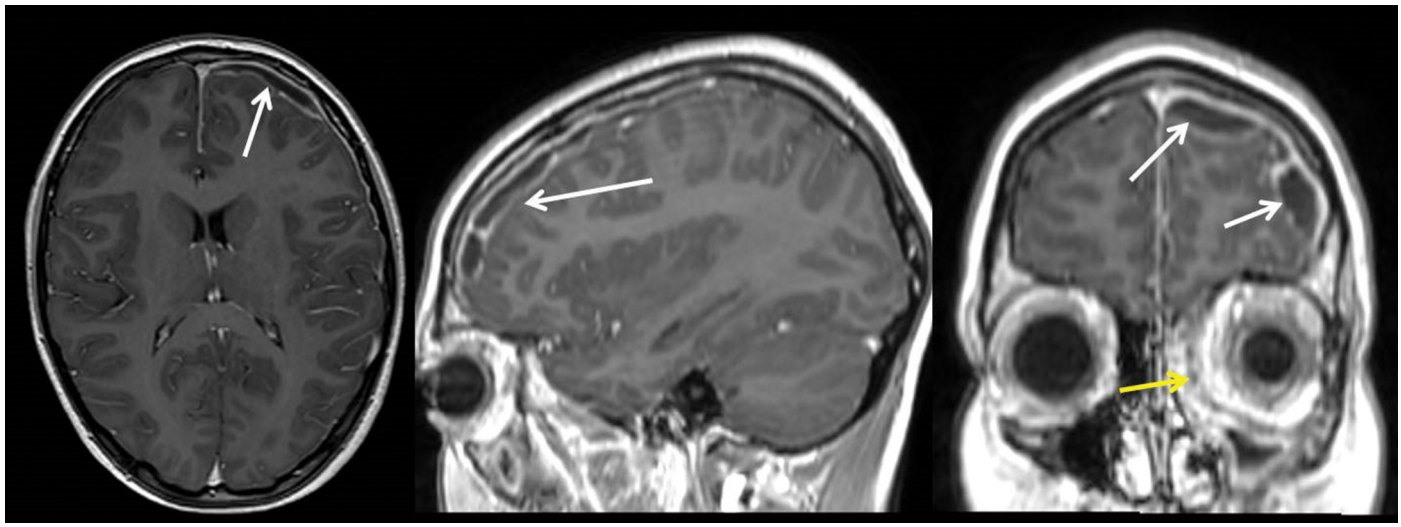

Figure 77.4 From left to right, axial, sagittal, and coronal T1-weighted MRI scans demonstrating ethmoid sinusitis (yellow arrow) and subdural empyema (white arrows).

Box 77.4 Typical examination findings in allergic rhinitis

- *Allergic shiners*: these occur because of infraorbital oedema from venodilation related to blood vessel changes in the context of allergic inflammation.
- *Dennie–Morgan lines*: these consist of increased folds or lines below the lower eyelid and are more common in patients with AR. The pathophysiology is not precisely understood. These lines do not always denote AR and can be more common in some ethnic groups without an increase in AR.
- *Allergic salute*: this is a behaviour related to nasal itching and rhinorrhoea consisting of repeated rubbing of the nose. The repeated pushing of the tip of the nose up with the hand leads to a transverse nasal crease.
- *Allergic facies*: typical allergic facies consist of a high-arched palate, mouth breathing, and dental malocclusion. This is generally seen in children with early-onset AR.
- *Pale nasal mucosa*: with anterior rhinoscopy, the nasal mucosa may appear pale and blue coloured with turbinate oedema. This may be accompanied by visible clear rhinorrhoea (anterior or posterior in oropharynx).
- *Cobblestoning*: the posterior oropharynx may develop hyperplastic lymphoid tissue leading to a 'cobblestone' appearance of the mucosa.

Classification

AR can be classified as seasonal (SAR) or perennial (PAR) depending on the allergens occurring seasonally (tree, grass, or weed pollen) or throughout the year (house dust mite, animal dander, or some moulds).

The Allergic Rhinitis and its Impact in Asthma (ARIA) group classifies AR dependent on the duration of symptoms (Figure 77.5).

Non-allergic rhinitis

This is a chronic rhinitis with at least two symptoms (nasal obstruction, rhinorrhoea, postnasal drip sneezing, or nasal/ocular itching), at least 1 hour daily for at least 12 weeks per year with all allergic tests being negative. Sub-phenotypes include hormonal, medication-induced, gustatory, or idiopathic rhinitis.

Pathophysiology

Antigen-presenting cells, such as dendritic cells, process allergens and present some peptides from them on the major histocompatibility complex class II molecules. This bound complex results in the

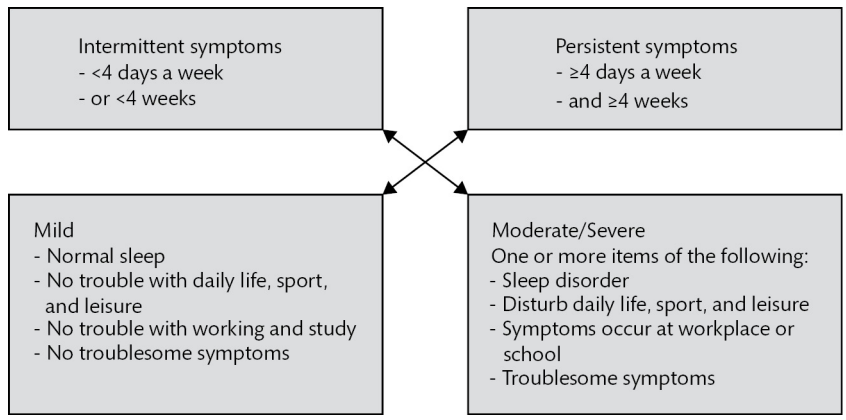

Figure 77.5 ARIA group classification of allergic rhinitis.

Reproduced from Klimek L, Bachert C, Pfaar O, et al. ARIA guideline 2019: treatment of allergic rhinitis in the German health system. *Allergol Select*. 2019;3(1):22–50, doi: 10.5414/ALX02120E (Creative Commons Attribution License).

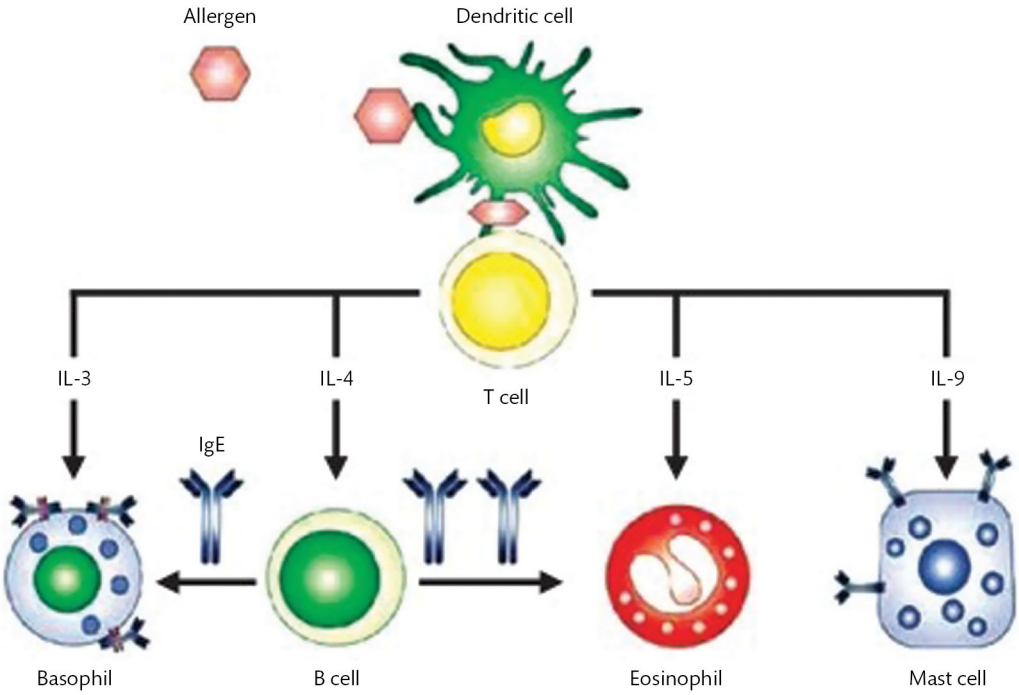

Figure 77.6 Initial response to allergen molecule. IL, interleukin.

Reproduced from Min YG. The pathophysiology, diagnosis and treatment of allergic rhinitis. *Allergy Asthma Immunol Res*. 2010;2(2):65–76, doi: 10.4168/aair.2010.2.2.65, with permission from The Korean Academy of Asthma, Allergy and Clinical Immunology.

differentiation of naïve CD4 T cells into allergen-specific Th2 cells. Activated Th2 cells secrete cytokines which induce B cells to produce IgE antibodies and promote the proliferation of eosinophils, mast cells, and neutrophils (Figure 77.6).

On exposure to allergens, two different patterns of allergic reactions may develop in patients with AR. An early response, characterized by sneezing and rhinorrhoea, occurs within 30 minutes and then subsides. This is thought to be due to stimulated mast cells secreting chemical mediators such as histamine, prostaglandins, and leukotrienes. In contrast, a delayed response of nasal obstruction occurs approximately 6 hours after exposure and subsides slowly. This is mediated by the migration of eosinophils, mast cells, and T cells to the nasal mucosa, where they contribute to tissue breakdown and remodelling (Figure 77.7).

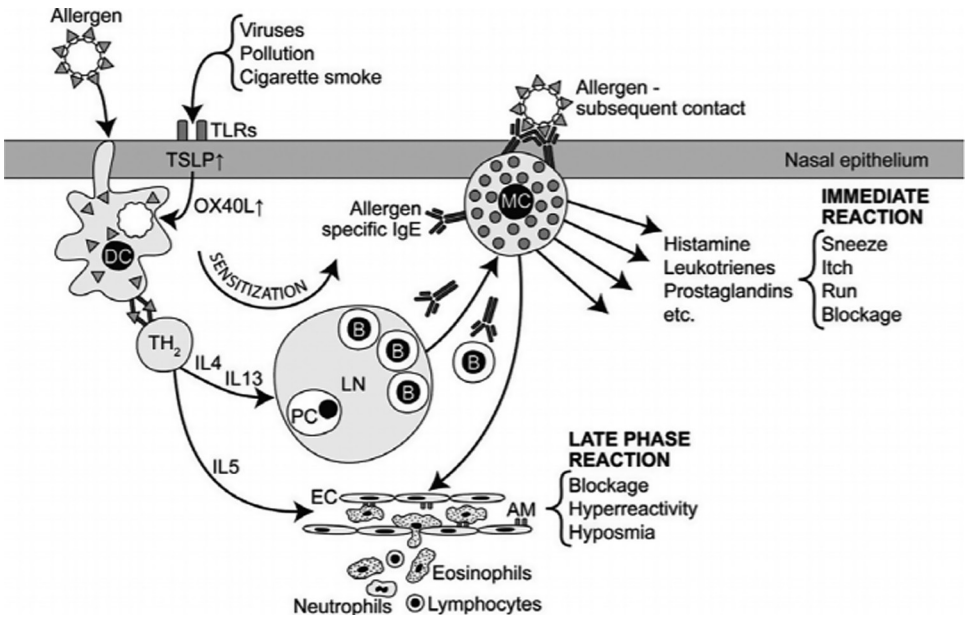

Figure 77.7 Immunological mechanisms of allergic rhinitis.

Reproduced from Scadding GK, Kariyawasam HH, Scadding G, et al. BSACI guideline for the diagnosis and management of allergic and non-allergic rhinitis (Revised Edition 2017; First edition 2007). *Clin Exp Allergy*. 2017;47(7):856–889, doi: 10.1111/cea.12953, with permission from John Wiley and Sons.

Investigations

Diagnosis is based either on skin prick tests or serum IgE levels of clinically relevant allergens using a radioallergosorbent test. A raised total IgE level is not exclusive to allergy and is not specific to AR. Skin prick tests can be influenced by antihistamines which the patient should stop 3 days before clinic appointments.

Treatment

Current treatment options include patient education/prevention, pharmacotherapy, and allergen immunotherapy.

Prevention

- A mobile phone app, MASK diary, freely available on Google and Apple stores, is useful in monitoring symptoms, medication usage, and quality of life more efficiently and in a timely manner.
- Avoiding active and passive tobacco smoke exposure helps with reducing AR. In children with AR, immunotherapy may prevent development of secondary sensitization.
- Avoidance of pets causing AR helps with controlling symptoms.

Medications

See Figure 77.8 and Table 77.3.

The British Society of Allergy and Clinical Immunology recommended the algorithm shown in Figure 77.9 for managing allergic rhinitis in 2017.

Recently, a combination of nasal corticosteroid (fluticasone propionate) and nasal antihistamine (azelastine) has been shown to be effective and might be better choice for AR treatment in children over 12 years of age.

Allergen immunotherapy

Conventional immunotherapy for AR includes subcutaneous immunotherapy and sublingual immunotherapy, and involves giving patients extracts containing allergens to which they produce specific IgE in order to induce immune changes and a desensitized state. Treatment needs to be commenced before the start of the season and continues for 3 years.

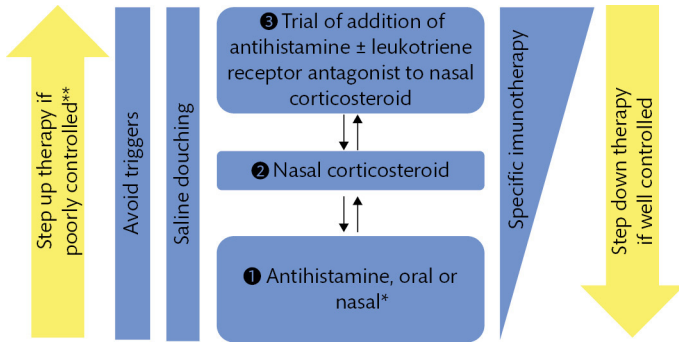

Figure 77.8 Approach to therapy for paediatric allergic rhinitis.
Reproduced from Scadding GK, Kariyawasam HH, Scadding G, et al. BSACI guideline for the diagnosis and management of allergic and non-allergic rhinitis (revised edition 2017; first edition 2007). *Clin Exp Allergy*. 2017;47(7):856–889, doi: 10.1111/cea.12953, with permission from John Wiley and Sons.

- *Subcutaneous immunotherapy*: consists of injecting subcutaneously, serially increasing concentrations of allergen until the maximum is reached and then spacing out the frequency of administration over time. This can be given for 3–5 years and is known to reduce the humoral response to allergen (IgE and subclasses of IgGs) and sensitivity of the end organs, and increases tolerance of allergens mediated by the T-cell response.
- *Sublingual immunotherapy*: a Timothy grass pollen extract is approved down to 5 years old. A five-grass extract is approved down to 10 years old. Dust mite and ragweed extracts are approved only starting at age 18 years.
- *Surgery*: surgery has no role in the treatment of allergic rhinitis. Short-term relief can be obtained by reducing the turbinates (submucosal diathermy or turbinoplasty).

Table 77.3 Commonly used medication for allergic rhinitis

Drug	Dosage	Administered	Adverse effect	Cautions
Cetirizine	Children aged 1 year: 250 mcg/kg twice daily (off-label) Children aged 2–5 years: 2.5 mg twice daily Children aged 6–11 years: 5 mg twice daily Children aged ≥12 years: 10 mg once daily	Orally	Blurred vision and dry mouth	Not licensed <1 year Renal impairment
Loratadine	Children aged 2–11 years: 5 mg once daily Children aged >12 years: 10 mg once daily	Orally		Not licensed <2 years Hepatic impairment
Azelastine hydrochloride	>6 years' age	Intranasal spray	Irritation of nasal lining	Bitter taste
Ipratropium bromide	>6 years' age: 2 puffs, three times daily	Intranasal spray	Epistaxis, nasal dryness	Cystic fibrosis
Sodium cromoglycate	1 puff, 3–4 times a day	Intranasal spray	Nil	Nil
Fluticasone	1–2 sprays, once daily, > 4 years	Intranasal spray	Epistaxis, irritation	Needs regular usage
Mometasone	1–2 sprays, twice daily, >3 years	Intranasal spray	Epistaxis, irritation	Needs regular usage
Montelukast	6 months–5 years: 4 mg once daily 6–14 years: 5 mg once daily >15 years: 10 mg once daily	Orally	Headaches, abdominal pains, Thirst	Advised in the evening Galactose intolerance
Saline washes	Three times daily	Intranasal	Epistaxis	Nil

Note: refer to *Children's BNF* for alternatives.

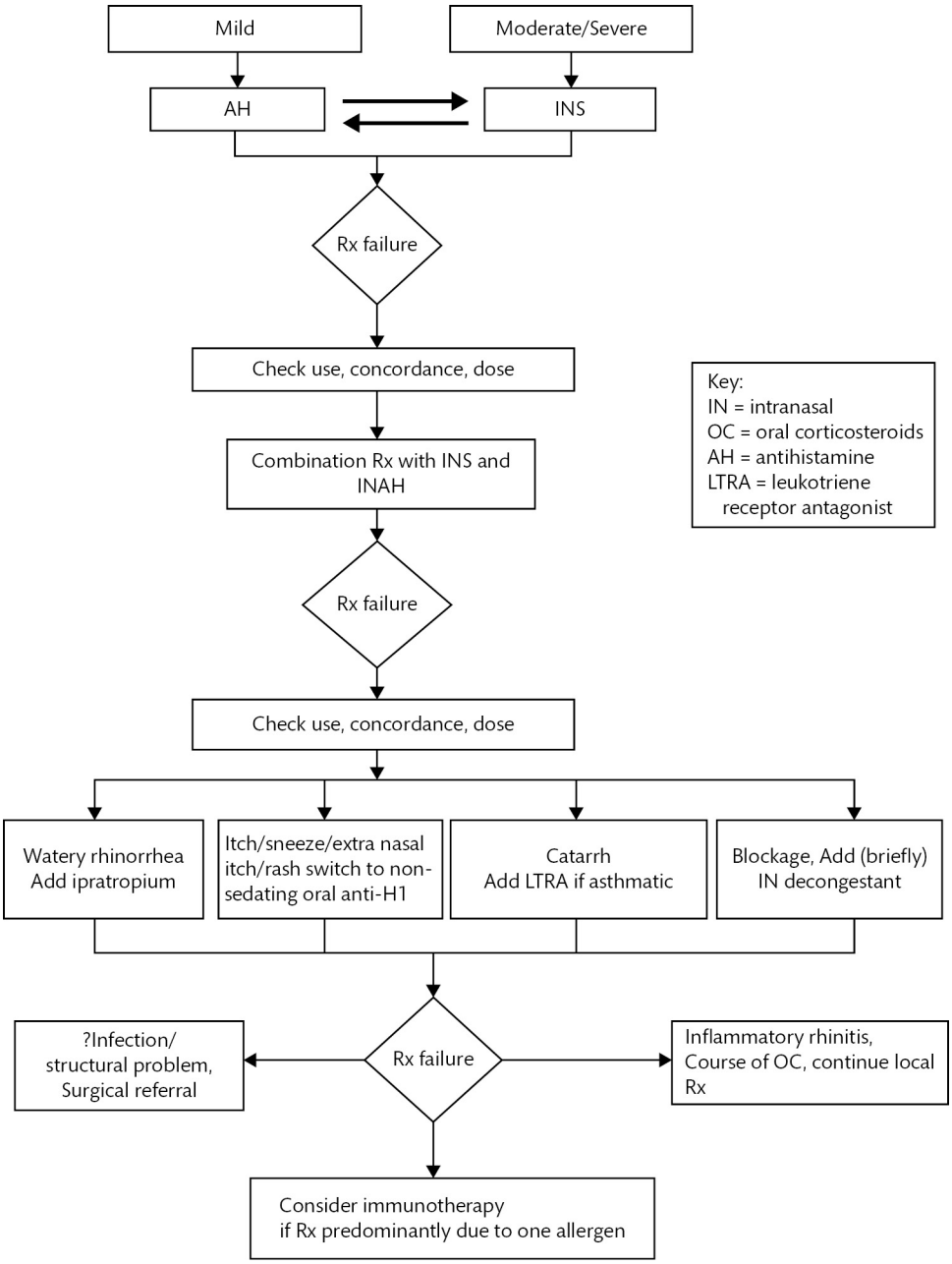

Figure 77.9 British Society of Allergy and Clinical Immunology allergic rhinitis treatment algorithm, 2017. INS, intranasal corticosteroids.

Reproduced from Scadding GK, Kariyawasam HH, Scadding G, et al. BSACI guideline for the diagnosis and management of allergic and non-allergic rhinitis (revised edition 2017; first edition 2007). *Clin Exp Allergy*. 2017;47(7):856–889, doi: 10.1111/cea.12953, with permission from John Wiley and Sons.

Epistaxis

Introduction

Epistaxis is very common in young children and in most cases is mild, intermittent, and settles without the need for anything more than first aid. The commonest cause is trauma in the area of the anterior septum. This might be from nose picking but bleeding can occur spontaneously either while the child is awake or sometimes while asleep. This can lead to significant parental alarm.

Epistaxis, however, can be a presenting symptom of either systemic pathology (e.g. coagulopathies), rarer conditions such as juvenile nasopharyngeal angiofibroma (JNA), or of a foreign body, or indeed non-accidental injury. It is therefore important to be aware of these possibilities and to be alert to other symptoms and signs that may point to a less benign cause (**Box 77.5**).

A child with epistaxis usually presents with a history of recurrent bleeds occurring from one or both nostrils that usually settle with first aid measures (lean forward, ice packs, pressure applied to both nasal alae). The source of bleeding is usually from the submucosal

Box 77.5 Symptoms that should trigger concern

- Severe bleeding with clots.
- Bleeding that does not settle after 20 minutes despite adequate first aid measures.
- Underlying bleeding disorders or symptoms suggesting an underlying coagulopathy or anaemia.
- Unilateral symptoms associated with obstruction, foul smelling discharge, or decreased facial sensation.
- Epistaxis in a young baby—consider non-accidental injury.
- Adolescent boy—exclude JNA.

arterial plexus at the anterior part of the septum (Little's area) or the retrocollumellar vein.

Management

If the epistaxis is severe, then resuscitation measures should be instituted. Packs such as Rapid Rhino (Smith & Nephew) can be used to control severe bleeds that do not respond to simple first aid measures. Once bleeding disorders have been excluded or treated, the pack can be removed after 24 hours and examination carried out, if necessary with the aid of a rigid or flexible nasendoscope to determine the source of the bleed and to exclude a mass. Investigations as dictated by the clinical presentation should be instigated.

However, severe epistaxis in children is rare and the vast majority of nose bleeds settle with pressure and first aid measures such as ice packs. Children usually present as outpatients referred with recurrent, relatively minor but troublesome bleeds. In this situation, having excluded symptoms that should trigger concern as detailed above, the child should be examined to identify the likely source of bleeding (usually anterior) (Box 77.6).

Box 77.6 Treatment options

- Conservative management with first aid measures alone and avoidance of trauma.
- A 10-day course of Naseptin (chlorhexidine hydrochloride and neomycin sulfate antiseptic cream), Vaseline (petroleum jelly), or Bactroban (mupirocin 2% ointment).
- If this is unsuccessful, then application of 75% silver nitrate cautery followed by a course of Naseptin.

Juvenile nasopharyngeal angiofibroma

JNA is a rare, benign, but locally invasive vascular tumour that occurs exclusively in males usually in their second or third decade of life.

Assessment

It usually presents with unilateral nasal obstruction or epistaxis that is severe. Headache and rhinorrhoea are other symptoms. The tumour is often large and invasive on presentation in which case symptoms might also include proptosis, cranial nerve involvement, visual disturbances, and facial swelling. The mass is usually visible on examination of the nose either with anterior rhinoscopy or endoscopic examination. It appears as a firm red or purple mass.

Investigation

Imaging involves contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI). CT scanning usually demonstrates an enhancing mass originating from the region of the sphenopalatine foramen. The mass might extend into the nasal cavity, the nasopharynx, the anterior cranial fossa, the pterygopalatine fossa, and the paranasal sinuses (Figures 77.10 and 77.11).

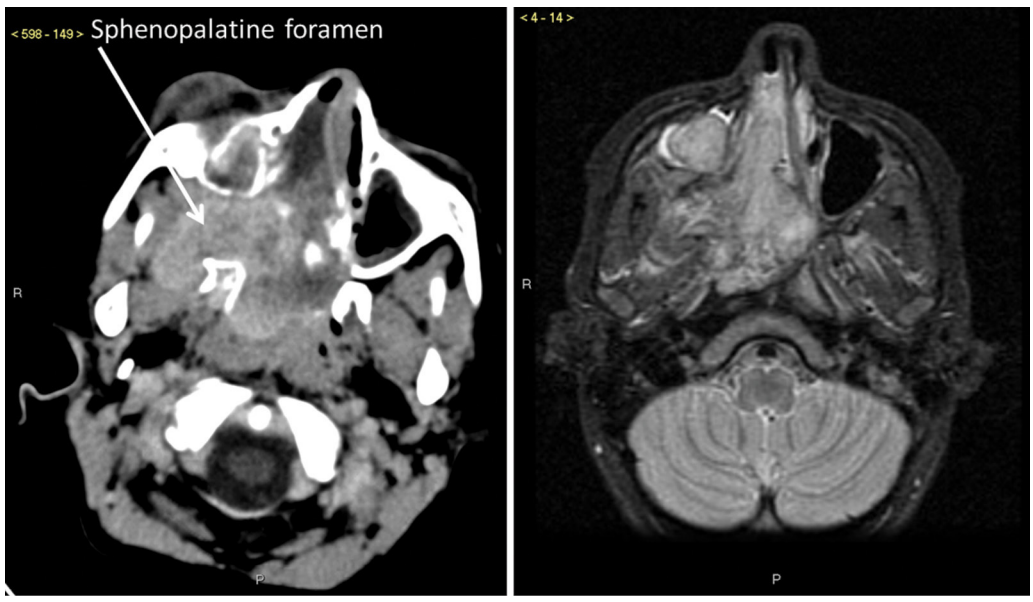

Figure 77.10 Axial contrast-enhanced CT and Stir sequence MRI showing widening of the sphenopalatine foramen occupying the nose, postnasal space, and extending lateral to the pterygoid plates arising from the sphenopalatine foramen.

Figure 77.11 Post-contrast T1-weighted MRI scan showing an extensive right-sided JNA.

CT angiography is useful to determine the vascularity and the feeder vessels that would require embolization prior to surgery (Figure 77.12).

Staging

Multiple staging systems have been proposed and used to aid in the management of this condition. The aims of a staging system are to direct management and/or to predict outcomes.

The staging system proposed by Radowski et al. in 1996 is commonly used (Table 77.4). Like other systems proposed at or before that time, it is based upon the extent of the tumour and sites of involvement. The mainstay of surgical treatment at that time was an open approach.

As endoscopic techniques have evolved, the endoscopic approach is used in almost all cases in combination with open approaches for more extensive tumours. Moreover, as perioperative embolization has become standard practice, staging systems have been modified to take this into account.

The University of Pittsburgh Medical Center staging system was proposed in 2010 (Table 77.5). Extension into a sinus was not felt to affect prognosis given the ability of endoscopic techniques to surgically access and clear disease from these areas. The response to embolization was felt to affect outcomes in terms of intra- and postoperative bleeding and tumour clearance and so has been incorporated into this system, which therefore can only be done post embolization (i.e. 1 day prior to surgery). This system helps to direct

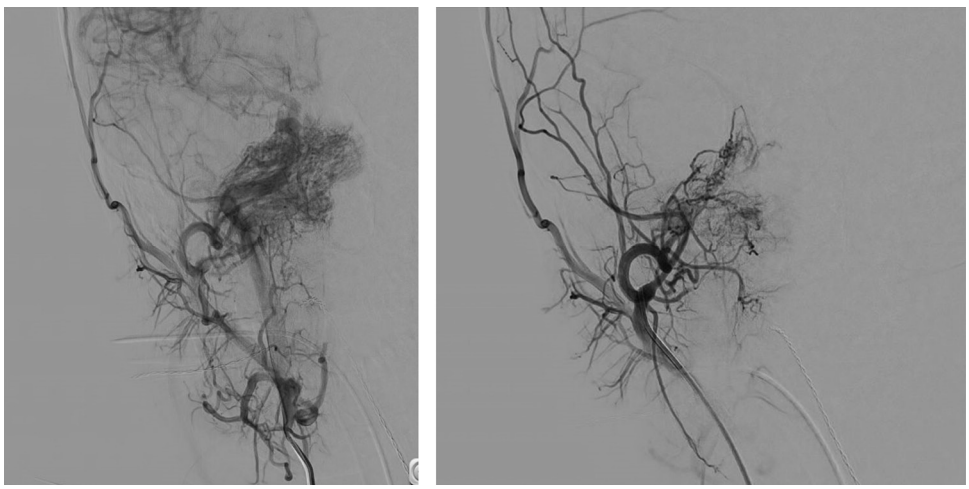

Figure 77.12 Pre- (left) and post-embolization (right). Note the reduction of the vascular 'blush' in figure on the right following embolization of the internal maxillary artery.

Table 77.4 Radowski's classification

Stage		
I	a	Limited to nose of nasopharynx
	b	Ia + extension to one or more sinus
II	a	Minimal extension through sphenopalatine foramen and into medial pterygomaxillary fossa
	b	Full occupation of pterygopalatine fossa, displacing posterior wall of maxillary sinus forward, orbit erosion, displacement of maxillary artery branches
	c	Extension into infratemporal fossa, cheek, posterior to pterygoid plates
III	a	Erosion of skull base with minimal intracranial extension
	b	Erosion of skull base with extensive intracranial extension +/− cavernous sinus

the surgical approach (stages I and II, endoscopic; stages III, IV, and V, combined), likelihood of intraoperative bleeding, and residual/recurrent disease.

Other staging systems have been proposed; these include the Janakiram staging system (2017) that classifies stages based upon preoperative CT scan findings and suggests a surgical approach for each stage without the use of preoperative embolization.

Treatment

The mainstay of treatment of these tumours is complete surgical excision with minimization of surgical morbidity. This is facilitated in many centres by preoperative embolization which, where accessible, has become an almost universal component of the surgical plan.

The choice of surgical approach has evolved as endoscopic approaches have been developed and, for smaller tumours, endoscopic excision alone results in clearance of the tumour (see Chapter 78). Nevertheless, open craniofacial approaches or combined approaches are sometimes required for access, recurrence, and extension into the anterior cranial fossa. These include transpalatal, lateral rhinotomy, midfacial degloving, and craniotomy. Instruments such as the harmonic scalpel, coblation, and laser have been utilized to help decrease intraoperative blood loss. A multidisciplinary approach to assessment and surgery is recommended. Management should be carried out in a centre with access to paediatric rhinologist/head and neck surgery, paediatric neurosurgery, paediatric intensive care, paediatric maxillofacial/craniofacial surgery, and preoperative embolization.

Table 77.5 University of Pittsburgh Medical Center classification of JNA

Stage	
I	Nasal cavity, medial pterygopalatine fossa
II	Paranasal sinuses, lateral pterygopalatine fossa; no residual vascularity
III	Skull base erosion, orbit, infratemporal fossa; no residual vascularity
IV	Skull base erosion, orbit, infratemporal fossa; residual vascularity
V	Intracranial extension, residual vascularity; M, medial extension; L, lateral extension

Blood loss remains the commonest complication of surgery and this is then associated with the need for transfusion, packing, and staged procedures. It is also associated with incomplete excision and recurrence. Other complications include trismus, infratemporal nerve injury, and palatal hypesthesia. There have been concerns about potential effects upon facial growth, particularly with extensive bone excision, but this is not an issue that is widely reported. Excision of tumours with intracranial extension can lead to complications as a result of haemorrhage in the areas dissected or direct injury of intracranial structures.

Non-surgical treatment options include radiotherapy, hormonal therapy, and Gamma Knife radiosurgery. These are reserved for recurrence or extensive disease where the risk of surgery (mainly from haemorrhage) is deemed too high.

Factors associated with recurrence or residual disease include tumour stage and intraoperative bleeding. Reported recurrence rates vary, but are typically quoted at around 30%. Recurrence rates can be as high as 50% for extensive tumours that extend into the infratemporal fossa, clivus, and cavernous sinus.

Controversies

Efficacy of silver nitrate cautery

Silver nitrate cautery might be effective in the treatment of epistaxis in children although its superiority over conservative treatment options has not consistently been borne out categorically in randomized controlled trials.

Silver nitrate cautery can be performed on both sides at the same time.

Traditional advice has been to cauterize only one side of the septum at a time. But some studies have demonstrated a lower risk of requiring further cautery and no increase in medium-term complications. No septal perforations were noted.

Preoperative embolization for treatment of juvenile nasopharyngeal angiofibroma

It has been shown that preoperative embolization results in a reduction in intraoperative blood loss that aids complete resection. However, other studies have demonstrated no difference in intraoperative blood loss between embolized and non-embolized groups. There are also reports of an increased risk of recurrence in some cases where preoperative embolization has been carried out, suggesting that it could result in some tumours not being identified and therefore being left behind. Moreover, embolization can cause complications such as cranial nerve deficits, retinal artery occlusion and blindness, pulmonary oedema, and common iliac artery thrombosis.

Single allergen immunotherapy versus multiple allergen immunotherapy

Although both options are available, there is a suggestion that single allergen therapy has an advantage over multiple allergen therapy, as dosing for the combination cannot be titrated.

Tips and pearls

- Clinicians should have a high index of suspicion of cystic fibrosis in children presenting with nasal polyps and sinus disease, especially with a history of poor weight gain, respiratory disease, and pancreatic insufficiency.
- Orbital abscess drainage can be done externally or endoscopically (for older children). Anterior maxillary artery is 24 mm from lacrimal crest; post ethmoidal artery a further 12 mm and optic nerve another 6 mm. Perform an antral washout at the end of an orbital drainage procedure. Suture the orbital septum back when closing.
- Escalate the treatment of allergic rhinitis in a stepladder fashion (**Figure 77.8**).
- Immunotherapy for failed medical treatment should be considered in patients with single-allergen sensitivity.
- Clinicians should have awareness of drug dosage and age for licensed use of steroid nasal sprays in children.
- Before performing silver nitrate cautery in a child, usually a local anaesthetic/decongestant such as Co-phenylcaine (lidocaine hydrochloride 5% w/v and phenylephrine hydrochloride 0.5% w/v topical solution) is applied topically on a piece of cotton wool. Beware, though, that it its use is off-licence in children less than 12 years of age and should be avoided in very young children. This is because phenylephrine can, on rare occasions, cause severe acute hypertension. There can be significant absorption from the nasal mucosa, particularly if it is broken.
- Following application of the silver nitrate, saline should be dabbed on the area to stop the chemical reaction. This also helps to prevent black nasal discharge and staining that can be distressing to both parent and child.
- Naseptin contains arachis oil (peanut oil) and should not be given to patients known to be allergic to peanuts. As there is a possible relationship between allergy to peanuts and allergy to soya, patients with a soya allergy should also avoid Naseptin.

Guidelines

ARIA guidelines are available online (https://euforea.eu/aria).

The 'European Position Paper on Rhinosinusitis and Nasal Polyps 2020' was published in *Rhinology* in 2020 (https://www.rhinologyjournal.com/Documents/Supplements/supplement_29.pdf).

Guidelines and clinical knowledge summaries for management of epistaxis have been published but these mainly focus on primary care management and referral criteria (https://cks.nice.org.uk/topics/epistaxis-nosebleeds/).

Acknowledgement

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78

Blocked nose

Neil K. Chadha

Introduction

Nasal blockage is a frequent symptom of childhood. Common aetiologies differ from those in adults, particularly in neonates and younger children. The clinical history will focus on onset, duration, associated symptoms, and functional impact on sleep, feeding, and growth. The physical examination will typically include flexible nasendoscopy and the role for imaging is dependent on the suspected aetiology. This chapter outlines the major differences between children and adults, diagnostic approaches, and management strategies for the most common causes of nasal obstruction.

Basic science: nasal anatomy and physiology

Physiological differences between children and adults

It has been widely stated in the medical literature that neonates must breathe exclusively through their noses from birth to somewhere between 6 weeks and 6 months of age. As opposed to being 'obligate nasal breathers', the term 'preferential nasal breathers' may be considered more accurate, as most infants are able to breathe through their mouth if the nose is partially blocked. The nose is the primary route of breathing in infancy because of its physiological benefits (ability to humidify, warm, decontaminate, and regulate the air coming into the lungs) and an anatomical configuration that allows the child to breathe and suck simultaneously for breastfeeding ensuring swallowing without aspiration. The posterior portion of the relatively large tongue presses upward on the soft palate, forming a seal that intermittently blocks the oral airway, combined with interlocking of the soft palate and the high epiglottis. This mechanism becomes less important as a child grows older, and mouth breathing becomes easier. Although most neonates can initiate oral breathing after nasal occlusion, the length of time from occlusion to initiation of oral breathing may be prolonged and associated with significant struggling or even oxygen desaturation. This is paramount in the typical presentation of severe bilateral nasal obstruction in neonates, which may have dire consequences.

Anatomical differences between children and adults

Sinus development

Although all the paranasal sinuses are usually present at birth, pneumatization occurs at different ages. The ethmoids and maxillary sinuses have usually completed pneumatization by approximately 15 years. The frontal sinus starts to pneumatize after the age of 4 years, not reaching its full size until the late teens. The sphenoid sinus pneumatizes towards the sella turcica by around age 7, and completes pneumatization around the age of 15. An awareness of the developing sinus anatomy is relevant for pathophysiology and planning surgical interventions—for example, the anterior border of the ethmoid perpendicular plate moves caudally during growth and cannot be used as a reliable landmark for the location of the anterior skull base.

Nasal septum

The majority of the nasal septum in a young child consists of cartilage, and this proportion decreases until puberty as the ossification of the vomer, the nasal bone, and the perpendicular plate of the ethmoid bone proceeds. Historically, otolaryngologists have been concerned about the potential detrimental effects of paediatric nasal surgery on the nasoseptal growth process. Anthropometric studies suggest nasal growth is completed around 14 years of age in females and 16 years in males. More recently, there has been mounting evidence that the septum can be repaired without compromising facial development. With appropriate indications and technique, paediatric septoplasty can therefore be performed safely before adolescence.

Another difference between the nasal septum of children and adults relates to the vascularized pedicled nasoseptal flap used for endoscopic reconstruction of anterior skull base defects. This flap consists of nasal septum mucoperichondrium and mucoperiosteum, and is based on the nasoseptal artery. Although the nasoseptal flap remains an excellent workhorse for endoscopic skull base surgery reconstruction in children, a reduced flap length and incomplete pneumatization of the sphenoid have led to some authors suggesting it may be of lesser value in younger children.

Adenoids

The lymphoid tissue pad in the nasopharynx, termed adenoids, undergoes a physiological hypertrophy, then atrophy across

childhood, affecting the adenoid size. There also exists further variability in adenoid size with external inflammatory and infective stressors. Although the peak tissue volume is typically around 6–10 years (with atrophy by adolescence), it is the relative size of the adenoids compared to the surrounding nasopharyngeal dimensions that is more important in the context of obstruction, and this is maximal at a younger age (2–6 years).

Assessment

History

Key factors when assessing nasal obstruction in a child include unilateral versus bilateral (to allow determination of anatomical location); timing of onset (as congenital causes present early and acquired causes present later); seasonality or environmental triggers (suggesting allergy); infective symptoms (such as purulence); features concerning for neoplasia (such as associated bleeding or neurological symptoms); previous trauma or surgery (suggesting septal deformity or long-standing polyposis); and quality-of-life effects, including sleep and tolerance of exertion (to help determine indications for intervention).

Clinical examination

The clinical examination of a child with nasal obstruction should specifically include an external assessment and anterior rhinoscopy, in addition to the remainder of a full head and neck examination.

Flexible nasendoscopy

Flexible nasendoscopic assessment of the nasal cavity and nasopharynx is typically more practicable in children than rigid nasal endoscopy, due to the narrowness of the nasal cavity and low tolerance for discomfort. Decongestion may help, and there is limited support for effectiveness of topical local anaesthesia.

Investigations

- *Allergy testing* (see Chapters 42 and 77).
- *Computed tomography* (CT): CT is the most valuable radiological investigation for paediatric nasal obstruction whenever a congenital nasal skeletal anomaly is identified and surgical intervention contemplated (e.g. choanal atresia, piriform aperture stenosis). Benign tumours (e.g. nasal polyposis, juvenile nasal angiofibroma) are also typically best characterized by CT, benefitting from clear anatomical delineations, particularly of bone and cartilage.
- *Magnetic resonance imaging* (MRI): the role of MRI is largely limited to looking for intracranial communication of intranasal lesions (e.g. in differentiating meningoencephalocele from glioma) or for the assessment of associated anomalies of the brain, spine, or orbit.

Differential diagnoses

Unilateral choanal atresia

In the absence of any abnormality of the contralateral nasal passage, unilateral choanal atresia will present later in childhood. Unilateral atresia is more common than bilateral choanal atresia and more likely to be an isolated anomaly. Diagnosis is based on a combination of single-sided history of thick, stringy nasal discharge with nasal obstruction in an infant or young child; the absence of airflow using a mirror; and the inability to pass a suction catheter or flexible nasendoscope. An unwitnessed nasal foreign body may have a similar presentation but is usually easily differentiated by the shorter symptom duration, purulent nasal discharge, and a nasendoscopic examination. Although the diagnosis of unilateral choanal atresia is definitively confirmed by CT imaging, it is recommended that CT be performed only as a preoperative planning investigation once surgery is intended, to avoid the need for unnecessary repeated radiation exposure and sedation or anaesthesia of a young child.

Bilateral choanal atresia

In contrast to unilateral choanal atresia, preferential nasal breathing in neonates results in bilateral atresia presenting at or very soon after birth with severe respiratory compromise requiring airway support. A neonate may exhibit cyclical cyanosis and oxygen desaturation, which may ultimately be fatal without oral intubation. Historically the obstruction has been described as bony, membranous, or mixed, but in reality, even with a membranous plate there generally exists some degree of bony narrowing from medialization of the medial pterygoids, creating a funnel-shaped posterior nasal cavity. Consideration should be given to the other associated genetic abnormalities, the most common being CHARGE (coloboma, heart defects, atresia choanae, retardation of growth, genito-renal defects, ear anomalies/deafness) syndrome (in which bilateral choanal atresia affects ~50%) or, less commonly, the craniosynostoses, such as Crouzon and Apert syndromes (see Chapter 90). Diagnosis is suspected by an inability to pass a suction catheter in either nostril, and is confirmed by nasendoscopy and CT scanning. Echocardiography should be performed early in addition to genetic testing for CHARGE syndrome.

Piriform aperture stenosis

Congenital piriform aperture stenosis is a rare clinical condition involving overgrowth of the nasal process of the maxilla, and may present in isolation or in association with craniofacial abnormalities such as holoprosencephaly, maxillary central incisors, or cleft palate. The presentation mimics bilateral choanal atresia, with early respiratory compromise requiring respiratory support and an inability to pass a suction catheter. Differentiation of piriform aperture stenosis from bilateral choanal atresia is made through nasendoscopy and CT imaging.

Mid-nasal stenosis

Narrowing of the mid-portion of the nasal cavity (mid-nasal stenosis) may exist with or without piriform aperture stenosis or choanal atresia. Significant stenosis of this region is very rare in isolation, more frequently occurring with syndromic craniosynostoses (such as in Apert and Crouzon syndromes, see Chapter 90), where global narrowing of the nasal cavity can exist.

Septal deviation

Congenital or acquired septal deviation may cause nasal obstruction in children, just as in adults. A quality-of-life impact may be more

difficult to assess in children, although the most common sequelae are disturbed sleep quality and reduced exercise tolerance. An added challenge in children is differentiating the contribution of septal deformity to nasal obstruction from other common sources of obstruction that may coexist, namely rhinitis and adenoid hypertrophy.

Antrochoanal polyp

An antrochoanal polyp presents most commonly in adolescence, although it may be identified earlier in childhood. This lesion differs significantly from the polyposis of inflammatory nasal diseases, being a single, large, macroscopically uniform polypoid mass. The lesion arises from the mucosa of the maxillary sinus wall, extending via its ostium into the nasal cavity, and at the time of presentation, often extends posteriorly into the nasopharynx presenting as a nasopharyngeal mass, or possibly further into the oropharynx itself. The lesion may arise from a mucocele of the acinous mucous glands of the maxillary sinus mucosa. An antrochoanal polyp grows slowly and progressively, first causing unilateral obstruction and then bilateral obstruction once the nasopharynx is filled. Definitive diagnosis is usually through nasendoscopy and CT, which demonstrates an opacified maxillary sinus and continuity of the lesion into the posterior nose and nasopharynx. There may be bony expansion of the maxillary sinus and nasal cavity, but no bony destruction or invasion should be seen.

Adenoid hypertrophy

See Chapter 75.

Rhinitis

Allergic rhinitis is common in childhood and adolescence, and a frequent cause of nasal obstruction and rhinorrhoea (see Chapter 77). A non-allergic form of rhinitis, termed neonatal rhinitis, manifests at birth or in the early weeks of the neonatal period. The typical presentation is noisy breathing and stertor, potentially with significant respiratory distress or apnoeas. The nasal cavity mucosa shows severe oedema and bogginess of the inferior turbinates with mucoid rhinorrhoea, and a nasal catheter easily passes ruling out atresia. The aetiology is unknown and may be multifactorial, with viral, inflammatory, vascular, and traumatic causes all implicated.

Inferior turbinate hypertrophy

Typically, interior turbinate hypertrophy in children is associated with allergic rhinitis (see Chapter 77).

Meningoencephalocele

A nasal meningoencephalocele is comprised of a herniation of the meninges into the nasal cavity through a congenital defect in the anterior skull base, containing non-functional cerebral parenchyma. If only cerebrospinal fluid (CSF) is contained, this is typically characterized as a nasal meningocele. In neonates and infants, the typical presentation is unilateral nasal obstruction, and a flesh-coloured, non-inflammatory, occasionally pulsatile lesion may be visible unilaterally on nasendoscopy. Spontaneous CSF leakage or bleeding may occur, but is rare. A definitive diagnosis is obtained by a combination of CT and MRI, with biopsy generally not indicated. Presentation may be delayed into early infancy as the lesion typically causes only unilateral nasal obstruction and a child may continue to thrive.

Glioma

In contrast to a nasal meningoencephalocele, a glioma consists of ectopic glial tissue, sometimes with a fibrous cord communicating towards the base of the skull. This lesion therefore does not communicate with cerebral parenchyma or contain CSF. Although typically presenting in the neonate, presentation may be delayed into infancy due to obstruction usually being unilateral. A nasal glioma usually appears as a greyish, firm, non-pulsatile mass, and before can be confused with a nasal polyp or meningoencephalocele. Once CT scanning and MRI are completed, the lesion can usually be differentiated from other nasal masses, although biopsy may be indicated for confirmation.

Juvenile nasal angiofibroma

Juvenile nasal angiofibroma (JNA; see Chapter 77), or nasopharyngeal angiofibroma, is a highly vascularized benign tumour typically originating in the posterior lateral part of the nasal cavity, centring on the sphenopalatine foramen. It is rare, almost exclusively affecting adolescent and pre-adolescent boys. The pathogenesis is unknown. JNA usually presents with unilateral recurrent epistaxis and progressive nasal obstruction. Nasendoscopy reveals a pinkish tumour with superficial vascular markings. Workup comprises of CT and MRI to determine anatomical relationships and extension. Bony remodelling and destruction are best depicted by CT, whereas MRI is best for assessing involvement of the cavernous sinus, orbital apex, relation with dura, and may be superior in demonstrating permeation of the pterygoid root, sphenoid greater wing, or clivus. Radiologically demonstrated anterior bowing of the posterior wall of the maxillary antrum (the Holman–Miller sign), with widening of the pterygomaxillary fissure and pterygopalatine fossa, is considered diagnostic, with tissue biopsy contraindicated because of the risk of severe bleeding.

Other nasal tumours

Other nasal tumours that may present as nasal obstruction in childhood include other benign lesions (e.g. neurofibroma, schwannoma, ossifying fibroma, osteoma, Langerhans cell histiocytosis, haemangioma, and dacryocystocele) and malignant lesions (e.g. rhabdomyosarcoma and lymphoma).

Treatment

Neonatal rhinitis

Medical treatment forms the mainstay of treatment, through the use of bulb suction, physiological saline rinses, short-term vasoconstrictors, hypotonic saline, or topical steroids. Topical nasal decongestants (e.g. xylometazoline or oxymetazoline) or diluted epinephrine (adrenaline) saline solution may be used for a few days or intermittently and are usually very effective, although prolonged use risks sensitization and the need for careful tapering. Topical steroids drops (e.g. betamethasone) may also be effective, but care must be taken to control dosing amid concerns of systemic absorption. Recognition and treatment of any suspected aggravating factor, such as gastro-oesophageal reflux, may be helpful. Symptoms may persist up to the age of 1 year, but long-term prognosis is excellent.

Meningoencephalocele

Consideration should be given to early vaccination against pneumococcus to reduce the risk of meningitis. The deformity is managed by surgical resection, which can often be exclusively endoscopic. The procedure consists of resection of the herniated meninges and their contents after sectioning at the neck, followed by repair of the skull base defect. Materials for repair include combinations of tissue autografts, such as cartilage or mucosa, vascularized local nasal septal flap based on the posterior nasal septal artery, or xenografts such as porcine collagen. A very large hernia with a substantial skull base defect may require a combined endoscopic and open neurosurgical approach, particularly to prevent or treat significant CSF leakage risk.

Glioma

The preferred approach to glioma is complete surgical excision, as an incomplete excision may result in recurrence. Endoscopic removal is favoured, but the approach is dependent on the anatomical location and extent. Where intracranial extension exists, a multidisciplinary team approach may be required with neurosurgical and/or transcraniofacial approaches required to ensure complete and safe excision.

Unilateral choanal atresia

Regular bulb suction and nasal irrigation can reduce the negative impacts of the unilateral nasal discharge. A decision on timing of elective surgical treatment of unilateral choanal atresia is dependent on the degree of impact on quality of life. Delaying surgical management until a child's nostril is wide enough to accommodate an effective endoscopic approach will be beneficial in reducing the likelihood of a need for an early revision, although revision surgery is required in the majority of cases, and over the full course of childhood multiple revision procedures may be required. A balance needs to be found between the impact of obstruction on an individual versus the technical ease of surgery with increasing age, and surgery is typically planned before school attendance at the age of 3–5 years. The anatomical abnormality usually consists of a mixture of a bony atretic plate, prominence of the medial pterygoids, and a widened thickened posterior nasal septum. With improved endoscopic instrumentation and image quality, a transpalatal approach to the atresia is now very rarely justifiable. A number of techniques have been described for the endoscopic opening of the choana, but all are essentially variations in positioning of the mucosal flaps to allow exposure of the atretic bone for removal. The goal is maximal preservation of the mucosal flaps for lining of the edges of the newly formed choanal bony opening once transnasal drilling is completed. Resection laterally of bone from the medial pterygoids, and superiorly from the floor of the sphenoid sinus, is less impactful in reducing restenosis than careful and complete removal of medial bone from the posterior nasal septum. Image-guided surgery may allow a better appreciation of the bone resection and increase safety and precision. A unilateral nasal stent is unlikely to provide adequate benefit to warrant its additional morbidity, but postoperative nasal saline irrigation and judicious use of topical nasal steroids are highly beneficial in reducing granulation tissue formation and early scarring.

Bilateral choanal atresia

Neonatal bilateral choanal atresia is considered an emergency and severe respiratory compromise is typical until a definitive airway is established. Initial management with an oral airway or McGovern nipple may be effective, although intubation is often necessary. Surgery is required to restore canalization. The narrow nostril makes transnasal visualization for the surgical approach challenging, but is ideally through a combination of transnasal and transoral endoscopy using 0° and 120° Hopkins rod telescopes, respectively. The transnasal approach has superseded the transpalatal approach outside of the very rare situation of a thick bony atresia. The atretic plate is identified transnasally, and is carefully punctured at its thinnest point (typically inferomedially) using suction or a dilator while under direct visualization via the transoral retrograde endoscopic view with the 120° endoscope. The size of the opening is then increased using serial dilatation with curved dilators (e.g. urethral dilators) or a guarded drill burr—especially to remove the midline posterior vomer. The inability to preserve mucosal flaps for coverage of newly exposed bone and concern about early restenosis has led some to advocate nasal stenting for 2–6 weeks, typically using modified folded and cut polyvinyl chloride endotracheal tubes. With or without stents, postoperative care includes regular suctioning, saline irrigation, and topical nasal steroids.

Piriform aperture stenosis

A trial of non-surgical management should be initiated, including topical steroids, decongestants, as well as oral airway devices or McGovern nipples. Surgery should be reserved for patients with persistent respiratory distress, or significant impacts on sleep, growth, or feeding. In most cases, failure of conservative management is identified within the first few weeks of treatment when weaning from airway support is failing. Children discharged without surgical intervention must be followed closely as they may have difficulties during respiratory infections, including severe obstructive sleep apnoea. For surgical treatment, the stenosed pyriform aperture is approached via a sublabial incision, elevating a mucoperiosteal flap to expose the bony stenosis bilaterally. A diamond burr is used to drill away excess bone from the inferior inlet along the floor of the nose, to the lateral process of the maxilla, taking care to avoid the nasolacrimal duct posterolaterally and the dental buds inferiorly. Nasal stents are often placed for 2–4 weeks, and postoperative care is similar to bilateral choanal atresia. More recently, primary treatment with endoscopic piriform aperture balloon dilatation has been reported with success.

Mid-nasal stenosis

Nasal obstruction from mid-nasal stenosis is often a result of a combination of the narrow nasal cavity width combined with oedema of the nasal lining. Initial treatments should include topical nasal steroids, decongestants, and antibiotics where indicated. The next step in conservative treatments includes the use of nasopharyngeal airway devices and continuous positive airway pressure ventilation. Where surgical treatment is indicated, bougie dilatation and balloon dilatation form the mainstay of treatment, and post-dilatation stenting may be required. Typically, multiple dilatations will be required. Any coexisting choanal atresia or piriform aperture stenosis increases the likelihood of failure of conservative therapy and may also require specific surgical treatment.

Septal deviation

In principle, septoplasty is possible at any age, but in most settings symptoms are not severe and surgery can be postponed until late

adolescence to minimize the risk of negatively influencing nasal facial growth. Early childhood mild septal deformity may also improve with nasal growth. Uncommonly, early surgical intervention is warranted due to severe quality-of-life impact or difficulty breathing at night resulting in obstructive sleep apnoea. In the adolescent, surgical approaches to nasal septal reconstruction are the same as in adults (see Chapter 40). When performed before adolescence, clinical indications must be well founded and surgery should be performed conservatively with cartilage-preserving techniques favoured, for example, limiting the procedure to resection of an obstructing spur or realignment of the septum with cartilage scoring and minimal resection. The aim is to reduce the risk of impairment of subsequent nasal or facial growth.

Antrochoanal polyp

The treatment of an antrochoanal polyp is exclusively surgical, because in contrast to inflammatory nasal polyposis there is generally no response to topical or systemic steroid therapy. Surgical treatment is performed endoscopically transnasally, and should aim for complete resection of the polyp including its base in the maxillary sinus. A wide maxillary antrostomy (often partly already present through the lesion's expansion), angled-view endoscopes up to 70°, and angled instruments, such as a 120° microdebrider blade, are important to facilitate access to the polyp base. Intraoperative image guidance may be helpful. Recurrence rates are high, particularly with incomplete resection of the polyp base. This is most likely to occur where the origin is anteriorly in the maxillary sinus which may be very challenging to adequately reach through an endoscopic approach. Access to this location may be facilitated by performing an inferior meatal antrostomy into the maxillary sinus; a trans-septal approach from the contralateral nostril; or an external anterior approach creating a window through the anterior canine fossa (the modified Caldwell–Luc approach). When performing the canine fossa approach, the bony window size can be minimized by using it for endoscopic lesion removal, combined with a transnasal route, thereby reducing the risk of injury to alveolar nerve branches and the infraorbital nerve.

Juvenile nasal angiofibroma

On the basis of the pattern of extension on CT and MRI, a JNA can be staged using one of many available staging systems (see Chapter 77), the more recent of which focus on the appropriateness of the lesion for endoscopic resection. The goal of surgical treatment is complete removal of the JNA, particularly as revision treatment can be very challenging due to scarring and loss of surgical landmarks. Endoscopic resection has become the mainstay of treatment over open surgical approaches such as transpalatal, transfacial, lateral rhinotomy, and mid-facial degloving, due to better visualization of tumour margins and anatomical structures, reduced haemorrhage, lower recurrence rates, and less postsurgical morbidity. Transarterial preoperative embolization, typically 24–48 hours before surgery, reduces intraoperative bleeding when combined with early detection and surgical control of the internal maxillary artery and other feeding vessels, including those from the internal carotid artery. A key principle of the endoscopic approach is to expose the lesion as extensively as possible without traumatizing its surface to minimize bleeding, with en bloc resection not usually feasible. Endoscopic exposure initially includes ethmoidectomy, sphenoidotomy, a large middle antrostomy, and a medial maxillectomy. Removal of the posterior maxillary wall allows for infratemporal fossa dissection; posterior septectomy facilitates exposure of nasopharynx; and the tumour is followed into the pterygopalatine fossa, the basisphenoid, and the upper parapharyngeal space, as required. A sublabial transmaxillary approach may also be helpful. Imaging-based surveillance after surgery is required as recurrences are often submucosal, and therefore not amenable to easy endoscopic identification. Non-surgical treatment modalities such as intensity-modulated radiotherapy may be considered, particularly for treatment of recurrence in critical areas.

Controversies/areas of interest for the future

Stenting

Widespread controversy exists over the benefits versus risks of postsurgical nasal stenting, in both unilateral and bilateral choanal atresia. Although many authors have published case series with and without stents for varying periods of time, recent data have suggested that there is no significant difference between these approaches and the need for revision surgery. As stents have an inherent morbidity and require significant maintenance by carers, together with the potential need for removal in the operating room, there is a trend towards a decrease in their usage. Novel designs of small stents have been developed for adult endoscopic sinus surgery, which elute steroids locally over a period of days to weeks. Early trials have investigated their potential to improve outcomes in choanal atresia repair, by reducing inflammation and granulation postoperatively and thereby hopefully reducing restenosis rates. No controlled study exists to date, but this will be an area of interest for the future.

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79

Paediatric dysphonia

Eaodaoin O'Cathain and Scott Rickert

Introduction

Voice disorders in the paediatric population are as varied as they are in the adult population. There is a wide spectrum of 'normal' voice in the paediatric population, which varies by age, sex, and pubertal development. It is important to note that different adults (parents, teachers, speech language pathologists, paediatricians) interacting with the child may perceive the child's voice differently and may not be in agreement as to whether the child is dysphonic and the degree of dysphonia. This range of opinion provides a complicating factor in discerning the presence of a voice disorder and how to manage it appropriately. It is estimated that dysphonia will affect between 4% and 23% of children at some stage in their development.[1] In contrast to their adult counterparts, children with dysphonia are likely to have a self-limiting benign aetiology. The widespread, simplistic view of the paediatric hoarse voice as something to outgrow and that requires no treatment nor follow-up can be a disservice to the patients and their families. The most common causes of dysphonia in children are due to vocal misuse and infectious causes, including human papillomavirus (HPV) causing papillomatosis. Most of these aetiologies will improve with input from speech and language therapists, and by taking a watch-and-wait approach. It is often the case that as a child grows, and the vocal cords change with puberty, these issues resolve. It has been reported that children with dysphonia can sometimes be unaware of their condition, which can cause problems when trying to motivate these children to complete treatment. It has also been shown that children with dysphonia can sometimes be judged more negatively than children with no voice disorders.[2]

It is therefore crucial to take a history from the patient's caregivers, including their teachers, as the reporting and assessment of severity of dysphonia has been shown to vary widely from patient to caregiver. It is also prudent to take a comprehensive history from the patient themselves, where appropriate. Sometimes there can be underlying anxieties and stressors that may precipitate or exacerbate dysphonia, for which the patient may need additional treatment and support. It is also important to understand that dysphonia in a child may cause social isolation and reduced involvement in school and sporting activities. It helps to be aware of the potential impact that dysphonia can have on a patient's academic, social, and emotional life.

Significant advances in paediatric voice over the past decade in evaluation, diagnosis, and management of paediatric voice disorders have improved both short-term and long-term outcomes for the dysphonic child.

Anatomy and physiology

The larynx grows and develops significantly from birth to adulthood. As a neonate, the larynx sits at the level of the C3–C4 vertebrae. By around the age of 15 years, the larynx sits at the level of C6–C7. In a neonate, simultaneous feeding and respiration are possible. In older children, the vocal fold consists of three layers. From deep to superficial, these consist of the vocalis muscle, the lamina propria (which consists of three layers: deep, intermediate, and superficial), and the epithelium. Newborn vocal cords differ from those of an older child or adult, in that the lamina propria consists of only one layer of loose collagen fibres between the vocalis muscle and the epithelium.[3] As children grow, their vocal folds also lengthen. It has been proven that there is no difference between male and female vocal cords in early childhood. However, this changes during puberty, when male vocal cords lengthen and the larynx grows more dramatically than in female adolescents.[4]

The superior and recurrent laryngeal branches of the vagus nerve innervate the laryngopharynx. The vocal cords are innervated by the recurrent laryngeal nerves. These branch from the vagus nerves in the upper thorax and re-enter the neck from the thoracic inlet. They travel in the tracheo-oesophageal groove. The right recurrent laryngeal nerve travels under the right subclavian artery and the left recurrent laryngeal nerve loops under the aortic arch. It is important to remember that mediastinal pathology or cardiac surgery in a baby can be a cause of iatrogenic vocal cord palsy. The superior laryngeal nerve travels along the pharynx and separates into the internal and external branches. These supply sensory innervation to the supraglottis and motor innervation to the cricothyroid muscle. The rest of the larynx is supplied by the recurrent laryngeal nerve for motor and sensory innervation.

Phonation occurs when exhaled air from the lungs passes between the vocal cords and causes oscillation of the mucosa of the vocal folds relative to the thyroarytenoid muscles and vocal ligaments. The intrinsic laryngeal muscles adjust the tension and position of

the vocal folds, which creates changes in pitch and frequency of the voice. Contraction of the thyroarytenoid and cricothyroid muscles shorten and lengthen the vocal folds respectively, creating lower and higher pitches of voice. Articulation is created through fine control of the tongue and lips. Resonance is created in the airway above the level of the vocal cords. The size of the pharynx, oral and nasal cavities, and larynx change with age and create changes in resonance, particularly in boys. Loudness of voice is dependent on the level of subglottic pressure involved during speech.

Assessment

A full history should include birth history, any history of birth trauma, any history of prematurity or intubation, history of tracheostomy placement, previous cardiac or mediastinal surgery, history of reflux, history of asthma or other chronic airway disease should be elicited. It is important to be aware that a child with chronic dysphonia may be deemed by their parents to have a normal voice. Therefore, referrals to ear, nose, and throat (ENT) may come from our colleagues in speech and language therapy initially.

On examination, general inspection should include assessment of ease of respiration and presence of stridor. A head and neck examination should rule out any evidence of craniofacial abnormalities or any syndromic features. A nasal and throat examination should also be performed to assess for nasal obstruction, adenotonsillar hypertrophy, or palatal insufficiency. The quality of the child's voice should be noted, to include any sign of a weak or breathy voice, coarseness, whisper, or strained voice. Resonance is usually described as either normal, or hyper- or hyponasal. There might be evidence of a variation in pitch, restricted range, decreased voice projection, voice breaks, or abnormal resonance.

Popular tools for parental or self-assessment of voice include the Consensus Auditory–Perceptual Evaluation of Voice (CAPE-V) questionnaire, and the Paediatric Voice Handicap Index (pVHI) (**Figure 79.1**).[5] The Paediatric Voice Related Quality of Life (PVRQOL) and Paediatric Voice Outcome Survey (PVOS) are other useful questionnaires.[6] It should be noted that some of these tools will require parental assessment of the child's quality of life and so may not be entirely accurate. These tools can be used at initial assessment and also following intervention, to determine whether the intervention has helped. All of these children should be seen by a team of professionals who are experienced in the management of paediatric dysphonia. These teams generally consist of a paediatric otolaryngologist in conjunction with a speech and language therapist, who performs aerodynamic and acoustic evaluation. They also assess the child's voice subjectively and objectively. Objective measures include assessment of pitch, range, loudness, shimmer, and jitter. These children will often have audiometric testing to ensure there is no element of hearing loss contributing to possible vocal misuse. Occasionally these children will also be assessed by a paediatric respiratory physician, an allergist, or a gastroenterologist.

Investigations

The initial investigation for any child with dysphonia is a flexible nasoendoscopy. This can be done in the clinic or by the bedside in the acute setting. Topical lidocaine with or without epinephrine (adrenaline) can be applied to the anterior nasal cavities via cotton buds or via spray application. In younger children, a caregiver will need to have the child seated on their lap, with both people facing the endoscopist. A tower plus camera attachment to the scope should be used for recording the scope and to aid in facilitating a safe distance between the ENT surgeon and the patient and caregiver (**Figure 79.2**). The smallest diameter of a flexible nasendoscope suggested for paediatric use is 2.1 mm. Typically the small-calibre scopes are more comfortable to the patient on examination. It is also important to note that the larger the diameter of the scope, the better the associated image quality, so it is an important balance to limit discomfort while not sacrificing image quality and limiting diagnostic abilities.

Stroboscopy should also be performed. This aids in assessing gross movement of the larynx, movement of the arytenoids, the microstructure of the vocal fold, and any evidence of lesion or scarring. In particular, it aids in the assessment of vocal cord symmetry, the mucosal wave, and glottis closure. A rigid strobe can be purchased which is of a smaller diameter than an adult strobe. To use this, a child must be very compliant, therefore it is more useful in older children or adolescents, although mature child as young as 3–4 years of age can tolerate a rigid examination well. The tongue is protruded and held with a gauze swab. Antifog solution is placed on the tip of the scope. When the patient opens their mouth, the rigid scope is passed through the mouth to sit over the larynx. It is often possible to use the intraoral rigid scope in a child without local anaesthesia, as their larynx is more superiorly situated than in an adult. The advantages of using the rigid strobe are that the image quality is excellent and that it has a zoom function. Disadvantages include possible discomfort while holding the mouth open, and potential damage to the teeth if a patient bites or is non-compliant. It is also possible to use a flexible strobe scope, which is passed in the usual transnasal approach, thereby being more readily tolerated for younger children. This method of stroboscopy is more easily tolerated, with no age restrictions. A microphone is also used. Stroboscopy uses flashes of light which synchronizes at a slightly slower speed to the vocal fold vibration, which is picked up by the microphone. This allows vocal fold vibration to be appreciated in what appears to be slow motion. The mucosal wave, symmetry, and glottic closure can all be appreciated in greater detail using stroboscopy.

Different sounds on vocalization should be performed to assess the voice. Sniffing causes abduction of the vocal cords, which allows assessment of the movement of the cords and arytenoids. In an older child, the recitation of a paragraph will allow for more in-depth analysis of the quality of the voice.

Laryngeal ultrasound is a newer technique for evaluating the anatomy and movement of the larynx (**Figure 79.3**). It can be a helpful diagnostic tool where a child is fearful of nasendoscopy or an examination is particularly difficult to obtain anatomically. It can be particularly useful when evaluating for a laryngeal cyst or distinguishing vocal cord palsy as patients are much more compliant with the ultrasound examination.

In some situations it may be necessary to further assess the airway under anaesthesia. This provides a microscopic viewpoint and the ability to palpate and examine any areas of concern. In these cases, one can perform a direct microlaryngoscopy as part of an endoscopic airway evaluation. One can also potentially include a flexible bronchoscopy with the same anaesthetic to assess the airway

Subject Number: _____ Date: _____

I would rate my/my child's talkativeness as the following (circle response)

1 2 3 4 5 6 7
Quiet Listener — Average Talker — Extremely Talkative

To be filled out by Staff:

F= _____
P= _____
E= _____
Total= _____

Talkativeness: _____

Instructions: These are statements that many people have used to describe their voices and the effects of their voices on their lives. Circle the response that Indicates how frequently you have the same experience.

0=Never 1=Almost Never 2=Sometimes 3=Almost always 4=Always

Part I - F

1)	My child's voice makes It difficult for people to hear him/her	0 1 2 3 4
2)	People have difficulty understanding my child in a noisy room	0 1 2 3 4
3)	At home, we have difficulty hearing my child when he/she calls through the house.	0 1 2 3 4
4)	My child tends to avoid communicating because of his/her voice.	0 1 2 3 4
5)	My child speaks with friends, neighbors, or relatives less often because of his/her voice.	0 1 2 3 4
6)	People ask my child to repeat him/herself when speaking face-to-face.	0 1 2 3 4
7)	My child's voice difficulties restrict personal, educational and social activities.	0 1 2 3 4

Part II - P

1)	My child runs out of air when talking	0 1 2 3 4
2)	The sound of my child's voice changes throughout the day	0 1 2 3 4
3)	People ask, "What's wrong with your child's voice?"	0 1 2 3 4
4)	My child's voice sounds dry, raspy, and/or hoarse	0 1 2 3 4
5)	The quality of my child's voice is unpredictable	0 1 2 3 4
6)	My child uses a great deal of effort to speak (e.g., straining)	0 1 2 3 4
7)	My child's voice is worse in the evening	0 1 2 3 4

0=Never 1=Almost Never 2=Sometimes 3=Almost always 4=Always

Figure 79.1 Paediatric Voice Handicap Index.

Reproduced from Zur KB, Cotton S, Kelchner L, Baker S, Weinrich B, Lee L. Pediatric Voice Handicap Index (pVHI): a new tool for evaluating pediatric dysphonia. *Int J Pediatr Otorhinolaryngol*. 2007;71(1):77–82, doi: 10.1016/j.ijporl.2006.09.004, with permission from Elsevier.

below the level of the glottis, to rule out any other dynamic tracheal or bronchial anomaly.

In the case of unilateral or bilateral vocal cord paralysis, it is important to perform imaging to rule out any underlying aetiology. Magnetic resonance imaging (MRI) of the brain can be helpful to rule out a Chiari malformation, which could cause a bilateral vagal nerve palsy. Imaging of the neck and thorax is also important to rule out any pathology along the course of the recurrent laryngeal nerve(s). MRI is preferable to computed tomography (CT) as it does not involve a radiation dose. It is worth noting that children older than a few months of age will likely need either sedation or a general anaesthetic to tolerate MRI or CT scanning. Dynamic voice CT is a newer investigation which is used in some children with previous history of multiple complex airway surgeries (Figure 79.4).

In children with a history of oesophageal reflux, or evidence of chronic laryngitis and erythema or oedema of the arytenoids, a contrast swallow study or oesophagoscopy can be performed. They may also require assessment by a gastroenterologist and a clinical assessment of swallow performed by a speech and language therapist.

Laryngeal electromyography (EMG) is helpful to assess vocal cord paresis versus palsy in adults. In adults and older children, one can perform this in the clinic setting with mild discomfort to the patient. In younger children, an awake laryngeal EMG is typically not tolerated and it is usually performed under a general anaesthetic and

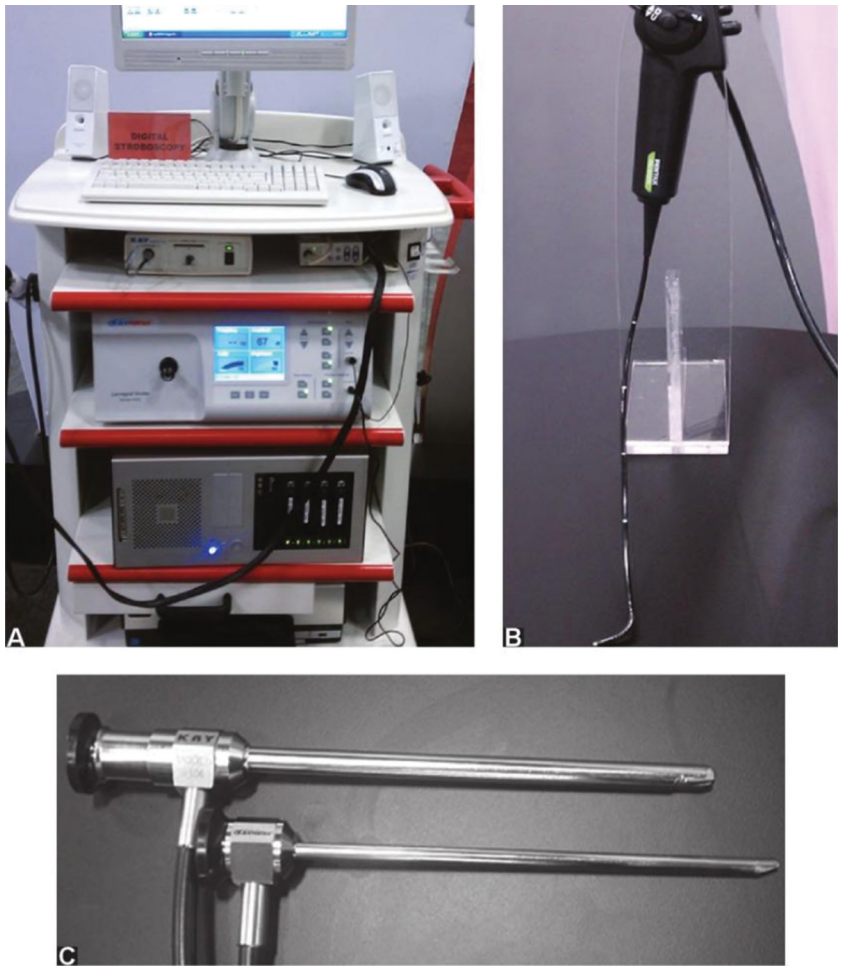

Figure 79.2 (a) Stack system used. (b) Flexiscope with chip-in-tip for stroboscopy. (c) Rigid stroboscopy scopes.
Adapted from Lalwani AK, Pfister MHF. *Recent Advances in Otolaryngology: Head and Neck Surgery*. Jaypee Brothers Medical Publishers; 2012.

with the aid of an electrophysiology physician. The data obtained under a general anaesthesia may not be as diagnostically distinct as an awake laryngeal EMG but can be used to assess and compare laterality in the case of vocal fold paresis/immobility. One can also perform a diagnostic laryngoscopic examination under anaesthesia at the same time to distinguish vocal fold immobility from neurogenic paresis.

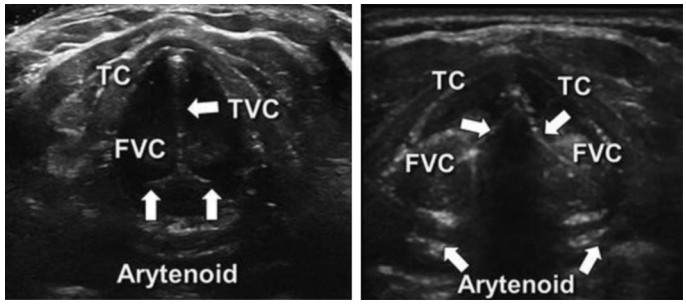

Figure 79.3 Laryngeal ultrasound.
Reproduced from Carneiro-Pla D. Transcutaneous laryngeal ultrasonography: technical performance and interpretation. In: Milas M, Mandel S, Langer J, eds. *Advanced Thyroid and Parathyroid Ultrasound*. Springer; 2017:351–354, https://doi.org/10.1007/978-3-319-44100-9_32, with permission from Springer Nature.

Differential diagnosis and treatment

Common causes of paediatric dysphonia

Laryngopharyngeal reflux

Laryngitis can be caused by laryngopharyngeal reflux (LPR). Unfortunately, there is no definite investigation to diagnose LPR. However, it is a cause of inflammation of the larynx. Symptoms can include globus pharyngeus and chronic cough or throat clearing. Signs on flexiscope can include oedema, erythema, and pachydermia. Treatment includes proton pump inhibitors, lifestyle modifications including weight loss, and referral to gastroenterology in recalcitrant cases for further workup including oesophagogastroduodenoscopy or impedance probe.

Vocal fold movement impairment

This is the second most common congenital laryngeal anomaly after laryngomalacia. It usually presents between birth and 2 months of age. It can be symptomatic of a multisystem anomaly involving the central nervous system and cardiopulmonary anomalies. Sometimes it is associated with laryngeal clefts or subglottic stenosis. Unilateral vocal fold movement impairment is more common than bilateral, and left more common than right. It can present as a weak or breathy

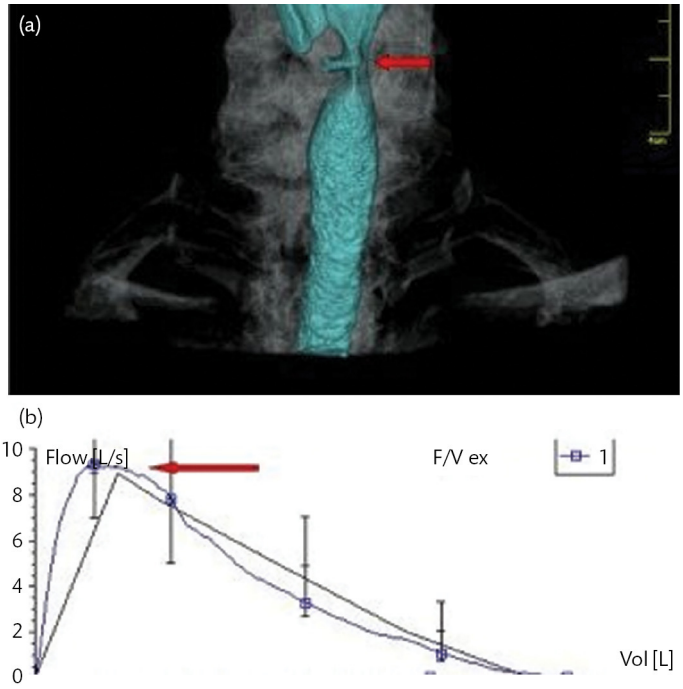

Figure 79.4 (a) Maximal intensity images reconstructed from the 320-slice CT image dataset displaying laryngeal airway in blue colour showed persistent marked laryngeal narrowing due to adduction of the vocal cords throughout the breathing cycle. (b) Flow–volume loop from the spirometry demonstrated flattening of the inspiratory and expiratory limbs, consistent with fixed upper airway obstruction.

Reproduced from Ruane LE, Lau KK, Low K, Crossett M, Vallance N, Bardin PG. Dynamic 320-slice CT larynx for detection and management of idiopathic bilateral vocal cord paralysis. *Respirol Case Rep*. 2014;2(1):24–26. doi: 10.1002/rcr2.37 with permission from John Wiley and Sons.

cry, or with respiratory distress and stridor if bilateral (Figure 79.5) (see Chapter 74). Sometimes unilateral vocal fold movement impairment will resolve with time. Observation is appropriate in cases where there is no known aetiology and no evidence of respiratory distress or difficulty feeding. In cases of vocal cord palsy caused by cardiac surgery, it has been shown that around 5% of babies or children undergoing surgeries of the aortic arch can be left with a left-sided vocal cord palsy.[7] Therefore, it is important to screen these children prior to discharge and to have a high index of suspicion in terms of diagnosis. Laryngeal EMG and endoscopic assessment have been found to be useful for assessing qualitative function of a vocal fold palsy and distinguish vocal fold immobility from neurogenic palsy.

In cases of non-resolving unilateral vocal cord palsy, medialization thyroplasty with injection or permanent implants such as Gore-Tex or silicone may be helpful. Occasionally, laryngeal reinnervation can be performed by anastomosing the ansa cervicalis to the recurrent laryngeal nerve on the affected side. Temporary laryngeal injection might be used prior to more permanent techniques, to test the possible outcomes if permanent surgery were to be performed. Following a first vocal cord injection, the patient should be admitted overnight to observe for any signs of respiratory compromise that may require treatment. If successful, vocal cord injection can be repeated, or more permanent surgery can be performed. Although there are no long-term studies to prove the benefits of early vocal cord injection

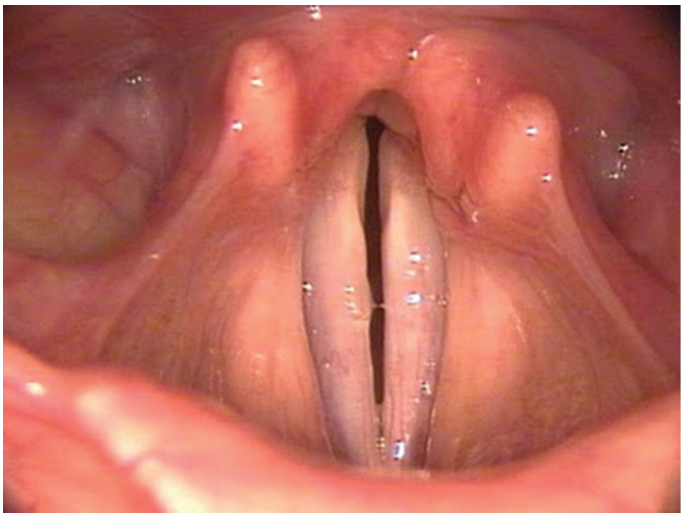

Figure 79.5 Bilateral vocal fold paresis seen with rigid stroboscopy.

following palsy, they have been shown in adults. Injectable materials include Gelfoam (6 weeks), Restalyne (3–6 months), and Cymetra (3–9 months). Longer-term options include Radiesse/Prolaryn (1–2 years), or fat (1–2 years or permanent). Choice of material used is influenced by the length of time desired for benefits to last, the risk/benefit profile of the material, and availability including cost.

Laryngeal webs

Laryngeal webs may be congenital or acquired and typically affect the anterior margin of the vocal fold. In tethering a portion of the vocal folds together, the vibration of the true margin of the vocal fold changes and prevents the normal propagation of the mucosal wave. Repeated microlaryngoscopy with intervention is the most common cause of acquired laryngeal webs, which tend to be thinner than congenital webs. Congenital airway webs, 75% which are glottic in location, are formed due to a failure in recanalization of the glottis during development (Figure 79.6). A narrowed cricoid is associated with congenital glottic webs.

Velocardiofacial syndrome, a genetic deletion of 22q (see Chapter 90), is also associated with congenital webs. Therefore, all congenital webs should have a genetic workup in their treatment plan.[8] Laryngeal webs are notoriously difficult to treat, as the goal is to restore a normal vibrating vocal fold with proper propagation of the mucosal wave. Typically, cold sharp dissection is used in an endoscopic fashion, with or without placement of a keel to try and prevent reformation of the glottis web. If the cricoid is severely narrowed, an anterior cricoid split (endoscopic or open) or laryngotracheoplasty may be indicated in addition.

Benign vocal cord lesions

Vocal cord nodules

These are the most common cause of dysphonia in children. They are caused by vocal misuse and abuse. They are bilateral and are found at the junction between the first third of the vocal fold and the subsequent two-thirds. The vocal fold closure is described as an hourglass closure configuration. On stroboscopy there can be a normal mucosal wave, or minimal changes of the mucosal wave. In general, vocal cord nodules respond very well to voice therapy.

Figure 79.6 Near complete congenital glottic web.

Vocal cord cysts

These are more often unilateral than bilateral, with a contralateral reactive lesion (Figure 79.7). The location of a vocal cord cyst will determine the impact on the voice. These are distinguished from vocal fold nodules by videostroboscopy as they impair the mucosal wave, depending on its size and location. These respond less predictably to voice therapy, and often requires surgery, which can be performed using the microflap technique.

Vocal cord polyp

Polyps are an uncommon finding in children. They generally present unilaterally and there can be an associated contact lesion on the ipsilateral cord. They are treated conservatively initially, with voice therapy. Surgery with minimal disruption to the underlying cord is kept in reserve for recalcitrant polyps.

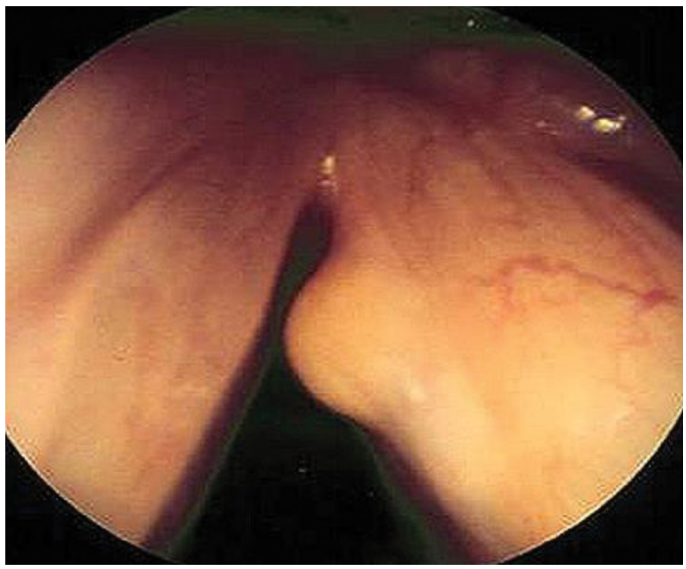

Figure 79.7 Vocal fold cyst.

Figure 79.8 Bilateral post-intubation granulomas seen within conventional flexible laryngoscopy.

Vocal cord granuloma

These can be associated with chronic cough or LPR. Occasionally they can occur as a result of previous intubation (Figure 79.8). These granulomas arise in the posterior third of the cord and present a few weeks post extubation. They can be treated conservatively or excised if they persist or cause airway stridor impacting breathing.

Recurrent respiratory papillomatosis

Recurrent respiratory papillomatosis (RRP) is the second most common cause of dysphonia in children (Figures 79.9 and 79.10). It is caused by HPV subtypes 6 and 11. Surgical debridement is the mainstay of treatment. Surgical debridement is most commonly performed using a microdebrider/skimmer blade as part of a microlaryngoscopy set-up. During the initial surgery, a biopsy should always be taken to confirm the diagnosis. The primary goals of surgery are to reduce tumour burden, decrease the chance of distal spread of disease, and to create a safe airway. It is also performed to improve voice quality, and to hopefully increase the time between surgeries. However, surgery is not curative. Most cases of RRP regress and 'burn-out' as the child grows and their immune system matures. Some adjuncts to surgery are KTP laser (532n), bevacizumab, and cidofovir.[9] Bevacizumab is a recombinant antibody designed to bind vascular endothelial growth factor A (VEGF-A) and inhibit VEGF receptor interaction. It is used as an intralesional adjunct in the management of RRP, although its use is off-label. Although no randomized control trials have occurred to date, results have been promising, with few side effects.[9] Cidofovir is a cytosine nucleotide analogue that inhibits the replication of DNA viruses by blocking viral DNA polymerase function. The mechanism of action in HPV and RRP is unclear; however, it is thought that it induces apoptosis and enhances the immune system. It is injected into the base of the papillomata once they have been debrided. Although it is well tolerated, and has been found to be of benefit at certain doses, there is a

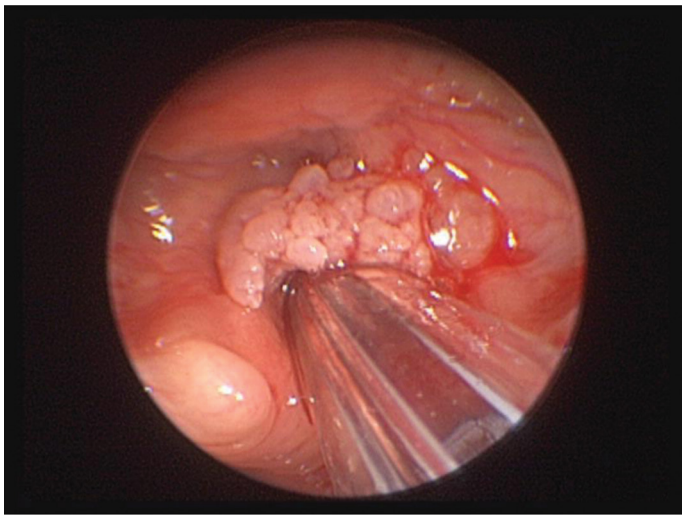

Figure 79.9 Obstructive papilloma of the larynx.

theoretical risk of malignant transformation.[9] Therefore the parents of a child undergoing potential cidofovir injection should be well counselled and provided with ample evidence prior to consenting to the treatment. Intravenous therapies such as bevacizumab and interferon can be considered in cases requiring more than four procedures per year; however, these are not instituted on a regular basis, as shown in a recent International Pediatric Otolaryngology Group consensus paper.

Functional voice disorders

Functional disorders are caused by dysregulated laryngeal function Up to 40% of adult patients have functional voice disorders[10] while only 7% of children have a functional aetiology.[11] Functional disorders affecting the voice include muscle tension dysphonia, psychogenic dysphonia, and puberphonia. This causes a strained voice with pitch changes and occasional periods of aphonia. Treatment in these cases is often behaviour modification in tandem with speech therapy. Occasionally Botox can be considered.

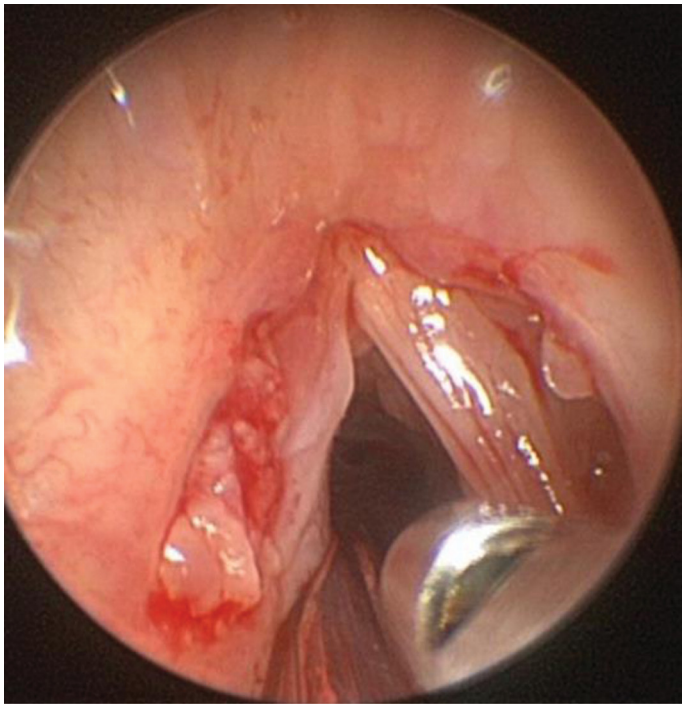

Figure 79.10 Non-obstructive papilloma of left true vocal fold and bilateral false vocal folds.

Paradoxical vocal fold dysfunction

This is also called paradoxical vocal cord movement, laryngospasm, and exercise-induced laryngeal obstruction. A typical patient is female, a high achiever in school, and an athlete. Frequently these patients have a history of asthma or exercise-induced asthma. They may also have a history of a psychological stressor or event which preceded or coincided with the onset of symptoms. Their symptoms of shortness of breath during paradoxical vocal fold movement classically do not improve with an inhaler. Episodes occur when the vocal cords adduct during inspiration, which causes stridor and 'air hunger' and associated panic. Future episodes can occur with minimal stimulation, and sometimes even without exercise. Pulmonary function tests can be performed, which may show a flattened inspiratory loop, or might be normal. Respiratory physicians, ENT, and speech and language therapist involvement are needed. During voice clinic evaluation, all routine investigations may appear normal. Treatment is often centred on the fact that it helps to diagnose the condition and acknowledge it. Breathing exercises with speech, and abdominal breathing with open-throat breathing, can be taught to break episodes when they occur. These skills can be taught with biofeedback, for example, on a treadmill during flexible endoscopy. It can be useful to enlist the help of a psychologist/sports psychologist in managing these patients. In recalcitrant cases, a psychiatry evaluation can be considered, as can acupuncture, or exercise stress laryngoscopy. Exercise stress laryngoscopy is a laryngeal evaluation during exercise, which attempts to recreate the physiological conditions experienced during episodes of this condition. The equipment needed to perform these investigations includes a treadmill, a flexiscope or strobe, a camera and tower, pulse oximetry, electrocardiography, spirometry, and concurrent measurement of vital signs. It is important to thoroughly explain the procedure and demonstrate all of the equipment prior to beginning the investigation.

Puberphonia

This is a disorder that occurs when there is a failure of the voice to alter from a higher pitch associated with early childhood to the lower vocal pitch of adolescence and adulthood. It may be due to a maladaptation to the psychological changes that occur during puberty. Hyperfunction of the cricothyroid muscle and muscular incoordination combined with the psychological uncertainties of puberty may be contributing factors. Children with this condition often have breathiness and decreased power of vocal projection. Treatment involves voice therapy and problem-directed psychotherapy.

Dysphonia in professional voice users

There is a particular subset of professional voice users who are children, who face challenges with dysphonia in tandem with their adult counterparts. This is a type of functional disorder which benefits from a multidisciplinary team approach by ENT, a voice therapist,

voice coach, and occasionally with a psychologist to best serve the demanding voice needs of these professional performers.

Less common causes of paediatric dysphonia

Dysphonia following airway reconstruction surgery

Laryngotracheal stenosis and reconstruction surgery is covered in Chapter 86. Premature births are on the rise in many countries.[12] As more babies are surviving premature births and requiring tracheostomies in their first few months of life,[13] it is important to recognize the potential impact of laryngotracheal reconstruction surgeries on voice. During these surgeries, the primary goal is improvement of and creation of a safe airway. Voice concerns are generally of secondary importance. These patients may have had multiple surgeries in the past, potentially creating scarring, bowing, or height mismatch of the vocal cords not allowing a consistent mucosal wave along the entire vocal fold yielding a dysphonic voice (**Figure 79.11**). If the posterior glottis is too wide, there can be the creation of a posterior glottic gap and inability to fully close the glottis effectively. This results in a supraglottic speech pattern and dysphonia secondary to it. Following these surgeries and decannulation of tracheostomies, these children will go on to have concerns around socializing, sports participation, and ultimately potential voice concerns affecting choice of occupation. One innovative way of assessing movement of the larynx in these patients is to perform a dynamic CT voice, which may help in diagnosing anatomical areas in need of further surgery or speech therapy. In these patients, the goal of voice 'restoration' involves changing to glottal from supraglottal voice, and improving loudness. Vocal fold injection may help to test for benefit of further surgery and may help glottic insufficiencies. However, it will not improve a posterior glottic gap. For posterior glottic gap post-laryngotracheal reconstruction, an endoscopic posterior cricoid reduction in an older child has been shown to reduce a posterior glottic gap.[14] This can be performed with a laser.

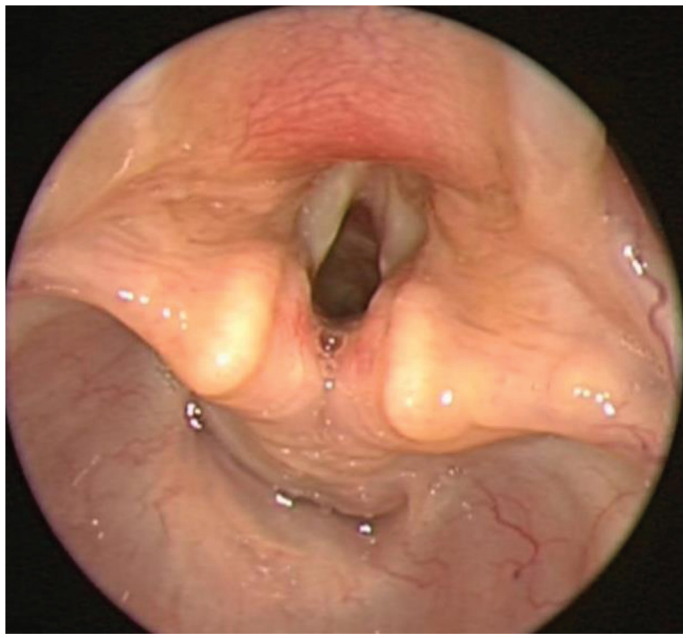

Figure 79.11 Post-reconstruction dysphonia with height mismatch of vocal folds.

Candidacy for this procedure is decided with voice evaluation in the clinic, followed by dynamic voice CT and a screening laryngoscopy and endoscopy.

Dysphonia in children with craniofacial anomalies

Children with craniofacial anomalies often develop dysphonia. In one study, 10% of these patients who were evaluated for dysphonia were found to have an organic vocal fold pathology causing their symptoms.[15] Half of these findings were attributable to iatrogenic causes. Issues with resonance caused by hyper- and hyponasality were also common. The pVHI score was found to be a helpful adjunct in diagnosing those children who would benefit from speech therapy.

Controversies in management of vocal fold pathologies

One of the biggest controversies in paediatric dysphonia is when to operate on children with vocal fold pathologies. There are no clear guidelines, however most practitioners feel that after the age of 7 years, recalcitrant benign lesions may require surgery. Of course, most practitioners adopt a conservative approach for as long as possible. It is sensible to consider the maturity of a child prior to suggesting surgery, as all postoperative patients will need to commit to a voice therapy programme. If the child is not considered mature enough to partake in a preoperative voice therapy programme, then surgery should be postponed until they are ready to engage with speech and language therapy.

There is no definitive decision on how much voice therapy is needed in most cases. However, it is reasonable to undergo a trial of ten sessions or up to 2 months of voice therapy at least, prior to reassessment and decision regarding further/different therapies or management strategies. Treatment of underlying LPR, gastro-oesophageal reflux disease, or eosinophilic oesophagitis is essential. When conservative management fails, it is pertinent to consider changing the type of voice therapy, or entertaining the possibility of unilateral vocal fold surgery if necessary initially, and choosing to operate based on the patient's and family's perceptions and opinions. It is important to stress to the patient and family that there is a mandatory voice rest period required following surgery. If this is not possible, then surgery may not be a wise choice. Surgery is always minimal, by use of subepithelial dissection techniques including the formation of microflaps.

Other relevant medications

Proton pump inhibitor

A recent survey of American Society of Pediatric Otolaryngology members showed a lack of consensus on proton pump inhibitor dosage and duration of treatment of gastro-oesophageal reflux disease in neonates and infants.[16] As with all medications in children, care should be taken to prescribe proton pump inhibitors based on age and weight.

Quadrivalent HPV recombinant vaccine

This is frequently used when systemic RRP treatment is deemed necessary. Systemic treatment is usually considered in cases of aggressive disease requiring more than four surgeries per year, or in cases of spread of disease in the distal airways, or rapid regrowth and airway compromise. A recent prospective study by Matsuzaki et al.

has shown significant reduction of RRP lesions in adults injected with the quadrivalent vaccine during their treatment.[17] Hopefully, with better access to the vaccine for children, there will ultimately be a reduction in the incidence of RRP. It is also helpful to know that vaccination during active RRP is beneficial in reducing disease burden.

Potential treatments modalities include the following:

- *Voice therapy with speech language pathologist*: vocal hygiene and abuse reduction, hydration, increase/decrease glottal closure, decrease muscular tension, respiration, easy onset, improve co-ordination of respiration and phonation, resonance/voice placement, decrease ventricular phonation, raise/lower pitch, increase/decrease loudness, lower laryngeal placement, reinforce reflux treatment.
- *Medical therapy and minimizing environmental factors*: hydration, environmental allergies, allergic rhinitis (avoidance of antihistamines, use of intranasal steroid sprays), asthma (inhaled steroid preparations), gastro-oesophageal reflux disease (basic precautions: antacids, prokinetic drugs (cisapride), H_2-receptor antagonists, proton-pump inhibitors (omeprazole)).
- *Surgical therapy when medical management/voice therapy fails*: conservative excision of benign lesions, 'cold' techniques, microlaryngeal instruments; avoidance of laser on true vocal folds; submucosal infusion technique, management of laryngeal papillomas, injection medialization laryngoplasty, non-selective recurrent laryngeal nerve reinnervation.

Surgeries

Unilateral vocal fold palsy

If, following voice therapy, a child's voice quality remains weak, or there are concerns about aspiration risk, vocal fold medialization surgeries can be considered. There are three main techniques in use currently.

Injection medialization

Although injection of material just lateral to the vocal fold can be performed awake as an outpatient procedure in adults, in children this procedure requires a general anaesthetic (**Figure 79.12**). None of the current synthetic materials are permanent, thus a patient may require multiple anaesthetics for repeated injections. However, injection medialization can be used as a temporary measure while waiting for full recovery of the nerve, or while assessing the potential benefit the patient may acquire from more permanent surgery in future. Younger children undergoing injection medialization should stay overnight for pulse oximetry and observation to ensure no airway compromise following the procedure.

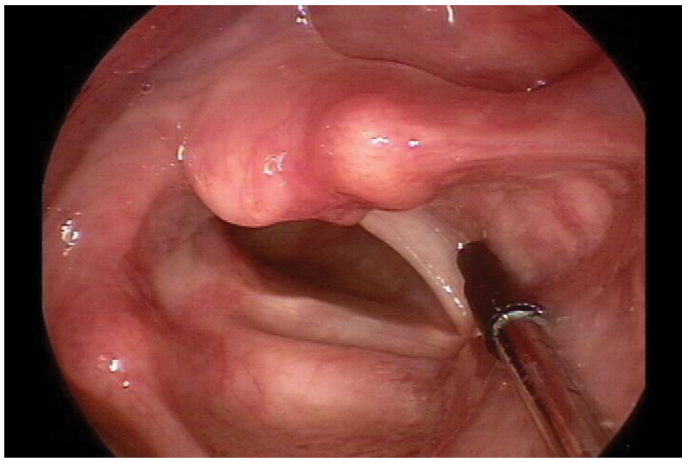

Figure 79.12 Injection medialization of vocal fold palsy.

Type I thyroplasty

In adults, type I thyroplasty is performed usually under light sedation to confirm the quality of phonation with the implant in place, so that adjustments can be made. However, children need a full general anaesthetic, which can make placement of the graft somewhat challenging. It is also worth mentioning that a younger child will outgrow the graft, and so may require revision surgery if there is ongoing atrophy of the vocal fold. Therefore, it may be prudent to postpone thyroplasty surgery until they are older.

Non-selective laryngeal nerve reinnervation

Laryngeal reinnervation has been proven to be beneficial in adults with chronic unilateral vocal cord paralysis. There are fewer studies in children. A review by Zur et al. in 2015 showed that the largest benefit from reinnervation surgery was seen in children from 1 to 2 years after the initial vocal cord palsy, in cases of unknown aetiology.[18] The advantages of reinnervation surgery include one procedure under general anaesthesia which is low risk, and does not preclude the use of medialization thyroplasty in future. Disadvantages include no long-term data, the need for an incision in the neck, and the need to wait months for an outcome. Preoperative workup includes a full voice evaluation as detailed previously. It is important to discuss potential outcomes and set reasonable expectations with parents prior to surgery, including the need to wait many months prior to hopefully having a successful improvement in voice quality. Laryngeal EMG may be helpful as an adjunct to diagnosis and planning; however, it may require a general anaesthetic in a child. It is important to explain to the parents that after the surgery the vocal cord may remain paralysed, but the tone will be improved, hence resulting in a better closure of the cords and improved acoustics (**Figure 79.13**).

Procedure for Removal of a Vocal Cord Cyst

- General anaesthetic, small endotracheal tube where possible or spontaneous ventilation without intubation.
- A submucosal infusion with saline can be helpful to hydrodissect the lesion from the underlying tissue.
- A sickle knife is used to lift the mucosa at the inferior aspect of the free edge of the vocal fold. The incision should be longer than the length of the cyst, for good exposure.
- Using an upwards 70° scissors and a microflap elevator, gently dissect out the cyst.
- Leave superficial lamina propria intact.
- Use a size 3 suction and pledget when needed.
- The cyst can rupture during microdissection. Be careful to gently peel the cyst away with an upwards pointing micro-cupped forceps.
- Ensure opposition of the incised mucosal edges upon closure.

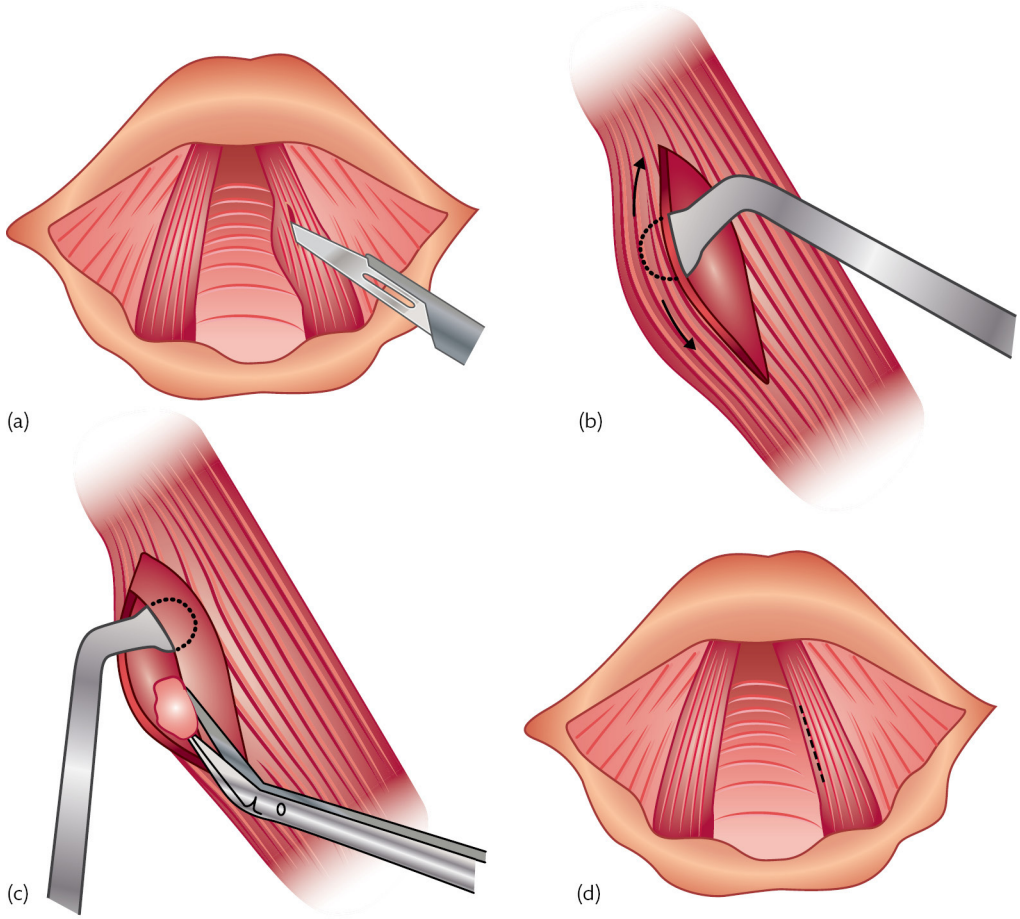

Figure 79.13 (a) initial mucosal incision on the lateral vocal fold superior to the vocal fold cyst; (b) sub mucosal dissection of the vocal fold cyst; (c) atraumatic removal of the vocal fold cyst within the vocal fold; (d) final incision view with vocal fold cyst removed, superior to striking edge of vocal fold.
From Sataloff RT, Chowdhury F, Hawkshaw MJ, Joglekar S. Vocal fold cysts. In: *Atlas of Endoscopic Laryngeal Surgery*. Jaypee Brothers Medical Publishers; 2014:57–68.

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80

Paediatric dysphagia

Michelle Wyatt and Vicky Thorpe

Introduction

Dysphagia is defined as a difficulty in or breakdown of the swallowing process. In order for infants and children to reach their growth potential, they need to be able to consume energy and nutrients in a safe, timely, and enjoyable manner. Swallowing difficulties can also have a detrimental effect on chest health and overall development. Studies have estimated that swallowing difficulties occur in approximately 1% of children in the general population, though the incidence rate is much higher in some clinical populations.[1]

There are a multitude of causes of paediatric swallowing disorders, viewed mainly as being structural, neurological, respiratory, or gastrointestinal. Of course, there can be overlap between these aetiologies and acquired psychological problems can also contribute. For the purposes of this chapter, the focus will be on structural issues considered relevant to the otolaryngologist.

Anatomy and physiology of the paediatric swallow

The normal swallow process

There are significant anatomical differences between an infant and an adult. Infants are obligate nasal breathers; their tongue fills the oral cavity and is positioned more anteriorly than in an adult. The larynx sits high up under the tongue base, providing the advantage of making the infant respiratory tract more protected than that of the adult.

Swallowing is a highly complex, integrated process involving a total of 31 pairs of striated muscles. There are motor and sensory components alongside voluntary and involuntary aspects. It is a progression from reflex behaviour to a learned patterned response and is susceptible to problems at all levels of the central nervous system. The swallow process is typically divided into four stages: oral preparatory, oral, pharyngeal, and oesophageal.

In neonates and young infants, all four components of swallowing are reflexive and involuntary. Babies breastfeed or bottle feed via suckling and (later) sucking patterns. This involves liquid being drawn into the mouth via a rhythmical action of the tongue. The tongue controls the fluid and transfers it to the posterior oral cavity. Later in infancy, the oral phase comes under voluntary control; with increasingly textured foods, children will learn to bite and chew and they develop more mature lateral tongue patterns. Typically, by 2 years, children will have developed mature oral stage skills, managing a wide range of textured/regular foods and further developed skills in self-feeding. It is important to note though, 'the readiness for transition feeding relates more closely to developmental levels in gross and fine motor skills than to a specific age'.[2]

The pharyngeal and oesophageal phases of swallowing are involuntary activities. The site of swallow initiation or trigger varies according to age and the consistency of food or fluid. It is largely seen at the level of the ramus of the mandible or within the valleculae space. There is complete velo-pharyngeal contact as the bolus passes into the pharynx. There is pharyngeal closure by wave-like contraction of the three paired pharyngeal constrictor muscles (superior, middle, and inferior) with the tongue base retracting and the posterior pharyngeal wall controlling the bolus with its wave motion. Laryngeal vestibule closure is achieved through (1) epiglottic inversion/downbending and arytenoids moving inwards; (2) the true and false vocal folds contracting, providing additional airway protection (with swallow apnoea); and (iii) the larynx elevating and moving anteriorly. There is complete passage of the bolus through the cricopharyngeal sphincter into the oesophagus through relaxation of the cricopharyngeus muscle and hyolaryngeal excursion. The pharyngeal stage or time for the bolus to move through the pharynx is 0.5–1 second.

The oesophageal stage is an involuntary phase with a duration of 2–20 seconds. The bolus moves through the oesophagus to the stomach by peristaltic action.

The abnormal swallow process

Difficulties at the oral stage can result in the following problems: (1) anterior loss; (2) reduced bolus formation, control, manipulation, and containment; (3) prolonged transit times with slow or ineffective transfer; and (4) oral residue post swallow.

Key physiological markers of abnormal swallowing at the pharyngeal stage are (1) timing of swallow initiation; (2) bolus transit and clearance in the pharynx; and (3) airway protection before, during, and after swallowing. Reduced laryngeal vestibule closure can result in epiglottic undercoating (material seen on the

undersurface of the epiglottis/coating the arytenoids), laryngeal penetration (material seen in the laryngeal vestibule up to the level of the vocal folds), and aspiration (material enters the airway below the level of the vocal folds). This can be with or without the presence of a reflexive cough.

Swallow assessment

Clinical—subjective

Clinical assessment is a key part of the overall dysphagia assessment. It provides detailed information on developmental level, feeding skills, swallowing skills, and the overall mealtime experience. It is particularly useful for reviewing oral stage skills.

A clinical assessment involves a detailed medical history (including information on birth history, neurodevelopmental level, chest health, and gastrointestinal/reflux symptoms) alongside discussion with key multidisciplinary team members and a detailed feeding history from parents/caregiver (including episodes of oral with or without tube feeding from birth). Prefeeding observations are carried out, including a developmental and communication screen. An oromotor assessment is performed informing on rate, symmetry, and strength of movement of face, jaw, lips, tongue, and palate. Voice production, secretion management, and spontaneous swallow is also noted. Assessment is typically informal; however, formal assessment tools are available such as the Schedule for Oral-Motor Assessment (SOMA).[3] Quality-of-life measures and dysphagia monitoring/outcomes tools are also available, for example, therapy outcome measures.[4]

Where pharyngeal or oesophageal stage difficulties are suspected (including aspiration risk), further instrumental assessment may be indicated.

Instrumental—objective

The two most common instrumental assessments of the paediatric swallow process are videofluoroscopic swallow study (VFSS) and fibreoptic endoscopic evaluation of swallow (FEES). The goals of the assessment are to (1) assess both swallow function and safety, (2) evaluate compensatory strategies, and (3) develop an appropriate feeding management plan. Both studies provide an accurate diagnosis of dysphagia in paediatric populations when performed, analysed, and interpreted by experienced clinicians.[5]

The VFSS (**Figure 80.1**) is a real-time fluoroscopic motion study to evaluate anatomy and to assess swallowing physiology. It provides a dynamic view of the swallowing process. FEES is an instrumental assessment of swallowing where a flexible nasendoscope is inserted transnasally to directly visualize nasal, oral, and laryngopharyngeal structures, secretions, sensory response, and pharyngeal swallow function. It focuses on the pharyngeal phase and provides real-time information on anatomy and function specific to airway protection. FEES (**Figure 80.2**) is comparable to VFSS in the assessment of the pharyngeal swallow process. Both studies present their challenges with compliance/tolerance in the paediatric population (e.g. ingesting barium, sitting in C-arm of X-ray machine, and having a nasendoscope passed). Miller and Willging[6] report that 'the FEES procedures performed jointly by an otolaryngologist and SLP [speech–language pathologist] offer a team approach to the interpretation and management recommendations'.

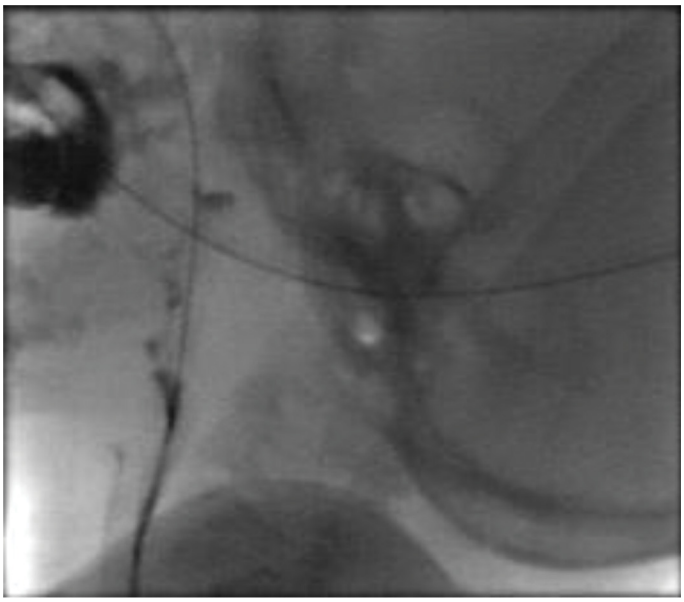

Figure 80.1 VFSS image showing an infant with an nasogastric tube *in situ* with material pooled in the oral cavity, material coating the hypopharynx, material in the pyriform space, and through the cricopharynx, and material in the airway coating the posterior tracheal wall.

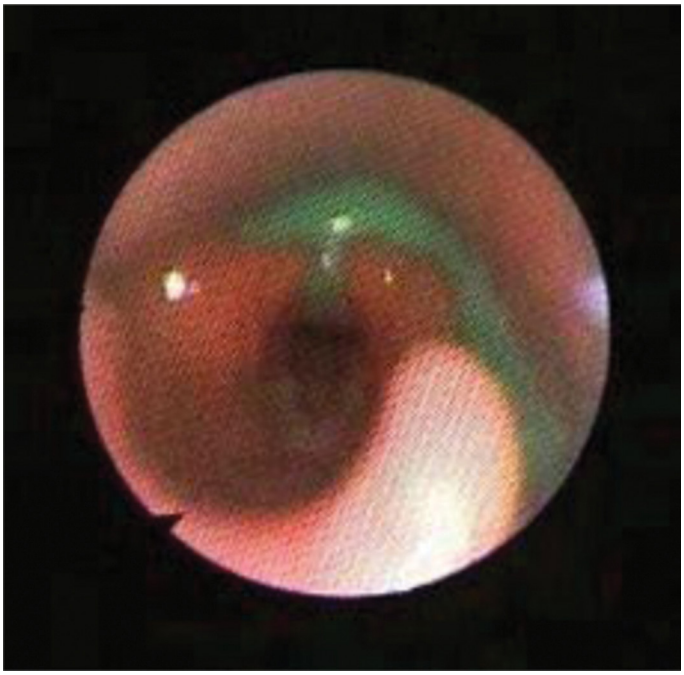

Figure 80.2 FEES image showing material in the left pyriform space and interarytenoid region with material overspilling into the posterior larynx.

Aetiology of dysphagia

Structural

Nose/nasopharynx

Babies are obligate nasal breathers for around the first 6 months of life and so any obstruction to the nasal airway can disrupt the

'suck–swallow–breathe' reflex. If there is an inability to breathe through the nose, then latching on to the breast or bottle to suck will be broken to take a breath via the mouth. The subsequent swallow of milk will be disrupted leading to possible aspiration, swallowing of air, inadequate oral intake, and even failure to thrive.

Causes of neonatal nasal obstruction are various. Masses such as gliomas or encephalocoeles are rare but must be excluded. MRI is the gold standard for diagnosis here.

The most common problem encountered in the general population is neonatal rhinitis (see Chapter 77). This is an inflammatory condition of the nasal mucosa presenting shortly after birth with blockage and clear nasal discharge. It is felt to be due to the passage of maternal hormones across the placenta. It generally improves by the age of 6 months but this may reflect the fact that the nose has grown and the baby has learnt to coordinate mouth breathing and feeding. Treatment depends on the whether the child shows any alarming respiratory problems or is failing to gain weight. A short course of steroid-based nose drops is therapeutic if needed.

Structural underdevelopment of the midfacial skeleton associated with certain syndromes, for example, the syndromic craniosynostoses (see Chapter 90), can also cause feeding issues due to nasal blockage in the first few weeks. Often these children will have more global issues and so a period of supplementary feeding may be required. If there are significant concerns about the obstruction a nasopharyngeal airway is sometimes placed to treat this (see Figure 80.3, nasopharyngeal airway shown). Once breathing improves, feeding usually follows but there can be early issues with regurgitation through the nasopharyngeal airway on swallowing.

Figure 80.3 Child with nasopharyngeal airway *in situ*.

Figure 80.4 Computed tomography image of mid-nasal stenosis.

This can be alleviated by withdrawing the nasopharyngeal airway slightly while the child feeds until they have learnt to coordinate their swallow with the tube in place.

A specific bony anomaly in Apert syndrome is mid-nasal stenosis (Figure 80.4). In this anomaly the distance between the lateral nasal walls in the mid portion of the neonatal nasal cavity is less than 6–7 mm compared to around 12 mm at the posterior choanae.

Pyriform aperture stenosis caused by bony overgrowth of the nasal process of the maxilla is another congenital bony difference which can result in breathing and then feeding difficulty. Again, if it is severe, supplementary feeding or surgical repair of the defect may be required.

From the age of 6 to 9 months the adenoid tissue in the baby will start to enlarge. This lymphoid tissue is situated in the nasopharynx and is the main cause of nasal blockage and feeding problems in a child in the first few years of life. Children with adenoid hypertrophy (Figure 80.5) have a classic open-mouth posture to allow them to breath. This leads to a reluctance to close the mouth to eat and a preference for fluids and softer food which can be moved from the oral to the pharyngeal phases of the swallow rapidly. Drooling is another side effect of adenoid enlargement. Medical therapy involves nasal steroid medication but symptoms often recur when this is discontinued. Surgical removal would be the definitive treatment (see Chapters 75 and 87).

Oral cavity/oropharynx

Feeding difficulties in babies with cleft lip and palate (see Chapter 89) are frequent and are largely due to reduced strength of suck/suction. The baby is unable to achieve sufficient negative intraoral pressure to aid with suction and efficient swallowing. Different feeding utensils are considered to promote weight gain and avoid failure to thrive in this population. Alternative feeding utensils can include squeeze bottles, specialist teats, spoon/cup feeding, and so on.[7] This

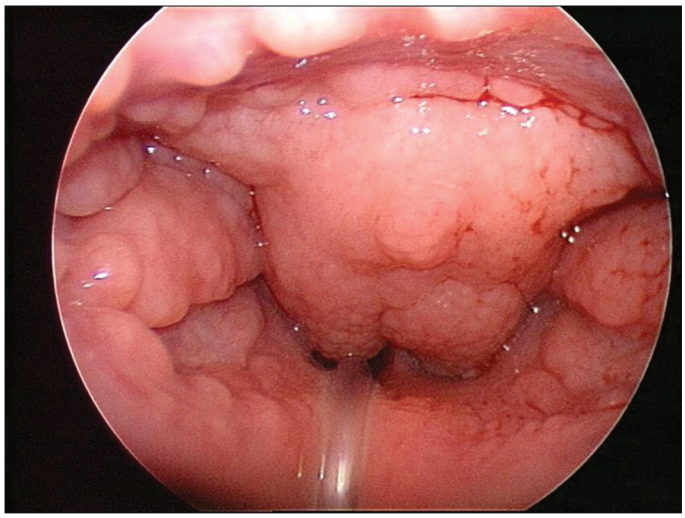

Figure 80.5 Adenoidal hypertrophy.

cohort tend to be under the care of specialist cleft multidisciplinary feeding teams.

Bony differences in this area, such as mandibular hypoplasia, generally cause feeding problems due to airway obstruction. A particular example is Pierre Robin syndrome (Figure 80.6) in which the retrognathia results in the tongue being more posteriorly placed in the pharynx.

An additional concern in this group is the coexisting cleft palate which arises *in utero* as the tongue cannot descend as it should due to the small mandible which then prevents the normal fusion of the palatal shelves. For the vast majority of these babies the mandible grows over the first year or two of life, allowing the tongue position and control to improve with breathing and feeding issues to settle. Again, the nasopharyngeal airway can be used to alleviate obstruction in this group. The feeding techniques used for babies with cleft palate described above are also helpful.

Significant tonsillar hypertrophy (grade 3 or 4) causes an obvious barrier to the food bolus moving from the oral to the pharyngeal phase of the swallow (Figure 80.7).

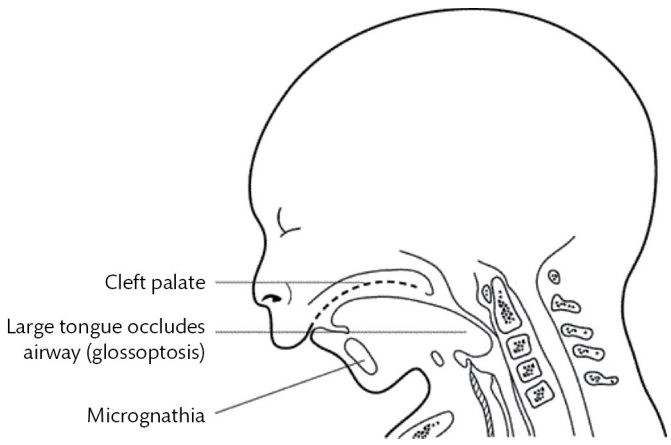

Figure 80.6 Pierre Robin syndrome.

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Figure 80.7 Tonsillar hypertrophy.

Children with this can present with prolonged chewing on food to make the bolus as soft as possible to push past the back of the oral cavity. Fluids can be preferred. Tonsillectomy for dysphagia is described and this seems more reasonable now with the intracapsular technique reducing the risk of significant secondary haemorrhage to such low levels (see Chapter 87).

Hypopharynx/larynx

Laryngeal cleft

Laryngeal cleft (see Chapter 86) is diagnosed in the postnatal period; the more severe the cleft, then the earlier this occurs. Clinical features suggestive of this pathology include stridor, respiratory distress, and feeding difficulties/aspiration.[8] The gold standard for diagnosis is microlaryngobronchoscopy and VFSS.

Type I laryngeal cleft According to the Benjamin–Inglis classification, a type 1 cleft involves a defect in the interarytenoid musculature without any extension below the level of the vocal cords or through the posterior cricoid lamina. An estimate is that at least 20% of these clefts are asymptomatic. However, problems such as choking with eating or drinking, a persistent wet cough, or recurrent chest infections can occur. Initial management would be conservative in the child with good chest health and no significant aspiration demonstrated on VFSS. This would include using a proton pump inhibitor to minimize reflux, the use of thickened feeds, and different feeding utensils (e.g. slow flow teat) to reduce aspiration risk. Surgical repair is recommended in children with persistent dysphagia, comorbid status, poor chest health, and/or difficulties with tolerance to conservative measures (e.g. thickened feeds).[9,10] It is the chapter authors' experience and that of other authors that a certain number of children need to continue with thickened feeds for up to 12–18 months post repair before they can safely move to typical/regular fluids.

The interesting group here is those individuals in whom there is a deep interarytenoid groove but this does not extend far enough to be defined as a true type 1 cleft. If these children are symptomatic (e.g. recurrent chest infections) and there is evidence of an impaired swallow or indeed frank aspiration on VFSS or FEES, then

a posterior laryngeal repair (same endoscopic procedure as a type I cleft repair) is reasonable to consider. This is also an option if there is significant clinical concern about the child, such as evidence of developing bronchiectasis or post lung transplant.

Type II–IV laryngeal cleft In these the cleft extends from below the level of the vocal cords, involving the cricoid plate and extending a variable distance with type IV being the longest. All children go forward for a surgical repair of their cleft. At the authors' centre, a VFSS is typically not performed prior to surgery. As in the type I cohort, children largely need to continue on a modified diet after surgery. They may also present with experiential or developmental feeding difficulties if they have had a prolonged period of tube feeding prior to surgery. They may also have feeding difficulties associated with an underlying or coexisting medical condition (e.g. Opitz G syndrome).

Vocal cord palsy

Aspiration can arise from vocal cord palsy (see Chapter 79). Interestingly, bilateral palsy can be associated with fewer swallow anomalies as the cords tend to sit in a more paramedian position. However, this is only the case in the presence of an adequate airway or a tracheostomy. If there is airway obstruction, feeding coordination can be more difficult. With a unilateral deficit, the cord is usually more lateralized, resulting in an increased likelihood of problems. Consideration as to the safety of the swallow must be given in all those with a diagnosis of vocal cord palsy. Particular high-risk groups are premature infants who have had a patent ductus arteriosus ligation and those post cardiac surgery where the recurrent laryngeal nerve is at risk. Ultrasound scanning has been highly effective at picking up this pathology and is quick to perform with no known side effects.[11]

Laryngomalacia

Laryngomalacia (see Chapter 74) is an established cause of stridor and sleep-disordered breathing in children. Swallowing/feeding difficulties are also reported in this cohort; however, there is variability in the literature as to its prevalence. Feeding difficulties are typically judged to be due to difficulties in coordinating breathing and swallowing. There is general agreement that supraglottoplasty is a safe and effective treatment technique which improves the airway and feeding outcomes and carries a low risk for persistent swallow dysfunction. The need for instrumental assessment in this population both pre- and postoperatively is questioned.[12,13]

It is also recognized that there can be transient difficulties in feeding after surgical intervention for laryngomalacia which usually settle and are felt to be related to oedema. Rarely, longer-term issues can arise due to scar tissue formation. Care should be taken therefore if there is a concern about underlying neurological problems or if a possible syndromic diagnosis exists. These are the individuals in whom such complications are more likely to arise.

Tracheostomy

There is limited evidence in the paediatric literature on the impact of a tracheostomy on a child's swallow physiology. Streppel et al.[14] reported that 70% of children with a tracheostomy tube presented with problems in the oral and/or the pharyngeal phase of swallowing, with 43% seen to aspirate on FEES. However, the primary considerations when determining risk of dysphagia are underlying airway pathology, primary/associated medical condition, and feeding skills. Much of the literature in the adult population demonstrates that a tracheostomy tube does not affect swallow physiology and there is no relationship between tracheostomy tube placement and dysphagia.[15]

Post-airway reconstruction

Transient and more persistent dysphagia has been reported in the literature after paediatric airway reconstruction (see Chapter 86). Duration and severity are largely dependent on airway status, efficacy of laryngeal closure, underlying medical aetiology, and feeding skills. Miller et al.[16] found duration of dysphagia to be increased in children undergoing anterior/posterior grafts with a tracheostomy *in situ*. Compensatory swallow strategies and diet modifications were treatments of choice. Due to the heterogeneity of this cohort, individual assessment (with or without instrumental assessment) is indicated.

Oesophagus

Repaired oesophageal atresia

Dysphagia is relatively frequent after oesophageal atresia repair with prevalence quoted as up to 50%. Both oral and pharyngeal phase problems can occur, with the latter being more common. Abnormal oesophageal motility is felt to be the underlying aetiological factor for this. Gastro-oesophageal reflux disease, which has a strong association, is also felt to play a role. Oral dysphagia may be due to the later onset of oral feeding in this group.

Anastomotic stenosis often occurs too (22–37% of cases) and this can make problems even more severe when superimposed on motility issues.

Gastrointestinal

Gastro-oesophageal reflux

Gastro-oesophageal reflux is the spontaneous return of gastric contents into the oesophagus. The main causes are felt to include transient and inappropriate relaxation of the lower oesophageal sphincter, transient increase in intra-abdominal pressure, and/or spontaneous free reflux from abnormally decreased lower oesophageal sphincter tone. Reflux can result in vomiting with failure to thrive, oesophagitis, aspiration, reactive airway disease (laryngospasm/bronchospasm), and episodes of bradycardia or apnoea. The association of gastro-oesophageal reflux with respiratory disease is strong, and if reflux develops, it can create a vicious cycle, leading to more reflux.

Detection of gastro-oesophageal reflux in babies with feeding problems is important due to the link with respiratory problems, the impact on an infant's desire to feed, and their weight gain. Treatment, if required, is initially with positional management (foam wedges under the mattress) and thickeners. If more severe, proton pump inhibitors are prescribed. Rarely, nasogastric or gastrostomy feeding is required, or surgery in the form of a Nissan's fundoplication.

Problems will resolve in 60–80% of infants by the age of 18 months with the biggest improvement seen at 8–10 months when the child sits up and has a more solid diet.

Psychological

Alongside physiological swallowing difficulties, infants and children can have experiential and/or behaviourally based feeding disorders.

Behavioural feeding difficulties (or food/fluid aversion) occur when a child is unwilling to consume a fluid/food despite sufficient physical skills to do so. Behavioural feeding disturbances may have a variety and combination of causes:

- *Reflux*: this can result in pain associated with eating and drinking and/or an altered sense of hunger.
- *History of long-term tube feeding*: this can result in reduced appetite for oral intake; reduced awareness/understanding of the sensation of hunger; minimal experience of oral intake, and subsequent experiential feeding difficulties.
- *Aversive experiences* in or around the mouth (e.g. tube feeding, suctioning), or *sensory disturbances* (e.g. oral hypersensitivity).
- *Experiences of forced-feeding*, leading to food refusal.

Children with entrenched psychological feeding difficulties need to be managed by a multidisciplinary team including a speech and language therapist, a dietitian, and a psychologist.

Treatment

Individualized and detailed differential diagnosis largely dictates treatment and management options. These may include the following:

- Utensils—slow flow teats.
- Consistencies—thickened fluids.
- Positional—elevated side lying in neonates; specialized seating.
- Compensatory swallow strategies and/or swallow exercises (used more in the older, cognitively able child).
- Alternative and/or augmentative feeding—orogastric tube/nasogastric tube/gastrostomy tube/nasojejunal tube/gastrojejunostomy tube.
- Surgery—such as laryngeal cleft repair.

Treatment options will vary for children and may include one or more of the above. Reassessment is ongoing and management will likely change over time.

Conclusion

This chapter has focused on different structural pathologies that contribute to eating/drinking and swallowing difficulties. The authors acknowledge that neurological impairment and cardiorespiratory conditions are also significant causes of dysphagia. It is beyond the scope of this chapter to discuss these aetiologies and their management in more detail. An interdisciplinary team approach is essential in the effective assessment and management of dysphagia in the medically complex child.

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81

Paediatric neck masses, cysts, and sinuses

Haytham Kubba and Tash Kunanandam

Introduction

This chapter covers a wide variety of lesions with very different presentations. This includes a child with a neck space infection who may be septic or develop airway obstruction, a child with a large neck mass whose parents are worried about cancer, and a child with nothing more than a tiny punctum on the tip of the nose whose parents are blissfully unaware that it connects to a large cyst in the frontal lobes and the risk of meningitis. Although each presentation is different, they all need to be handled sensitively and carefully; while a thorough knowledge of the pathology is essential, the skills to handle worried parents and unwell children are equally important.

This chapter describes the relevant embryology and anatomy, discusses the common congenital and acquired neck lesions, proposes a guide for assessment, and considers some of the controversies in this field.

Anatomy and embryology

From the fourth week of embryonic life, paired ridges are visible on each side of the embryo between the brain and heart. These ridges, or branchial arches, go on to form many of the important structures in the head and neck. The arches are numbered 1–6 from cranial to caudal but, in the human embryo, only four are visible externally: the fifth involutes, the sixth fuses with the fourth.

The arches consist of mesenchyme which gives rise to muscle, cartilage, and bone, together with an artery and nerve for each arch. Abnormalities of the arches therefore give rise to predictable clinical presentations, Treacher Collins syndrome being an example of first and second arch malformations (see Chapter 90).

On the external surface of the embryo, between the arches, are ectoderm-lined grooves known as the branchial clefts. Only the first cleft persists, as the ear canal. Any abnormality of a branchial cleft therefore presents as a congenital sinus opening on (ectodermally derived) skin. Second branchial cleft sinuses are relatively common, first cleft sinuses are rare.

Inside the primitive pharynx of the embryo, between the arches, are endoderm-lined outpouchings—the pharyngeal pouches. Only the first pharyngeal pouch persists as a tract, the Eustachian tube. The second contributes to the tonsil, the third contributes to the inferior parathyroid glands and thymus, and the fourth to the superior parathyroid glands, calcitonin-secreting cells of the thyroid, and parts of the larynx. Any abnormality of pharyngeal pouches will present as a congenital sinus opening on (endodermally derived) mucosal surfaces. A fourth pouch sinus is the more common.

Between weeks 4 and 6 of embryonic life, the frontonasal process gives rise to medial and lateral nasal processes. These fuse around the nasal pits to form nostrils as the maxillary processes grow in from the side to form the upper lip. The palatal shelves grow in from the sides, fuse in the midline, and form the palate between weeks 6 and 12. These structures are all derivatives of the first branchial arch. Failure of fusion of the palatal shelves leads to a cleft palate; this may be associated with a cleft lip if the maxillary processes fail to fuse with the frontonasal process. Any such abnormalities of the fusion lines of mesenchymal processes can result in facial clefts, and epithelial entrapment here can lead to dermoid cysts.

The pinna forms from six protrusions of mesenchyme (the hillocks of His) situated around the first branchial cleft, three on each of the first and second arches, fusing around week 12. Abnormalities here can lead to microtia, accessory auricles, or preauricular sinuses.

The majority of the thyroid gland develops from the foramen caecum at the base of tongue, and descends during early fetal life to the anterior neck. The thyroglossal duct (tract) disappears around weeks 8–10, but persistence can lead to thyroglossal duct cyst formation. The process of thyroid gland descent is often not complete until infancy and it is not uncommon to find the thyroid isthmus at the level of the larynx during open neck surgery in premature babies. Complete failure of descent results in a lingual thyroid, visible on tongue protrusion.

Congenital lumps

Dermoid cysts

Dermoid cysts are evident at birth. They consist of squamous epithelium filled with keratin debris and are common in the head and neck; the usual sites are either end of the eyebrow, postauricular sulcus, floor of mouth (**Figure 81.1**), nasolabial fold, and the midline of the neck. Dermoid cysts at these sites rarely have a sinus opening

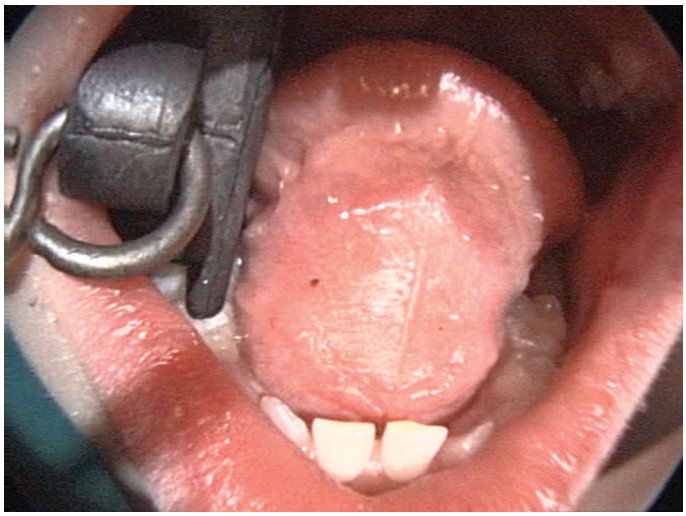

Figure 81.1 A dermoid cyst in the floor of the mouth.

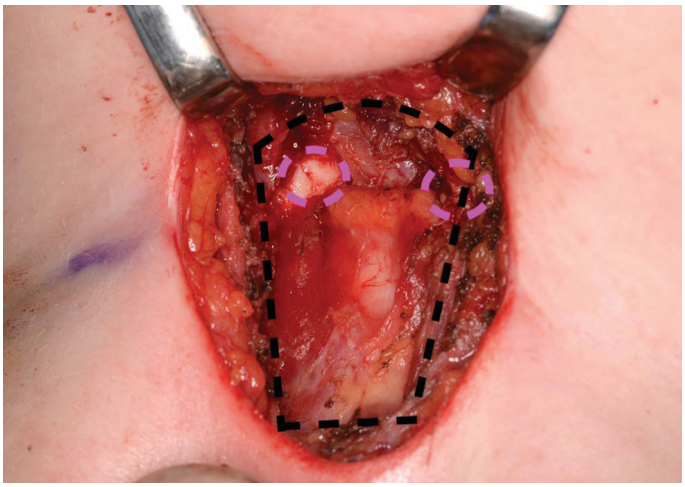

Figure 81.2 The neck following thyroglossal duct cyst excision: the excision margins are marked in black and the cut ends of the hyoid outlined in pink.

and the risk of infection is therefore very low. They are often excised for cosmesis alone.

Midline nasal dermoids

These usually present as a small sinus opening on the nasal dorsum or tip, occasionally with protruding hairs. The sinus gives little clue to the potentially extensive nature of the lesion. The sinus tract leads to a cyst (or chain of multiple cysts), which can extend under the nasal bones, and potentially into the skull base at the root of the nose anterior to the crista galli. The cyst may lie on the dura, or extend into the frontal lobe of the brain. The presence of an external sinus means that bacterial infection is almost inevitable given time. Early magnetic resonance imaging (MRI) scanning is essential to define the extent of the lesion and plan surgery (e.g. neurosurgical input). Excision is often achieved via an external rhinoplasty approach, although a vertical incision on the nasal dorsum may be justified, particularly with extensive skin loss from infection.

Thyroglossal duct cysts

Midline neck lumps are usually thyroglossal duct cysts (~75%) although differentials include dermoids (20%), pretracheal lymph nodes (4%), and rarities such as thyroid isthmus nodules, foregut duplication cysts, and thymic tumours (1%). They are not often apparent at birth but appear during early childhood. Once they present, they may fluctuate in size but rarely disappear completely. They have a high risk of infection over time as bacteria from the pharynx may pass down the tract from the foramen caecum and surgery is often recommended. An ultrasound scan of the neck should be obtained before surgery to ensure that there is a normal thyroid gland separate from the lesion. Simple cyst excision results in high recurrence rates and surgery should involve removal of the tract in its entirety. The tract itself is a fine, indistinct epithelial structure with multiple branches and so surgery involves excising widely in normal tissue. This should include a strip of midline tissue from thyroid isthmus to foramen caecum including pretracheal fascia, central part of the hyoid, and some muscle from tongue base (Figure 81.2).

Meningoencephalocoeles and gliomas

Abnormalities of anterior skull base formation can lead to frontonasal meningoencephalocoeles; these present as intranasal soft masses or extranasal ones at the root of the nose. The mass may consist of dura alone (meningocoele) or a combination of dura and brain tissue (meningoencephalocoele). Classically, such lesions expand on crying, confirming their connection to intracranial contents. If the skull base forms completely during fetal life, a meningoencephalocoele may be 'cut off' from its attachment to the brain, resulting in a mass of ectopic brain tissue (glioma) within the nose or on the nasal dorsum. Masses around the root of the nose and nasal dorsum require imaging with MRI (+/– computed tomography) to exclude a connection to the brain before contemplating surgery. Gliomas are simple to excise: endoscopic resection is often feasible for intranasal ones with external rhinoplasty approach suiting many extranasal gliomas. Meningoencephalocoeles may be approached similarly for excision but with repair of the skull base defect undertaken in conjunction with neurosurgeons.

Haemangiomas

Congenital infantile haemangiomas ('strawberry naevi') are common, affecting approximately 1% of Caucasian infants. These benign tumours of endothelium secrete their own growth factors and follow a characteristic natural history: they are absent at birth but grow rapidly over the first few weeks of life, plateauing around 6–9 months of age, before slowly involuting. The majority disappear completely, although some may take 3–5 years to resolve, depending on the lesion size. Extensive lesions can result in permanent changes in skin pigmentation or atrophy of subcutaneous fat and, even on resolution, can leave behind lumps of fibrofatty stroma.

A child with five or more haemangiomas should undergo liver ultrasound scanning to exclude haemangiomas here. Children with extensive haemangiomas of the face and neck should be investigated for PHACE syndrome, which includes abnormalities of the heart, brain, vasculature, and eyes. It carries a risk of stroke and is therefore important to identify. Investigations should include MRI brain, echocardiography, and thyroid function.

Box 81.1 Absolute and relative indications to treat haemangioma

Absolute indications

- Risk of airway obstruction (lesion within nose, pharynx, larynx, or trachea, or causing external compression).
- Risk of amblyopia (lesion in front of the eye).
- Risk of dysphagia.
- Ulceration.

Relative indications

- Risk of large, cosmetically disfiguring lesion.
- Failure to involute.

Box 81.3 Protocol for initiating propranolol therapy for haemangioma

- Admit for 24 hours if aged less than 8 weeks or weight less than 3 kg.
- Consider electrocardiography and cardiology opinion if family history of heart anomalies, bradycardia (<100 bpm), hypotension, heart murmur, or arrhythmia.
- Start propranolol orally at 1 mg/kg/day in three divided doses.
- Steroid cover may be required initially if symptoms from the haemangioma are severe.
- Check glucose if any change in infant's condition.
- Stop drug if hypoglycaemic, bradycardic, or hypotensive.
- Increase to 2 mg/kg/day after 1 week if tolerated.

Many haemangiomas will resolve naturally without treatment. Many will require treatment (Box 81.1) and propranolol is invariably the first-line therapy. Propranolol is usually very effective and well tolerated, but is required for 12–18 months (Box 81.2). Not all children tolerate propranolol, and not all haemangiomas respond, so other treatments will occasionally be required (Box 81.3). A small lesion in the nostril, for example, may be best served by surgical excision. Haemangiomas also respond rapidly and predictably to corticosteroids, which can be used as initial treatment for symptoms of stridor. Bevacizumab is a VEGF inhibitor which can be used for haemangiomas.

Box 81.2 Treatment modalities for haemangioma

- Propranolol.
- Corticosteroids.
- Vincristine.
- Interferon alpha.
- Bevacizumab.
- Surgical excision.
- Tracheostomy.

Vascular malformations

Vascular malformations are present at birth and grow in proportion to the child. They are named according to their vessel of origin. A reference to the full classification of vascular lesions of the head and neck is given at the end of this chapter in the 'Further reading'.

Capillary malformations (port wine stains) are mostly a cosmetic concern unless associated with Sturge–Weber syndrome (cerebral angiomas, seizures, intellectual disability, glaucoma). Cosmetic treatment involves depigmentation with pulsed-dye laser.

Arteriovenous malformations are thankfully rare. They result in progressive tissue hypertrophy with extensive disfigurement and bleeding. Management is complex: embolization and surgical excision are often incomplete and inadequate in preventing progressive growth of the lesion. Trametinib is a MEK inhibitor which may have some effect in managing arteriovenous malformations.

Lymphatic malformations present as soft, compressible, multicystic masses in the head and neck (Figure 81.3). They arise from a developmental abnormality of lymphatics leading to fluid accumulation within thin-walled epithelial cysts. Although congenital, they may only present much later on in life as the lesion swells after minor trauma, a viral infection, or other trigger. Generally, lymphatic malformations below the hyoid bone tend to be macrocysts

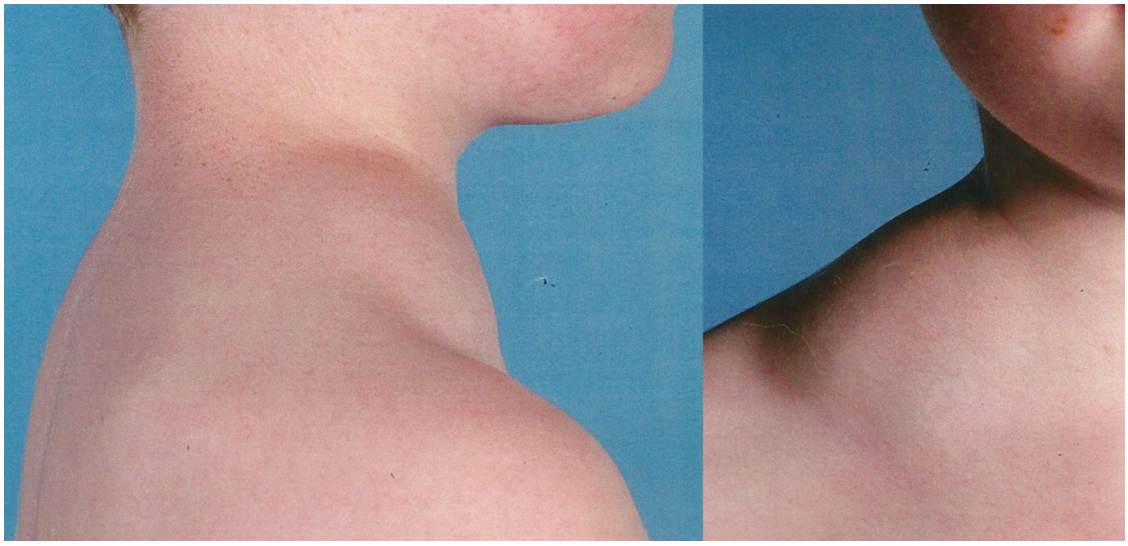

Figure 81.3 A lymphatic malformation of the posterior triangle of the neck.

with microcystic disease more prevalent above the hyoid. Once apparent, lymphatic malformations may fluctuate in size with episodic swelling due to infection, bleeding (from associated venous elements), or without a discernible cause. They can be sufficiently large to cause cosmetic and functional (e.g. airway compromise) concerns. Treatment options are discussed later in this chapter but include mammalian target of rapamycin (mTOR) inhibitors such as sirolimus, injection sclerotherapy, and surgery.

Congenital sinuses

Preauricular sinuses

Preauricular sinuses are very common. They arise due to abnormalities of fusion of the hillocks of His and are distinct from true anomalies of the first branchial cleft. Most are asymptomatic and require no treatment. Recurrent infections, however, can lead to extensive scarring and skin loss so excision should be recommended if infections are occurring. Preauricular sinuses have a very typical location immediately adjacent to the helix of the pinna at its most anterior point. The epithelial tract is very fine, branches frequently, and passes close to (and occasionally through) the cartilage of the helix and ends blindly, superficial to the plane of the parotid fascia, zygomatic arch, and temporalis fascia. Surgery consists of wide excision in normal tissue, rather than attempts to identify and dissect the tract.

Second branchial cleft sinuses

Congenital sinuses of the second branchial cleft are relatively common. They typically occur as small openings along the anterior border of the lower third of sternomastoid. They rarely lead to infection but can be associated with continuous clear discharge due to the presence of ectopic salivary tissue in the tract wall. When symptomatic and surgery is being considered, the tract here is usually thick and easy to dissect due to a muscular layer; it follows a very predictable course superiorly, over the hypoglossal nerve, between internal and external carotid arteries, ending a few millimetres medial to the carotid bifurcation at the superior constrictor of the pharynx. True fistulae extending through into the tonsil fossa are rare and excision of the tonsil is rarely needed.

It is worth noting that many older textbooks will show drawings (never photographs) of second branchial cleft sinuses together with *branchial cysts*. These two pathologies rarely occur together. Branchial cysts are very rare in children, tend to occur in the third and fourth decades of life, are unlikely to be branchial in origin, and may not even be congenital. They will not be considered further in this chapter. It is also worth noting that second branchial cleft sinuses will often be erroneously referred to as *branchial arch fistulae* by surgeons wishing to demonstrate their ignorance of embryology.

First branchial cleft sinuses

Congenital sinuses of the first branchial cleft are much rarer and can be very challenging to manage. They vary from superficial epithelial structures, which do not involve the facial nerve, to large cartilage tubes lined with squamous epithelium and full of keratin debris, which attach to the inferior surface of the cartilaginous part of the external auditory meatus and extend anteriorly for a variable distance with a varying and often complex relationship to the facial nerve. The superficial epithelial lesions may have a sinus opening in the concha of the pinna or ear canal. The cartilaginous ones often have a sinus opening around the angle of the mandible or along the jawline. There may also be a fibrous band running from the floor of the ear canal to the handle of malleus. Infection is common and excision is usually required. The relationship of the lesion to the facial nerve is unpredictable so detailed imaging is required and surgery should include facial nerve dissection with nerve monitoring.

Fourth pharyngeal pouch sinuses

Recurrent abscesses in the region of the left thyroid lobe can be due to a congenital sinus originating in the pharynx at the apex of the left piriform fossa—a congenital anomaly of the fourth pharyngeal pouch (Figure 81.4). Diagnosis requires pharyngoscopy under anaesthetic with options for concurrent treatment by means of endoscopic cautery of the tract. Recurrent cases may require excision with left hemithyroidectomy.

Midline cervical cleft

Midline cervical clefts are very rare. There is usually a vertical midline patch of red discoloured skin with a small sinus at either end: the inferior one leads towards the sternal notch, and the superior one to an indistinct subcutaneous cord attached to the inner surface of the mandible. Treatment involves excision of the skin lesion and associated subcutaneous cord coupled with z-plasty wound closure to prevent web contractures. Surgery is usually performed early to prevent impaired mandibular growth.

Acquired sinuses

Persistent discharge occurs when a *pre-existing lesion* has been infected (a thyroglossal cyst or a dermoid, for example) or when there is *non-tuberculous mycobacterial infection* in a lymph node.

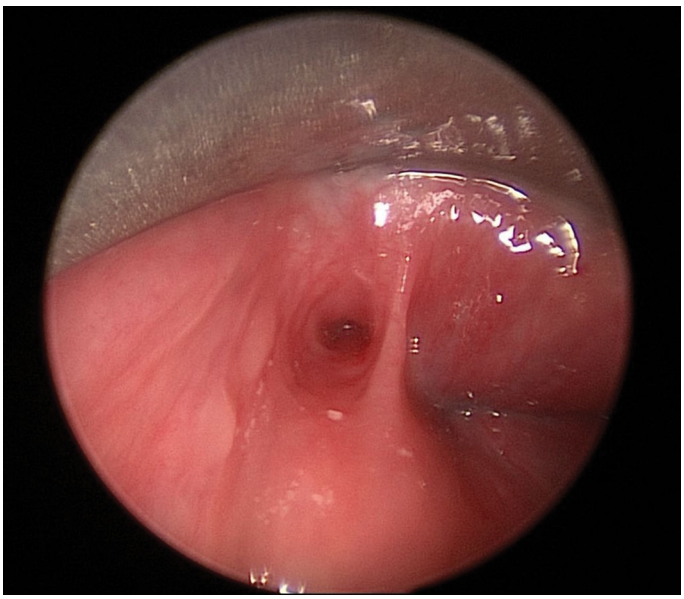

Figure 81.4 A fourth pharyngeal pouch sinus visible at the apex of the left pyriform fossa.

Spontaneous discharge from an *acute bacterial lymph node abscess* results in healing within a few days.

Acquired neck lumps with fever and malaise

Lymphadenitis and lymph node abscess

A neck lump in the presence of fever and malaise suggests infection. The lymph nodes may be enlarged as part of the physiological response to any local infection in the head and neck area but *acute bacterial lymphadenitis* occurs when infection spreads from the primary source to the nodes. This is one of the most common emergency presentations to any paediatric otolaryngology department. Lymphadenitis can progress with central necrosis of the node to form an *abscess*. These are most commonly located superficially in the submandibular triangle or adjacent to the upper third of sternomastoid. Lymph nodes in less accessible areas can give rise to *deep neck space infections* such as *retropharyngeal* and *parapharyngeal abscesses*. *Quinsy (peritonsillar abscess)* also occurs in children, usually without much recurrent tonsillitis. Surgical drainage is required for any significant collection, although quinsy in young children will respond well to intravenous antibiotics.

Kawasaki's disease

Consider *Kawasaki's disease* (mucocutaneous lymph node syndrome) in children under 5 years with cervical lymphadenopathy. This autoimmune vasculitis presents with fever, oedema of the hands and feet, erythema of the conjunctiva, and erythema of the oral mucosa. Arthritis, aseptic meningitis, and myocarditis may also occur in the early stages. Desquamation of skin on the hands and feet occurs later. Diagnosis is important as coronary aneurysms can occur and be fatal without treatment. Treatments include aspirin, intravenous immunoglobulins, and steroids.

Periodic fever, aphthous stomatitis, pharyngitis, and adenitis (PFAPA) syndrome

PFAPA is of unknown aetiology. It presents in young children as episodes of high fever with mouth ulcers, pharyngitis, and tender cervical lymphadenopathy occurring every 3–5 weeks for at least 6 months. The child is perfectly well in between episodes. Treatment may be supportive alone, but active options include steroids and tonsillectomy.

Acquired neck lumps without fever

Lymphadenopathy

It is normal to be able to feel cervical lymph nodes in preschool children, which are small (<5 mm), ovoid, soft, and mobile. Nodes located over firm muscle (trapezius) or bone (lower border of mandible) are particularly easy to feel.

Local inflammatory stimuli often lead to temporary enlargement of nodes according to normal physiological responses. Common stimuli include upper respiratory infections, dental decay, tonsillitis, adenoiditis, head lice, and eczema. A well, young child with small nodes that fluctuate in size is most likely to have *reactive cervical lymphadenopathy* and requires no investigation, only reassurance and possibly attention to the underlying inflammatory focus. If investigation cannot be avoided, ultrasound can demonstrate nodal features specific for reactive hyperplasia (ovoid shape, echo texture, hilar architecture, blood flow pattern on Doppler).

Rapidly growing nodes, supraclavicular fossa nodes, and nodes measuring 2 cm or more in maximum diameter require urgent tissue sampling.

A suggested algorithm for the management of cervical lymphadenopathy in children is included in the 'Further reading'.

Non-tuberculous mycobacterial infection

Mycobacteria are widely present in the environment and are a common cause of cervical lymphadenitis in preschool children. Common organisms include *Mycobacterium scrofulaceum*, *M. bovis*, and *M. avium intracellulare*. In the UK these are commoner than tuberculosis, but a history of foreign travel and potential tuberculosis contacts should always be sought. Non-tuberculous mycobacterial infection initially presents as enlarged nodes, typically in the parotid or submandibular areas, progressing to violet-red skin discolouration, then a fluctuant subcutaneous abscess, skin breakdown with a discharging sinus, and finally resolution with a small puckered scar (Figure 81.5). This process can take anything from 6 months to 3 years. Non-tuberculous mycobacterial infection can be distinguished from acute bacterial lymph node abscess due to lack of malaise or fever, and the longer history (few weeks rather than a couple of days). Incision and drainage are to be avoided as they lead to skin breakdown and prolonged discharge, potentially compromising the ultimate cosmetic result. Diagnosis is clinical, although a chest X-ray and tuberculin skin test are useful to help rule out tuberculosis. Samples of any discharge may show acid-fast bacilli on Ziehl–Neelsen staining but most often these are absent, and culture identifies the organism less than half the time. Treatment

Figure 81.5 Non-tuberculous mycobacterial infection of the neck.

options include watchful waiting, antibiotics, curettage, and surgical excision, which are discussed later in this chapter.

Cat scratch disease

The faeces of cat fleas are the source of this bacterial infection, caused by *Bartonella henselae*. It is usually asymptomatic, benign, and self-limiting in immunocompetent individuals. Positive serology can avoid the need for excision biopsy of lymph nodes so the diagnosis is always worth considering.

Specific viral infections

HIV can cause a generalized, diffuse lymphadenopathy with adenotonsillar hypertrophy soon after seroconversion. It usually settles spontaneously but obviously it is an important diagnosis to make. It is worth considering HIV-associated lymphoma if a lump is rapidly growing.

Cytomegalovirus and Epstein–Barr virus can also cause prolonged cervical lymphadenopathy. Again, positive serology can avoid the need for invasive tissue sampling.

Other inflammatory conditions

Langerhans cell histiocytosis is a disease of unknown aetiology presenting as bilateral cervical lymphadenopathy in preschool children. It is an aggressive, multifocal disease with an appreciable mortality. Excision biopsy of an affected node is required for diagnosis.

Castleman's disease is a B-cell lymphoproliferative disorder. In children, it most commonly presents as a mass of nodes in one area of the neck. Diagnosis is histological and treatment may be limited to excision of the affected nodes. A more aggressive, multifocal form occurs mostly in adults, often in association with HIV infection.

Tumours

Thankfully, childhood cancer is much rarer than in adults but it is still the second commonest cause of death in childhood (after trauma). Around 12% of all malignancies in children arise in the head and neck. Diagnostic delay is unfortunately common. The nature of childhood malignancies is very different to those in adults. The commonest malignancies are lymphoma (27%), neural tumours (23%), thyroid tumours (21%), and soft tissue sarcomas (12%). Squamous carcinoma is extremely rare. Any large, rapidly growing or unusual lump should lead to urgent imaging (often MRI) and tissue sampling. It is important to liaise early on with oncologists and histopathologists for advice on the ideal investigations and preparation of tissue samples.

Assessment

Any encounter with a child in the acute setting should begin with rapid initial assessment for life-threatening airway obstruction, looking for signs of effort of breathing (stridor, stertor, respiratory rate, pulse rate, accessory muscle use, subcostal indrawing, intercostal recession, tracheal tug) and effectiveness of breathing (colour, conscious level, demeanour, pulse oximetry). Dehydration, pain, and sepsis should also be recognized and managed.

A comprehensive history should cover features of the lesion (duration, changes in size) but also associated features such as foreign travel, HIV or tuberculosis contacts, night sweats, and weight loss.

Examination should include ears, nose, oral cavity (including teeth), and the entirety of the neck, before moving on to axillae, groins, abdomen, heart, and hands. Repeat the examination after pain, fever, and dehydration have been corrected, as the signs will often have changed and may be much easier to appreciate.

Trismus is a sign of irritation of the pterygoid muscles from infection, usually quinsy or a parapharyngeal abscess. Trismus can also occur with malignancy and trauma. It is measured as the maximum distance between the upper and lower incisors, measured in relation to the child's fingers: normal mouth opening is equivalent to the width of three fingers. It is not only a clue to the location of the pathology, but also a warning of a potentially difficult airway for the anaesthetist. Other potential signs of deep neck space infection include torticollis, palatal bulging, retropharyngeal swelling, and stertor.

Awake transnasal fibreoptic laryngoscopy can be very useful but is only really practical in infants and children over the age of 7 years and also carries the risk of precipitating complete loss of the airway.

Investigations

Blood tests

Laboratory investigations for any child presenting with fever and malaise should include full blood count and C-reactive protein.

Imaging

Ultrasound scanning is quick, painless, and easy to perform, rendering it the first-line investigation in the evaluation of most neck lumps and infections. It can distinguish solids from liquid, is useful for diagnosing the presence and location of pus, and in distinguishing cystic lesions from solid tumours. Ultrasound is also essential to confirm the presence of a normal thyroid gland when surgical excision of a thyroglossal duct cyst is being considered.

Computed tomography scanning is useful in suspected deep neck space infections providing superior anatomical information compared to ultrasound for locating a collection of pus and planning surgery. If general anaesthesia is required, the surgical team should be prepared to proceed to theatre under the same anaesthetic for surgical drainage.

MRI gives excellent soft tissue detail and is essential in planning treatment for tumours, lymphatic malformations, first branchial cleft anomalies, meningoencephalocoeles, and midline nasal dermoids. It can be noisy, time-consuming, and unpleasant to undergo an MRI scan, but most children aged 7 years or older will tolerate the procedure awake. Neonates (<6 weeks of age) can undergo the procedure under natural sleep after a good feed. Children between these ages will often need a general anaesthetic to get good-quality scan pictures.

Other investigations

A plain chest X-ray and a tuberculin skin test should be considered for children with a clinical diagnosis of non-tuberculous mycobacterial infection of cervical lymph nodes. This is especially important if the child has recently travelled to an area where tuberculosis is endemic or has had contact with a family member who has tuberculosis. Non-tuberculous mycobacterial infection will usually give a weakly positive reaction to tuberculin.

Urgent echocardiography and the opinion of a cardiologist should be sought if Kawasaki's disease is suspected.

Preauricular sinuses and second branchial cleft sinuses are easy to diagnose on examination alone, and they have a very predictable anatomical course. Neither requires any investigation for diagnosis or to plan surgical excision. However, children with preauricular sinuses may have branchio-oto-renal syndrome, particularly if the preauricular sinuses are bilateral and associated with second branchial cleft sinuses. A hearing test and an ultrasound of the kidneys should be requested.

Controversies

Congenital lesions and timing of surgery

There is some evidence that general anaesthesia in early infancy can have long-lasting effects on neurodevelopment. It is therefore wise to defer non-urgent surgery until after the first or second year of life. However, some conditions necessitate earlier surgery. Second branchial cleft sinuses can be easily excised through a single incision in preschool children. Once children reach school age, the distance from the sinus to the carotid bifurcation and pharyngeal muscle increases and more than one neck incision may be required ('step-ladder'). Similarly, for nasal dermoids, once the midface has started to grow during the school years, the distance from columella to root of nose becomes too much of a challenge for an external rhinoplasty approach. The fact that some lesions communicate with the outside world (via the sinus opening of nasal dermoids or foramen caecum for thyroglossal duct cysts) means that early surgery is also necessary to prevent infection. Dermoids in other sites, preauricular sinuses, and second branchial cleft sinuses, on the other hand, have a much lower risk of infection and can often be managed conservatively.

Surgeon-performed ultrasound

Although commonplace in much of Europe and North America, it is uncommon in the UK at present for surgeons to perform ultrasound scans in the outpatient clinic as part of routine physical examination. It is a useful skill that every head and neck specialist should consider learning.

Excision biopsy or core biopsy for lymph nodes

Core biopsy allows sampling of a suspicious lymph node, avoiding surgical scars and minimizing risk of damage to adjacent structures such as cranial nerves. In adolescents it may be possible without general anaesthesia. The tissue sample is large enough to retain many of the architectural features of the lymph node and allow immunohistochemistry; both are essential for diagnosis in lymphoma and other tumours, although diagnosis will still be easier with a complete lymph node. The decision is really one for the pathologist rather than the surgeon, to be able to confirm diagnostic certainty on the sample provided by a core biopsy.

Non-tuberculous mycobacterial infection

Management here is controversial and none of the treatment options are ideal. Complete surgical excision often requires quite extensive surgery (parotidectomy and/or selective neck dissection) for essentially benign, self-limiting disease in a young child. The facial nerve and its mandibular branch in particular are very much at risk. Antibiotic therapy (single agent or multiagent) is often no better than conservative management. Allowing the disease to run its course avoids the disadvantages of surgery but does leave the child with an unsightly, discharging sinus for many months, which may prevent attendance at day care. The final scar varies but can be particularly unsightly in cosmetically sensitive areas such as the cheek. If the skin has already broken down, a conservative approach is recommended. If the skin is still intact, and the lesion can be accessed via a cosmetic incision away from the inflamed skin then curettage of the necrotic subcutaneous tissue could be undertaken, preserving the skin but with no attempts to excise the underlying lymph node and increase surgical morbidity.

Management of lymphatic malformations

Injection sclerotherapy has many potential advantages over surgery for lymphatic malformations, not least avoiding a scar and the risk of damaging major neurovasculature. However, each cyst must be individually punctured, drained, and injected under ultrasound guidance. Large lesions will require multiple procedures (often a course of three at 3-month intervals) each requiring a general anaesthetic. Each procedure runs the risk of swelling before subsequent scarring. This is dangerous for lesions adjacent to the airway. The ideal lesions for sclerotherapy are those formed of a small number of large cysts, below the hyoid. Of course, these lesions are also easiest to excise surgically. Suprahyoid microcystic lesions are often very extensive and may involve important and inaccessible areas such as the tongue base and parapharyngeal space; unfortunately, injection therapy has little to offer here.

Regardless of location, surgical excision is usually very challenging. The lesions can be very extensive and tend to wrap around major vessels and nerves with no respect for fascial planes. It is simply not acceptable to leave a child with nerve palsies and disfigurement from ill-judged surgery to address benign disease. Surgery should be performed by those with experience in this field. Perioperatively, it is always preferable to leave a small amount of disease behind rather than risk permanent neurovascular injury. Prolonged periods of lymph drainage and seromas are common after surgery and should be managed expectantly.

There has been growing excitement about the mTOR inhibitor drug *sirolimus*. So far, evidence suggests it may arrest growth or cause limited shrinkage of some malformations. Its exact role is not yet clear.

Alpelisib is a PIK3K inhibitor which may have some use in managing lymphatic malformations.

PFAPA syndrome

Trials suggest that tonsillectomy leads to fewer episodes of fever, but the condition settles spontaneously making observation a perfectly reasonable treatment. The benefits of surgery are unknown.

Hot tonsillectomy in quinsy

Quinsy is often a presumptive diagnosis in an ill child with fever, sore throat, and trismus, unable to cooperate with examination. Most settle with antibiotics, rehydration, and analgesia, but a few will require assessment under general anaesthesia. Needle aspiration of the quinsy is unreasonable in this circumstance due to high recurrence rates. Surgical drainage of the abscess is acceptable, but once the abscess is drained, hot tonsillectomy could be performed to prevent recurrence completely. Removing the contralateral tonsil is

more controversial with the increased risk of postoperative haemorrhage for little benefit.

Relevant guidelines

There are no agreed guidelines as such but a suggested algorithm for managing cervical lymphadenopathy in childhood is listed in 'Further reading'.

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82

Paediatric drooling and salivary glands

Anna Harrison and Neil Bateman

Introduction

Drooling, or sialorrhoea, is defined as the spillage of saliva due to either excessive production or decreased swallowing. It could be considered physiological in children up to 4 years and drooling tends to resolve in those without underlying neuromuscular comorbidities by this age. The ability to control saliva develops alongside feeding and other motor skills.

Drooling has significant implications for patient and family and can result in the need to changes clothes repeatedly, damage to computer keyboards, cause irritation to skin, as well as cause significant embarrassment, especially in older children. The inability to manage saliva can result in 'posterior drooling' with the resulting aspiration and the need for regular pharyngeal suction.

Salivary gland diseases, apart from viral conditions, in the paediatric population are rare but often present to an otolaryngologist. Differential diagnosis can include congenital abnormalities, viral and bacterial infections, autoimmune disease, and tumours.

Anatomy and physiology

There are three major paired salivary glands (parotid, submandibular, and sublingual) and up to 1000 minor salivary glands in the oral cavity and pharynx. The largest gland is the parotid, located on the side of the face and upper neck. This pyramid-shaped gland produces watery, serous saliva via a 5–6 cm long excretory duct (Stenson's duct) opening opposite the second upper molar. The facial nerve passes through the gland before dividing into its five main divisions. The submandibular gland lies on the hyoglossus muscle with the duct (Wharton's) opening in the floor of the mouth. It produces thick, mucoid saliva. The lingual and hypoglossal nerves lie in close apposition to the deep surface of the gland. The smallest sublingual glands are located in the sublingual fossa, superior to the mylohyoid muscle. The secretory duct may empty into Wharton's duct or separately into the oral cavity. Up to 1.5 litres of saliva are produced in a day when fully grown. The majority, 70–75%, from the submandibular gland, 20–25% from the parotid, and the remaining from the sublingual glands. Saliva is made up of 99.5% water and the rest is a combination of proteins, electrolytes, and bactericidal/antimicrobial factors. The protein content of saliva is vital for dental caries prophylaxis in children. The mucous component of saliva prevents the oral mucosa from drying out aiding swallowing and chewing. Saliva production is stimulated by parasympathetic muscarinic innervations directly to the major saliva glands.

Drooling

Assessment and history

Drooling-rating systems do exist and have a useful role in assessing the efficacy of interventions. These include the Teacher's Drooling Score and Blasco's scale. The history has an important role in determining the optimal treatment options. The key points are summarized in Box 82.1.

In salivary gland diseases, history should cover onset and location of swelling, associated pain, erythema, exacerbating factors such as chewing/eating, and systemic symptoms. Always remember immunization status, especially measles, mumps, and rubella.

Examination and investigation

The examination of a child with drooling should include the assessment of posture and a general assessment of the child's neurological status, a careful examination of the oral cavity to assess for dental

Box 82.1 History and assessment of drooling: key points

- Assess the frequency and severity of drooling (number of bibs or clothing changes) including situations which increase drooling such as watching television.
- Evidence of chronic aspiration such as coughing/choking at night, recurrent chest infections.
- Symptoms of gastro-oesophageal reflux (severe reflux can cause hyperstimulation, and therefore hypersalivation).
- Feeding, speech, and general development, especially neurological development and posture.
- Review medication. Medication with cholinergic effects such as anticonvulsants may cause hypersalivation.
- Assess the effects of drooling on the child and family including skin changes and embarrassment levels.
- History of dental disease, malocclusion, crossbite, or overbite.
- Previous treatments or investigations.

disease or tonsillar hypertrophy, and palpation of the neck and salivary glands. When considering treatment options, it is often helpful to consider the amount of mouth opening a child has. Children who are gastrostomy fed not infrequently have significant limitations of mouth opening which can render intramural procedures more challenging.

Differential diagnosis

Drooling secondary to impaired swallowing

- *Neurological/muscular*: dysfunction of the oral and pharyngeal phases of swallowing, structural or functional inability to close the lips, impaired coordination, and/or poor posture seen commonly in cerebral palsy.
- *Physiological*: associated with nausea, anticipated pain, and teething.
- *Acute infections*: for example, epiglottitis, tonsillitis, and dental infections.

Hypersalivation secondary to medication

Less often prescribed in children but should be considered. Include antipsychotics such as clozapine, cholinergic agonists, and anticonvulsants.

Treatment/management—medical and surgical

Drooling is best managed using a holistic, stepwise approach. The following is a suggested framework. It is important to have an honest discussion with carers, and patient when appropriate, in order to manage the expectations they may have from treatment.

1. *Review of posture and positioning* in conjunction with physiotherapy/speech and language as appropriate. Adapt posture, especially those in a wheelchair to prevent forward positioning.
2. *Oral awareness and oral motor skills training*: this may reduce drooling by improving jaw stability and closure; increasing tongue mobility, strength, and positioning; improving lip closure, especially during swallowing; and raising awareness of the issue in the patient. This may take some time and is best performed by a therapist with specific expertise and experience in the area, but can have significant benefit in those aware enough to follow instructions.
3. *Orthodontic treatment of malocclusion and caries*: significant dental disease should be addressed. Palatal training devices may increase tongue movement and promote the initiation of swallowing although their use is not commonplace in UK practice.
4. *Pharmacotherapy*: anticholinergic medication can block the parasympathetic stimulation of the glands. Oral glycopyrronium bromide or hyoscine hydrobromide patches are commonly used. Generally, they have good efficacy in many children although their use is not infrequently limited by side effects.
5. *Botulinum toxin*: botulinum toxin inhibits the release of acetylcholine at the presynaptic neurosecretory junctions within the saliva glands. Botox A is available for clinical use. This should be administered under ultrasound guidance either under local or general anaesthetic or sedation. As a general rule, the maximum doses of Botox A per gland could be considered to be 20 units for each submandibular gland and 10 units for each parotid gland. In general, the submandibular glands have the greatest effect on drooling. It is limited by the duration of effect which is generally between 3 and 6 months. The most concerning side effect in the case of children with drooling would be dysphagia which occurs if there is misplacement of the Botox or leakage from the glands. If it occurs, it is generally temporary but can require a period of tube feeding. Botox has limited application in children who require a general anaesthetic for its use as a general anaesthetic every 3 months is, for most children, not an acceptable treatment option.
6. *Surgery*: a variety of surgical options exist depending on the patient and the presence of aspiration after a trial of more conservative measures.
   - *Salivary duct ligation*: two-duct ligation, both submandibular ducts; three-duct ligation, both submandibular ducts and one parotid duct; or four-duct ligation of both submandibular and both parotid ducts. The submandibular ducts can be easily identified in the floor of the mouth and Ligaclips used for ligation. The parotid ducts can be cannulated using a lacrimal probe and an incision made around the duct orifice. The duct can then be dissected out until a clip or tie is placed around it. This offers a simple, safe option to drooling and can be performed as a day case in the majority of children. In a small number of children, gland swelling can occur but is usually managed effectively with antibiotics and observation. In a proportion of children recurrence of drooling may occur.
   - *Salivary duct relocation*: relocation of the submandibular ducts is a well-established surgical treatment for drooling. An island of mucosa incorporating the opening of both submandibular ducts is created and the ducts mobilized. A tunnel is created along the floor of the mouth to the inferior pole of the tonsil and the duct sutured to the inferior pole. This can be performed along with excision of the sublingual glands in an attempt to reduce the incidence of ranula formation. This procedure has demonstrated efficacy but is associated with more dissection in the floor of the mouth and has increased postoperative pain as well as risks of bleeding and swelling. Duct relocation can lead to aspiration and this option would not be suitable for children with impaired swallowing ability or a history of aspiration.
   - *Salivary gland excision*: in patients who aspirate or have significant posterior drooling, bilateral submandibular gland excision offers a surgical alternative. Success rates have been reported of around 60%. Risks of submandibular excision include injury to the marginal mandibular, hypoglossal, and lingual nerves.

Salivary gland disease in children

A detailed clinical history will often guide diagnosis in salivary gland conditions. In the majority of cases the presenting symptom is swelling of one or more salivary gland and it is important to ascertain the duration, progression, and any fluctuation occurring with this. It is also important to obtain a history of the child's general health and any systemic diseases which may impact the diagnosis. A full ear, nose, and throat/head and neck examination is important including

a careful intraoral examination. Always examine and document the status of the facial nerve in those with parotid abnormalities.

Congenital salivary gland disease

Hypoplasia or aplasia of salivary glands is uncommon and can affect one or more of the salivary glands. Congenital salivary cysts may occur in the parotid gland and may be difficult to distinguish from a branchial anomaly. The diagnosis is made by imaging in the form of ultrasound or magnetic resonance imaging (MRI) and treatment options include aspiration sclerotherapy or excision.

Infections

Viral

- *Mumps*: the previously common childhood illness of mumps is caused by a paramyxovirus and characterized by bilateral parotid swelling and fever. The diagnosis is generally clinical but serum immunoglobulin M can be tested (30% false negative). Long-term complications include epididymitis and orchitis with potential subfertility, mumps meningitis, and encephalitis. The treatment is symptomatic.
- *Human immunodeficiency virus* (HIV): the salivary glands may be involved in HIV infection and the diagnosis should be considered in all patients presenting with salivary gland disease. Bilateral cystic and painless enlargement of the parotid glands is pathognomonic for HIV infection. Ultrasound shows typical hypoechoic and anechoic structures representing epithelial cysts and lymph nodes. Diagnosis is via serology and treatment is symptomatic.

Bacterial

- *Neonatal suppurative parotitis*: acute sialadenitis may occur immediately after birth and is associated with *Staphylococcus aureus* infection. It is not uncommonly seen in children on the neonatal intensive care unit when it occurs. It is generally treated with systemic antibiotics and attention paid to maintaining appropriate levels of hydration.
- *Suppurative parotitis*: this is uncommon in children but may present with a painful swollen parotid gland with purulent secretions visible at the orifice of Stenson's duct. It is often associated with significant comorbidities. Treatment is with antibiotics (co-amoxiclav) and imaging such as ultrasound to exclude an associated abscess.
- *Tuberculosis and other mycobacteria*: the lymph nodes surrounding and within the salivary glands may be affected by tuberculosis or non-tuberculous mycobacterial infection (see Chapter 81).

Other granulomatous and inflammatory diseases

- *Cat scratch (Bartonella henselae) disease*: may result from a cat scratch or bite and may occasionally present as parotid or submandibular area swelling near the site of the original inoculation.
- *Sarcoidosis*: sarcoid is rare in children but can present with parotid swelling. The combination of anterior uveitis, parotid gland enlargement, facial palsy, and fever is referred to as *uveoparotid fever* or *Heerfordt syndrome.*

Juvenile recurrent parotitis

This uncommon condition is characterized by episodes of swelling of the parotid gland. The pathophysiology is unknown and does not appear to be infective or obstructive. This may be associated with skin erythema and mild pyrexia. These episodes usually last for 24–48 hours but can persist for as long as 2 weeks. The condition is most commonly unilateral. The typical age of onset is between 3 and 6 years. In between episodes, patients are symptom free. The symptoms resolve in the majority of patients after puberty. In the majority of children the diagnosis can be made clinically. Ultrasound may be used to exclude a mass within the gland but no other investigations are necessary to make the diagnosis.

In the first instance, the treatment is symptomatic with analgesia and anti-inflammatory drugs. Antibiotics are often used empirically. For recurrent episodes, sialendoscopy is a minimally invasive option for treatment. The orifice is usually easily cannulated and the ducts often have a pale, featureless appearance. Irrigation of the gland with saline is easily facilitated during the procedure and installation of steroids in the form of triamcinolone 40 mg has been reported by multiple authors to result in resolution of the episodes or an episode-free period. It is very rare to have to consider parotidectomy in these patients and this should be considered a last resort.

Salivary gland stones

Salivary gland stones are uncommon in children but their presentation mimics that in adults which will be familiar to most otolaryngologists and they typically present with gland swelling on salivation. They are more common in the submandibular gland. In children they are typically located in the distal submandibular duct and are therefore usually amenable to intraoral removal. Sialendoscopy is now well established as a minimally invasive approach to manage paediatric salivary gland stones. Where this is technically not possible (for instance, with large impacted stones) excision of the submandibular gland may be required (**Figure 82.1**).

Mucus retention cysts

Mucus retention cysts are true epithelial-lined cysts usually involving the minor salivary glands and are commonly seen on the lips, buccal

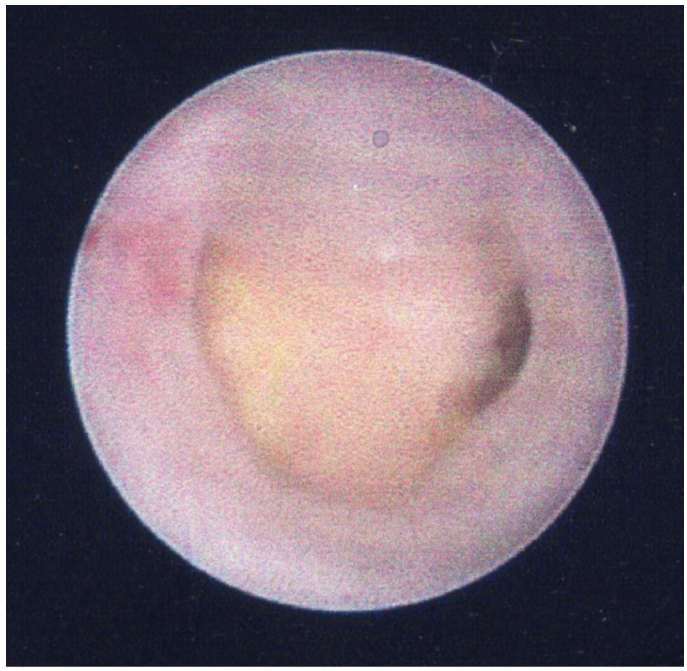

Figure 82.1 Sialendoscopic picture of a salivary gland stone.

mucosa, and tongue. They are diagnosed clinically and the treatment consists of surgical removal when they are problematic.

Ranula

Ranulas are extravasation mucoceles, arising from the sublingual gland. As such, they are said to lack an epithelial lining and represent 'pseudocysts'. They typically present as a bluish swelling in the floor of mouth, from which they derive their name. Other causes of ranula formation include scar tissue around the duct secondary to surgery, chronic inflammation, or infection. 'Plunging' ranulas present as neck masses as the ranula expands inferiorly, through the mylohyoid muscle, into the neck. Definitive treatment involves excision of the sublingual gland, and excision or marsupialization of the ranula. Exploration of the neck is rarely necessary.

Tumours

Tumours of the salivary glands can be divided into vasoformative tumours, neoplasms of salivary gland origin and tumours arising from perisalivary lymphoid tissue, such as lymphoma.

Haemangioma

Infantile haemangioma is the most prevalent benign tumour of the parotid gland, accounting for 90% of salivary gland lesions that occur in children before 1 year of age. About a third are present at birth. They are composed of vascular endothelium, which undergoes a rapid growth phase; they proliferate rapidly during the first 1–2 months of life, and often a second growth spurt occurs between 4 and 6 months of age, followed by a slow spontaneous involution, which occurs gradually over several years and is generally complete by the end of the first decade. The diagnosis should be made clinically with reference to imaging, most commonly in the form of MRI scanning (Figure 82.2). Treatment decisions should be made in the context of the knowledge that spontaneous resolution without long-term sequelae is the norm. With this in mind, where a decision has been made to pursue active treatment, propranolol therapy is well recognized as being effective in the majority of children (see Chapter 81). Surgery should be reserved for cases where the diagnosis is in doubt or when there is a persisting need for active intervention when propranolol has been ineffective.

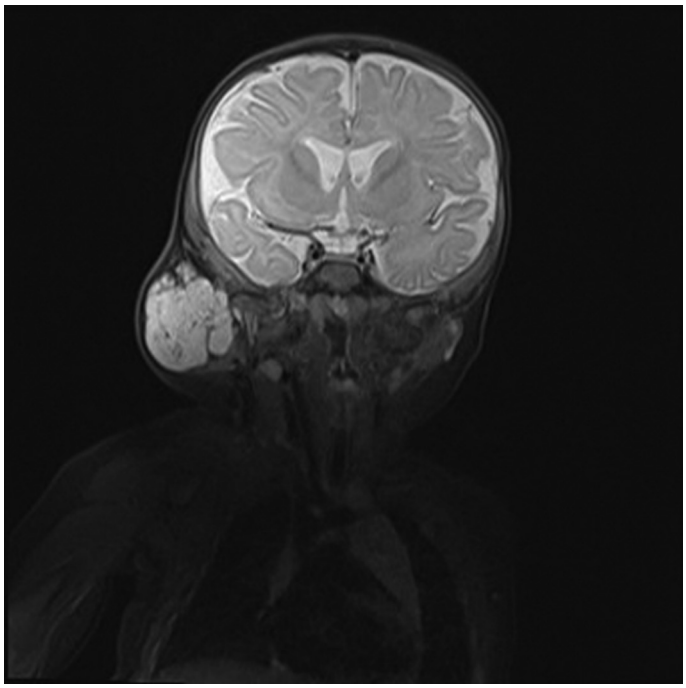

Figure 82.2 MRI of a parotid haemangioma.

Tumours of salivary origin

Salivary gland neoplasms in children are uncommon. The majority are epithelial (95%), with the remaining being mesenchymal. Compared with adults, the proportion of malignancies among all salivary gland neoplasms is higher in children, although the majority are still benign and the commonest tumour is a pleomorphic salivary adenoma. The commonest malignant tumour of salivary origin is mucoepidermoid carcinoma. A high index of suspicion is required when presented with a child with a solid mass in the salivary gland and they should all be treated as potentially malignant. An ultrasound scan is easily obtainable and can distinguish between cystic and solid lesions. MRI scanning can determine the extent of the lesion. Fine needle aspiration cytology is less well established in paediatric practice compared to adults as it is more difficult to interpret and it is often necessary for children to have general anaesthetic for this. Often, therefore, it is necessary to proceed directly to a surgical excision.

The management of benign salivary tumours in children is analogous to their management in adults (see Chapter 58). Malignancies are similarly managed with a multidisciplinary team approach.

Lymphoma

Lymphoma of the parotid gland often manifests as a painless progressively enlarging mass that is rarely suspected before biopsy or surgical removal. It can manifest as focal or diffuse, single or multiple, and unilateral or bilateral masses.

Immune conditions

Juvenile Sjögren's syndrome

Autoimmune disease characterized by progressive lymphocytic and plasmacytic infiltration, mainly of the salivary glands and lacrimal glands. In children parotid swelling seems to predominate, with less frequent oral and ocular symptoms. The typical diagnostic criteria are less relevant in children and the diagnosis should be considered in recurrent sialadenitis especially with a history of conjunctivitis. Treatment is based on symptomatic relief with salivary substitutes and regular dental checks and management should be in conjunction with a paediatric rheumatologist.

Surgical techniques

Parotidectomy

Parotidectomy in children is generally indicated for the excision of solid tumours. The facial nerve in children is more lateral and more superficial than in adults. These anatomical differences must be noted to avoid injury to the nerve. Facial nerve monitoring and magnification should be used. The posterior belly of the digastric muscle provides a reliable landmark for identification of the facial nerve with the nerve emerging just deep to the muscle at the point

where the superior margin of the muscle meets the mastoid process. Parotid surgery in children is uncommonly performed and should only be performed by surgeons with significant training and experience.

Sialendoscopy

Sialendoscopy allows the visualization of the ductal anatomy as well as the ability to remove stones and carry out ductal dilatation. In adults, the commonest indication is sialolithiasis, whereas in children, the commonest indication is juvenile recurrent parotitis. In children, the overwhelming majority require a general anaesthetic for the procedure.

Controversies

The treatment of drooling in an otherwise developmentally normal child is controversial. While drooling can be socially embarrassing, the treatment options, especially surgery, may cause more harm than good and the risks must be fully justified. While the stepwise management is typically agreed throughout the ear, nose, and throat community, when it comes to the choice of surgical procedure and the use of botulinum toxin, practice may still vary between clinicians.

Tips and pearls

- Drooling is common in children under the age of 2 years, and usually resolves before the age of 5 years without intervention.
- Sialorrhea is seen mainly in patients with neurological disorders and is mostly caused by an inability to control oral secretions rather than an overproduction of saliva.
- Treatment options for sialorrhea include rehabilitation (positioning, oral motor, behaviour therapy), medical treatment (anticholinergics, botulinum toxin injection), and/or surgery (submandibular gland excision, duct ligation or relocation).
- Salivary gland swelling in children is often inflammatory in origin.
- Salivary gland malignancy is rare, but solid salivary masses in children are more likely to be malignant than in an adult and all should be treated as potentially malignant.
- Sialoendoscopy is a minimally invasive and effective tool for the diagnosis of salivary gland ductal pathology (inflammation, stenosis, stricture) and for the treatment of sialolithiasis and juvenile recurrent parotitis.

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83

Paediatric hearing loss

Jaya Nichani, Adam Walker, and Iain A. Bruce

Introduction

Hearing impairment in children has been classified by the World Health Organization as hearing thresholds worse than 25 dB in the better hearing ear. Different classifications of severity exist, such as *mild* (26–40 dB), *moderate* (41–60 dB), *severe* (61–80 dB), and *profound* (>81 dB). With the introduction of newborn hearing screening programmes, infants with a permanent moderate hearing loss can be identified within 4–5 weeks of birth. The prevalence of hearing loss of moderate grade or worse has been reported as 1.1/1000 at birth, increasing to 3.5/1000 in adolescents. This increased prevalence rate in older children reflects the progression of mild hearing loss over time, late-onset genetic causes, and acquired causes. Temporary hearing loss, however, has a higher incidence and prevalence in childhood and can have a significant impact on speech and language development. The most common cause of temporary hearing loss in children is otitis media with effusion (OME) (see Chapter 88). OME affects 20% of 2-year-olds and has a cumulative incidence of 80% in the first 8 years of life. It is associated with mild to moderate hearing loss. Most children will have a fluctuating hearing loss, with treatment being reserved for children with speech and language delay, with a persistent hearing loss, those children with associated comorbidities, and when recurrent ear infections are associated with OME.

Timely management of permanent bilateral congenital hearing loss of moderate severity or worse is important in preventing the negative impact upon development of a child. Management strategies for children with a congenital hearing loss will depend on the severity of the hearing loss, age at diagnosis, if it is asymmetrical or symmetrical, and the nature (conductive, sensorineural, or mixed) of the hearing loss. It is well documented that the earlier a child is given access to speech and language, the greater their potential for development of receptive and expressive language. Due to neural plasticity, the 'cut-off' period for speech and language development is considered to be around 4 years of age. *Neural plasticity* refers to the neural connections and networks that develop in the brain in response to auditory stimulation and access to speech and language. A lack of access to speech and language beyond this age will severely impact the child's ability to develop spoken language. Inadequate or lack of stimulation for prolonged periods will hinder the development of these connections and eventually these areas will be used by other senses which are intact. This will eventually prevent any meaningful benefit from delayed hearing (re)habilitation.

Anatomy and embryology of the ear in infants and children

The ear is divided into three parts, with each associated with a particular type of hearing loss. Pathology affecting the outer and middle ear will result in a conductive hearing loss (CHL) and that affecting the inner ear will result in a sensorineural hearing loss (SNHL).

Each part of the ear has a distinct embryological origin. The outer and middle ear develop from the first and second branchial arches at 5 weeks of gestation. The pinna develops from the first and second branchial arch from the six nodular swellings called *hillocks of His*. The external auditory canal is formed from the first pharyngeal cleft while the first pharyngeal pouch contributes to the development of the Eustachian tube and the middle ear. The malleus and incus arise from the first pharyngeal arch and the stapes superstructure from the second arch. The otic placode, which develops from the ectodermal cells of the neural plate, forms the inner ear. The otic placode forms the otic vesicle which develops into the cochlea and the semicircular canals. The vestibulocochlear–facial ganglion complex originates from the hindbrain which lies close to the developing otic vesicle. In a normally developed ear, the position of the facial nerve at birth is almost similar to the adult anatomy except for its position at the more superficially located stylomastoid foramen, which results from mastoid process underdevelopment. This needs to be taken into account when performing ear surgery in an infant.

Abnormalities affecting the embryogenesis of the vestibulocochlear–facial nerve complex are associated with the development of the otic vesicle. The facial nerve embryogenesis and development occurs concurrent to the embryogenesis of the ear, hence the point at which a defect occurs in the gestational period will result in the eventual abnormality that manifests in the neonatal period. Most infants born with a permanent hearing loss will have normal embryogenesis. However, infants with microtia and ear canal abnormalities will have a distinct CHL with normal sensorineural hearing thresholds due to the nature of embryogenesis.

Clinical assessment

Examination of the ears can be challenging in children, particularly in the very young and those with neurodevelopmental disorders. That notwithstanding, the clinician can acquire important information on inspection prior to physical examination. Dysmorphism should be documented with particular attention to the position, shape, and development of the pinnae. Syndromes such as Down syndrome and CHARGE (Coloboma, Heart defects, Atresia choanae, Retardation of growth, Genito-renal defects, Ear anomalies/deafness) are associated with characteristic appearances of the *external ears*. Although the majority of preauricular sinuses (**Figure 83.1**) are isolated developmental anomalies of the hillocks of His, they should prompt examination of the neck for branchial anomalies and inquiry about coexisting renal disease that might suggest the diagnosis of branchio-oto-renal (BOR) syndrome. Preauricular appendages should be documented. Syndromes such as Goldenhar, CHARGE, Down, and Treacher Collins are associated with characteristic anomalies of the *eyes and eyelids*. Examination of the *hands* may demonstrate specific digital anomalies that in combination with an abnormally shaped head may suggest a craniosynostosis. During inspection of the *face*, particular attention should be given to identifying facial asymmetry and macrostomia (Goldenhar syndrome, also known as oculo-auriculo-vertebral spectrum), malar hypoplasia and micrognathia (Treacher Collins), and facial movements. Facial nerve examination may be limited due to the child's inability to cooperate with formal examination, and in such circumstances it is important to provide a narrative description of facial movements (e.g. at rest and when crying or smiling) and adequacy of eye lid closure. CHARGE syndrome can be associated with facial nerve palsy and various middle and inner ear abnormalities. This description is not exhaustive, and clinicians should be aware that syndromic deafness may be sensorineural, conductive, or mixed type, and can be caused by numerous recognized *chromosomal* (e.g. Down syndrome, Turner syndrome, and 22q11.2 copy number variations—both microdeletion (Di George syndrome) and microduplication) and *single gene* disorders (e.g. Noonan, Stickler, BOR, Treacher Collins, and CHARGE syndromes). A referral to a clinical geneticist is mandatory whenever a syndromic cause is suspected (see Chapter 90).

Figure 83.1 Left preauricular sinus with associated skin inflammation.

Otoscopy may be limited due to the size of the external auditory canal, wax, and the presence of canal atresia. The appearance and health of the *external auditory canal* and *tympanic membrane* should be documented, with particular reference to OME as the commonest cause of hearing loss in childhood. Retractions and perforations should be described, and an attempt made to grade the severity (**Figure 83.2**). Cholesteatoma in children is often found in the posterosuperior quadrant of the pars tensa. When available, digital images and/or video of the tympanic membrane should be taken. The *anterior nasal space* should be examined for signs of rhinitis and the *oral cavity* for palatal anomalies, including bifidity of the uvula which may be associated with submucous cleft palate. Although the aetiology of OME is multifactorial, Eustachian tube dysfunction and/or immaturity is an important causal factor. Palatal anomalies are associated with Eustachian tube dysfunction and may indicate increased risk of the development and persistence of OME (see Chapter 88).

Age-appropriate hearing assessment

The accuracy of any hearing assessment is dependent upon the suitability of the test for the age and development of the child. The most suitable method of assessment is referred to as 'age-appropriate hearing assessment'; therefore, the 'appropriate' method of assessment will differ from child to child and cannot be determined solely from chronological age.

Electrophysiological

In the infant and certain other populations, the most suitable method of assessment is via 'electrophysiological' measurements that do not require the subject to physically respond when a sound is heard. The most commonly used electrophysiological measurements include auditory brainstem response (ABR), cortical audiological evoked potential (CAEP) and otoacoustic emissions (OAEs) (see Chapter 5).

Auditory brainstem response

An ABR is a series of several 'voltage deflections' caused by responses from several sites along the auditory response pathway, from the VIIIth cranial nerve to the brainstem. These deflections create a characteristic 'pattern' to the ABR trace, and their presence can be used as evidence of a response to an auditory stimulus. The stimulus can be delivered using either headphones or a bone vibrator, meaning that an accurate and detailed assessment of hearing is possible in very young children. In the newborn population, ABR is usually performed in natural sleep, though in some cases (such as older children that are unable to perform behavioural assessment) it can be performed under sedation or general anaesthetic. The ABR is recorded by attaching recording electrodes to the patient's skin: one

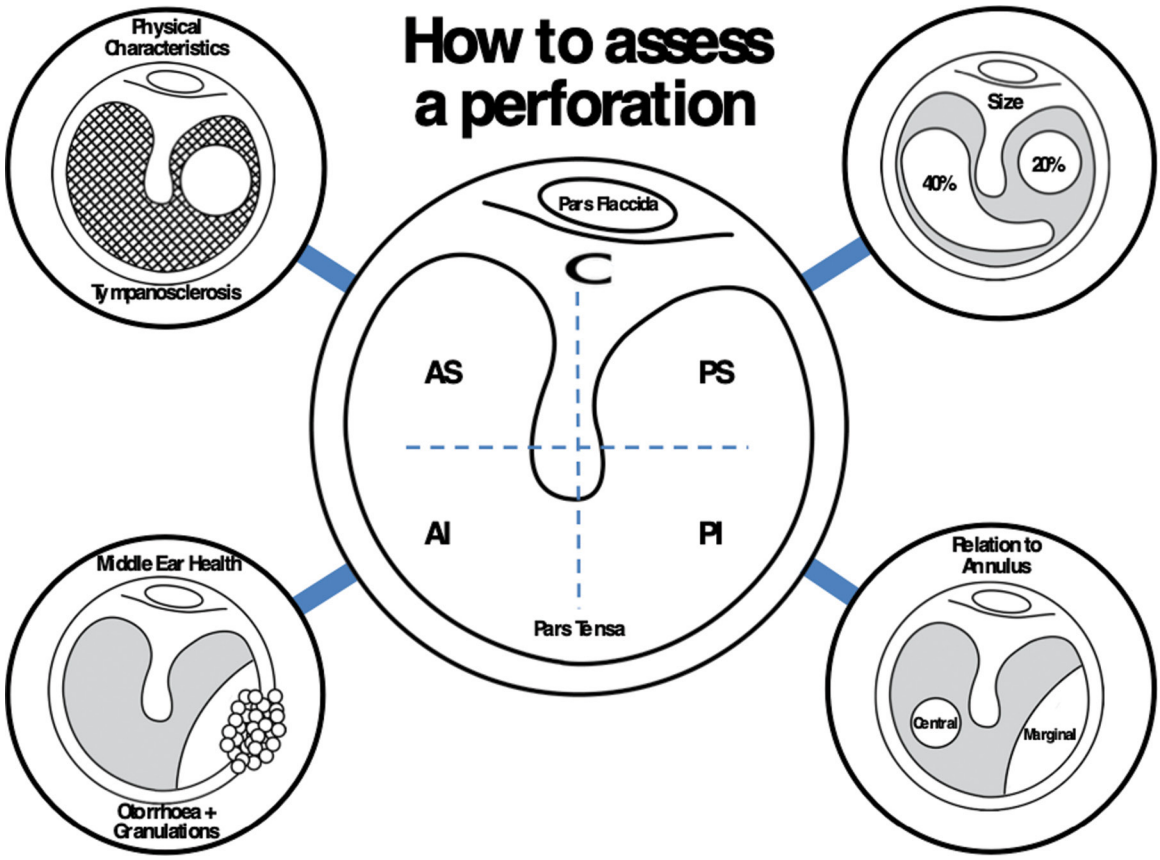

Figure 83.2 How to assess a perforation. Quadrants of the pars tensa: anterosuperior (AS), anteroinferior (AI), posterosuperior (PS), posteroinferior (PI).

on each mastoid, and one on the high forehead (while taking care to avoid the fontanelle in newborns). Frequency-specific auditory stimuli are then delivered to the ear using either headphones or a bone vibrator and the response to each of the stimuli is 'averaged' to create the waveform for each stimulus run (which will ideally contain around 3000 individual responses). Averaging of the responses, combined with replication of traces, is vital to allow for the cancellation of noise, and for ensuring a high reproducibility of any response present. The clinician will then determine whether there is sufficient evidence of an auditory response in the final trace before changing frequency, intensity, or transducer as they deem appropriate.

Cortical auditory evoked potential

The CAEP is a similar response to the ABR but with a later latency which is a result of its generation site further along the auditory pathway, in the auditory cortex. The CAEP matures with age and can therefore be used as a reasonably accurate method of assessing auditory thresholds in the adult population when pure tone audiometry is not possible or reliable. Aided CAEP can be used as a method of assessing access to speech (either aided or unaided) in young children.

Otoacoustic emissions

OAEs are low-intensity 'sounds' in the ear canal which are a by-product of sensory activity in the inner ear. OAEs originate in the outer hair cells of the cochlea and are transmitted through the middle ear to the ear canal. Recordable OAEs are seen as an indication of 'satisfactory' outer hair cell function, but are *not* a test of hearing levels. There are two main types of OAEs that are measured:

Transient evoked otoacoustic emissions (TEOAEs)

TEOAEs are elicited by a click stimulus presented via a probe that is placed into the ear canal. As in ABR, multiple stimuli are delivered via the ear canal and the response is averaged to allow for the cancellation of noise, to ensure a high reproducibility of any response present. The use of a click stimulus means that most of the stimulus energy is within the frequency range of 1.5–4 kHz. The 'response' is analysed to ensure it is more than 6 dB greater than the level of background noise at a set number of frequencies in order to elicit a 'pass'. TEOAEs are the first method of assessment in many newborn hearing screening programmes.

Distortion product otoacoustic emissions (DPOAEs)

DPOAEs are elicited by simultaneous presentation of two pure tone stimuli (referred to as f1 and f2) that are a fixed frequency ratio apart, with f2 being the main 'test' frequency. The ratio of frequency separation means that the two tones are relatively close together in frequency, and so the combination of both tones produces distortion at various sites along the cochlea during presentation. The largest of these 'distortion products' can be determined via the mathematical equation $2f1 - f2$, and as such will be relatively close in frequency to f2. Therefore, the presence of DPOAEs is seen as an indication of satisfactory cochlear function at the frequency of f2. DPOAEs

are much more frequency specific than TEOAEs and results can be plotted and compared to thresholds measured via an audiogram.

Behavioural

Distraction test

Historically, in the UK the distraction test was performed by health visitors at the child's '2-year check' appointment. This practice was stopped as a result of the advent of newborn hearing screening which brought a higher specificity and sensitivity of testing and resultant earlier diagnosis of hearing loss. Distraction testing has been largely superseded by visual reinforcement audiometry (VRA), but can be used to obtain some information when other methods have failed. Distraction testing should be viewed as a measure of hearing sensitivity, rather than absolute hearing thresholds. A distraction test is performed using 'soundfield' stimuli (i.e. stimuli are not presented via headphones) and so does not provide ear specific information. The stimuli used may be pure/warble tones (presented from a handheld warbler), 'musical' (e.g. chime bars or a high-frequency rattle), or live voice (e.g. an 's' or 'humming' sound, to represent high and low frequencies, respectively). The level of frequency specificity is dependent on the stimuli chosen and its method of presentation by the tester. To perform a distraction test requires two testers, one to present the stimuli and the second to 'distract' the child, who is sat on a parent's knee or in a highchair facing the second tester. The second tester will engage the child in low-level play, such as with a spinning toy, and then phase down/stop the play as tester one is about to present the stimulus. The premise of the test is that the child will orientate to the stimulus by turning to look, at which point tester one will 'reward' the child. This can be repeated using different stimuli/levels as required. The act of presenting the stimulus is not without error, hence the reason VRA is a more favoured assessment method, as the child may gain cues that the stimulus is about to be presented from several sources during the test technique, thus leading to false-positive results. Distraction testing can be performed as soon as the child has sufficient head control and is able to independently turn (~7 months). The upper age limit for distraction testing is around 15 months of age, though it can be used for older children with additional needs.

Visual reinforcement audiometry

The principle of VRA is based on orientation to auditory stimuli, as in distraction testing; however, the premise of VRA is to attempt to 'condition' the child to the reward. The test setup is similar to distraction testing in that tester one will present the sounds (usually from an adjoining 'observation room') while tester two engages the child in low-level play with the child sat on a parent's knee or in a highchair facing the second tester. The difference between VRA and distraction testing is that the play remains continuous and is not stopped or phased down prior to the stimulus presentation. Initially the stimulus and reward are presented together, and the reward pointed out to the child in order to gain their attention and help them link the sound to the reward. The rewards in question can be animated toys, flashing lights, or animations on a screen. Once the child has 'conditioned' and linked the presentation of the stimuli to the activation of the reward then the stimuli can be presented alone, a turn towards the reward signifies that the child has heard the sound and then the intensity of the sound can be decreased. Using this stepwise manner, accurate hearing thresholds can be obtained at multiple frequencies. There is a risk of false-positive 'turns' to the sound and testers should be experienced in order to perform the test accurately. Stimuli are, in most cases, warble tones which are highly frequency specific. Stimuli can be presented in the soundfield (via a loudspeaker and so not ear specific) or via headphones, including via a bone conduction headband to provide more accurate information regarding hearing levels and type of hearing loss. As in distraction testing, the child needs to be old enough to have independent head control but not old enough that they are too engrossed in the play of the second tester. The optimal age range for VRA is 8 months to just over 2 years of age, although testing becomes increasingly difficult as the child approaches 2 years of age.

Play audiometry

Play audiometry becomes the most appropriate test from the age of 2 years upwards, although in practice there is a 'grey area' around 24–30 months in which some children may favour VRA and some play audiometry. The principle of play audiometry is more akin to 'regular' pure tone audiometry as performed in adults; however, rather than pressing a button, the child is conditioned to perform a game-like task (such as placing a wooden ball on a stick or a building a tower brick by brick) when they hear the stimulus. Therefore, the child is actively 'listening' for the stimulus rather than having their play interrupted as is the case in VRA. Play audiometry can be performed by one tester alone or by two testers in a similar set up to VRA. Again, the child is 'conditioned' to the task by presenting the stimulus and prompting the child to perform the action simultaneously. Once the tester is happy that the child is conditioned then the intensity and frequency can be adjusted to measure the child's hearing thresholds. As in VRA, stimuli are frequency-specific warble tones presented in the soundfield or via transducers such as headphones or a bone conductor.

Permanent childhood hearing impairment

Paediatric childhood hearing impairment may be congenital or acquired. Acquired hearing loss may be due to infection, ototoxic drug use, trauma, or be idiopathic.

Congenital hearing loss

Congenital hearing loss is conductive if the pathology affects the outer and middle ear. This is often as a result of microtia, with or without canal atresia, canal stenosis, or ossicular fixation. Congenital CHLs are often sporadic pathologies, but bilateral and certain unilateral CHLs may be associated with identified syndromes, including Treacher Collins syndrome, Goldenhar syndrome (**Figure 83.3**), BOR syndrome, CHARGE syndrome (**Figure 83.4**), and trisomy 21 (see Chapter 90).

Congenital SNHL may be genetic or acquired. Genetic SNHL may be non-syndromic or syndromic. Non-syndromic congenital SNHL is associated with a mutation in the gene coding for the portion of connexin 26 protein in up to 80% of the patients. Common syndromic forms may be autosomal recessive in nature (Usher syndrome, Pendred syndrome, Jervell and Lange-Nielsen syndrome), autosomal dominant (Waardenburg syndrome, Stickler syndrome, BOR syndrome, neurofibromatosis type 2), or X-linked (Alport syndrome, oto-palato-digital syndrome).

Figure 83.3 Dysplastic pinna and preauricular appendages in a child with Goldenhar syndrome.

Congenital infections are the most common causes for acquired congenital hearing losses. The most common infection is cytomegalovirus infection, with other implicated infectious agents including *Toxoplasma gondii*, rubella, and herpes simplex viruses.

Figure 83.4 Dysplastic right pinna in CHARGE syndrome, demonstrating the characteristic anomalous development of the antihelix.

Acquired hearing loss

Infections in the postnatal period can also cause hearing losses. Localized ear infections may result in permanent SNHL or a CHL due to a tympanic membrane perforation or ossicular erosion. Systemic infections which result in a SNHL may be caused by pathogens such as bacteria (meningococcus, pneumococcus), viruses (including herpes simplex viruses, herpes zoster, HIV, rubella, and mumps), or parasites (schistosomiasis, malaria). Hearing loss may be due to the ototoxic effects of toxins, infection-induced neuropathy, or cochlear fibrosis and ossification. Cochlear obliteration (fibrosis or ossification) may be seen in children with meningitis. Early identification is crucial as continued obliteration may prevent the use of cochlear implantation in those with severe-to-profound hearing loss.

Ototoxic drugs are another cause of permanent childhood hearing impairment. Drugs implicated in this process are aminoglycoside and macrolide antibiotics, quinine, certain non-steroidal anti-inflammatory drugs, loop diuretics, certain antimalarial drugs, and platinum-based chemotherapy drugs. Platinum-based chemotherapy drugs cause irreversible hearing loss and regular monitoring is needed to identify developing hearing loss and when to consider discontinuation of this group of chemotherapeutic agents. In other ototoxic drugs the ototoxic effect may be reversed if it is identified early and the treatment discontinued. Serum peak and trough levels need to be monitored when using most of these drugs to ensure safe serum levels are maintained to prevent ototoxicity. However, certain individuals have a genetic susceptibility to ototoxicity and need regular hearing assessment in addition to the monitoring of blood levels.

Broadly, traumatic hearing loss in childhood occurs either due to the severity and nature of the forces exerted, or as a consequence of increased susceptibility to damage associated with certain congenital inner ear abnormalities. Hearing loss due to noise exposure or barotrauma is less common in children as compared to adults, although there is some concern regarding the potential negative impact of the long-term use of personal over- or in-ear headphones at raised volumes. Hearing loss secondary to physical trauma may go unnoticed due to the over-riding need to manage any associated brain injury or cranial fracture. However, it is important to consider hearing loss after a head injury in a child, especially when there is radiological confirmation or clinical suspicion of a base of skull or temporal bone fracture. Wide vestibular aqueduct (WVA or enlarged vestibular aqueduct) is the commonest inner ear abnormality associated with SNHL. It is characterized by an abnormally wide connection between the endolymphatic sac and the vestibule of the inner ear. Although, the exact mechanism for inner ear damage is not fully understood, WVA is often associated with a 'stepwise' reduction in hearing thresholds, which may be attributable to known episodes of relatively minor head trauma or certain physical activities (e.g. trampolining). Vertigo may be associated with hearing loss after head trauma in affected children. Advice regarding avoidance of certain physical activities in children with WVA will remain controversial until more is known about the pathogenesis of the inner ear damage. The severity and timing of hearing loss associated with WVA varies considerably, with some infants and young children diagnosed following magnetic resonance imaging, mandated as part of the aetiological investigations undertaken after the identification of permanent childhood hearing impairment at newborn hearing screening.

Multidisciplinary management of childhood hearing loss

The downstream consequences of hearing loss mandate that a multidisciplinary approach is often needed to limit the consequences upon child development. The potential negative impact upon speech and language development, communication, educational performance, and psychosocial development necessitates that paediatric hearing services should have access to audiology, speech therapy, specialist teaching, and psychology professionals. Multidisciplinary working is central to *hearing implant* (cochlear implant and bone conduction hearing devices) and *microtia and ear reconstruction* services. Where available, guidelines should be used to structure the multidisciplinary discussion, such that parents are reassured that the recommendation is individualized, coherent, and based on best evidence. For example, decisions regarding hearing implants in children with congenital canal atresia must be made collectively, to ensure that the choice and positioning of a bone conduction implant does not prohibit autologous ear reconstruction at a later date.

Management options

Listening tactics

Any child, but particularly those with hearing loss (whether aided or unaided), will benefit from the employment of *listening tactics*. Listening tactics are methods of increasing the 'signal-to-noise ratio', which is the level of the input (e.g. teacher's voice) above the level of background noise. In a classroom environment, the signal-to-noise ratio can easily be reduced via a number of simple strategies such as sitting the child closer to the teacher and making sure that they can see the teacher's face (to aid with visual clues/lip reading). In the home environment this can be achieved by making sure the background noise is kept to a minimum while conversing with the child or ensuring that people take turns to speak around the dinner table for example. 'Assistive listening devices' can also help reduce the signal-to-noise ratio by 'artificially' bringing the sound signal closer to the listener. Assistive listening devices work by receiving the signal from the speaker (often by a microphone worn around the neck or on a lapel) and transmitting to a receiver which can be attached to a hearing aid (for children with hearing loss), or worn independently of a hearing aid (for children that have significantly difficulty hearing in background noise).

Conventional hearing aids

Children with a hearing loss are usually first provided with a behind-the-ear (BTE) hearing aid. In a small number of clinical situations, a BTE hearing aid may not be suitable, and children may be initially fitted with a bone conduction hearing aid (non-surgical). BTE hearing aids are worn behind the pinna and are connected to an 'earmould' that sits in the child's ear. In young children, earmoulds are made from a soft silicone material for comfort and to reduce potential damage to the ear from bangs to the ear. As children get older, the earmould can be changed to a hard acrylic or even a 'slim-tube' for comfort or aesthetics. In simple terms, a hearing aid detects the incoming signal via a microphone; this converts the sound to electricity. This is then made 'louder' by an amplifier, before the amplified electrical signal is then converted back to sound by a receiver. It is important to note that modern hearing aids do not simply make all sounds louder, but rather they amplify the sound in accordance with the level and configuration of hearing loss as well as the original intensity of the input. On the whole, hearing aids are well tolerated in young children given the wide range of colours available; in older children the preference often changes towards a desire for more discrete styles of device and configuration.

Bone conduction hearing devices

Bone conduction hearing devices (see Chapter 16) are reserved for children who are unable to wear conventional BTE hearing aids. There are three main indications: (1) chronic ear conditions with ear discharge, where a BTE hearing aid aggravates the discharge (e.g. chronic suppurative otitis media and otitis externa); (2) anatomical considerations (stenotic/atretic ear canals and microtia) that make fitting BTE hearing aids difficult; and (3) unilateral SNHL. Suitability and candidacy for these devices should be assessed by a multidisciplinary team with active involvement of the parent(s)/carer(s) and the child when possible. This decision will be based on multiple factors which include individual patient factors, ear health, and hearing considerations.

Factors influencing decision-making in each of these scenarios will include age of the child, associated comorbidities, type of hearing loss, bone conduction thresholds, parent/patient willingness to undergo surgery, and individual surgeon preference.

Options available include *non-surgical* devices which may be worn on a band or with an adhesive device. These are best suited for children who are young, those that prefer a non-surgical option, where hearing loss is likely to be temporary, and where surgery is contraindicated. Surgical options include active and passive devices which may be trans- or percutaneous. With the continuing evolution in the technology, the future is promising for transcutaneous active devices which are likely to provide maximum benefit and wearability.

Cochlear implantation

Cochlear implantation is fully established as a treatment option for severe-to-profound hearing loss in children (see Chapter 16). Auditory plasticity dictates that prelingually deafened children should be implanted before 4.5 years of age, with emerging evidence that 'early' implantation in late infancy gives better outcomes. Postlingually deafened children and young people, along with those whose hearing loss is progressive, may be suitable candidates for implantation providing there is evidence of auditory learning reflecting compliance with conventional hearing aids or a preceding period of better hearing.

Pivotal to the counselling of parents and determining candidacy for cochlear implantation is the multidisciplinary team approach to shared decision-making. Assessment can be particularly challenging in children with borderline hearing thresholds, cognitive impairment, and the autistic and auditory neuropathy spectrum disorders. When measuring benefit following cochlear implantation in children with additional needs, care must be taken to contextualize performance to the underlying condition and to avoid placing undue reliance on outcome measurement instruments designed specifically for children who are otherwise developing normally.

Initially, technological advances and greater understanding of programming and (re)habilitation led to an evolution in the audiological

criteria to include individuals with potentially useable residual hearing and/or evidence of benefit from conventional hearing aids. The subsequent development of 'soft surgery' techniques for insertion of the electrode array has enabled cochlear implantation candidacy to be extended further to include those with normal or near-normal residual low-frequency hearing (hearing preservation).

While the success of attempted hearing preservation surgery has improved, issues remain regarding the longer-term maintenance of natural residual hearing after CI. This has led to the ongoing development of technologies to deliver 'drugs, cells, and genes' to the inner ear, with the aim of preserving and ultimately multiplying the population of residual sensory cells.

84

The painful and discharging ear in children

Ricky Pal and Mark Felton

Introduction

This chapter discusses the aetiology, diagnosis, and management of the painful and discharging ear in children. There are a range of causes of paediatric otorrhoea and otalgia, with significant overlap with the adult chapters on cholesteatoma (see Chapter 19), tympanic membrane perforation (see Chapter 22), and otitis externa (see Chapter 8). In this chapter the focus will be on the specifics of paediatric pathology, namely acute otitis media (AOM), recurrent acute otitis media (rAOM), and congenital cholesteatoma, and the decision-making process and divergence from adult practice when managing tympanic membrane perforation and cholesteatoma in children.

Basic science: anatomy and physiology

The anatomy of the ear canal, tympanic membrane, and middle ear have been covered in previous chapters (see Chapters 5 and 83). In children, there is a greater degree of Eustachian tube (ET) dysfunction related to a more horizontally orientated ET, adenoidal obstruction in the nasopharynx, and inflammation of the ciliated columnar epithelium, commonly related to virally mediated upper respiratory tract infection.

Assessment

Initial assessment of the discharging or painful ear is with clinical examination, focusing on otoscopic or microscopic evaluation of the external auditory canal and tympanic membrane. Facial nerve function should be assessed. Causes for referred otalgia should also be sought if ear examination is normal. When otorrhoea obscures an assessment of the tympanic membrane, aural mopping or microsuction should be undertaken to clear the discharge. On occasion, examination under general anaesthesia is required to facilitate this in children with chronic otorrhoea that fails to respond to medical treatment, or if an obstructing aural polyp is present.

Age-appropriate hearing assessment and tympanometry should be undertaken where there is clinical concern for hearing and prior to surgical intervention. Details of age-appropriate assessment are covered in Chapter 83 on clinical and audiological assessment of hearing and paediatric hearing loss.

Investigations

AOM is a clinical diagnosis and acute investigations are rarely indicated. When intratemporal or intracranial complications of AOM are suspected, appropriate imaging is indicated, most commonly a contrast-enhanced computed tomography (CT) scan of the brain and temporal bone. Further intracranial assessment may be undertaken with magnetic resonance imaging. This is discussed in further detail later in the chapter.

Classifications

There are a variety of terminologies used to describe middle ear disease and otitis media. This chapter divides these into acute versus chronic otitis media and non-suppurative versus suppurative, and focuses on suppurative disease manifesting with a painful or discharging ear. Chronic suppurative otitis media may further be divided into mucosal versus squamous disease, which may be active or inactive.

Differential diagnosis

Paediatric otalgia and otorrhoea may manifest acutely and resolve or persist and become chronic, with symptoms lasting more than 6 weeks.

Acute otitis media

Presentation and diagnosis

The most common cause of an acutely painful or discharging ear in children under the age of 5 years is AOM. The incidence corresponds with seasonal increase in winter months of viral upper respiratory tract infections, and decreases with increasing age, being most common in children under 2 years of age. A small subset of children will develop rAOM. Rarely, following an episode of AOM, a child may present with a complication of AOM.

AOM is defined as the first 3 weeks of acute inflammation of the middle ear space. It typically occurs in a child under the age

of 6 years with a reported incidence of 256/1000. Presentation varies with age, with irritability, pyrexia, otalgia, ear tugging, and hearing loss. In cases of acute tympanic membrane perforation, a mucopurulent discharge and exudate is seen. Diagnosis is based on clinical examination with otoscopy and further investigations, such as tympanometry or imaging are rarely indicated. Guidelines for initial management are numerous. Diagnostic action statements from the American Academy of Paediatrics guidelines include the following[1]:

- AOM should be diagnosed when there is moderate to severe tympanic membrane bulging or new-onset otorrhoea not caused by acute otitis externa.
- AOM may be diagnosed from mild tympanic membrane bulging and ear pain for less than 48 hours or from intense tympanic membrane erythema; in a non-verbal child, ear holding, tugging, or rubbing suggests ear pain.
- AOM should not be diagnosed when pneumatic otoscopy and/or tympanometry do not show middle ear effusion.

Aetiology

The initial infectious pathogen is typically viral, commonly following an upper respiratory tract infection. Transmission from the nasopharynx via the ET occurs, with stasis, inflammation, and resultant alteration in middle ear ventilation and pressure. As a consequence, the normally sterile middle ear space is colonized by nasopharyngeal microbes resulting in an acute inflammatory cascade within the middle ear cleft with vasodilatation, exudation, and leucocyte invasion. Persistence and suppuration with coexistent bacterial pathogens occurs, most commonly with *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Moraxella catarrhalis.*

Management and guidelines

Given the initial inflammation is commonly viral and spontaneous resolution frequently occurs, current 2018 National Institute for Health and Care Excellence guidance[2] in the UK is for conservative management and self-care with analgesia and hydration in unilateral, non-severe AOM in children aged 6–23 months or non-severe AOM in older children. A back-up antibiotic prescription may be considered should symptoms persist for more than 72 hours. The American Academy of Paediatric guidelines (2013)[1] suggests a joint decision-making process with the parents, and may be managed either with antibiotics or with close follow-up and withholding of antibiotics unless the child worsens or does not improve within 48–72 hours of symptom onset. The number needed to treat to avoid a single episode of mastoiditis is 4831. In the primary care setting, an immediate antibiotic prescription is recommended for children under 2 years of age with bilateral AOM, otorrhoea after tympanic membrane perforation, or if the child is systemically unwell or is at high risk of complications. Groups defined as higher risk of complications are:

- children aged under 6 months
- immunocompromised children—acquired or congenital, most commonly immunoglobulin G subclass deficiency.

Recommended initial therapy is with oral amoxicillin (macrolide if penicillin allergic). Should there be persistence of symptoms, high-dose co-amoxiclav is recommended.

Prevention

At a population level, vaccination with the 13-valent pneumococcal conjugate vaccine and *Haemophilus influenzae B* vaccine has been shown to reduce the incidence of pneumococcal and *Haemophilus* AOM. At an individual level, risk factors include daycare attendance, parental smoking, and pacifier use, while breastfeeding is known to be protective.

Recurrent acute otitis media

The definition of rAOM is three or more distinct episodes within 6 months or four or more episodes within 12 months. Infants who have their first episode of AOM before 6 months of age or who have siblings with severe and rAOM are at risk for more severe rAOM. Individual risk factors, immunization history, and presence of persistent otitis media with effusion should be sought. Children with cleft palate, craniofacial abnormalities, and trisomy 21, who have an increased incidence of otitis media with effusion, are predisposed to AOM. There is increasing evidence to suggest a genetic predisposition to rAOM, with strong familial associations demonstrated in studies.

Conservative measures aimed at reducing modifiable individual risk factors, such as parental smoking and pacifier use, with parental education should be encouraged. Level 1 evidence demonstrates the efficacy of appropriate pneumococcal conjugate and *Haemophilus influenzae B* vaccination in keeping with childhood immunization schedules at reducing pneumococcal and *Haemophilus*-mediated AOM. On occasion, despite appropriate vaccination, a child may not mount an adequate humoral response to infection causing serotypes. In recalcitrant cases, functional antibody serology may be requested, and if an inadequate humoral response is identified, a booster or vaccination with the 23-valent pneumococcal vaccine may be recommended.

There is evidence confirming the efficacy of chemoprophylaxis with medium- to long-term antibiotics. The commonest recommended regimens are with amoxicillin twice a day at half dose for appropriate age/weight or azithromycin 10 mg/kg three times a week taken for 2–3 months. This will vary based on local microbiology recommendations and patterns of local antibacterial resistance. The protective effect ceases once chemoprophylaxis is stopped. In a meta-analysis of 14 randomized trials (1461 children) comparing long-term antibiotics (>6 weeks) with placebo or no treatment for the prevention of acute and chronic suppurative otitis media,[3] antibiotic prophylaxis reduced the occurrence of any episode of AOM (37% vs 56%; pooled risk ratio 0.65, 95% confidence interval (CI) 0.53–0.79%). Approximately five children would need to be treated to prevent one child from experiencing AOM while on treatment and the number of episodes of AOM while on antibiotics reduced from 3 to 1.5 per year (incidence rate ratio 0.51, 95% CI 0.39–0.66).

In those children with persistent otitis media with effusion with rAOM, myringotomy and ventilation tube (grommet) insertion is supported as an alternative to chemoprophylaxis. Typically, a ventilation tube will reduce pain and severity of episodes. Should a breakthrough episode of AOM occur, then otorrhoea via the tube may occur. This then allows treatment with non-ototoxic topical preparations, such as quinolone ear drops. The American Academy of Otolaryngology-Head and Neck Surgery Foundation offers the

following guidance on the use of tympanostomy tube insertion for children with rAOM[4]:

- Tympanostomy tube insertion should not be performed in children with rAOM who do not have middle ear effusion in either ear at the time of assessment for tube candidacy.
- Bilateral tympanostomy tube insertion should be performed in children who have unilateral or bilateral middle ear effusion at the time of assessment for tube candidacy.

A 2018 Cochrane systematic review identified five randomized trials that compared bilateral tympanostomy tube placement (with or without adenoidectomy) with no surgery (i.e. active monitoring, antibiotic prophylaxis, or placebo medication) in children less than 16 years of age.[5] However, the included studies were conducted before the introduction of routine pneumococcal conjugate vaccine and were at significant risk of bias. As a consequence, low-quality evidence suggests that children receiving tympanostomy tubes are less likely to have recurrence of AOM than those managed with active monitoring and placebo medication (approximately one less episode at 6 months and a less noticeable effect by 12 months).

Controversies: prophylactic antibiotics versus ventilation tube insertion for rAOM

Based on the above review, the evidence directly comparing antibiotic prophylaxis and tympanostomy tube placement is uncertain. One trial of 264 children up to 3 years old demonstrated antibiotic prophylaxis was more effective[6] when randomly assigned to three groups (amoxicillin prophylaxis, myringotomy, and tympanostomy tube placement, or placebo) and followed for 2 years. The average rate of new episodes of AOM was decreased in the amoxicillin group compared with the tympanostomy and placebo groups (0.6, 1.02, and 1.08 new episodes of AOM or otorrhoea per child per year, respectively).

Adenoidectomy may be performed in children with repeat tube insertion following recurrence of symptoms and otitis media with effusion. However, the literature does not support its use as a primary preventative measure.

Complications of acute otitis media

Complications of AOM occur rarely in comparison to the pre-antibiotic and pre-pneumococcal vaccination era, with studies demonstrating an incidence of between 0.05% and 0.25%. Complications may be divided into intratemporal and extratemporal and may occur following direct extension, haematogenous spread, or thrombophlebitis. Intratemporal complications include ongoing hearing loss, mastoiditis, facial palsy, and labyrinthitis. Extratemporal complications include intracerebral complications, such as meningitis, empyema, brain abscess, and lateral sinus thrombosis. A low index of suspicion is required, especially in cases where signs and symptoms, such as a spiking pyrexia, persist despite appropriate antimicrobial therapy. In cases where complications are suspected, fine-cut CT of the petrous bone and of the brain with contrast is the emergency imaging modality of choice.

Acute mastoiditis and subperiosteal abscess

Mastoiditis is the commonest complication of AOM and is defined as suppuration within the mastoid air cells from the middle ear space, with subsequent osteitis and periostitis. It presents in less than 0.4% of cases of AOM cases, most commonly under the age of 2 years.

Presenting features include ongoing pain and pyrexia following AOM with associated, erythema, and tenderness over the mastoid process, blunting of the postauricular sulcus, pinna protrusion, and, if this progresses to a subperiosteal abscess, swelling and fluctuance. Abscess formation typically occurs following a coalescent mastoiditis with destruction of the bony air cell septa and breach of the lateral mastoid cortex.

Management is guided by the severity of disease, patient characteristics, presence of other complications, and response to previous measures. Antimicrobial therapy is the mainstay of treatment and usually is the only intervention required in uncomplicated cases. Oral antibiotics have often been prescribed in the community prior to presentation in secondary care and intravenous antibiotics directed towards pathogens associated with AOM (*S. pneumoniae*, *S. pyogenes*) and based on local microbiology sensitivities are instituted. This approach is supported by a systematic review and meta-analysis assessing the management of paediatric mastoiditis, which estimated the success of medical treatment with antibiotics alone at 72.9%. Small subperiosteal abscesses in children who respond to prompt medical therapy may also be managed conservatively or with needle aspiration. Intravenous therapy may need to be continued for 5 days or longer dependent on response and presence of complications. Resolution of pyrexia, otalgia, postauricular erythema, swelling, and tenderness should be sought prior to stepping down to oral antibiotics. A full blood count, C-reactive protein, and, if pyrexial, blood cultures are recommended, with serial monitoring of inflammatory markers allowing monitoring of response to treatment.

In cases refractory to parenteral antibiotic treatment, or where complications have developed, surgical intervention is indicated. A CT scan of the petrous bones and brain with contrast should be considered in cases that are refractory to medical treatment, where an intracranial or temporal bone complication is suspected or prior to acute mastoid surgery.

Surgical intervention comprises initially myringotomy, tympanocentesis, and grommet insertion (ventilation tube), to drain the middle ear, obtain samples for microbiology, and allow the addition of non-ototoxic topical antibiotics, and, where indicated, drainage of a subperiosteal collection via a postauricular incision and cortical mastoidectomy. In young children, due to incomplete development of the mastoid process, the facial nerve exits the stylomastoid foramen in a more superficial position than in adults and this needs to be accounted for when making a postauricular incision. The mastoid air cells are commonly filled with granulation tissue, making identification of the aditus ad antrum and mastoid anatomy difficult. Dissection and microsurgical drilling should proceed cautiously, with a more conservative cavity being created in many instances, to reduce the risk of surgical complications. Samples should be sent for microbiology and histology and a corrugated or yates drain may be placed at the end of surgery.

Considerations for informed consent

Potential complications to discuss when consenting for surgery include ongoing infection, bleeding, hearing loss, tinnitus, vertigo, facial weakness and altered taste, a more prominent ear, and cerebrospinal fluid leak.

If inadequately treated, an acute mastoiditis may develop into a subacute, masked mastoiditis, which may require a prolonged course of antibiotics or surgery. Audiological follow-up should be undertaken in children.

Intratemporal complications

Less common local complications of AOM and acute mastoiditis can include abscess extension inferiorly to sternocleidomastoid (Bezold's abscess), posteriorly with associated osteomyelitis (Citelli's abscess), or medially to the petrous air cells. Gradenigo syndrome, as a consequence of petrous apex infection/petrositis, presents with a triad of discharging ear, retro-orbital pain, and ipsilateral VIth (abducens) cranial nerve palsy. Suppurative labyrinthitis with direct bacterial invasion from AOM via an acquired or congenital fistula or dehiscence in the labyrinth presents with a permanent profound hearing loss and vestibular symptoms.

Facial palsy

Facial palsy is a rare complication of AOM and is presumed to occur due to localized inflammation and oedema of the tympanic and mastoid sections of the nerve within the fallopian canal or a dehiscent facial nerve. Treatment should be with antimicrobial therapy, and myringotomy drainage with or without ventilation tube, to obtain specimens for culture and to allow the middle ear drainage of pus. Oral steroid therapy (1 mg/kg), in cases without evidence of disseminated infection or sepsis, is indicated.

The majority of cases respond well to the above-described management. Extremely rarely, in cases of complete facial paresis with poor response to medical treatment and evidence of subacute mastoiditis, a more aggressive approach with mastoidectomy and facial nerve decompression may be considered. Electroneuronography after 72 hours may be undertaken, with greater than 90% degeneration suggesting a poorer outcome.

Intracerebral complications

A range of intracerebral complications may be seen following AOM, including most commonly meningitis and more rarely empyema, intracerebral abscess, and otitic hydrocephalus. Individual management is outside the scope of this chapter, and medical and neurosurgical input is indicated. The commonest route of spread is haematogenous rather than direct. Intracranial complications may manifest with headache, photophobia, meningism, focal neurological signs, seizures, drowsiness, or acute mental state change in addition to pyrexia. Cases presenting with these features should be appropriately investigated with a CT brain with contrast with or without lumbar puncture and referred for urgent assessment to the appropriate medical or neurosurgical team.

Cerebral venous sinus thrombosis

The incidence of cerebral venous sinus thrombosis (CVST) is low following AOM and mastoiditis, and is seen in less than 3% of cases of acute mastoiditis. Mastoid inflammation leads to dural and intimal inflammation and mural thrombus within the lateral sigmoid sinus.

Guidance from the British Society for Haematology on CVST is not specific to ear-related pathology. A systematic review of the management of otogenic CVST identified 190 patients with level 4 evidence.[7] All children were treated with antimicrobial therapy, with the majority (91.6%) requiring surgical intervention, most commonly with cortical mastoidectomy and ventilation tube insertion with or without lateral sinus decompression or thrombectomy.

Controversies: anticoagulation for otogenic CVST

Currently, there is no clear guidance of the benefit of anticoagulation, and a shared decision-making process is advised based on the individual case and response to medical and surgical interventions. The aim of anticoagulation is to prevent propagating thrombus. This may be identified on initial CT imaging, with an appropriate venogram phase study, or following interval imaging. Gadolinium-enhanced magnetic resonance venography is the modality of choice, and is more sensitive than CT. A decision to anticoagulate should be made in conjunction with neurology and haematology input, after a full assessment of the benefits and risks, and is normally reserved for patients with evidence of thrombotic propagation to the intracranial venous sinuses and/or jugular vein, or in the presence of neurological sequelae and signs or embolic events (**Figure 84.1**).

Otitis externa

Acute inflammation of the external auditory canal is a less common cause of a painful discharging ear in children. Otitis externa presents acutely with pain, blockage due to debris, itchiness, and often discharge. As covered in Chapter 8, the most common bacteria are *Staphylococcus aureus* and *Pseudomonas aeruginosa*.

Topical treatment

A Cochrane review has demonstrated topical antimicrobials containing steroids were significantly more effective than placebo drops: odds ratio 11 (95% CI 2.00–60.57). In general, no clinically meaningful differences were noted in clinical cure rates between the various topical interventions reviewed. Typically, a 7- to 10-day course is prescribed on an empirical basis. There is little evidence to support the routine use of microbiology swabs to guide the choice of treatment. Water precautions are required until the infection is completely treated.

Ciprofloxacin 0.3% with or without steroid (Ciloxan or Ciprodex/Cilodex), Sofradex, and Gentisone HC preparations are commonly used with quinolone preparations being the safest if a tympanic membrane perforation cannot be excluded as they are non-otoxic. Alternatively, antiseptic preparations such as 2% acetic acid or astringent agents such as aluminium acetate ear drops are also available. Previous antiseptic preparations such as glycerol ichthammol are no longer readily available.

Children who fail to respond to topical treatment and/or if the canal is completely occluded should be reviewed in secondary care for aural toilet with further topical treatment and oral antibiotics for severe cases. Intravenous antibiotics may be indicated if there is evidence of spreading cellulitis or perichondritis. Caution should be taken in prescribing systemic oral ciprofloxacin in children due to the risk of arthropathy. If the canal is occluded from oedema an otowick should be inserted, with removal planned at 48–72 hours. A microbiology swab is indicated for severe or prolonged cases not improving on topical treatment.

Fungal otitis externa may be a cause for persistence despite antimicrobial topical drops. Fungal spores may be seen on otoscopy

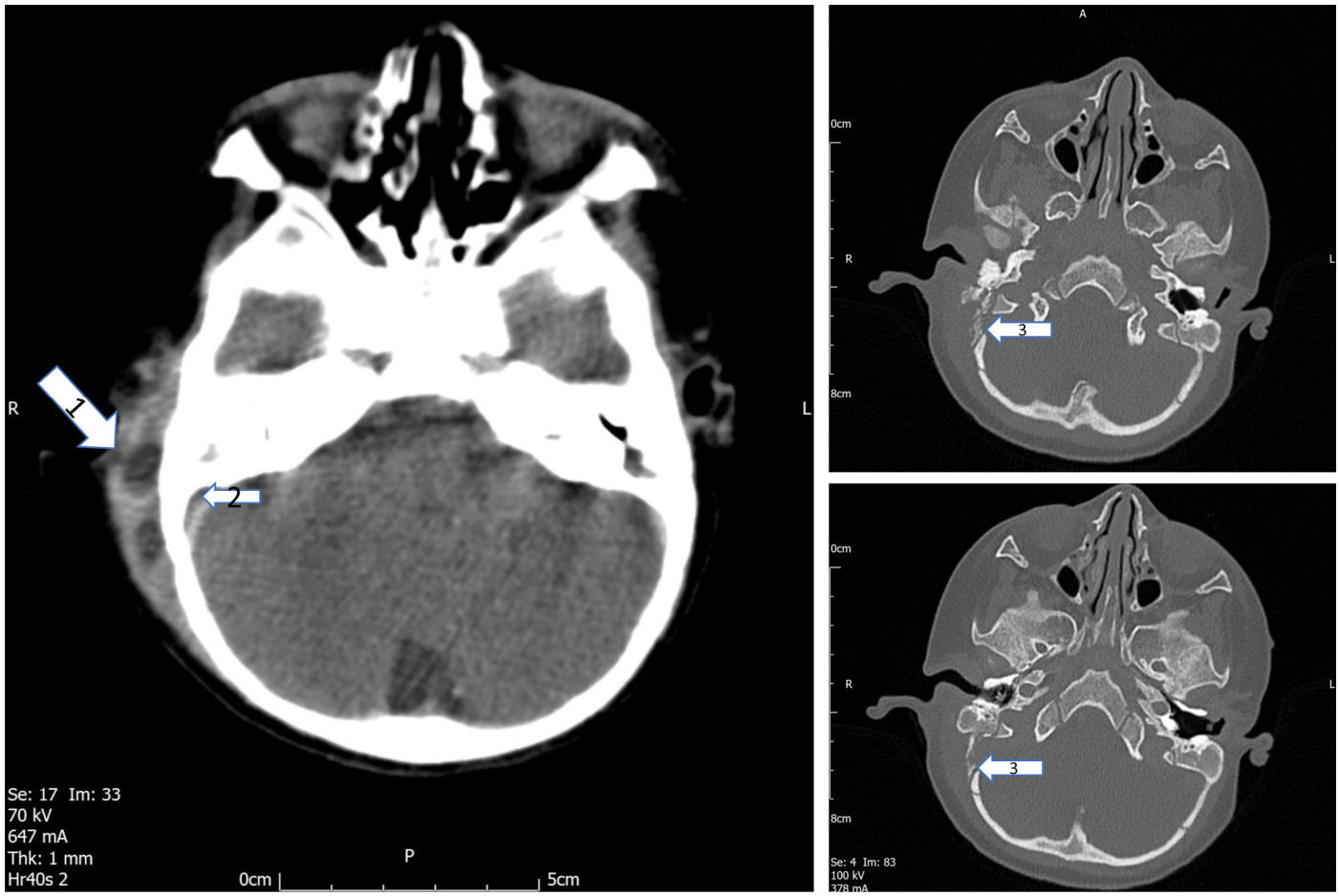

Figure 84.1 Acute mastoiditis with subperiosteal abscess (1), perisinus collection (2), and osteomyelitic changes of temporal and parietal bone (3).

and microbiology culture demonstrates *Aspergillus niger/fumigatus* or *Candida* spp. Prolonged and extensive use of topical antimicrobial or corticosteroid treatment may affect the flora in the ear canal, increasing the risk of fungal infections. If a mild to moderate, uncomplicated fungal infection is suspected in the context of chronic otitis externa, a topical antifungal such as clotrimazole 1% solution (Canesten HC) or clioquinol and a corticosteroid can be offered. Sensitivity to topical ear preparations may also occur, specifically to the preservative carrier benzalkonium chloride, especially with prolonged or recurrent use. This can be identified by dermatological patch testing.

Foreign body

Foreign bodies within the external auditory canal are common in children. They are commonly asymptomatic, but may cause pain or discharge, especially if the foreign body is organic.

Foreign bodies can often be removed in the ear, nose, and throat clinic using the microscope and instruments such as a wax hook. For foreign bodies that are impacted and too painful to remove or in the non-compliant child, general anaesthetic removal is needed. Inert foreign bodies such as plastic beads can be removed on an elective list within 6 weeks of being seen. Organic foreign bodies need removal on a semi-urgent basis. A battery foreign body should be removed as an emergency.

Neoplastic disease

Very rarely, persistent otorrhoea or otalgia non-responsive to appropriate medical management may be related to an underlying neoplastic cause. Children may present with chronic otorrhoea with evidence of granulomatous changes within the auditory canal or middle ear or an aural polyp. On occasion, cranial nerve involvement may be a presenting feature. In such cases there should be low threshold for biopsy and appropriate temporal bone imaging.

The most common paediatric malignant middle ear tumour is rhabdomyosarcoma (embryonal parameningeal subtype) with a mean age of presentation of 5 years old. Langerhans histiocytosis is the next most common tumour seen and is a heterogeneous single or multisystem condition. It is characterized by an idiopathic monoclonal proliferation of Langerhans' cells. In approximately 15% of cases temporal bone involvement is seen. Diagnosis is with biopsy, including for staining for S100 and CD1a. Temporal bone imaging shows characteristic osteolytic lesions on CT and, when involved, an expansile soft tissue mass of the middle ear or mastoid.

The mainstay of treatment of both conditions is with multimodality therapy with chemotherapy and/or radiotherapy undertaken by the paediatric oncology team in a multidisciplinary setting.

Chronic otitis media

Tympanic membrane retraction

A retracted tympanic membrane is caused by chronic ET dysfunction and therefore is more prevalent in children prone to this—more frequently in children with a craniofacial diagnosis, cleft palate (see Chapter 89), or trisomy 21 (see Chapter 90).

The history may be suggestive of ET dysfunction, with popping sensations, struggling with changes in pressure, being unable to perform a Valsalva manoeuvre to equalize middle ear pressure, hearing loss (often intermittent), intermittent otalgia, and discharge (recurrent infection).

Retractions can be graded by the Sade (pars tensa) and Tos (pars flaccida) classification. Pars tensa retractions are more common in children than pars flaccida retractions. Potential complications of a retracted tympanic membrane include ossicular erosion (posterior–superior retraction with incus erosion) with permanent conductive hearing loss, recurrent infections, and progression to cholesteatoma.

Treatment

Although the pathophysiology of tympanic membrane retraction is well understood, there is no consensus on treatment. Options for treatment include conservative with active monitoring, medical (debridement and topical antibiotic drops for infection), or surgical.

Controversies: conservative versus surgical management of tympanic membrane retractions

A Cochrane review examining surgery for tympanic membrane retraction pockets in both adults and children did not show evidence either to support or refute the role of surgical intervention.[8] There is controversy and variation in practice regarding when to intervene for a tympanic membrane retraction. Some authors have shown progression of retractions without intervention, whereas others have advocated for a watchful approach due to a low risk of progression in the cohorts they have observed.[9,10]

A proportion of children will improve their middle ear ventilation with age (especially towards the teenage years), and tympanic retraction will spontaneously resolve. In asymptomatic cases with normal hearing, a policy of watchful waiting may be more prudent. Surgery may be indicated if the child is significantly symptomatic, to prevent progression of a retraction and complications from a retraction. Alternatively, complications of retractions may be treated surgically if and when they develop.

Preventative surgical options include myringotomy and ventilation tube insertion or cartilage tympanoplasty. Ventilation tube insertion can be challenging due to limited middle ear space, but intraoperative nitrous oxide can help elevate the tympanic membrane from the promontory. Tubes can also extrude earlier with ongoing ET dysfunction. Multiple ventilation tubes or long-term T-tubes may need to be undertaken. Incus erosion is not uncommon with posterior–superior retraction and incudopexy. Tympanoplasty for those with incudopexy without erosion is unlikely to adversely affect postoperative hearing outcomes and hearing may improve postoperatively for those with incus erosion. Cartilage tympanoplasty can be challenging as elevating a significantly retracted or atelectatic tympanic membrane risks damage to the ossicular chain and implantation cholesteatoma.[11] Use of the KTP laser in the middle ear at the time of surgery will reduce the risk of implantation cholesteatoma. Tympanomastoid surgery and ossicular chain reconstruction is indicated to treat complications (Figure 84.2).

Tympanic membrane perforation

Recurrent or relapsing otorrhoea is a presenting feature of infection with an underlying tympanic membrane perforation. Persistent perforations in children most commonly occur following tympanic membrane rupture during AOM or iatrogenically, following myringotomy and ventilation tube insertion.

Treatment

Acute infections are most successfully treated with aural toilet/microsuction and appropriate non-ototoxic topical antimicrobial

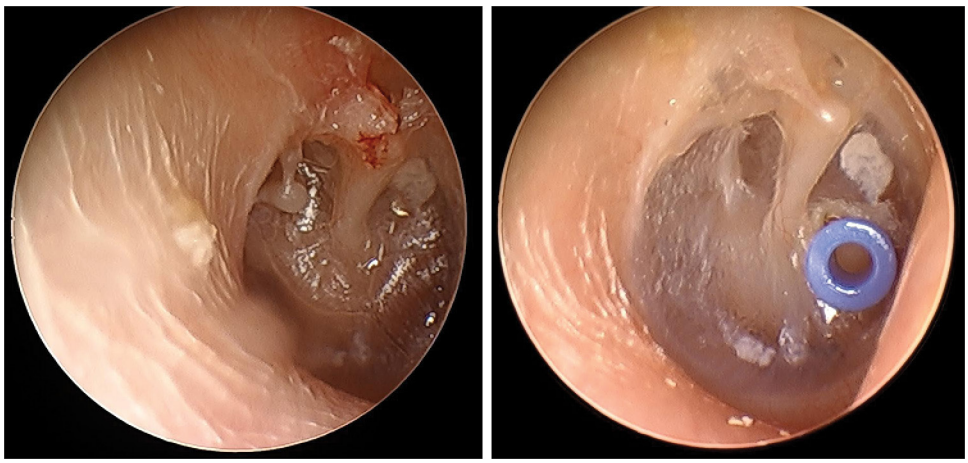

Figure 84.2 Tympanic membrane retraction; before and after ventilation tube insertion with correction of grade 3 retraction.

therapy. Indications for surgical closure (myringoplasty/type 1 tympanoplasty) are patients wishing to reduce the cycle of recurrent otorrhoea and infections, desire to cease water precautions, and, less commonly, when a significant conductive hearing loss is present, usually associated with a subtotal perforation, to improve the hearing.

Details regarding technique, graft materials, and outcomes are discussed in Chapter 22 on tympanoplasty and will not be repeated. Briefly, various approaches and graft placement techniques are available to the surgeon, including a traditional postauricular or endaural incision, a per-meatal approach, and, more recently, an endoscopic approach. Various autologous biological graft materials may be used, including temporalis fascia, perichondrium, tragal or conchal cartilage, fat, or non-autologous graft materials such as Biodesign. Grafts are most commonly placed using an underlay technique, but butterfly, push-through, overlay, and fasciaform techniques are routinely practised. There is no equivocal evidence to suggest performing a cortical mastoidectomy concurrently improves the success rate.

Controversies: timing of surgery

In children, the primary dilemma is regarding the timing and age of the child at which tympanic perforation repair is undertaken to maximize the chance of successful repair. This is based on patient factors, such as a history of cleft palate or craniofacial abnormalities, past otologic history, and traditionally at an age when upper respiratory tract symptoms and ET function improve secondary to anatomical development and adenoidal regression. The age when rAOM or otitis media with effusion are no longer prevalent and the above factors are met is most commonly after the age of 7 years. Age has been the only factor shown by meta-analysis to affect outcome, with success improving every year up to the age of 13 years.[12] In case of a unilateral perforation, the contralateral ear may be used as an indicator of middle ear function, especially if ventilation tubes have previously been inserted or there is a significant otologic history.

A systematic review and meta-analysis of 2609 cases identified tympanoplasties performed on larger perforations or in children with abnormal contralateral ear findings were more likely to fail. Adenoidectomy, graft position, and active infection were not found to significantly alter the overall success rate of 83%.[13]

Cholesteatoma

Cholesteatoma is defined as keratinizing squamous epithelium in the middle ear cleft. It is a three-dimensional epidermal structure that exhibits independent growth, replaces middle ear mucosa, and reabsorbs bone. Cholesteatoma affects 5–15/100,000 population, presenting in males more than females (ratio 3:1).[14]

Risk factors for cholesteatoma in children include chronic otitis media and conditions predisposing to ET dysfunction, including cleft palate, trisomy 21, and children with craniofacial anomalies. The largest proportion of cases is seen in the second decade of life, followed by the first.

Physiology

Cholesteatoma can be congenital or acquired, with the vast majority of cases being acquired (98%). Primary acquired cholesteatoma occurs in a retraction pocket as a consequence of chronic ET dysfunction and negative chronic middle ear pressure, which is more prevalent in a paediatric population. The retraction pocket expands over the malleus head to erode the scutum developing into attic cholesteatoma. Secondary acquired disease occurs following migration of epithelial cells through a perforation or implanted via trauma or iatrogenically.

Congenital cholesteatoma represents 2% of paediatric cholesteatoma cases and tends to occur in the first decade of life with a mean age of 4.5 years. Classically, congenital cholesteatomas are described in the anterior–superior quadrant of the middle ear, near the manubrium of the malleus, with minimal ossicular involvement. This classical presentation is more common in younger children. However, disease can variably occupy the middle ear and mastoid. Posterior–superior quadrant disease is also commonly seen (especially with increasing age), with disease near to the incudostapedial joint frequently with ossicular erosion.[14,15]

The Derlacki and Clemis (1965) criteria for congenital cholesteatoma require a[16]:

- white mass medial to an intact tympanic membrane
- normal pars tensa and flaccida
- no previous ear surgery, otorrhoea, or perforation.

Controversies: aetiology of congenital cholesteatoma

Theories for congenital cholesteatoma include:

- embryonic epithelial rests in the middle ear (failure of involution and growth of epidermoid formation, derived first branchial groove ectoderm), most accepted theory[14]
- epithelial cells present in the amniotic fluid may enter the middle ear cleft and subsequently grow into a cholesteatoma
- squamous metaplasia of the middle ear mucosa.

Management

Classifying congenital cholesteatoma disease extent aids surgical planning and helps predict outcomes from surgery. A number of classifications exist.

Nelson classification[17]:

- Type 1—confined to the middle ear and does not involve the ossicles.
- Type 2—involves the posterior superior quadrants and attic, the site of the ossicular chain.
- Type 3—involve the sites of type 1 and 2 as well as the mastoid.

Potsic stages[18]:

- Stage I—limited to one quadrant.
- Stage II—involving multiple quadrants without ossicular involvement.
- Stage III—ossicular involvement without mastoid extension.
- Stage IV—mastoid involvement.

Increasing age in childhood is associated with more extensive disease and higher classification grades. Closed-type cystic cholesteatoma is more commonly seen in younger children, with older children having open-type cholesteatoma. More extensive initial disease and ossicular erosion is associated with a higher proportion of residual disease.[19,20]

Features of acquired cholesteatoma specific to children include slow growth initially; however, once the matrix is breached, they behave more aggressively. Compared to adults, children have higher rates of pars tensa disease, less erosion of the lateral semicircular canal and bone canal of the facial nerve, and more extensive due to a more pneumatized mastoid allowing more rapid epithelial growth. Comparison of disease between adults and children has shown that paediatric cholesteatomas may produce a more inflammatory response. Granulation tissue associated with cholesteatoma is more common in children than in adults who tend towards keratin matrix alone. The association of granulation tissue with cholesteatoma may reflect more aggressive disease and a higher risk for residual disease.[21,22]

Management in children is often more challenging as it more frequently involves the extremes of the middle ear space leading to a higher risk of residual disease, while persistent ET dysfunction in children predisposes to an increased rate of recurrent disease.

Assessment

Examination under anaesthetic may be required if the diagnosis is uncertain or the child is unable to comply with examination to adequately visualize the entire tympanic membrane. Audiometry may show a conductive or mixed loss.

Investigations

Preoperative imaging, most commonly with CT, is increasingly undertaken with the aims of delineating the anatomy, specifically regarding the level of the tegmen and sigmoid sinus, assessing for complications and extent of disease, allowing patient-specific preoperative counselling, and to guide preoperative planning regarding the surgical approach, such as the feasibility of a canal wall up procedure or per-meatal endoscopic approach to isolated middle ear disease. In some circumstances medico-legal aspects may be an indication for imaging. In children, the short duration of CT is generally tolerated well without need for anaesthesia, and low-dose protocols are able to minimize the radiation dose children are exposed to (0.25–2.5 mSv).

Diffusion-weighted echo-planar magnetic resonance imaging can be less rarely used as a preoperative scanning modality or more commonly as an alternative to second look in combined-approach tympanoplasty patients. It has the advantage over CT of being able to distinguish between inflammatory tissue and cholesteatoma, where other imaging modalities cannot, due to the duration of imaging typically requiring anaesthesia or sedation under the approximate age of 7 years.

Treatment of cholesteatoma

Surgery is indicated for all cholesteatomas unless the child/patient is not fit for anaesthetic. Medical treatment involves symptom control while waiting for surgery with microsuction to remove debris and topical antibiotics/steroids if there is infection.

The aim of surgery is to remove disease, prevent recurrence, have a dry self-cleaning, infection-free ear, and restore hearing. The type of surgery depends on patient factors, surgeon experience, and disease extent. Technical aspects of cholesteatoma surgery are described in Chapter 19 and include minimally invasive procedures (endoscopic), and canal wall up and canal wall down procedures. To prevent children requiring a lifetime of aural care and water precautions, canal wall up surgery is increasingly preferred, but on occasion canal wall down surgery is necessary (Figure 84.3 and Table 84.1).

Tips and pearls—cholesteatoma

Consent

A surgeon performing cholesteatoma surgery should discuss complications related directly to surgery but must also discuss the high possibility of residual/recurrent disease in paediatric cases, possible hearing outcomes, need for future imaging (diffusion-weighted magnetic resonance imaging), and a high likelihood of future surgery including second-look surgery and ossiculoplasty.

Endoscopic surgery

The authors advocate the use of endoscopes in all paediatric cholesteatoma surgery. Modern 3 mm otoendoscopes are now commonly used in paediatric otology, increasing the number of cases that can be performed transcanal and with a minimally invasive approach.

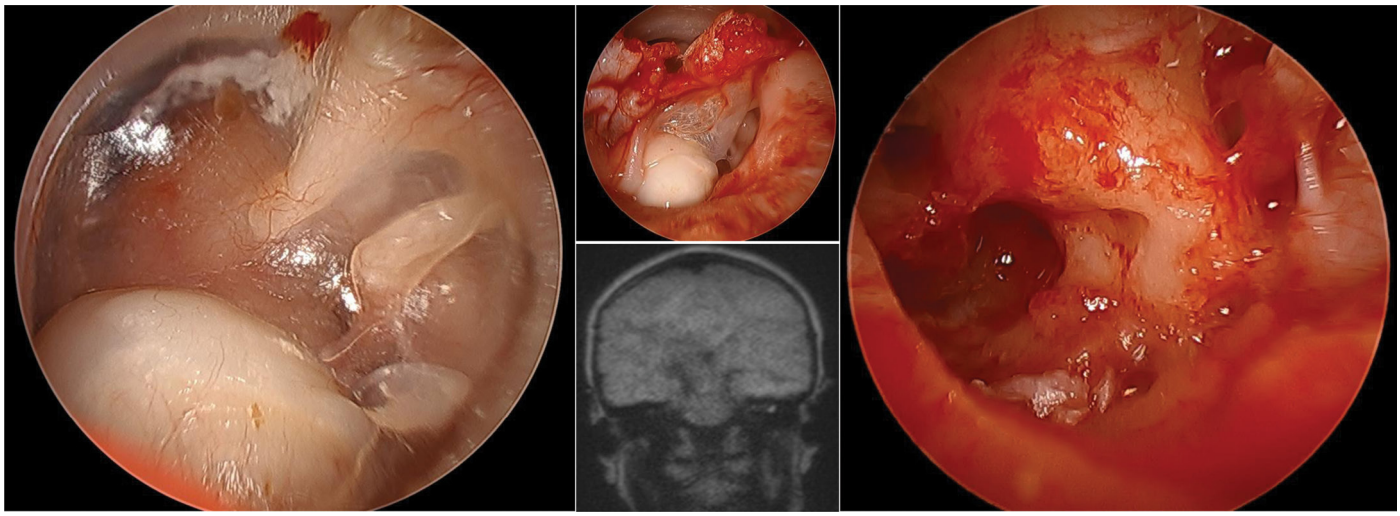

Figure 84.3 Congenital cholesteatoma, with preoperative diffusion-weighted imaging cholesteatoma protocol magnetic resonance imaging scan and perioperative endoscopic images. The disease was amenable to transcanal endoscopic removal.

Table 84.1 Pros and cons of canal wall up versus canal wall down cholesteatoma surgery

	Canal wall up	Canal wall down
Advantages	• Physiological tympanic membrane position • Better acoustic properties • No mastoid bowl—reduced aural care and infections • Favourable for hearing reconstruction later	• Ability to deal with disease in one setting • Ability to see recurrent disease more easily • Lower incidence of recurrent disease
Disadvantages	• Second stage often required • Higher rates of residual and recurrent disease	• Altered anatomy • Life-long cleaning • Higher rate postoperative discharge • Greater difficulty in reconstructing hearing

The authors aim to perform total endoscopic ear surgery for those children with disease limited to the middle ear and combined microscopic and endoscopic for those with extension to the mastoid beyond the lateral semicircular canal. Endoscopes allow for visualization of tympanomastoid recesses not visible by the operating microscope. The authors recommend the use of dedicated 3 mm otoendoscopes (0°, 30°, and 45°) and appropriate curved dissecting instruments.

A systematic review of the literature has shown that endoscopic ear surgery, either alone or in combination with the microscope, is associated with lower residual disease compared to microscopic techniques.[23]

The authors commonly use a transcanal endoscopic approach in children over 3 years; however, not all children's ear canals are wide enough to accommodate both an endoscope and instruments, especially children with craniofacial anomalies. In these cases, the initial approach is microscopic, with the endoscope being used as an adjunct once sufficient access is achieved.

Laser-assisted surgery

The KTP laser as an adjunct to the endoscope and microscope has been shown to reduce residual disease.

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85

Dizziness in children

Alok Sharma and Gundula Thiel

Introduction

Childhood dizziness is a common problem with an estimated incidence of between 5% and 18%.[1–3] However, children all develop abilities at different ages and to different extents and teasing out what is abnormal from normal can also be quite tricky.

A good history is essential to allow diagnosis. The child's ability to communicate can pose significant challenges. Thus the history is often taken primarily from the parent but it is important to elicit as much information as possible from the child.

Similar difficulties can be encountered during a complete otoneurological evaluation. It must be noted that children have a remarkable ability to compensate for and adapt to static vestibular defects.

The differential diagnosis of a vestibular disorder in a child encompasses medical subspecialties as diverse as ear, nose, and throat, neurology, medical paediatrics, and audiology.

We describe in this chapter a practical approach to dizziness in childhood to aid diagnosis of commonly encountered causes for vestibular disturbances in children (see Chapter 7 for adult vertigo management). This will be helpful in both acute and chronic presentation of vertigo. Variations of this approach are used worldwide and are adapted from Halmagyi.[4]

History

See Table 85.1.

How does it feel?

Is there an illusion of movement or is it just light-headedness?

It is important to find out if the child is describing true vertigo (illusion of movement such as rotation), disequilibrium (a to-and-fro movement in the vertical or horizontal plane), light-headedness (feeling 'fuzzy'), *or* presyncope (feeling faint).

True vertigo is most indicative of a vestibular disorder; however, there is significant overlap between symptoms of peripheral and central dysfunction.

Central neurological causes should be excluded (hereditary ataxia, posterior fossa disease, hydrocephalus).

Table 85.1 History

Question	Answer	Possible differential diagnosis
How does it feel?	Illusion of movement	
	Light-headedness	
How long does it last?	Seconds	BPVC (migraine)
		BPPV (vestibular)
	Minutes to hours	Vestibular migraine
		Ménière's disease
	Hours to days	Labyrinthitis/vestibular neuronitis
What makes it worse?	Movement	Peripheral cause
	Rapid position change to upright	Orthostatic reaction
	Rolling over/looking up	BPPV
	Straining or noise	Dehiscent superior semicircular canal syndrome
Associated symptoms?	Hearing loss:	
	Conductive	Otitis media with effusion
		Chronic suppurative otitis media
		Third-window effect
	Sensorineural	Cochleovestibular anomalies
		Infective
Milestone delay	Yes	Congenital or severe vestibular loss
Head injury	Yes	Temporal bone fracture
		Post-concussion syndrome
Ototoxic drugs	Aminoglycoside, platinum, antimalarials	
Headaches	Yes	Vestibular and basilar migraine
Accompanying aura	Visual, auditory	Migraine
		Seizures
Visual problems		Refraction/vergence
Mental health		Can be cause of or reaction to dizziness
Ataxia		Cerebellar/posterior column

In children who present without a history of 'true vertigo', common medical conditions (anaemia, diabetes mellitus) should be excluded first. Those who give a history of presyncope, cardiac (e.g. arrhythmias), and circulatory (postural hypotension) causes should be investigated.

How many episodes and how often?

Multiple, short episodic vertigo?

Acute episodic vertigo for seconds (30–60) is indicative of benign paroxysmal vertigo of childhood (BPVC) (also known as benign recurrent vertigo of childhood or Basser's syndrome) or benign paroxysmal positional vertigo (BPPV).

BPVC is a migraine variant whereas BPPV results from either canalithiasis or cupulolithiasis.

BPPV versus BPVC

BPPV is rare in children, typically only occurring following head trauma or surgery (e.g. cochlear implantation) though one of the most common aetiologies of vertigo in the adult population. However, BPVC is one of the most common causes of vertigo in children between 2 and 6 years of age, causing up to 35% of cases of childhood dizziness.[5]

The clinical presentation of BPVC is short-lived episodes of intense vertigo lasting seconds, which is not positional and has no precipitating factors (unlike BPPV). It can be associated with nystagmus and postural imbalance. In some cases nausea and vomiting occur, but no cochlear symptoms are present. Children do not show any signs of vestibular dysfunction between episodes. Half of these children will go on to develop migraine in adolescence and most have a family history of migraine.[6]

Motion intolerance or 'car sickness' is also a feature associated with migraine variants and a risk of migraine later on in life. Other forms of migraine variant may also be present including benign paroxysmal torticollis (<2 years of age) and cyclical vomiting.

Multiple episodes: minutes to hours

Multiple episodes of vertigo lasting longer than 20 minutes, and up to several hours in duration, are most commonly due to vestibular migraine and, less commonly, Ménière's disease. Childhood or adolescent onset of Ménière's disease has clinical features similar to adults (see Chapter 14); however, early onset tends to be associated with more aggressive disease and increased likelihood of early bilateral involvement.[7]

Single episodes: hours to days

Acute-onset vertigo that is severe and incapacitating, and lasting hours to days, is in keeping with a diagnosis of labyrinthitis or vestibular neuronitis. Often these children will need to be admitted acutely to hospital and will be investigated for central causes, although clinical history and examination will be consistent with a peripheral origin. As in adults, vestibular neuronitis in children presents with acute severe rotatory vertigo and postural imbalance with nausea, vomiting, and oscillopsia but normal hearing. The duration of vertigo is often days though children recover more quickly from this disorder than adults due to their ability to compensate. A prodromal history may support a viral cause for acute vestibular failure, which may (labyrinthitis—when the inner ear is involved) or may not (vestibular neuronitis—when the vestibular nerve only is involved) be associated with a hearing loss.

What makes it worse?

Movement

Classically, a peripheral-caused vertigo will be exacerbated by any movement. Note that it needs to be differentiated from motion sickness by a careful history. Children between the age of 4 and 10 years are most susceptible and it is a common finding in migraineurs.[8]

Orthostatic hypotension can cause vertigo when rapidly moving from lying down to standing or after prolonged standing and can be accompanied by a sensation of 'feeling faint'. This can be seen in children undergoing rapid growth around puberty and corresponds to a transitory poor adaptation of the cardiovascular system to sudden changes in body position.

Rolling, bending, looking up?

This may underline BPPV, which is detailed above.

Straining or noise?

Dehiscent superior semicircular canal syndrome has been described in children though is rare. Tullio phenomenon (vertigo precipitated by sound) and Hennebert's sign (positive fistula test with normal middle ear) are typically found. Three-quarters of patients also experience chronic disequilibrium, which is often the most debilitating symptom.[9]

Any associated symptoms/problems?

Hearing loss?

In vertigo with accompanying hearing loss:

Conductive hearing loss?

Otitis media with effusion (glue ear) may be detected in the clumsy child with poor balance, typically with accompanying hearing loss. This has been quoted[10] as the most common cause of vestibular complaints in children and the symptoms are 'unsteadiness' and 'clumsiness' rather than true vertigo. Golz et al.[11] postulated that toxins present in middle ear fluid enter the inner ear fluid and cause serous labyrinthitis. Other researchers[12] have proposed that pressure changes in the middle ear cause displacements of the round and oval windows, leading to secondary movement of labyrinthine fluids. Squamous chronic suppurative otitis media (cholesteatoma) may be accompanied by vertigo and hearing loss with a possibility of fistula to the lateral semicircular canal or oval window leading to labyrinthitis. History and clinical examination will help reach a diagnosis.

Other causes of conductive hearing loss and vertigo or disequilibrium include perilymphatic fistula (described in detail below) as well as any other condition that produces a third-window effect (e.g. enlarged vestibular aqueduct, superior canal dehiscence).

Sensorineural hearing loss?

Concurrent abnormalities of the vestibular portion of the inner ear occur in up to 70% of children with permanent hearing loss.[13,14] Children with cochlear vestibular loss present with motor delay or disequilibrium as opposed to true vertigo. However, some children with cochleovestibular anomalies can present with episodic and progressive vestibular loss which is characterized as sudden-onset acute vertigo lasting hours to days with or without changes in hearing.[15] Cochleovestibular anomalies include, for example, enlarged vestibular aqueduct alone or in combination with the spectrum of

incomplete partitions (IP-1, IP-2, and IP-3). These anomalies can be non-syndromic or syndromic (e.g. Waardenburg, Pendred, or branchio-oto-renal syndromes—see Chapter 90). Episodic vertigo due to cochlear-vestibular abnormalities can be mistakenly diagnosed as Ménière's disease in children which, as previous explained, is rarer. Another syndrome with cochleovestibular loss is Usher syndrome, which is further compounded by progressive visual impairment due to retinitis pigmentosa. Identification of vestibular dysfunction in a child with profound hearing loss of unknown aetiology should initiate a work-up for Usher syndrome which typically includes electroretinography (see Chapter 90). In addition, there are a number of acquired or congenital infectious causes of combined cochleovestibular loss including meningitis and cytomegalovirus—see Chapter 83.

Delay in developmental milestones?

A thorough review of motor milestones should be performed in all children presenting with vertigo. While children with episodic vertigo will rarely have delay in their milestones, children with congenital or acquired severe vestibular loss most certainly will. Specifically, inquiring whether head control was achieved by 6 weeks, sitting by 6–9 months, and walking between 12 and 15 months can contribute to the suspicion of vestibular impairment. Additionally, inquiring about balance-intensive activities, such as the age at which the child was able to ride a bicycle without training wheels, can also signal vestibular impairment.

Head injury?

Head injury causing a temporal bone fracture involving the labyrinth or a perilymphatic fistula can cause acute vertigo with tinnitus and hearing loss. The history characteristically helps in the diagnosis. More generally, post-concussive syndrome may also include symptomatic vertigo not associated with hearing loss and these children are also at higher risk of BPPV.

History of ototoxic drug use?

Previously commonly prescribed topical aminoglycoside antibiotics, especially gentamicin, are vestibulotoxic and can cause marked disturbance of vestibular dysfunction with acute vertigo and ataxia. Thankfully this is not usually associated with hearing loss whereas antimalarials and platinum-based drugs can cause ototoxicity in the form of high tone hearing loss and tinnitus. While vertigo is rarely reported, the vestibulotoxic effects are likely underappreciated as screening protocols rarely assess vestibular function.

Associated headaches? Vestibular and basilar migraine?

Over 50% of dizzy children have an association with headache[1,16] and vestibular migraine accounts for 20–36% of headache in children.[16–19] The typical presentation is episodic attacks of rotatory vertigo, lasting minutes to hours, accompanied or followed by headache and often vomiting, typically with sensitivity to light and sound. An important clue in the diagnosis is the family history; 85% have a history of a first-degree relative suffering from migraine.[6]

Vestibular migraine should be distinguished from basilar migraine where the audiovestibular symptoms may be part of the aura and there may be other symptoms like paraesthesia, ataxia, and loss of consciousness. The distinction is important because the treatment of the two conditions is different.

Accompanying aura?

Vertigo with aura may be part of vestibular migraine, but may also be indicative of seizures. In temporal lobe or occipital lobe epilepsy, the child may describe a sensation of movement with visual or auditory hallucinations.[18]

A comparison of visual hallucinations in patients with migraine and occipital epileptic seizures showed that in epileptic seizures they are multicoloured with circular or spherical patterns as opposed to the black and white linear patterns of migraine; this may be helpful in distinguishing between the two.[20] Similarly, symptoms of temporal lobe seizures can be similar to BPVC; therefore, if aura is present or there is prolonged recovery which may represent a post-ictal state, then electroencephalography and referral to neurology is warranted.

Accompanying ophthalmological symptoms?

It is reported that visual problems alone lead to vertigo in children aged over 5 years in 10% of cases.[21–23] Specifically, brief recurrent vertigo may be caused by errors in refraction (myopia, hypermetropia, or astigmatism) or anomalies in ocular vergence. These episodes are associated with fatigue (end of day, or after long sessions on computer or television screens) and can often happen with position changes of the head or gaze; they are often accompanied with headaches and nausea, but rarely vomiting.[24] This vertigo may be being seen increasingly, associated with the increased availability and usage of screen devices by children and prolonged personal device screen time.

Mental health symptoms?

Childhood mental health issues may also manifest in complaints of 'dizziness'. Mental health symptoms can result from long-standing peripheral or central vestibular dysfunction that has been slow to resolve (e.g. prolonged post-concussive symptoms), or vertigo may represent the physical symptoms associated with a primary diagnosis of depression or anxiety. Prolonged school avoidance is a marker of potential associated mental health symptoms. Medically unexplained vertigo is a diagnosis of exclusion. Thorough workup is required to identify organic causes of vertigo that require other treatment. Multidisciplinary assessment is key and the majority of patients are effectively treated with therapy-based interventions while a minority require pharmacological therapy. An appropriate treatment plan can be effective in symptom reduction and improved quality of life.[6]

Ataxia?

Ataxia is a common mode of presentation of cerebellar and posterior column disease in children. Conditions that may present with ataxia in children include Chiari malformations, hereditary cerebellar ataxia, Refsum's disease, viral encephalitis, and posterior fossa tumours. Associated localizing neurological signs are often present.

What not to miss

While extremely rare in children presenting solely with dizziness, a space-occupying lesion should be considered in the differential diagnosis.

It is important to remember that it is not only hearing loss that distinguishes auditory neuropathy patients from their normal counterparts; there is mounting evidence emerging that this group

of patients with 'auditory neuropathy' may also demonstrate 'vestibular neuropathy' and may demonstrate abnormal findings on balance function tests as well.[25]

What you should cover on examination

The completeness and thoroughness of your examination will depend on the cooperation of the patient. For each child you will need to gauge the degree of cooperativeness and grade your examination, obtaining the most relevant information first. While striving for a complete head and neck and neurotological examination is ideal, in children it is often incomplete. Children-friendly adaptions can be made through games and play and a degree or patience.

A few key manoeuvres can help efficiently identify those children at risk of impairment. These include three things:

1. The milestones outlined in the history section, above.
2. Having the child stand on one foot (30 months—briefly, 36 months—2 seconds, 4 years—5 seconds, 5 years—10 seconds).
3. Head thrust testing—while this is the minimum in children presenting with vertiginous complaints or hearing loss, a more detailed evaluation is outlined below.

General physical examination should include evaluation of head and neck for trauma, congenital anomalies, neck mobility/tenderness, and temporomandibular joints.

Examine the ears

To exclude middle ear disease, especially otitis media with effusion and cholesteatoma.

Assess nystagmus

The presence of nystagmus may indicate peripheral or central pathology. As peripheral vestibular nystagmus is suppressed by visual fixation, it is ideal to prevent fixation by using Frenzel glasses or infrared goggles. Spontaneous nystagmus should be assessed in all positions except extremes of gaze (>30°) to prevent physiological nystagmus secondary to extraocular muscle elasticity in children.[26] (Fistula testing in children is best done with electronystagmography and specific measurements of pressure levels using an immittance meter.)

Neurological examination

The neurological examination may be divided into examination of children under 4 years and over 4 years of age. In children under 4 years most information is obtained through observation, play, and reflex behaviour; tonic neck reflexes are present from 4 months of age. Newborn infants may focus but do not follow objects. By 2 weeks of age, visual following occurs and by 6 weeks, following both vertically and horizontally should be present. In children older than 4 years the general neurological examination is comparable to that of adults.[27]

Examination of cranial nerves and cerebellar function

Focal neurological signs may be detected in patients with brainstem or cerebellar lesions.

Performing balance tests

Romberg's and Unterberger's (or Fukuda's) tests are not specific and may be abnormal in peripheral or central lesions. They are, however, a useful way to assess balance with vision eliminated. Ataxia due to a central cause may be more severe (e.g. patient unable to stand) than that from a peripheral cause.[28]

Head thrust test

The child is asked to fixate on a distant object (like a cartoon character) while the examiner turns the head rapidly by 15° in one direction and then the other. While turning to the normal side, the child's gaze remains fixed on the distant object. While turning to the abnormal side, the vestibulo-ocular reflex fails and the child's gaze turns with their head; there is then a voluntary saccadic movement of the eyes back onto the target.

Head shaking nystagmus test

Ask the child to close his/her eyes (or use Frenzel lenses), tilt the child head-down 30° then oscillate their head 20 times horizontally. Observation of nystagmus after head shaking indicates a vestibular imbalance.

Gait and gross motor assessment

Subjective evaluation of age-appropriate gait can be tested in clinic to look for gait asymmetry and ataxia. Age-appropriate motor assessments are also available.

Other considerations

The positioning test (Dix–Hallpike) may be used depending on the history although classic BPPV in children is uncommon.

What management should you offer?

Investigations

Evaluation of hearing

Audiological and tympanometric testing is vital and may include a pure tone audiogram with acoustic reflexes or auditory brainstem response depending on the age of the child.

Blood tests

Routine blood tests are rarely of value; specific blood tests may be considered as dictated by history/examination.

Imaging

While often low in yield, the threshold for imaging of the brain and internal auditory canal in any child with an unexplained vestibulocochlear abnormality or asymmetry is low. Imaging, however, may offer significant reassurance to both the child and the parent. Magnetic resonance imaging is ideal to exclude central causes and inner ear malformations, but may require a general anaesthetic in younger children. Computed tomography or magnetic resonance imaging may be used to investigate middle ear disease.

Vestibular function tests

They are especially helpful to determine if unilateral or bilateral loss has occurred and whether there is a resulting absence of function or

hypofunction. Testing is also indicated in balance disorders when it is important to differentiate between poor balance resulting from visual, vestibular, or proprioceptive disorders.[29]

Video-oculography

Video-oculography (VOG) has now replaced conventional electronystagmography and is useful in measuring spontaneous nystagmus, positional nystagmus, and horizontal canal function in response to caloric stimulation. Adaptations of VOG techniques, such as using interesting cartoon characters instead of light, have been described. These assess both peripheral and central vestibular system through the vestibulo-ocular reflex.

Other tests

Horizontal canal function can also be assessed using a computerized rotary chair. This can help clarify questionable VOG results and can be readily adapted to children by modifying adult techniques. Children less than 6 months of age do not have the capacity to supress their vestibulo-ocular reflex and therefore horizontal canal function can be probed in these children by seating them in their parent's lap and manually rotating the chair and looking for appropriate per and post-rotary nystagmus. This technique can also be used in older children in the clinic but requires the use of Frenzel lenses or VOG goggles. Computerized dynamic posturography can be useful in assessing functional balance and has been performed in children as young as 3 years.

Saccular function can be assessed by vestibular evoked myogenic potentials. In children, this can be performed by adapting standard auditory brainstem response equipment using insert earphones.[30]

Treatment

The majority of parents and children require reassurance and an explanation for the cause of vertigo. Treatment is very much dependent on cause and degree of symptoms.

For acute episodes, vestibular sedatives can be helpful; phenothiazines in children should be used with caution as they may cause extrapyramidal side effects and should not be used for prolonged periods.

For conditions like vestibular neuronitis, Ménière's disease, and BPPV the principles of treatment are similar to adults.

The following are some of the treatments that can be offered for the common causes of vertigo in children.

Vestibular migraine

Treatment of migraine in children should be managed by a paediatrician, neurologist, or physician with experience of migraine treatments in children rather than a general otolaryngologist. However, initially trigger factors (ophthalmological, sleep, stress, dietary, psychological) can be identified and addressed, accompanied by simple analgesics (paracetamol, non-steroidal anti-inflammatory drugs). If this proves insufficient an ergot preparation can be effective; in adolescents (>12 years of age), sumatriptan nasal spray is effective and should be considered for the acute treatment of migraine.[31]

Where there are more than three episodes per month, prophylaxis should be considered with propranolol or pizotifen. It should be noted that ergot preparations are contraindicated in basilar migraine for fear of causing severe vasoconstriction and stroke.[29]

Benign paroxysmal vertigo of childhood

It is important to inform and reassure parents of the benign nature of this condition of unknown cause that does not need any other specific treatment. There is a suggestion that creatine kinase levels are elevated in BPVC and its measurement may help diagnosis.[32] BPVC has a long-term favourable outlook and long-term follow-up studies have shown complete resolution.[33]

If the vertigo does not improve or progresses, magnetic resonance imaging should be considered to exclude a central cause.

Referral to a tertiary centre

A small number of children will require referral to a tertiary centre for further evaluation and treatment. This includes children who do not respond to initial management or need specialist surgical input by a neuro-otologist.

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86

Acquired paediatric airway stenoses and congenital laryngotracheal disorders

Ben Hartley and Claire Frauenfelder

Introduction

Fixed airway obstruction and congenital airway anomalies are rare but life-threatening problems affecting less than 1/10,000 children. They are usually detected in the first year of life and presentation may be insidious. The index of suspicion should remain high until the airway has been properly inspected, and the threshold for referral to a centre capable of endoscopically evaluating the airway should be low. This chapter discusses assessment and management of acquired paediatric airway stenoses and congenital laryngotracheal anomalies.

Airway development

Development of the airway and lungs begin in the third and fourth weeks of life and continues throughout the prenatal period. After the laryngotracheal sulus arises in the proximal primitive foregut in week 3, it extends caudally. Concurrently a pair of lateral tracheo-oesophageal ridges rise cranially. The resulting ventral outpouching tissue develops into the trachea and lung buds. The tracheo-oesophageal septum separates the developing trachea from the oesophagus and has formed by week 6. In week 8 a slit in the median of the primordial pharyngeal floor forms between the fourth and sixth branchial arches, giving rise to the glottic inlet, and the cartilaginous framework of the glottis then forms from within these fourth and sixth arches.

Interrupted development of the embryological aerodigestive tract gives rise to a range of abnormalities. Congenital abnormalities in the airway are rarely isolated, most frequently associated with other anomalies. Independent development of different aerodigestive tract subsites at different rates result in a variable collection and combination of possible anomalies. For example, incomplete or failure of glottic inlet canalization results in laryngeal webs or atresia; failure of closure of the posterior cricoid lamina results in laryngeal cleft; and developmental interruptions and abnormalities in the trachea and trachea-oesophageal septum result in long laryngeal clefts, trachea-oesophageal fistulae, long segment tracheal stenosis, and, in severe forms oesophageal or tracheal atresia or agenesis.

Patient assessment

A thorough medical history should be obtained including any comorbidities, obstetric history, and neonatal course (see Chapter 73 for assessment of paediatric patients for ear, nose, and throat (ENT) problems). A history of prematurity or neonatal interventions such as respiratory support, intubation, or general anaesthetics lower the threshold to inspect the airway endoscopically under anaesthetic.

Understanding the child's development is also important. Failure to thrive, poor feeding, aspiration events, recurrent chest infections, unexplained increase in work of breathing, blue spells, or frequent re-presentation for emergency department assessment by concerned parents all warrant investigation.

Delay in diagnosis may occur in these children and it is possible that the otolaryngologist is the first clinician to recognize the red flags for a serious underlying airway condition. When a presentation is concerning, admission for monitoring and assessment should be considered and involvement of paediatrician colleagues is encouraged.

Emergency presentation and airway assessment

Earlier chapters have discussed assessment and management of acute airway distress in a child (see Chapter 74). Where a child presents in extremis, a low threshold for concurrent endoscopic examination of the airway in theatre and intubation should be considered. The most experienced paediatric anaesthetist available should be involved, along with trained ENT operating theatre staff. A clear team brief should be performed, taking time to ensure all staff are aware of planned steps that will be performed should a difficult airway be encountered. Microlaryngobronchoscopy instrument set-up should be ready and tested prior to induction of anaesthesia. A rigid bronchoscope and a blade for emergency front-of-neck airway access should also be available. Procedural consent should include

emergency tracheostomy, postoperative transfer to intensive care, and prolonged intubation.

Investigations

The most important investigation of a suspected airway abnormality is endoscopic inspection under general anaesthetic. Where the patient's presentation is not emergent, additional preoperative investigations may be beneficial. A standardized and reproducible technique for microlaryngobronchoscopy should be used to record findings for all patients (Figure 86.1).

Respiratory function

In neonates or infants, overnight oximetry may be available; however, interpretation of this is challenging with poor sensitivity and specificity. Formal polysomnography and input of a paediatric respiratory physician is the standard and will determine any oxygen support required while investigation of the child's airway is underway. Respiratory symptoms may warrant lung function testing, although in a practical sense, these are more useful for older children where an acquired airway abnormality is suspected.

Swallow function

Aspiration should also be excluded as recurrent airway soiling will exacerbate respiratory dysfunction. Where history obtained from the parents raises concern regarding dysphagia or aspiration, the child's swallow should be clinically assessed by a speech and language therapist, and a videofluoroscopic swallow considered. Establishing the child's baseline swallow function is useful, particularly when assessing for a laryngeal cleft and when deciding if operative intervention is indicated.

Operation: All Procedures
Date: ??/??/????
Surgeon: Primary Surgeon — CODING(from database/PIMS)
Assistant: Assisting Surgeon(s) & Consultant-in-charge — e.g. MLB = E50.9 + E38.9
Anaesthetist: Consultant Anaesthetist

INDICATION:	Indication for the procedure Previous ENT procedures (including dates) Relevant past medical history e.g. Ex-prem (weeks/40); previous intubation; dates of tracheostomy/LTR	
PROCEDURE:	Anaesthetic details e.g. topical treatments to larynx Type of laryngoscope used +/– suspension Details of surgical intervention e.g. angle of Hopkins rod; instruments used	
FINDINGS:	SUPRAGLOTTIS View (Grade)	
		Laryngomalacia (e.g. short AE folds, arytenoid prolapse)
		Morphology (e.g.?-shaped epiglottis)
		Oedema/LPR changes
	GLOTTIS	Vocal cord appearance (e.g. nodules, papillomas)
		Laryngeal cleft
		Crico-arytenoid joint mobility
		Dynamic view
	SUBGLOTTIS	Appearance/shape e.g. cysts?
		Size (sized with ET tube and SGS grading)
		LPR changes
	TRACHEA/ CARINA	Appearance
		Tracheomalacia
		Tracheo-oesophageal fistula/pit
		Stomal assessment if required
	BRONCHI	LEFT.....& RIGHT..... (must be specifically mentioned e.g. "trachea to bronchi normal")
IMPRESSION:	Diagnosis/Summary of pertinent findings	
PLAN:	- Post-operative instructions (e.g. routine obs, humidification, steroids) - Investigations/reviews to be requested (e.g. videofluoroscopy, sleep study)-in patient vs outpatient and urgency - Discharge plan (e.g. day of discharge, antibiotic and duration) - Further procedures requested (e.g. repeat MLB) - Out patient follow up	

Figure 86.1 Great Ormond Street Hospital for Children microlaryngobronchoscopy proforma.

Reproduced from Alli A, Hewitt R, Jephson C, et al. Operative documentation in paediatric microlaryngoscopy and bronchoscopy: an audit of practice at Great Ormond Street Hospital for Children. *Otorhinolaryngologist*. 2013;6(1):64–67, with permission from Rila Publications Ltd.

Radiology

Radiological examination is rarely indicated prior to airway inspection. A chest X-ray may be useful to exclude a vascular ring or to assess lung fields.

For patients with tracheobronchomalacia, bronchoscopy and bronchogram can provide dynamic assessment of the extent of airway collapse and also be used to assist titration of airway ventilation pressure setting. Specialized centres now also incorporate dynamic or four-dimensional computed tomography scanning of the thorax when assessing for possible operative interventions (e.g. aortopexy, posterior tracheopexy, and splints).

Differential diagnoses

Acquired stenosis of the airway

Iatrogenic stenosis of the paediatric airway is a modern entity which has arisen following improvement in neonatal mortality and the advent of neonatal intensive care units in the 1960s. Prior to this, premature and unwell neonates were unlikely to survive and few were intubated for a prolonged period of time. As medical techniques improved survival of neonates, a range of airway sequalae became apparent. Intubation with large, rigid tubing resulted in injury to the larynx and vocal cords, directly causing glottic stenosis and vocal cord palsies, and to the epithelium of the airway, leading to subglottic and tracheal stenoses.

Prolonged intubation, especially if sedation is lightened and the child is able to move around, is associated with friction of the endotracheal tube against the airway mucosa, shearing it and resulting in scar formation or fixation of the cricoarytenoid joints or vocal cords themselves. The pseudostratified columnar epithelium of the subglottis is thin and delicate, and easily injured during intubation or by prolonged pressure exerted by an endotracheal tube (worse if a cuff present and inflated). Trapped between firm plastic and the underlying cartilaginous framework of the larynx and subglottic tracheal rings, pressure necrosis results, followed by oedema and ulceration, and there may be damage to the underlying and poorly vascularized cartilage. As the mucosal insult heals by secondary intention, granulation may form, followed by fibrosis and, later, mature scar.

Older children may develop stenosis following laryngeal or airway surgery such as tracheostomy; injury such as burns or blunt or penetrating trauma to the larynx or trachea; inflammatory processes associated with infection; severe reflux; or underlying conditions such as granulomatosis with polyangiitis, pemphigus, sarcoidosis, systemic lupus erythematous, or epidermolysis bullosa.

Presentation with airway symptoms usually occurs in the weeks following extubation but may occasionally be delayed and become apparent months or years later. Symptoms are usually progressive and worse on exertion. They include shortness of breath, stridor, hoarse or altered voice or cry, dysphagia, or change in swallow function due to shortness of breath while feeding or during activity.

Congenital stenosis of the larynx and trachea

Airway distress associated with congenital airway stenosis is usually noted in the first months of life unless the defect is relatively minor. Obstruction greater than 50% of the airway will be symptomatic and, depending on the subsite, may present differently.

Respiratory compromise at birth is associated with laryngeal webs and atresia. Complete laryngeal atresia is fatal unless recognized during antenatal investigation and an *ex utero* intrapartum treatment (EXIT) procedure is performed or a patent tracheo-oesophageal fistula (TOF) is present (**Figure 86.2**).

Partial occlusion by laryngeal webs or subglottic scar or cysts may present with dysphonia or altered cry, stridor, or airway distress.

Congenital subglottic stenosis is diagnosed when there is a narrowing of the subglottis in the absence of previous intubation and occurs when the airway does not accept an age-appropriate endotracheal tube with a leak at normal ventilation pressures. It is graded from 1 to 4 using the Cotton–Myer system (**Figure 86.3**). The cricoid may be elliptical in association with several syndromes (most commonly trisomy 21) or the stenosis may be membranous.

Congenital tracheal stenosis is more likely to have an insidious presentation as occlusion of 50% may remain asymptomatic, while up to 75% may be present before dyspnoea at rest is apparent. As the neonatal and infant airways are so small, even a minor change to the airway lumen will restrict movement of air. Great Ormond Street Hospital published a classification system of congenital tracheal stenosis based on anatomical variations (**Figure 86.4**). Complete tracheal rings are the most common cause and may be associated with short- or long-segment stenosis, depending on the number of affected rings. Concurrent cardiovascular anomalies such as pulmonary artery sling are common in long-segment stenosis, present in more than two-thirds of patients. Occasionally, complete absence of one or more tracheal rings will result in stenosis.

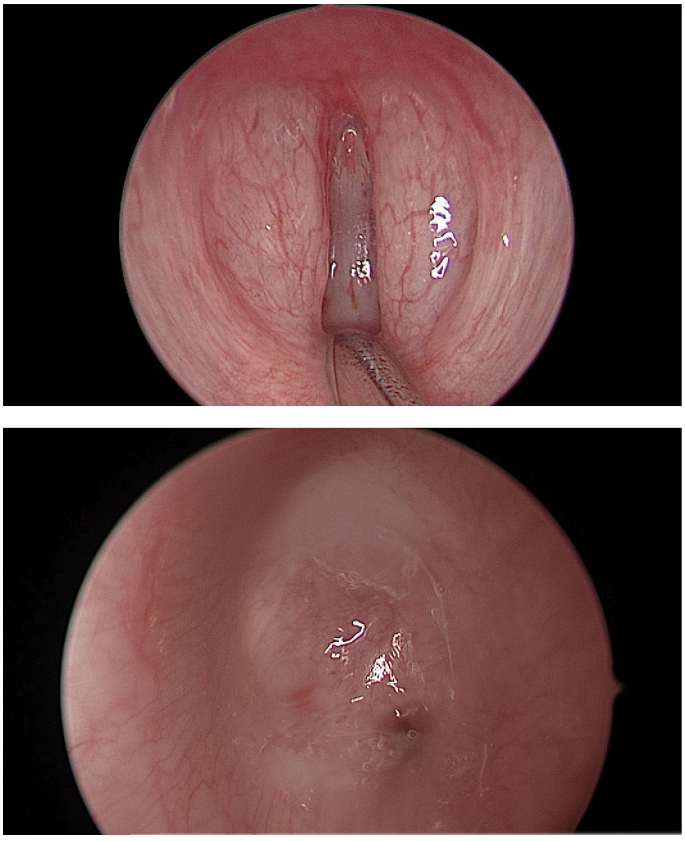

Figure 86.2 Endoscopic images of laryngeal atresia.

Classification	From	To
Grade I	No Obstruction	50% Obstruction
Grade II	51% Obstruction	70% Obstruction
Grade III	71% Obstruction	99% Obstruction
Grade IV	No Detectable Lumen	

Figure 86.3 Cotton–Myer grading of airway stenosis.

Reproduced from Hartley BE, Cotton RT. Paediatric airway stenosis: laryngotracheal reconstruction or cricotracheal resection? *Clin Otolaryngol Allied Sci*. 2000;25(5):342–349, doi: 10.1046/j.1365-2273.2000.00399.x, with permission from John Wiley and Sons.

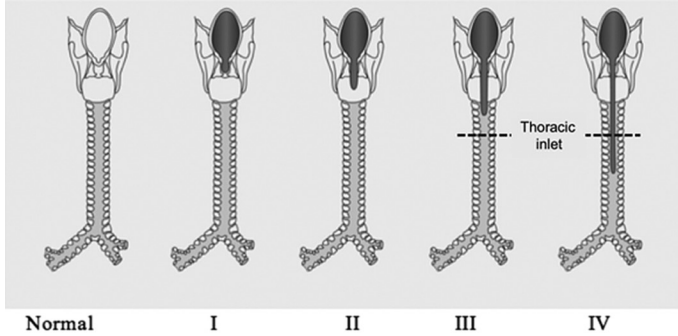

Figure 86.5 Benjamin–Inglis classification of laryngeal clefts.

Reproduced from Varela P, Torre M, Schweiger C, Nakamura H. Congenital tracheal malformations. *Pediatr Surg Int*. 2018;34(7):701–713, doi: 10.1007/s00383-018-4291-8 with permission from Springer Nature.

Laryngeal clefts

Laryngeal clefts are a defect in the posterior wall of the larynx and, in long clefts, the trachea. Clefts are classified using the Benjamin–Inglis classification (Figure 86.5) and over 50% of patients with long clefts have concurrent congenital abnormalities of some kind. Clefts are usually suspected in a patient with dysphagia, aspiration, or recurrent chest infections and short clefts may have a delayed presentation in older children.

Laryngeal clefts are diagnosed at microlaryngobronchoscopy by inspection and palpation of the posterior glottic commissure. Care should be taken when palpating as excessive pressure on the soft tissue of the posterior commissure will lead to overdiagnosis (Figure 86.6).

Type 1 clefts are present when the posterior glottic commissure is lower than normal but does not extend beyond the level of the true vocal folds. This may be difficult to distinguish from a deep interarytenoid groove. Type 2 clefts are a partial defect in the posterior cricoid lamina.

Type 3 clefts extend through the cricoid and into the cervical trachea, while a type 4 laryngeal cleft extends into the thoracic trachea and on occasion, beyond into a bronchus. The long clefts are usually identified in the early neonatal period presenting with airway

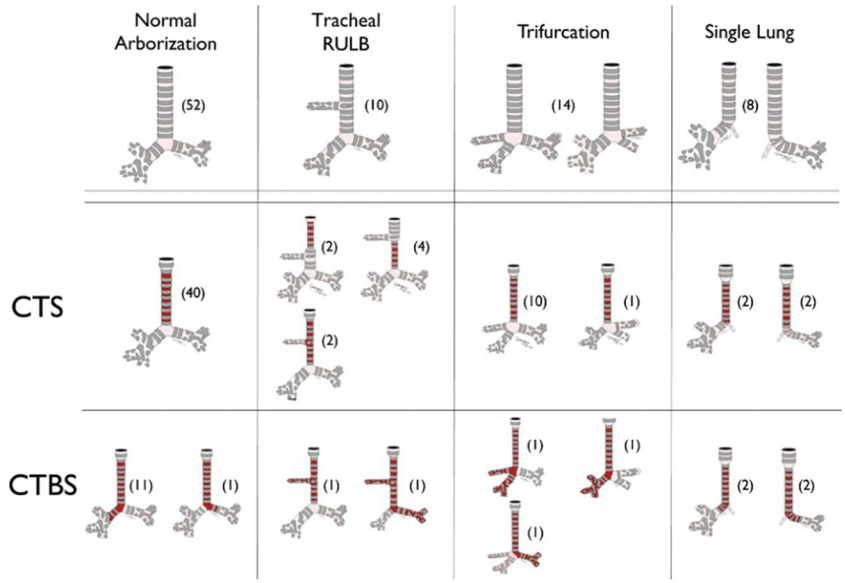

Figure 86.4 Great Ormond Street Hospital for Children classification of congenital tracheal stenosis, according to tracheobronchial arborization. The stenotic segment is pictured in red. CTBS, congenital tracheobronchial stenosis; CTS, congenital tracheal stenosis; RULB, right upper lobe bronchus.

Reproduced from Speggiorin S, Torre M, Roebuck DJ, McLaren CA, Elliott MJ. A new morphologic classification of congenital tracheobronchial stenosis. *Ann Thorac Surg*. 2012;93(3):958–961, doi: 10.1016/j.athoracsur.2011.12.019, with permission from Elsevier.

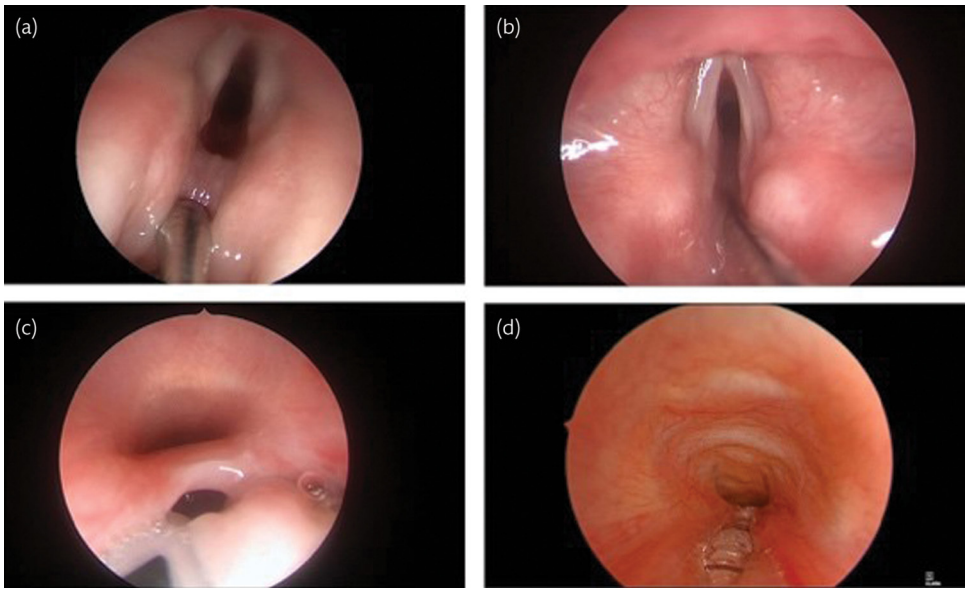

Figure 86.6 Laryngeal clefts. (a) Type 1; (b) type 2; (c) type 3; (d) type 4.

distress and severe reflux of feeds. They are commonly present in conjunction with other syndromes (Pallister–Hall, Optiz–Frias, CHARGE, or VACTERL). Due to the poor structural integrity of the trachea, airway malacia is an ongoing problem for patients with long clefts even after surgical repair. The malacia may be severe and patients frequently require non-invasive ventilation as they grow with or without tracheostomy.

Tracheo-oesophageal fistulas

TOFs with oesophageal atresia (OA) are a common congenital abnormality, occurring in approximately 1/3000 live births and associated with other congenital abnormalities in over 50% of cases. Different combination of TOF/OA were classified by Gross (Figure 86.7). Isolated TOFs without associated OA (H-type) are rare, found only in less than 4% of cases. Neonates with TOF/OA are most commonly diagnosed antenatally when polyhydramnios is seen, or in the first days of life with excessive drooling or poor feeding and inability to pass a nasogastric tube. This pathology is typically managed by specialized neonatal and paediatric surgeons (SNAPS); however, otolaryngologists often perform microlaryngobronchoscopy to assess for concurrent abnormalities and may cannulate the TOF to aid the SNAPS team's localization of TOF at the time of repair (Figure 86.8).

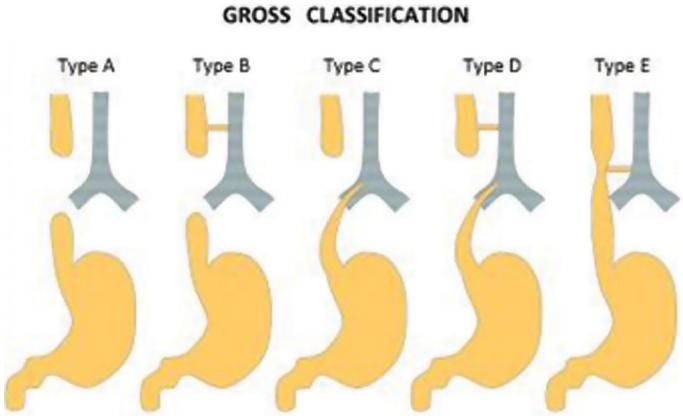

Figure 86.7 Gross classification of oesophageal atresia with or without trachea-oesophageal fistula.
From Gross RE. *The Surgery of Infancy and Childhood*. WB Saunders; 1953:76.

Tracheobronchomalacia

Tracheobronchomalacia is excessive compliance of the lower airway and may be segmental or affect the entire airway. During dynamic respiration, variable luminal airway pressures are exerted on the tracheal walls and, in areas where the cartilaginous framework is weak, collapse of the airway results. This structural abnormality of the trachea with or without bronchi may be congenital or may

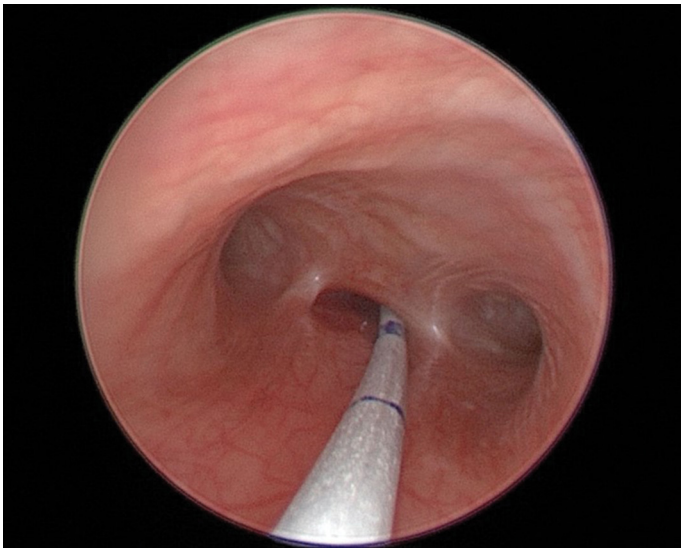

Figure 86.8 Tracheo-oesophageal fistula. Large distal TOF cannulated with a ureteric stent during microlaryngobronchoscopy to facilitate localization by SNAPS team during thoracoscopic ligation of TOF.

arise after surgery or trauma affecting the airway. Symptoms and presentation range from mild, intermittent stridor to severe with recurrent life-threatening events. Assessment of severity of airway collapse is made by a combination of direct inspection during microlaryngobronchoscopy, radiological assessment via bronchoscopy and bronchogram, or increasingly via dynamic three-dimensional computed tomography scanning of the chest.

Treatment

Techniques for surgical management of subglottic stenosis

Surgical management of paediatric subglottic stenosis can be understood by appreciating the development of operative techniques over time.

After the introduction of prolonged intubation of premature infants in the neonatal intensive care unit, high rates of glottic and subglottic stenosis were initially managed using tracheostomy to bypass the occlusion and waiting until the child outgrew the problem.

It was recognized that by improving airway management decisions in neonatal intensive care units that the rates of stenosis developing could be decreased. Rigid red rubber endotracheal tubes were exchanged for softer polyvinylchloride and cuffed tubes continue to be discouraged. Smaller tubes are selected to decrease ischaemic pressure necrosis of the glottic and subglottic mucosa. Tube taping for security and to prevent shearing movement against the airway mucosa has improved. Nasotracheal intubation is also encouraged and prolonged intubation avoided where possible. Although a slightly higher rate of posterior glottic injury and scar has resulted from the alternative tube positioning, overall incidence of postintubation injury in neonates has improved.

Airway reconstruction: decision-making and application of current techniques

Systematic endoscopic evaluation of the entire airway is important prior to airway reconstructive surgery. Airway stenosis should be sized and graded using an endotracheal tube and length of the stenosis segment(s) recorded along with its precise anatomical location. Previous surgical sites and presence of any supra-stomal stenosis or malacia should be noted. All other airway subsites from nose to carina should be assessed and all problem areas taken into consideration when planning operative intervention. Prior to major reconstructive surgery, any other subsites should be addressed if a single-stage procedure is to be undertaken, for example, adenotonsillar hypertrophy, nasal obstruction, or craniofacial abnormalities.

Early cricoid split and laryngotracheal reconstruction techniques for subglottic stenosis

Rethi was one of the earliest surgeons to describe laryngeal reconstructive techniques. In the early 1950s he described splitting the cricoid cartilage in adults who had suffered laryngeal trauma and developed stenosis to facilitate tracheostomy decannulation. Grahne, a Finnish surgeon, was the first to describe the use of this technique in children. He split the cricoid and inserted a metal stent while protecting the patient's airway with a tracheostomy.

Fearon and Cotton in Toronto in the 1970s reported a laryngotracheal reconstruction (LTR) technique achieved by augmentation of the anterior cricoid split with a cartilage graft to maintain addition luminal width. Use of both auricular cartilage with a skin island, and rib cartilage with buccal mucosa with a stent were described for airway expansion. Meanwhile in London, Evans described a laryngotracheoplasty performed by dividing the trachea from the cricoid and reconstructing it around a rolled silastic sheet using a castellated incision. Following stent removal and successful healing of the airway after either technique, the patient's tracheostomy was decannulated weeks to months later. Both techniques are variations of airway lumen expansion using cartilage. Over time the LTR became the favoured technique and decannulation rates from tracheostomy were over 90% following these procedures.

Cricoid split

Following an astute suggestion from a paediatrician colleague in 1977, Cotton successfully performed an isolated cricoid split as an alternative to tracheostomizing a neonate who had failed extubation. By undertaking the expansion procedure and stenting the airway for several days using the endotracheal tube, Cotton demonstrated it was possible to perform laryngotracheal surgery without a covering tracheostomy.

In the following decade, 70% of neonates with subglottic stenosis treated with a cricoid split were successfully extubated and did not require further airway surgery. It remains a common and practical procedure to avoid tracheostomizing neonates with isolated subglottic stenosis where there are no underlying respiratory comorbidities or oxygen requirement.

Single-stage laryngotracheal reconstruction

Once use of the endotracheal tube as a temporary stent had been established by the cricoid split procedure, it quickly followed that an airway could be expanded and reconstructed without leaving a tracheostomy *in situ*, and where a patient already had a tracheostomy, it could be permanently removed during the reconstructive procedure.

Cartilage is harvested for graft material, most commonly from the costal margin, but if only a small graft is required, it may be taken from the thyroid ala. Anterior and/or posterior cartilage grafts are placed at the site that will address the stenosis, expanding the airway lumen to accommodate an age-appropriate endotracheal tube. Typically, anterior stenosis is addressed with an anterior graft, posterior grafts for posterior glottic and subglottic stenosis, and dual grafts employed for more luminal expansion.

A midline anterior neck incision is made, incorporating the tracheostomy stoma if present and the airway exposed. The thyroid isthmus is divided for access. The airway is opened vertically at the area of stenosis inspected. If a posterior graft is required, a temporary tracheostomy can be placed or the patient ventilated via a cutdown endotracheal tube via the surgical field, facilitating access to incise the posterior wall of the airway. A limited pocket is fashioned, and the graft is placed. The patient is nasally intubated from above in a single stage procedure, or a stent placed in two-stage case and tracheostomy remains *in situ*. The anterior graft is secured, and drains placed in the neck to allow escape of air as the airway heals and prevent trapped subcutaneous emphysema or pneumothorax.

The LTR is technically robust and is reproducible between surgeons and units with a broader range of experience. It is suitable for grade 1–3 subglottic stenosis, and occasionally more severe stenosis

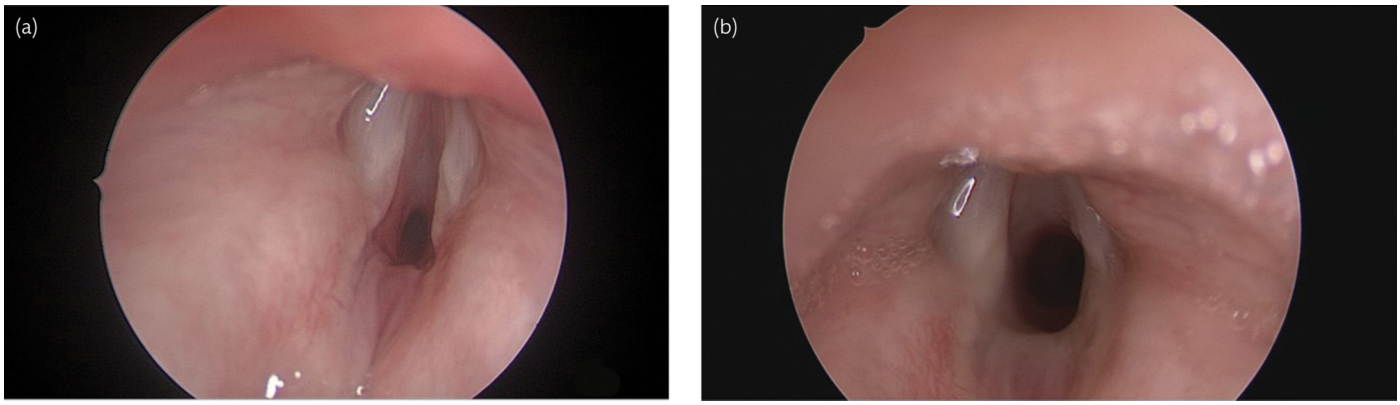

Figure 86.9 Subglottic stenosis (grade 3). (a) Preoperative endoscopic view; (b) 9 months post single-stage LTR with anterior graft.

that comes within 3 mm of the vocal cords, allowing for more precise graft placement to address reconstruction at the anterior commissure (Figure 86.9).

Following single-stage LTR, the patient remains intubated in the intensive care unit for 1–2 weeks postoperatively. Many children will tolerate a nasotracheal tube *in situ* while awake if sedation is weaned carefully in the days following surgery. This is helpful to encourage carefully supervised mobilization and deep breathing to prevent postoperative atelectasis and pneumonia. The patient is then returned to the operating theatre for microlaryngobronchoscopy to inspect the repair and downsizing of the endotracheal tube, prior to extubation the following day with systemic corticosteroid prophylaxis for 24 hours.

Cricotracheal resection

In the early 1990s, Monnier in Switzerland published a series describing the cricotracheal resection, an alternative technique taken from adult practice to increase the lumen in paediatric subglottic stenosis by resecting the stenotic segment and the anterior cricoid arch while also thinning down the posterior plate of the cricoid. The distal trachea is pulled up, telescoped over, and secured to the remaining cricoid plate posteriorly and the thyroid cartilage anteriorly. The technique is more technically challenging and results variable, with stronger outcomes achieved in higher-volume centres. While cricotracheal resection can be used in primary cases of grade 3 or 4 stenosis greater than 3 mm below the vocal folds, it usually reserved for revision cases where primary LTR has been unsuccessful.

Endoscopic surgical techniques

The re-emergence of endoscopic treatment of subglottic stenosis in recent times can be attributed to the improvement in instruments and endoscopes for visualization. Division of subglottic stenosis by cold steel or using laser coupled with balloon dilation has been successfully described. There has also been recognition that early, immature stenosis (glottic, subglottic, or tracheal) that is soft, oedematous, and yet to form fibrotic scar tissue, is amenable to serial balloon dilation. The advantages of this technique are that it can be performed at the time of endoscopic airway evaluation, the balloon exerts radial force along the length of the stenosis, and the force applied can be titrated specifically to the stenosis under direct vision and with direct haptic feedback to the surgeon as the balloon inflates. Intralesional infiltration with corticosteroid is felt by some to be beneficial, although the evidence base for this is not strong.

Endoscopic LTR with placement of a posterior graft has also been described as a single- or two-stage procedure.

Serial endoscopic inspections of the repaired subglottis should be performed regardless of reconstructive technique used to address subglottic stenosis. This method is the most effective way to monitor progress of the repair and to facilitate removal of granulation or gentle balloon dilation to address early postoperative scar tissue formation.

Medical adjuncts

Subglottic stenosis is a surgical disease; however, some medical therapies play a role as adjuncts to the surgical management.

Systemic corticosteroids to optimize conditions in the 24 hours prior to extubation have been shown to be of benefit in avoiding reintubation particularly in ex-premature infants and those with multiple comorbidities. They also play a role at the time of acute respiratory infection leading to worsening of airway symptoms in a patient with known airway stenosis, although these patients should be monitored closely as they are likely to obstruct their airway more quickly than other patients of the same age.

Optimization of any gastro-oesophageal reflux is important to protect healing of airway surgical sites, particularly in severe gastro-oesophageal reflux seen with severe structural or functional oesophageal abnormalities in children with multiple comorbidities.

Techniques for surgical management of tracheal stenosis

There is a wide variation of presentations of tracheal stenosis in terms of length and degree of stenosis. Each patient's operation is ultimately dictated by the anatomy of the stenosis, the severity of their symptoms, and their comorbidities, especially when there are cardiovascular anomalies which require concurrent surgery.

Multidisciplinary team

Thorough assessment of patients with tracheal stenosis (congenital or acquired) requires input from many health professionals, ideally coordinated by a multidisciplinary team including otolaryngologists, cardiothoracic surgeons, SNAPS, anaesthetists, interventional radiologists, respiratory physicians, physiotherapists, specialized nursing staff, respiratory technicians, and speech and language therapists.

Conservative management

Where symptoms are mild, there is justification to watch and wait for the child to grow as some studies indicate that tracheal diameter may improve with age and symptoms also improve. Concurrent vascular malformation is an indication to proceed with surgery even if symptoms are mild.

Endoscopic treatment including balloon dilation

Endoluminal division of tracheal stenosis is most effective in thin, immature acquired tracheal stenoses but may be effective in short-segment tracheal stenosis; however, it is unlikely to be of benefit where complete tracheal rings are present. Division of stenotic tissue by cold steel or laser should be undertaken with caution and may be augmented by balloon dilation. In the authors' institution, intralesional corticosteroid is infiltrated into the stenosis at each treatment.

Tracheal resection and anastomosis

Excision and end-to-end anastomosis is the treatment of choice for an isolated short-segment stenosis involving up to half of the trachea and not amenable to an endoscopic approach. In most cases the operation can be achieved via a transcervical approach as the paediatric trachea is shorter and higher within the mediastinum until approximately 10 years of age. A sternotomy can be avoided, and additional tracheal mobility can be achieved by mobilizing the posterior pericardial reflections behind the left atrium. Cardiopulmonary bypass is not usually required and, when working in conjunction with an experienced paediatric airway anaesthetist, transient intubation of the distal trachea is a suitable way to continue ventilating the patient intraoperatively after division of the trachea prior to re-anastomosis of the airway.

Slide tracheoplasty

While either a transcervical or thoracic approach can be used for slide tracheoplasty, most centres perform a sternotomy and place the patient on cardiopulmonary bypass. The stenotic tracheal segment is mobilized, including division of the thyroid isthmus to relieve tension across the final anastomosis. After reflecting away the recurrent laryngeal nerves bilaterally, the midpoint of the tracheal stenosis is transected and an oblique anastomosis is fashioned with a split of the stenosed tissue and all complete cartilage rings. This preserves the patient's own tissue, expands the intraluminal airway, but shortens the overall length of the trachea. The segments are anastomosed using everting PDS sutures and reinforced with fibrin sealant. Prior to cardiac decannulation protamine is administered and the lumen of the airway inspected using a flexible bronchoscope; once off bypass, the patient is returned to the intensive care unit. For 48 hours postoperatively the mediastinum is irrigated via drains with dilute aqueous betadine and antibiotic prophylaxis given. The airway continues to be toileted via the endotracheal tube for 24 hours then the patient is extubated where possible after re-inspection with a flexible bronchoscope.

Techniques for surgical management of laryngeal clefts

Multidisciplinary team

All patients with laryngeal clefts are in need of assessment by a multidisciplinary team. For patients with short clefts and minimal symptoms this may be a one-off review with a speech and language therapist. For other children, particularly those with long clefts and complex comorbidities that frequently require other surgical procedures, the team may involve otolaryngologists, speech and language therapists, anaesthetists, interventional radiologists, SNAPS, cardiothoracic surgeons, intensive care physicians, respiratory physicians, dieticians, physiotherapists, and specialized nursing staff among others.

Swallow dysfunction is the indication for surgical intervention in a short laryngeal cleft and is universal in child with longer clefts. In longer clefts most will have a nasogastric tube in place for feeding prior to repair and all will have ongoing need for an alternative enteral feeding route for some time due to the abnormal or absent peristatic function of the oesophagus. Consideration should be made for insertion of a gastrostomy or jejunostomy at the time of, or prior to, a long cleft repair.

Endoscopic repair of laryngeal clefts

Type 1 and type 2 laryngeal cleft repairs are usually approached endoscopically unless there are anatomical considerations that limit the feasibility of this (e.g. poor transoral access, hypermobility of the midface, etc.). Some centres also report successful endoscopic repair of type 3 clefts.

For endoscopic cleft repair the patient is anaesthetized, and the airway opened via suspension laryngoscopy using an appropriately sized Lindholm laryngoscope. The patient should be breathing spontaneously, and oxygen delivery can be via nasal prong, cut-down endotracheal tube in the oropharynx to the side of the laryngoscope, or using THRIVE (transnasal humidified rapid insufflation ventilatory exchange). Jet ventilation is rarely used in children. Edges of the cleft are resected, and the cleft is closed using a double layer of absorbable Vicryl suture on a short-half needle. An intraoperative dose of corticosteroid is followed by an additional dose during the evening postoperatively. It is ideal to avoid intubating these patients in the postoperative period where possible to protect the repair. For type 1 and type 2 clefts where the patient was having oral intake preoperatively (usually with some form of modification), their usual diet is recommenced immediately postoperatively and swallow function reassessed at 6 weeks.

Open repair of laryngeal clefts

Where cardiopulmonary bypass is not required to support an unstable patient, the patient undergoing open laryngeal cleft repair is intubated transorally. Endoscopic inspection of the airway at the beginning of the case is helpful as it is common for the endotracheal tube to displace posteriorly into the oesophagus through the cleft. Via either a transcervical approach or sternotomy, the trachea is exposed, the thyroid isthmus divided, and a partial laryngofissure is performed. A temporary distal tracheostomy can be made, or a cut-down endotracheal tube used to ventilate the distal tracheal segment via the laryngofissure; this allows the surgeon to remove and replace the tube as required during the procedure to access the distal end of the cleft during the repair. This requires close communication between anaesthetist and surgeon throughout the surgery.

The edges of the cleft are bulky and redundant tissue is infiltrated with local anaesthetic containing epinephrine (adrenaline) for haemostasis prior to its excision. Once the cleft edges have been excised, a multilayer closure is performed. The oesophageal mucosa is inverted into its lumen posteriorly and closed with a running suture, ensuring no leak. Some centres advocate interposition of graft material such as mobilized sternocleidomastoid muscle as an additional

layer between trachea and oesophagus to prevent leak, although this is not the authors' practice. Using skin hooks to secure the arytenoid cartilages, the suture line is brought to the level of the posterior commissure. The tracheal mucosa incorporating any remnant trachealis fibres are close using absorbable interrupted Vicryl sutures everting the suture line into the tracheal lumen with a tension-free closure. The patient is then reintubated with a transnasal endotracheal tube passed beyond the cleft repair. The laryngofissure is then closed directly around the tube, drain placed, and skin closed.

Early postoperative extubation is ideal to limit pressure on the healing repair and, where ongoing intubation is required, a nasal tube is preferred as it places less pressure on the repair site. Where airway malacia remains, a significant concern following cleft repair and nasal/facial delivery of non-invasive ventilation is not suitable, a delayed tracheostomy is favoured to allow for complete cleft repair healing before the tracheostomy tube is placed.

Management of tracheobronchomalacia

Treatment options for tracheomalacia and tracheobronchomalacia are dependent on the significance of airway collapse. In some patients it spontaneously resolves with growth and improved structural integrity of the tracheal cartilage with maturity. Treatment of concurrent reflux can decrease reactive inflammation in the airway, and prophylactic antibiotics can decrease frequency of respiratory infections which exacerbate respiratory dysfunction or severe malacic spells.

Non-invasive ventilation is important to maintain positive pressure within the dynamic airway and delivery can be titrated to an individual patient's needs. A tracheostomy can be used to mechanically maintain some additional integrity in the airway or as an adjunct to facilitate long-term non-invasive ventilation delivery.

The airway can be stented, however permanent stents are problematic and are avoided in children due to crusting and migration, and, as the child grows, they are unlikely to continue to be adequate. Absorbable stents play a role both as a treatment or as a temporary 'proof of concept' prior to other surgical intervention. External splinting of the airway has been used in selected cases to improve the decrease in the compliance of the airway and is advantageous in avoiding opening the airway; however, it does disrupt the tissue planes within the mediastinum.

Aortopexy and posterior tracheopexy

Operative techniques to suspend and tension the trachea externally have been successful in improving the severity of tracheomalacia. Aortopexy elevates the anterior tracheal wall via the pretracheal fascia attached to the aorta. The aorta is suspended anteriorly to the sternum, while posterior tracheopexy fixes the pars membranacea (soft tissue of the posterior airway) to the anterior spinal ligament. The airway lumen is inspected during the procedure as the endotracheal tube is withdrawn, allowing for re-tensioning until an optimal result is achieved.

Controversies/areas of future interest

Type 1 laryngeal cleft versus deep interarytenoid groove: when to operate?

Diagnosis and management of these patients may pose a challenge as many children with type 1 clefts remain asymptomatic, while conversely those with a deep interarytenoid groove may symptomatically aspirate. Decision-making about operative intervention for the posterior glottis in these two groups must include functional assessment of swallow in conjunction with a speech and language specialist.

The management options are as follows:

- *Watch and wait*: if the child remains clinically well, has no significant comorbidities, has a robust chest, and is suffering no sequelae from minor aspiration it is reasonable to defer or hold off any operative intervention.
- *Thickened fluids*: ongoing use of thickened fluids in conjunction with a speech and language therapist will decrease risk of chest infection due to aspiration but is inconvenient for families and poorly tolerated by a subset of children.
- *Temporary posterior glottic augmentation*: injection of short-acting hyaluronic acid will bulk up the tissue of the posterior commissure for several weeks and allow for assessment of the swallow with addition tissue mass at the posterior commissure. While it is not a direct comparison to surgical fixation of this area, it can inform operative decision-making and any effect from the injection will dissipate over 8–12 weeks.
- *Endoscopic repair*: may improve glottic closure during a disordered swallow and prevent soiling of the airway even if a classic laryngeal cleft is not present. Can be considered in high-risk patients where chronic airway soiling (even if small volume) may be critical to overall patient prognosis (e.g. lung transplant).

When has endoscopic treatment failed? Timing of open surgery for subglottic stenosis

Endoscopic treatment of subglottic stenosis may require several treatments before a patient improves and graduates from their treatment course. Other patients may have no response to endoscopic treatment and clearly need to proceed to open surgical reconstructive options.

There is a group of patients that fall between those who have a response to endoscopic management which is not sustained, requiring further general anaesthetics. While this course of action is reasonable for several treatments, the question should arise: when has endoscopic treatment failed?

For these patients, consideration of all aspects of their individual situation needs to be given:

- Anatomy and severity of the stenosis.
- Response to endoscopic treatment including time to recurrence of symptoms between treatments.
- Comorbidities which would increase risk associated with open surgery (e.g. craniofacial abnormalities, syndromes, other staged surgeries).
- Social circumstances and access to emergency management between endoscopic treatments.
- Parental understanding of risk versus benefit when persisting with repeated endoscopic treatments and ongoing hospital admissions, repeat general anaesthetics, and liaison with treating otolaryngologist and team.
- Option of a tracheostomy tube to bypass the airway lesion until the child grows or becomes suitable for open procedure (including

responsibility and care needs required to care for a child with a tracheostomy).

Discussion of the case with specialized paediatric otolaryngological colleagues or within an airway multidisciplinary meeting should also be considered.

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87

Tonsillectomy and adenoidectomy

Keith G. Trimble

Introduction

Historical literature on tonsillectomy reflects developments in surgical instrumentation and devices, partial versus complete removal of tissue, and observational cohort studies comparing postoperative complications between techniques. Recent publications show a shift to surgery in younger children and an increased use of specialist children's hospitals.[1,2] Drivers towards partial tonsillectomy techniques include data from the large Swedish National Patient Register showing partial tonsillectomy as equally effective in treating sleep-related breathing disorder from adenotonsillar hypertrophy[3]; the unexpected deaths of five children in Austria caused by postoperative haemorrhage in 2006[4]; and the large Evalina study showing negligible complication rates[5] and warnings about the unpredictable pharmacogenetics of codeine metabolism in children following tonsillectomy, prompting interest in a less painful surgical technique.[6] The modern trend towards faster day-case discharge (4 hours or less) and partial tonsillectomy techniques brings us nearly full circle to the early twentieth century when partial tonsillectomy and surgery on the kitchen table were accepted practice. It is easiest to consider the surgical techniques under the categories of extracapsular (total tonsillectomy) and intracapsular (partial tonsillectomy or tonsillotomy).

Tonsillectomy technique

A survey of UK surgeons in 2002 found that 12.5% still performed guillotine tonsillectomy (**Figure 87.1**), popularized in the late 1800s.[7] More recently, a large cohort study of paediatric tonsillectomy in England showed an increase in radiofrequency controlled ablation (COBLATION, referred to as 'coblation' from now onwards) usage from 7% in 2008/2009 to 27% in 2018/2019.[8] When George Waugh published his technique of complete enucleation of the tonsils or 'cold' dissection in 1909 with notes on 900 cases,[9] this was seen as an advance over the guillotine method where some tonsillar tissue remained. In his technique, the faucial pillar is incised with fine-toothed forceps to expose the white tonsil capsule superiorly, tonsils are dissected with metal instruments by blunt dissection with bleeding controlled by packing the tonsillar fossae, and the surgery is completed with an average operating time of 3 minutes.[10] One hundred years on, the National Prospective Tonsillectomy audit in England and Northern Ireland recommended all trainee ear, nose, and throat surgeons should still achieve competency in the cold steel technique and haemostasis with ties,[11] before learning other techniques. Many refinements of the extracapsular technique have developed including the introduction of 'hot' diathermy techniques, which improved the operator's experience (reducing intraoperative bleeding, and decreasing the need for ligation with point coagulation of bleeding sites) and reduced operative time for the patient. The ability to dissect and control intraoperative bleeding with the same instrument was probably the most important factor contributing towards the popularity of hot techniques, despite early suggestions that rates of secondary haemorrhage and postoperative pain were increased. There is wide variation in the cost of single-use equipment for tonsillectomy, with cold steel and monopolar cautery being least expensive by two orders of magnitude.[12] However, the most significant healthcare costs relate to operative time and postoperative readmission rate.

Figure 87.1 Tonsillectomy guillotine with sharp cutting and blunt crushing blades, introduced by Otto Popper in 1929: pictured Popper device still used in Belfast in 2020.

Extracapsular tonsillectomy

Monopolar diathermy was the most frequently reported method for tonsillectomy in the US in 2007 where more than 250,000 tonsillectomies in children occur each year.[13] Other hot techniques reported for extracapsular tonsillectomy include bipolar diathermy forceps, bipolar diathermy scissors, harmonic scalpel ultrasonic wand, carbon dioxide laser beam, KTP laser fibre (Figure 87.2), argon plasma coagulator, and bipolar radiofrequency ablation (also known as cold ablation or coblation).

Monopolar diathermy applies high-frequency alternating current, generating temperatures of 400–600°C directly to the tonsillar area, facilitating virtually bloodless removal of the tonsil from the underlying muscle. The superior pole of the tonsil is grasped with a Luc's forceps or straight Allis clamp and retracted medially to identify the lateral edge of the tonsil. A finger-switching diathermy pencil or a Colorado tip electromicrodissection needle (Figure 87.3c) is used to incise the anterior faucial pillar and the entire tonsil is dissected from the fascia overlying the peritonsillar vessels and constrictor muscle. Monopolar diathermy reduces intraoperative blood loss and the risk of immediate postoperative haemorrhage but increases the risk of secondary haemorrhage compared with cold dissection. Although rare, intraoral burns, airway fire, and other fire-related injuries are a potential risk of using monopolar in an oxygen-rich environment.

Microbipolar diathermy forceps are claimed to have better tissue effects than monopolar but increase surgical time as char needs to be cleaned from the bayonet tips. In bipolar diathermy the current passes through the tissue between electrodes in the tips of forceps or scissors, causing less lateral thermal injury (Figure 87.3e). Some operators use Ethicon PowerStar bipolar scissors to reduce operative time and intraoperative bleeding[14]; however, intraoral burns and increased secondary haemorrhage rates are associated with use of this device[15] (Figure 87.3d).

Figure 87.2 KTP laser extracapsular tonsillectomy performed by Mr S. Kaluskar FRCS, Tyrone County Hospital, Northern Ireland, 1999.
Courtesy of Mr S. Kalusakar.

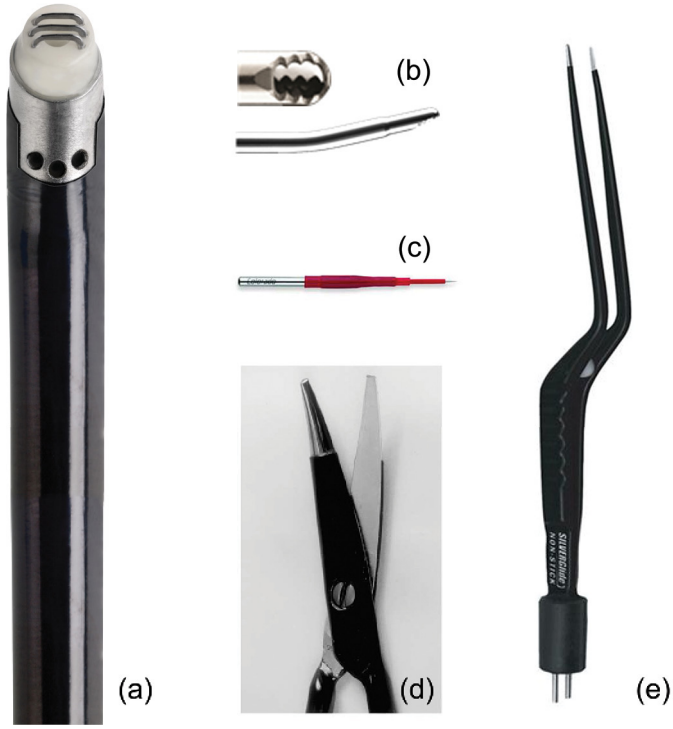

Figure 87.3 Mechanized or electrosurgical tonsillectomy wands used for intra- and extracapsular tonsillectomy by the author. (a) Procise XP Coblation wand (Smith & Nephew). (b) A 4.0 mm tonsillectomy blade for M5 microdebrider (Medtronic). (c) Colorado microdissection diathermy needle (Stryker). (d) BP520 PowerStar Bipolar Scissors, Metzenbaum 7 inch (Ethicon). (e) Bipolar diathermy Silverglide non-stick bayonet forceps (Stryker).

Intracapsular tonsillectomy

Coblation uses bipolar radiofrequency energy, rather than heat, to remove tissue (Figure 87.3a). This technique ablates and coagulates soft tissue by generating a high-energy plasma field. The ionized plasma field is generated by a radiofrequency current that passes through a normal saline medium between the device tip (electrode) and the tissue. The ionized field of sodium molecules breaks molecular bonds and produces a melting tissue effect at a much lower temperature of 40–70°C compared with monopolar. Some studies suggest that the lower temperature used in coblation results in reduced thermal damage to surrounding tissue and muscle, which in turn results in less postoperative pain.[16] Although the coblation wand was originally reported for extracapsular tonsillectomy, the combination of coblation technology with intracapsular tonsillectomy has shown most benefit with 2/500 readmission rate in a series of 500 children.[5] A Procise EZ View wand with suction channel and saline flow is used on coblate setting with a Mollison pillar retractor to protect the anterior pillar and medialize the tonsil. The tonsil tissue is coblated from medial to lateral using a light contact technique and avoiding suction blockage with denatured tissue fragments. The coblation setting is reduced when the capsule is approached and with experience a near-total technique can be attempted (Figure 87.4). Coagulation setting can be used to seal small bleeding points or denature islands of lymphoid remnants overlying

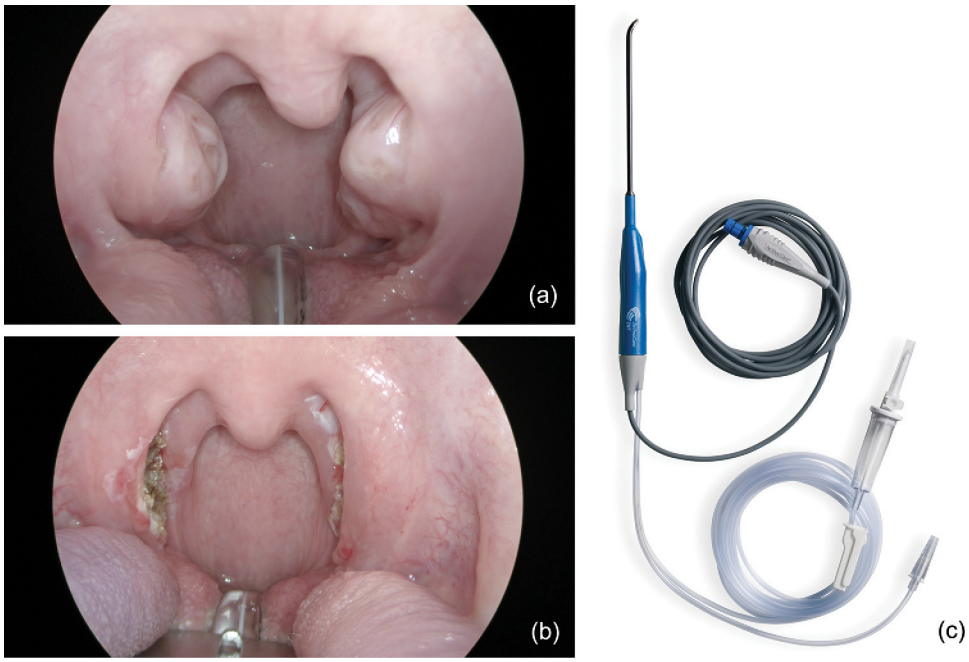

Figure 87.4 Intraoperative oropharynx images showing intraoperative pre- and post-coblation intracapsular tonsillectomy, captured using 30° 5 mm Storz telescope on Olympus VISERA 4K UHD Camera Control Unit.

the capsule. Buried tonsil tissue can cause some frustration as the view is limited unless some of the anterior pillar is trimmed; even with experience a near-complete excision onto the capsule feels slow but the objective of reducing all tissue below the tonsil crypts should be achievable.

The microdebrider was introduced for intracapsular tonsillectomies by Peter Koltai in 2002.[17] This technique removes about 90% of the tonsillar tissue with a rapidly rotating blade (**Figure 87.3**b) that suctions out excised tissue while bathed in saline and preserving the tonsillar capsule. Microdebrider tonsillectomy is associated with decreased postoperative morbidity but greater intraoperative blood loss compared with electrocautery tonsillectomies. Similar intraoperative blood loss was seen when comparing the use of the microdebrider for adenoidectomies with the adenoid curette.[18]

The National Prospective Tonsillectomy Audit reported increased haemorrhage rates for diathermy dissection tonsillectomy (adjusted odds ratio 2.47, $p < 0.0001$) and coblation extracapsular tonsillectomy (adjusted odds ratio 3.07, $p < 0.0001$) compared to the reference category of cold steel tonsillectomy.[11] Use of bipolar diathermy for haemostasis only carried an intermediate risk of haemorrhage (adjusted odds ratio 1.57, $p = 0.004$). Further investigation of diathermy power settings demonstrated a quantitative relationship between increasing power usage and subsequent haemorrhage.

Adenoidectomy

Adenoidectomy is bundled with tonsillectomy in more than 50% of cases.[8] Adenoidectomy is indicated in children when there is evidence of enlarged adenoids causing symptoms such as blocked nose, mouth breathing, or obstructive sleep apnoea. The traditional technique for removal of the adenoids continues today with blind curettage to the adenoid vegetations using a St. Clair Thompson stainless steel curette: this was the most common (70%) method reported in the National Tonsillectomy audit.[11] Suction diathermy under indirect vision with a dental mirror was popularized as a safer, faster alternative by Great Ormond Street Hospital in 1998[19] and this along with coblation are the popular methods used in specialist children's hospitals today. Power-assisted total or partial adenoidectomy with a transoral approach along with an indirect visualization through a laryngeal mirror was described in 2007 but is less popular.[13]

Anaesthesia

Day surgery is optimal for most children and there are clear guidelines for the provision of paediatric anaesthesia, which can be delivered in district general hospital for a large cohort of otherwise well children.[20] However, anaesthesia for tonsillectomy in a children's hospital can be challenging for the child with low weight, craniofacial syndrome, cardiac anomalies, or severe obstructive sleep apnoea with pulmonary hypertension. Good communication between the surgeon, anaesthesiologist, and the parents of the child is essential for a smooth perioperative journey. The goals are good preparation of the child with familiarization or premedication as required beforehand, good oropharyngeal access for the surgeon in the 'shared airway', prevention of postoperative nausea, optimization of perioperative analgesia, and minimizing risk of laryngospasm or distress on extubation. The anaesthetist should be prewarned of the severity of obstructive sleep apnoea syndrome and vigilant for respiratory complications.[21] Although it is better to postpone surgery for a child with

intercurrent respiratory illness, infection is common in the winter season and airway reactivity can impact the anaesthetist's decision to extubate deep or awake. Intravenous anaesthetic drugs, avoidance of blood and secretions in the upper airway with good surgical technique, careful suctioning, and communication with the anaesthetist can facilitate a smooth emergence. Incidence of laryngospasm in these children is higher outside the children's hospital centres.[22] High rates of day-care tonsillectomy are expected in children and this may involve increasing use of the preoperative assessment team to facilitate patient selection. Liberalization of preoperative fasting regimens with fluids up to 1 hour beforehand can reduce dehydration. Paracetamol and non-steroidal anti-inflammatory drugs are commonly used and supplemented with short-acting intraoperative opioids. Dexamethasone is effective for postoperative pain, nausea, and vomiting.

Pain

Oropharyngeal pain is significant and near universal regardless of the method used for surgery. It can lead to poor oral intake and dehydration. It can also delay discharge from the hospital or readmission for pain control and intravenous fluids until the patient can maintain adequate fluid intake. Patients are usually sent home on oral paracetamol and non-steroidal anti-inflammatory drugs including ibuprofen or diclofenac. Following warning about opioids in children with sleep apnoea symptoms, codeine is banned in children aged under 18 years having tonsillectomy for obstructive sleep apnoea.[23] This is attributed to the genetic variation of the cytochrome P450 enzyme CYP2D6 and its association with ultrarapid metabolism of codeine, leading to increased levels of its active metabolite, morphine. These increased levels of morphine can cause respiratory depression and death; a US Food and Drug Administration adverse event review revealed 13 cases of paediatric deaths (ten) or respiratory depression (three) attributed to therapeutic use of codeine.[6]

Complications

Tonsillectomy and adenoidectomy are relatively safe procedures, but immediate, early, and late complications do occur. Postoperative complications include anaesthetic and airway risks, mandible dislocation, oral burns, pulmonary oedema, aspiration, postoperative haemorrhage, atlantoaxial subluxation, Eustachian tube injury, nasopharyngeal stenosis, velopharyngeal insufficiency, and taste disturbances. Mortality rates for tonsillectomy have been estimated between 1/16,000 and 1/35,000 and these deaths are attributed to bleeding, aspiration, cardiopulmonary failure, electrolyte imbalance, or anaesthetic complications. Retrospective review of more than 300,000 paediatric tonsillectomy cases in England showed mortality of 1:27,000.[8] Complications are more common in patients with craniofacial disorders, Down syndrome, cerebral palsy, major heart disease, or bleeding disorders and in children younger than 3 years old with proven obstructive sleep apnoea. A 2007 systematic review identified uncommon postoperative complaints including odynophagia, nausea, vomiting, referred otalgia, fever, and dehydration. Referred otalgia and loss of taste over the posterior third of the tongue is secondary to temporary effects on the glossopharyngeal nerve. Other rare complications from a 2017 systematic review include subcutaneous emphysema, mediastinitis, Eagle syndrome, and atlantoaxial subluxation. These unusual events should be considered during patient-focused consent discussions.[24] Both Austria and the UK have published increased postoperative readmission rates for unknown reasons.[4] In England, the 2019 Getting it Right First Time (GIRFT) report noted high 30-day readmission rates after tonsillectomy in children: 9.4% of 48,000 procedures in over a 17-month period starting April 2015.[25] A Scottish study in 2021 showed increases in 30-day readmission rates (12.1%) and return to theatre rates (1.7%) compared to 15 years earlier.[26]

Haemorrhage

The most frequent serious complication of tonsillectomy is post-tonsillectomy haemorrhage, occurring at a rate between 0.28% and 20%. Retrospective review of more than 300,000 paediatric tonsillectomy cases in England showed a 0.4% rate of primary haemorrhage requiring return to theatre and a 5.1% rate of secondary haemorrhage requiring return to theatre.[8] The wide range of post-tonsillectomy haemorrhage rates reflects the diversity in the otolaryngological community on how to define significant post-tonsillectomy haemorrhage. All patients with bleeding after tonsillectomy should return for clinical evaluation and any patients requiring intervention should be recorded as having a significant haemorrhage.[27] Haemorrhage is divided into primary haemorrhage within 24 hours of surgery and secondary haemorrhage occurring after 24 hours, most commonly on postoperative days 5–10. The cause of primary haemorrhage is generally attributed to inadequate haemostasis during the procedure. Secondary haemorrhage is thought to result from sloughing of the superficial eschar from the tonsillar fossa and/or infection. Postoperative haemorrhage after tonsillectomy may or may not require haemostasis in the operating room. Immediate postoperative bleeding after adenoidectomy is very unusual following suction diathermy or coblation techniques. However, patients with significant bleeding should be taken back to the operating room for examination of the nasopharynx and haemostasis.

Postoperative pulmonary oedema

This can occur in patients with a history of obstructive sleep apnoea and cor pulmonale secondary to adenotonsillar hypertrophy or, more often, following severe laryngospasm as a negative pressure phenomenon. These patients require close observation postoperatively and continuous pulse oximetry.

Hypernasal speech

Hypernasal speech immediately after surgery is not uncommon. This occurs secondary to pain, which restricts movement of the tonsillar pillars and soft palate; this is usually transient and resolves within several weeks.

Velopharyngeal insufficiency

This is an unusual complication related to adenoidectomy. Patients present with hypernasal speech and reflux of fluids through the nose. Patients may require evaluation and management by speech pathology. Submucosal cleft palate is defined as the triad of a bifid uvula, notching of the posterior border of the hard palate, and a muscular diastasis of the velum (zona pellucida) (see Chapter 89). If there is any doubt about the function/structure of the palate, a

superior adenoidectomy is performed where only the choanal part of the adenoid tissue is removed. This helps to relieve adenoid obstruction and at the same time helps to preserve palatal function.[28] Children with a history of cleft palate or submucosal cleft should only undergo adenoidectomy after full discussion with family about risk/benefit profile. Surgical intervention for velopharyngeal insufficiency is reserved for patients who do not respond to speech therapy for at least 1 year.

Nasopharyngeal stenosis

This is a rare complication after adenoidectomy/tonsillectomy and in almost all cases is related to excessive monopolar diathermy at the time of surgery. Patients with nasopharyngeal stenosis have scarring of the uvula extending to the posterior pharyngeal wall, resulting in partial or complete obstruction of the nasopharyngeal airway. Care must be taken when removing lymphoid tissue from the torus tubarius and lateral nasopharynx during an adenoidectomy.

Atlantoaxial subluxation (Grisel syndrome)

This is a rare complication of adenoidectomy/tonsillectomy, resulting from laxity of the anterior transverse ligament between the atlas and axis in the cervical spine. Patients can present with a stiff neck and typically hold their head to one side with slight rotation towards the opposite side. However, the symptoms and signs can be insidious over weeks or months afterwards with reduced mobility or long tract signs such as ankle clonus/hyperreflexia. Radiographic evaluation of the anteroposterior and lateral cervical spine with flexion–extension may aid in the diagnosis. Computed tomography or magnetic resonance imaging may be necessary to confirm. Most cases of atlantoaxial instability are secondary to infection or trauma. Management may require intravenous antibiotics and possible cervical stabilization. Patients with Down syndrome are more susceptible to traumatic atlantoaxial subluxation after tonsillectomy. Extra care is taken with cervical spine manipulation during surgery in patients with Down syndrome. Cervical flexion/extension films are not indicated in these patients if the neurological examination is normal before surgery; however, the family should be counselled to look for symptoms afterwards.

Intracapsular versus extracapsular debate

Despite the volume of published studies, there is still no consensus on the best method of tonsillectomy. The death of five children in Austria around 2007 prompted a prospective study of all surgeries in that country over 9-month period.[4] It identified postoperative haemorrhage in 15% children after extracapsular tonsillectomy and 2% after intracapsular tonsillectomy. In 2017, a Cochrane review included 29 studies in children and adults and concluded that current evidence for techniques of tonsillectomy was of very low quality, therefore it was uncertain whether or not the coblation technique had any advantages over traditional tonsillectomy techniques.[16] There was no clear difference in adverse events in the included studies. Randomized controlled trials using consistent, validated outcome measures were recommended to establish whether the coblation technique had a benefit over other methods. However, the Cochrane review only included adult patients and specifically excluded the intracapsular tonsillectomy technique, probably the main variable contributing to reduced postoperative pain and bleeding in young children. In 2017, a single centre prospective study of 500 cases using coblation intracapsular tonsillectomy was published, showing very low rates of complications, mostly in very young children.[5] In 2017, a meta-analysis of randomized controlled trials concluded that, when compared to extracapsular tonsillectomy, intracapsular tonsillectomy demonstrated a reduced incidence of postoperative haemorrhage, reduced postoperative pain, reduced need for analgesics, more rapid rate of return to normal diet, and fewer days before returning to normal activity.[29] Despite this evidence, a 2020 survey of American Society of Pediatric Otolaryngology members who primarily treated children showed only 20% of those responding performed intracapsular tonsillectomy, mostly using the microdebrider blade.[30] It should be noted that the survey response rate was only 42% so the percentage performing intracapsular tonsillectomy may have been even lower. Notably 50% of members of the American Society of Pediatric Otolaryngology still prescribed opioid analgesia for postoperative pain in children despite earlier clinical practice guidelines.

Conclusion

Arguably the main goal for quality improvement in children with tonsil conditions should be to educate clinicians, patients, and/or caregivers regarding the indications for tonsillectomy and the natural history of tonsil conditions. However, when surgery is indicated, the tide in Europe is swinging back to partial tonsillectomy/intracapsular technique in children, obviating the need for take-home opioid prescriptions and facilitating day-case procedures and earlier return to normal activity. Paediatric tonsillectomy is being performed in younger children, increasingly for sleep-related breathing disorders and increasingly in specialist children's hospitals. GIRFT has published UK guidance on which children should be safely treated closer to home and which should be transferred to tertiary centres. There is a paradoxical increase in hospital readmission and return to theatre rates, and it may be that increasing expectations of families, reduced use of codeine-based analgesia, constitutional differences in patients, and/or surgical technique are contributing. Patient-focused discussions on consent should now include detail of tonsillectomy technique, uncommon but significant complications, and local surgeon complication rates, which should be audited annually. There should be clear local guidelines for day-case adenotonsillectomy to decrease the rate of unscheduled admissions. With the increasing evidence for partial tonsillectomy techniques, there is room for a large observational study to confirm minimal complications and faster return to normal activity for children meeting criteria for tonsillectomy.

UK surgeons should continue to audit their tonsillectomy results.[31]

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88

Otitis media with effusion

Mat Daniel

Introduction

Otitis media with effusion (OME, glue ear) is characterized by the presence of fluid in the middle ear cavity in the absence of symptoms or signs of acute inflammation. It is a common cause of hearing loss in children, and its treatment with ventilation tube (VT, grommet) insertion is one of the commonest surgical procedures in children.

Epidemiology

OME is a common condition. In the general population peak prevalence is at about 2 years of age, when it is estimated that as many as 20% of screened children will have OME; there is a second smaller peak at about 5 years when 17% will have OME.[1] Before school age, up to 90% of children will have at least one episode of OME, and they develop on average four episodes per year.[2]

Risk factors

- Age.
- Male sex.
- Ethnicity: Alaskan Eskimo, American Indian, Australian Aboriginal.
- Atopy.
- Day care attendance.
- Family history.
- Cleft palate.
- Down syndrome.
- Seasonal variation—worse in colder months.
- Second-hand cigarette exposure.
- Premature birth.
- Lack of breastfeeding.

OME is much more common in children with Down syndrome (see Chapter 90) or cleft palate (see Chapter 89), with prevalence reaching between 60% and 85%. In general, the presence of an additional sensory, physical, behavioural, or cognitive impairment may lead to OME having a disproportionally large impact on the child's overall functioning.

Rates of VT insertion vary widely, and concerns about overuse are a source of much political interest. In Denmark, 29% of children will receive VTs (for any indication) before their fifth birthday,[3] but in the UK, epidemiological modelling suggests that only half the number of children that might benefit from VTs actually receive them.[4]

Natural history

For most otherwise healthy children, OME is a temporary phenomenon and most episodes resolve within 3 months. The mean duration of unilateral effusions is about 5–6 weeks, and the mean duration of bilateral effusion is 6–10 weeks.[5] When discovered on screening of asymptomatic children, OME resolves in 63% by 3 months and in 88% by 1 year.[6] However, about a third will have repeated episodes, and 5–10% of episodes last at least 1 year.[2]

OME is defined as chronic after 3 months. Once bilateral OME has been present for 3 months, the spontaneous resolution rate is much lower,[6,7] only 27% at 6 months; half will still have OME 3 years later, and the median duration of OME is 7.8 years.

Aetiology

Pathologically, OME is a chronic inflammatory condition affecting the middle ear mucosa resulting in mucus secretion that accumulates in the middle ear cleft.[8] The middle ear mucosa constantly secretes mucus, which is then moved down the Eustachian tube. The mucus has a protective function, as it provides a physical barrier and prevents adhesion of bacteria to mucosa. Upon irritation, the mucosa responds by producing an increased amount of mucus, and by producing mucus that is more viscous; OME is an exudate, not a transudate.

The aetiology of OME is complex and multifactorial. A variety of stimuli have been studied as possible initiators of the inflammation,[9] including acute bacterial infection, chronic bacterial biofilms, allergy, reflux, viruses, and exhaust fumes.

A wide range of bacteria have been implicated in OME, including *Streptococcus pneumoniae*, *Moraxella catarrhalis*, *Haemophilus influenzae*, and *Staphylococcus aureus*; bacteria existing as biofilms (groups of bacteria attached to a surface), as opposed to free-floating

planktonic bacteria, are thought to be a key driver of inflammation. Hypoxia might initiate inflammation, or may itself develop as a result of inflammation. Genetic polymorphism will lead to variability in how an individual responds to inflammatory stimuli.

Eustachian tube dysfunction may lead to impaired mucus clearance from the middle ear, or paradoxically it may allow reflux of nasopharynx bacteria into the middle ear instead of preventing it. Eustachian tube function will be influenced by age and anatomy, and affected by passive smoking and respiratory infections. The adenoid serves as a reservoir of nasopharynx bacteria, from where bacteria may leave to ascend to the middle ear via a dysfunctional Eustachian tube.

Diagnosis

Children with OME present with hearing loss, speech delay (typically mispronunciation rather than lack of vocabulary), educational concerns, poor balance, ear discomfort, and sometimes associated with episodes of acute otitis media.[2] Less commonly, OME may be associated with structural tympanic membrane changes.[2]

Presentation in primary and secondary care can be quite varied. Because OME is so common, any symptom attribution needs to be correlated with hearing level and OME persistence. Clinicians should enquire about length of symptoms, degree of concern, previous hearing tests/concerns, risk factors, family history, past medical and birth history, and presence of other ear, nose, and throat issues. Clinical examination can also note mouth breathing, rhinorrhoea, or craniofacial abnormalities.

OME has a significant impact on the quality of life of the child and their family, over and beyond just hearing. For example, when prospectively recorded, three-quarters of children are noted to have ear pain, two-thirds sleep disturbance, half behavioural problems, and 15% poor balance.[2]

The classic otoscopy findings in OME are a dull, yellowish drum with radial injection of the drum vasculature, but there is a wide range of clinical appearances. Diagnosing OME on otoscopy can be difficult, especially in an uncooperative child. Otoscopy can reliably make the diagnosis if carried out by an experienced operator, as can pneumatic otoscopy and tympanometry; tympanometry and pneumatic otoscopy both have sensitivity in excess of 90%, but tympanometry has lower specificity (50–75%) than pneumatic otoscopy (80%).[2] On audiometry, OME may cause conductive loss with thresholds ranging from normal to moderate loss (0–55 dB hearing level (HL)). On average, OME causes hearing loss of about 28 dB HL, but approximately a fifth of children have hearing loss worse than 35 dB HL.[2]

Shared decision-making

OME is a common condition, with a fluctuating and eventually self-resolving course. The functional impact that hearing impairment causes is highly variable, and dependent on the environment that a child is growing up in. Available treatments are not risk free, and often effective for short time periods only.

Thus, any decision-making needs to see beyond the hearing test, and take account of the child and their environment as a whole. As might be expected, parents/guardians value printed information and ability to discuss with friends and family, and dislike paternalistic attitudes[10]; parents/guardians are especially influenced by efficacy and the speed at which benefit is noted, risks, social implications, and personal experiences. Presenting parents/guardians with a full range of treatment options suitable and individualized for their child, including conservative management if appropriate, with attendant benefits and risks, will allow them to make a decision that is in the child's best interest.

Management

Management of OME has been the subject of much research including meta-analysis, and translated into national guidelines such as the UK National Institute for Health and Care Excellence (NICE)[11] and American Academy of Otolaryngology–Head and Neck Surgery Foundation/American Academy of Pediatrics/American Academy of Family Physicians guidelines.[2]

This chapter deals with management of isolated OME in children aged up to 12 years. Treatment of OME associated with acute otitis media or retraction or tympanic atrophy is not considered here, and may be different to what is discussed here.

Management is dictated by the presence of hearing loss and developmental/educational effects of that hearing loss. In the absence of hearing loss and developmental/educational concerns, treatment would not usually be required unless there are other conditions/symptoms associated with OME.

Some children are more at risk of the effect of OME on speech, language, or learning, for example, those that have associated sensory, cognitive, physical, or behavioural impairment. Such children may benefit from screening for the presence of OME, and more close observation than for otherwise healthy, typically developing children. Screening for OME is not recommended in otherwise healthy, typically developing children[2] due to the common and spontaneously resolving nature of OME.

It is often asked whether OME leads to long-term tympanic membrane changes, and if so do VTs act to prevent tympanic membrane structural changes. However, meta-analysis has shown that VTs actually increase tympanic membrane pathology.[12] Therefore, it is not appropriate to suggest treatment to try and affect the tympanic membranes in the long term, treatment of OME should be focused on the here and now; note this may not apply to management if OME is already associated with structural tympanic membrane changes or repeated infections.

Watchful waiting

Given that OME commonly resolves spontaneously, a 3-month period of observation is recommended by guidelines and backed up by evidence of the natural course of the disease. Even after 3 months, if OME is still present but hearing is unaffected, or there is little functional impairment, continued observation is appropriate for most knowing that OME is not leading to adverse educational/developmental effects or significant symptoms.

During watchful waiting, the following strategies minimize the effect of OME on the child:

- Attract the child's attention before speaking.
- Be close to the child when speaking.
- Face the child when talking.
- Reduce background noise.
- Use short, simple sentences.
- Talk at a slightly slower pace than normal.
- Read to and with the child.
- Repeat words when misunderstood.
- Sit the child at the front of the class and ensure teachers are aware.

Ventilation tubes

When a decision is made to manage OME surgically, VT insertion is recommended by NICE.[11] However, American guidelines recommend VTs in children aged under 4 years, but VTs or adenoidectomy, or both, in children aged 4 years and older.[2]

VT insertion is a relatively simple procedure. Typically it is carried out under general anaesthesia, although some perform surgery without, and newer insertion devices may make surgery even quicker.

VTs have a small effect on hearing (when measured using standard hearing tests), which persists for the duration of tube stay time. Once the tube extrudes, hearing is where it would be if no treatment had taken place.[13] Hence, VTs are a 'temporary solution for a temporary problem'. At least a quarter of children will require further surgical treatment within 2 years of their first VT insertion.[14]

Complications of VT insertion are relatively common.[12] Otorrhoea occurs in 16% of patients in the postoperative period, and in 26% during later periods. Up to 7% of tubes become blocked, 5% develop granulation tissue, and 0.5% migrate medially. Tympanosclerosis is seen in 32%, focal atrophy in 25%. Perforation rates with short-stay tubes are in the region of 2.2%, but as high as 17% with long-stay tubes.

A Cochrane review[15] concluded that postoperative otorrhoea can be reduced by saline washouts at surgery, a single application of antibiotic/steroid drops, prolonged application of drops, or prolonged application of oral antibiotics. Postoperative otorrhoea rates are variable, so if any prevention is needed because of local high rates, this should be with saline irrigation or a single application of drops.

The treatment of VT-associated otorrhoea is most effective when topical antibiotic or antibiotic/steroid drops are given, rather than with systemic antibiotics.[16,17]

In most children, water precautions have no impact on VT-associated otorrhoea.[18] While ear plugs may reduce infection rates, the effect is small, and it is estimated that a child would have to wear ear plugs every day for 2.8 years to prevent one episode of otorrhoea. On balance, the cost and inconvenience of water avoidance do not appear to justify any small benefit.

Adenoidectomy

Adenoidectomy has been investigated as a sole treatment for OME, or as an adjunct to VT insertion (see also Chapter 87).

When used in addition to VT insertion, adenoidectomy roughly halves the need for further surgery.[14,19] However, it is not simple to extrapolate this and state that everyone should have adjuvant adenoidectomy, because adjuvant adenoidectomy was not necessary in those who didn't have OME recurrence, nor effective in those who did have recurrence but adenoidectomy did not prevent it. Previous guidelines recommended adding adenoidectomy to a second set of VTs when OME returned, but this appeared to be an extrapolation of evidence rather than the result of actual studies.

A Cochrane review in 2010 concluded that adenoidectomy has a significantly beneficial effect on OME resolution, although only a small benefit to hearing.[20] More recent attempts to determine who benefits from adenoidectomy have identified that adenoidectomy is most effective against OME in children aged 4 years and over, and not in younger children[21]: this meta-analysis showed that 49% of children aged 4 years or over that had adenoidectomy for OME were symptom-free at 12 months, compared to only 30% of those who did not have adenoidectomy. Note that even with adenoidectomy, 51% of children in this meta-analysis still had symptoms at 12 months after treatment.

The American guidelines recommend considering adenoidectomy in addition to VTs in children aged 4 years or over, as a means of reducing OME recurrence. In younger children, adjuvant adenoidectomy is not recommended unless there are specific non-otitis symptoms or signs to warrant it.[2] In contrast, current UK guidelines recommend considering adding adenoidectomy to VTs, unless assessment indicates an abnormality with the palate.[11] Both guidelines emphasize the importance of shared decision-making, given that adenoidectomy has risks additional to VTs alone. Further, the benefits of adenoidectomy have been studied at population level, but it is currently not possible to predict exactly whether a specific child in clinic would benefit.

Autoinflation

The opening of the Eustachian tube in order to aerate the middle ear can be achieved in a variety of ways, including the Valsalva manoeuvre, the Otovent device, and the EarPopper device. A Cochrane review has concluded that autoinflation is effective in the short term,[22] and NICE recommends that autoinflation can be considered in the 3-month active observation period in children who are likely to be able to cooperate.[11] A recent study in children aged 4–11 years found that those using a nasal balloon were more likely to have normal tympanometry at 1 month (47% vs 36%) and at 3 months (50% vs 38%), with compliance being 89% at 1 month and 80% at 3 months.[23]

Hearing aids

Hearing aids are a valid non-surgical alternative to treat OME, and are recommended by NICE when surgery is contraindicated or not acceptable to the child/parents/guardians.[11] While hearing aids may help hearing, they may not help all symptoms of OME such as ear discomfort. Hearing aids appear to be well accepted by children.[24]

The fluctuating nature of hearing loss in OME may make fitting of air conducting hearing aids difficult, and the use of a commercial bone conducting headset has recently been described as an alternative for children with OME.[25]

Not recommended treatments

The following are not recommended as management of OME:

- Systemic corticosteroids.
- Intranasal corticosteroids.
- Antibiotics—a Cochrane review concluded that oral antibiotics are associated with OME resolution, but effect on hearing was uncertain, and they are also more likely to lead to adverse effects. The risks outweigh any small benefits.[26]
- Antihistamines.
- Decongestants.

Conclusion

OME is a common condition with a fluctuating course that often causes no significant symptoms. Practice guidelines advise on recommended treatments, with VTs, hearing aids, adenoidectomy, autoinflation, as well as watchful waiting being reasonable choices.

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89

Cleft and craniofacial abnormalities

Louisa Ferguson

Introduction

Cleft lip and/or palate (CL±P) is the commonest congenital craniofacial malformation and has an incidence of around 1:700, resulting in about 1000 new cases in the UK each year. Treatment of patients with a CL±P is a lifelong commitment and is best done in a multidisciplinary setting, containing surgeons, speech therapists, orthodontists, dentists, psychologists, otorhinolaryngologists, and audiologists. An understanding of the diagnosis and treatment of children born with CL±P is important for the ear, nose, and throat (ENT) surgeon as there are multiple ENT issues including hearing, airway, sleep, speech, and nasal issues with which these patients can present.

The formation of the lip and palate involves a complex set of steps which start early in gestation and are complete by week 12. Different anatomical variations of CL±P are possible from a developmental standpoint and various systems are used for classification. The most widely used in the UK is the LAHSHAL system, which involves starting with the right lip and marking the cleft status back to the palate and then forward to the left lip. Each area is marked as complete (C), incomplete (I), or no cleft (X) (**Figure 89.1**). It is important to accurately record the type and extend of the cleft, especially for epidemiological and surgical outcome reasons. Broadly speaking, clefts can be of the lip, palate, or both; they can be bilateral or unilateral; and complete or incomplete. Typically, the distribution of babies born is 46% cleft lip and palate together, 33% isolated cleft palate, and 21% isolated cleft lip.

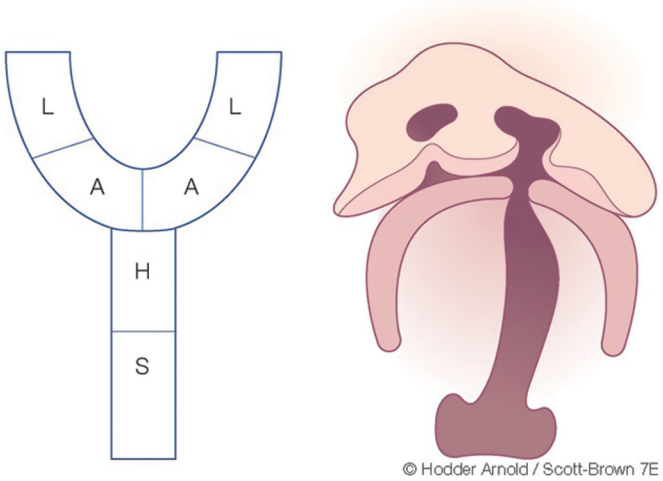

Figure 89.1 LAHSHAL system. Involves starting with the right lip and marking the cleft status back to the palate and then forward to the left lip.

Reproduced from Wynne D, Ferguson L. Cleft lip and palate. In: Watkinson JC, Clarke RW, eds. *Scott-Brown's Otorhinolaryngology and Head and Neck Surgery*. 8th ed, vol 2. CROC Press; 2018, with permission from CRC Press.

Functional anatomy

An understanding of the normal anatomy of the lip, nose, and palate is essential for returning form and function, which are the primary goals in the repair of any CL±P.

Lip

The oral sphincter is formed by the orbicularis oris, which forms a complete ring under the mucosal covering of the lip. In a cleft lip this complete sling is disrupted, resulting in abnormal insertion of the orbicularis oris muscle, and therefore lack of integrity of the sphincter mechanism. This is seen in **Figure 89.2**. The lip is well defined into three areas: the cutaneous skin of the upper lip, the dry mucosa of the vermilion, and the moist mucosa of the sulcus of the internal lip. There is loss of continuity of the vermilion border and shortening of the philtrum on the cleft side, resulting in malrotation of cupid's bow. In children with a bilateral cleft lip, the orbicularis oris muscle is deficient in the central prolabium. As the central premaxilla and prolabium are essentially free of muscle detachments, this area protrudes forward unopposed.

Nose

Due to the abnormal insertion of the orbicularis oris muscle, the nose takes on a typical deformity associated with cleft lip. The lower lateral cartilage on the cleft side is pulled out by the abnormal insertion of the orbicularis oris into the nasal base on that side. This results in a flattened lower lateral cartilage, and the alar base placed more lateral and inferior than it should be on the cleft side. The lower lateral cartilage takes on a different shape, with the medial crus shortened and the lateral crus longer and flatter. These abnormalities result in a lower dome on the cleft side and a wider nasal base. The columella

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Figure 89.2 (a) Nasolabial muscle ring in unilateral cleft lip. M, myrtiform head of nasalis muscle; O, oblique head of orbicularis oris; T, transverse head of nasalis muscle. (b) Photograph of a child with cleft lip.

(a) Reproduced from Wynne D, Ferguson L. Cleft lip and palate. In: Watkinson JC, Clarke RW, eds. *Scott-Brown's Otorhinolaryngology and Head and Neck Surgery*. 8th ed, vol 2. CROC Press; 2018, with permission from CRC Press.

is shortened on the cleft side and the anterior cartilaginous columella is deviated to the non-cleft side, due to the unopposed pull of the abnormally inserted orbicularis oris into the columella. The posterior septum is deviated to the cleft side, resulting in an S-shaped deformity. An example of this is seen in Figure 89.2.

Palate

The palate is made up of the hard palate and the soft palate. The hard palate consists of the fused lateral palatal shelves that join anteriorly at the premaxilla. The vomer sits on the palatine shelves, and is typically not seen intraorally; however, if the cleft of the palate extends into the hard palate it can be seen through the hard palate cleft. The velum, otherwise known as the soft palate, extends from the posterior aspect of the hard palate to the uvula. Anteriorly (closest to the hard palate) is the palatine aponeurosis, which is suspended from the skull base by the tensor veli palatini. The posterior velum consists of four muscles which contribute to movement of the palate. The most important is the levator veli palatini which is the primary elevator of the palate. It originates from the petrous temporal bone and cartilaginous Eustachian tube (ET) and enters the velum between the two heads of the palatopharyngeus muscle, joining with its partner from the opposite side behind the aponeurosis to create the levator sling. In patients with a cleft palate, the levator veli palatini no longer has this transverse orientation, but instead the muscle is longitudinally orientated and inserts into the bony cleft margin and posterior palatine bones. The palatoglossus and palatopharyngeus arise from the back of the palatal aponeurosis and maxillary tuberosity. They act as depressors and, along with the levator veli palatini, these muscles act to lengthen the velum. Closure of the velopharyngeal port is important to achieve adequate speech and swallowing. See Figure 89.3.

Submucous cleft palate

A submucous cleft palate occurs when the muscles of the velum are abnormally positioned, but the overlying mucosa is intact. Clinical findings are the classic triad of bifid uvula, zona pellucida (diastasis of soft palate musculature with overlying intact mucosa), and hard palate notch. Studies have shown that a relatively large number of patients with a submucous cleft palate are actually asymptomatic. Treatment, in the form of palatoplasty to realign the muscles, is only required if symptoms of velopharyngeal insufficiency (VPI) are present. It is, however, important to recognize a submucous cleft, especially when considering adenoidectomy.

Management

Up to 75% of cases of CL±P are now picked up antenatally on ultrasound, with isolated cleft palate being the most difficult to diagnose before birth. Following diagnosis, the parents are referred to the cleft multidisciplinary team so that appropriate support can be provided. Advice can be given on feeding, expectations of treatment protocols, and details of parental support groups.

Surgical management is dependent on the surgical deformity present. Table 89.1 shows an expected surgical timeline.

Principles of lip repair

Both functional and cosmetic issues are addressed during lip repair. In both unilateral and bilateral lip repairs the aims are the same, to provide continuity of the muscle and a symmetrical-looking lip with the minimum of scarring. In a bilateral cleft lip this can be more of a challenge as muscle has to be repaired across the projected premaxilla. Strapping the lateral lip segments to the premaxillary segment can facilitate easier lip repair. There are multiple approaches to reconstructing the skin of the lip, with the Millard repair and the Fisher repair being the two most commonly practised in the UK.

Most techniques follow the same general principles:

- Detachment of the muscles from their abnormal insertion.
- Repair of the muscles to their anatomical position.
- Lengthening of the philtrum on the cleft side.

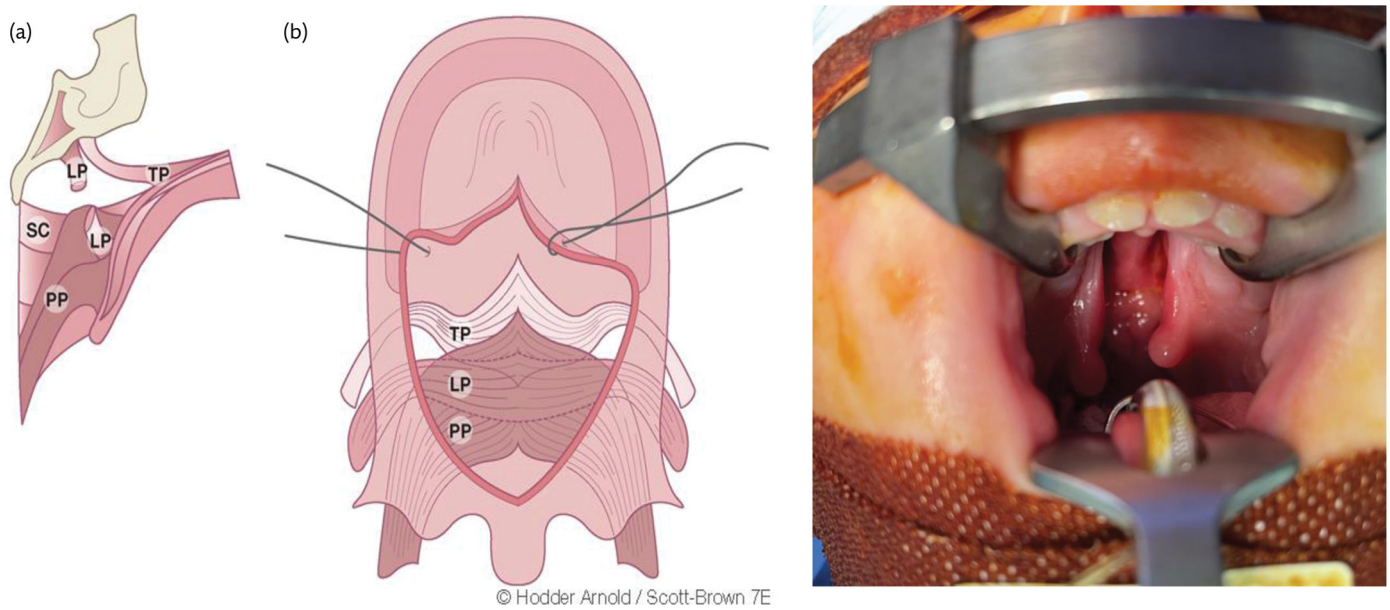

Figure 89.3 Velum muscles: (a) sagittal view from midline; (b) inferior view. (c) Photograph of a child with a cleft palate. LP, levator palatini; PP, palatopharyngeus; SP, superior constrictor; TP, tensor palatini.

(a, b) Reproduced from Wynne D, Ferguson L. Cleft lip and palate. In: Watkinson JC, Clarke RW, eds. *Scott-Brown's Otorhinolaryngology and Head and Neck Surgery*. 8th ed, vol 2. CROC Press; 2018, with permission from CRC Press.

- Accurate apposition of the white roll, vermilion border, and wet/dry mucosal junction.
- Symmetry of cupid's bow.
- Seal off the nasal cavity from the oral cavity.

Primary rhinoplasty can be performed at the time of lip repair, or as a formal rhinoplasty in adulthood. Patients with CL±P are known to be prone to reduced nasal airway, so correcting the septal deformity, along with the aesthetic deformity is key when rhinoplasty is performed.

Principles of palate repair

Closure of the hard palate with a vomerine flap at the time of lip repair in a complete cleft lip and palate is popular within the UK. In patients with an isolated cleft palate, the vomerine flap can be used to help close the nasal layer at the time of palate surgery. The favoured method of soft palate repair within the UK is generally the intravelar veloplasty, popularized by Brian Sommerlad. This technique recreates the transverse orientation of the muscles by radical dissection of the velar muscles:

- Tension-free closure of the nasal and oral layer.
- Release of the abnormal palatal muscle insertions from the hard palate and hamulus.

Table 89.1 An expected surgical timeline

3–6 months	Primary lip +/– hard palate +/– nose repair
9–13 months	Palate repair +/–
3 years old and onwards	Secondary speech surgery (if required)
8–12 years	Alveolar bone graft (if required)
16 years+	Rhinoplasty (if required) Orthognathic surgery (if required)

- Reorientation of the palatal muscles.
- Repair of the muscles posteriorly within the palate.

Alveolar bone grafting

Alveolar bone grafting is used in children with a cleft involving the alveolus to stabilize the alveolar arch and allow eruption of the lateral incisor and canine teeth. It also aids in orthodontic treatment in the future. Timing of the bone graft is usually determined by the development of adult teeth.

Genetics and associated syndromes

There are many different syndromes associated with CL±P. Cleft lip and palate may be syndromic or non-syndromic. Most are non-syndromic and occur as an isolated anomaly. Isolated cleft palate is more commonly associated with a syndrome than cleft lip with or without cleft palate.

Table 89.2 shows some of the commoner genetic conditions you may come across linking ENT issues with CL±P (see Chapter 90).

The 22q11 deletion is a particular genetic consideration in cleft as up to 50% can have palatal abnormalities, including cleft palate, submucous cleft, and non-cleft velopharyngeal dysfunction.

Airway

Pierre Robin sequence

Neonates are obligate nasal breathers due to the superior position of the infantile larynx. This high-positioned larynx allows the epiglottis to interlock with the soft palate when breastfeeding,

Table 89.2 Common genetic conditions linking ENT issues with cleft lip and/or palate

Cleft palate only	Cleft lip and/or palate
Stickler syndrome	Van der Woude syndrome
Treacher Collins syndrome	CHARGE syndrome
Kabuki syndrome	Trisomy 21
22q11 deletion	Trisomy 13
Nager acrofacial dysostosis	Goldenhar (oculo-auriculo-vertebral spectrum) syndrome
	Opitz syndrome

allowing infants to breathe and swallow simultaneously. When the nasal flow of air is blocked, this can have a significant impact on both airway and feeding. Pierre Robin sequence (PRS) is a clinical diagnosis made on the presence of micrognathia, glossoptosis and upper airway obstruction. About 90% of cases of PRS are associated with a cleft palate. As the tongue is retrodisplaced into the posterior pharynx, varying levels of airway obstruction can ensue. This can be apparent at birth, or progressively worsen over the first 6 week of life, hence close monitoring of any child with PRS is imperative. Many children with PRS experience improvement of their airway symptoms over time, and are ready to have their palate repaired between 9 and 13 months, as per guidelines. Improved airway is due to a combination of mandibular growth and neuromuscular development allowing the tongue to move forward with more ease. Failure to thrive and poor feeding can be an indication of airway obstruction, and the link between weight gain and severity of airway obstruction has been found in longitudinal studies. Treatment of airway obstruction in PRS can be non-surgical or surgical; in a significant proportion of children with PRS the airway can be managed non-surgically. Lateral position, nasopharyngeal airway, or non-invasive ventilation in the form of continuous positive airway pressure are all conservative options. Lateral positioning while asleep is controversial in neonates and is an area that is currently under further investigation on its effectiveness and safety in this population (SLUMBRS2 study). Children requiring nasopharyngeal airways can be managed effectively at home with the support of specialist nurses. Rarely, children who do not manage with conservative measures may require surgical intervention. This can take the form of tongue lip adhesion, tracheostomy, or mandibular distraction. Treatment choice depends on many factors and will be decided by the multidisciplinary team. Approximately half of children who have PRS have an associated syndrome, and genetic assessment is recommended. Around 25% will have Stickler syndrome, which is associated with auditory, ocular, and musculoskeletal issues (see Chapter 90).

Sleep-disordered breathing

Children with CL±P are at increased risk of having sleep-disordered breathing and sleep apnoea; obstructive sleep apnoea (OSA) is discussed in more detail in Chapter 75. Studies have found 8.5% of CL±P children have polysomnography-confirmed OSA, with as many as 40% showing symptoms of sleep-disordered breathing. This is approximately three times the general paediatric population. Postoperative correction of CL±P increases the risk of developing OSA and special attention must be paid to patients following surgery. Increase in nasal resistance due to deviated septum, along with narrowing of the pharyngeal airway following palate repair, can increase the risk. Pharyngoplasty, which may be performed for VPI, is also an increased risk factor for OSA as this procedure changes the anatomy of the posterior pharyngeal wall. Assessment and treatment of OSA in the cleft population are similar to those in the general population. Nasopharyngeal airways may be used in children with PRS as above, and can be used temporarily following postoperative palate repair. Tonsillectomy with or without adenoidectomy can be used to improve the upper airway; however, special consideration should be given to adenoidectomy, and an inferior cuff of adenoid tissue should be left intact to maintain the velopharyngeal port for speech.

Middle ear disease and hearing

The prevalence of otitis media with effusion (OME) is around 75% in children with cleft lip and palate compared with 20% in children without clefts. Children with only a cleft lip have the same risk of OME as the general population. OME is discussed in depth in Chapter 88; however, a variety of factors contribute to this high incidence in children with a cleft palate. Studies have shown abnormalities of ET function, primarily the failure of the ET tube opening mechanism, as one of the primary drivers of ET dysfunction in this population. Abnormal insertion of the muscles that aid ET dilation in children with a cleft palate, mainly the tensor veli palatini, also play a role. Abnormalities of the ET itself, including shorter length of ET tube, abnormalities of the ET cartilage, and differences in relationship between the tensor veli palatini and the ET cartilage all contribute. The relationship between structure and function of the ET tube is complex, and evidenced by the fact that once palatoplasty has been performed, the natural history of resolution of OME is still significantly longer than in the general population. The literature has been mixed on the improvements afforded to palatoplasty on resolution of OME, and is certainly an area for future study, as this would help guide management of these children.

Options for management of OME in children with a cleft palate include hearing aids and grommets.

Currently, decisions in regard to need and timing for grommets are taken by the multidisciplinary team looking after the child's care. There is not sufficient current evidence in the literature to recommend a single approach. Grommets were traditionally placed at the time of palate surgery; however, more recently a more conservative approach has been taken. All children with a cleft palate undergo frequent routine audiological assessment until the age of 5 years, and therefore this should guide intervention.

Complications of middle ear disease are higher within the cleft population, including chronic otitis media, tympanic membrane perforation, and cholesteatoma formation. These middle ear complications seem to be of similar incidence whether children have had grommets or not, and likely just reflect the complexity of ear disease found in this population. This reinforces the need for specialist ENT input in this population, with regular surveillance and audiological assessment.

Velopharyngeal insufficiency

The velopharyngeal port is a three-dimensional structure that functionally separates the oropharynx from the nasopharynx during speech and swallowing. VPI is defined as the incomplete closure of the velopharyngeal port. This can be due to structural abnormalities of the palate, or can be caused by non-cleft-related aetiologies such as neurological disorders, 22q11 deletion syndrome, or post adenoidectomy. Elevation of the soft palate to approximate with the lateral and posterior pharyngeal walls creates the velopharyngeal port. This sphincter-like mechanism stops air flowing into the nose on certain sounds requiring oral pressure. If closure is insufficient for any reason, air escapes into the nose, resulting in specific speech characteristics—most notably hypernasal resonance, nasal emission, and turbulence.

Assessment of VPI is best performed by a multidisciplinary team. Detailed developmental and medical history along with examination of the palate to check for clefting or submucous clefting should be undertaken. In children with no obvious diagnosis for their velopharyngeal dysfunction, advice from a neurologist and geneticist should be sought. Standardized speech assessment to assess articulation and intelligibility are performed by specialist speech and language therapists.

Investigations include speech videofluoroscopy, which shows closure of the palate against the posterior pharyngeal wall. If tolerated, flexible nasoendoscopy can also give a view of the velopharyngeal sphincter. Treatment decisions come down to underlying symptoms and the size and location of the velopharyngeal gap. Palatoplasty may be indicated to treat VPI, either as a re-repair of a primary cleft palate, or in the case of a submucous cleft. Pharyngoplasty involves altering the shape of the pharyngeal wall to allow closure of the velopharyngeal port. This can be done either by using flaps from the midline of the pharyngeal wall (midline pharyngoplasty) or by medial transposition of flaps from the lateral pharyngeal wall (sphincter pharyngoplasty). Pharyngoplasty is the first-line treatment for non-cleft VPI requiring surgery.

Other craniofacial considerations

Microtia

Microtia is a spectrum from complete absence of auricular tissue (anotia) to a near-normal ear with only minor abnormalities. A number of different classifications have been proposed; one widely adopted is that by Weerda, who classified microtia from grade I to III, with grade IV being anotia (Table 89.3). Microtia is nearly twice as frequent in males and the ratio of right:left:bilateral is 6:3:1.

Microtia should be treated in a multidisciplinary setting. Due to the embryology of the ear, external ear abnormalities are often associated with middle ear abnormalities, and therefore hearing often needs to be considered and managed appropriately.

Treatment options for microtia include:

- no treatment
- prosthetic ear
- surgical reconstruction.

Table 89.3 International consensus recommendations on microtia, aural atresia and functional ear reconstruction

Grade I	Smaller than normal ear, but essentially normal features
Grade II	Auricle that is rudimentary and malformed but contains recognizable components
Grade III	Small lump of tissue containing some cartilage, 'peanut ear'
Grade IV	Anotia—absence of auricle

Adapted from Zhang TY, Bulstrode N, Chang KW, et al. International consensus recommendations on microtia, aural atresia and functional ear reconstruction. *J Int Adv Otol*. 2019;15(2):204–208.

Auricular reconstruction normally takes the form of autologous costal cartilage grafting. This is performed once the costal cartilage is of adequate development for framework carving, often from 6 years old onwards. There are a number of different surgical techniques which can be used, including single-stage or two-stage procedures. The choice of technique is often dependent on the deformity needing correction, patient selection, and surgeon preference.

Otoplasty

Prominent ears can cause significant psychological and social distress to individuals. Research shows that children often start to develop self-awareness around the age of 6 years, which is typically when otoplasty is considered. Ideally the child will have an understanding and wish for the surgery, as this will help engagement, which is especially important in the postoperative course.

Understanding the normal anatomy of the auricle is paramount to any surgeon wishing to undertake otoplasty. Surgery should be tailored to the deformity that is being corrected. The three areas that often need to be considered are:

- unfolded antihelix
- enlarged conchal bowl
- prominent lobule.

Preoperative photographs are important, as is assessing the position of the ear in the Frankfurt plane. The pinna should sit with the top of the helical rim at the level of the lateral brow. The vertical axis of the ear should incline 20° posteriorly from the vertical plane. The width of the ear is around 55% of the length, which is typically around 6 cm. The auricle protrudes 20–30° from the skull. When planning for otoplasty, the protrusion of the pinna from the mastoid is important. The superior portion of the helical rim typically measures 1–1.2 cm from the scalp. At the midpoint of the ear this increases to 1.6–1.8 cm and at the lobule 2–2.2 cm. Patients often want symmetry between the two ears, which is why initial assessment and measurements are so important. Of course, symmetry is what every surgeon is aiming to achieve; however, it is important to counsel patients that ears are never truly symmetrical and that the ears will likely be siblings rather than twins. Achieving symmetry in unilateral otoplasty is more difficult to achieve than in bilateral otoplasty.

There are a number of surgical techniques available for otoplasty, including anterior scoring, suture techniques, and incisionless pinnaplasty. It is outside the scope of this chapter to describe each of these in detail; however, surgery should be tailored to the deformity which is being corrected. Postoperative management and complications are based on which type of surgery is performed. For open

techniques, a head bandage is required postoperatively to avoid accidental disruption to the ear. Dependant on the age of the patient, surgical technique and surgeon preference, the head bandage can remain for 24 hours or up to 5 days. It is then recommended that a soft headband is worn at night and during outdoor activities for 8 weeks. Most surgeons recommend no contact activities for 8 weeks following the procedure.

Complications

- Bleeding/haematoma—may require drainage.
- Infection/abscess formation—antibiotics and/or drainage.
- Suture protrusion.
- Cartilage deformity.
- Unsatisfactory result.

Controversies/areas of interest for the future

Further research is required to identify best practice for management of OME in children with a cleft palate.

Standardized assessment and recording of severity of PRS, would benefit management and outcomes in management of PRS.

The SLUMBRS2 study will look at the effectiveness of side lying while asleep in children with a cleft palate and its impact on OSA.

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90

The syndromal child

Ravi Thevasagayam and Sanjeev Gupta

Introduction

Children with congenital anomalies comprise almost half of all hospital admissions. These children are encountered by ear, nose, and throat (ENT) surgeons on a daily basis. Conditions encountered in textbooks will be seen particularly in paediatric ENT clinics. There are many hundreds of syndromes and so it is unrealistic and unhelpful to memorize the features of each. That said, there are themes and syndromes more commonly seen in ENT and so it is helpful to have a working knowledge of these and a willingness to reference.

Fortunately, information about the various syndromes is now readily available online. The parents of syndromal children are often well informed and knowledgeable about their child's condition and they usually welcome conversations that acknowledge this. They may have expectations about the consultation informed by accessing online information, online contact with other parents, and interactions with other clinicians. It is often helpful to make a direct inquiry about parental perspectives and expectations.

A *syndrome* relates to an error of morphogenesis in which the presence of more than one malformation is known or assumed to be the result of a single cause. The term *sequence* is used when a series of anomalies is caused by a cascade of events initiated by a single malformation, deformation, or disruption. An *association* is the non-random occurrence in two or more individuals of multiple anomalies not known to be a syndrome or a sequence.

ENT surgeons may come across, as a first presentation, a condition such as hearing loss that may in fact be part of an as-yet undiagnosed syndrome. It is therefore helpful to have a working knowledge of syndromes associated with conditions often seen in ENT. A high index of suspicion and an awareness of targeted examination may help prompt diagnosis and onward referral. In this phase, clinicians should endeavour to have sympathetic interactions with parents and carers for whom this may be a frightening and distressing time.

The following is a list of conditions commonly seen in ENT clinics. These conditions may be a presentation of an underlying syndrome and, if associated with other features, should prompt investigations to exclude one.

Syndromes found in ENT by condition

Conductive hearing loss

- Down.
- Goldenhar.
- Treacher Collins (TCS).
- Turner.
- Kartagener.

Sensorineural hearing loss

- Usher.
- Pendred.
- Jervell and Lange-Nielsen.
- Waardenburg.
- Branchio-oto-renal.
- Alport.
- CHARGE.
- Alstrom.

Nasal obstruction

- Choanal atresia.
- CHARGE.
- Piriform aperture stenosis.
- Solitary median maxillary central incisor syndrome.

Tongue base and pharyngeal obstruction

- Beckwith–Wiedemann.
- Mucopolysaccharidosis (MPS).
- Pierre Robin sequence.
- Stickler.
- Craniosynostosis.

Laryngeal cleft

- Opitz GBBB/Opitz–Frias.

Tracheo-oesophageal fistula

- Opitz GBBB/Opitz–Frias.
- Pallister-Hall.

Laryngeal web

- Velocardiofacial syndrome.

Key syndromes and congenital disorders to know about: major and minor

Major syndromes

Down syndrome

Incidence

1 in 700, making this the most common chromosomal disorder and cause of learning disabilities in children.

Genetics

Down syndrome is not inherited in most cases. It is typically caused by spontaneous errors in cell division involving chromosome 21 during early development of the fetus. These cell division abnormalities result in an extra partial or full chromosome 21.

There are three types of Down syndrome:

1. Trisomy 21 (95%)—three copies of chromosome 21.
2. Mosaic Down syndrome—rare, only some cells have three copies of chromosome 21, the other cells are normal.
3. Translocation Down syndrome (3–4%)—a portion of chromosome 21 becomes attached onto another chromosome before or at conception, and is the only type that may be inherited.

Figure 90.1 Child with Down syndrome—features apparent include up-slanting palpebral fissures, epicanthic folds, flattened nasal bridge, and Brushfield spots on iris.

ENT manifestations

Facial

- External facial features are characteristic—flattened nasal bridge and facial bones, tongue protrusion, telecanthus (widely spaced eyes), up-slanting palpebral fissures, epicanthic eye folds, Brushfield spots in the iris (Figure 90.1).

Ears

- Small ears.
- Narrow ear canals: lead to wax build-up as well as difficult access for grommet insertion.
- Hearing loss: typically conductive, due to otitis media with effusion (OME) secondary to Eustachian tube dysfunction (see Chapter 88), occurring in most children with Down syndrome, persisting to an older age than in other children. Sensorineural hearing loss is less common and may be related to inner ear pathology, including a large vestibule, and changes in the otolithic organs as well as semicircular canals and their ampullae. Age-related sensorineural hearing loss occurs at an earlier age.
- Ventilation tubes (grommets) are effective in correcting hearing loss related to OME in children with Down syndrome but access may be difficult due to narrow ear canals and the benefit may be shorter-lived, with increased risk of subsequent otorrhoea, early extrusion of the tubes, and recurrence of OME. Hearing aids may therefore be offered in the first instance. However, chronic otorrhoea may prevent the wearing of hearing aids, as may poor compliance with keeping hearing aids on. Regular follow-up is recommended given the risk of progression of chronic otitis media to cholesteatoma. Specialist multidisciplinary assessment and management is essential between ENT and paediatric audiology, paediatricians, speech and language therapists, and teachers for the deaf.

Airway

- Obstructive sleep apnoea (OSA): midfacial hypoplasia and relative macroglossia (tongue appearing large related to the smaller size of the oral cavity and oropharynx) reduce upper airway dimensions. This, combined with hypotonia, obesity, and adenotonsillar hypertrophy, leads to OSA in 60% of children. Preoperative overnight oximetry, preferably full polysomnography, should be obtained prior to consideration for adenotonsillectomy to determine severity of OSA and requirement for high dependency care postoperatively as up to 75% need postoperative breathing support. Indeed, adenotonsillectomy alone may not be curative and some will need ongoing nocturnal non-invasive ventilation (e.g. continuous positive airway pressure) or secondary surgery for residual OSA, with multidisciplinary input (see Chapter 75)
- Subglottic stenosis: congenital, usually grade 1 (mild). The expected endotracheal tube size is typically 0.5 size smaller than the expected age-appropriate tube size.

Head and neck

- Brachycephaly and flat occiput.
- Short neck with thick nuchal folds.
- Atlantoaxial instability: 20%, risk of serious spinal injury (1–2%) with overextension of the neck (e.g. in surgery such as adenotonsillectomy), and rarely non-traumatic atlantoaxial subluxation (Grisel syndrome) associated with overuse of diathermy in adenoidectomy. Cervical spine X-rays are not sensitive to

predict those at risk, and therefore care should be taken to protect the cervical spine of every patient instead.

General features

- Other characteristic phenotypic features: single palmar crease, short stature with small hands and feet, sandal-gap between first and second toes.
- Learning difficulties and developmental delay—in 99% but can range from mild to severe, with increasingly recognized co-occurring autism in 5–10%.
- Hypotonia and excessive joint flexibility.
- Hypothyroidism.
- Cardiac abnormalities: 40%—atrioventricular septal defect, ventricular septal defect, patent ductus arteriosus, and tetralogy of Fallot.
- Gastrointestinal defects: constipation (common), congenital duodenal atresia, Hirschsprung disease.
- Relative immunodeficiency: low immunoglobulin A levels, increased risk of respiratory tract infections.
- Obesity, especially in older age.
- Leukaemia: 10–15 times more common, but a lower risk of solid tumours.
- Dementia: Alzheimer's disease.
- Eyesight: strabismus and myopia.
- Dental anomalies and periodontal disease, but fewer dental caries in children.
- Shorter life expectancy—but improved over the years, now over 60 years.

Risk factors

- Advancing maternal age.
- Having one previous child with Down syndrome.
- Carriers of genetic translocation.

Goldenhar syndrome

Also known as hemifacial microsomia, Goldenhar is a common birth defect involving the first and second branchial arch derivatives.

Incidence

1 in 5,600–26,500. 50% more in males than females.

Genetics

Most cases are sporadic, with an unknown cause. Inheritance tends to be autosomal dominant, although some are autosomal recessive. Multifactorial aetiology, possibly related to early injury to the stapedial artery, affecting first and second branchial arch structures. Environmental factors such as maternal drug use increase risk.

ENT manifestations

- Usually unilateral, most commonly affecting right side.
- Microsomia of hemiface affecting mandible and temporomandibular joint, orbit, and muscles of mastication and facial muscles.
- Preauricular appendages.
- Microtia/anotia, ear canal atresia.
- Conductive hearing loss: ossicular malformation or absence.
- Sensorineural hearing loss: abnormal development of cranial nerve (CN) VII, stapedius, semicircular canals, oval window.

General features

- 'OMENS+' grading system: **O**rbit, **M**andible, **E**ar, **N**erves, **S**oft tissues, + (extra-craniofacial features).
- Ocular abnormalities—epibulbar dermoid, coloboma of upper eyelid (vs lower eyelid in TCS).
- Vertebral abnormalities—fusion or absence of cervical vertebrae.
- Mild developmental delay.
- Central nervous system defects—hydrocephalus, occipital encephalocele, and cerebellar hypoplasia.
- Cardiac defects—atrioventricular septal defects.
- Radial anomalies.

Treacher Collins syndrome

TCS, also known as mandibulofacial dysostosis, is a disorder of maldevelopment of the first and second branchial arches in which there is a very characteristic facial appearance. TCS tends to be bilateral and symmetrical compared to Goldenhar syndrome, which is usually unilateral and asymmetrical.

Incidence

1 in 50,000.

Genetics

Mostly autosomal dominant inheritance, although 50% have no family history and are therefore new mutations.

There is some genetic heterogeneity in TCS, with four types identified, including the main form, TCS1—caused by mutations in the *TCOF1* 'treacle' gene (chromosome 5q32); TCS2—mutation in *POLR1D* gene (chromosome 13q12, autosomal dominant/recessive); TCS3—mutation in *POLR1C* gene (chromosome 6p21, autosomal recessive); and TCS4—mutation in *POLR1B* gene (chromosome 2q14, autosomal dominant).

ENT manifestations

- External ear abnormalities with microtia (see Chapter 89), anotia, and canal atresia. This is associated with a conductive hearing loss. Bone-anchored hearing aids may be often considered (**Figure 90.2**).
- A third have an associated cleft palate.
- Micrognathia and mandibular hypoplasia. This, particularly if associated with choanal atresia, will result in airway issues at birth which may necessitate tracheostomy. The mandibular defect may require osteotomies or distraction osteogenesis. OSA may be an issue later.
- These patients have significant issues with regard to difficult intubation. This is due to the steep angle of the dental plane, dental crowding, and significant retrognathia. Problematic intubation into adulthood should always be anticipated with experienced anaesthetic and ENT cover available when general anaesthesia is contemplated.

General features

- Poorly developed midface with absence or hypoplasia of the zygomatic bones. There are usually eyelid abnormalities and mandibular hypoplasia.
- Down-slanting palpebral fissures with colobomas of the lower eyelid common.

Figure 90.2 Child with Treacher Collins syndrome—features apparent include down-slanting palpebral fissures, midfacial hypoplasia, micrognathia and mandibular hypoplasia, and low-set microtia (with canal atresia). This child has a tracheostomy *in situ* due to airway issues at birth. He is also fitted with a bone-anchored hearing aid.

Turner syndrome

This condition affects girls and is caused by a completely or partly missing X chromosome, with a 45,XO karyotype. Short stature and loss of ovarian function are the most common features. About a third have webbing of the neck.

Incidence

1 in 2,500 live female births.

Genetics

Most cases are not inherited. The 45,XO chromosomal abnormality occurs as a random event in the formation of the reproductive cells of a parent.

ENT manifestations

Recurrent OME and conductive hearing loss is common in children. There is a higher incidence of cholesteatoma with 25% having bilateral disease. These patients require close surveillance and surgery should be performed by experienced ear surgeons. High-frequency hearing loss is described in adulthood.

Waardenburg syndrome

Waardenburg syndrome is the most common type of autosomal dominant syndromic hearing loss. It is caused by mutations in any of several genes that affect division and migration of neural crest cells. These cells may form cells in the cochleae and melanocytes, resulting in the hearing loss and pigmentary abnormalities.

Incidence

1 in 42,000. An estimated 2–5% of congenitally deaf children have Waardenburg syndrome.

Genetics

Inheritance is usually autosomal dominant but may also be autosomal recessive. Waardenburg syndrome has been classified into four main phenotypes. Types 1 and 3 are caused by a mutation in the *PAX3* gene (chromosome 2q36). Type 2 is most commonly caused by a mutation in *MITF* (chromosome 3p13) and type 4 is most commonly caused by a mutation in *SOX10* (chromosome 22q13).

General features

Congenital sensorineural hearing loss and a characteristic white forelock is seen in all four types (**Figure 90.3**):

- Type 1 (50% of cases): characterized by a white forelock or premature greying. There may also be heterochromia iridis (different coloured eyes) or brilliant blue eyes. There may also be dystopia canthorum or telecanthus (wide gap between each medial canthus of the eye, i.e. a broad nasal root, but with a normal interpupillary distance). There may be a high nasal bridge, flattened nasal tip, unibrow (synophrys), or a smooth philtrum.
- Type 2 (30%): differs from type 1 in that dystopia canthorum is not seen and the hearing loss tends to be more severe.
- Type 3 (<2%, aka Klein–Waardenburg syndrome): has dystopia canthorum and additional upper limb abnormalities; considered a more severe form of type 1.
- Type 4 (20%, aka Waardenburg–Shah syndrome): associated with Hirschsprung disease.

ENT manifestations

The main issue relates to this as a cause of sensorineural hearing loss. It is not uncommon for the diagnosis to be made in adulthood with the hearing loss as the presenting feature. Adults may dye their hair, making the diagnosis challenging. While amplification aiding is most commonly all that is needed, cochlear implantation has been used with very good results.

Figure 90.3 Child with Waardenburg syndrome with a white forelock and heterochromia iridis of the right iris (https://casereports.bmj.com/content/2013/bcr-2013-201140).

Branchio-oto-renal syndrome

This condition is characterized by abnormalities in the neck, ears, and kidneys, and is the second most common type of autosomal dominant syndromic hearing loss after Waardenburg syndrome. Hypoplastic kidneys or absent kidneys may be present, resulting in chronic renal failure requiring dialysis and, potentially, ultimately renal transplantation.

Incidence

1 in 40,000.

Genetics

Autosomal dominant inheritance, caused by mutations in the *EYA1* gene (chromosome 8q13) in 40%, and *SIX5* gene (chromosome 19q13) in 5%. A related disorder, branchio-otic syndrome, exists without renal anomalies and is associated with the *SIX1* gene (chromosome 14q23).

ENT manifestations

- Ear abnormalities including preauricular pits, preauricular skin tags, dysplastic pinnae, and middle ear abnormalities including canal atresia (**Figure 90.4**).
- Inner ear abnormalities include Mondini dysplasia.
- Conductive, sensorineural, or mixed hearing loss.
- Neck anomalies include branchial cysts and fistulas.
- Children with branchial abnormalities and hearing loss should be have renal function checked and possibly renal ultrasound.

Usher syndrome

Usher syndrome is comprised of congenital sensorineural hearing loss, progressive visual loss due to retinitis pigmentosa, and vestibular dysfunction. It is the most common type of autosomal recessive syndromic hearing loss and the most common cause of deaf-blindness, with a dual sensory impairment.

Figure 90.4 Child with branchio-oto-renal syndrome with right preauricular sinus and two branchial fistulae.

Incidence

3–5 per 100,000.

Genetics

Inheritance is usually autosomal recessive. Mutations in at least 11 genes have been identified giving rise to the four different types of Usher syndrome and their numerous subtypes. The most common gene mutations are in *MYO7A* (chromosome 11q13) in type 1, *USH2A* (chromosome 1q41) in type 2, *CLRN1* (chromosome 3q25) in type 3, and *ARSG* (chromosome 17q24) in type 4.

ENT manifestations

There are four main types of Usher syndrome:

- Type 1—children are born profoundly deaf and begin to lose their sight in the first decade. They also have balance problems with delayed walking milestones due to vestibular hypofunction. These form 90% of Usher patients.
- Type 2—generally associated with moderate to severe hearing loss which tends to be non-progressive. Vestibular problems are unusual. Visual problems manifest in the teens.
- Type 3—children are typically not born deaf but have progressive hearing loss and about half have vestibular problems.
- Type 4—similar to type 2 with late-onset, progressive, moderate to severe hearing loss without vestibular involvement and is X-linked recessive.

Pendred syndrome

This is the second most common type of autosomal recessive syndromic hearing loss after Usher syndrome. It causes a congenital bilateral sensorineural hearing loss and goitre, with patients usually euthyroid or hypothyroid (**Figure 90.5**).

Incidence

1 in 15,000 but may account for up to 10% of all congenital hearing loss.

Genetics

Autosomal recessive inheritance resulting in mutations in the *SLC26A4* (formerly known as *PDS*) gene on chromosome 7q22 which codes for pendrin, an anion transporter protein expressed mainly in the thyroid but also in the cochlea and kidney.

ENT manifestations

Bilateral congenital progressive sensorineural hearing loss. There may be an enlarged vestibular aqueduct. There may be cochlear abnormalities such as Mondini dysplasia and these can be detected on computed tomography and magnetic resonance imaging. Diagnosis is via the pendrin gene although the perchlorate discharge test is sometimes done.

General features

Thyroid dyshormogenesis which often causes thyroid hyperplasia. This may not be apparent until the second decade when a raise in

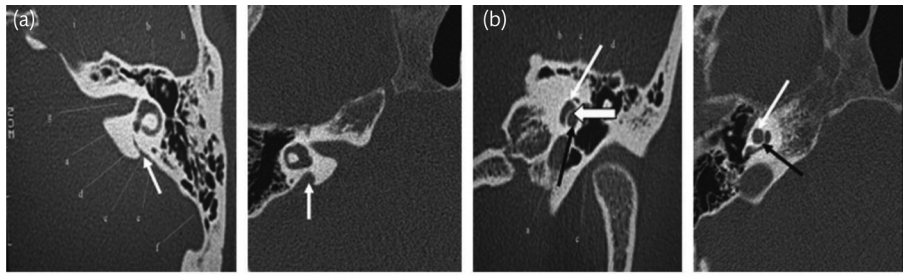

Figure 90.5 Otological features of Pendred syndrome with enlarged vestibular aqueduct (a), and Mondini defect (b) compared to normal anatomy.

Reproduced from Kandasamy N, Fugazzola L, Evans M, Chatterjee K, Karet F. Life-threatening metabolic alkalosis in Pendred syndrome. *Eur J Endocrinol*. 2011;165(1):167–170, doi: 10.1530/EJE-11-0101, with permission from Oxford University Press.

thyroid-stimulating hormone may be detected with normal thyroxine levels.

Jervell and Lange-Nielsen syndrome

This is a rare type of long QT syndrome associated with severe bilateral congenital sensorineural hearing loss. It is associated with arrhythmias that may manifest as syncope and even sudden death. It is the third most common type of autosomal recessive hearing loss (Figure 90.6).

Incidence

1 in 166,000—625,000.

Genetics

Autosomal recessive inheritance caused by mutations in the *KCNQ1* (chromosome 11p15) and *KCNE1* (chromosome 21q22) genes.

ENT manifestations

Children with severe sensorineural hearing loss should have an electrocardiogram where a prolonged QT interval (>500 msec is typical) is found. A family history of syncope, arrhythmias, or sudden unexpected deaths should arouse suspicion and prompt thorough cardiac evaluation.

Stickler syndrome

A clinically variable and genetically heterogeneous connective tissue disorder characterized by ocular, auditory, skeletal, and orofacial abnormalities caused by mutations in genes involved in the formation of collagen.

Incidence

1 in 10,000.

Genetics

There are currently four main types of Stickler syndrome:

- Type 1—membranous vitreous type, solely or predominantly ocular, autosomal dominant, caused by mutation in *COL2A1* gene (chromosome 12q13)—75% of Stickler cases.
- Type 2—beaded vitreous type, autosomal dominant, mutation in *COL11A1* gene (chromosome 1p21).

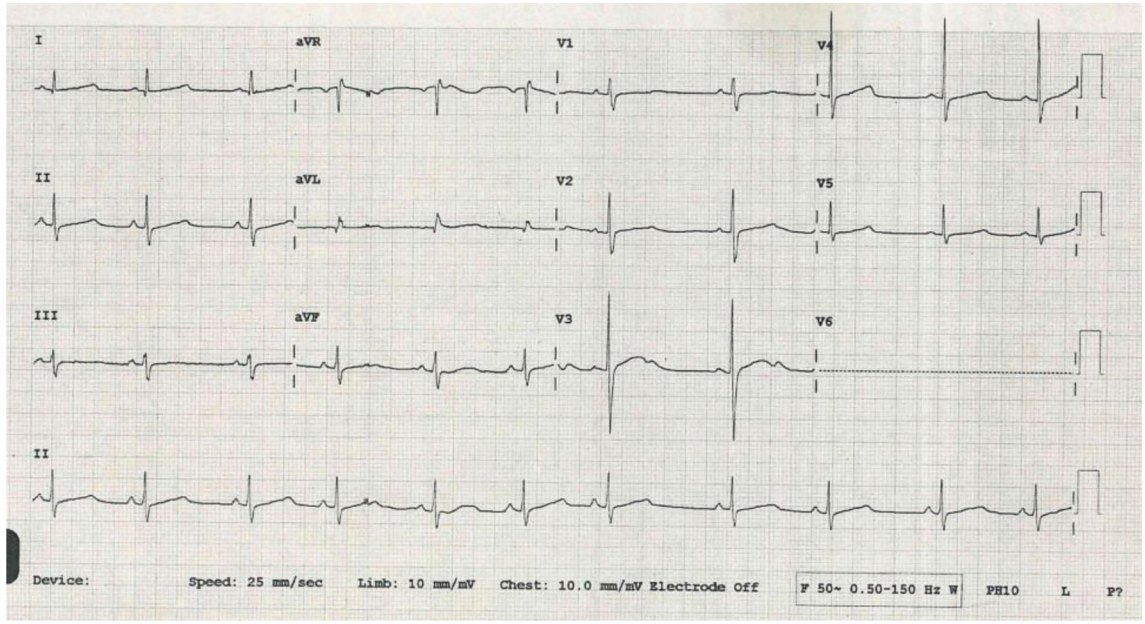

Figure 90.6 Electrocardiogram of child with Jervell and Lange-Nielsen syndrome with prolonged QT interval.

- (Previously designated Type 3—now reclassified as otospondylomegaepiphyseal dysplasia.)
- Type 4—autosomal recessive, *COL9A1* mutation, chromosome 6q13.
- Type 5—autosomal recessive, *COL9A2* mutation, chromosome 1p34.

ENT manifestations

- Sensorineural or conductive hearing loss, especially at high frequency.
- Midfacial hypoplasia and flat midface.
- Midline clefting—cleft palate or bifid uvula, secondary to Pierre Robin sequence or independently.
- Pierre Robin sequence (see Chapter 89).

General features

- Ocular—high myopia, vitreoretinal degeneration, retinal detachment, and cataracts.
- Skeletal—mild spondyloepiphyseal dysplasia and early-onset osteoarthritis

Alport syndrome

Syndrome comprising sensorineural hearing loss and nephritis, often progressing to renal failure.

Incidence

1 in 5,000–10,000.

Genetics

Three types of Alport exist, all involving mutations in genes encoding type IV collagen, which is found in the ears, eyes, and kidneys:

- Type 1: mutation in type IV collagen alpha-5 gene (*COL4A5*), X-linked.
- Type 2: mutation in *COL4A3* or *-A4*, on chromosome 2q36, autosomal recessive.
- Type 3: mutation in *COL4A3*, on chromosome 2q36, autosomal dominant.

ENT manifestations

Slowly progressive sensorineural hearing loss, especially affecting high frequencies (in about 55% of males and 45% of females)—due to bilateral degeneration of organ of Corti and stria, presenting in first decade of life.

General features

- Progressive renal disease and nephritis: glomerular basement membrane defect.
- Renal dysplasia or agenesis.
- Ocular disorders: myopia, cataracts.
- Hypertension.

Neurofibromatosis type 1

The peripheral type of neurofibromatosis (NF), previously also known as von Recklinghausen disease.

Incidence

1 in 2,500—3,000.

Genetics

Autosomal dominant disorder caused by a mutation in the *NF1* gene on chromosome 17q11, which encodes neurofibromin. Signs are often noticeable at birth or shortly afterward, and almost always by age 10.

ENT manifestations

- Benign tumours in the airway (e.g. larynx) can cause airway obstruction and present with stridor, dysphonia, and/or dysphagia.
- Unilateral vestibular schwannoma: 5% risk—causing retrocochlear sensorineural hearing loss.

General features

- Hyperpigmented café-au-lait spots and fibromatous tumours of the skin.
- Lisch nodules in the eye, which are melanocytic iris hamartomas that do not affect vision.
- Increased susceptibility to the development of benign and malignant tumours.
- Bone deformities: scoliosis, bowing of lower limb.
- Optic glioma, usually appear by age 3 years.
- Learning difficulties.
- Large head size.
- Short stature.

Neurofibromatosis type 2

Central neurofibromatosis, much less common than NF1, characterized by tumours of the vestibulocochlear nerve CN VIII (usually bilateral) causing hearing loss, meningiomas of the brain, and schwannomas of the dorsal roots of the spinal cord.

Incidence

1 in 25,000.

Genetics

Autosomal dominant disorder caused by a mutation in the *NF2* gene—which encodes merlin, a tumour suppressor protein—on chromosome 22q12; 50% of cases are due to a spontaneous non-inherited mutation.

ENT manifestations

- Bilateral CN VIII acoustic neuromas (vestibular schwannoma)—95% risk.
- Gradual hearing loss, average age of onset 20 years.
- Tinnitus, poor balance, headaches.
- Facial palsy.

General features

- Meningiomas of the brain.
- Schwannomas of the dorsal roots of the spinal cord.
- Dysphagia.
- Visual loss.
- Limb atrophy.
- Ataxia.

CHARGE syndrome

The term 'CHARGE' is an acronym for the main features of:

- **C**oloboma
- **H**eart defects
- **A**tresia choanae
- **R**etardation of growth
- **G**enito-renal defects
- **E**ar anomalies/deafness.

See Figure 90.7.

Incidence

1 in 10,000–15,000.

Genetics

The debate about whether this was an association or a genetic abnormality ended in 2004 with the identification of mutations and deletions in the *CHD7* gene, on chromosome 8q12, in the majority of patients. A minority of patients do not have an identifiable abnormality in the gene and some with the pathogenic genetic abnormality do not fulfil all the diagnostic criteria or have only a single feature. Inheritance is autosomal dominant.

ENT manifestations

- Choanal atresia (60%) or stenosis—unilateral or bilateral. Bilateral choanal atresia typically presents as an emergency in the first days of life as neonates are obligate nasal breathers (Figure 90.8). CHARGE should be considered in all patients with choanal atresia, with genetic evaluation if suspected. Consideration should be given to cardiac ultrasound and hearing evaluation prior to atresia correction.
- Ear anomalies—microtia, preauricular skin tags, sensorineural/conductive deafness.

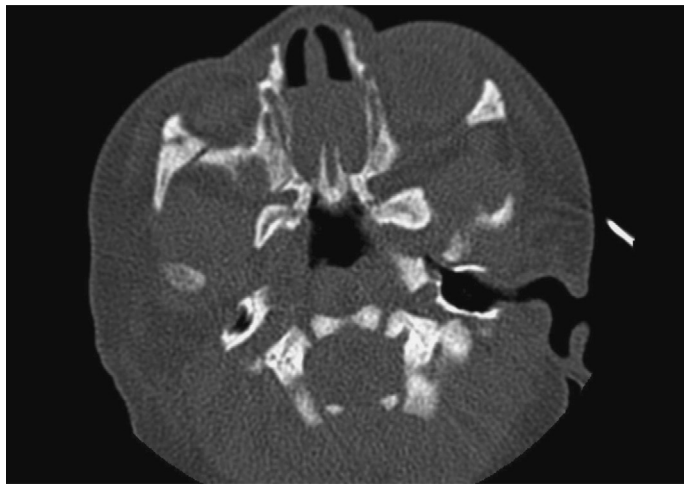

Figure 90.8 Bilateral choanal atresia in neonate with CHARGE syndrome.

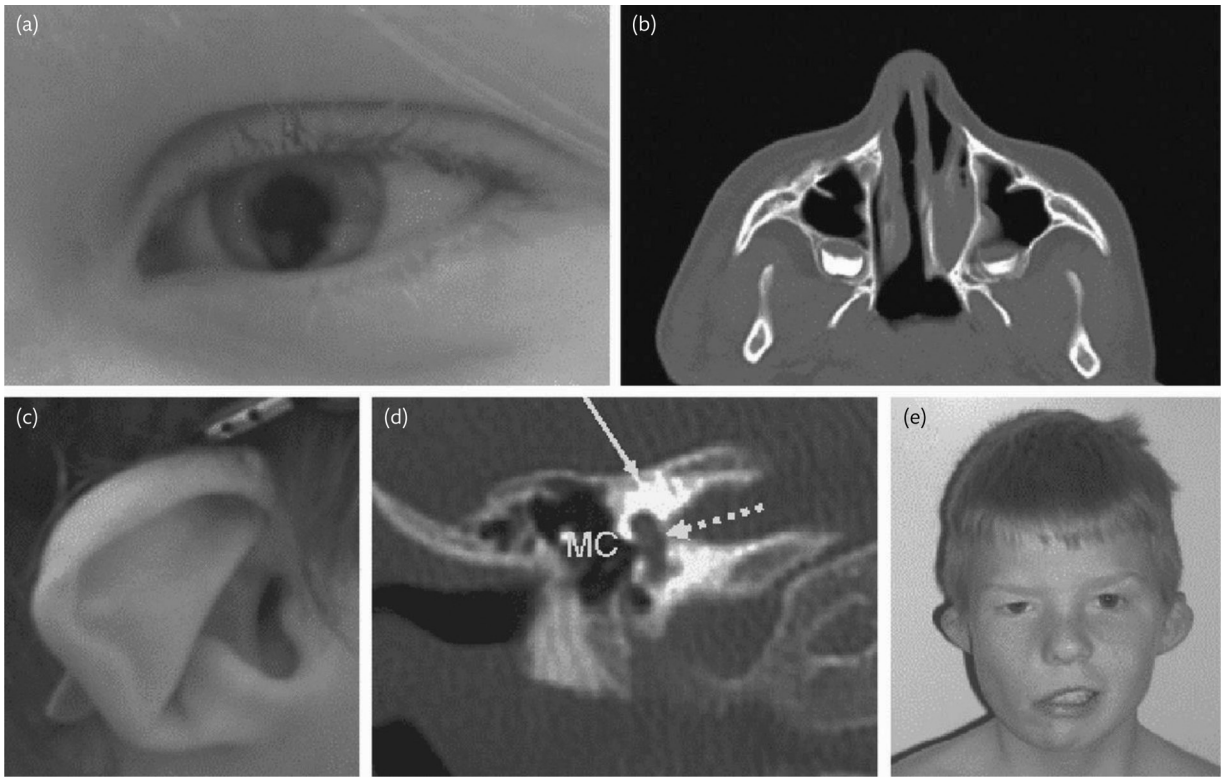

Figure 90.7 Some clinical features of CHARGE syndrome. (a) Coloboma of the right iris. (b) Unilateral choanal atresia. (c) Pinna abnormality and deafness. (d) Agenesis of the semicircular canals (dashed arrow points toward the basal curve of the cochlea, full arrow points toward the place where semicircular canals are normally visible); MC, middle ear cavity. (e) Right-sided facial palsy.

Reproduced from Martin DM, Van Ravenswaaij Arts CMA. CHD7 and CHARGE syndrome. In: Erickson RP, Wynshaw-Boris AJ, eds. *Epstein's Inborn Errors of Development: The Molecular Basis of Clinical Disorders of Morphogenesis*. 3rd ed. Oxford University Press; 2016:995–1000, https://doi.org/10.1093/med/9780199934522.003.0148, with permission from Oxford University Press.

- Macrostomia.
- Laryngeal cleft.

General features

- Coloboma (80%)—commonly iris.
- Heart defects—any, but commonly atrial septal defect/ventricular septal defect/patent ductus arteriosus.
- CN dysfunction, facial palsy, anosmia.
- Retarded growth/short stature.
- Genital/renal hypoplasia.
- Developmental delay.
- Tracheo-oesophageal fistula (TOF).

Craniosynostosis

This term relates to a group of conditions in which there is premature fusion of cranial sutures (Figure 90.9). This results in disordered cranial development with a misshapen skull and often midface abnormalities.

Incidence

1 in 1,800–3,000. Three in four affected are boys.

Genetics

Craniosynostosis occurs as part of a syndrome in between 15% and 40%, and inheritance is autosomal dominant. Fibroblast growth factor receptor genes, *FGFR1*, *FGFR2*, and *FGFR3*, and *TWIST* gene mutations are implicated. Mutations in the *FGFR2* gene (chromosome 10q26) occur in 90% of syndromic cases.

The main syndromes include the following:

- Crouzon syndrome—shallow orbits resulting in proptosis. Cleft palate and ear canal atresia/stenosis is seen.
- Apert syndrome—syndactyly (fusing of fingers and toes) is the striking feature. Midfacial hypoplasia is particularly marked, with up to a third having a cleft palate.
- Pfeiffer syndrome—short, broad thumbs and first toes are seen. There is an association with choanal atresia and tracheal abnormalities.

ENT manifestations

- All patients have OME and middle ear abnormalities as well as sensorineural hearing loss. The ENT team is responsible for regular hearing assessments and hearing rehabilitation/surgical intervention.

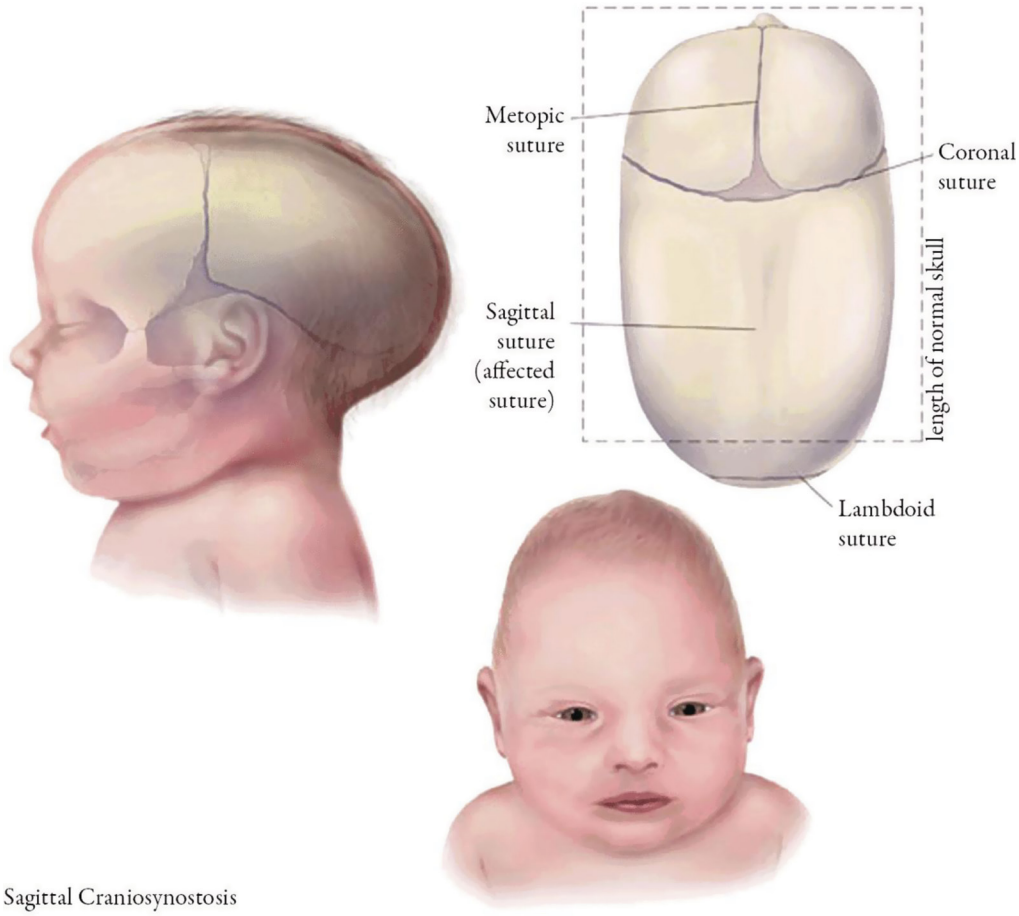

Figure 90.9 Sagittal craniosynostosis with premature fusion of sagittal suture.

Reproduced from Hsieh H, Thornton L, Mann G. Craniosynostosis and anesthetic management for cranial vault remodeling. In: Traul DE, Osborn IP, Anitescu M, eds. *Neuroanesthesia: A Problem-Based Learning Approach*. Oxford University Press; 2018:128–130, https://doi.org/10.1093/med/9780190850036.003.0015, with permission from Oxford University Press.

- Up to half of all patients will develop OSA due to midfacial hypoplasia. There should be a low threshold for overnight oximetry/polysomnography to assess severity. Distraction osteogenesis surgery may be an option and patients should be assessed jointly by specialist facial surgeons and ENT. A tracheostomy is sometimes required.

Achondroplasia

The most frequent form of short-limb dwarfism.

Incidence

1 in 77,000.

Genetics

Autosomal dominant with complete penetrance; however, 85% are not inherited and are the result of a new mutation. It is caused by a mutation in the fibroblast growth factor receptor-3 gene (*FGFR3*) on chromosome 4p16, which causes abnormality of type II collagen production.

ENT manifestations

- Recurrent acute and chronic otitis media with conductive hearing loss, resulting in speech delay.
- Temporal bone abnormality with dehiscence or prominence of the jugular bulb in middle ear cavity.
- OSA due to midfacial hypoplasia resulting in relative adenotonsillar hypertrophy, hypotonia, and obesity. Central apnoeas may also occur as a result of foramen magnum compression, which is responsible for a sudden infant death rate of between 2% and 5% in achondroplasia. In severe cases, growth may lead to raised airway resistance and severe reductions in respiratory compliance, resulting in cor pulmonale with terminal cardiorespiratory failure. Polysomnography is therefore a very useful investigation in this group.
- Gastro-oesophageal reflux may also significantly contribute to airway disease.
- Upturned nasal tip.

General features

- Long, narrow trunk with short limbs, particularly in the proximal (rhizomelic) segments.
- Characteristic facies with megalencephaly, frontal bossing, and midfacial hypoplasia.
- Abnormalities at the craniocervical junction, the commonest being foramen magnum compression, with risk of hydrocephalus and cervical cord compression. Progressive hydrocephalus is also attributed to jugular foramen stenosis resulting in raised intracranial venous pressure.
- Exaggerated lumbar lordosis.
- Limitation of elbow extension.
- Genu varum—bowing of the legs.
- Trident hand.
- Hypotonia.
- Normal intelligence unless hydrocephalus present and untreated.

Velocardiofacial syndrome (VCFS)

VCFS is part of a spectrum of disorders caused by deletions in chromosome 22q11, including chromosome 22q11.2 deletion syndrome, DiGeorge syndrome, and Shprintzen syndrome. VCFS is characterized by abnormalities of the velum (palate), cardia (heart), and facies (face).

Incidence

1 in 4,000–6,000.

Genetics

VCFS is caused by mutations in the *TBX1* gene on chromosome 22q11. Autosomal dominant inheritance occurs in 10–15% of cases, otherwise most are a sporadic deletion with neither parent affected.

ENT manifestations

- Facial anomalies: elongated face, almond-shaped eyes, long-wide nose with bulbous tip, malar flattening, hypoplastic mandible, open mouthed.
- Ears: low set with overfolded helix or other auricular anomalies, conductive hearing loss due to associated palatal anomalies and frequent infections secondary to immune deficiency, speech delay.
- Palate: Pierre Robin sequence resulting in cleft palate (and/or cleft lip) with velopharyngeal insufficiency (poor mobility of the soft palate and lateral pharyngeal walls)—leading to hypernasality and risk of nasal regurgitation.
- Larynx: anterior glottic web (15%), laryngomalacia.

General features

Best remembered with the 'CATCH22' mnemonic:

- **C**ardiac: congenital heart disease, conotruncal defects—malformations of the infundibulum (conus arteriosus) and great arteries (truncus arteriosus).
- **A**bnormal facies.
- **T**hymic aplasia: resulting in T-cell deficiency.
- **C**left lip/palate.
- **H**ypoparathyroidism: with hypocalcaemia due to agenesis of the parathyroid glands.
- **22**q11.

Others:

- Learning difficulties.
- Behavioural problems: including anxiety, attention deficit hyperactivity disorder, autism, and depression.
- Medialized internal carotid artery: care must be taken during adenoidectomy
- Skeletal anomalies and short stature.
- Renal and genital anomalies.

Minor (but still important) syndromes

Beckwith–Wiedemann syndrome

A paediatric overgrowth disorder involving a predisposition to tumour development. The hallmarks of this condition include exomphalos, macroglossia, and gigantism.

Incidence

1 in 13,000.

Genetics

Caused by mutation or deletion of genes within chromosome 11p15, including tumour suppressor p57 (*KIP2*/*CDKN1C*), *H19*, insulin-like growth factor 2 (*IGF2*), and *LIT1* (*KCNQ1OT1*). Autosomal dominant inheritance.

ENT manifestations

- Hemihypertrophy of tongue, and/or macroglossia, which can lead to OSA. Relative adenotonsillar hypertrophy can be treated with adenotonsillectomy. Some children require anterior tongue reduction.
- Conductive hearing loss due to stapes fixation.
- Linear ear lobe creases and posterior helical ear pits.

General features

- Abdominal wall defects including exomphalos.
- Gigantism, with pre- or postnatal growth over 90th centile, but adult heights are generally in the normal range.
- Neonatal hypoglycaemia.
- Visceromegaly involving liver, spleen, pancreas, kidneys, or adrenals. Serial abdominal and renal ultrasound is advocated.
- Hemihypertrophy of limbs—asymmetrical enlargement.
- Predisposition to embryonal malignancies, most commonly Wilms' tumour and hepatoblastoma.

Cri-du-chat syndrome

Incidence

1 in 20,000–50,000 births.

Genetics

Most are not inherited and are a result of new mutations causing deletion of the short arm of chromosome 5 (5p15) with deletion of the telomerase reverse transcriptase (*TERT*) gene. 12% result from an unbalanced translocation involving one parent.

ENT manifestations

- High-pitched cat-like cry in neonates, due to cranial base malformation affecting neurons to the larynx, with a diamond-shaped endolarynx and interarytenoid muscle paralysis.
- Microcephaly and facial dysmorphism with round face, hypertelorism, micrognathia, epicanthal folds, low-set ears.
- Ear infections and conductive hearing loss.
- Bifid uvula, cleft palate, cleft lip.

General features

- Severe psychomotor and intellectual difficulties.
- Hyperactivity or self-abusive behaviour.
- Hypotonia, and then hypertonia in older age.
- Congenital heart defects: patent ductus arteriosus most common.

Möbius syndrome

This is a rare congenital neurological disorder that is characterized by usually bilateral facial nerve (CN VII) and abducens nerve (CN VI) paralysis. Affected individuals have an expressionless face and usually inability to move eyes from side to side.

Incidence

1 in 50,000–500,000.

Genetics

Most cases are sporadic. There is evidence that a gene responsible for Möbius syndrome is located on chromosome 13q12–q13. Sporadic mutations in the *PLXND1* and *REV3L* genes have also been identified in a number of patients. Inheritance is autosomal dominant. The pathogenesis is unclear but is thought to be caused by a vascular disruption in the basilar artery antenatally.

ENT manifestations

- Facial paresis, usually bilateral, with mask-like facial appearance.
- Flattened nasal bridge.
- Micrognathia and microstomia.
- Palatal dysfunction.
- Tongue hypoplasia, asymmetry, and paresis (unilateral).
- Patients may have deafness if auriculotemporal nerve (CN VIII) is involved.
- There may be swallowing and speech issues at birth.
- Speech and language therapy can be effective if patients have intelligible speech.

Mucopolysaccharidosis

MPS is a group of inherited metabolic disorders caused by a lack of specific enzymes involved in breaking down glycosaminoglycans (GAGs), or mucopolysaccharides. The accumulation of partially degraded GAGs within lysosomes causes interference with cell, tissue, and organ function. Lysosomes are compartments in the cell that digest and recycle different types of molecules. MPS is classified as a lysosomal storage disorder.

Incidence

- Severe MPS I: 1 in 100,000; attenuated MPS I: 1 in 500,000 births.
- MPS II: 1 in 100,000–170,000 males.
- MPS III: 1 in 70,000 births.

Genetics

Seven distinct forms of mucopolysaccharidosis—MPS I, II, III, IV, VI, VII, and IX—and numerous subtypes have been identified. Most are of autosomal recessive inheritance, although MPS II (Hunter syndrome) specifically follows an X-linked pattern of inheritance.

The more commonly recognized syndromes include the following:

- MPS I—two subtypes: *severe MPS I* and *attenuated MPS I*. Previously subdivided by syndromes with decreasing severity: *Hurler* (most severe with death in first decade), *Hurler–Scheie*, and *Scheie* (least severe). Mutation of *IDUA* gene on chromosome 4p16 causing deficiency of alpha-L-iduronidase.
- MPS II—*Hunter syndrome*. Mutation of *IDS* gene on chromosome Xq28 causing deficiency of iduronate-2-sulfatase.
- MPS III—*Sanfilippo syndrome*. Most common form of MPS. Four subtypes, each due to the deficiency of a different enzyme: heparan *N*-sulfatase (type A, *SGSH* gene, chromosome 17q25); alpha-N-acetylglucosaminidase (type B, *NAGLU* gene, chromosome 17q21); acetyl CoA:alpha-glucosaminide acetyltransferase (type C, *HGSNAT* gene, chromosome 8p11); and *N*-acetylglucosamine

6-sulfatase (type D, GNS gene, chromosome 12q14). MPS IIIA and IIIB are much more common than MPS IIIC and IIID.

- MPS IV—*Morquio syndrome*. 2 types: type A—deficiency of galactosamine-6-sulfate (*GALNS* gene, chromosome 16q24) and type B—deficiency of beta-galactosidase (*GLB1* gene, chromosome 3p22).

ENT manifestations

MPS I, Hurler syndrome:

- Dwarfism.
- Facial dysmorphism, macroglossia, hearing loss, short neck, recurrent upper respiratory tract infections and otitis media, adenotonsillar enlargement with OSA in more than 80%, vocal cord enlargement with deep hoarse voice.

MPS II, Hunter syndrome:

- Dwarfism.
- Macrocephaly.
- Broad features with low nasal bridge, short neck, bulging forehead, macroglossia, recurrent upper respiratory tract infections and otitis media, adenotonsillar enlargement, mucosal membrane hypertrophy, vocal cord enlargement with deep hoarse voice.

MPS III, Sanfilippo syndrome:

- Less pronounced than other types of MPS.
- Short stature.
- Macrocephaly.
- Coarse facial features (**Figure 90.10**), recurrent upper respiratory tract infections and otitis media, hearing loss, sleep disturbance.
- Delayed speech and behavioural problems.

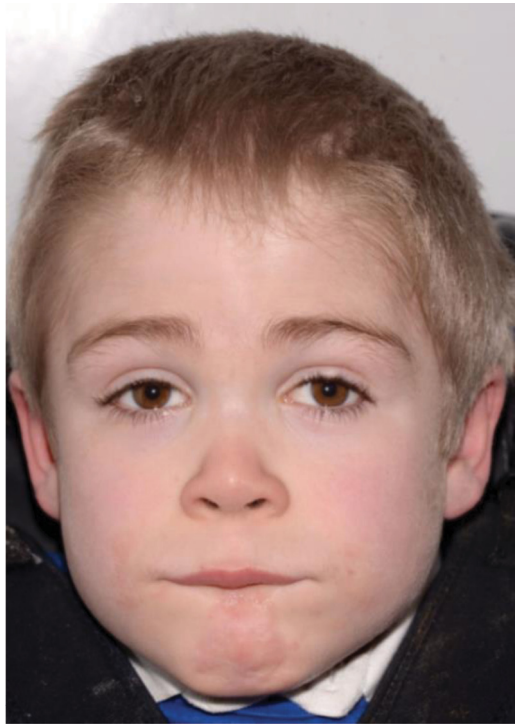

Figure 90.10 Child with Sanfilippo syndrome (MPS III)—features apparent include macrocephaly and coarse facial features.

General features

- Initial normal development in most types of MPS.
- Gradual decline in appearance, physical abilities, organ and system functioning, and cognitive development during first decade of life.
- Clouding of cornea causing severe visual loss.

Osteogenesis imperfecta

This is a group of conditions, also known as brittle bone disease, that are characterized by bone fragility. At least eight main types are now identified with type 2 being the most severe. The underlying condition is caused by a lack of type I collagen.

Incidence

1 in 20,000.

Genetics

More than 90% of cases are caused by a mutation in *COL1A1* or *COL1A2* genes, on chromosome 17q21 and 7q21, respectively. Inheritance is autosomal dominant.

ENT manifestations

- The main issue is deafness, often conductive but also mixed and sensorineural hearing loss. The issues occur at the stapes footplate which may be thickened. There may be crural fractures. Other temporal bone anomalies exist including deficient ossification around the facial canal and cochlea.
- Children with osteogenesis imperfecta require regular routine audiological testing and impairment into adulthood is common.

General features

- Repeated bone fractures.
- Abnormal dentition.
- Blue sclera (**Figure 90.11**).

Prader–Willi syndrome

This is a genetic disorder which leads to children being constantly hungry. Children may have short stature, hypogonadism, learning difficulties, and behavioural problems. The constant hunger means that morbid obesity is a significant risk. Feeding difficulty is common in first 2 months with hypotonia and may require nasogastric feeding.

Incidence

1 in 10,000–25,000.

Genetics

Most commonly arises from a lack of expression of paternal inherited genes on chromosome 15q11–q13. *NDN* and *SNRPN* gene mutations have been implicated. Inheritance is autosomal dominant.

ENT manifestations

The main issue is in terms of management of OSA. While adenotonsillectomy can be helpful/curative, multilevel obstruction is common particularly when obesity is present. This is most commonly at the level of the tongue base. Drug-induced sleep endoscopy is helpful. Central apnoeas are also common so preoperative polysomnography is important to determine likelihood of surgical success. Complication rates are higher post adenotonsillectomy,

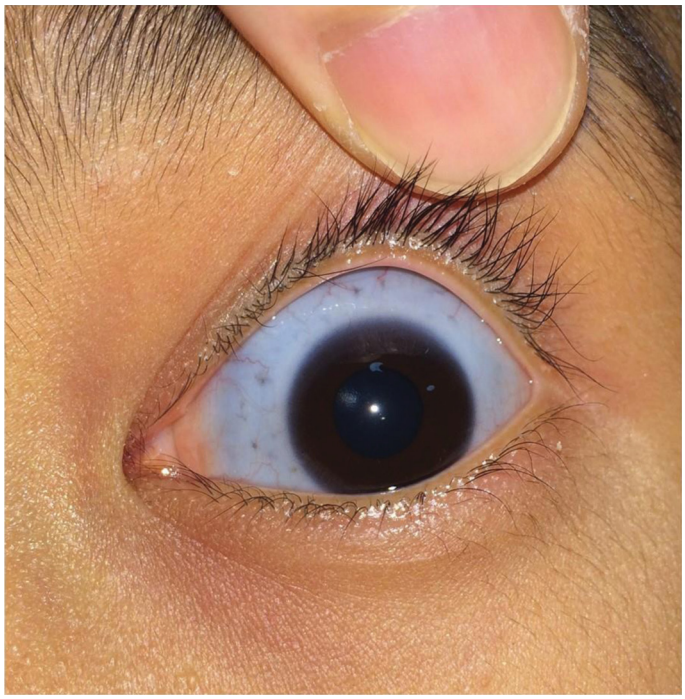

Figure 90.11 Blue sclera in osteogenesis imperfecta.
Reproduced from Mitaka H. Osteogenesis imperfecta and blue sclera. *QJM*. 2018;111(9):665, doi: 10.1093/qjmed/hcy060, with permission from Oxford University Press.

Sequence

Pierre Robin sequence

A sequence of features occurring *in utero*:

1. Micrognathia, *leading to*
2. Relative glossoptosis: posterior positioning of the tongue in the oropharynx, *leading to*
3. Cleft palate: as a result of failure of fusion of the soft palate.

Incidence

1 in 8,500–14,000.

Associations

- Velocardiofacial syndrome.
- Stickler syndrome.

ENT manifestations

- OSA symptoms due to upper airway obstruction caused by the tongue. This tends to improve with age with mandibular growth and increased pharyngeal muscular tone.

Association

VACTERL association

This is a non-random collection of the following anatomical defects, of which three or more being present confirms diagnosis:

- **V**ertebral defects.
- **A**nal atresia.
- **C**ardiac defects (ventricular septal defect, atrial septal defect, tetralogy of Fallot, patent ductus arteriosus, or valve problems).
- **T**racheo-oesophageal fistula (TOF).
- o**E**sophageal atresia.
- **R**enal/**R**adial dysplasia.
- **L**imb anomalies (extra digits, shortened limbs).
- (Single umbilical artery, sometimes included in the classification.)

Incidence

1 in 10,000–40,000 live births.

Genetics

It is typically sporadic and not inherited. There may be a genetic basis for the disorder in some families, with vertebral features most commonly found in relatives. Gene alterations, including duplications or deletions (copy number variation), and mitochondrial dysfunction have been proposed. Causative environmental factors such as maternal diabetes have also been linked.

ENT manifestations

The most common congenital defect in VACTERL is TOF. There may be resulting symptoms of aspiration, such as choking and cough with feeding and recurrent chest infections. ENT may be involved and a microlaryngoscopy and rigid bronchoscopy can identify a TOF and rule out a laryngeal cleft. Patients with a TOF typically have associated tracheomalacia and may present with biphasic stridor and a loud, barking cough, often colloquially called a 'TOF cough'.

Conclusion

There is a myriad of syndromes that are seen within the context of a paediatric ENT clinic. Having a working knowledge of common syndromes and a willingness to reference resources is extremely helpful. These children face many challenges in life, and it is incumbent on clinicians to provide high-quality and sympathetic holistic care for them. This patient group often remain within ENT for long periods and having a good relationship with the patient and carers is desirable and rewarding. ENT care may well provide significant contributions to the quality of life of these children particularly regarding communication, breathing, and sleep.

FURTHER READING

National Institute for Health and Care Excellence. Otitis media with effusion in under 12s. NICE guideline [NG233]. 2023. https://www.nice.org.uk/guidance/ng233

Online Mendelian Inheritance in Man®. OMIM®: an online catalogue of human genes and genetic disorders. https://www.omim.org/

Index

For the benefit of digital users, indexed terms that span two pages (e.g., 52–53) may, on occasion, appear on only one of those pages.

Note: Tables, figures, and boxes are indicated by an italic *t*, *f*, and *b* following the page number.

22q11 deletion syndrome 687, 688*t*, 689
360 feedback 4, 5

abducens nerve *see* cranial nerves: VI
abscess
  Bezold's 653
  Citelli's 653
  deep neck space *see* deep neck space infections
  intracerebral 653
  intracranial 273
  lymph nodes 633–34
  mycobacterial 467, 468
  orbital 272
  parapharyngeal *see* parapharyngeal abscess
  peritonsillar 370–71, 384, 575
  retropharyngeal 465, 466, 467, 468–69, 634
  submandibular 465, 466, 469
  submental 465, 466, 469
  subperiosteal (Pott's puffy tumour) 272, 273, 598, 599, 652, 654*f*
  thyroid 466
abuse of child 563
acceptance and commitment therapy 42
achalasia 359
achondroplasia 700
acinic cell carcinoma 524*t*, 524*f*
acoustic neuromodulation 42
acoustic reflex 27–28, 32–34, 33*f*, 40
acoustic rhinometry 189, 238, 239–40
acoustic shock 125, 130
acoustic trauma, acute 125, 129
Actinomyces spp. 372, 591
Adam's apple 348
adenocarcinoma 293, 294, 295*f*, 522, 523, 525
adenoidal hyperplasia 314, 317
adenoid cystic carcinoma 292–93, 295, 524*t*, 524*f*, 525
  imaging 530–31, 531*f*, 535*f*, 536
adenoidectomy
  cleft lip and/or palate 688, 689
  Down syndrome 692
  obstructive sleep-disordered breathing 583, 585, 586, 587
  otitis media
    with effusion 683
    recurrent acute 652
  paediatrics 588, 677, 678–79
  rhinosinusitis, chronic 598
adenoids
  anatomy and physiology 588, 590, 608–9
  rhinosinusitis, chronic 598
  size 580, 582, 608–9
    hypertrophy 580, 584, 585, 626, 627
adolescents
  adenoids 608–9
  airway obstruction, acute 572, 574–75
  anaesthesia 14
  assessment 561
  consent 564–65, 566
  dysphonia 614
    see also puberphonia 354, 553, 557, 619
  hearing loss 643
  lymph node biopsy 636
  migraine 660, 663
  nasal blockage 610, 611–12
adrenal insufficiency 56
advance care plan 8–9
aerosol-generating medical procedures (AGMPs) 20–22, 21*t*
age factors
  adenoid size 608–9
  allergic rhinitis 314
  anaesthesia 636
  benign paroxysmal positional vertigo 112
  choanal atresia 611
  cholesteatoma 656
  chronic refractory cough 555
  dysphonia 614, 615–16, 620
  epiphora 231*t*
  epistaxis 198
  head and neck cancer 494–95, 496
  hearing assessment 644–46, 650, 662
  hearing loss, paediatric 643
  hyperparathyroidism 470, 471, 473
  hypopharyngeal cancer 450, 453–54
  juvenile nasopharyngeal angiofibroma 604
  juvenile recurrent parotitis 640
  laryngeal anatomy and physiology 613
  laryngeal cancer 411, 415
  nasal cycle 237
  nasal growth 608
  neck lumps 380, 635
    branchial cleft cyst 508
    paraganglioma 516
    schwannoma 515
    thyroglossal duct cyst 514, 515
  neck trauma 400
  necrotizing otitis externa 176
  non-allergic rhinitis 315
  obstructive sleep apnoea 580, 583, 584
  olfactory disorders 195
  oral ulceration 406
  oropharyngeal cancer 419, 420–21
  osteogenesis imperfecta 149
  otitis media
    acute 650–51
    with effusion 681, 683
  otoplasty 689
  otosclerosis 148
  peak nasal inspiratory flow 240
  puberphonia 354, 553, 557, 619
  rhinorrhoea 208
  stapes surgery 153
  swallowing process 624
  temporal bone trauma 166
  thyroid cancer 445, 446
  tinnitus 37–38, 129
  tympanic membrane perforation surgery 656
  tympanoplasty 160
  vasomotor rhinitis 315
  see also adolescents; infants; neonates
ageing process 309, 309*b*, 310*f*–11*f*
  dysphonia 354
  facial rejuvenation *see* face: non-surgical rejuvenation; face: surgical rejuvenation
  submandibular salivary gland 379
ageusia 192*t*
air conduction audiometry 30
air conduction hearing aids *see* hearing aids
airway assessment in children 566–67
airway compromise, acute 570, 571, 578
  anatomy and physiology 570
  assessment 570
  differential diagnosis/associated conditions 571–75
  investigations 571
  medical treatments 575
  surgical management 575–78
airway development 665
airway management, anaesthesia 12–13
airway obstruction 391, 395
  anatomy 391, 391*f*
  controversies 395
  differential diagnoses 392
  evaluation and assessment 391–92
  guidelines 395
  holding measures 392–93, 393*b*
  human factors and non-technical skills 394–95
  impending, signs of 392*b*
  interventions 393–94, 394*f*
  investigations 392
  paediatric 665
    assessment 665–66
    controversies and future 673–74
    differential diagnoses 667–70, 667*f*–69*f*
    investigations 666–67, 666*f*
    treatment 670–73, 671*f*
  physiology 395
  science 391
  tips and pearls 395
airway reconstruction
  dysphagia after 628
  dysphonia after 620
alcohol intake
  anaesthesia 13
  dizziness 57
  by doctors 6
  head and neck cancer 344, 346
    hypopharyngeal 450–51
    laryngeal 411
    nasopharyngeal 456
    oral 428, 431, 432, 436
    oropharyngeal 427
    squamous cell carcinoma 521
  neck lumps 375, 376, 382–84
  vestibular migraine 109
Alexander law 48–49
allergic rhinitis (AR) 208–9, 209*f*, 314, 318
  adenoidal hyperplasia 317
  asthma 317, 318
  central compartment atopic disease 285
  controversies 213
  differential diagnosis 314–15, 317–18, 610
  examination findings 315, 315*f*
  guidelines 213–14
  investigations 315–16, 315*f*–16*f*
  nasal obstruction 237, 238, 240
  paediatric 599–603, 606, 607, 610

allergic rhinitis (AR) (*cont.*)
pathogenesis 314
rhinosinusitis, chronic 317, 598
tips and pearls 317–18
treatment 316–17
allergy
nasal complaints 184, 188
neck lumps 376
rhinitis *see* xix
rhinosinusitis, chronic 286–87, 597–98
allografts 335, 337*t*, 339
Alport syndrome 646, 697
alternative medicine 42
Alzheimer's disease 195
American Academy of Facial Plastic and Reconstructive Surgery (AAFPRS) 307
American College of Radiology Thyroid Imaging, Reporting and Data System (TIRADS) 445, 536–37
American Joint Committee on Cancer (AJCC) 421, 434, 458, 461, 521
aminoglycoside eardrops, in presence of open middle ear 67
amyotrophic lateral sclerosis 363
anaemia 320, 322, 323, 660
anaesthesia 12
airway 12–13
airway obstruction 393–94
endoscopic sinus surgery 275
hereditary haemorrhagic telangiectasia 320
laryngeal cancer 412
laryngotracheal, head, and neck surgery 17–18
local 14, 15, 16
otology 16
paediatrics 14, 15–16, 636
choice of anaesthesia 15
day-case tonsillectomy 14*b*
otology 16
pre-assessment 14–15
premedication 15
tonsillectomy 14, 14*b*, 15
upper respiratory tract infection 15*b*
phonosurgery 549–50, 549*f*, 552
preoperative preparation and risk assessment 13–14, 13*b*
procedural 15–16
regional 15
rhinology 16–17
rhinoplasty 249
throat packs 17*b*
tonsillectomy 677–78
topical 16
tracheostomy 540
tubeless 17
anaphylaxis 392
angiofibroma
juvenile nasal *see* xx
sinonasal 291, 292, 294*f*
staging 291*t*, 291
treatment 293–94
angiography 291, 392, 399, 401, 402, 517
anosmia 19–20, 184, 192*t*, 195, 196
anotia 97, 98
anterior neck abscess 465, 466, 468
anterior skull base
endoscopic approaches *see* endoscopy: anterior skull base approaches
reconstruction 331, 333
complications 338–39
controversies 339
decision-making 333, 334*f*, 336*f*, 337*t*, 337*b*
free grafts 333–37, 337*t*
postoperative care 338
principles 333
securing the reconstruction 338
tips and pearls 339
vascularized flaps 337–38
anterolateral thigh fasciocutaneous flap 338
anterolateral thigh flap (ALT flap) 485, 487, 488
antrochoanal polyp 610, 612
anxiety
dizziness 56, 57, 661
dysphonia, paediatric 613
neck lumps 375–76
paediatrics 568, 613, 661
parental 568, 582–83
sound tolerance disorders 38
throat pain 373
tinnitus 38, 39, 40, 42
aortopexy 673
Apert syndrome 609, 626, 699
aphonia 557
apnoea 582
apnoea–hypopnoea index (AHI) 387, 390, 582
appraisals, professional 3, 6
arachnoid cysts 135–36, 138*t*
Arcanobacterium haemolyticum 592
areca nut use 428, 450
Arnold's nerve 27
arteriovenous malformations (AVMs) 319, 320, 322, 323–24, 323*t*, 632
arytenoid cartilages 348
Aspergillus spp.
flavus 62
fumigatus 653–54
niger 62, 63*f*, 653–54
assistive listening devices 36, 41, 648
asthma
airway compromise, acute 573–74
allergic rhinitis 317, 318
dysphonia, paediatric 619
obstructive sleep-disordered breathing 583
rhinosinusitis, chronic 598
astigmatism 661
ataxia 661, 662
atlantoaxial subluxation 679, 692–93
audiometry
dizziness, paediatric 662
dysphonia, paediatric 614
noise-induced hearing loss 126, 127*f*, 128*f*, 129
otalgia and otorrhoea 61, 65
otitis media
with effusion 682
recurrent acute 65
otosclerosis 148–49
pure tone *see* pure tone audiometry
sensorineural hearing loss 78–79, 79*f*
speech 84
temporal bone trauma 170–71
tinnitus 40
vertigo 53
audiovestibular implants 119, 124
guidelines 119–20
implant team 119
outcomes 123–24
paediatrics
active middle ear implants 120
auditory brainstem implants 123
bone conduction hearing devices 120
cochlear implants 122–23
guidelines 119–20
outcomes 123
patient selection 119
see also 36; ; ; middle ear implants
auditory brainstem implants (ABIs) 119, 120, 123–24
auditory brainstem responses (ABRs) 34–35, 35*f*
functional hearing loss 35
paediatrics 644–45, 662
sensorineural hearing loss 78
temporal bone trauma 170–71
auditory evoked potentials 34–35, 35*f*
auditory nerve *see* cranial nerves: VIII
auditory neuropathy 35, 661–62
auditory pathways 27–28, 29
auditory steady-state response 35
auditory system, functional elements 38–39, 39*f*
tinnitus 38–39, 39*f*
auditory testing *see* hearing assessment
audits 4–5
augmented reality 276
aura 661
aural atresia 97–98, 98*t*
acquired 102
audiovestibular implants 123
congenital 99–102, 100*f*–1*f*
aural fullness 69, 70
anatomy 59
balloon dilation Eustachian tuboplasty 70
causes 59*t*
dizziness 48
Eustachian tube 69–70, 70*t*
sensorineural hearing loss 78
auricle *see* pinna
auriculotemporal syndrome (Frey syndrome) 439, 439*t*, 441, 442
autism 563
autografts 335, 337*t*, 339
autoimmune conditions
differential diagnosis 288
inner ear disease 56
sensorineural hearing loss 78
autoinflation, otitis media with effusion 683
autophony 48

bacterial tracheitis 574
Bacteroides spp. 591
balance 45–46, 45*f*, 662
balloon dilation Eustachian tuboplasty 70
bamboo nodules 547, 550
Barbeque manoeuvre *see* Lempert 360˚ roll manoeuvre
barium swallow
branchial cleft cyst 512
dysphagia 360–61
modified *see* xxi
barotrauma, otic 66, 66*t*
Barrett's oesophagus 361
Bartonella henselae 635, 640
basal cell carcinoma 377, 481, 524*t*
basilar membrane, anatomy and physiology 28–29
Basser's syndrome *see* xix
Battle's sign 171*b*
Beckwith–Wiedemann syndrome 700–1
Bell's palsy 73–74, 73*t*, 137
benign paroxysmal positional vertigo (BPPV) 57, 112, 116–17
anatomy and physiology 112
associated conditions 114
differential diagnosis 106, 114, 115*t*
duration of dizziness 46–48
Epley manoeuvre 55
investigations 53, 112–14, 113*f*
medical treatments and regimens 114–16, 116*f*
outpatient examination 51–52
paediatrics 660, 661, 662, 663
surgical treatments 116
symptoms 46
temporal bone trauma 174
triggers 48
vitamin deficiencies 57
benign paroxysmal vertigo of childhood (BPVC) 660, 663
betel nut use 450–51
Bezold's abscess 653
bi-lobed flap 482, 483
biomarkers
Ménière's disease 110–11
tinnitus 42
vestibular migraine 110–11
biopsy
deep neck space infections 467
fine needle aspiration *see* fine needle aspiration
head and neck pathology 344, 520
laryngeal cancer 412
lymph nodes 636
meningoencephalocele 610
nasal 186–87, 246
nasal septum 205, 207
nasopharyngeal carcinoma 523
neck lumps 379, 380, 381, 382, 384
oral cancer/precancer 431, 432–33, 437
oral ulceration 409
oropharyngeal cancer 420–21
paraganglioma contraindication 516
phonosurgery 550
sentinel node 434
sinonasal tumour 291
thyroid cancer 447–48
bispectral index (BIS) 388

blepharoplasty
  lower 301–4, 303*f*, 304*f*, 305*f*
  upper 300, 301*f*, 302*f*
body mass index (BMI)
  sleep-related breathing disorders 386–87, 388, 389, 390
  see also obesity
bone conduction audiometry 30
bone conduction hearing devices (BCHDs) 119, 120
  aural atresia 102
  cholesteatoma 144
  guidelines 119
  magnetic resonance imaging 123–24
  ossicular chain abnormalities 156
  otitis media with effusion 683
  otosclerosis 149
  outcomes 123
  paediatric 648
  Treacher Collins syndrome 693, 694*f*
bone diseases, differential diagnosis and treatment 89
Borrelia burgdorferi 74
botulinum neurotoxins (BoNTs), facial rejuvenation 309–10, 311*b*, 311*t*, 313
  complications 310
  regulations 313
brachytherapy 434, 436–37, 460
brainstem infarcts 46–48
brainstem stroke 48
branchial cleft
  cyst 508
    anatomy and embryological considerations 508, 509*f*, 510*t*, 510*f*
    assessment 508–11, 511*f*
    controversies and future 513
    differential diagnoses 511
    imaging 537, 538*f*
    investigations 511–12, 511*f*
    neck lumps 380, 384
    tips and pearls 512–13, 512*f*–13*f*
    treatment 512, 512*f*
  sinuses 630, 633, 636
branchial cysts 633
branchio-oto-renal (BOR) syndrome 636, 646, 695, 695*f*
  dizziness 660–61
  hearing loss 644, 646
breastfeeding
  dysphagia 625–26
  nasal anatomy 608
  swallowing process 624
British Committee for Standards in Haematology 202, 202*b*
British Thyroid Association
  Thy score 446
  U classification system 445, 536–37, 536*f*, 536*t*
brittle bone disease *see* xx
bronchoscopy
  COVID-19 20
  neck lumps 382
  paediatrics 571, 575, 576, 578, 614–15
brow lift *see* eyebrows: non-surgical rejuvenation; eyebrows: surgical rejuvenation
bruxism 67
bullying 5
burnout 6

cacosmia 184, 192*t*, 195, 284
caesaraean section 578
caffeine 109
caloric testing 53–54
canalplasty 161
canal wall down (CWD) surgery, cholesteatoma 144, 145
canal wall up (CWU) surgery, cholesteatoma 143*f*, 144–45, 146
Candida spp.
  albicans 62
  hypopharyngeal cancer differential diagnosis 451
  otitis externa 653–54
  otomycosis 62
  pharyngitis 592
  throat pain 371
candour, duty of 5
cannabis oil vaping 373
cantholysis 276–77, 278*f*
canthotomy, lateral 276–77, 278*f*
carbon dioxide laser ablation 20–21
carbon ion radiotherapy 458
carbon monoxide poisoning 57
cardiac disease 13–14
Care Act (2014) 10
Care Quality Commission 5, 9
carotid artery
  aberrant 82*t*
  external 200
  internal 168
Carrau–Hadad flap 337
Castleman's disease 381, 635
cat scratch disease 635, 640
cauliflower ear 64, 64*f*, 171
cavernous sinus thrombosis 272
cellulitis
  orbital 272, 274
  postseptal 272
  preseptal 272
Centor criteria 594
central compartment atopic disease (CCAD) 284–85, 286–87
cerebellar function 52, 662
cerebellar haemorrhage 46–48
cerebral venous sinus thrombosis (CVST) 653
cerebrospinal fluid (CSF)
  leak
    anterior skull base reconstruction 333, 338–39
    differential diagnosis and treatment 82*t*, 85–87, 87*f*
    endoscopic interventions 277, 326, 327–28, 329–30, 331–32
    grading 337*t*
    meningoencephalocele 610
    midface fracture 404
    olfactory disorders 194
    otorrhoea 66
    rhinorrhoea 208, 210–13, 212*f*–13*f*
    sinonasal tumour surgery complication 295
    temporal bone trauma 169, 171, 172*f*, 172–73, 174
  production reduction 338
cerumen impaction 94–97, 96*f*
cervical adenopathy 384
Chandler classification, orbital complications 272
CHARGE syndrome 698–99, 698*f*
  cleft lip and/or palate 688*t*
  differential diagnosis 609
  hearing loss 644, 646, 647*f*
  laryngeal clefts 668–69
chemical peels, facial rejuvenation 312
chemodectoma *see* paraganglioma
chemotherapy
  head and neck cancer 491
    hypopharyngeal 452, 453
    laryngeal 413, 414, 415, 416*b*, 416–17
    nasopharyngeal 458–62
    neoadjuvant/induction chemotherapy 495
    oral cancer 434, 436, 437
    oropharyngeal cancer 421, 423, 424*b*, 425, 426, 522–23
    palliative use 496
    radiotherapy with concomitant chemotherapy 492–95
  head and neck reconstruction 490
  Langerhans histiocytosis 655
  rhabdomyosarcoma 655
Chiari malformation 571–72, 615
children *see* infants; neonates; paediatrics; toddlers
Chlamydia pneumoniae 592
choanal atresia 572–73, 611
  differential diagnosis 609
  rhinorrhoea 210
  stenting 612
cholesteatoma 83*f*, 141, 147, 650, 656
  acquired 141, 142*f*
  assessment 657
  atresia, acquired 102
  classification 141
  clinical assessment and investigations 142–44, 144*t*
  conductive hearing loss 81
  congenital 89, 90*f*, 141, 141*f*
  definition 141
  differential diagnosis 82*t*, 177–78
  dizziness 660, 662
  external ear 103–4, 103*t*, 103*f*
  facial paralysis 74
  hearing loss 141, 142, 144*t*, 644
  investigations 657
  management 144, 656–57
  middle ear and mastoid anatomy 142*f*–43*f*
  non-retraction pocket 141
  ossicular chain abnormalities 154, 155, 156
  otalgia 147, 656–58
  otorrhoea 103, 141, 142, 144*t*, 145, 147, 656–58
  pathophysiology 141–42
  petrous apex 138
  physiology 656
  postoperative surveillance 146
  recurrent, avoidance and management of 146–47
  retraction pocket 141
  surgical options 144–46, 145*t*, 146*t*
  temporal bone trauma 174
  tips and pearls 657–58
  treatment 657, 657*f*, 658*t*
  Turner syndrome 694
cholesterol granuloma 137, 138*t*
chondrosarcoma 138*t*, 138, 329
chorda tympani 152, 158–59
chordoma 138*t*, 138, 329
chromoendoscopy, virtual 344
chylous fistula 436
ciliary dysfunction disorders 287–88
ciliary dyskinesia, primary 598
Citelli's abscess 653
cleft lip and/or palate (CL±P) 61, 687, 690
  adenoidectomy 678–79
  airway 687–88
  alveolar bone grafting 687
  cholesteatoma 656
  dysphagia 626–27
  functional anatomy 681, 686*f*, 687*f*
  genetics and associated symptoms 687, 688*t*
  LAHSHAL classification system 685, 685*f*
  management 686–87, 687*t*
  middle ear disease and hearing 688
  otitis media
    with effusion 681
    recurrent acute 651
  submucous 686
  Treacher Collins syndrome 693
  tympanic membrane
    perforation 656
    retraction 655
  velopharyngeal insufficiency 689
Clostridium botulinium 309
cluster headache 210, 220, 222*t*, 223
cocaine 204, 207, 243–44, 244*f*, 245
cochlea
  anatomy and physiology 27–29, 29*f*
  balance 46
  embryology 643
  noise-induced hearing loss 125
  synaptopathy, noise-induced 127
  vertigo 55
cochlear implants (CIs) 119, 121
  anaesthesia 16
  guidelines 119–20
  implant design and surgical technique 121–22
  magnetic resonance imaging 123–24
  Ménière's disease 106
  ossicular chain abnormalities 156
  otosclerosis 85, 150
  outcomes 123
  paediatrics 16, 648–49
  sensorineural hearing loss 78, 80
  speech processing strategies 122–23
  temporal bone trauma 170–71, 174
  tinnitus 41
  Waardenburg syndrome 694
cochlear microphonics (CM) 35
cochlear obliteration 647

cochlear stimulation, direct acoustic 150
cochleovestibular nerve *see* cranial nerves: VIII
Code of Practice 1972 (occupational noise) 126
Cogan syndrome 56
cognitive behavioural therapy (CBT)
  dizziness 56
  head and neck cancer rehabilitation 41
  psychogenic voice disorders 558
  tinnitus 42
cognitive decline 79
collagen fillers 311*t*, 312
colleagues, working with 5
  see also multidisciplinary team
common cold 210, 267, 597
communication tactics, hearing rehabilitation 36
complaints 5
complementary and alternative medicine 42
computed tomography (CT)
  4D 472, 475
  airway abnormality, paediatric 667
  airway obstruction 392, 571
  allergic rhinitis 314, 315, 315*f*, 316
  angiography 392, 399, 401, 402
  atlantoaxial subluxation 679
  cholesteatoma 143–44, 147, 657
  cone beam 84, 149, 154, 187, 492
  deep neck space infection 468, 592–93, 635
  dizziness, paediatric 662
  dysphagia, paediatric 626
  dysphonia, paediatric 615, 617, 620
  Eagle syndrome 593
  endoscopic sinus surgery 274–75, 275*t*, 277
  epiphora 231
  facial pain 225
  facial trauma
    mandible fracture 403
    white eye blow out 403
    zygomatic complex fracture 403
  fluorodeoxyglucose 179, 434 *see* fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT)
  glioma 631
  headache 220, 225
  head and neck 527–29, 528*f*, 530
    radiotherapy 491
  high-resolution *see* high-resolution computed tomography
  hyperparathyroidism 471–72, 475
  hyperthyroidism contraindication 537
  hypopharyngeal cancer 532–35
  juvenile nasopharyngeal angiofibroma 604–5
  laryngeal cancer 411, 532–35, 534*f*
  medullary thyroid cancer 537
  meningoencephalocoele 631
  nasal obstruction 238, 609, 610
    glioma 610
    juvenile nasal angiofibroma 610, 612
    meningoencephalocele 610
  nasopharyngeal cancer 458, 460
  neck lumps 379, 380, 381, 382, 384
    branchial cleft cyst 511, 528*t*
    dermoid cyst 518
    paraganglioma 516
    schwannoma 515
    thyroglossal duct cyst 513
  necrotizing otitis externa 178*f*, 178, 179, 180
  nose and paranasal sinuses 187
  obstructive sleep apnoea 585
  olfactory disorders 192–93
  oral cancer 433, 434*t*, 533*f*
  oral ulceration 409
  oropharyngeal cancer 419–20, 421, 422*f*–23*f*
  ossicular chain abnormalities 154, 155*f*
  otitis media, acute 650, 652
  papillary thyroid cancer 537*f*
  and PET *see* positron emission tomography: and computed tomography
  rhinorrhoea 210, 211–12
  rhinosinusitis
    acute 269, 269*f*, 273
    chronic 286, 596, 597
  salivary glands, major 536
  sinonasal tumour 291, 292–93, 294, 294*f*, 295*f*
  skull base and sinonasal disease 530
  stapes surgery 149, 151, 152*f*
  temporal bone trauma 171, 172*t*
  throat pain 370, 372, 590
  thyroid cancer 447–48
conductive hearing loss (CHL) 77, 81, 643–44, 646, 647
  assessment 81–84
  audiovestibular implants 120
  aural atresia 102
  causes 120*b*
  cholesteatoma 140
  differential diagnoses and treatment 82*t*, 84–89
  dizziness, paediatric 660
  external ear malformations 98
  investigations 84
  keratosis obturans 103
  myringitis 65
  ossicular chain abnormalities 154, 156, 158
  otalgia and otorrhoea 61
  otic barotrauma 66
  otosclerosis 148–49, 150
  paediatric syndromes 691, 693, 694
  pure tone audiometry 31
  stapes surgery, revision 151
  temporal bone trauma 174
  tinnitus 38
  tympanic membrane perforation 160
  see also mixed hearing loss
cone beam computed tomography 84, 149, 154, 187, 492
confidentiality 5–6
conflicts of interest 6
congenital high airway obstruction syndrome (CHAOS) 572
conjunctivo-dacryocystorhinostomy 233
Consensus Auditory-Perceptual Evaluation of Voice (CAPE-V) 350, 614
consent
  anaesthesia 14
  anterior skull base reconstruction 339
  cholesteatoma surgery 657
  endoscopic sinus surgery 275
  head and neck cancer 345
  head and neck reconstruction 490
  mastoiditis surgery 652–53
  mental capacity assessment 7, 8
  myringoplasty 161
  neck lumps, clinical examination 377
  ossiculoplasty 156
  otosclerosis surgery 150
  paediatrics 561, 564–65, 566
    airway compromise, acute 575
    airway obstruction management 665–66
  rhinoplasty 249, 262
  sphenopalatine artery, endoscopic ligation 200
  thyroidectomy 446
  tonsillectomy 679
continuous positive airway pressure (CPAP) 387, 389, 390, 586
continuous professional development 4
contrast swallow 366, 615
  see also barium swallow
Control of Noise at Work Regulations 2005 126, 130
core biopsy, lymph nodes 636
corniculate cartilages 348
coronal brow lift 298, 299*f*
coronaviruses (CoVs) 19
  see also COVID-19
cor pulmonale, decompensated 13
cortical audiological evoked potential (CAEP) 35, 644, 645
Corynebacterium diphtheria 592, 593
Cottle's test 185
cough
  chronic (cough hypersensitivity syndrome) 555
    clinic 345
    differential diagnosis 216–17
    neck lumps 381
    postnasal drip 215–16
    refractory (CRC) 554–55
  inducible laryngeal obstruction 555
  reflux-related 217
  rhinosinusitis, acute 268
COVID-19
  aerosol- and droplet-generating medical procedures 20–21
  anosmia 19–20
  future of otolaryngology 22
  good medical practice 3, 6
  infection mechanism 19
  Liberty Protection Safeguards 9
  nose and paranasal sinuses, clinical assessment 183, 184, 189
  olfactory disorders 194, 196
  origin and source of transmission 19
  paediatric assessment 561
  personal protective equipment 21–22, 21*t*, 545
  testing 20
  tonsillectomy 595
  tracheostomy 540, 545
cranial nerves (CNs)
  III 329
  V (trigeminal) 132, 168
  VI (abducens) 132, 168, 329
  VII *see* facial nerve
  VIII (auditory/cochleovestibular) auditory nerve (cranial nerve VIII) 29, 48, 55, 132, 168
  IX (glossopharyngeal) 132, 168, 451
  X (vagus) 27, 132, 168, 451
  XI (spinal accessory) 132, 168
  XII (hypoglossal) 132, 390, 439*t*
  anatomy 132
  dizziness, paediatric 662
  dysphagia 359, 359*t*
  lower CN schwannomas 138*t*
  nasopharyngeal cancer 457
  necrotizing otitis externa 177, 179
  otalgia 60*f*
  salivary surgery risk 439, 439*t*
  temporal bone trauma 168, 169
  throat pain 368
craniofacial abnormalities
  cholesteatoma 656
  complications 690
  dysphonia 620
  microtia 689, 689*t*
  otitis media, recurrent acute 651
  otoplasty 689–90
  tympanic membrane perforation 656
    retraction 655
  see also 629
craniopharyngioma 328
craniosynostoses 609, 626, 644, 699–700, 699*f*
cricoid cartilage 348, 401
cricoid split 670
cricopharyngeal dysfunction 361, 362
cricothyroidotomy 392, 393, 394*f*, 394
cricotracheal resection 671
cri-du-chat syndrome 701
critical incident reports 12
Crouzon syndrome 609, 699
cuneiform cartilages 348
cystic fibrosis 287–88, 598, 607
cytology samples, head and neck pathology 520
cytomegalovirus 591–92, 635, 647, 660–61

dacryocystitis 229
dacryocystography 229–31
dacryocystorhinostomy (DCR) 229, 231–34, 232*t*, 233*f*, 234*t*, 235
dacryoscintigraphy 229–31
deep head hanging manoeuvre 116
deep neck space infections 465, 469
  anatomy 465
  clinical presentation and acute management 466–67
  enteral feeding 469
  investigations 467–68

paediatrics 590, 592–93, 634, 635
surgical approaches 468–69
throat pain 370, 371
tonsillectomy 469
tracheostomy 469
deep vein thrombosis (DVT)
hereditary haemorrhagic telangiectasia 320, 324
oral cancer surgery complication 436
prophylaxis 320, 436
dehiscent superior semicircular canal syndrome 660
delirium 8–9
deltopectoral flap 482
dementia 8–9, 123
Dennie–Morgan lines 600
dental disease
otalgia 67
pain 224
sore throat 593
dental procedures
COVID-19 21
extraction 491
depression
dizziness 56, 661
rhinosinusitis, chronic 284
sound tolerance disorders 38
tinnitus 38, 39, 40, 42
Deprivation of Liberty Safeguards (DoLS) 9–10
dermal fillers *see* fillers (facial rejuvenation)
dermatochalasis 298
dermoid cyst 381, 381*t*
anatomy, embryological considerations, and assessment 517–18, 518*f*
assessment 517–18
differential diagnoses 518
embryology 517–18
investigations 518
paediatrics 630–31
treatment 518
developmental milestones 562
dizziness, paediatric 661
deviated nasal septum (DNS)
allergic rhinitis 317
Cottle's classification 238, 239*f*
nasal obstruction 237, 238, 240–41
paediatrics 609–10, 611–12
rhinoplasty 262
diabetes mellitus
anaesthesia 14
dizziness, paediatric 660
neck lumps 376, 382–84
necrotizing otitis externa 63, 176, 179, 180
neuropathy 52
otorrhoea 61
skull base osteomyelitis 138
dietary factors
dizziness 48
food allergies 213
hypopharyngeal cancer 451
Ménière's disease 110
nasopharyngeal cancer 456
postnasal drip 216
reflux-related cough 217
tinnitus 42
vestibular migraine 109
Difficult Airway Society 393, 395
Di George syndrome 644
diphtheria 593, 595
direct acoustic cochlear stimulation 150
disequilibrium 46, 47*f*, 48
see also dizziness
distraction test (hearing assessment) 646
diverticuloscopy 452
Dix–Hallpike manoeuvre 51–52, 51*f*
benign paroxysmal positional vertigo 112–14, 117, 662
temporal bone trauma 171
dizziness
duration 46–48
general observations 46
history taking 46–48
paediatrics 659
examination 662
history 659–62, 659*t*
investigations 54
management 662–63
migraine 106
treatment 663
persistent postural-perceptual 56
scope of the problem 45
see also disequilibrium; light-headedness; vertigo
Down syndrome 644, 679, 692–93, 692*f*
see also trisomy 21
drooling 638
assessment and history 638, 638*b*
controversies 642
differential diagnosis 639
examination and investigation 638–39
tips and pearls 642
treatment/management 639
droplet-generating medical procedures (DGMPs) 20–21
drug-induced sleep endoscopy (DISE) 388, 389, 390, 584–85, 587
drug use by doctors 6
dry eye 228, 231
dual-energy X-ray absorptiometry (DXA) 471, 475
duty of candour 5
dysdiadochokinesia 52
dysgeusia 192*t*
dysosmia 184
dysphagia 356
airway compromise, acute 574
anatomy and physiology 357–58, 358*f*
causes 361–66, 363*f*–65*f*
classification 356–57, 356*f*
clinical assessment 358–60, 359*t*, 360*f*
controversies 366
deep neck space infections 466
diagnostic pathway 356–57, 357*f*
guidelines 366
head and neck cancer rehabilitation 498–99, 501, 504
high 366
hypopharyngeal cancer 451
investigations 360–61, 361*f*, 362*f*
laryngeal cleft 672
laryngeal movement disorders 556
oesophageal 356–57, 356*f*
oropharyngeal 356–57, 356*f*
paediatric 624, 629
aetiology 625–29
anatomy and physiology 624–25
assessment 625
drooling 639
treatment 629
red flags 357*t*
tips and pearls 366
Dysphagia Handicap Index (DHI) 360
dysphonia 348, 354
causes 350–54, 350*b*, 351*f*–53*f*
clinical assessment 349–50
controversies 354
guidelines 354
paediatric 613
assessment 614
differential diagnosis and treatment 616–22
investigations 614–16, 617
spasmodic 354

Eagle syndrome (stylohyoid syndrome) 372, 372*f*, 593
ear
anatomy and physiology 27–30, 27*f*–28*f*, 29*f*–30*f*, 643, 650
embryology 643
tumours 525
ear canal malignancy 64
ear defenders 42, 129
eardrops
cerumenolytic 96
open middle ear 67
ear drum *see* tympanic membrane
ear wax impaction 94–97, 96*f*
Eating Assessment Tool (EAT-10) 360
eating disorders 382–84
echocardiography 609, 631, 636
e-cigarettes and vapes 373
elderly people
benign paroxysmal positional vertigo 112, 115
deep neck space infections 469
dizziness 45, 48, 56–57
dysphagia 361, 363
facial paralysis 74
head and neck cancer 496, 498
hyperparathyroidism 473
migraine 106
necrotizing otitis externa 179, 180
non-allergic rhinitis 217
oral cancer 436
otitis externa, necrotizing 63
paraganglioma 517
perforated tympanic membrane 161
postnasal drip 217
rhinorrhoea 208
tinnitus 129
elective lymph node irradiation 493–94
electrocautery 20–21
electrocochleography (ECochG) 34, 55, 55*f*
electroencephalography 661
electromyography (EMG) 73, 171, 615–17, 621
electroneuronography (ENoG) 73, 171, 653
electronystagmography 53
empty nose syndrome 237, 243, 246
empyema
acute bacterial rhinosinusitis complication 273
acute otitis media complication 653
chronic rhinosinusitis complication 598, 600
encephalitis 640
encephaloceles 86–87, 87*f*
endolymphatic hydrops 106, 110–11, 111*f*
endoscopy
airway abnormality, paediatric 666
anterior skull base approaches 326, 327*f*, 332, 333–39
complications 331–32
controversies 332
coronal plane 327*t*, 329–31, 330*t*, 330*f*
reconstructive options 331
sagittal plane 326–29, 327*t*, 328*f*–29*f*
branchial cleft cyst 512, 513
brow lift 299*f*, 299–300
cerebrospinal fluid rhinorrhoea 213, 214
choanal atresia 611
cholesteatoma 144, 657–58
COVID-19 20, 189
drug-induced sleep (DISE) 388, 389, 390, 584–85, 587
dysphagia 360*f*, 360–61, 365, 366
epiphora 229, 231–32, 233–34, 235
facial pain 224
glioma 611
head and neck pathology 344, 520
hereditary haemorrhagic telangiectasia 320, 321
hypopharyngeal cancer 451, 452
juvenile nasal angiofibroma 612
juvenile nasopharyngeal angiofibroma 605, 606
laryngeal cleft 672, 673
nasal crusting 245
nasal obstruction 240
nasal septal perforation 206
nasopharyngeal cancer 460
neck lumps 380
neck trauma 401
obstructive sleep apnoea 388, 389
ossiculoplasty 156
parathyroidectomy 473
rhinoplasty 262
rhinosinusitis
acute 268
chronic 284–85, 284*f*–85*f*
sinonasal tumours 270, 292, 293, 294
sinus surgery (ESS) 274, 530
complications 276–77, 277*t*, 278*f*
consent 275
controversies 279
image guidance 276

endoscopy (*cont.*)
indications 274
outcomes 278
packing and stents 276
perioperative interventions 275
postoperative medication 277–78
preoperative preparations 274–75, 275*t*
recovery 278
revision rates 278–79
rhinosinusitis, chronic 284–85, 284*f*–85*f*
technique 275–76, 275*f*, 276*f*, 277*f*
tips and pearls 279
sphenopalatine artery ligation 200, 201*f*, 201
subglottic stenosis 671
three-dimensional 279
tracheal stenosis 672
tympanoplasty 161, 162*f*, 162
see also nasendoscopy
enteral feeding 469
environmental issues, and anaesthesia 15, 16
eosinophilic granulomatosis with polyangiitis (EGPA) 242, 243*t*, 245, 288
eosinophilic oesophagitis 358, 359, 360–61
epidermoids 135–36, 138*t*, 138
epididymitis 640
epiglottis, anatomy 348
epiglottitis 573, 592, 595
epilepsy 661
epiphora 227
assessment and investigation 228–31, 231*t*, 232*f*
causes 228, 230*f*
controversies 233–34
tips and pearls 234–35
treatment 231–33, 232*t*, 233*f*–34*f*, 234*t*
see also lacrimation
episodic ataxia type 2 58, 106
epistaxis 198, 202
acute management 198–99
anatomy 198, 198*f*, 596
causes 199*b*
drugs implicated in 199*b*
nasal sprays as cause 243
controversies 201–2, 606
COVID-19 20
differential diagnosis 200
guidelines 202*b*, 202
hereditary haemorrhagic telangiectasia 319, 320, 321, 322, 323, 324–25
investigations 199–200
juvenile nasopharyngeal angiofibroma 604
paediatrics 596, 603–4, 606, 607
treatment 200, 200*b*, 201*f*
Epley manoeuvre 112, 114–15, 116*f*, 116, 117
Epstein–Barr virus (EBV)
cervical lymphadenopathy 635
glandular fever 591–92
nasopharyngeal cancer 456, 458–60, 523, 525
squamous cell carcinoma 521
Epworth Sleepiness Scale 386, 387
for Children and Adolescents (ESS-CHAD) 583
erythema multiforme 410
erythroplakia 428, 429*f*
Escherichia coli 142
essential tremor 354
ethesioneuroblastoma *see* olfactory neuroblastoma
ethical issues 10
ethmoid arteries 200, 324
ethmoid sinusitis 600
ethnicity factors, obstructive sleep apnoea 580, 585
European Academy of Facial Plastic Surgery (EAFPS) 307, 313
European Board for certification in Facial Plastic and Reconstructive Surgery (EBFPRS) 307
Eustachian tube (ET)
anatomy and physiology 27–28, 69
dysfunction
aural fullness 69–70, 70*t*
cleft lip and/or palate 688
Down syndrome 692
hearing loss 644
otitis media with effusion 682
tympanic membrane retraction 655
paediatrics 650
patulous 32, 33*f*, 37, 40, 41, 69
Ewald's laws 50, 112–13
excision biopsy 521, 521*f*, 636
exercise-induced laryngeal obstruction (paradoxical vocal fold dysfunction) 555, 619
exercise stress laryngoscopy 619
exostoses, external auditory canal 94*f*, 94, 95*f*–96*f*
external beam radiotherapy (EBRT) 434, 436–37, 460
external ear 92
anatomy and physiology 27, 27*f*, 28*f*, 131–32
atresia
acquired 102
congenital 99–102, 100*f*–1*f*
cerumen impaction 94–97, 96*f*
cholesteatoma 103–4, 103*t*, 103*f*
conductive hearing loss 84
exostoses and osteomas 94*f*, 94, 95*f*–96*f*
foreign body 92–93, 92*f*, 93*f*, 654
hearing loss 643, 644
keratosis obturans 102–3
malformations 97–98
differential diagnosis and associated conditions 98–102, 99*f*–101*f*
Jahrsdoerfer grading system 98*t*
see also microtia
otalgia and otorrhoea 61–64
temporal bone trauma 167–68
see also pinna
extranasal pedicled vascular flaps 337–38
extra-oesophageal reflux disease (EORD) 287
ex utero intrapartum treatment (EXIT) 578, 667
eyebrows
non-surgical rejuvenation 309
surgical rejuvenation 298*f*, 299*f*, 300
eyelids, surgical rejuvenation
lower eyelid 300–4, 302*b*, 303*f*, 304*f*, 304*b*, 305*f*
upper eyelid 300, 301*f*, 302*f*

face
analysis, rhinoplasty preparation 247–49, 249*b*
elevator and depressor muscles 311*t*
examination 184–85
non-surgical rejuvenation 309, 310*t*, 311*f*, 312, 313
botulinum toxin 309–10, 311*b*, 311*t*
fillers 310–12, 311*t*
regulations 313
rhytids 309, 311*f*, 311*b*
surgical rejuvenation 297, 297*f*, 307
lower face and neck 304–7, 306*f*–7*f*
upper face 298*f*, 298–304, 299*f*, 301*f*, 302*f*, 303*f*, 304*f*, 305*f*
facelift 305–7, 306*f*–7*f*
facial artery myomucosal flap 482, 483–84
facial nerve (cranial nerve VII) 72
acoustic reflex texting 32–34
anaesthesia 16
anatomy 72, 305*f*
atresia surgery 101–2
dehiscence 149, 150*f*
facial therapy and reanimation 75–76
haemangioma 138*t*
hearing loss, paediatric 643–44
House–Brackmann grading 73*t*, 73, 169, 170*t*
identification 441*t*
jugular foramen paraganglioma 136
lateral skull base disease 132
necrotizing otitis externa 179
otitis media, acute 652
palsy 60
parotid gland surgery 440–41, 441*f*, 442, 641–42
pathology 73–75, 73*t*, 75*f*
physiology and special investigations 72–73, 73*t*
salivary gland surgery risk 439
schwannoma 138*t*
sound tolerance disorders 38
Sunnybrook grading system 169, 170*t*
surgical facial rejuvenation 304, 305, 307
trauma 402
temporal bone 168, 173, 173*f*, 174, 175
tumours 137
differential diagnosis 82*t*
facial pain 183–84, 220, 225
assessment 220, 222*t*
atypical (persistent idiopathic facial pain) 222*t*, 224, 224*b*
clinical features and treatment 223–25, 224*b*
controversies 225
differential diagnosis 221, 221*f*
endoscopic sinus surgery 274
guidelines 225
investigation 220, 221*f*
midfacial segment pain 222*t*
rhinosinusitis
acute 270
chronic 284
facial palsy 653
facial trauma 402
fractures of facial skeleton 403–4, 404*f*
paralysis 74
soft tissue injury 402–3
failure to thrive 563, 564, 583, 625–26, 665, 687–88
falls 45, 112
familial hypocalciuric hypercalcamia (FHH) 471, 473
FeverPAIN criteria 594
Feyh–Kastenbauer retractor 452
fibreoptic endoscopic evaluation of swallow (FEES) 625, 627–28
fibreoptic endoscopy 451
fibreoptic nasopharyngolaryngoscopy 392, 393
fibular osteocutaneous flap 485, 488
fillers (facial rejuvenation) 310–12, 311*t*, 313
complications 312
regulations 313
fine needle aspiration 520
branchial cleft cyst 512
neck lumps 384
oropharyngeal cancer 420–21
paraganglioma contraindication 516
salivary gland tumours 439, 641
thyroglossal duct cyst 514
thyroid 445, 446, 447–48, 536–37
finger–nose–finger test 52
first aid, epistaxis 199
first bite syndrome 372, 439, 439*t*, 442
fistulography 512, 513
flamingo flush 148
flexible endoscopic evaluation of swallowing (FEES)
dysphagia 361, 362*f*, 362, 363, 365
head and neck cancer rehabilitation 499, 500
Floseal 201
fluorescein, intrathecal 212
fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT) 179, 434
head and neck 530
nasopharyngeal cancer 460
fluoroscopy 527
flying
after stapes surgery 153
after tympanoplasty 165
Fontaine's sign 379–80
forehead
flap 482
surgical rejuvenation 298*f*, 298

foreign body
  airway obstruction, acute 571, 573, 578
  dacryocystorhinostomy silastic tubes, and epiphora 234
  deep neck space infections 466
  dysphagia 358
  epistaxis 603
  external auditory canal 92–93, 92*f*, 93*f*
  external ear 92–93, 92*f*, 93*f*, 654
  nasal blockage 609
  nasal crusting 243
  rhinorrhoea 210
  throat pain 372
  tympanic membrane perforation 160–61
free flaps, pedicled 338
free radial forearm flap (FRAFF) 485, 487, 488, 489
Frey syndrome (auriculotemporal syndrome) 439, 439*t*, 441, 442
Friedman mouth opening score 386, 389
Fukuda's test 662
functional endoscopic sinus surgery *see* endoscopy: sinus surgery
functional hearing loss 35
furunculosis 62–63
Fusobacterium necrophorum 592, 593
future of otolaryngology 22

gait assessment 52, 662
gait ataxia 52
gastric cancer 361
gastro-oesophageal reflux disease (GORD)
  differential diagnosis 217
  dysphagia 359, 361, 362, 628
  dysphonia 620
  rhinosinusitis, chronic 598
gaze-evoked nystagmus (GEN) 49
gaze stabilization 45, 45*f*, 46
gender factors
  airway compromise, acute 574
  Alport syndrome 697
  benign paroxysmal positional vertigo 112
  branchial cleft cyst 508
  cholesteatoma 656
  cough, chronic refractory 555
  craniosynostosis 699
  dysphagia 358, 359
  dysphonia 619
  epiphora 227
  Goldenhar syndrome 693
  head and neck reconstruction 488
  hereditary haemorrhagic telangiectasia 323
  hyperparathyroidism 471
  hypopharyngeal cancer 450
  juvenile nasopharyngeal angiofibroma 604
  laryngeal anatomy and physiology 613–14
  masseter hypertrophy 309–10
  medication overuse headache 225
  microtia 689
  migraine 220, 221–22
  nasal crusting 244
  nasal growth 608
  oropharyngeal cancer 419
  otosclerosis 148
  paraganglioma 516
  peak nasal inspiratory flow 240, 240*t*
  phonosurgery 553
  puberphonia 354, 557
  rhinorrhoea 208
  sensorineural hearing loss 79
  sinonasal tumours 292
  surgical facial rejuvenation 300, 307
  thyroid cancer 444
  tinnitus 37–38
  Turner syndrome 694
gender reassignment, male-to-female 553
general anaesthesia *see* anaesthesia
General Medical Council (GMC), 'Good Medical Practice' 3, 6
  domains 3, 4*t*
  evaluation, reflection, and then action 4
  roles and responsibilities 3
genetic factors
  achondroplasia 700
  Alport syndrome 697
  Beckwith–Wiedemann syndrome 701
  branchio-oto-renal syndrome 695
  CHARGE syndrome 609, 698
  cleft lip and/or palate 687, 688*t*
  craniosynostosis 699
  cri-du-chat syndrome 701
  dizziness 57, 58
  Down syndrome 692
  episodic ataxia type 2 106
  Goldenhar syndrome 693
  head and neck cancer 411
  hearing loss 643, 644, 646
    conductive 81–84
    sensorineural 77, 77*t*
  hereditary haemorrhagic telangiectasia 319, 320*t*, 323, 324
  hyperparathyroidism 470, 473
  Jervell and Lange-Nielsen syndrome 696
  Möbius syndrome 701
  mucopolysaccharidosis 701–2
  nasopharyngeal cancer 456
  neurofibromatosis
    type 1 697
    type 2 697
  osteogenesis imperfecta 702
  otitis media
    with effusion 681–82
    recurrent acute 651
  otosclerosis 148
  paraganglioma 516
  Pendred syndrome 695
  Prader–Willi syndrome 702
  rhinosinusitis, chronic 598
  schwannoma 515
  Stickler syndrome 696–97
  thyroid cancer 447–48
  tinnitus 37–38
  Treacher Collins syndrome 693
  Turner syndrome 694
  Usher syndrome 695
  VACTERL association 703
  velocardiofacial syndrome 617, 700
  vestibular migraine 106
  Waardenburg syndrome 694
Gillick competence 564–65, 566
glandular fever 370, 591–92, 595
Glasgow–Edinburgh Throat Scale 365
glioma 610, 611, 631
Global Tracheostomy Collaborative 543
globus pharyngeus 358, 361–62, 451
glomangiopericytoma 292
glomus tumour *see* paraganglioma
glossectomy, median 389
glossopharyngeal nerve *see* cranial nerves: IX
glottoplasty 553
glue ear *see* otitis media: with effusion
goitre 365–66, 365*f*, 384
Goldenhar syndrome 644, 646, 647*f*, 688*t*, 693
good medical practice 3, 6
  appraisal and revalidation 3
  care of self and others 6
  domains 4*t*
  evaluation, reflection, and then action 4
  keeping up to date 4
  probity 5–6
  review and audit of practice 4–5
  roles and responsibilities 3
  working with colleagues and patients 5
  see also General Medical Council (GMC), 'Good Medical Practice'
gout typhus 82*t*
Gradenigo syndrome 653
grade of hoarseness, roughness, breathiness, asthenia, strain (GRBAS) 350
granuloma
  cholesterol 137, 138*t*
  laryngeal 353, 353*f*
  lethal midline 245
  post-traumatic 574–75
  vocal cord 618
  vocal process 550
granulomatosis, orofacial 410
granulomatosis with polyangiitis (GPA)
  differential diagnosis 288
  dizziness 56
  nasal crusting 242–43, 243*t*, 244, 244*f*, 245–46
  nasal septal perforation 205, 207
  oral ulceration 410
granulomatous disease 381
Grisel syndrome (atlantoaxial subluxation) 679, 692–93
grommets *see* ventilation tubes
gross motor assessment, paediatric dizziness 662
group A beta-haemolytic *Streptococcus* (GABHS) 590, 591, 592, 594
growth charts 563, 564
Gufoni manoeuvre 115–16

Hadad–Bassagasteguy flap 337
haemangioma 631–32
  airway compromise, acute 573, 576–77
  facial nerve 137
  facial paralysis 74
  salivary glands 641, 641*f*
haemangiopericytoma 292
Haemophilus influenzae
  epiglottitis 592, 595
  otitis media
    acute 64, 651
    with effusion 681–82
  pharyngitis 592
  prophylaxis 172–73, 174
  rhinosinusitis
    acute 267, 597
    chronic 598
  tonsillitis 591
haemorrhage, tracheostomy complication 541
halo sign, temporal bone trauma 171
harassment 5
Hashimoto's disease 384
Hayes Martin manoeuvre 436
headache 220, 225
  assessment 220, 222*t*
  clinical features and treatment 221–23, 222*t*, 223*b*, 224–25, 224*b*, 225*b*
  cluster 210, 220, 222*t*, 223
  controversies 225
  differential diagnosis 221, 221*f*
  dizziness, paediatric 661
  guidelines 225
  investigation 220, 221*f*
  medication overuse 223, 224*f*, 225
  red flags 109*b*
  tension-type 222*t*, 223, 223*b*
  see also migraine
head and neck anatomy 527, 528*f*, 528*t*, 529*f*
head and neck cancer (HNC) 343
  anaesthesia 13, 14
  clinic set-up in secondary care 343
  diagnostic stage 346
  dysphagia 358–59, 364*f*, 364
  ear tumours 525
  examination 344
  functional outcome measures 498*t*
  future 347
  history 343–44, 344*b*
  imaging sequence 345
  joint clinic models 345
  lymphomas 525
  malignant mesenchymal tumours 526
  metastatic carcinoma of unknown primary 525
  mucosal malignant melanoma 525–26
  multidisciplinary team 344, 345–46, 345*b*, 346*b*, 346*t*
  neck lumps 380, 381, 382–84
  non-surgical treatments 491
    acute toxicities and supportive care 495
    assessment and post treatment surveillance 495–96
    future developments 496
    neoadjuvant/induction chemotherapy 495
    palliative systemic therapies 496
    radical re-irradiation 496

head and neck cancer (HNC) (*cont.*)
radiotherapy 491–94
radiotherapy, palliative 496
radiotherapy with concomitant chemotherapy 493–95
one-stop lump clinic 344–45
oral ulceration 406, 409
paediatrics 635
post-treatment and follow-up stage 346–47
pre-habilitation 499
red flags 371
rehabilitation 498, 504
assessment 498–99, 498*t*, 499*t*
interventions 499–501, 500*t*, 501*t*
laryngectomy 502–4, 502*t*, 503*f*
late effects 501–2
salivary gland 438, 523–25, 524*t*, 524*f*–25*f*
sinonasal tumours 523
squamous cell carcinoma 521
laryngeal 522, 522*f*
oropharyngeal 522–23, 523*f*
symptoms 343, 343*b*
throat pain 369–70, 371–72
treatment stage 346
urgent referral pathway 343
see also head and neck reconstruction; hypopharyngeal cancer; laryngeal cancer; nasopharyngeal cancer; oral cancer; oropharyngeal cancer; pharyngeal cancer; thyroid cancer
head and neck pathology 520, 526
cancer *see* head and neck cancer
interpreting the pathology report 521
pathology request form 520
sample acquisition 520
sample pathway 520–21, 521*f*
head and neck reconstruction 477–78
assessment and preparation 477
common complications and their prevention 490
controversies 489
decision-making 478
flaps
free 485, 487–88
local skin 481–84
regional 482–86
future 490
individual techniques 489
pharyngeal reconstruction 488, 489
principles 477
'reconstructive ladder' 478–81
rehabilitative team roles 478
safety measures 477
tips and pearls 490
head impulse test (HIT) 50
video 50, 54
head injury
cerebrospinal fluid rhinorrhoea 211
dizziness, paediatric 661
tinnitus 38
head shaking nystagmus test 662
head thrust test 662
Health Education England 313
hearing aids 36, 119
aural atresia 102
cholesteatoma 144
conductive hearing loss 85
Down syndrome 692
external ear malformations 98
as foreign bodies 92, 92*f*
manufacturers, and cochlear implants 122
Ménière's disease 110
mixed hearing loss 85
ossicular chain abnormalities 89, 156
osteosclerosis 149, 150, 153
otitis externa 62
otitis media with effusion 683, 688
otomycosis 62
paediatric 648
perforated tympanic membrane 161
sensorineural hearing loss 79, 80
stapes surgery 153
temporal bone trauma 89, 174
tinnitus 41, 42, 80
hearing assessment
age-appropriate 644–46, 650, 662
sound field audiometry 30
see also pure tone audiometry
hearing implants *see* audiovestibular implants
hearing loss
cholesteatoma 141, 142, 144*t*, 644
dementia 123
dizziness 48, 660–61
Down syndrome 692
epidermoids 135–36
functional 35
Ménière's disease 105, 109, 110
meningiomas 134–35
necrotizing otitis externa 177
osteogenesis imperfecta 149
otitis media with effusion 682
otosclerosis 89, 148–49, 153
paediatric 643
assessment 644–46
clinical assessment 644, 644*f*, 645*f*
management options 648–49
multidisciplinary management 648
permanent 646–47, 647*f*
tinnitus 38, 41, 42
Turner syndrome 694
tympanoplasty complication 163
vestibular migraine 105
vestibular schwannoma 133
see also xix; mixed hearing loss; noise-induced hearing loss; sensorineural hearing loss
hearing protection devices 42, 129
hearing rehabilitation 35–36
hearing tests *see* hearing assessment
hearing therapy 35
heel-to-shin test 52
Heerfordt syndrome 640
helicotrema 28–29
hemifacial microsmia *see* Goldenhar syndrome
Hennebert's sign 53, 84, 171, 660
hepatitis 376
hereditary haemorrhagic telangiectasia (HHT) 320, 324–25
controversies 323–24, 323*t*
demographics 319
diagnosis 319–20, 319*b*, 319*f*–20*f*, 320*t*
differential diagnosis 320
guidelines 324, 324*b*
management 320–23, 321*f*, 322*b*, 322*f*, 323*b*
nomenclature 319
herpes simplex virus (HSV) 406, 409–10, 647
herpes zoster oticus (HTO) 48, 66
high-resolution computed tomography (HRCT)
cerebrospinal fluid rhinorrhoea 211–12
hereditary haemorrhagic telangiectasia screening 324
temporal bone trauma 171, 172*t*, 174
'HINTS' and 'HINTS Plus' acronyms (acute vertigo) 55
Hippocratic oath 3, 375
HIV
lymphadenopathy 635
neck lumps 376, 377, 382–84
necrotizing otitis externa 176, 179
salivary glands 438, 640
vertigo 48
Hodgkin lymphoma 525
honesty 5–6
hospitality 6
House–Brackmann classification 169, 170*t*
human immunodeficiency virus *see* HIV
human papillomavirus (HPV)
dysphonia 613, 618–19
head and neck cancer, non-surgical treatment 495
hypopharyngeal cancer risk 451
neck lumps 376, 380, 384
oropharyngeal cancer 419
de-escalation controversy 426
non-surgical treatment 494–95
squamous cell carcinoma 522–23, 525, 531–32
symptoms 371
recurrent respiratory papilloma 352–53
squamous cell carcinoma (SCC) 521
head and neck pathology 523
oropharyngeal 522–23, 525, 531–32
vaccine 620–21
Hunter syndrome 701, 702
Hurler–Scheie syndrome 701
Hurler syndrome 701, 702
hyaluronic (HA) fillers (facial rejuvenation) 310–11, 311*t*, 312
hydrocephalus 52
hyperacusis 37
associations 38
conductive 48
epidemiology 38
hearing protection devices 42
investigations 40
malleolar sign 52
treatments 41, 42
see also sound tolerance disorders
hyperbaric oxygen 179–80
hyperglycaemia 56
hypermetropia 661
hyperosmia 192*t*
hyperparathyroidism
primary (PHPTH) 470
aetiology 470
assessment 471–72
controversies 475
genetic testing 473
revision surgery 474–75
treatment 473–74, 475
secondary 470, 475
tertiary 470, 475
hypertension
epistaxis 199
idiopathic intracranial 56, 86–87
hyperthyroidism 537
hyperventilation 57
hyperventilation-induced nystagmus 53
hypoglossal nerve *see* cranial nerves: XII
hypoglycaemia 56
hypoparathyroidism 475
hypopharyngeal cancer 450
assessment and investigations 451
differential diagnoses 451
guidelines 454
prognosis 450, 454
risk factors 450–51
treatment 452–54
hypopharynx (laryngopharynx)
anatomy 368, 369*t*, 451
assessment 370
dysphagia, paediatric 627–28
imaging 532–35
physiology 451
hypopnoea 582
hyposmia 192*t*
hypotension
orthostatic 56, 660
spontaneous intracranial 213
hypothyroidism 377, 384
hypoventilation, sleep-related 585

idiopathic intracranial hypertension (IIH) 56, 86–87
image-guided radiotherapy (IGRT) 492
imaging 527, 537
airway obstruction, acute 571
anatomy 527, 528*f*, 528*t*, 529*f*
cerebrospinal fluid rhinorrhoea 211–12
conductive hearing loss 84
deep neck space infections 467–68
dizziness, paediatric 662
endoscopic sinus surgery 276, 279
facial pain 225
head and neck cancer 345
hyperparathyroidism 471–72
laryngeal cancer 411–12
lateral skull base disease 132–33
lymph nodes and cystic neck masses 537, 537*f*–38*f*, 539*b*
modalities 527–30

see also xix; echocardiography; magnetic resonance cisternography; magnetic resonance imaging; positron emission tomography; single-photon emission computed tomography; ultrasound; X-rays
necrotizing otitis externa 178*f*, 178–79
nose and paranasal sinuses 187
olfactory disorders 192–93
oropharyngeal cancer 419–21
otitis media, acute 650–51
paraganglioma 516
pharynx 531–35, 533*f*–34*f*
rhinoplasty 249
rhinosinusitis, chronic 286, 287*f*
salivary glands, major 535–36, 535*f*
skull base and sinonasal disease 530–31, 530*f*–31*f*, 532*t*–30*t*
sleep-related breathing disorders 387
thyroid 536–37, 536*f*, 536*t*
immunization history 562
immunocompromised patient
anterior skull base reconstruction 333
nasal crusting 242
necrotizing otitis externa 176–77
oral ulceration 406, 409–10
otitis externa, necrotizing 63
otitis media, acute 651
rhinosinusitis, acute 268–69, 271, 273
skull base osteomyelitis 74, 138
immunoglobulin G4 disease 379
immunotherapy
allergic rhinitis 602, 606, 607
head and neck cancer 496
nasopharyngeal cancer 462
incudostapedial joint, reconstruction 156–57, 156*f*
incus
anatomy and physiology 27–28
dislocation 152
embryology 643
eroded 152*f*, 152
independent mental capacity advocate (IMCA) 9
inducible laryngeal obstruction (ILO) 554*f*, 555, 574
infants
airway abnormality 666, 667
airway obstruction, acute 572, 573
dysphagia 625–26, 628
dysphonia 616–17
hearing loss 643
nasal anatomy and physiology 608
nasal blockage 609, 610
salivary glands 641
swallowing process 624
see also paediatrics
infectious mononucleosis *see* glandular fever
inferior turbinate
flap, posteriorly pedicled 337
hypertrophy 610
reduction 218
informed consent *see* consent
infratemporal fossa 330
inner ear
anatomy and physiology 27, 27*f*, 28–30, 29*f*–30*f*
autoimmune inner ear disease 56
balance 46
temporal bone trauma 168
insomnia 39, 40, 48
Insomnia Severity Index 40
intensity-modulated radiotherapy (IMRT) 323, 491, 492, 496
nasopharyngeal cancer 457, 458, 459, 460
International Union Against Cancer (UICC) 458, 461
internuclear ophthalmoplegia 50
intracerebral abscess 653
intranasal flaps 337
Iowa Oral Performance Instrument 499
iron deficiency anaemia 451
Islam 161
island nasolabial flap 481
Ixodes tick 74

jaw tumour syndrome 470
jejunal flap 488, 489
Jerger classification system 32
Jervell and Lange-Nielsen syndrome 643–49, 696, 696*f*
joint voice clinics 345
Jones 1 and 2 tests 229
jugular bulb
high 82*t*, 88
temporal bone trauma 169
jugular foramen tumours 136–37
juvenile nasopharyngeal angiofibroma (JNA) 603, 604–6
airway compromise, acute 574
controversies 606
differential diagnosis 610
treatment 612
juvenile recurrent parotitis 640, 642

Kabuki syndrome 688*t*
Kadish staging, sinonasal tumour 291
Kallman syndrome 195
Karapandižić technique 482, 484
Karl Storz Steiner distending laryngoscope 452
Kartagener's syndrome 598
Kawasaki's disease 634, 636
keratosis obturans (KO) 64, 102–3
Khalfa Hyperacusis Questionnaire 40
Kiesselbach's plexus 198
Klebsiella spp.
ozaenae 244
pneumoniae 142
Klein–Waardenburg syndrome 694
kyphosis 363

labyrinthitis 653, 660
LacQ questionnaire 229, 232*f*
lacrimation
excessive *see* epiphora
science 227–28, 228*f*–29*f*
lagophthalmos 229
Langerhans cell histiocytosis 635, 654
laryngeal atresia 667, 667*f*
laryngeal cancer 354, 411
examination 411
history 411
investigation 411–12
management 412–17, 412*b*, 413*f*, 413*b*, 414*b*, 415*f*, 415*b*, 416*b*
risk factors 411
laryngeal cleft 668–69, 668*f*–69*f*
dysphagia 627–28
management 672–73
paediatric syndromes 691
timing of surgery 673
laryngeal dysplasia 417
laryngeal dystonia 354
laryngeal framework surgery 552–53
laryngeal granuloma 353, 353*f*
laryngeal nerve
recurrent (RLN)
anatomy 349, 613
hyperparathyroidism 475
paralysis 552, 556
phonation 349
thyroid gland 444, 448
trauma 401
reinnervation 621, 622
superior 349, 556, 613
laryngeal web 572, 617, 618, 692
laryngectomy
head and neck cancer rehabilitation 499, 501–4, 502*t*, 503*f*
hypopharyngeal cancer 453
laryngeal cancer 412–13, 414
laryngeotracheobronchoscopy 584–85
laryngitis 353, 392
laryngocele 380–81
laryngomalacia 571–73, 575–77, 628
laryngopharyngeal reflux (LPR) 217, 353, 593, 616
laryngopharyngectomy 450
laryngopharynx *see* hypopharynx
laryngoscopy
airway compromise, acute 570, 575–78
airway obstruction 393
exercise stress 619
laryngeal cancer 411, 412
laryngeal movement disorders 555, 557, 558
neck masses, cysts, and sinuses 635
video 393
laryngospasm 46, 554*f*
laryngostroboscopy 349–50
laryngotracheal disorders, congenital 665
assessment 665–66
controversies and future 673–74
differential diagnoses 667–70, 667*f*–69*f*
investigations 666–67, 666*f*
treatment 670–73, 671*f*
laryngotracheal injury 399–401, 400*f*
Schaefer Classification System 400*t*, 400
laryngotracheobronchitis 392
laryngotracheoplasty 617
larynx
anatomy and physiology 348–49, 391, 391*f*, 613–14
artificial 502*t*, 502, 504
congenital stenosis 667, 667*f*–68*f*
cysts, benign 572–73
dysphagia 627–28
dystonia 556
exercise-induced laryngeal obstruction 555, 619
granuloma 574
imaging 532–35, 534*f*
inducible laryngeal obstruction 554*f*, 555, 574
movement disorders 354, 554, 558
functional voice disorders 557–58, 557*f*
neuromuscular conditions 555–57
upper airway 554–55, 554*f*
non-selective reinnervation 553
papillomatosis 550
phonosurgery *see* phonosurgery
selective adductor denervation-reinnervation 553
squamous cell carcinoma 522, 522*f*
trauma 574
laser ablation, carbon dioxide 20–21
laser peels 312
laser surgery 412, 413*f*
lasting power of attorney 8–9
lateral nasal wall flap, posteriorly pedicled 337–38
lateral skull base disease 131, 139
anatomy 131–32, 131*f*, 132*t*
arachnoid cysts 136
diagnostic evaluation 132
epidermoid 135–36
facial nerve tumours 137
jugular foramen tumours 136–37
lipoma 135, 135*f*
meningioma 134–35, 135*f*
petrous apex lesions 137–38, 138*t*
petrous apicitis 139
skull base osteomyelitis 138
vestibular schwannoma 132–34, 133*f*, 135*f*
Le Fort fractures 404*f*, 404
legal claims 5
leishmaniasis 244
Lemierre's syndrome 465, 593
Lempert 360˚ roll manoeuvre 115–16, 117
leprosy 244
lethal midline granuloma 245
leukaemia 382–84
leukoplakia 351*f*, 351, 428, 429*f*
Liberty Protection Safeguards 9
lichen planus 410, 429*f*
light-headedness 46, 47*f*, 48
see also dizziness; vertigo
limb coordination tests 52
limb dysmetria 52
Limberg flap 481, 482
lingual nerve 439*t*
lingual thyroids 574
lipoma 135, 135*f*, 138*t*, 381
lipreading 36
listening tactics 648
Little's area 198, 198*f*, 199
local anaesthesia 14, 15, 16

log roll manoeuvre *see* Lempert 360° roll manoeuvre
long-segment tracheal stenosis, congenital 572
lordosis 363
loss of consciousness 48
Ludwig's angina 466–67
lumbar drain 331–32, 338, 339
Lund–Kennedy Endoscopic Score (LKES) 189
Lund–Mackay Score (LMS) 189
lupio pernio 245
Lyme disease 48, 57, 74
lymphadenectomy 439
lymphadenitis 634–35
lymphadenopathy 378–79, 634, 635
lymphatic malformations 632–33, 636
lymph nodes
  abscess 633–34
  elective irradiation 493–94
  imaging 537
lymphocele 381
lymphoma 378–79, 381, 384, 525
  HIV-associated 635
  salivary glands 641
  sore throat 593, 594
  thyroid cancer 445
lymphoscintigraphy 381

magnetic resonance cisternography 212
magnetic resonance imaging (MRI)
  airway compromise, acute 572
  airway obstruction 392
  atlantoaxial subluxation 679
  audiovestibular implants 123–24
  branchial cleft cyst 511
  cholesteatoma 144, 146, 147, 657
  dermoid cyst 518
  diffusion-weighted imaging (DWI) 146, 147
  dizziness 662
  dysphagia 365, 366, 626
  dysphonia 615
  epiphora 231
  first bite syndrome 372
  headache 220
  head and neck 527, 529*f*, 529
  head and neck cancer 493, 635
  hereditary haemorrhagic telangiectasia screening 324
  hyperparathyroidism 475
  hyperthyroidism 537
  hypopharyngeal cancer 532–35
  juvenile nasopharyngeal angiofibroma 604–5
  laryngeal cancer 532–35
  lateral skull base disease 137, 138*t*
  nasal obstruction/blockage 238, 609
    glioma 610
    juvenile nasal angiofibroma 610, 612
    meningoencephalocele 610
  nasopharyngeal cancer 458, 460, 532, 534*f*
  neck lumps 379, 380, 381, 382, 384, 635
    glioma 631
    haemangioma 631
    meningoencephalocele 631
    midline nasal dermoids 631
  necrotizing otitis externa 178*f*, 178–79, 180
  nose and paranasal sinuses 187
  obstructive sleep apnoea 585
  olfactory disorders 193, 195
  oral cancer 434
  oral ulceration 409
  oropharyngeal cancer 419–20, 421, 533*f*
  otitis media, acute 650
  paraganglioma 516, 516*f*, 517
  rhinorrhoea 210
  rhinosinusitis
    acute 269, 270*f*, 273
    chronic 286
  salivary glands 535*f*, 536
    haemangioma 641, 641*f*
    paediatrics 640
    tumours 641
  schwannoma 515
  sinonasal tumour 291, 294
  skull base and sinonasal disease 530–31, 530*f*
  temporal bone trauma 171, 172*t*
  thyroid cancer 447
  thyroglossal duct cyst 513
  trigeminal neuralgia 223
  wide vestibular aqueduct 647
malleolar sign 52
malleostapedotomy 85
malleus 27–28, 643
malnourished patients 13
maltreatment of child 563
mandible fracture 403–4
mandibulofacial dysostosis *see* Treacher Collins syndrome
manometry, high-resolution 361
Marie–Foix manoeuvre 169, 171*b*
masseter hypertrophy 309–10
mastoidectomy
  cholesteatoma 103, 143*f*
  COVID-19 20–21
  facial paralysis 74
  jugular foramen paraganglioma 136
  nasopharyngeal cancer 460
  necrotizing otitis externa 177
  otitis media, acute 652, 653
mastoiditis 652–53, 654*f*
maxillary artery 200
MD Anderson Dysphagia Index (MDADI) 360
measles 148
medication overuse headache 223, 224*f*, 225
medullary thyroid cancer 537
melanoma, malignant 377, 525–26
membranous labyrinth 28
Ménière's disease (MD) 105
  auditory evoked potentials 34
  diagnosis 105–6, 105*b*
  differential diagnosis 106
  dizziness 46–48, 53, 55, 57
  endolymphatic hydrops 106, 110–11, 111*f*
  examination 109
  future 110–11
  guidelines 111
  history 108
  investigations 109
  natural history and epidemiology 106–8, 107*f*
  paediatrics 660–61, 663
  pathophysiology 106
  patient experience 110
  sensorineural hearing loss 78–79
  tinnitus 38, 40, 42
  treatments 109, 110
meningeal artery, middle 169
meningioma 134–35, 135*f*, 138*t*, 332
meningitis
  acute bacterial rhinosinusitis complication 273
  acute otitis media complication 653
  anterior skull base reconstruction complication 339
  cerebrospinal fluid rhinorrhoea 210–11, 212–13
  dizziness 660–61
  hearing loss 647
  prophylaxis 611
  temporal bone trauma 169
meningocoele 169, 631
meningoencephalocoele 169, 631
  anterior cranial fossa 291
  conductive hearing loss 87
  cribriform 337
  differential diagnosis 82*t*
  nasal blockage 610, 611
Mental Capacity Act (MCA, 2005) 7, 7*b*, 9
mental capacity assessment 7–9, 8*b*
mental health/illness in doctors 6
mesenchymal tumours, malignant 526
microlaryngobronchoscopy 627, 666, 666*f*, 671
microscope, operating 161, 162*f*
microtia 97, 97*f*, 689, 689*t*
  hearing loss 643
  treatment 98–99, 99*f*
middle ear (tympanic cavity)
  anatomy and physiology 27*f*, 27–28, 69, 142*f*–43*f*
  conductive hearing loss 84
  implants *see* middle ear implants
  malignancy 82*t*, 88, 654
  masses 87, 88*f*
  myoclonus 32, 40
  open, and eardrop usage 67
  otalgia and otorrhoea 64–66
  reflexes 31–34, 33*f*
  surgery 16, 20–21, 74
  see also otitis media
middle ear implants
  active (AMEIs) 119, 120, 121*f*, 122*f*
    guidelines 119
    ossicular chain abnormalities 156
    outcomes 123
  passive 120
Middle Ear Risk Index (MERI) 155, 155*t*
Middle East respiratory syndrome (MERS) 19
middle turbinate flap, posteriorly pedicled 337
middle turbinate flip flap 337
midface fracture 404*f*, 404
midline cervical cleft 633
midline nasal dermoids 631
mid-nasal stenosis 609, 611, 626
migraine
  assessment 220
  clinical features and treatment 221–23, 222*t*
  dizziness 46, 48
    investigations 54
    paediatric 660, 661, 663
    psychiatric disorders 56
  familial hemiplegic 106
  guidelines 111
  sound tolerance disorders 38
  tinnitus 38
  vestibular 48, 57, 105, 663
mindfulness 42
misokinesia 37
  see also sound tolerance disorders
misophonia 37
  see also sound tolerance disorders
mixed hearing loss
  assessment 81
  audiovestibular implants 120
  differential diagnoses and treatment 84, 85, 89
  nasal crusting 242
  otosclerosis 148–49
  paediatrics 644
  pure tone audiometry 31
  stapes surgery 153
  temporal bone trauma 174
Möbius syndrome 701
modified barium swallow impairment profile (MBSImp) 361
Moffett's solution, and modification 275
mononucleosis 573–74
Moraxella catarrhalis
  otitis media
    acute 64, 651
    with effusion 681–82
  rhinosinusitis
    acute 267, 597
    chronic 598
Morquio syndrome 702
mosaic Down syndrome 692
motion sickness 660
motivational interviewing 501
motor neuron disease 556, 557
mouthwashes, throat pain caused by 372–73
mucocele 274
mucociliary flow 189
mucoepidermoid carcinoma 523–25, 524*t*, 524*f*, 530–31, 536, 641
mucopolysaccharidosis (MPS) 701–2, 702*f*
mucosal malignant melanoma 525–26
mucosectomy 380, 426
mucus retention cysts 640–41
Multidimensional Voice Programme (MDVP) 350
multidisciplinary team (MDT) 556–57
  airway obstruction 394–95
  audiovestibular implants 119
  cleft lip and/or palate 685, 686, 688, 689
  dizziness 661
  dysphagia 625, 629

dysphonia 349, 614, 619–20
epiphora 227, 234
glioma 611
good medical practice 5
head and neck cancer 344, 345–46, 345*b*, 346*b*, 346*t*
non-surgical treatment 495
rehabilitation 498, 499, 500, 504
hearing loss 648
hyperparathyroidism 474–75
hypopharyngeal cancer 452
imaging 527
juvenile nasopharyngeal angiofibroma 606
laryngeal cancer 413–14, 416
laryngeal clefts 672
microtia 689
neck lumps 380, 384
neck trauma 402
necrotizing otitis externa 179, 180
oral cancer 434, 437
oropharyngeal cancer 421
otitis media with effusion 688
paediatrics 564–66
rhinosinusitis, acute 273
salivary gland tumours 438
sinonasal tumour 294
sleep-related breathing disorders 390
thyroid cancer 445, 446, 447
tracheal stenosis 671
tracheostomy 543–44
see also colleagues, working with
multiple endocrine neoplasia (MEN) 470, 475
multiple sclerosis 50, 556
mumps 640
muscle tension dysphonia (MTD) 556, 557, 557*f*
myasthenia gravis 50, 556
mycobacterial abscess 467, 468
Mycobacterium spp.
avium intracellulare 634–35
bovis 634–35
scrofulaceum 634–35
tuberculosis 409
Mycoplasma pneumoniae 592
myoneurectomy 550, 553
myopia 661
myringitis 64–65, 65*f*
myringoplasty 16, 66, 160, 655–56
myringostapediopexy 157
myringotomy 16, 74, 651–52, 653, 655

Nager acrofacial dysostosis 688*t*
nasal acoustic analysis 241
nasal airway physiology 236–37, 237*f*
nasal cautery 200, 201
nasal crusting 183, 242
examination 245
history 242–45, 242*t*–43*t*, 244*f*–45*f*
investigations 245–46
tips and pearls 246
treatment 246
nasal cycle 237
nasal dermoids 636
nasal obstruction (NO) 183, 184, 241
aetiology and pathophysiology 237–38, 239*f*
assessment 238–40, 239*t*, 240*t*
controversies 241
definition 236
nasal crusting 242
paediatrics 608
assessment 609
controversies and future 612
differential diagnoses 609–10
syndromes 691
treatment 610–12
prevalence 236
treatment 240–41
Nasal Obstruction Symptom Evaluation (NOSE) 189, 238
nasal packing 200, 200*b*, 201
nasal polyps
chronic rhinosinusitis with (CRSwNP) 274, 276, 278–79
endoscopic sinus surgery 274, 276, 277, 278–79
facial pain 224
nasal crusting 245
nasal polyp score 189
nasal septum
anatomy and physiology 608
deformities 185
deviated *see* deviated nasal septum
examination 185, 186*f*
perforation 204, 207
aetiology 204–5, 204*b*
avoidance 206–7
conservative management 205
investigations 205
malignancy 204, 205
nasal crusting 243
obturators 205
surgical treatment 205–6, 206*f*–7*f*
symptoms 205*b*, 205
nasendoscopy 185–86, 187*f*
cleft lip and/or palate 689
COVID-19 189
deep neck space infections 467
dysphagia 360, 360*f*, 363, 364, 365–66
dysphonia 614, 616
epistaxis 604
laryngeal movement disorders 558
Lund–Kennedy Endoscopic Score 189
nasal blockage 608, 609, 610
neck lumps 382, 382*f*
obstructive sleep apnoea 388, 584–85
rhinosinusitis, chronic 597
nasolabial flap 482
nasolaryngoscopy 380–81, 564
nasopharyngeal airway (NPA) 586, 587
nasopharyngeal cancer (NPC) 456, 462, 523, 525
histological classification 456
initial assessment and investigations 457–58
management 458–62
non-surgical treatment 495
pathology and risk factors 456
prognosis 458
research questions 462
spread, pattern of 456–57
staging 458
symptoms 457
nasopharyngeal stenosis 679
nasopharynx
anatomy 368, 369*t*
assessment 370
dysphagia, paediatric 625–26, 627
nasoseptal flap, pedicled 337
National Tracheostomy Safety Project 395, 541, 542*f*, 543, 544*f*, 544–45
nebulizers, and COVID-19 20
neck
anatomy 527, 528*f*, 528*t*, 529*f*
dissection 495
hypopharyngeal cancer 453
oral cancer 434, 436
oropharyngeal cancer 421, 424, 425, 426
salivary gland tumours 440, 441–42
thyroid cancer 446, 447, 448
lumps 375, 384, 508
blood/pathology tests 382–84, 383*t*
clinical examination 377–82, 377*f*, 378*f*, 379*f*, 381*t*, 382*f*
cystic 537, 538*f*, 539*b*
general appearance and first impressions 375
history taking 375–77, 376*b*
nasopharyngeal cancer 457–58
presentations 376*t*
see also branchial cleft: cyst; dermoid cyst; paediatrics: neck masses, cysts, and sinuses; paraganglioma: neck; schwannoma: neck; thyroglossal duct cyst
trauma 397, 397*f*
airway management 398–99
causes of death 397*b*
clinical assessment 397–98, 398*f*
controversies and future 401–2
initial approach and stabilization 398
management principles 399*t*, 399–401, 399*f*, 400*t*, 400*f*, 402*f*
tips and pearls 402
see also deep neck space infections; head and neck pathology; head and neck reconstruction
necrotizing otitis externa (NOE) 63, 176, 176*f*, 180, 525
anatomy 176, 177*f*
diagnosis 176–77, 177*t*
differential diagnosis 177–78
histology 177
management 179–80, 179*t*
microbiology 177, 177*t*
monitoring and outcomes 180
radiology 178*f*, 178–79
needle cricothyroidotomy 393
neglect/maltreatment of child 563
Neisseria spp.
gonorrhoeae 592
meningitidis 592
neonates
airway abnormality 666, 667, 669, 670
airway compromise, acute 570, 571–73, 578
auditory brainstem responses 34–35
cleft lip and/or palate 687–88
dysphagia 625–26
epiphora 231*t*
external ear malformations 97
hearing screening 123, 562, 643, 644–45, 646
laryngeal anatomy 613
nasal anatomy and physiology 608
nasal blockage 608, 609, 610, 611
neck masses, cysts, and sinuses 635
neurological examination 662
obstructive sleep apnoea 586
otoacoustic emissions 34
rhinitis 610, 626
sensorineural hearing loss 78
suppurative parotitis 640
swallowing process 624
see also paediatrics
neoplasia 245, 392, 654–55
neurilemmoma/neurinoma *see* schwannoma
neuroblastoma, olfactory 291, 292, 293, 295, 523, 530*f*
neuroendocrine carcinoma 292, 522
neurofibromatosis (NF)
type 1 697
type 2 646, 697
neuroma *see* schwannoma
neurosyphilis 57
neurotological examination 48–49, 49*b*
nocturnal enuresis 582–83
Noise at Work Regulations 1989 126
noise-induced hearing loss (NIHL) 125
definitions 125
diagnosis 126–27, 127*f*, 128*f*
history 125
occupational 125, 126
quantification 128–29
tinnitus 129
non-allergic rhinitis *see* rhinitis: non-allergic
non-Hodgkin lymphoma 525
non-organic hearing loss 35
non-tuberculous mycobacterial infection 634–35, 636, 640
Noonan syndrome 644
nose
airway *see* nasal airway physiology
allergy *see* allergic rhinitis
anatomy and physiology 189, 608–9
blocked *see* nasal obstruction
cleft lip and/or palate 685–86
clinical assessment 183
clinical scores 189
controversies and uncertainties 189
examination 184–85
history 183–84
interventions 185–87, 186*f*, 187*f*
investigations 187–89
'red flags' 183*t*
closure 322, 322*f*
crooked 259–62, 263*f*
crusting *see* nasal crusting
dysphagia, paediatric 625–26, 627
examination 192
fracture 403
functions 236

nose (*cont.*)
midline nasal dermoids 631
obstruction *see* nasal obstruction
saddle 262, 265*f*
nuclear medicine 527, 530
see also positron emission tomography; radioiodine; technetium-99m sestamibi (TC99m MIBI) parathyroid scintigraphy
NUT carcinoma, sinonasal 292
nystagmus
benign paroxysmal positional vertigo 112–15, 113*f*, 117
dizziness, paediatric 662
Ménière's disease 109
vertigo
acute 55
investigations 53–54
outpatient examination 48–49, 50, 51–52, 53

obesity
dizziness 56
head and neck reconstruction 488
paediatrics 563
paediatrics 580, 583, 585
tracheostomy 541
obstructive apnoea–hypopnoea index (oAHI) 582
obstructive sleep apnoea (OSA) 386, 390, 580, 587
anatomy 580
assessment 583
cleft lip and/or palate 688, 690
clinical assessment and suitability for surgical intervention 386
controversies 586–87
definition 582
differential diagnoses 585
dizziness 57
Down syndrome 692
guidelines 587
investigations 387, 583–85
physiology 582
septal deviation 611–12
terminology 580
tonsillectomy 677–78
treatment 387–90, 388*b*, 585–86
obstructive sleep-disordered breathing (oSDB)
anatomy 580, 581–82
assessment 582–83
investigations 583, 585
physiology 580
terminology 580
treatment 585–86
see also obstructive sleep apnoea
occlusion imaging 517
occupational factors
cough, chronic 216–17
hypopharyngeal cancer 451
laryngeal cancer 411
nasal crusting 242*t*, 244
neck lumps 376–77
non-allergic rhinitis 217
nose and paranasal sinuses, clinical assessment 184
postnasal drip 216
throat pain 373
occupational noise-induced hearing loss 125, 126
ocular torsion reaction test 50–51
oculo-auriculo-vertebral spectrum *see* Goldenhar syndrome
odontogenic sinusitis 269, 270*f*
odynophagia 356, 371, 451
oesophageal atresia (OA) 669
oesophageal stenoses, upper 451
oesophagitis 359
oesophagogastroduodenoscopy 361
oesophagoscopy 344, 345, 366, 382, 451, 615
oesophagus
dysphagia 628
perforation, tracheostomy complication 540
Ohm's law 238*b*
olfactory bulbs (OBs) 191, 195
olfactory disorders *see* smell, sense of: disorders
olfactory groove meningioma 332
olfactory neuroblastoma 291, 292, 293, 295, 523, 530*f*
olfactory sensory neurons (OSNs) 191, 194
oligometastatic disease 496
olivopontocerebellar atrophy 50
Ondine's curse 585
one-stop neck lump clinic 344–45
Onodi cell 294–95
operating microscope 161, 162*f*
Optiz–Frias syndrome 668–69
Opitz G syndrome 628
Opitz syndrome 688*t*
oral cancer 428, 432, 437
assessment 432, 433*t*, 433*f*
investigations 432–34, 434*t*
management 434–37, 435*f*, 436*f*
potentially malignant disorders (PMDs) 428–32, 429*f*, 430*t*
staging 434
symptomatology 432
oral cavity
anatomy 428
disease 379
dysphagia 626–27
hearing loss, paediatric 644
imaging 531, 533*f*
sore throat 593
oral ulceration 406
causes 406, 407*t*, 409–10
clinical examination 406–9
diagnostic algorithm 406, 408*f*
history 406
malignancy 406, 409
orbital cellulitis 272, 274
orbital complications, acute bacterial rhinosinusitis 272
orbital floor fracture 403
orchitis 640
organ of Corti 29
organs at risk (OARs), head and neck cancer radiotherapy 492
oropharyngeal cancer 419
assessment and investigations 420–21
de-escalation controversy 426
management 421–26, 422*f*–23*f*, 423*t*, 424*b*, 424*f*–25*f*, 425*b*
non-surgical treatment 494–95
prognosis 426
risk factors and HPV role 419
staging 421
symptoms 371, 419–20
unknown primary 426
oropharyngeal dysphagia 361–66, 363*f*–65*f*
oropharynx
anatomy 368, 369*t*, 419, 420*f*
dysphagia 626–27
imaging 531–32, 533*f*
squamous cell carcinoma 522–23, 523*f*, 525
orthopantomography 379, 403, 421, 466, 468, 527
orthostatic hypotension 56, 660
oscillopsia (Tullio phenomenon) 46, 48, 53, 81–84, 171, 660

Osler–Weber–Rendu syndrome *see* hereditary haemorrhagic telangiectasia
osseous labyrinth 28
ossicular chain
abnormalities *see* ossiculoplasty/ossicular chain abnormalities
anatomy and physiology 27–28
disconnection 82*t*, 85, 86*f*
temporal bone trauma 168
see also incus; malleus; stapes
ossiculoplasty/ossicular chain abnormalities 154, 159
acquired 154
aetiology 154
assessment 154–56, 155*f*, 155*t*
Austin classification 155, 155*t*
cholesteatoma 146*t*, 146
congenital 89, 154
definition 154
future research 159
guidelines, lack of 159
surgical management 156–58, 156*f*–58*f*, 157*t*
postoperative complications 158–59
temporal bone trauma 174
treatments 156
osteitis 273, 598
osteogenesis imperfecta (OI) 702, 703*f*
conductive hearing loss 89
differential diagnosis 149
surgery 151
osteomas 94*f*, 94, 95*f*–96*f*
osteomeningeal breach 169
osteomyelitis
acute bacterial rhinosinusitis complication 273
chronic rhinosinusitis complication 598
mycobacterial 465
necrotizing otitis externa 177
skull base 74, 138
osteophytes 363, 363*f*
osteoradionecrosis (ORN) 421, 434, 491
otalgia 59, 66, 650
anatomy 59, 60*f*
assessment 59–61, 650
barotrauma 66, 66*t*
causes 59*t*
cholesteatoma 147, 656–58
clinical examination 61
controversies 67
dental disease 67
differential diagnosis 650
external ear pathology 61–64
foreign body 654
herpes zoster oticus 66
investigation 61
malignancy 67
necrotizing otitis externa 176, 177
neoplastic disease 654–55
neuralgia 67
otitis externa 653–54
otitis media
acute 650–53
chronic 655–56
referred 60*f*, 66–67, 368
temporomandibular joint dysfunction 67
tips and pearls 67–68
tympanic membrane and middle ear pathology 64–65
otic barotrauma 66, 66*t*
otitic hydrocephalus 653
otitis externa (OE) 59, 61, 653–54
acute 61–62, 62*f*
chronic 63
fungal (otomycosis) 62, 63*f*
necrotizing/malignant *see* necrotizing otitis externa
topical treatment 653
otitis media
acute (AOM) 59, 64, 650
aetiology 651
complications 652–53, 654*f*
facial paralysis secondary to 74
investigations 650
management and guidelines 651
presentation and diagnosis 650–51
prevention 651
recurrent (rAOM) 64–65, 650, 651–52
chronic 65, 154, 655–56, 655*f*, 692
classifications 650
with effusion (OME) 652, 662, 681, 684
adenoidectomy 683
aetiology 681–82
autoinflation 683
cleft lip and/or palate 688, 690
diagnosis 682
differential diagnosis and treatment 82*t*, 85–86
dizziness 660
Down syndrome 692
epidemiology 681
hearing aids 683
hearing loss 89, 643, 644
management 682
natural history 681
obstructive sleep apnoea 583
shared decision-making 682
treatments not recommended 684
Turner syndrome 694
ventilation tubes 683
watchful waiting 682–83
suppurative 48
otoacoustic emissions (OAEs) 34, 34*f*, 644, 645–46
distortion product (DPOAEs) 645–46
functional hearing loss 35

sensorineural hearing loss 78
tinnitus 37, 40
transient evoked (TEOAEs) 645
otolithic membrane 30*f*, 30
otomycosis 62, 63*f*
oto-palato-digital syndrome 646
otoplasty 689–90
otorrhoea 59, 650
anatomy 59
assessment 59–60, 61, 650
causes 59*t*
cerebrospinal fluid 66
cholesteatoma 103, 141, 142, 144*t*, 145, 147, 656–58
clinical examination 61
controversies 67
differential diagnosis 650
external ear pathology 61–64
foreign body 654
investigation 61
necrotizing otitis externa 176
neoplastic disease 654–55
otitis externa 653–54
otitis media
acute 650–53
chronic 655–56
tips and pearls 67–68
tympanic membrane and middle ear pathology 64–66
ventilation tube complication 683
otosclerosis 83*f*, 148, 153
aetiology 148
assessment 148
conductive hearing loss 81
controversies 153
definition 148
differential diagnosis and treatment 82*t*, 84–85, 85*f*, 149–50
far-advanced 150
genetics 148
guidelines, lack of 153
hearing loss 89, 148–49, 153
incidence 148
investigations 84, 148–49, 149*f*, 150*f*
natural history 148
surgical management 150–52, 151*f*, 152*f*
tinnitus 38, 40, 148, 150
unilateral 153
otoscopy 154, 160, 650–51, 682
ototoxic drugs 647
outer ear *see* external window
oval window 27–28, 29, 53, 89
oximetry 583–84, 587, 666
oxygen, hyperbaric 179–80
ozaena (primary atrophic rhinitis) 244

pachydermia 353
paediatrics
adenoidal hyperplasia 314, 317
allergic rhinitis 314, 316, 317
assessment 569
airway 566–67
clinic environment 561
complex children 564
decision-making and consent 564–65, 566
examination 563–64
history taking 562, 563, 569
observation 563
perioperative support 568
play therapy 568–69
pre-hospital programmes 568
preoperative 568
sepsis 567, 568
seriously ill 'sick' child 565–66
shock 567
tips and pearls 561
aural fullness 69
deep space neck infections 465, 466, 468
ear anatomy and embryology 643
epiphora 231*t*
epistaxis 198, 199
external ear
congenital atresia 99
exostosis 98
foreign body 92, 93
malformations 98
facial paralysis secondary to acute otitis media 74
facial trauma 403
Kallman syndrome 195
maltreatment 563
nasal septal perforation avoidance 207
neck masses, cysts, and sinuses 630
acquired lumps with fever and malaise 634
acquired lumps without fever 634–35
acquired sinuses 633–34
anatomy and embryology 630
assessment 635
branchial cleft cyst 508, 513
congenital lumps 630–33
congenital sinuses 633
controversies 636–37
guidelines 637
investigations 635–36
thyroglossal duct cyst 513
olfactory neuroblastoma 292
oral ulceration 406, 409–10
otalgia and otorrhoea 59, 64, 65
otitis media
acute 64, 65
with effusion 89
otosclerosis 153
peak nasal inspiratory flow 240*t*
phonosurgery 553
pure tone audiometry 30
rhinorrhoea 210
rhinosinusitis
acute 270, 272, 273
chronic 317
sensorineural hearing loss 78
syndromes *see* achondroplasia; Alport syndrome; Beckwith–Wiedemann syndrome; branchio-oto-renal syndrome; CHARGE syndrome; 644; cri-du-chat syndrome; Down syndrome; Goldenhar syndrome; Jervell and Lange-Nielsen syndrome; Möbius syndrome; mucopolysaccharidosis; neurofibromatosis; osteogenesis imperfecta; Pendred syndrome; Prader–Willi syndrome; Stickler syndrome; Treacher Collins syndrome; Turner syndrome; Usher syndrome; velocardiofacial syndrome; Waardenburg syndrome
throat pain 371, 372, 580–87
tinnitus 37–38
tympanometry 31–32
see also adolescents; airway compromise, acute; airway obstruction: paediatric; anaesthesia: paediatrics; infants; neonates; toddlers
Paediatric Septic Six 567, 568
Paediatric Sleep Questionnaire (PSQ) 583, 585
Paediatric Voice Handicap Index (pVHI) 614, 615, 620
Paediatric Voice Outcome Survey (PVOS) 614
Paediatric Voice Related Quality of Life (PVRQOL) 614
Paget disease 81–84
Pagnini–McClure manoeuvre 171
palatal myoclonus 41
palate
cleft *see* 629
reduction, anterior 389
palatopharyngoplasty 389
palatoplasty 689
palliative care 496
hypopharyngeal cancer 452, 453–54
laryngeal cancer 416–17
nasopharyngeal cancer 156
oral cancer 434
oropharyngeal cancer 425–26
thyroid cancer 447–48
Pallister–Hall syndrome 668–69
panda eyes 171*b*
panendoscopy, rigid 366
papillary carcinoma 515
papillary cystadenoma lymphomatosum *see* Warthin tumour
papillary thyroid cancer 537*f*, 537
papilloma 291, 292, 352–53, 353*f*, 523
papillomatosis 552, 613, 618–19, 620–21
paradoxical vocal fold dysfunction (exercise-induced laryngeal obstruction) 555, 619
paradoxical vocal fold motion *see* inducible laryngeal obstruction
paraganglioma
anatomy and embryological considerations 516
assessment 516
controversies and future 517
differential diagnosis 82*t*, 516
facial paralysis 74
investigations 516–17, 516*f*
jugular foramen 136
magnetic resonance imaging 138*t*
neck 379–80
surgical tips 517
treatment 517, 517*f*
paranasal sinuses
anatomy and physiology 282–83, 282*f*, 283*f*–84*f*
paediatrics 596, 608
clinical assessment 183
clinical scores 189
controversies and uncertainties 189
examination 184–85
history 183–84
interventions 185–87, 186*f*, 187*f*
investigations 187–89
'red flags' 183*t*
endoscopic surgery *see* endoscopy: sinus surgery
paraneoplastic syndrome 57
parapharyngeal abscess 465, 466, 468
paediatrics 634, 635
throat pain 371, 371*f*
parapharyngeal tumour 364*f*, 364
parathyroidectomy 473–74, 475
minimally invasive (MIP) 471, 473, 474
minimally invasive video-assisted (MIVAP) 473
remote access 473–74
parathyroid glands
anatomical locations 471
disorders 470, 475
cancer 470
see also hyperparathyroidism; hypoparathyroidism
Parkinson's disease 50, 52, 195, 556, 557
parosmia 192*t*, 194, 195–96
parotidectomy 641–42
parotid gland 380
anatomy and physiology 638
cyst 382–84
drooling management 639
haemangioma 641, 641*f*
juvenile Sjögren's syndrome 641
lymphoma 641
tumour 377, 438, 438*t*, 439
facial paralysis 74
imaging 535, 535*f*
surgery 440–42, 440*f*, 441*t*, 441*f*
parotitis
juvenile recurrent 640, 642
suppurative 640
paroxysmal hemicrania 222*t*, 223
partial ossicular replacement prostheses (PORPs) 157
passive smoking
allergic rhinitis 602
otitis media
acute 651
with effusion 682
rhinosinusitis, chronic 598
Paterson–Brown Kelly (Plummer–Vinson) syndrome 364–65, 451
pathology report interpretation 521
pathology request form 520
patients, working with 5
patients' rights 3
Deprivation of Liberty Safeguards 9–10
ethical approaches to medicine 10
Mental Capacity Act 7, 7*b*
mental capacity assessment 7–9, 8*b*
safeguarding adults 10

peak nasal inspiratory flow 189, 238, 240, 240*t*
pectoralis major myocutaneous (PMMC) flap 485, 486, 488, 490
pemphigoid, mucous membrane 410
pemphigus vulgaris 410
Pendred syndrome 646, 660–61, 695–96, 696*f*
Penetration–Aspiration Scale (PAS) 361
perichondritis 63–64
  relapsing 64, 64*f*
perilymphatic fistula 58, 660, 661
periodic fever, aphthous stomatitis, pharyngitis, and adenitis (PFAPA) syndrome 594, 634, 636
peripheral vestibular dysfunction 46–48, 49, 50, 52, 55
peritonsillar abscess 370–71, 384, 575
persistent idiopathic facial pain (PIFP) 222*t*, 224, 224*b*
personal protective equipment (PPE)
  Covid-19 20, 21–22, 21*t*, 545
  epistaxis 199
petrosectomy 74
petrous apex lesions 137–38, 138*t*
petrous apicitis 139
PFAPA syndrome 594, 634, 636
Pfeiffer syndrome 699
PHACE syndrome 631
phantosmia 184, 192*t*, 195–96
  post-traumatic 194
pharyngeal cancer 411
  see also oropharyngeal cancer
pharyngeal lymphatic ring *see* Waldeyer's ring
pharyngeal obstruction 691
pharyngeal pouches 451, 630, 633, 643
pharyngeal stenoses 451
pharyngectomy 453
pharyngitis 384, 592
pharyngolaryngectomy 453–54
pharyngo-laryngo-oesophagectomy 453
pharyngo-oesophageal injury 401, 402*f*
pharyngoplasty 689
pharyngotomy 388–89, 453
pharynx
  anatomy 368, 369*f*–70*f*
    contents and boundaries 369*t*
    fascial layers 368*t*
  imaging 531–35, 533*f*–34*f*
  reconstruction 488, 489
phonation 349
  see also dysphonia
phonophobia 37
  see also sound tolerance disorders
phonosurgery 547
  principles 549–53, 549*f*, 550*f*, 551*f*–52*f*, 553*b*
  vocal fold lesions 547, 548*f*
  vocal fold paralysis 547–49
photodynamic therapy 432
photophobia 46
physical fitness, and anaesthesia 13
physical illness in doctors 6
Pierre Robin sequence (PRS) 627, 687–88, 690, 703
pinna (auricle)
  anatomy and physiology 27, 28*f*
  cellulitis 63–64
  embryology 630, 643
  malformations 97–102, 99*f*–101*f*
  otoplasty 689
  see also external ear
piriform aperture stenosis 609, 611
pituitary gland lesions 326, 328–29, 328*f*
platysmal banding 310
play audiometry 646
play therapy 568–69
pleomorphic adenoma 438, 441, 442, 524*t*, 525*f*, 641
  imaging 535–36, 535*f*
Plummer–Vinson (Paterson–Brown Kelly) syndrome 364–65, 451
pneumatic otoscopy 682
pneumocephalus 339
pneumococcal vaccination 175
pneumomediastinum 541
pneumothorax 541
Pocket SIT 192
Poiseuille's law 238*b*
polyarteritis nodosa 56
polychondritis, relapsing 245
polypharmacy side effects 48, 57
polyps *see* nasal polyps; vocal cords/folds: polyps
polysomnography (PSG)
  airway abnormality 666
  obstructive sleep apnoea 584, 585, 586–87
    cleft lip and/or palate 688
port wine stains 632
position testing 51–52, 51*f*
Positive Outcome and Experience Management Strategies (POEMS) 568
positron emission tomography (PET) 527, 530
  and computed tomography (PET/CT)
    fluorodeoxyglucose *see* fluorodeoxyglucose positron emission tomography-computed tomography
  hypopharyngeal cancer 532–35
  laryngeal cancer 534*f*
  nasopharyngeal cancer 534*f*
  nose and paranasal sinuses 187
  oral cancer 533*f*
  oropharyngeal squamous cell carcinoma 531–32
  salivary glands, major 535
  sinonasal tumour 291, 295
head and neck cancer 495
hyperparathyroidism 475
laryngeal cancer 416
nasopharyngeal cancer 458
neck lumps 380, 381
oropharyngeal cancer 419–20, 421
sinonasal tumour 291
post-concussive syndrome 661
post-cricoid web 364–65, 365*f*
post-headshake nystagmus test 51
postnasal drip (PND) 183, 215
  anatomy 215
  assessment 216
  causes 216*b*
  controversies 218
  differential diagnosis 216–17
  guidelines 219
  investigations 216
  pathophysiology 215
  physiology 215, 216*f*
  rhinosinusitis, chronic 284
  tips and pearls 219
  treatments 217–18, 218*t*, 219*f*
post-traumatic olfactory dysfunction 194, 195
post-traumatic stress disorder 38
postural orthostatic tachycardia syndrome 56
posture tests 52
posturography, computerized dynamic 54, 54*f*
postviral olfactory dysfunction (PVOD) 194, 195
Pott's puffy tumour (subperiosteal abscess) 272, 273, 598, 599, 652, 654*f*
power of attorney 8–9
Prader–Willi syndrome 702
preauricular sinuses 633, 636, 644, 644*f*
pregnancy 56, 223, 324
prematurity 583, 620, 628, 665, 670
premedication 15
presbycusis 78–79
presbyphonia/presbylaryngis 354
pressure sores 10
presyncope 56, 659, 660
primary ciliary dyskinesia (PCD) 288
probity 5–6
professional voice users 619–20
progressive supranuclear palsy 50
prostheses
  anterior skull base reconstruction 335–37, 337*t*
  auricular 99, 99*f*
  obstructive sleep apnoea 387
  ossiculoplasty 156–57, 157*f*, 157*t*, 158*f*, 158, 159
    complications 159
  voice 504
Proteus mirabilis 142
proton beam therapy 458, 496
pseudohypoacusis 35
Pseudomonas aeruginosa
  cholesteatoma 142
  otitis externa 61, 653
    necrotizing 176, 177, 179, 525
  perichondritis 63–64
  petrous apicitis 139
  pinna cellulitis 63–64
  rhinosinusitis, chronic 598
  skull base osteomyelitis 138
psychogenic voice disorders 557, 558
psychological factors
  dysphagia, paediatric 628–29
  inducible laryngeal obstruction 555
  tinnitus 42
  see also anxiety; stress
puberphonia 354, 553, 557, 619
pulmonary oedema 678
punctoplasty 233, 234*f*
pure tone audiometry 30–31, 31*f*, 32*f*
  acoustic trauma, acute 129
  cholesteatoma 143
  conductive hearing loss 84
  dizziness, paediatric 662
  Ménière's disease 105, 109
  noise-induced hearing loss 127
  ossicular chain abnormalities 154
  otosclerosis 148–49
  sensorineural hearing loss 79
  tinnitus 40
  tympanic membrane perforation 81, 160
pyriform aperture stenosis 626

quinolone eardrops 67
quinsy 592, 594, 595, 634, 635, 636–37

raccoon eyes 171*b*
radial forearm fasciocutaneous flap 338
radiation exposure 376
radioallergosorbent test (RAST) 188, 314, 315, 316, 317, 318
radiofrequency devices, facial rejuvenation 312
radioiodine 446–47, 530, 537
radionuclide cisternography 212
radiotherapy (RT) 491
  acute toxicities and supportive care 495
  carbon ion 458
  concomitant chemotherapy 492–95
  external beam 434, 436–37, 460
  future developments 496
  head and neck cancer rehabilitation 500, 501, 504
  hypopharyngeal cancer 452, 453–54
  image-guided 492
  intensity-modulated *see* intensity-modulated radiotherapy
  juvenile nasal angiofibroma 612
  Langerhans histiocytosis 655
  laryngeal cancer 412–13, 414–17, 414*b*, 415*f*, 415*b*, 416*b*
  nasopharyngeal cancer 457–61
  oral cancer 434, 436–37
  oropharyngeal cancer 421–23, 422*f*–23*f*, 423*t*, 424*b*, 426, 522–23
  palliative radiotherapy 496
  paraganglioma 517
  practical considerations 491–92
  principles 491
  radical 492–94
  radical re-irradiation 496
  rhabdomyosarcoma 655
  salivary gland tumours 442
  schwannoma 515
  stereotactic ablative body 496
  thyroid cancer 447
  treatment assessment and post treatment surveillance 495–96
Ramsey Hunt syndrome 48, 66, 74
ranula 641
Rathke's cleft cyst 328
reactive cervical lymphadenopathy 634
reanimation surgery 75–76

recruitment 37
see also sound tolerance disorders
rectus abdominis myocutaneous flap 338
recurrent aphthous stomatitis (RAS) 406, 409
recurrent parotitis 640, 642
recurrent respiratory papillomatosis 618–19, 620–21
reflective practice 4
reflex decay testing 32–34
reflux
extra-oesophageal reflux disease 287
gastro-oesophageal *see* gastro-oesophageal reflux disease
laryngopharyngeal 217, 353, 593, 616
Reflux Symptom Index 217
regional anaesthesia 15
Reinke's oedema 350, 550
Reissner's membrane 28–29
religious factors 339
ReSPECT (Recommended Summary Plan for Emergency Care and Treatment) process 14
respiratory disturbance index (RDI) 582
respiratory effort-related arousal (RERA) 582
respiratory failure 57
retropharyngeal abscess 465, 466, 467, 468–69, 634
revalidation, professional 3, 5
reviews of practice 4–5
rhabdomyosarcoma 654
rheumatoid nodules 547, 550
rhinitis
allergic *see* allergic rhinitis
chronic 314, 315, 318
differential diagnosis 284
hearing loss 644
medicamentosa 217, 243, 246
nasal crusting 243, 244, 246
neonatal 610, 626
non-allergic (vasomotor) 209, 315, 318
differential diagnosis 217
with eosinophilia syndrome (NARES) 209
nasal obstruction 237, 238, 240
paediatrics 600
obstructive sleep apnoea 585
primary atrophic (ozaena) 244
secondary atrophic (empty nose syndrome) 237, 243, 246
toxic 195
Rhinobase 247–49
rhinology 16–17
rhinomanometry (RMM) 238–39, 239*t*
anterior 189
active (AAR) 239, 239*t*
rhinometry, acoustic 189, 238, 239–40
rhinoplasty 247
anaesthesia 249
cleft lip and/or palate 687
closure of incisions, packing, and splints 259
crooked nose 259–62, 263*f*
endoscopic 262
nasal fracture 403
osteotomies 254–56, 255*b*, 255*f*
postoperative follow-up 262–66
preoperative
preparation 247–49, 249*b*
photography 247, 249*b*
self-evaluation form 248*b*
preservation 254
profile alignment 249–54, 252*f*, 253*b*–54*b*
revision 262
saddle nose 262, 265*f*
surgical approaches and incisions 249, 249*b*
tip surgery 256–59, 256*f*, 258*b*, 258*f*, 260*f*
rhinorrhoea 183, 208, 214
allergic rhinitis 316
anatomy 596
bacterial 210
bilateral
infectious 210
non-infectious 208–10, 209*f*
controversies 213, 606
differential diagnosis 208*t*
guidelines 208, 209*f*, 213–14, 607
tips and pearls 607
unilateral 210–13, 212*f*–13*f*
vasomotor rhinitis 315
viral 210
rhinoscleroma 244, 245*f*
rhinoscopy, anterior 185, 245, 268
rhinosinusitis 596–98
acute (ARS) 267
aetiology 267
anterior skull base reconstruction 339
bacterial (ABRS) 210, 267, 268*t*, 268–69, 271–73
and chronic rhinosinusitis 317
classification 267
clinical features and treatment 224
diagnostic approach 267–69, 268*t*, 268*f*, 269*f*–70*f*
differential diagnosis 269, 270*f*
endoscopic sinus surgery 274
paediatrics 596–97
pathophysiology 267, 268*f*
prognosis 273
tips and pearls 273
treatment 269–71
anaesthesia 16
chronic (CRS) 183, 281, 597–98
and allergic rhinosinusitis 317
anatomy and physiology 282–83, 282*f*, 283*f*–84*f*
anterior skull base reconstruction 339
assessment 283–85, 285*f*
classification 189, 267, 285–86, 285*f*
clinical features and treatment 224, 224*b*
clinical scores 189
differential diagnoses and associated conditions 193–94, 287–88, 314
endoscopic sinus surgery 274, 276, 286, 288, 289*b*, 289
imaging 225
investigations 192–93, 220, 286–87, 287*f*
nasal crusting 242
nasal obstruction 237, 238, 240, 241
pathophysiology 281–82, 281*f*
postviral olfactory dysfunction 194
primary versus secondary 286
prognosis 282
smell, sense of 184
tips and pearls 289
treatment 282, 288–89, 289*b*
with nasal polyps (CRSwNP) 274, 276, 278–79
without nasal polyps (CRSsNP) 274, 276, 278–79
complications 598, 599–600
history 184
subacute 267
rhinospirometry 189
rights of the patient *see* patients' rights
Rinne test 48, 148
Romberg test 52, 662
rotary chair testing 54
round window 29, 53
Royal College of Anaesthetists 12, 13, 14
rubella 647

saccades 50, 53, 54, 55
saccule 29–30, 45, 46, 54–55, 112
safeguarding
adults 10
children 563, 565
salivary ducts 639
salivary fistula 436
salivary glands 638, 639–40
anatomy and physiology 638
assessment and history 638
benign inflammatory disease 382–84
bulk, reduction of 310
congenital disease 640
excision 639
haemangioma 641, 641*f*
imaging 535–36, 535*f*
immune conditions 641
infections 640
juvenile recurrent parotitis 640
mucus retention cysts 640–41
ranula 641
stones 640, 640*f*
submandibular 379
surgical techniques 641–42
tips and pearls 642
tumours 384, 438, 641
adjuvant treatment 442
diagnostic testing and workup 439
lymphoma 641
minor salivary glands 438, 438*t*
neck dissection consideration 441–42
neoplasms 523–25, 524*t*, 524*f*–25*f*
outcome 442
preoperative counselling 439, 439*t*
presentation 438, 438*t*
prognosis 442
surgical complications 442
surgical planning 439–41, 440*f*, 441*t*, 441*f*
Sanfilippo syndrome 701–2, 702*f*
sarcoidosis
differential diagnosis 288
nasal crusting 242, 243*t*, 244, 245, 246
nasal septal perforation 205
paediatrics 640
salivary gland enlargement 438
sarcoma 293
scala media 28–29
scala tympani 28–29
scala vestibuli 28–29
Schedule for Oral-Motor Assessment (SOMA) 625
Scheie syndrome 701
schwannoma
neck 379–80
anatomy and embryological considerations 515
assessment 515
differential diagnoses 515
investigations 515
treatment 515–16
facial nerve 137
facial paralysis 74, 75*f*
jugular foramen 136–37
vestibular *see* vestibular schwannoma
Schwartze's sign 148
scleroderma 359
Scottish Intercollegiate Guidelines Network (SIGN) 202*b*, 202, 594
screening programmes
neonatal hearing 123, 562, 643, 644–45, 646
oral cancer 432, 437
otitis media with effusion 682
scuba diving, after stapes surgery 153
self-care, good medical practice 6
semicircular canals (SCCs)
anatomy and physiology 28, 29, 30*f*
balance 45, 46
dehiscence (SCCD) 106
differential diagnosis and treatment 82*t*, 87, 88*f*
imaging 149, 149*f*
investigations 53, 84
vertigo 48, 52, 53, 54–55, 57–58
embryology 643
vertigo 48, 53
benign paroxysmal positional vertigo 51–52
head impulse test 50
Semont manoeuvre 114–15, 117
sensorineural hearing loss (SNHL) 77
aetiology 77, 77*t*, 121*b*
assessment 81
audiometry 78–79, 79*f*
audiovestibular implants 120
aural atresia 101–2
aural fullness 69
clarity or fidelity of hearing 79
dizziness, paediatric 660–61
external ear malformations 98

sensorineural hearing loss (SNHL) (*cont.*)
inner ear developmental abnormalities 77*b*
Ménière's disease 109
myringitis 65
nasal crusting 242
noise-induced hearing loss 126, 128–29
ossiculoplasty complication 159
otic barotrauma 66
otosclerosis 148, 150
paediatrics 643, 644, 648
acquired SNHL 647
congenital SNHL 646
syndromes 691
presentation 78
prevention 79–80
progression 79
pure tone audiometry 31
recruitment 37
stapes surgery complication 152
temporal bone trauma 174
tinnitus 38, 40
treatment and management 80
see also mixed hearing loss
sensory testing, vertigo 52
sentinel node biopsy 434
sepsis
deep neck space infections 466, 468
head and neck reconstruction 490
neck lumps 384
paediatrics 567, 568
throat pain 370, 594
septectomy 612
septicaemia 406
septodermoplasty 321–22, 324
septoplasty
endoscopic sinus surgery 276
nasal crusting after 243
nasal obstruction 240–41
paediatrics 608, 611–12
postnasal drip 218
septorhinoplasty 240, 241, 265
septum *see* nasal septum
seroma 436
severe acute respiratory syndrome (SARS) 19
severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) *see* COVID-19
sexual history 344, 346, 376
shingles 409–10
Shipman Inquiry 3
shock 567
hypovolaemic 567
Short Form Health Surveys
SF-6D 238
SF-36 238
sialadenitis, chronic sclerosing 379
sialadenosis 382–84
sialendoscopy 438, 640, 642
sialography 527
sialolithiasis 642
sialorrhoea *see* drooling
sigmoid sinus 169
significant incident reports 5
silastic septal button 204, 205
single-photon emission computed tomography (SPECT) 471–72, 475
sinonasal disease 530–31, 530*f*–31*f*, 532*t*
Sino-Nasal Outcome Tests 22 and 23 (SNOT-22 and SNOT-23) 189, 238, 274, 278
sinonasal tumours 292, 523
anatomy 290, 290*f*
assessment 290–91
controversies and future 295–96
differential diagnoses and associated conditions 291–93
investigations 291
staging 291*t*, 291
tips and pearls 295
treatment 293–95, 293*b*
sinonasal undifferentiated carcinoma (SNUC) 290–91, 292, 295
Sinus and Nasal Quality of Life Survey (SN-5) 597
sinuses, paranasal *see* paranasal sinuses
sinusitis 183, 220, 221*f*, 315
Sistrunk technique, thyroglossal duct cyst 514, 514*f*
Sjögren's syndrome 438, 525, 641
skew deviation
test 50–51
vertigo, acute 55
skin prick tests 188
rhinitis 209
allergic 314, 315–16, 316*f*, 317, 318, 602
skull base
endoscopic surgery 608
imaging 530–31, 530*f*–31*f*, 532*t*
disease *see* lateral skull base disease
osteomyelitis 74, 138
sleep apnoea 48, 386
obstructive *see* obstructive sleep apnoea
sleep disorders/impairment
breathing-related *see* sleep-related breathing disorders
insomnia 39, 40, 48
nasal obstruction 184
non-breathing-related 585
tinnitus 41
sleep-related breathing disorders (SRBDs) 386, 390
cleft lip and/or palate 688
clinical assessment and suitability for surgical intervention 386–87
investigations 387
treatment 387–90, 388*b*
see also obstructive sleep apnoea; obstructive sleep-disordered breathing; sleep apnoea; snoring
sleep studies 387, 583–84, 586
slide tracheoplasty 672
SMART acronym, good medical practice 4
smell, sense of 184
anatomy and physiology 191, 192*f*
disorders
assessment 192, 193*f*
controversies and future research 196
differential diagnoses 193–95, 193*t*, 194*f*
guidelines 196
investigations 192–93
rhinosinusitis, chronic 284
terminology 192*t*
tips and pearls 195–96
treatments 195, 196*f*
tests 187–88, 188*f*, 598
training 195
smoking
allergic rhinitis 602
cessation 13
dysphonia 351
head and neck cancer 344
laryngeal cancer 411, 522
laryngeal dysplasia 417
nasopharyngeal cancer 456
neck lumps 375, 376
oral cancer 428, 432
oropharyngeal cancer 419
rhinosinusitis, chronic 598
throat pain 371
vapes and e-cigarettes 373
see also passive smoking
smooth pursuit 49–50, 53
Sniffin' Sticks 188, 192, 598
snoring 386, 390, 580
assessment 582–83
terminology 580
treatment 585
social media 5–6
socioeconomic factors
hypopharyngeal cancer 451
laryngeal cancer 411
nasal crusting 244
oropharyngeal cancer 419
SOCRATES mnemonic (otalgia) 60–61
sound field audiometry 30
sound therapy 41
sound tolerance disorders 44
associations 38
defined 37
epidemiology 38
investigations 40
see also tinnitus
space-occupying lesions 661
spasmodic dysphonia *see* larynx: dystonia
specialized neonatal and paediatric surgeons (SNAPS) 669
speech
cleft lip and/or palate 689
delay 682
head and neck cancer rehabilitation 498–99, 498*t*, 500, 501, 504
hypernasal 678
motor neuron disease 556
multiple sclerosis 556
oesophageal 502*t*, 502, 504
speech and language therapists (SLTs)
cleft lip and/or palate 689
dysphonia, paediatric 613, 614, 619, 620, 621
dysphagia, paediatric 629
head and neck cancer rehabilitation 498, 499, 500–1, 504
laryngeal movement disorders 556–57, 558
tracheostomy patients 544
speech audiometry 84
sphenopalatine artery 198, 200, 201*f*, 201, 324
spinal accessory nerve *see* cranial nerves: XI
spinal pathology 363, 363*f*
spindle cell carcinoma, laryngeal 522, 522*f*
spinocerebellar ataxia 50
SPITE system (ossiculoplasty) 155
squamous cell carcinoma (SCC) 521
cystic neck masses 537, 538*f*
ear tumours 525
facial paralysis 74
head and neck reconstruction 479–81, 482, 485
hypopharyngeal 450
laryngeal 412*b*, 413*b*, 522, 522*f*, 534*f*
lymph nodes 537
nasopharyngeal 458, 523, 532, 534*f*
neck lumps 377, 380, 381, 384
oral (OSCC) 409, 410, 428, 431, 432, 531, 533*f*
investigations 432–33
management 434, 435*f*
potentially malignant disorders 428, 429–30
oropharyngeal (OPSCC) 419–26, 522–23, 523*f*, 531–32, 533*f*
salivary glands 438
sinonasal 290–91, 292, 523
thyroglossal duct cyst 515
vertigo 53
stapedectomy 40, 85, 150
stapedial artery, persistent 82*t*
stapedius muscle 27–28
stapedotomy
complications 152
controversies 153
direct acoustic cochlear stimulation 150
ossicular chain discontinuity 85
osteogenesis imperfecta 89
osteosclerosis 150–51, 151*f*
reverse 151
revision surgery 151, 152*f*
tympanosclerosis 85
vertigo 150, 152
stapes
abnormalities 149*f*, 149
anatomy and physiology 27–28, 29
embryology 643
fixation 149, 150, 151, 154
Staphylococcus spp.
aureus
cholesteatoma 142
furunculosis 62–63
methicillin-resistant (MRSA) 138
neonatal suppurative parotitis 640
otitis externa 61, 653
otitis media with effusion 681–82
petrous apicitis 139
rhinosinusitis, chronic 288, 598
epidermis 288
epiglottitis 592
tonsillitis 591
stents, neck trauma 401, 402

stepping test 52
stereotactic ablative body radiotherapy 496
stereotactic radiosurgery 292, 460, 515, 517
stertor 566, 580, 583
Stickler syndrome 644, 646, 687–88, 688*t*, 696–97
strawberry naevi 631
Streptococcus spp.
  anginosus 267, 271
  pneumoniae
    cerebrospinal fluid rhinorrhoea 211
    otitis media, acute 64, 651, 652
    otitis media with effusion 681–82
    prophylaxis 174
    rhinosinusitis, acute 267, 597
  pyogenes 652
  tonsillitis 591
stress 568
  dysphonia 613, 619
  post-traumatic stress disorder 38
  throat pain 373
  tinnitus 39, 41
  vestibular migraine 109
stridor 566, 570, 594
stroboscopy 614
stroke, dysphagia after 360, 363
Sturge–Weber syndrome 632
stylohyoid syndrome *see* Eagle syndrome
subglottic stenosis 667, 668*f*, 670–71, 671*f*
  acquired 573
  congenital 572
  Down syndrome 692
  timing of open surgery 673–74
sublingual glands 438, 438*t*, 638, 639
submandibular abscess 465, 466, 469
  see also Ludwig's angina
submandibular gland
  anatomy and physiology 638
  drooling management 639
  stones 640
  tumour 438, 438*t*, 439–40, 440*f*, 441–42
submental abscess 465, 466, 469
  see also Ludwig's angina
submental flap 482–85
submucous fibrosis 428, 431
subperiosteal abscess (Pott's puffy tumour) 272, 273, 598, 599, 652, 654*f*
suicidal ideation 41
sulcus vocalis 352*f*, 352
SUNA 222*t*, 223
SUNCT 222*t*, 223
Sunnybrook grading system 169, 170*t*
superficial musculoaponeurotic system (SMAS) 304–5, 305*f*, 306*f*, 306, 307
supine roll test 114
suppurative parotitis 640
supraclavicular flap 485
supraglottic cancer 371
supraglottitis 370, 371, 392
supraglottoplasty 575–76, 577, 628
Susac syndrome 56
swallowing
  airway abnormality 666
  anatomy and physiology 357–58, 358*f*, 368, 624–25
  FEES *see* flexible endoscopic evaluation of swallowing
  head and neck cancer
    characteristics dependent on site and relocation of resection 500*t*
    pre-habilitation 499
    rehabilitation 498–99, 498*t*, 501, 501*t*, 504
  neuromuscular conditions 555, 556, 557
  tracheostomy patients 544
  VFSS *see* videofluoroscopy swallow study
  visual assessment 360
  see also dysphagia
Swallowing Quality of Care (SWAL-CARE) questionnaire 360
Swallowing Quality of Life (SWAL-QOL) questionnaire 360, 365
Sydney Swallow Questionnaire (SSQ) 360
syncope 56
syphilis 48, 243*t*
systemic lupus erythematosus (SLE) 56, 243*t*, 244, 246, 359

tandem gait test 52
taste alterations
  Bell's palsy 73
  cochlear implants 122
  obstructive sleep apnoea complication 389–90
  and olfactory disorders 192, 193*f*
  ossiculoplasty complication 158–59
  otosclerosis 150
  and smell, sense of 184
  tympanoplasty complication 163
tearing, excessive *see* epiphora
technetium-99m sestamibi (TC99m MIBI) parathyroid scintigraphy 469, 471–72, 473, 530
tectorial membrane 29
telemedicine 22
temporal bone
  anatomy 131–32, 166, 166*f*
  squamous cell carcinoma 177
  trauma 166, 175
    clinical aspects 169–71, 170*t*, 172*t*
    controversies 174–75
    fracture 74, 89
    guidelines 175
    imaging findings 172*t*
    physiopathology and classification 167*t*, 167–69, 167*f*–68*f*
    tips and pearls 171*b*
    treatment 171–74, 172*f*, 173*f*
temporomandibular joint (TMJ) dysfunction 38, 67, 220, 224–25
tension-type headache 222*t*, 223, 223*b*
tensor tympani 27–28
third-window abnormalities 87, 88*f*
thoracic duct cyst 381
throat packs 17*b*
throat pain/soreness 368, 373
  acute 370–71, 371*f*
  anatomy and physiology 368, 368*t*, 369*t*, 369*f*, 370*f*, 588, 589–90
  assessment 368–73, 371*f*, 372*f*, 588–90
  chronic 372–73, 372*f*
  controversies and future 595
  differential diagnoses/associated conditions 590–94
  foreign bodies and trauma 372
  malignancy 369–70, 371–72
  management 373, 373*f*
  paediatrics 580–87
  tips and pearls 373, 595
  treatment 594
thyroglossal duct cyst 381–82, 381*t*
  anatomy and embryological considerations 513, 513*f*
  assessment 513, 514*f*
  carcinoma 515
  controversies and future 515
  differential diagnoses 513
  investigations 513–14
  paediatrics 631
  surgical tips 514–15
  treatment 514, 514*f*
thyroid cancer 444, 448
  anaplastic 444–45, 447–48
  assessment 445
  controversies 448
  differentiated 444–45, 446–47, 448
  follicular 444–45
  Hurtle cell 444–45
  imaging 537*f*, 537
  investigations 445, 446
  management 438
  medullary 444–45, 447
  neck lumps 382
  papillary 444–45, 446, 448
  pathology 444–45
  treatment 446–48
thyroidectomy 431, 446, 447, 515
thyroid function tests 513
thyroid gland
  abscess 466
  anatomy and physiology 444
  cartilage 348
  embryology 630
  goitre 365–66, 365*f*, 384
  imaging 536–37, 536*f*, 536*t*
  nodules 381*t*, 381–82, 384, 536–37, 536*f*, 536*t*
  tracheostomy 540, 541
Thyroid-Specific Patient-Reported Outcome (ThyPRO) 365
thyroplasty 552–53, 553*b*, 617, 621
tinnitus 44
  acoustic trauma, acute 129
  associations 38
  defined 37
  dizziness 48
  epidemiology 37–38
  examination 39–40
  guidelines 42, 43*b*
  heart synchronous 39–40
  heterogeneity 42
  history 39–40
  investigations 40
  Ménière's disease 110
  noise-induced hearing loss 129
  non-pulsatile 40
  non-rhythmical 37
  ossicular chain abnormalities 154
  otosclerosis 38, 40, 148, 150
  pathophysiology 38–39, 38*b*, 39*f*
  persistent spontaneous 41–42
  pulsatile 37, 37*b*, 40, 41, 88
  rhythmical 37, 41
  sensorineural hearing loss 78, 80
  somatosensory 37
  stapes surgery complication 152
  syndromic 38, 40
  tips and pearls 42
  treatments 40–42
Tinnitus Functional Index (TFI) 40
Tinnitus Handicap Inventory (THI) 40
Tinnitus Questionnaire (TQ) 40
tissue samples, head and neck pathology 520–21
TNM staging, sinonasal tumour 291
tobacco use
  hypopharyngeal cancer 450–51
  laryngeal cancer 411
  neck lumps 376
  oral cancer 428, 431, 432, 437
  oropharyngeal cancer 419
  squamous cell carcinoma 521
  see also smoking
toddlers
  airway obstruction, acute 572, 573
  obstructive sleep apnoea 586
  see also paediatrics
tongue base
  obstruction 691
  surgery
    mucosectomy 380, 426
    obstructive sleep apnoea 388–89, 388*b*
tonsillectomy 594, 595, 675, 679
  anaesthesia 14, 14*b*, 15, 677–78
  cleft lip and/or palate 688
  complications 678–79
  deep neck space infections 469
  Down syndrome 692
  dysphagia 627
  extracapsular (total) 675–76, 676*f*, 679
  glandular fever 591–92
  hot 675
  intracapsular (partial/tonsillotomy) 675, 676–77, 677*f*, 679
  intracapsular versus extracapsular debate 679
  neck lumps 380
  obstructive sleep apnoea 388
  obstructive sleep-disordered breathing 583, 585, 586, 587
  pain 678
  PFAPA syndrome 636
  quinsy 636–37
  technique 675–77, 675*f*–77*f*
  throat pain 370–71
  tonsillolith prevention 372
tonsillitis 370–71, 384, 588, 590–92, 594
  airway compromise, acute 573–74
  controversies 595
tonsilloliths 372

tonsils
anatomy and physiology 588, 589–90
size 580, 581, 583
dysphagia 627
obstructive sleep apnoea 580, 582, 584, 585
toothache 224
topical anaesthesia 16
total ossicular replacement prostheses (TORPs) 157
toxic rhinitis 195
Toxoplasma gondii 647
trachea
congenital stenosis 667, 668*f*
identification 391*f*
stenosis 671–72
stoma 414
transection 401
tracheobronchomalacia 573, 667, 669–70, 673
tracheocutaneous fistula, tracheostomy complication 543
tracheoinnominate fistula, tracheostomy complication 543
tracheomalacia 673
tracheo-oesophageal fistula (TOF) 543, 669, 669*f*, 691
tracheo-oesophageal puncture (TOP) 504
tracheopexy, posterior 673
tracheostomy 540, 545
acquired stenosis of the airway 667
airway compromise, acute 577–78
airway obstruction 393, 394, 395
anatomy 540
blocked tube 541, 544–45
complications 541–43, 542*f*
dysphonia 620
COVID-19 20–21, 540, 545
deep neck space infections 469
dysphagia 628
head and neck cancer rehabilitation 500
high versus low 541
hypopharyngeal cancer 450, 453
laryngeal cancer 412, 414, 416–17
neck trauma 398–99, 401
obstructive sleep apnoea 586
patient care 543–45, 544*f*
patient considerations 540
prophylactic 414
subglottic stenosis 670
technique 540–41
thyroid cancer 431
tracheobronchomalacia 673
transclival endoscopic surgery 329*f*, 329
transcranial magnetic stimulation 42
transcribriform endoscopic surgery 326, 328*f*
transfrontal pericranial flap 338
transient ischaemic attacks 46–48
translocation Down syndrome 692
transnasal humidified rapid insufflation ventilatory exchange (THRIVE) 549–50
transnasal oesophagoscopy (TNO) 361, 366
transodontoid endoscopic surgery 330–31, 330*f*
transoral laser microsurgery 424, 452
transoral robotic surgery (TORS)
hypopharyngeal cancer 452
nasopharyngeal cancer 460
obstructive sleep apnoea 388, 389, 390
oropharyngeal cancer 424, 425, 522–23
transorbital endoscopic surgery 329
transorbital neuroendoscopic surgery (TONES) 329
transplanum endoscopic surgery 327–28, 328*f*
transpterygoid endoscopic surgery 329–30
Carrau–Prevedello–Kassam classification 330*t*
transpterygoid temporoparietal fascial flap 338
transsphenoidal endoscopic surgery 326–29, 328*f*
trauma
airway obstruction 392, 571, 573–74
cerebrospinal fluid rhinorrhoea 211
deep neck space infections 466
dizziness 661
epiphora 231*t*
epistaxis 603
hearing loss 647
nasal crusting 243, 245
nasal obstruction 241
neuropathic facial pain after 225
olfactory disorders 194, 195
oral ulceration 406, 409
ossicular chain abnormalities 154
psychogenic voice disorders 558
throat pain 372
see also facial trauma; neck: trauma; temporal bone trauma
Treacher Collins syndrome (TCS) 630, 644, 646, 688*t*, 693, 694*f*
Treponema pallidum 409
trichophytic brow lift 298–99, 299*f*
trigeminal autonomic cephalgias 223
trigeminal nerve *see* cranial nerves: V
trigeminal neuralgia 220, 222*t*, 223–24
trigeminovascular reflex 106
trismus 595, 635
trisomy 13 688*t*
trisomy 21 646, 651, 655, 656, 688*t*, 692
see also Down syndrome
trust 4*t*, 5
tuba aperta *see* Eustachian tube: patulous
tubeless anaesthesia 17
tuberculosis (TB) 48, 243*t*, 244, 640
Tullio phenomenon *see* oscillopsia
tuning forks 48
turbinates 185, 186
turbinectomy, inferior 243
Turner syndrome 644, 694
tympanic cavity *see* middle ear
tympanic membrane (TM, ear drum)
anatomy and physiology 27–28, 28*f*, 160
conductive hearing loss 81, 84
eardrop usage in presence of open middle ear 67
hearing loss, paediatric 644
otalgia and otorrhoea 64–66
otitis media with effusion 682
otosclerosis 148
perforated 160, 655–56
anatomy 160
assessment/investigations 160
conductive hearing loss 81
emergency treatment 160–61
hearing loss, paediatric 644, 645*f*
retraction 655, 655*f*
tympanometry 31–34, 33*f*, 650
conductive hearing loss 84
dizziness 662
ossicular chain abnormalities 154
otalgia and otorrhoea 61
otitis media
acute 650–51
with effusion 682
otosclerosis 148–49
perforated tympanic membrane 160
recurrent acute otitis media 65
tinnitus 40
tympanoplasty 160
cholesteatoma 143*f*
consent 161
controversies 165
definition 154
general considerations 162
graft choice 162
indications 161
positioning 161
preparations 162
surgical options 161, 162*f*
tympanic membrane perforation 655–56
tympanic membrane retraction 655
tympanomeatal flap not raised 163*f*, 163
tympanomeatal flap raised 163–65, 164*f*–65*f*
tympanosclerosis
differential diagnosis and treatment 82*t*, 85, 85*f*
investigations 84
tympanostomy 651–52

ulceration, oral *see* oral ulceration
ultrasonic devices, facial rejuvenation 312
ultrasound (US)
airway obstruction 392, 572
branchial cleft cyst 511, 513
CHAOS 572
deep neck space infections 466, 467, 468
dysphagia 628
glandular fever 592
head and neck 520, 527, 529–30
hyperparathyroidism 471–72, 475
larynx 411, 614, 616
lymph nodes 537
neck lumps 379, 379*f*, 380, 381, 382, 384, 635, 636
oral cancer 533*f*
oropharyngeal cancer 419–21, 533*f*
salivary glands 438, 535–36, 535*f*, 640, 641
thyroglossal duct cyst 513
thyroid gland 537*f*
cancer 445, 447–48
nodules 536–37, 536*f*, 536*t*
unconscious patient 8
Union for International Cancer Control (UICC) 521
United States Food and Drug Administration (FDA) 313
University of Pennsylvania Smell Identification Test (UPSIT) 188*f*, 188
University of Pittsburgh Medical Center (UPMC)
angiofibroma staging system 291*t*, 291
juvenile nasopharyngeal angiofibroma staging 605–6
Unterberger's test 662
upper airway cough syndrome *see* postnasal drip
upper airway disorders 554–55, 554*f*
upper airway resistance syndrome 580
upper respiratory tract infection 15*b*, 210
UPSIT 192
Usher syndrome 646, 660–61, 695
utricle 29–30, 45, 46, 54–55, 112
uveoparotid fever 640
uvulopalatopharyngoplasty 388, 586

VACTERL association 668–69, 703
vagus nerve *see* cranial nerves: X
vallecular cyst 574
Van der Woude syndrome 688*t*
vapes and e-cigarettes 373
varicella zoster virus 409–10
VASCERN 324, 324*b*
vascular anomalies 88, 89*f*, 632–33
vasomotor rhinitis *see* rhinitis: non-allergic (vasomotor)
velocardiofacial syndrome (VCFS) 572, 617, 700
velopharyngeal insufficiency (VPI) 678–79, 686, 688, 689
veloplasty, intravelar 687
ventilation tubes (VTs, grommets)
Down syndrome 692
otitis media, acute 651–52
complications 652, 653
otitis media with effusion 681, 682, 683
cleft lip and/or palate 688
tympanic membrane retraction prophylaxis 655
ventricular cysts 550
vertigo 46, 47*f*, 58
acute 55
conductive hearing loss 81–84
defined 46
differential diagnosis 56–58, 106
Eustachian tube dysfunction 69
history 109*b*
investigations 53–55, 54*f*, 55*f*
Ménière's disease 105, 106, 110
myringitis 65

ossicular chain abnormalities 154
ossiculoplasty complication 158
otic barotrauma 66
outpatient examination 48–53, 49*f*, 49*b*, 51*f*
patient experience 110
post-traumatic 48
stapedotomy 150, 152
temporal bone trauma 174
tips and pearls 58
vestibular migraine 105, 110
see also benign paroxysmal positional vertigo; dizziness
vestibular aqueduct, enlarged 82*t*, 87, 88*f*
vestibular evoked myogenic potentials (VEMPs) 54–55
vestibular function tests 54, 662–63
vestibular labyrinth 46
vestibular migraine (VM) 48, 57, 105, 663
diagnosis 105–6, 105*b*
differential diagnosis 106, 114
examination 108, 109
future 110–11
guidelines 111
history 108
investigations 109
natural history and epidemiology 106, 108
pathophysiology 106
patient experience 110
treatments 109–10
vestibular neuritis 46–48
vestibular neuronitis 660, 663
vestibular paroxysmia 58
vestibular schwannoma 132–34, 133*f*, 135*f*
differential diagnosis 135
magnetic resonance imaging 138*t*
tinnitus 38, 40
vestibular system, anatomy and physiology 29–30, 29*f*–30*f*
vestibular testing 84
vestibule, anatomy and physiology 28
vestibulo-ocular reflex (VOR) 45, 46, 50
vestibulopathy (vestibular hypofunction) 57
vibroplasty 120
videofluoroscopic swallow study (VFSS)
dysphagia 361, 361*f*, 362, 363, 365
head and neck cancer rehabilitation 499
paediatrics 625, 627–28
video head impulse test (vHIT) 50, 54
video laryngoscopy 393
videonystagmography (VNG) 50, 53, 53*f*
video-oculography (VOG) 663
vidian neurectomy 218
virtual chromoendoscopy 344
visual acuity, dynamic 51
Visual Analogue Scale (VAS), nasal symptoms 189, 238
visual reinforcement audiometry (VRA) 646
vitamin deficiencies 57
vocal cords/folds
airway compromise, acute 571–72
anatomy and physiology 613–14
benign lesions 617–19
cold steel versus laser surgery 354
cyst 351, 351*f*
intracordal 547, 548*f*
paediatrics 618
dysfunction *see* inducible laryngeal obstruction
dysphonia 350, 352, 352*b*
muscle tension 557
paediatric 616–17, 620
granuloma 618
injection augmentation 552*f*, 552
laryngeal cancer 413*f*
laryngeal webs 617
lesions 547, 548*f*
leukoplakia 351*f*, 351
nodules
bamboo 351, 351*f*
dysphonia 352, 352*f*
paediatrics 617
phonosurgery 547, 550
surgical intervention timing 354
palsy
causes 350*b*
dysphagia 364, 364*f*, 628
dysphonia 350
paediatrics 616–17, 621, 622
paradoxical dysfunction (exercise-induced laryngeal obstruction) 555, 619
paradoxical movement *see* inducible laryngeal obstruction
paralysis 555–56
dysphonia 353–54, 353*f*
paediatrics 615–16
phonosurgery 547–49
paresis 615–16, 617
polyps 352, 352*b*, 618
phonosurgery 547, 548*f*, 552*f*
Reinke's oedema 550
scar 350
surgery, voice rest after 354
voice
functional disorders 557–58, 557*f*, 619–20
head and neck cancer rehabilitation 498–99, 498*t*, 499*t*, 500, 501, 504
inducible laryngeal obstruction 555
motor neuron disease 556
myasthenia gravis 556
Parkinson's disease 556
surgical voice restoration (SVR) 502*t*, 502–4, 503*f*
tremor 556
vocal fold paralysis 555–56
see also dysphonia; phonosurgery
Voice Handicap Index (VHI) 350
Paediatric 614, 615, 620
Voice Scoring System (VoiSS) 350
von Recklinghausen disease (neurofibromatosis type 1) 697
vulnerable patients, safeguarding 10

Waardenburg–Shah syndrome 694
Waardenburg syndrome 646, 660–61, 694, 694*f*
Waldeyer's ring 368, 588, 589
Warthin tumour 384, 524*t*, 525*f*
imaging 535, 535*f*, 536
Weber test 48, 84
Weerda diverticuloscope 452
Wegener's granulomatosis *see* granulomatosis with polyangiitis
wheezing 566
white eye blow out 403
wide vestibular aqueduct (WVA) 647
Williams–Beuren syndrome 38
wisdom teeth 466
World Health Organization (WHO) 5, 19

xenografts 335, 337*t*
X-rays
airway abnormality 667
airway obstruction 392, 571
dysphagia 363*f*, 365–66
head and neck 527, 635
cancer 492, 499
deep neck space infections 467
neck trauma 398, 403
nasal crusting 246
obstructive sleep apnoea 387, 584, 585
oral cancer 434
rhinosinusitis, acute 269
throat pain 372

Yacovino manoeuvre 116

Zenker diverticulum 359, 366
Zurich smell diskettes 188
zygomatic complex fracture 403