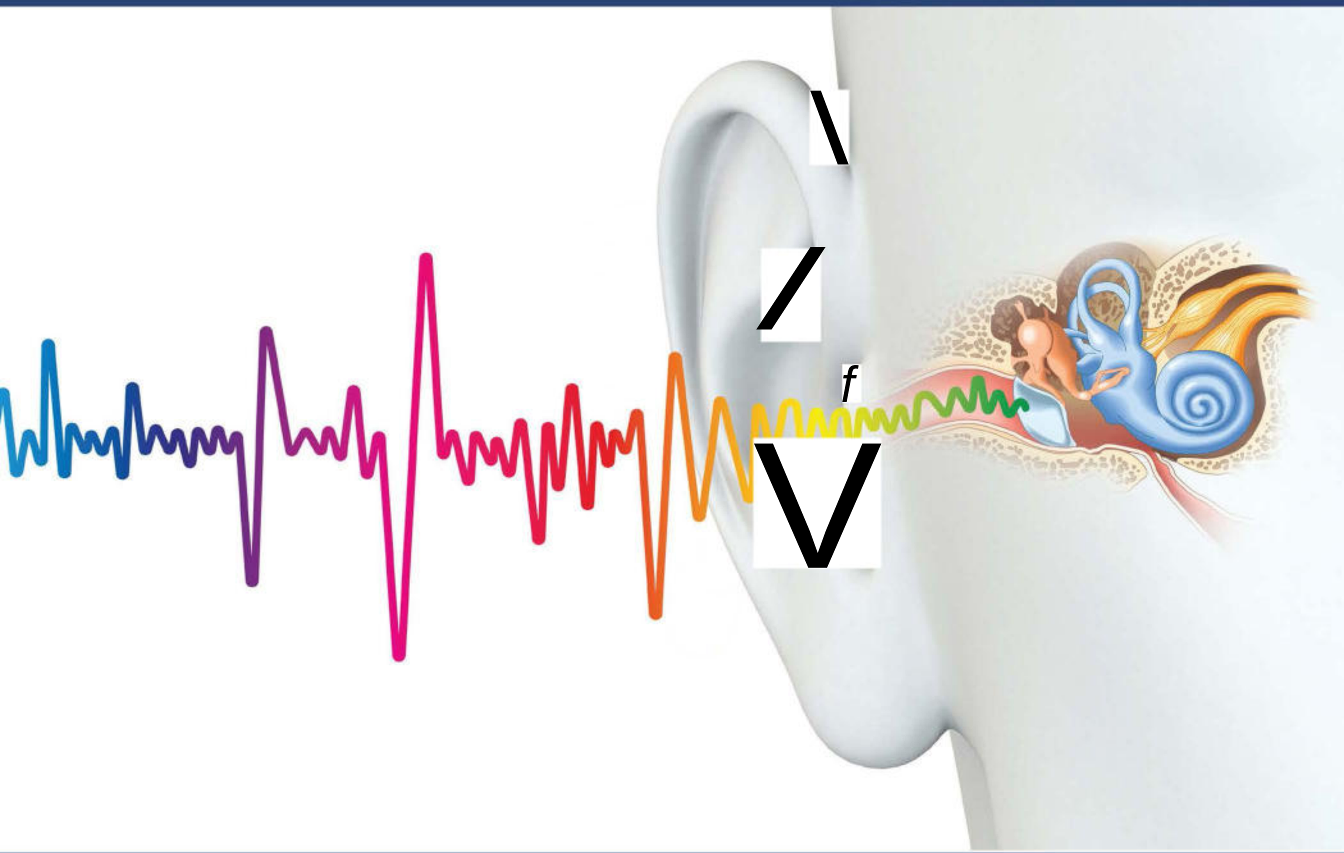
Browning's
Audiology for Clinicians

THIRD EDITION



George G Browning
Emma Stapleton
Haytham Kubba

Browning's Audiology for Clinicians

This book is a practical guide to interpreting audiometric tests, alongside the clinical assessment, to make sound evidence-based decisions in the clinical management of adults and children with ear conditions.

Audiology is central to clinical decision making, but it is not always taught well to Otorhinolaryngology surgeons, and often not taught at all. This third edition of a popular and classic text is an essential revision aid for Otorhinolaryngology trainees preparing for specialist examinations including the FRCS (ORL-HNS).

Fully updated and well-illustrated, it provides up-to-date content on the management of a wide range of ear conditions and will be of interest to the whole multidisciplinary team including audiologists, audiological scientists, audiological physicians, private hearing aid providers and speech and language therapists.

Browning's Audiology for Clinicians

Third Edition

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Contents

Preface		**vii**
Acknowledgements		**ix**
Authors		**xi**
1	Sound and human hearing	1
2	Clinical evaluation of ears and hearing	12
3	Pure tone audiometry	30
4	Conductive and mixed hearing loss	48
5	Behavioural hearing testing in children	62
6	Speech audiometry	68
7	Objective testing	74
8	Otitis media with effusion	86
9	Sensorineural hearing impairment in children	93
10	Sensorineural hearing impairment in adults	100
11	Non-surgical management of hearing impairment	114
12	Surgical management of hearing impairment	131
13	Cochlear implants	145
14	Outcomes, audit and consent	152
Appendix 1: UK population distribution of FFA by age and sex		**159**
Appendix 2: Example of patient information regarding hearing aids		**173**
Index		**179**

Preface

In the mid-1970s, ear surgery was evolving aided by the microscope and the drill. Audiometry was replacing tuning fork tests in hearing assessment. Mr Sandy Doig, an otologist at Glasgow Royal Infirmary, was a strong supporter of these developments and was instrumental in establishing the Scottish Section of the newly created MRC Institute of Hearing Research in Glasgow. Professor Stuart Gatehouse was appointed Scientist in Charge, with me, as an otologist. We helped design the UK National Study of Hearing, prospectively recorded our surgical outcomes and completed a number of randomised clinical trials. Mr Iain Swan later joined the team as a second otologist, helping expand the scope of clinical assessment and the development of outcome measures.

In the 1980s, we realised that there were many excellent textbooks covering ear diseases, but none were written as guides to practising clinicians, the majority of whom are generalists. This led to the first edition in 1986 of *Clinical Otology and Audiology*. This book was popular and was made available at the annual courses of the same title, and at the twice-yearly Glasgow Temporal Bone Dissection Course.

By 1998, copies of the first edition were difficult to find. Hence, an updated second edition was published whose scope was expanded to include hearing problems in infants and children. In addition, considerable advances had been made in the radiological diagnosis of vestibular schwannoma, which up till then had been a mostly audiometric pursuit. Cochlear implantation was being developed, both in children and adults, as were bone-anchored hearing aids.

At that time, the Scottish School of Audiology had moved to Glasgow Royal Infirmary under our direction. The role of the audiologist was changing from being one of diagnosis alone to a much wider role in clinical management including the social, psychological and symptom-based aspects of hearing loss and tinnitus.

Inevitably, in the early twenty-first century, the second edition also sold out and photocopies had to be provided at the courses which had continued in the UK, South Africa and elsewhere.

Since 1998, there have been considerable changes in the sound processing capabilities of digital hearing aids. Their provision commercially, outside the publicly funded health system, has expanded considerably especially to those with age-related hearing loss. There is now a more multi-disciplinary team approach to those with hearing problems. Hence, the potential readership of the third edition has expanded considerably from otolaryngology trainees to a much broader team including audiologists, audiological scientists, audiological physicians, private hearing aid providers and speech and language therapists. The third edition's title, *Browning's Audiology for Clinicians*, is more appropriate in 2025.

This is not, and was never intended to be, a comprehensive textbook of audiology or of otological surgery. Surgeons who want to know how to do a stapedectomy procedure should look elsewhere, as should audiologists interested in the prescription of hearing aids. This book is intended as a practical guide to interpreting audiometric tests, alongside the clinical assessment, in order to make sound, evidence-based decisions in the clinical management of patients.

George G Browning
Emeritus Professor

Acknowledgements

The three contributing authors to the Third Edition have been a well-coordinated team, supported by Miranda Bromage and Georgia Thompson from CRC Press/Taylor & Francis.

In addition, each author has been supported by advice and suggestions from their colleagues and I wish to personally thank:

- Professor Claude Laurent MD PhD on sections of Chapters 2 and 4
- Dr Jack Holman PhD on sections of Chapter 11
- Dr William H Whitmer PhD on sections of Chapter 11.

The first and second editions of 'Clinical Otology and Audiology', published in 1986 and 1998 respectively, were materially aided by the late Professor Stuart Gatehouse, the Audiological consultant in charge of the MRC CSO Scottish Institute of Hearing Research latterly at Glasgow Royal Infirmary until 2007. The main purpose of this Institute, whose headquarters were at Nottingham University, was to carry out a UK National Study of Hearing that has been cited extensively in this edition and is now readily available in an electronic version.

Mr Iain RC Swan was subsequently appointed as a second otologist to the group and one of Stuart's aims was to prospectively utilise the patients attending both the ORL and Audiological departments at Glasgow Royal Infirmary, and indeed the whole of Scotland, in well-designed and analysed studies. This included the development of the now, internationally used Glasgow Benefit Inventory, and supported Mr Haytham Kubba, then a surgical registrar, in developing a children's version.

The outcomes of GGB's and IRCS's ear surgery were also used in prospective studies, with perhaps the most influential one of the Glasgow Benefit Plot having been suggested by Stuart Gatehouse.

My research involvement with them and with other colleagues, particularly at the MRC IHR Headquarters in Nottingham, has, I hope, added to knowledge but also increased my understanding of the problems that occur at the interface between otology and audiology. This was particularly so with my material involvement with TARGET, the national randomised controlled trial of the management of children with bilateral persistent otitis media with effusion.

George G Browning

Authors

George G Browning qualified in 1964 and was awarded FRCS Surgery in 1970 from the University of Edinburgh, and later FRCS Otorhinolaryngology from Glasgow in 1976. Dr Browning has worked in a number of countries through fellowship training and visiting professorships, including at Harvard University, the University of Cape Town, Flinders University Adelaide and the University of Toronto. He has received multiple awards, including the Walter Jobson Horne Prize from the British Medical Association in 1998 and the George Davey Hotwells Memorial Prize from the University of London in 2000. In 2005, he became an Honorary Member of the Swedish Medical Association, and in 2009 he became an Honorary Member of the South African Otolaryngological Society. During his career, Dr Browning has served on multiple training and examining boards, such as being Vice Chairman of the SAC in Otolaryngology (1997–1999) and the SAC in Audiological Medicine (1998–2000). He was on the Intercollegiate Board in Otolaryngology (1997–2001), and the Final FRCS in both Edinburgh (1979–2002) and Glasgow (1981–2003).

He was President of the Royal Society Section of Otology (1999–2000); Chairman of the Academic Board of the Presidents of the 54 Sections (2001–2003); and Vice-President of the Royal Society of Medicine (2005–2007). From 2004 to 2014, he was the Editor of *Clinical Otolaryngology*, and at the end of his tenure, the impact factor of the journal had risen from near the bottom to being sixth of 34 ORL journals. He was the Otology Editor for the 2008 Seventh Edition of *Scott Brown's Otolaryngology, Head and Neck Surgery*, for which the Glasgow Royal Infirmary ORL team wrote a total of nine chapters. He is currently Visitor MRC/CSO Scottish Section Hearing Sciences, Royal Infirmary, Emeritus Professor in Otorhinolaryngology, University of Glasgow and Honorary Consultant Otolaryngologist, Royal Infirmary, Glasgow.

Emma Stapleton qualified from the University of Edinburgh in 2000. She was inspired by George Browning very early in her career, enjoying his unique yet accessible lectures at various events, and attending his Clinical Otology and Audiology Course in Glasgow. She has received multiple awards including the Hallpike Prize, George Seed Prize, Hunterian Society Prize, Philip Stell Prize, RSM Otology Prize, BACO Audiology Prize, Rowena Ryan Prize, and the Hunter Doig Medal from the Royal College of Surgeons of Edinburgh. Following advanced fellowship training in Glasgow, Manchester, Australia and Canada. She is now a Consultant Otorhinolaryngologist and Lead Auditory Implant Surgeon at the Manchester Royal Infirmary, and Clinical Lead of the Manchester Cochlear Implant Programme. She is President of the North of England Otolaryngology Society, Past Chair of Women in ENT UK, editorial board member of several ENT journals, and is a Trustee of the TWJ Foundation, the *Journal of Laryngology and Otology* and the British Cochlear Implant Group. She is invited to lecture nationally and internationally on her areas of expertise and is pursuing a part-time PhD in Audiology at the University of Manchester.

Haytham Kubba qualified in 1992 from the University of Newcastle upon Tyne. He moved to Glasgow as a Registrar in 1998. He first met George Browning when he was working for his MD at the Medical Research Council's Institute for Hearing Research. He finished his training as clinical fellow in Paediatric Otolaryngology at Great Ormond Street Hospital in London. He has been a consultant in children's ENT in Glasgow since 2003, with a brief interlude for an 18-month sabbatical at the Royal Children's Hospital in Melbourne. He is an Honorary Associate Professor and Subdean for Child Health at Glasgow University. He has been President of the British Association for Paediatric Otolaryngology since September 2024 and was previously on the Council of ENT Scotland and the Otorhinolaryngological Research Society. He was editor of the paediatric section of *Scott Brown's Otolaryngology Head and Neck Surgery* Eighth Edition. He is a regular speaker at international conferences and has organised and taught a number of postgraduate courses, including George Browning's Clinical Otology and Audiology Course.

1

Sound and human hearing

WHAT IS SOUND AND HOW DO WE MEASURE IT?

Sound is the vibration of the molecules of a medium. That medium is usually air but it could be water or, in the case of seismic waves from an earthquake, solid rock. Whatever the medium, the vibration is passed from one molecule to the next, and the sound propagates as a wave of alternate compression and rarefaction. Sound travels at around 343 m/s in air (depending on temperature), but much faster in water (1481 m/s) because the molecules are much closer together and hence the vibration behaviour is passed on more quickly. Sound travelling in air towards a fluid interface, such as the surface of a swimming pool, will almost all bounce off the surface rather than entering the water. In fact, more than 99.9% bounces off the surface. This is because the rarefied molecules of the air struggle to impart enough energy to the much more densely packed molecules of water. This *impedance mismatch* is a very important concept which we will come to again when we discuss the middle ear.

There are two important properties of a sound wave which we describe: *frequency* and *intensity*. Frequency is the speed of the vibration, measured in Hertz (Hz, or cycles per second). We perceive the frequency of a sound as *pitch*, ranging from the deep notes of a double bass all the way up to the squeakiest high pitch notes on a violin. The range of human hearing in a healthy young adult is around 20–20 000Hz.

Intensity is the amplitude of the vibration. In other words, it is the amount of energy carried by the sound wave. We perceive it as *loudness*. The range of human hearing is so enormous that it is hard to comprehend: the loudest thing we can hear is around 20 trillion times louder than the quietest thing we can hear. That range is impossible to work with, so we make it manageable by converting it to a logarithmic scale called the decibel (dB). That turns a 1–200 trillion scale into a much more manageable 0–140 scale but introduces some mathematical quirks which we need to take a minute to understand. A difference of 3 dB equates to double the amount of energy in the wave. A difference of 10 dB equates to 10 times the energy in the wave but we perceive it as twice the loudness (**Table 1.1**). When we are discussing the amount of energy in a sound wave, we use a version of the decibel scale called dB SPL (decibels sound pressure level). For example, we might say a guitar produces 80 dB SPL. This scale is based on the logarithm of the ratio of the sound intensity to a reference value of 20 micro-pascals of sound pressure, which is about the quietest thing that can be heard.

The quietest sound that can be consciously perceived at any given frequency is called the *threshold*. Human hearing is most sensitive around the centre of the range, at the frequencies most important for understanding speech, namely 500–4000 Hz. Above and below this range, it takes a lot more sound energy for a sound to be loud enough to be heard. If we plot a person's hearing using the dB SPL scale we get u-shaped curves like those in **Figure 1.1**. This is not very easy to interpret in clinical practice, so instead we plot the dB HL (decibels hearing loss) which is the threshold referenced to the mean threshold at each frequency of an idealised sample of normal-hearing people. If the person's threshold at a particular frequency is the same as the reference sample, we plot it at 0 dB. If the sound has to be 20 dB louder for the person to hear it compared to the reference, we plot at 20 dB on the chart. If the person hears better than the reference sample, we

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Table 1.1 An illustration of the logarithmic dB scale

Sound	Decibel level	Musical dynamics	Number of times louder than threshold
Threshold of Hearing	0		1
Normal Breathing	10		10
Leaves Rustling	20		100
Empty Theatre	30	ppp	1,000
Mosquito Buzzing	40	pp	10,000
Quiet Restaurant	50	p	100,000
Normal Conversation	60	mp	1,000,000
Traffic	70	mf	10,000,000
Vacuum Cleaner	80	f	100,000,000
Truck Engine	90	ff	1,000,000,000
Subway Train	100	fff	10,000,000,000
Rock Band	110		100,000,000,000
Threshold of Pain	120		1,000,000,000,000
Machine Gun	130		10,000,000,000,000
Jet Engine	140		100,000,000,000,000

plot at −10 or −20 dB. Note that dB HL and dB SPL are both logarithmic ratios.

Other decibel scales exist for use in specific circumstances, such as dB A which is a sound pressure scale weighted according to how much sound is likely to reach the cochlea (low-frequency sounds have to be louder to reach the cochlea than high-frequency sounds). The dB A scale is used in relation to industrial noise exposure.

THE OUTER EAR

The outer ear consists of the pinna and ear canal. The pinna acts like a funnel to collect sound from a large area and channel it into the much smaller ear canal. The canal itself resonates in a way that selectively amplifies sounds around 2000–4000Hz and this, plus the large funnel shape of the pinna, serve to amplify the incoming sound by around 20 dB.

The pinna also aids in sound localisation. In most circumstances in day-to-day life, localisation is most important in the horizontal plane. If a sound source is located to one side, it reaches the ipsilateral ear sooner than the contralateral ear (inter-aural time difference) and is also slightly louder at the ipsilateral ear (inter-aural loudness difference), and these two phenomena allow the source of the sound to be localised (**Figure 1.2**). The forward-facing cupped shape of the pinna means that sounds originating from in front sound louder and contain more high-frequency notes than sounds from behind: this helps localise sounds in the front-to-back plane. The pinna is shaped differently in its superior part from its inferior part and this also affects the frequency distribution of sound collected from above as compared with sound from below, mostly through effects at 7 kHz, aiding localisation in the vertical plane.

Contrary to popular teaching, it is possible to localise sound with one ear, as the necessary variations in sound pressure level can be achieved by minor head movements which change the relative position of the pinna and the ear to the sound.

THE MIDDLE EAR

The middle ear is an air-filled space, connected to the nasopharynx by the Eustachian tube. Sound is collected by the tympanic membrane at the proximal end of the ear canal and transmitted across the middle ear space by three articulated bones (malleus, incus and stapes) to the oval window of the cochlea. This is a complex arrangement which deserves some explanation.

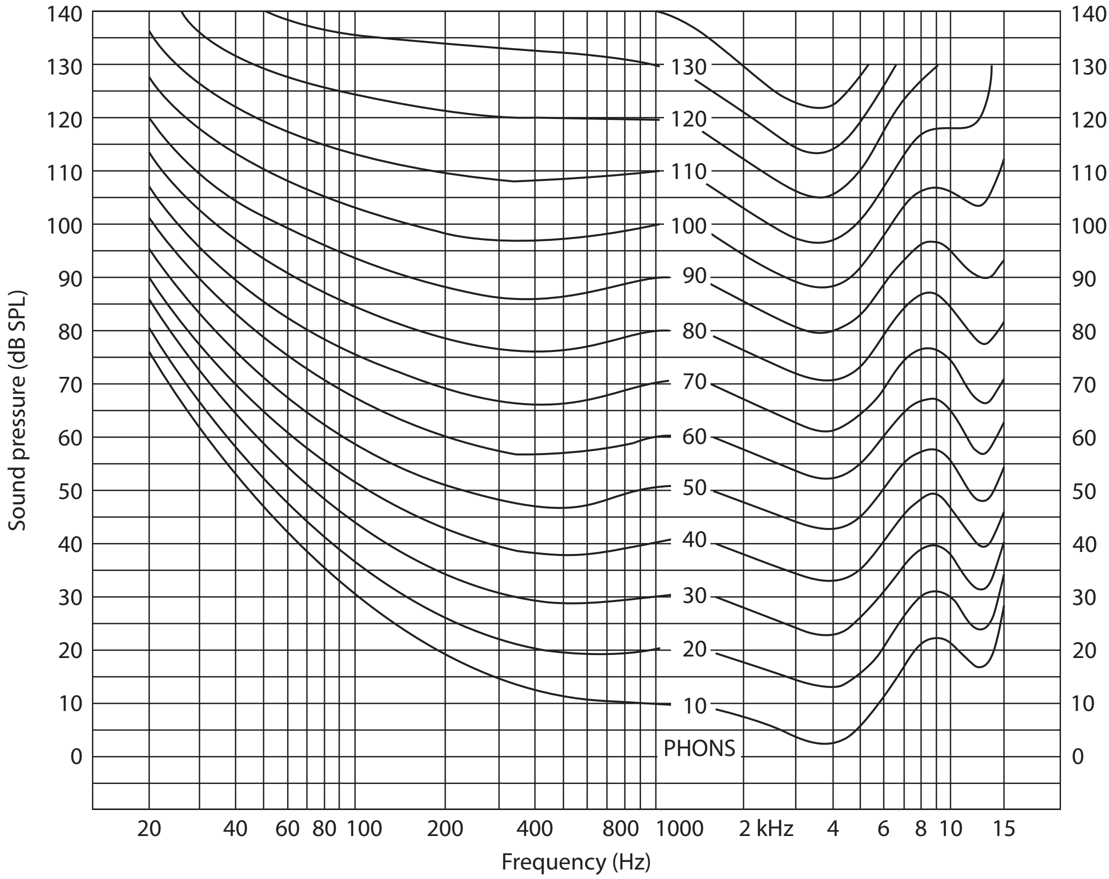

Figure 1.1 The sensitivity of the human ear to sound as measured in dB SPL.

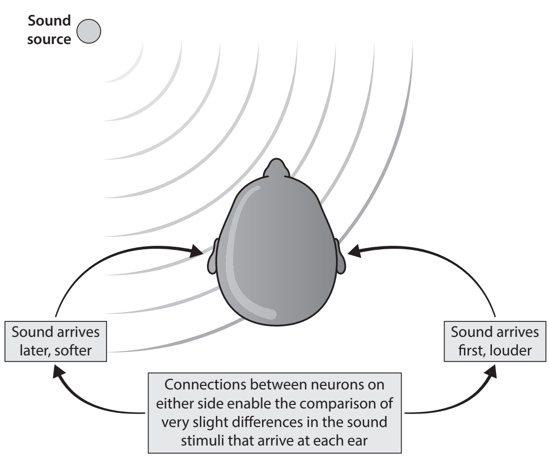

Figure 1.2 Sound localisation in the horizontal plane.

Fish have a series of pores along the side of their body called the lateral line. Within each pore is a patch of sensory epithelium containing sensory hair cells similar to those of the mammalian inner ear. Fish use these to sense sound and vibrations in the sea water which displace stereocilia on the sensory hair cells. When the primitive ancestor of land animals first crawled into land, its world will have suddenly gone very quiet: the impedance mismatch between air and water (see above) means that sound travelling through the air would simply bounce off the fluid-filled sensory apparatus and hearing would no longer be possible. A mechanism to overcome the impedance mismatch evolved in the form of a simple middle ear. The first part of this is a tympanic membrane to collect sound over a very large area. By channelling this onto a much smaller oval window, the amount of energy is effectively amplified (by around 25 dB). Connecting the two is a small bone: it is small enough to vibrate with the tympanic membrane in response to sound, but dense enough to transfer energy to the fluid of the cochlea. This single-ossicle system is the arrangement found in many amphibians and birds and it works well.

Snakes sense vibration by resting their jaws on the ground. They can detect the footprints of a mouse perhaps a mile away and they can identify the direction very accurately. They can do this because the two halves of the jaw are completely separate, allowing "stereo" hearing. Each side of the jaw is made of three articulated bones, the most proximal of which is in contact with the stapes in the middle ear (**Figure 1.3**). In mammals, the two halves of the jaw are fused to form a single mandible, and the two more proximal jaw bones on each side have migrated into the middle ear to become ossicles. This is why the malleus and incus have their embryological origin from the first (mandibular) branchial arch, while the stapes is formed from the second arch.

By strict definition, the tympanum is the anatomical term for the tympanic membrane but by common usage, it has come to include the middle ear space and the ossicular chain. The tympanic membrane is shaped like a curved cone and is lined by a single layer of squamous epithelium on the canal side and a single layer of mucosa on the middle ear side. In between is a fibrous tissue layer whose collagen fibres are arranged in two main directions. There are radial fibres which run centrally and surround the handle of the malleus and there are circular fibres, a localised concentration of which forms the annulus (**Figure 1.4**). This fibrous tissue layer is the functional key to the tympanic membrane, making it a mobile but relatively inelastic structure that allows the sound pressure to be transmitted along the full length of the handle of the malleus.

Stroboscopic studies at high sound pressure levels have shown that there are two areas of

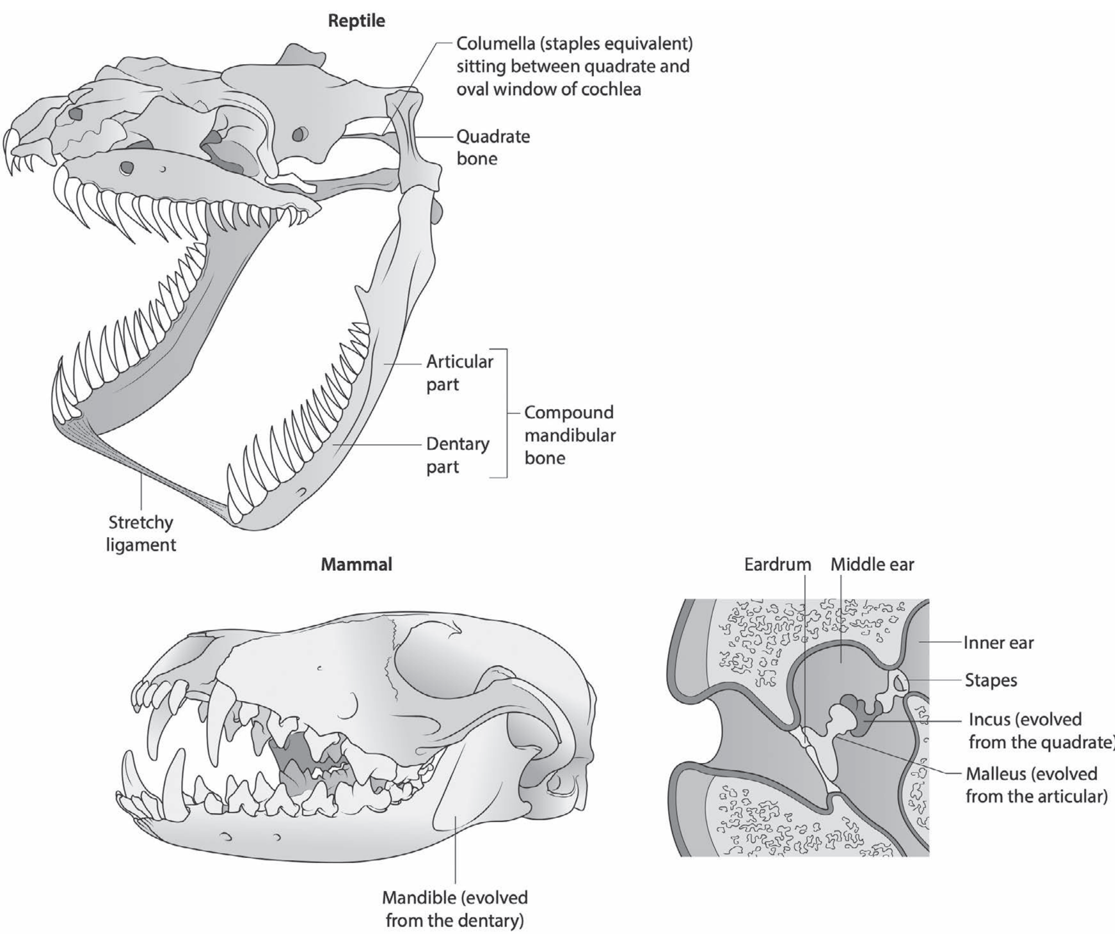

Figure 1.3 Bones of the jaw and middle ear in snakes versus mammals.

Figure 1.4 Tympanic membrane fibres (1).

tympanic membrane movement, one on either side of the malleus handle (**Figure 1.5**). Because of the physical properties of a curved membrane, the force from the movement of these two areas is transmitted to the malleus handle along its full length (**Figure 1.6**). The malleus handle is, therefore, the key to picking up sound energy in a piston system and the suspensory ligaments suspend the ossicular chain to allow movement in a piston-like, rather than a rotatory manner (**Figure 1.7**). In humans, the incudo-malleolar joint would appear to have no function apart from holding the two bones together as there is no movement at the joint. The incudo-stapedial joint, on the other hand, functions as a true synovial joint. The bodies of the incus and malleus probably act as counterweights to the system, which functions optimally at frequencies up to 4 kHz. Above that, the movement of the tympanic membrane becomes uncoordinated, the piston system becomes less efficient and some rotatory or levering movements occur. When there is a tympanic membrane defect or middle ear disease, the major effect on sound transmission will thus be at the lower as opposed to the higher frequencies. This is the main reason why an air–bone gap is greater at the low as opposed to the high frequencies.

The 3-ossicle system provides a small additional amount of amplification through leverage (around 2 dB HL overall), and the main effect is at high frequencies. This, together with some changes in the cochlea, means that that the frequency range of mammals (up to 20 000Hz) extends much higher than that of lizards, for example (up to 4000 Hz).

The role of the stapedius and tensor tympani muscles is usually described as preventing the inner ear being damaged by external noise. This may be partly true for the stapedius muscle but is

Figure 1.5 Areas of maximum tympanic membrane movement.

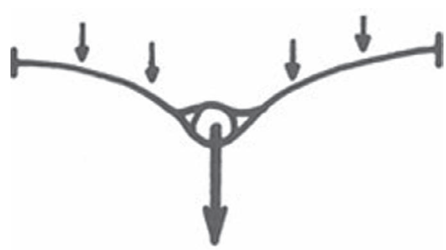

Figure 1.6 Physical properties of the tympanic membrane which transfers all its force of movement to the handle of the malleus along its entire length.

completely untrue for the tensor tympani muscle. In humans, the tensor tympani does not contract in response to sound. It may only be there to stop the tympanic membrane being blown out by sudden increases in middle ear pressure, as may occur during sneezing. The stapedius muscle does contract in response to sound, but this only attenuates the lower frequencies. The contraction occurs too slowly to be of value for impulse noise, and fatigues too quickly to be of value for sustained loud noise. Their surgical division, as in stapedectomy or tympanoplasty, is likely to be of little consequence.

THE INNER EAR

The scala vestibuli and scala tympani are in continuity, forming one long fluid-filled tube with the oval window at one end and the round window at the other. The stapes sits in the oval window and when it pushes in, the fluid in the scala vestibuli/tympani is pushed in and the round window bulges out (**Figure 1.8**). In the opposite phase of

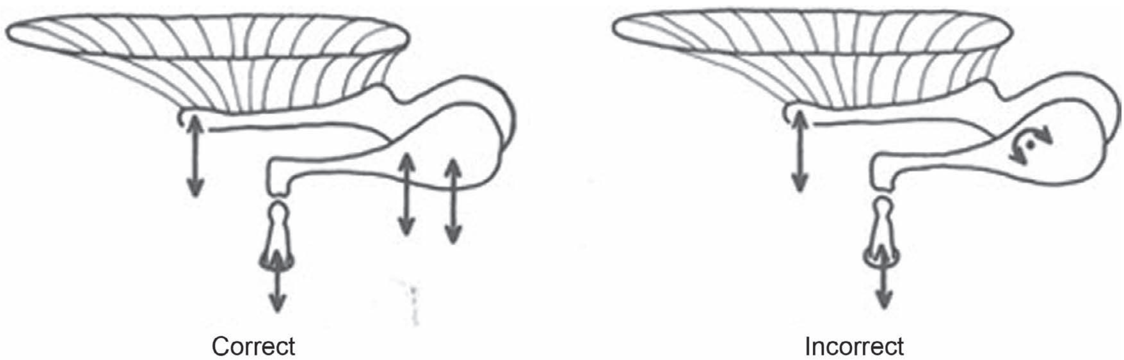

Figure 1.7 The main movement of the ossicles is piston-like, as shown in the figure on the left, certainly at low frequencies. A rotatory or levering action, shown on the right, is much less important.

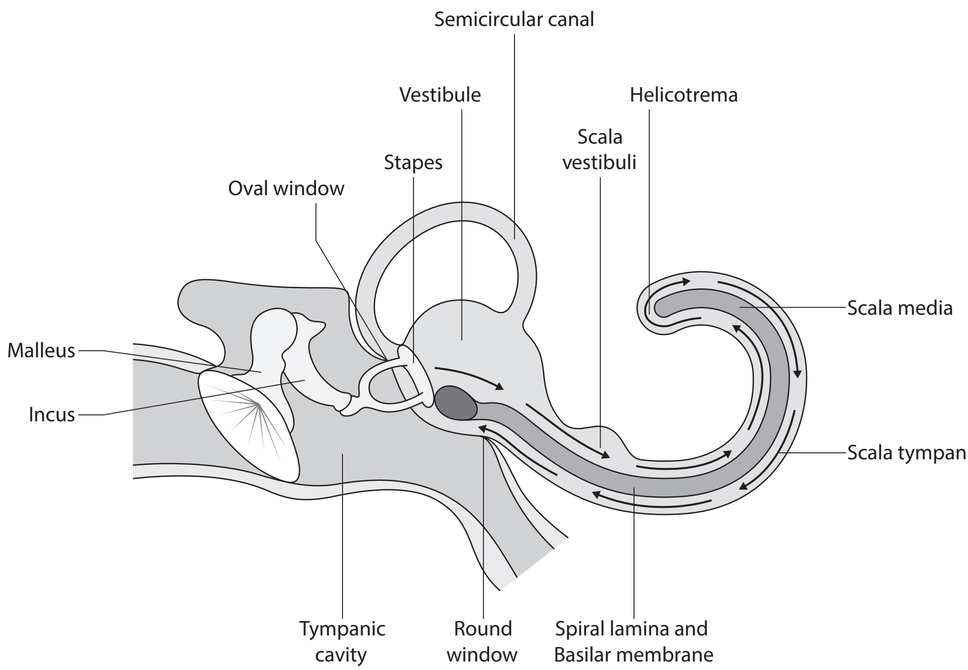

Figure 1.8 Vibrations of the stapes are transmitted through the oval window to the perilymph of the scala vestibuli and scala tympani.

each vibration, the round window moves in and the oval window bulges out. Fluid is incompressible, so unless there are two windows, no vibration can occur. Vibration of the stapes leads to vibration of the fluid (*perilymph*) in the scala vestibuli/tympani. This in turn leads to vibration of the basilar membrane which separates the scala tympani from the scala media.

The scala media is a completely separate channel which is filled with *endolymph*. Endolymph has an unusual composition, having a high concentration of potassium and a low concentration of sodium. Perilymph, in contrast, is a pretty standard extracellular fluid, similar to plasma and cerebrospinal fluid (CSF), being rich in sodium and poor in potassium. The endolymph is produced, and its ionic composition maintained, by the stria vascularis.

Sitting on the basilar membrane are the hair cells of the organ of Corti (**Figure 1.9**). There is one row of *inner hair cells* and there are three rows of *outer hair cells*. The hair cells have bundles of *stereocilia* projecting from their superior surface, embedded within the stiff *tectorial membrane*. When the basilar membrane is vibrated by the movement of perilymph in the scala tympani, the hair cells themselves are vibrated. The stereocilia, being fixed in the stiff tectorial membrane, are deflected as the cell bodies are vibrated. This causes depolarisation of the inner hair cells. Impulses travel along the axons in the cochlear nerve, whose cell bodies are in the spiral ganglion in the centre of the spiral of the cochlea. The inner hair cells are the *sound receptors*, acting as *transducers* converting mechanical energy from vibration into electrical energy in the cochlear nerve.

Pitch is encoded by position in the cochlea. The basilar membrane is *stiff* and *light* at the end nearest the oval and round windows, but *flexible* and *heavy* towards the apex of the cochlea. This difference in the physical properties of the basilar membranes means that its resonant frequency varies along its length: high-frequency sounds will vibrate at the base and low-frequency sounds at the apex (**Figure 1.10**).

The outer hair cells are not sound receptors: their function is much more interesting. They act as a *cochlear amplifier* because they contract in response to sound. At the point of maximal vibration of the basilar membrane, the outer hair cells contract and, in doing so, amplify the vibration by up to 50 dB. At adjacent points on the basilar membrane, they contract out of phase to dampen down the vibration, thus increasing the sound specificity and discrimination of the cochlea. The noise that the outer hair cells make as they contract is known as the *cochlear microphonic* and it can be measured as a test of cochlear function (*see* **Chapter 7**).

The axons in the cochlear nerve synapse in the *cochlear nucleus* of the pons. From here, some fibres pass directly to the ipsilateral *superior olive complex*, and some cross the midline of the pons (decussate) in the *trapezoid body* to reach the contralateral superior olive complex. This is where inter-aural time and loudness differences are processed for sound localisation. Axons continue up through a tract (axon bundle) called the *lateral leminiscus* to reach the *inferior colliculus* of the midbrain and then the *medial geniculate body of the thalamus*. Electrical activity in this pathway can be measured (*see* **Chapter 7**).

The first part of the cortex to receive sound information is the *primary auditory cortex* in the

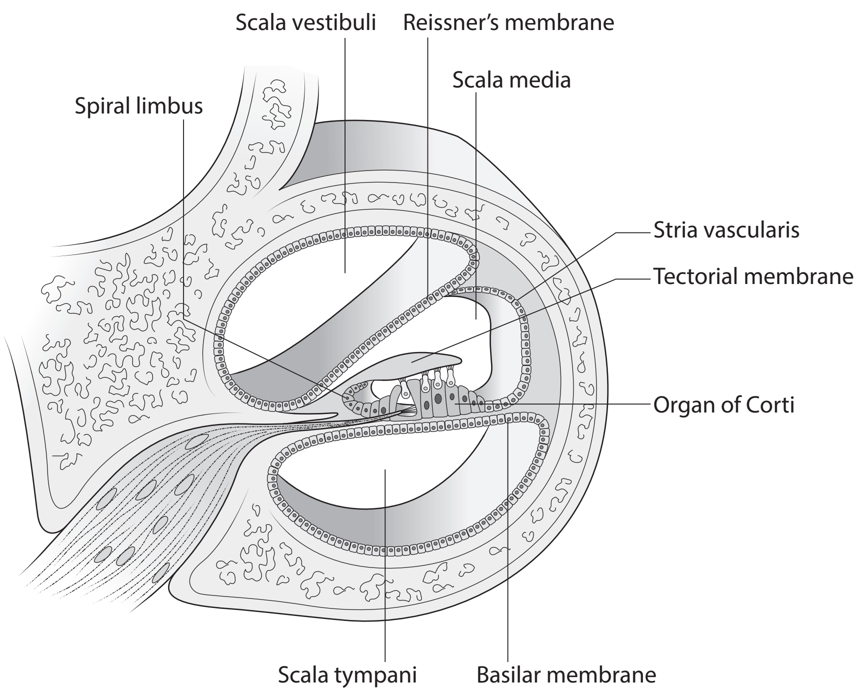

Figure 1.9 Cross section of the cochlea.

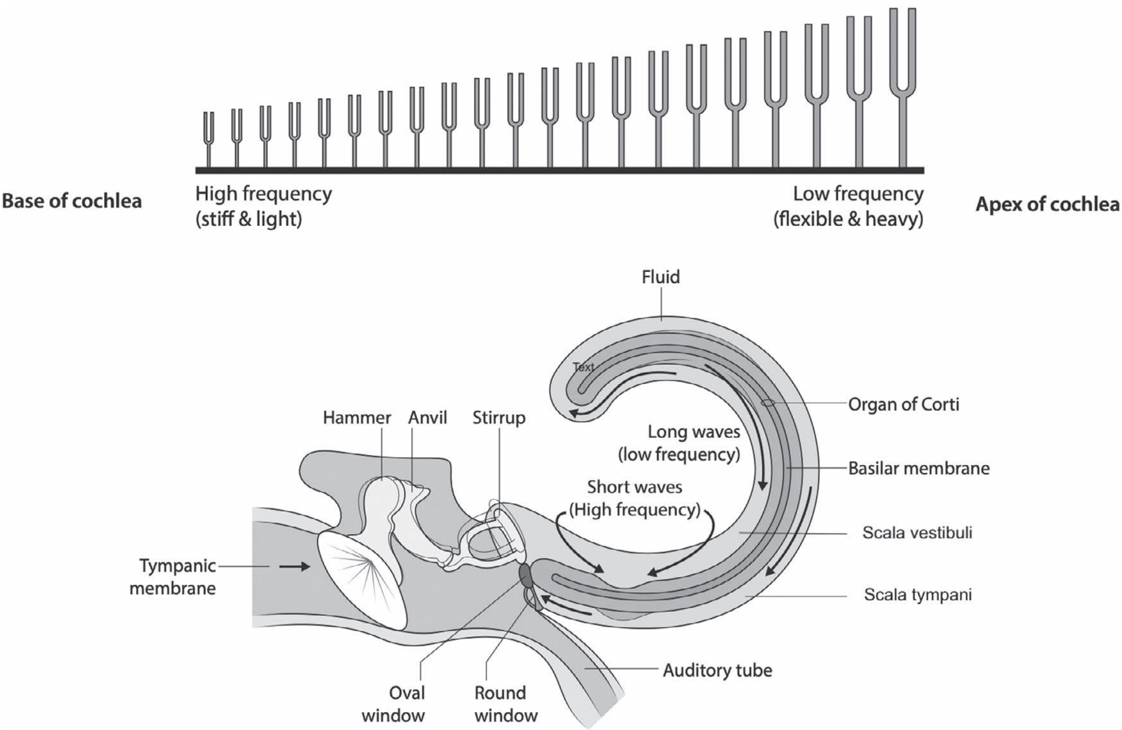

Figure 1.10 Pitch is encoded by place along the basilar membrane, whose physical properties and therefore resonant frequencies vary from basal end to apex.

superior temporal gyrus. This is where we first become consciously aware of sound.

NORMAL DEVELOPMENT AND AGEING

The middle ear and cochlea are well formed in the fetus by 15 weeks' gestation. The neural pathways are in place by 20 weeks and the hearing system is functional by 25 weeks' gestation. Between 25 weeks' gestation and 6 months of age, the auditory system undergoes a period of rapid development during which the response to frequency and intensity of sound is shaped in the cochlea, cochlear nerve and auditory cortex. This requires sound input including speech, music and environmental noise, and cannot occur if there is continuous background noise of 60 dB or more (which can be a major issue on a neonatal intensive care unit, NICU). Note that this process starts *in utero* and the effect on sound processing is lifelong.

In children with congenital hearing loss, central auditory pathways will not develop unless sound stimulation is restored by some means before approximately 18 months of age. Beyond this age, the central pathways will have been repurposed for other forms of sensory processing and will be permanently unavailable for hearing.

The ear canal increases in length and volume over the first 2 years of life, and the middle ear volume increases until adolescence. These changes will have some effect on frequency response. The most important changes that occur in childhood, however, are those that affect concentration, motivation and cognition, as it is these that determine the limits of behavioural testing in children (*see* **Chapter 5**).

Hearing thresholds for high-frequency sounds increase predictably with age. While most children can hear sounds up to 20 kHz, the upper limit of hearing is usually around 17 kHz at age 25 years, 16 kHz at 30 years, 15 kHz at 40 years, 12 kHz at 50 years and 8 kHz thereafter.

EPIDEMIOLOGY OF HEARING LOSS

At birth, around 1.3 per thousand children in the UK will have a permanent bilateral hearing loss of 40 dB HL or more (**Figure 1.11**) (3). Around one-third have environmental causes, of which the most common is congenital cytomegalovirus (CMV) infection, accounting for around 20% of congenital deafness overall. Others include toxoplasmosis, rubella, syphilis and herpes. Two-thirds are genetic in origin. Syndromes account for 20% of these, with Pendred being the most common. Around 80% of genetic deafness is nonsyndromic, meaning the hearing loss is an isolated finding in a child who is healthy in all other respects. Of these, 80% are autosomal recessive (so a clear family history of deafness is unlikely), 19% are autosomal dominant and 1% are X-linked or mitochondrial in inheritance (**Figure 1.12**). *GJB2* (connexin 26) mutations account for the greatest number of children with autosomal recessive nonsyndromic deafness, and they constitute around 20% of all congenital cases of deafness (**Table 1.2**).

By 9-years-old, the prevalence rises to at least 2 per 1000 (4). Some of these are acquired, mostly from bacterial meningitis. The rest are progressive hearing losses of congenital origin which may be genetic (most commonly Pendred) or environmental (mostly CMV).

The prevalence of hearing loss increases with age, with 26% of 50-year-olds having a hearing loss of at least 25 dB HL, rising to 71% of 70-year-olds (5). Age-related hearing loss (formerly known as presbycusis) is the most common reason, but noise-induced hearing loss, ototoxic drugs, skull fractures, infection and tumours all contribute. This equates to around 12 million people with hearing loss in the UK and the number is rising. Hearing loss can lead to social withdrawal and isolation, and people with hearing loss are more likely to develop depression and dementia (6).

Age-related hearing loss is not completely understood but loss of hair cells, atrophy of the stria vascularis and loss of cochlear neurons all play a part. There are genetic factors involved, as well as environmental factors such as cigarette smoking, noise exposure and head trauma (7).

In the developing world, the burden of hearing loss is much greater. Our best current estimates are that around 1.57 billion people (20% of the world's population) have a hearing loss of some degree. It is of moderate or worse severity in 430 million people (5% of the world's population) and that number is rising (8). Many of the causes of hearing loss are more prevalent in developing world settings including pre- and peri-natal complications, chronic otitis media, industrial noise trauma and exposure to ototoxic drugs.

These issues will be discussed more in **Chapters 9** and **10**.

CONCLUSIONS

- The frequency of a sound wave (which we perceive as pitch) is measured in Hz and the range of human hearing is around 20–20 000 Hz.
- The intensity of a sound wave (which we perceive as loudness) is measured in decibels and the range of human hearing is 0–140 dB HL. A difference of 10 dB is 10 times the amount of sound energy, but we perceive it as twice the loudness.
- The middle ear is a mechanism for overcoming the impedance mismatch between sound travelling through air and sound travelling through fluid.

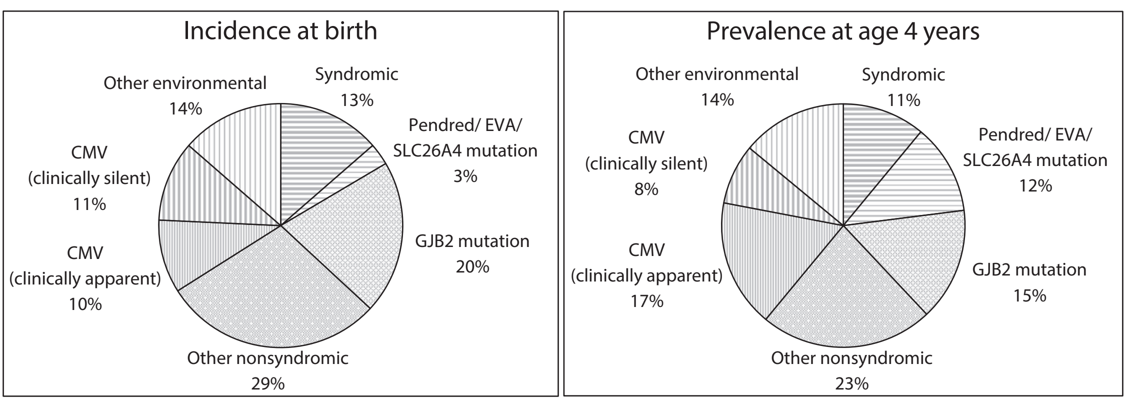

Figure 1.11 Permanent bilateral hearing loss in children by aetiology at birth and age 4 (after Morton & Nance [2]).

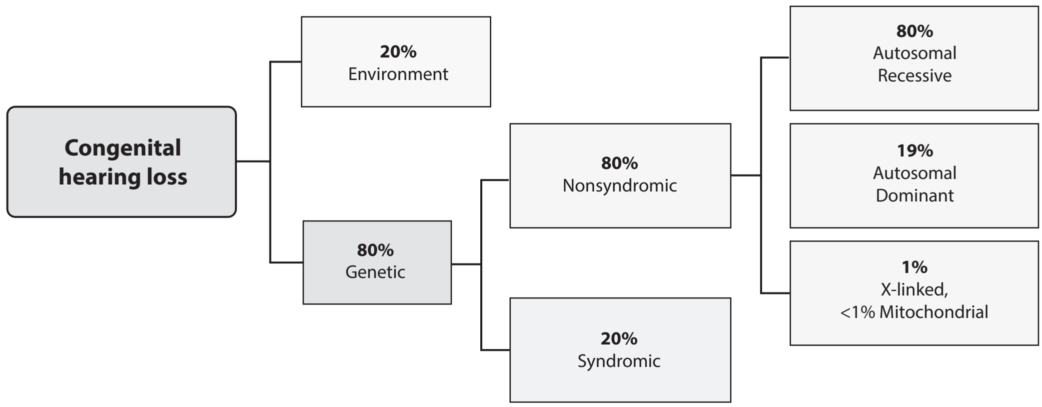

Figure 1.12 Causes of congenital hearing loss.

Table 1.2 Frequent aetiologies of syndromic and nonsyndromic hearing loss

Clinical feature	Mode of inheritance	Syndrome or gene
Syndromic hearing loss	Autosomal dominant	• Neurofibromatosis 2 • Branchio-oto-renal syndrome • Treacher Collins • Stickler syndrome • Waardenburg syndrome
	Autosomal recessive	• Pendred syndrome • Jervell and Lange-Nielsen syndrome • Usher syndrome • Refsum disease
	X-linked dominant	• Alport syndrome
	Mitochondrial	• MELAS • MERRF
Nonsyndromic hearing loss	Autosomal dominant	• *WFS1* • *TECTA* • *COCH* • *KNCQ4*
	Autosomal recessive	• *GJB2* • *SLC26A4* • *MYO15A* • *OTOF* • *CDH23* • *TMC1*

Abbreviations: CDH23, cadherin-related 23; *COCH*, cochlin; *GJB2*, gap junction protein beta 2; *KNCQ4*, potassium voltage-gated channel subfamily Q member 4; MELAS, mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes; MERRF, myoclonic epilepsy with ragged red fibers; *MYO15A*, myosin XVA; *OTOF*, otoferlin; *SLC26A4*, solute carrier family 26 member 4; *TECTA*, tectorin alpha; *TMC1*, transmembrane channel like-1; *WFS1*, wolframin ER transmembrane glycoprotein.

- The inner hair cells are the sound receptors; the outer hair cells are the cochlear amplifier.
- The prevalence of permanent hearing loss of 40 dB HL or more is around 0.13% at birth, rising to 0.2% in childhood. Eighty per cent of cases are genetic in origin, and most of these are not associated with a syndrome.
- The prevalence of at least 25 dB HL hearing loss rises with age to 26% at age 50, and 71% at age 70.

FURTHER READING

For more information on working with decibels, please see: www.animations.physics.unsw.edu.au/jw/dB.htm (Accessed 17 July 2024).

For more information about the epidemiology of hearing loss please see: Choe G, Park SK, Kim BJ. Hearing loss in neonates and infants. *Clin Exp Pediatr* 2023; 66(9): 369–76.

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2

Clinical evaluation of ears and hearing

INTRODUCTION

When there are abnormalities in the auditory system, patients usually present with a hearing impairment, and otolaryngologists usually devote most of their attention to identifying the site affected and the pathology responsible. Though this is important, it is perhaps more important to assess the resultant disability because, at present, there is no specific therapy to halt or reverse most of the pathological conditions that affect the inner ear. As such, in most instances the management is of the disability rather than that of the underlying condition. Unfortunately, methods of assessing the disability tend to have been neglected. This makes it difficult to gauge the relative benefits of surgery or the provision of a hearing aid in relieving the disability.

Hearing Pathology

Pathology can affect any anatomical part of the auditory pathway (**Figure 2.1**), singly or in combination. The two most common combinations are 1) when inflammation in the middle ear spreads to the inner ear and brain and 2) when ageing of the inner ear is associated with more general neurological pathology.

Pathology may or may not be symptomatic. Post-mortem temporal bone histology frequently identifies otosclerotic foci or vestibular schwannomas that were clinically silent. However, with these two exceptions, the majority of ear pathology is symptomatic. History taking of symptoms has the dual objective of helping identify the pathology and of assessing the associated disability (see below). In those with ear pathology, hearing impairment is the commonest and usually the most dominant symptom. In addition, tinnitus is frequently, either intermittently or continuously and the disability associated with its presence varies greatly with multiple contributing mental factors.

Hearing Impairment

Audiometry is the main scientific method of assessing the hearing. Air conduction threshold averages on pure tone audiometry (PTA), or less frequently speech audiometry, are used to determine the degree of impairment (**Table 2.1**). The addition of bone-conduction thresholds on audiometry, along with otoscopy, helps define the pathological site and resultant type of the impairment.

- A conductive impairment is due to pathology in the external or middle ear, and primarily attenuates sound in terms of its loudness.

Classification									
Anatomical	Outer ear	Middle ear	Inner ear	VIII nerve	Low brainstem	High brainstem	Cerebellum	Auditory reception	Non-auditory reception
Physiological	Peripheral				Central				
Pathological	Conductive		Sensorineural		→	?	Brain		

Figure 2.1 Auditory pathway.

DOI: 10.1201/b23377-2

Table 2.1 WHO Classification system used in this text to describe the severity of a hearing impairment (the pure tone averages are of 0.5, 1, 2 and 4 kHz in the better hearing ear)

Pure tone average (dB HL)	Severity of impairment
0–25	None
26–40	Mild
41–60	Moderate
61–80	Severe
81–110	Profound
110+	Total

(After WHO [2].)

Table 2.2 UK adult population prevalence (%) of hearing impairment in the better hearing ear

PTA (dB HL)		Population prevalence	Cumulative prevalence
None	<25	84.0	84
Mild	25–29	4.9	89
	30–34	3.0	92
	35–39	2.4	94
Moderate	40–44	1.9	96
	45–54	1.8	98
	55–64	1.0	99
Severe	65+	1.0	100

(After Davis [1].)

- A sensory impairment is due to pathology in the inner ear, most commonly loss of hair cells and, apart from attenuating the loudness of the sound, there is often a loss of frequency discrimination and timing (temporal) abilities. Clinically and audiometrically, there may be evidence of recruitment, when sounds can appear abnormally loud.
- A neural impairment is due to pathology in the cochlear division of the eighth cranial nerve and, because it can affect both the afferent and efferent nerve fibres, has the same effect as a sensory impairment.
- Central impairments are caused by pathology in the brainstem or higher audiological centres where the afferent neurological input from both ears is processed for localisation and interpretation such as speech. Audiometric assessment of central auditing function is difficult and primarily of research interest rather than of clinical or diagnostic value.

Prevalence of Hearing Impairment

Ascertaining the prevalence of hearing impairment in the population has to take into account that humans have two ears. In medical practice, when the prevalence of pathology is being determined, it is usual to take it as the percentage of individuals with a disease in one or both ears, with the ratio of the population with unilateral/bilateral disease being stated.

Establishing the occurrence of hearing impairment can be done in a similar manner, after defining what constitutes a hearing impairment. This is usually taken from the four-frequency average (FFA) of the pure tone thresholds over 0.5, 1, 2 and 4 kHz. The average that defines a hearing impairment is usually taken as 25 dB HL.

For clinical purposes, different averages are used that reflect the most affected frequencies for that condition. For example, in conduction impairments a three-frequency average over 0.5, 1 and 2 kHz and, for noise trauma, a three-frequency average over 1, 2 and 3 kHz can be used.

It is also possible to state the prevalence of different degrees of impairment using the WHO classification of impairment (**Table 2.2**) Using such systems can lead to confusion in two aspects. The first when there is a difference in the degree of impairment in the two ears. Thus, an individual with a severe impairment in one ear and mild hearing impairment in the other ear is not severely deaf, as disability is primarily determined by the hearing in the better hearing ear (see below). The WHO classification makes this interpretation clear by saying that their classification applies to the better hearing ear. The second confusing aspect is that the best hearing category is called ‘None’ rather than ‘Normal’ (2).

The prevalence of hearing impairment needs to be known for the better and poorer hearing ears. **Table 2.3** shows the prevalence of hearing impairment in the adult British population as assessed in the MRC National Study of Hearing (3). Overall, 14% have an impairment of 25 dB HL or poorer in their better hearing ear, but this increases to 42% in those over the age of 60 years. Impairments are

Table 2.3 Amount (dB) the head shadow attenuates sound coming via air from the contralateral ear

	Frequency (kHz)						
	0.25	0.5	1	2	3	4	8
Mean	1	1	1	7	10	13	10
Maximum	2	4	5	11	15	18	15
Minimum	0	0	0	5	9	10	5

(After Shaw [20].)

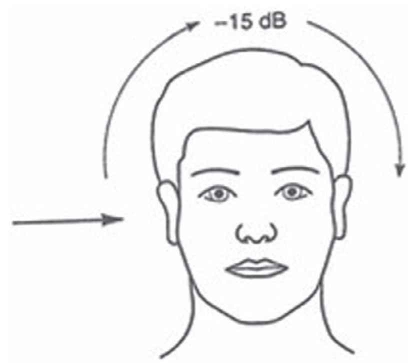

Figure 2.2 Average transcranial attenuation of tree-field sound in a quiet environment.

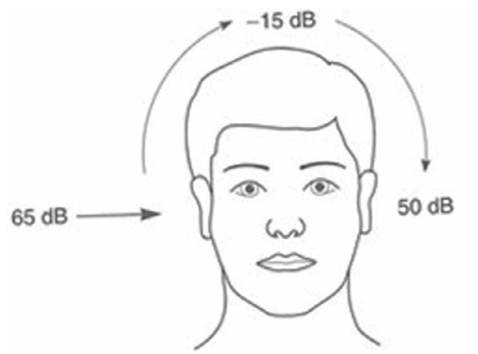

Figure 2.3 Example of transcranial attenuation.

commoner in men and those in manual occupations. Overall, sensorineural impairments are more common than conductive impairments when defined as an air–bone gap of 15 dB or greater over 0.5, 1 and 2 kHz. Conductive impairments constitute a higher proportion in the poorer hearing ear and in the young. Prevalence data for children are discussed in **Chapter 8** (Otitis Media with Effusion) and **Chapter 9** (Sensorineural Hearing Impairment).

Hearing Disability

In most circumstances, a patient's hearing disability is determined by the hearing in their better hearing ear. This is because sound, even though presented on the poorer side, will be heard by the other better hearing ear because sound can go round the skull, albeit attenuated in its loudness (**Figure 2.2**). The amount of attenuation varies with frequency (**Table 2.3**) but for the speech frequencies, it is usually taken as −15 dB. Thus, a patient totally deaf in one ear, but with no impairment in the other, will hear a speaker on their dead ear side at 65−15 = 50 dB SPL. This is sufficiently loud for speech to be understood (**Figure 2.3**).

Thus, if one wishes to know the population prevalence of hearing-impaired individuals the prevalence of hearing impairment in the better ear is looked at. In these circumstances, it is useful to grade the severity of the impairment because the main use of such data is for policy making (**Table 2.3**). If one wishes to know the prevalence in the population to determine the needs for hearing aids, the majority of individuals do not seek advice until their thresholds are poorer than 45 dB HL even though an individual with a 25 dB HL threshold in their better hearing ear would benefit from a hearing aid. Two per cent have an impairment greater than 45 dB HL in their better hearing ear which is thus the relevant figure to take rather than the prevalence of hearing impairment greater than 25 dB HL in the poorer ear (**Table 2.4**).

CLINICAL EVALUATION OF AN ADULT WITH HEARING SYMPTOMS

History

History taking in a subject with hearing symptoms can:

- help to diagnose a pathology
- assess the patient's symptoms and consequent disability
- formulate management options with knowledge of what has already been tried.

Because of our ability to diagnose the majority of external and middle ear conditions by otoscopy, the history is often of little additional value in

Table 2.4 Population prevalence (%) in adults of a hearing impairment* in the better and poorer hearing ears, overall and in 20-year age bands, broken down into sex and occupational groups and with the ratio of sensorineural (SN) to conductive (CON) impairments

	Prevalence impairment in better ear	SN/CON ratio	Prevalence impairment in poorer ear	SN/CON ratio
Overall prevalance	14 (13–15)	6.5:1	18 (16–19)	2:1
Age (years)				
18–40	1.5 (1–3)	4:1	3.5 (2–5)	1:1
41–60	11 9–13)	4:1	17 (15–20)	2:1
61–80	42 (38–46)	11:1	45 (41–50)	3:1
Sex				
Women	12 (11–14)	5:1	15 (13–17)	2:1
Men	16 (14–17)	8:1	21 (19–23)	2.5:1
Occupational grouping				
Non-manual	12 (11–14)	7:1	17 (15–19)	3:1
Manual	15 13–17)	6:1	19 (17–21)	2:1

*: A hearing impairment is defined as a pure tone threshold averaged over 0.5, 1, 2 and 4 kHz of 25 dB HL or poorer. The 95% confidence limits are given in parentheses (after Browning and Gatehouse [3].)

arriving at a diagnosis. Thus, in a patient with otoscopic evidence of chronic otitis media, the object of taking a history is not to make the diagnosis but to assess the resultant disability caused by the symptoms and to plan management. Equally, if an audiogram is available when a patient is interviewed, history taking is very different in those with a sensorineural as opposed to a conductive impairment. The temptation then is not to carry out the clinical assessment in the traditional order of history followed by examination. Indeed, the order of partial history followed by examination, including audiometry followed by further history has much to commend it.

The History in Diagnosis

Table 2.5 is a list of the commoner ear conditions, with those that can be diagnosed by otoscopy being asterisked. These comprise most external and middle ear conditions. Inner ear conditions are mainly diagnosed from the age of the patient with additional potential aetiologies enquired about. How a specific sensorineural diagnosis is made is discussed in more detail in **Chapter 10**.

Knowledge of previous ear surgery along with identification of ear scars can aid interpretation of the otosopic findings and clarify future management options.

Table 2.5 Main method of diagnosis

Commoner otological conditions	Main method of diagnosis	
	History	Otoscopy
Wax		*
Otitis externa		*
Acute otitis media		*
Otitis media with effusion		*
Chronic otitis media		*
Otosclerosis	*	
Traumatic ossicular disruption	*	
Sensorineural impairment:	*	
Age related	*	
Noise trauma	*	
Drugs	*	
Head injury	*	
Vestibular schwannoma	*[a]	
Vestibular syndromes	*	

[a]: + radiology

History to Assess Hearing Disability

Patients with hearing impairment vary considerably in the degree and circumstances where it is disabling. They not infrequently also have tinnitus. Correspondingly, the degree and symmetry of the loss, along with the patient's lifestyle, and motivation, combine to give different degrees of disability. With additional symptoms such as ear discharge or discomfort, the factors that contribute to the disability are less recognised but psychosocial factors have a considerable impact overall.

A fuller discussion of how to assess hearing disability is in **Chapter 11**, but it is important to realise that disability is usually what determines management rather than the diagnosis or symptoms *per se*. Appropriate management strategies are covered in **Chapters 11** and **12**.

OTOSCOPY

How to perform otoscopy and interpret the findings would take a textbook to cover (see, for example, Wormald and Browning 1996 [4] and Browning and Wormald 2019 [5]) and will only be partially covered in this text. Suffice to say that the likely diagnosis varies with the patient's age and symptoms. Thus, a young adult with a hearing impairment is more likely to have chronic otitis media than a sensorineural impairment, whilst an elderly patient is more likely to have a sensorineural impairment than chronic otitis media, though in both cases they may have a mixed impairment.

Methods

In the majority of patients, an adequate view of the ear canal and tympanic membrane including the attic can be performed with a hand-held otoscope with the largest speculum 'possible'. Removal of wax or pus to visualise the tympanic membrane is mandatory if there is a conductive impairment so that an otoscopic diagnosis can be made. This is best done with careful suction under microscopic vision.

An otoscope attached to a smart phone or endoscope are alternative techniques, which can also take photographs for record-keeping purposes or to send to colleagues for their opinion. Increasingly, image analysis of digital photos is being used for automated screening and training purposes and this will continue to expand with artificial intelligence (AI) (6).

Otoscopic Pathology

The otologist, when they see a patient with a hearing impairment, will diagnose by otoscopy the majority of pathologies of the external auditory canal and middle ear, the main exception being otosclerosis. On some occasions, the tympanic membrane may be difficult to assess and tympanometry can be of value (*see* **Chapter 7**). In those with evident canal or middle ear pathology, the air–bone gap in PTA will quantify the magnitude of the conduction defect. The degree of impairment will be assessed from the air conduction (AC) thresholds. Sometimes, when otoscopy is normal, the audiogram will suggest a conduction defect. The task then is to confirm that this is indeed the case and reach a diagnosis.

Prevalence of Middle Ear Pathology

The British MRC National Study of Hearing assessed the prevalence, in adults over the age of 18 years, of middle ear disease in the early 1980s (**Table 2.6**). It is unlikely to have changed since then. Otoscopically, 12% (95% confidence interval (CI) 10.2–13.6) had healed otitis media, 2.6% (CI 1.8–3.4) had inactive chronic otitis media and 1.5% (CI 1.1–1.9) had active chronic otitis media. Unfortunately, the patients with active chronic otitis media could not be broken down into cholesteatoma and mucosal disease. Defining otosclerosis as a normal tympanic membrane and normal middle ear pressure, but with an air–bone gap of 15 dB over 0.5, 1 and 2 kHz, 2% (CI 1.5–2.7) of the population had these findings.

Wax (Cerumen)

The otoscopist has to be aware that what sometimes appears to be wax is not wax. Dried pus and squamous debris are frequently brown and can be confused with wax, particularly in a mastoid cavity.

The inexperienced frequently consider wax to be the cause of a conductive hearing loss. Wax is secreted by the ceruminous glands in the cartilaginous, outer one-third of the canal. Normally, wax is shed along with dead squamous epithelium from the skin of the canal by migration of the epithelium. Sound vibrations are not usually impeded in the canal unless it is so occluded that the vibrations cannot pass through a gap. Ear-plugs worn for sound protection work in this manner, provided they are pressed right in. Wax in the outer third of the canal is never sufficiently occlusive to achieve this effect. However, if impacted by a finger, cotton

Table 2.6 Prevalence in adult population (%) of chronic otitis media and otosclerosis (the 95% confidence limits are given in parentheses)

	Chronic otitis media		
	Active	Inactive	Otosclerosis
Overall prevalence	2.5 (2–3.5)	1.5 (1–2)	2 (1.5–3)
Age grouping			
18–40 years	2.5 (1–4)	1 (0.5–1.5)	1.5 (1–3)
41–60 years	2 (1–3)	2 (1–3)	2 (1–3)
61–80 years	3 (2–4)	2 (1–3)	3(1–4)
Sex grouping			
Women	2.5 (2–3)	1 (0.5–1.5)	2 (1–3)
Men	3 (1.5–4)	2 (1–3)	2 (1–3)
Occupational grouping			
Non-manual	2 (1–3)	1 (0–1)	1.5 (1–2)
Manual	3 (2–4)	2 (1.5–3)	3 (2–4)

(After Browning and Gatehouse [3].)

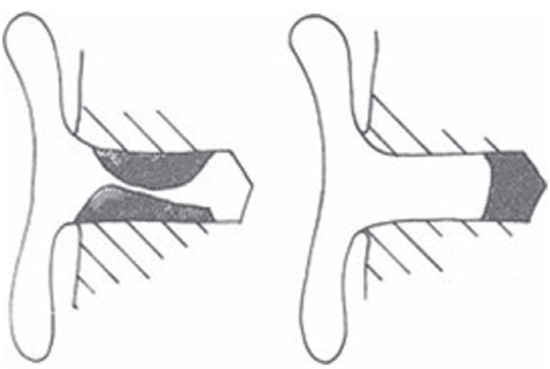

Figure 2.4 The wax on the left would not cause an acoustic obstruction, whereas that on the right would.

bud or mould of a hearing aid deep in the canal, it may do so (**Figure 2.4**). Impacted wax against the tympanic membrane may also sometimes directly inhibit its mobility. Debris on the tympanic membrane may affect its loading and its frequency response but seldom causes a hearing loss. This is in contradistinction to a drop of water, which will immobilise it by surface tension, in the same way as middle ear fluid does in otitis media with effusion.

Otitis Externa

Otitis externa seldom causes a significant hearing loss. It only does so when the canal is narrow and there is retention of debris deep in the canal. Sometimes, the tympanic membrane itself is involved and becomes oedematous and less efficient at transferring sound.

Otitis Media with Effusion

Various terms have been used to describe this condition in the past, such as serous otitis media, secretory otitis media, non-purulent otitis media, chronic non-suppurative otitis media and glue ear, but otitis media with effusion (OME) is the preferred term. It is defined as the presence of a middle ear effusion of fluid for at least 3 months in the absence of overt signs of infection, and it is most prevalent in pre-school age children. Aetiology and management are discussed in **Chapter 8**. This section discusses diagnosis.

Clinicians will vary in their otoscopic ability to diagnose OME but the overall 'hit rate' appears to be remarkably poor unless tympanometry is also performed (*see* **Chapter 7**). Otoscopic features include a dull tympanic membrane which may be grey or yellow in hue, with prominent blood vessels running radially (like spokes on a wheel) and a retracted position such that the malleus handle appears shortened and horizontal. The antero-inferior light reflection may be broken up (**Figure 2.5**).

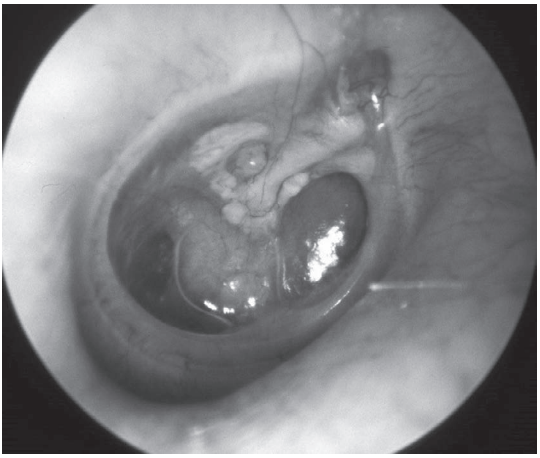

Figure 2.5 Otitis media with effusion.

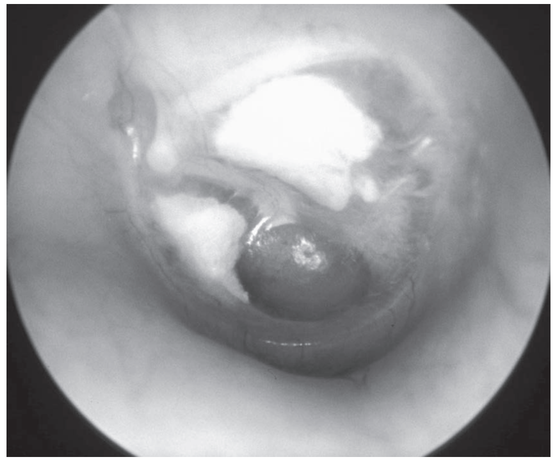

Figure 2.6 Healed chronic otitis media.

For those less well trained, video otoscopy, using a smart phone, and cloud-based otitis media diagnosis is developing (6, 7).

Chronic Otitis Media

The diagnosis of chronic otitis media implies a permanent abnormality of the pars tensa or flaccida. In most patients, the initiating factors are unknown but are likely to include acute otitis media, OME and surgery, especially grommet insertion.

There is usually little difficulty in diagnosing chronic otitis media once the external auditory canal has been cleansed of debris and any associated otitis externa allowed to settle. This can take several visits. There is usually no need to give the patient an anaesthetic to clean out the ear to assess the pathology, except perhaps in some children. There are several classifications of chronic otitis media but, from the point of view of management, most would agree that three decisions have to be made:

- Are the pars tensa and flaccida intact?
- Is the disease active or inactive?
- If active, is the inflammation confined to the mucosa or is there squamous epithelium involved as well? In other words, is there a cholesteatoma?

Once the above decisions have been made, the chronic otitis media can be classified as being one of the following types.

Healed Chronic Otitis Media (Figure 2.6)

If the tympanic membrane and pars flaccida are intact and in their normal positions, the pathology can be considered healed and unlikely to become active. However, there may be a conduction defect due to ossicular chain disruption or tympanosclerosis of the tympanic membrane or ossicles.

The best way of confirming that the tympanic membrane is intact is by pneumatic otoscopy when it will be seen to be mobile. An alternative is to perform tympanometry. When a defect is present, the most usual result is a flat tympanogram (type B) with a high volume which represents the combined volume of the canal and the middle ear space. The less frequent alternative is that the Eustachian tube allows the air to escape and a seal cannot be obtained (*see* **Chapter 7**).

Inactive Chronic Otitis Media (Figure 2.7)

Here, there is either a permanent perforation of the pars tensa or a retracted area in the middle ear or attic. At the time of examination, there is no evidence of inflammation ('activity').

In such an ear, there are two main concerns, namely any associated hearing impairment and the possibility of future activity. The likelihood of the latter occurring in an ear that is inactive is unknown. It is usually considered that, provided drainage of mucus from the middle ear and mastoid spaces past the ossicles to the Eustachian tube is occurring and there is free exit of epithelial

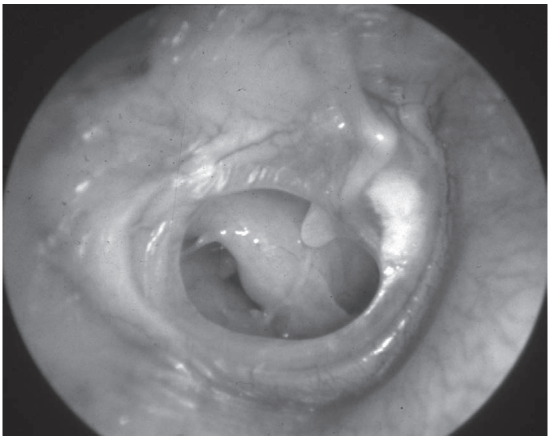

Figure 2.7 Inactive chronic otitis media.

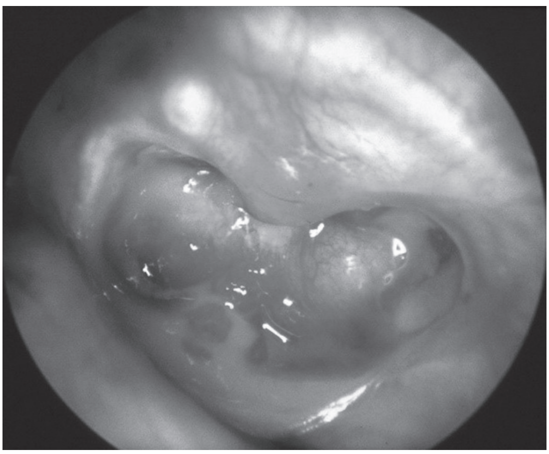

Figure 2.8 Active mucosal chronic otitis media.

debris from any retracted area, future activity is unlikely. Although advice is often given to avoid water getting into the ears, there is no evidence that this increases the likelihood of activity developing.

Active Chronic Otitis Media (Figure 2.8)

An ear is active if there is pooling of secretions, either mucopus or mucus, in the middle ear or external auditory canal. Provided that all debris is removed from the ear and every part examined, there should be little difficulty in deciding if an ear is active or not. The inexperienced often neglect what they think is a piece of wax postero-superiorly in the attic region. This is often not wax but dried pus overlying active disease. As such, it is important to remove it to fully examine the pars flaccida.

Table 2.7 Relative risks of complications occurring in ears with cholesteatoma compared to those with active mucosal disease

Complication	Cholesteatoma versus mucosal disease
Conductive impairment	No difference[1]
Sensorineural impairment	No difference[1]
Labyrinthitis	Not known
Facial nerve palsy	Mucosal commoner[2]
Meningitis	Mucosal commoner[2]
Intracranial abscess	No difference[2,3]
Lateral sinus thrombosis	No difference[2]

[1] Glasgow Royal Infirmary data (personal communication, based on authors own data); [2] Singh and Maharaj (1993) (8); [3] Nunez and Browning (9).

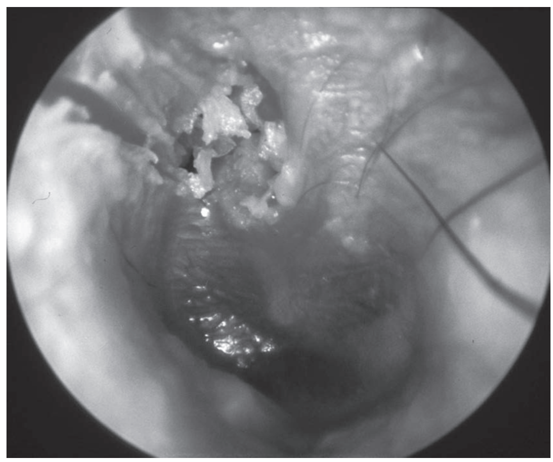

Figure 2.9 Active squamous chronic otitis media.

It is not necessary that the patient has noticed an aural discharge for an ear to be active. It all depends on the volume of the secretions and whether they can drain down the Eustachian tube. Consequently, the length of time a patient has had a discharging ear is not a reliable indicator of the duration of activity.

There are two main variants of activity, the first being where the inflammation is primarily of the mucosa of the middle ear, antrum and mastoid air cells. This type of disease can be referred to as active mucosal disease. The second type is where, in addition to the mucosal inflammation, there is a retracted pocket of squamous epithelium filled with epithelial debris; that is a cholesteatoma **(Figure 2.9)**.

Many otologists would consider it is particularly important to identify a cholesteatoma because it would be more likely to be associated with life-threatening complications, such as an intracranial abscess. In the past, it was common for an ear with mucosal disease to be called 'safe' and one with cholesteatoma to be called 'unsafe'. The relative chances of complications occurring in mucosal disease as opposed to ears with a cholesteatoma have not been defined, mainly because of the difficulty of assessing the incidence of the two main types of active chronic otitis media in the community. What can be done is to analyse the incidence of active squamous disease against active mucosal disease in patients that present clinically with complications (**Table 2.7**).

Complications can be categorised in different ways but, for clinical practice, it is better to think of them in terms of likelihood of occurrence. This is particularly helpful when considering management. A conductive hearing impairment is the commonest complication. Labyrinthitis and sensorineural impairment are not uncommon whilst facial nerve palsy, meningitis, intracranial abscess and lateral sinus thrombosis are thankfully rare. This order of rank is based upon clinical reports rather than hard scientific evidence. The only incidence data are on intracranial abscesses associated with active chronic otitis media. In the UK, the chances of active chronic otitis media causing an intracranial abscess are 1 in 10 000 per year (10). Taken over a lifetime the chances are greater, so that a patient aged 20 has a 1 in 167 chance over the next 60 years of developing an intracranial abscess.

Table 2.7 lists the complications and the relative likelihood of them occurring in cholesteatomatous ears against those with mucosal disease, as evaluated from the literature. As can be seen, there is no evidence that non-cholesteatomatous ears are 'safe'. Surgery is not without its risks and indeed can result in the same complications as the disease process itself. Again, the relative risks, especially in the hands of the average surgeon, are not known, which makes it extremely difficult to assess the advantages of surgery in preventing complications.

It would be helpful if radiology could assess the extent and type of activity. Radiology can identify anatomical defects in the temporal bone provided they are big enough and the correct view has been taken. For example, it is possible to identify defects in the attic but radiology cannot determine the cause. Such defects can be caused by mucosal disease, cholesteatoma or surgery. The ability of mucosal disease to cause bone erosion cannot be doubted because ossicular chain disruption is so frequently associated with it. Whilst CT can identify bony anatomy and bony erosion in detail, non-EP diffusion-weighted MRI is required to differentiate between cholesteatoma and mucosal disease and can identify cholesteatoma pearls 2 mm or larger (10). Imaging can ascertain whether the mastoid air cell system is sclerotic but this will almost invariably be the case if the disease process has been present for any length of time.

Otosclerosis

Otosclerosis is a diagnosis made by exclusion of other common causes of a conductive defect. When the tympanic membrane is normal in appearance on otoscopy with no suggestion of middle ear fluid, the only alternative diagnoses to be considered are malleus head fixation, congenital middle ear abnormalities and traumatic ossicular chain discontinuity. In comparison to otosclerosis these are uncommon, but it would be valuable to be able to diagnose otosclerosis apart from using surgical exploration of the middle ear. In some ears with otosclerosis, a pink (flamingo) flush may be seen through the tympanic membrane but this is a relatively uncommon finding and is subject to considerable observer error. Tympanometry is unhelpful in making this distinction (as discussed in detail in **Chapter 7**).

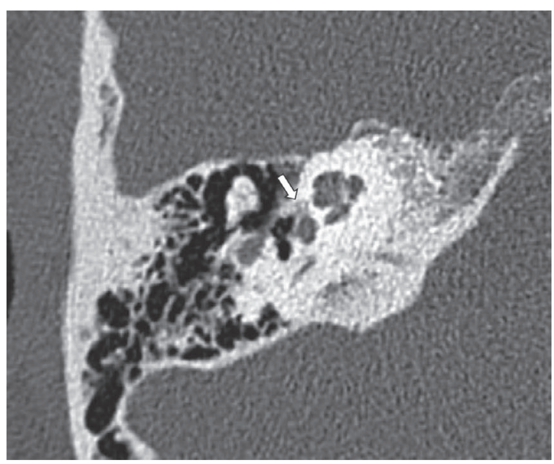

Figure 2.10 High resolution CT scan, with a focus of antefenestral otospongiosis marked with an arrow, in a patient who had subsequent successful stapes surgery.

A clinical diagnosis of otosclerosis can be confirmed by high-resolution CT scan (11) or cone beam CT. The classic finding described by radiologists as 'antefenestral otospongiosis' (**Figure 2.10**) represents a focus of demineralised bone which has rendered the stapes footplate immobile. It should be noted that otosclerotic stapes fixation can exist in the presence of a normal CT scan, and exploratory tympanotomy may be required to confirm the diagnosis. Equally, otospongiotic foci can be identified on imaging in the presence of normal hearing, for example, there is often bilateral radiological otosclerosis on CT scans of patients with a unilateral conductive hearing loss. Radiological investigations should therefore be used judiciously in combination with the wider clinical picture as it has a 20% false negative as well as a 10% false-positive rate (11).

Ossicular Chain Disruption

Disruption of the ossicular chain following trauma is relatively uncommon and most individuals will clearly relate their hearing impairment to the head injury. The tympanic membrane will be normal so, in the presence of a conductive hearing impairment, the main alternative diagnosis is otosclerosis. Unfortunately, tympanometry is of no value in making this diagnosis because of the reasons discussed above (and discussed in detail in **Chapter 7**).

Discontinuity of the ossicular chain can be identified on high-resolution CT imaging or cone beam CT imaging, which can identify subtle defects of the ossicles, their positions and articulations. This can be useful to identify the cause of the conductive hearing loss, and to ascertain the risks and likely outcomes of surgery.

Patients with conductive hearing loss due to an ossicular chain which is not transmitting sound effectively due to otosclerosis or ossicular discontinuity, have several options. They can do nothing; trial an air-conduction hearing aid; trial a bone-conduction hearing aid with a view to considering an implanted bone-conduction device in future, or they can proceed to definitive surgery. It is generally accepted practice that a conservative approach is adopted in the first instance, prior to proceeding to surgery which carries significant risks and cannot be guaranteed to be successful.

CLINICAL ASSESSMENT OF HEARING THRESHOLDS IN ADULTS AND CHILDREN

Air-Conduction Masking

In clinical testing of hearing, it is important to mask out hearing in the non-test ear. The rules and methods involved are different from those in audiometry. When using tuning forks to identify the better hearing ear with the Weber test and to clarify whether an ear has a conductive defect with the Rinne test, air-conduction masking is important but seldom carried out.

Sounds arriving by air at the ear on one side will also be heard by the ear on the other side (*see* **Figure 2.2**). However, the head-shadow will attenuate the sound by an amount depending on its frequency (*see* **Table 2.2**). As the minimal attenuation of the four-frequency average over 0.5, 1, 2 and 4 kHz is 4 dB HL, the rule should be that the non-test ear should always be masked when clinically testing by air conduction. This rule does not apply in audiometry (*see* **Chapter 3**).

Methods of Air-Conduction Masking

Unfortunately, no single method can produce the required range of masking sound levels that might be required. Correspondingly, no one method is suitable for all situations (**Table 2.8**). For example, there will be occasions such as when tragal rubbing provides insufficient masking. On the other hand, in most circumstances, a Barany box will produce too much masking and will mask the test ear as well as the non-test ear (*see* **Figure 2.13**). There are three commonly used methods.

Table 2.8 Appropriate method of masking the non-test ear when voice testing

Whispered voice	At 2 feet / 60 cm	Tragal rubbing
Whispered voice	At 6 inches / 15 cm	Tragal rubbing
Conversational voice	At 2 feet / 60 cm	Tragal rubbing
Conversational voice	At 6 inches /15 cm	Tragal rubbing
Loud voice		Barany box

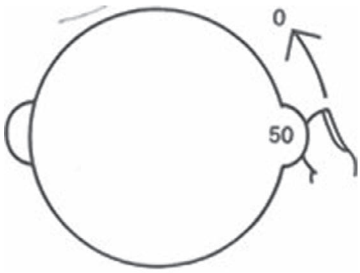

Figure 2.11 Effective masking level (dB) of tragal rubbing.

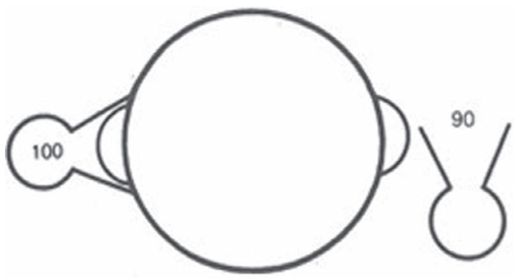

Figure 2.12 Effective masking level (dB) of a Barany box held over (100 dB A) and held at right angles to the ear (90 dB A).

Tragal Rubbing

Occluding the external auditory canal by putting a finger on the tragus only attenuates sounds by ~10 dB and so is of no value. On the other hand, if the tragus is rubbed at the same time (**Figure 2.11**), speech will be attenuated by ~50 dB.

The advantage of tragal rubbing over other methods is that the masking sound is produced within the external auditory canal and there is no risk of the sound crossing over and masking the test ear. However, there is a danger of under-masking if the test speech is louder than 60/70 dB A. This means that a Barany box will be necessary when using a loud voice in free-field speech testing.

Barany Box

For many years, a Barany noise-box was the standard method of all clinical masking. These produce a broad-band noise although in some boxes there can be marked dips in the frequency spectrum and the noise can be irregular. The maximum sound output varies from box to box, but a lower limit of 90 dB A can be assumed when a

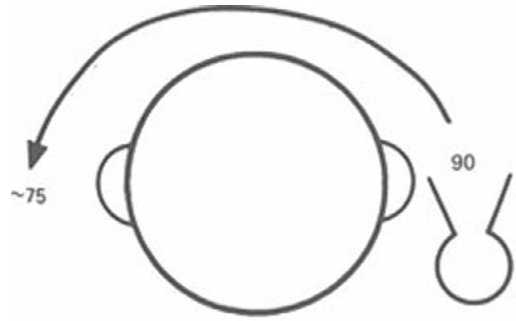

Figure 2.13 Mean potential level (dB) of cross-masking that can occur with a Barany box.

box is held at right angles to the ear and 100 dB A when held over the ear (**Figure 2.12**). These levels are sufficient to mask in all practical circumstances but there is a danger that because the sound can travel round the skull, the test ear will also be masked (**Figure 2.13**). Eighty per cent of normal hearing ears will be masked by a Barany box on the other ear as evidenced by the test ears inability detect a whispered voice at 60 cm (12). Because of this, a Barany box should only be used in free-field speech testing when testing an ear with a potentially severe or profound impairment, i.e. one that cannot hear a conversational voice at 15 cm (see below).

FREE-FIELD VOICE TESTS OF HEARING

It is remarkable how often clinicians omit to assess a patient's hearing by free-field voice tests. Assessments more often done are the Rinne and Weber tuning fork tests that aim to determine whether an impairment is conductive or sensorineural. How tuning fork tests can be interpreted in the absence of knowledge of the degree of impairment in each ear is a mystery. Free-field voice testing takes only 1–2 minutes and, though not as reliable as pure tone or speech audiometry, is of value for the following reasons.

Firstly, audiometry may either not be available or not necessary. When audiometry is not available to assess the air-conduction thresholds, such as in primary care or in a developing country, free-field voice testing using a whispered voice can be used as a screening tool in both adults and children (12, 13). It will reliably detect whether there is a hearing impairment in one or both ears. The decision then is whether an audiogram would give additional guidance on management. That very much depends on the otoscopic findings and the age of the patient. Having an audiogram with air-conduction thresholds does not aid the fitting of a hearing aid. Having an audiogram with bone-conduction thresholds is necessary to determine the magnitude of the air–bone gap in those with otoscopic evidence of ear pathology. Evaluation of bone-conduction thresholds is not generally available outside of a specialist audiology department. Even if audiometry is available, to obtain valid results requires a trained tester working with calibrated instruments in a sound deadened area using techniques that take into account the subject's age.

Secondly, it can be used as a check on audiometry. Following otoscopy and free-field voice testing, it should be possible to predict the type and severity of any impairment in each ear. Comparison can then be made with the audiogram and, where discrepancies arise, one or both must be invalid. Motivation of the subject can be very different between subjects.

Method

Free-field voice testing is primarily a method of detecting whether a hearing impairment is present in one or both ears and, when this is the case, to determine its severity. This is achieved by assessing the threshold for understanding speech by varying the vocal effort and the distance from the ear to produce a range of speech sound levels.

It is first explained to the patient that they are expected to repeat back, as accurately as possible, what words they hear being said by the examiner. The examiner then stands behind the patient to obviate visual speech reading and says a test word loudly enough to ensure that the task is understood.

Thereafter, combinations of numbers and letters (5 B 3) are frequently used as this gives a high variety of combinations. Bi-syllable words (e.g. cowboy, football) are often used for children. It is normal to test the better hearing ear first if there is one. The non-test ear is then masked by tragal rubbing. The patient's free-field threshold is the voice and distance level at which they get more than 50% correct. It is usual to assess the thresholds by gradually increasing the relative loudness of the voice, so that the examiner starts using a whispered voice 2 feet (60 cm) away from the patient, which is the furthest away he can reach when masking the non-test ear.

Provided a whisper is used, and this is best achieved by exhaling first, a patient who does not hear a whispered voice at arm's length is hearing impaired in that ear, that is, would benefit from amplification (pure tone average [PTA] ≥ 25 dB HL). If the patient can repeat this back this does not actually mean that the hearing is 'normal', as a normal hearing ear in a teenager will hear a whisper at least 12 feet (4 m) away. Rather it means that the likely associated disability is insufficient to merit management.

If the patient fails to hear a whispered voice at 2 feet (60 cm) the relative sound level is increased in steps to a whispered voice at 6 inches (15 cm), to a conversational voice at 2 feet (60 cm), to a conversational voice at 6 inches (15 cm), to a loud voice at 2 feet (60 cm) and finally to a loud voice at 6 inches (15 cm). For each new presentation, the number/letter combinations should be changed to avoid the patient recognising them from previous presentations. The test is terminated when a patient repeats 50% of the words correctly at any one voice and distance level. When using a loud voice, a Barany box must be used as tragal rubbing provides insufficient masking. If there is any doubt concerning a threshold, the examiner can test again at a lower voice level.

Clinical voice tests have been criticised because the sound level of speech varies between examiners and there is a considerable difference in the sound levels that are produced by an examiner on different occasions, particularly in whispering. To avoid this, whispering should be done after full expiration.

Despite the above criticisms, in the hands of experienced otologists, monaural, free-field speech can reliably screen individuals for a hearing impairment greater than 25 dB HL and can grade the severity of a hearing impairment into normal, mild/moderate and severe/profound.

Screening for Hearing Impairments Using a Whispered Voice

Adults and children with a speech frequency average greater than 30 dB HL will be unable to hear a whispered voice 2 feet (60 cm) from the test ear. In a systematic review of four adult studies (14), the sensitivity ('hit rate') of this is 90–100% and the

Table 2.9 Sensitivity and specificity of a hearing impairment being detected by an individual's inability to hear a whispered voice at 2 feet (60 cm)

Impairment	Percentage	
	Sensitivity	Specificity
PTA over 0.5, 1 and 2, 4 kHz		
≤ 25 dB HL	91	96
≤ 30 dB HL	96	91
≤ 35 dB HL	98	86

(After Browning et al. [15]).

false-positive rate was 70–87% when compared with audiometric assessment.

Hence, if an adult patient can hear a whispered voice 2 feet (60 cm) from his or her ear, the clinician can be fairly certain that the pure tone thresholds will be better than 30 dB HL and, in many instances, this will make an audiometric evaluation unnecessary (**Table 2.9**).

The sensitivity in four studies of the whispered voice in children ranged from 80% to 90% with the specificity being above 90% (14).

Grading the Severity of an Impairment

A comparison is made in **Table 2.10** of the free-field thresholds assessed by skilled testers, against the mean FFA average in 200 adult ears, along with the 5th and 95th percentiles (14). Though there is some overlap, patients can be divided into three groups by free-field voice testing 1) FFA less than 30 dB HL; 2) FFA 30–70 dB HL; and 3) FFA greater than 70 dB HL. This corresponds to normal, mild/moderate and severe/profound impairments, respectively.

Qualification

When comparing the results of voice testing with those of audiometry, the levels of skill and the test environment can vary considerably and repeat testing of one or both of these techniques should be carried out again.

TUNING FORK TESTS

Rinne Test

The Rinne test is the most frequently used tuning fork test, its stated role being to identify a conduction defect.

When an abnormality has been otoscopically detected in an ear, such as a perforation, there will be a conductive defect and a Rinne test has no potential value. In these cases, what is required is knowledge of the magnitude of the conductive hearing defect measured audiometrically by the air–bone gap.

In those with a normal tympanic membrane and a hearing impairment on free-field testing, the Rinne test has the potential to identify a conductive defect due to ossicular chain conditions such as otosclerosis.

CHOICE OF TUNING FORK

Forks of 512 Hz are usually preferred but there is evidence (15) that a 256 Hz fork is more accurate. There is the potential problem that forks with a

Table 2.10 Comparison of free-field voice thresholds and pure tone average (PTA) over 0.5, 1, 2 and 4 kHz

Voice level	Distance	PTA Mean (dB HL)	Percentiles	
			5th	95th
Whisper	2 feet / 60cm	12	–	27
	6 in / 15 cm	34	20	47
Conversation	2 feet / 60 cm	48	38	60
	6 in / 15 cm	56	48	67
Loud	2 feet / 60 cm	76	67	87

(After Swan and Browning [12]).

frequency lower than 256 Hz can make it difficult for a patient to distinguish between hearing the sound and feeling it by vibration. It is difficult to sufficiently activate forks with a frequency higher than 512 Hz for them to be heard by those with a moderate or severe impairment.

The correct method of performing the Rinne test is well described elsewhere but some practical points are worth making. Activating a 512 Hz fork by compressing the tines between the fingers produces a sound level of around 70 dB SPL whereas hitting it against a knee or elbow, without causing pain, produces a sound level of around 90 dB SPL.

Tuning forks should be as heavy as possible as the sound level produced is more sustained. Though forks vary, a decay of 10 dB HL every 10 seconds is the slowest that can be anticipated. The sound level produced by light forks decays rapidly which is a disadvantage as there is inevitably a time delay between asking the patient to compare the loudness by air and bone conduction.

When testing air conduction, the tines of the fork should be held directly in line with the external auditory canal, as holding it at an angle diminishes the sound level.

In testing bone conduction, it is important to make good contact with the skull. This is not achievable on the mastoid tip, the best position being on the flat bone just superior and posterior to the external canal. It is also important that firm pressure is used because the sound level can vary by as much as 15 dB with different degrees of pressure. As the patient's head tends to move away from the tuning fork, it is best to hold the head steady on the contralateral side using the examiner's free hand.

The patient can be asked to make a comparison between the relative loudness of the air and the bone conduction. Alternatively, the sound in one of the test modes can be allowed to decay until it is no longer heard and the patient is then asked if they can hear it by the other mode. In general, loudness comparison techniques identify smaller air-bone gaps than decay methods (16,17). By convention, the results are reported as Rinne positive when the air conduction is louder than the bone conduction and Rinne negative when the bone conduction is louder than the air conduction. Many, including the author, find it difficult to remember which is which, so reporting the results as to which route is louder bypasses this problem. This can be done with symbols such as BC > AC and AC > BC.

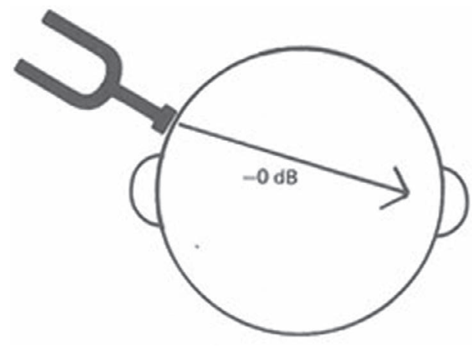

Figure 2.14 For practical purposes, the clinician should use the concept that the bone conduction from an activated tuning fork placed on one side of the head will be heard just as loud on the other side.

Bone-Conduction Masking

It is theoretically important to mask the non-test ear whenever the bone conduction is being tested, particularly when testing the poorer hearing ear. This is because the bone conduction on the poorer hearing side may be heard by the other, better ear and give a false-negative BC > AC Rinne (**Figure 2.14**). The only way to mask the bone conduction is by making a sound at the external auditory meatus in the non-test ear. If there is a hearing impairment in the non-test ear, tragal rubbing and even a Barany box may be insufficiently loud. If a Barany box was used routinely there would also be a considerable danger of masking the bone conduction in the test ear. Knowledge of the degree of impairment by free-field voice testing helps decide which method to use, but even then there is often some uncertainty. Because of this, most otologists omit to mask tuning fork tests unless there is gross asymmetry in the hearing.

VALIDITY

It is generally held that the Rinne test will reliably detect a conductive defect of 20 dB or greater. This is not so! This is the level at which 50% of patients with an air–bone gap of 20 dB will be AC > BC (Rinne positive) and 50% will be BC > AC (Rinne negative) (16–18).

The value of the Rinne test is best assessed on data which has analysed the proportion of ears with different sizes of air–bone gap which are Rinne positive and negative. The false-negative and false-positive diagnosis rates can then be calculated for each size of air–bone gap. An example

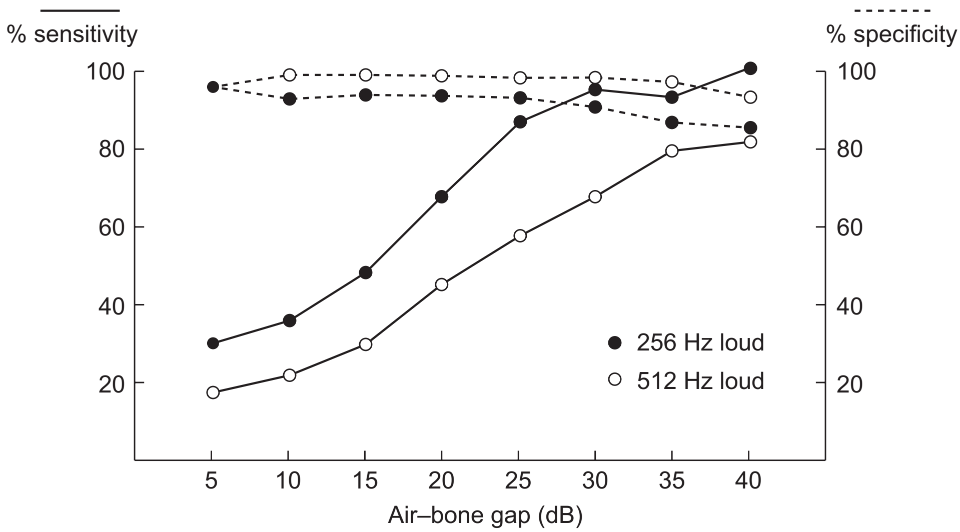

Figure 2.15 Sensitivity and specificity of 256 Hz (•) and 512 Hz (o) tuning forks in detecting differences in air and bone conduction of various magnitudes using loudness comparison in Rinne test (after Browning et al. [15]).

of such an analysis is shown in **Figure 2.15**. From this and other studies it is possible to calculate (**Table 2.11**) (18–22) the size of air–bone gap that will be correctly identified 50, 75 and 90% of the time, the first value being included as this is the size of air–bone gap that would be correctly detected at the same level by tossing a coin. Though each of the studies can be criticised in different ways, the overall conclusion is inescapable: the Rinne test will not reliably detect (i.e. on 90% of occasions) a conduction defect unless there is an air–bone gap of at least 30–40 dB.

An alternative and more encouraging way to look at the specificity data, is that if the bone conduction is louder than the air conduction then there is likely to be an air–bone gap of 10 dB or more. This is different from saying that the Rinne test will detect a conductive defect, as it is only when the air–bone gap is greater than 40 dB that it will be detected on 90% of occasions. The reason why practising otologists have come to believe that the Rinne test is more reliable than this is that when the bone conduction is louder than the air conduction (Rinne negative) there is usually an air–bone gap. The above figures support this but what the otologist tends to forget is the considerable number of occasions when there is an air–bone gap and a Rinne negative BC > AC response is not obtained.

Table 2.11 Magnitude of air–bone gap (dB) which would be correctly identified by Rinne test on various percentages of occasions

	Confidence limits		
	50%	75%	90% +
256 Hz fork			
Crowley and Kaufman (18)	25		30
Gelfand (19)		40	
512 Hz fork			
Crowley and Kaufman (18)	25		30
Wilson and Woods (21)			40
Gelfand (19)		40	
Golabek and Stephens (16)	19		
Browning and Swan (17)	20		45

Clinical Role

What the otologist needs to know in a patient with a hearing impairment is whether there is a conduction defect and, if so, what is its magnitude? The latter question can only be determined by PTA with masking. The Rinne test does not help determine the magnitude of an air–bone gap, it is only an aid to determining whether there is a conduction defect. In patients with otoscopic evidence of middle ear pathology, such as chronic otitis media, by definition there must be a conduction defect and the Rinne test adds nothing. However, when the tympanic membrane is normal and the bone conduction is louder than the air conduction (Rinne negative) there is most likely a conduction defect. In adults, this would suggest otosclerosis. If the air conduction is louder than bone conduction (Rinne positive) in these circumstances there could still be a conductive defect due to otosclerosis of a magnitude that could benefit from surgery.

Weber Test

Why this test is so popular is uncertain. One of the main problems with this test is that the response is not reproducible, as can be verified by retesting a patient. Different results are frequently obtained if the base of the fork is positioned on the nasion or on the upper lip rather than on the vertex.

Most publications would agree that the results of the Weber test are difficult to interpret when there is a bilateral hearing impairment. In 30% of such patients, the test will be referred to the midline so the result cannot be interpreted as being correct or incorrect. Of the 70% who do refer the test to one ear, around 25% refer it to the incorrect ear (23). It is concluded that the Weber test is likely to add little to the assessment in the majority of patients.

Other Tuning Fork Tests

Many other tests have been described and are well summarised by Hinchcliffe (1981) but are infrequently used (24). This is mainly because these tests were developed before audiometric testing was possible. Now that accurate audiometry is almost universally available, they are of historical interest only. The Weber test in the author's opinion comes in the historical interest category.

CONCLUSIONS ON OTOSCOPY

- Otoscopy will diagnose most conditions of the external auditory canal and middle ear. The main exception to this is otosclerosis.
- Wax or debris frequently needs to be removed to fully assess middle ear pathology particularly if there has been previous ear surgery.
- Wax is infrequently a cause of a hearing impairment unless it is impacted deep in the canal affecting the vibration of the tympanic membrane.
- Otitis externa will be associated with a mild hearing loss at the very most.
- Chronic otitis media is primarily a condition of adults, affecting about 6% of the UK adult population. At any one time, the ear may be active or inactive and the condition may be pathologically associated with mucosal or squamous disease. The conductive hearing loss in such ears will be created by defects in the tympanic membrane and erosion or fixation of the ossicular chain.
- Otosclerosis is the presumptive diagnosis in those with a normal tympanic membrane associated with a conductive hearing defect. It affects about 2% of adults. Tympanometry is of no value in confirming this diagnosis.

CONCLUSIONS ON CLINICAL ASSESSMENT OF HEARING

- Clinical assessment of a patient's hearing by free-field speech can be helpful in many ways. It is wise to have the severity of an impairment clinically assessed as audiometry can on occasions be inaccurate or not available.
- Experience and a quiet environment improve its validity in both adults and children.
- In many instances, the degree of accuracy that audiometry gives may be unnecessary, for example, in screening the elderly for hearing aid provision. Exaggerated thresholds may be missed if suspicion is not aroused by clinical testing. In addition, it considerably aids the interpretation of tuning fork tests if these are carried out.
- Masking is as important in clinical testing as it is in audiometric testing.
- Tragal rubbing is the easiest and most appropriate method of masking to use routinely as it requires no instruments and there is no risk of over-masking.
- A Barany box can potentially over-mask the test ear. In free-field voice testing it should only

be used when there is a unilateral profound loss of hearing and in tuning fork testing when a bilateral conductive impairment is likely.

- If a patient can hear a whispered voice at a distance of 2 feet (60 cm), his or her pure tone average threshold is likely to be better than 30 dB HL.
- Free-field speech testing in adults by skilled testers, can divide the severity of a hearing impairment into three bands: normal; mild/moderate; severe/profound. This is sufficiently accurate for many purposes.
- The Rinne test is less reliable in detecting a conductive defect than we would like to believe. Though the cross-over point from Rinne positive to negative is an air–bone gap of ~20 dB, this means that 50% of individuals with a 20 dB gap of will be Rinne positive (air conduction louder than bone conduction) and 50% will be Rinne negative (bone conduction louder than air conduction).
- The Rinne test will not reliably detect (90% confidence) an air–bone gap until it is 40 dB or greater.
- On the other hand, if the bone conduction is louder than the air conduction (Rinne negative) there will be an air–bone gap of 10 dB or greater. However, a considerable proportion of ears with clinically important air–bone gaps will not give this result.
- The Weber test adds little to the assessment of the hearing because of its non-repeatability and error rate compared with quality pure-tone audiometry with appropriate masking.
- In the detection and quantification of a conductive defect, reliance will have to be primarily on pure tone audiometry rather than on tuning fork tests.

FURTHER READING

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3

Pure tone audiometry

MANUAL DIAGNOSTIC AUDIOMETRY

Pure tone audiometry is the basic audiometric method of clinically assessing the thresholds for air and bone conduction for each ear, over a range of pure tones. These thresholds are obtained by a trained individual with the subject in a sound-proofed environment such as a booth. The audiometric electric equipment, headsets and bone-conduction vibrators are regularly calibrated. The air and bone thresholds are obtained following a set technique (BSA Recommended procedure pure tone audiometry) (1) and are then charted as an audiogram preferably on a 'Recommended format for audiogram form (1918) from the British Society Association or a similar version (**Figure 3.1**). This can then be used: (a) to assess the degree of impairment in each ear; and, if one is present, (b) to ascertain whether it is conductive, sensorineural or mixed in type. To do the latter often requires masking of the non-test ear, which needs a skilled individual to recognise when it is required and then carry it out appropriately.

One aspect that is, unfortunately, not recorded on this form is where the testing was carried out regarding the background noise level (see 'Test Environment').

AUTOMATED SCREENING OR MONITORING AUDIOMETRY

As trained audiometricians with appropriate test facilities are not universally available, automated approaches have been developed, often based on universally available smart phones or laptops.

When calibrated and used in a soundproof booth, automatically obtained air-conduction thresholds are not materially different from manual assessments (2). This makes them potentially useful as screening tools in a sound-deadened environment to identify adults with hearing impairment and also for basic hearing aid fitting. Without being able to assess bone-conduction thresholds, automated procedures cannot determine whether an impairment is sensorineural or conductive. This means that for children they are only really useful for triaging referrals to specialist centres.

Inevitably, with time, automated audiometry will develop so that bone-conduction thresholds can be accurately assessed, with relevant masking. The placement of headsets and bone-conduction vibrators can be done by minimally trained assistants.

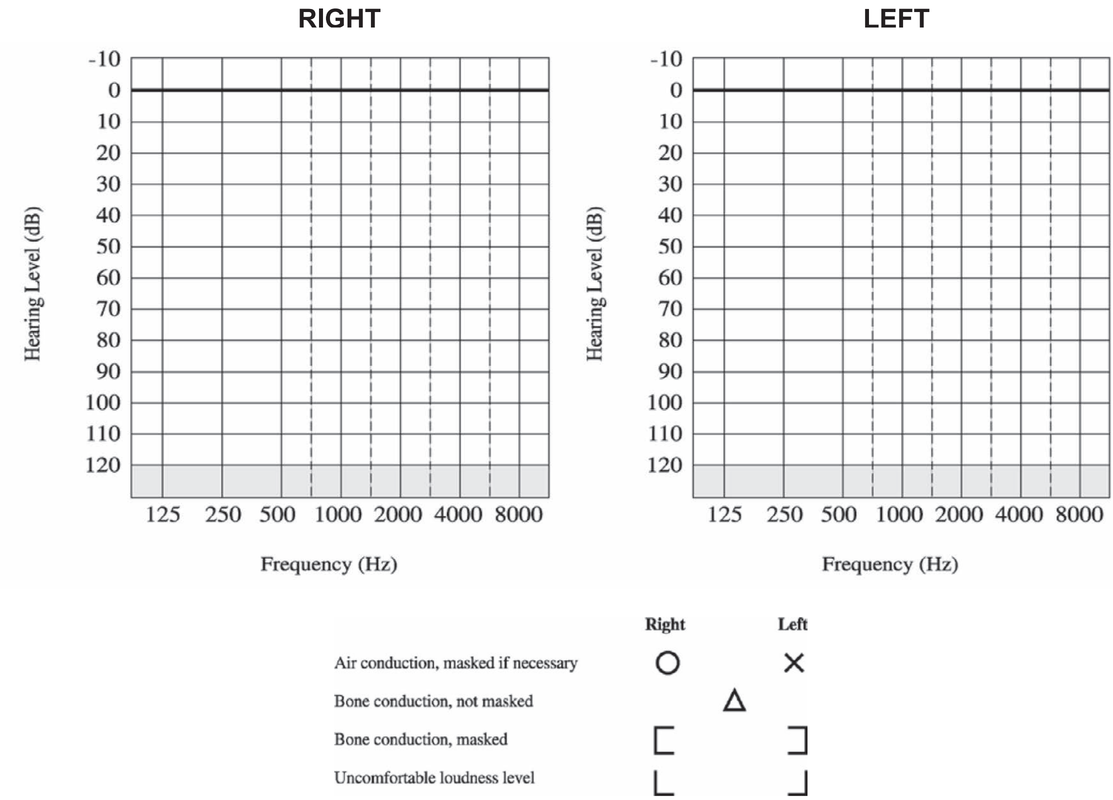
DIAGNOSTIC PURE TONE AUDIOGRAM CHART

The recommended format for an audiogram form from the British Society of Audiology (*see* **Figure 3.1**) has the test frequencies on the horizontal axis and the thresholds of hearing on the vertical axis. The test frequencies are at octave intervals from 0.25 to 8 kHz (250–8000 Hz). Additional frequencies can also be tested, with 3 and 6 kHz being the commonest when assessing sensorineural impairments. The vertical axis is in decibels hearing level (dB HL) with a range from –10 to 120 dB HL. The latter is the loudest sound level that can be delivered over headphones to test the air-conduction thresholds with most audiometers. Some audiometers, particularly portable ones, are less powerful.

DOI: 10.1201/b23377-3

Name: **Date:**

Date of birth: **Case No:**



Audiometer type & serial number:____

Earphone type:____

Date of last objective calibration:____

Tester:_________________________________Signature:____________________

Comments:____

Figure 3.1 Standard BSA pure tone audiogram template.

Figure 3.2 shows the level of detection of sounds in decibels of sound pressure level (dB SPL) of the human ear. In this SPL decibel scale, there is an equal amount of energy (in dynes/cm^2) at each frequency. Because of the U-shaped nature of the SPL audiogram, it was decided that the use of a dB SPL scale in a clinical audiogram would make abnormalities difficult to identify especially over the speech frequencies of 0.5, 1, 2 and 4 kHz. Hence, a biological scale of human hearing was developed, so that 0 dB HL would be the expected threshold of detection of a pure tone air and bone threshold, irrespective of its frequency. The amount of energy at 0 dB HL at each frequency is therefore not the same, the 'normal' levels being obtained by testing 'otologically normal', young adults. Unfortunately, the 'normal' levels were not the same in young adults in every country. The differences between countries were resolved by the International Standards Organisation in 1964 (5) and their standards are now universally used. So in the dB

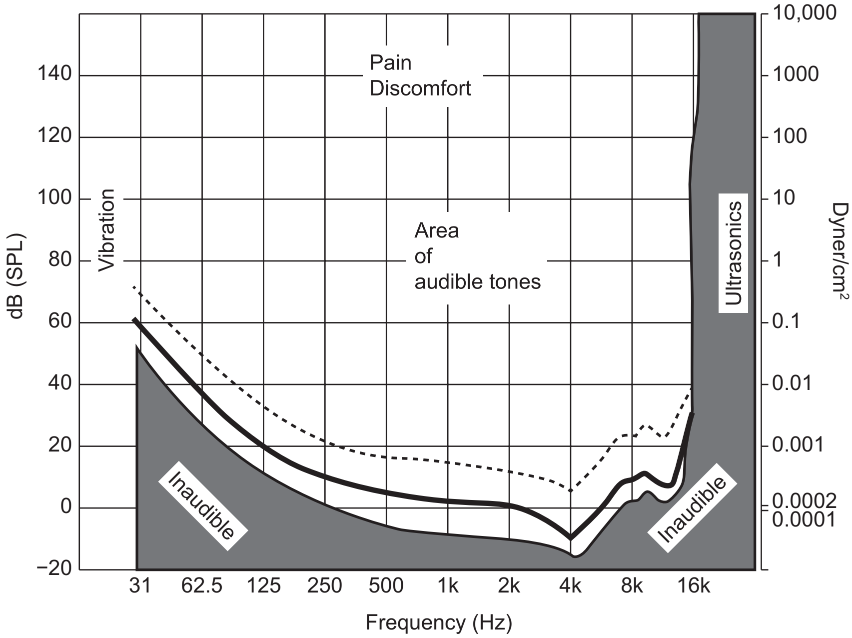


Figure 3.2 Normal sound pressure levels (dB SP) of detection of pure tones in humans. Solid line = median levels; broken line = upper 90th percentile (after Robinson and Dadson [3]).

HL scale 'normal' hearing adults would be expected to have a flat audiogram, with the mean level being 0 dB HL over any combination of frequencies with the not-masked bone conduction being no different from the air conduction (**Figure 3.3**).

Portable instruments are used to measure noise levels in industrial situations, but they, like the ear, do not measure as efficiently at some frequencies as others. Over the years, several weighted dB noise scales have been used for these instruments but the current one is dB A.

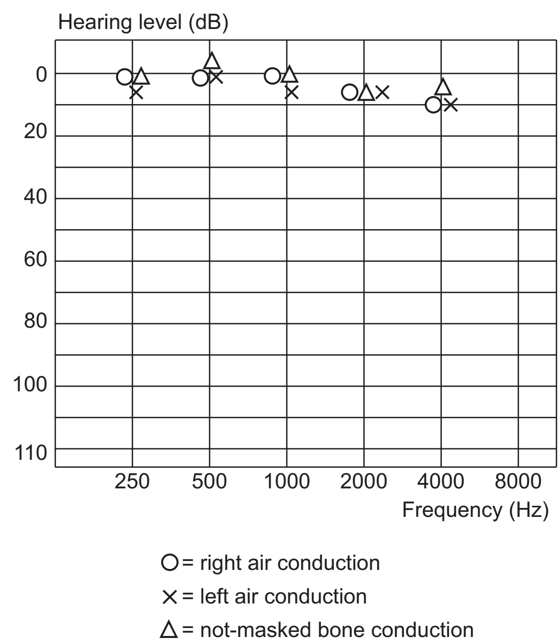


Figure 3.3 Normal hearing in both ears, i.e. the air-conduction thresholds are less than 25 dB HL. The not-masked bone-conduction thresholds are less than 10 dB different from any of the air-conduction thresholds. There is thus no potential air–bone gap and no need to mask the bone conduction.

RECOMMENDED MANUAL DIAGNOSTIC TECHNIQUE

Several ways of manually assessing the air- and bone-conduction thresholds but the British Society of Audiology's recommended procedures (**Table 3.1**) would be acceptable in most departments, both in and outside the UK. Many aspects require some comment.

Test Environment

Without spending a considerable amount of money, it is not possible to exclude all extraneous noise

Table 3.1 BSA 2018 recommended procedure pure tone audiometry

Method of finding air- and bone-conduction thresholds
1. Following a satisfactory positive response, reduce the level of the tone in 10-dB steps until no further response occurs.
2. Increase the level of the tone in 5-dB steps until a response occurs.
3. After the first response using an ascending approach, decrease the level by 10 dB and begin another ascending 5-dB series until the subject responds again.
4. Continue to decrease the level by 10 dB and increase by 5 dB until the subject responds at the same level on two out of two, three or four (i.e. 50 % or more) responses on the ascent. This is the hearing threshold level. Threshold is defined as the lowest level at which responses occur in at least half of a series of ascending trials with a minimum of two responses required at that level.
5. Proceed to the next frequency, starting at a clearly audible level (e.g. 30 dB above the adjacent threshold, but see notes on familiarisation in Section 6.6) and use the 10-dB-down, 5-dB-up sequence described in Step 4 until the threshold criterion is satisfied.

from the test environment. The sound level in a 'quiet' room is seldom less than 30 dB A and often 40–50 dB A. Background noise acts as a masking noise and can appreciably influence the results in normal individuals, especially bone conduction at low frequencies. The commonest cause of a low-frequency hearing loss is thus insufficient exclusion of background noise from the test environment (**Figure 3.4**).

Background noise can be lessened by several means:

- sound attenuating headphones
- sound deadening of the test room
- use of an acoustic booth.

Calibration

The sound level output of both the air- and bone-conduction modes of an audiometer can change with time. This means that an audiometer needs to be calibrated and adjusted regularly, preferably at least every 6 months. A recommended daily practice of setting up of the equipment is also suggested.

Threshold Determination

The term threshold means the minimum sound level at which a pure-tone signal can be detected. However, these can differ depending on the techniques being used to determine them. Firstly, the thresholds obtained will depend on whether they are assessed by ascending from the inaudible until they become audible or descending from the audible until they become inaudible. The recommended procedure is a tracking one, ascending and descending about the threshold until positive responses have been made on 50% or more of the occasions, the signal has been presented at a specific dB HL. Secondly, it is important to instruct the patient to respond, 'at the least suggestion of a signal' rather than 'when the signal is definitely heard'. If the patient responds in the second manner, the threshold can be 5–15 dB greater.

Reproducibility of Air-Conduction Thresholds

In testing an air-conduction threshold, the test/retest variation is no greater than ± 5 dB HL. So, when averaging, say over four frequencies, any change in the pure tone average of 5 dB HL or greater between two test occasions is significant. This is provided both test machines have been calibrated and the test situation is similar.

Validity of Air-Conduction Thresholds

In the majority of ears, the air-conduction thresholds do not require masking. The few that do are discussed below.

It is from an average of the air-conduction thresholds of the 'speech frequencies' of 0.5, 1, 2 and 4 kHz that one can predict the likely degree of disability the patient is likely to have in communication with others. The World Health Organisation (2014) has recommended a classification system

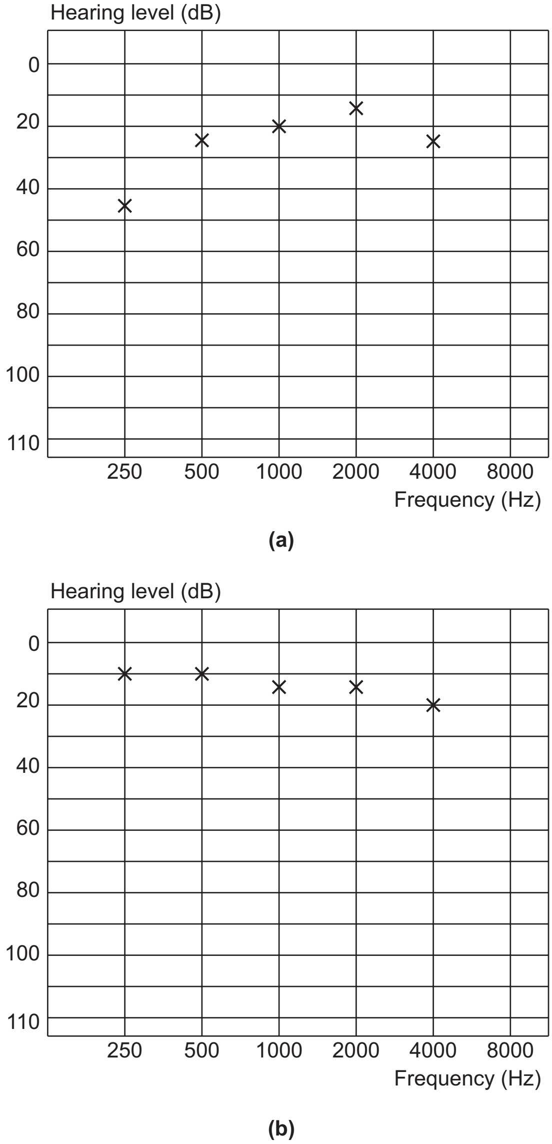


Figure 3.4 Effect of noisy test situations on hearing in normal individuals. **(a)** Upper 95th percentile of hearing in a group of patients tested in a noisy casualty ward; **(b)** upper 95th percentile of hearing in these same patients when tested in an acoustic booth.

of these averages that are predicted by the better hearing ear (**Table 3.2**) (5).

Assessment of Bone-Conduction Thresholds without Masking

Not-masked bone-conduction thresholds have to be obtained once the air-conduction thresholds have been assessed. These are obtained using the same technique as described in **Table 3.1**, vibrating the skull with the vibrator most commonly placed on the mastoid of the worse-hearing ear.

If the not-masked bone-conduction thresholds are the same as the air-conduction thresholds in both ears, there is no conductive defect in either ear (**Figure 3.5**). There is no requirement to test the bone-conduction thresholds any further. It is only when the not-masked bone-conduction thresholds are poorer than the air-conduction thresholds in one or both ears that masking is required.

Masking to Determine Masked Bone-Conduction Thresholds

The bone-conduction vibrator vibrates the entire skull and, hence, both cochlea (**Figure 3.6**). It is

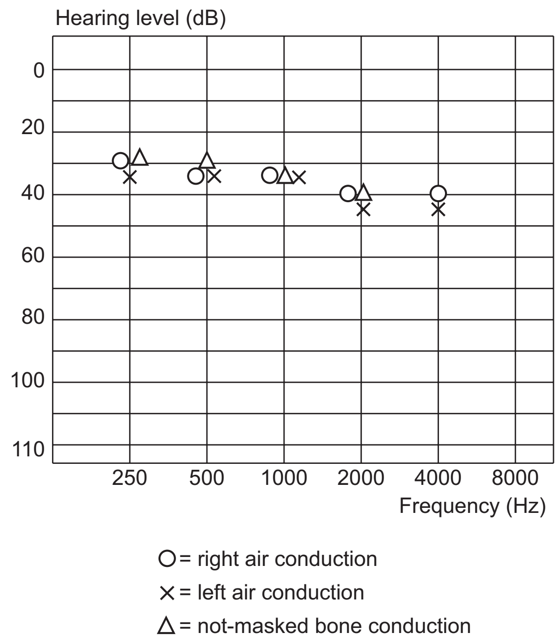


Figure 3.5 Bilateral, symmetrical sensorineural impairment. The pure tone average of the air-conduction thresholds in the right ear is 35 + 35 + 40 + 50 = 160 ÷ 4 = 40 dB HL and in the left ear is 35 + 35 + 45 + 50 = 165 ÷ 4 = 41 dB HL. That is, both ears have a mild hearing impairment. At all frequencies, the not-masked bone-conduction thresholds are less than 10 dB different from the air-conduction thresholds in both ears. There is thus no potential air–bone gap in either ear and no need to mask the bone-conduction thresholds.

Table 3.2 WHO Grades of hearing impairment in better hearing ear

Grade of impairment	Average 0.5, 1, 2, 4 kHz	Performance	Recommendations	Comments added to the previous classification
0: No impairment	25 dB or better	No or very slight hearing problems. Able to hear whispers	None	20 dB also recommended. People with 15–20 dB levels may experience hearing problems. People with unilateral hearing losses may experience hearing problems even if better ear normal
1: Slight/mild impairment	26–40 dB	Able to hear and repeat words spoken in normal voice at 1 m	Counselling. Hearing aids may be needed	Some difficulty in hearing but can usually hear normal level of conversation
2: Moderate impairment	41–60 dB	Able to hear and repeat words using raised voice at 1 m	Hearing aids usually recommended	None
3: Severe impairment	61–80 dB	Able to hear some words when shouted into better ear	Hearing aids needed. If no hearing aids available, lip-reading should be taught	Discrepancies between pure tone thresholds and speech discrimination score should be noted
4: Profound impairment including deafness	81 dB or greater	Unable to hear and understand even a shouted voice	Hearing aids may help in understanding words. Additional rehabilitation needed. Lip-reading and sometimes signing essential	Spoken speech distorted, the degree depending on the age at which hearing was lost

Abbreviations: dB: decibel; Hz: Hertz; ISO: International Organisation for Standardisation; m: metre (After International Organisation for Standardisation [5].)

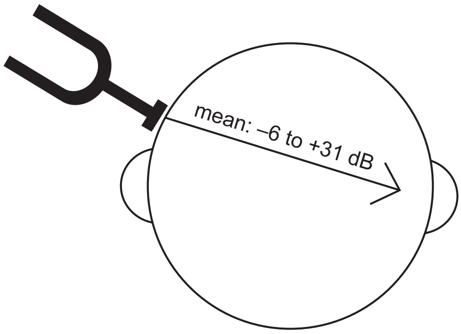


Figure 3.6 Trans-skull attenuation of bone conduction. Though there is a range of levels of attenuation across the human skull, both positive and negative (**Table 3.3**) for practical purposes when performing (and interpreting) an audiogram, the implications are such that it has to be assumed that the bone conduction measured in one ear will be heard in the other ear. Hence, audiometric masking of the bone conduction in the non-test ear is always essential, whenever there is a potential conduction defect suggested by there being an air–bone gap (difference) indicative of a conductive defect in the test ear.

agreed that, regardless of where the bone vibrator is placed, there is no way of knowing which ear is being tested unless masking is used. Some suggest that the not-masked bone-conduction thresholds be assessed with the vibrator on the vertex to remind the tester of this but, in many instances, this can lead to considerably higher thresholds (4). **Table 3.3** shows the transcranial attenuation of bone conduction from one side of the skull to the other. The range of values is quite large and indeed can be better for the ear on the opposite side to the one being stimulated. As it is not possible to predict what the value will be in an individual patient, the minimum values have to be assumed. These are less than 0 dB at every frequency. Hence, whenever one wishes to assess the bone conduction in an ear with a potential air–bone gap, it is necessary to mask the bone conduction in the other ear. This cannot be done by bone conduction, as this will also mask the bone conduction in the test ear. Hence, it has to be masked by air conduction, usually with an ear insert rather than headphones.

Masking Techniques

Several different methods of masking have been proposed but the preferred method is shadow masking and to record the thresholds on a masking chart (**Figure 3.7**). The masking signal is a narrow band of noise in the region of the test frequency delivered by air-conduction to the non-test ear. It is usual to start with the masking sound at its threshold of detection in the non-test ear. If the AC or BC threshold in the test ear deteriorates with this level of masking, cross-hearing is occurring, and the level of masking is increased by 10 dB and the threshold reassessed. This procedure is repeated until the thresholds are no longer affected by an increase in masking level. The true threshold is the level where three sequential masking levels give rise to thresholds that are within 5 dB (**Figure 3.7**). If the masking sound is increased any further, the thresholds will start to deteriorate, initially because of central masking and then because of cross-masking. The reason why the central masking occurs is uncertain, but its contribution is unimportant in practice compared with cross-masking when the masking sound is sufficiently loud that it also masks the test ear.

In those whose air-conduction thresholds suggest asymmetric hearing, if the not-masked bone-conduction threshold is similar to the air-conduction thresholds in the better ear, the not-masked bone-conduction thresholds apply to that ear. This requires the bone-conduction thresholds to be assessed in the poorer ear with air-conduction masking of the better hearing ear.

Table 3.3 Transcranial attenuation (dB) of audiometric bone conduction

Bone conduction	Mean	0.5 kHz	1 kHz	2 kHz	4 kHz
Minimum	–6	–10	–5	–5	–5
Maximum	31	25	25	35	40
Median	10	10	5	10	15

(After Snyder, 1973 [7].)

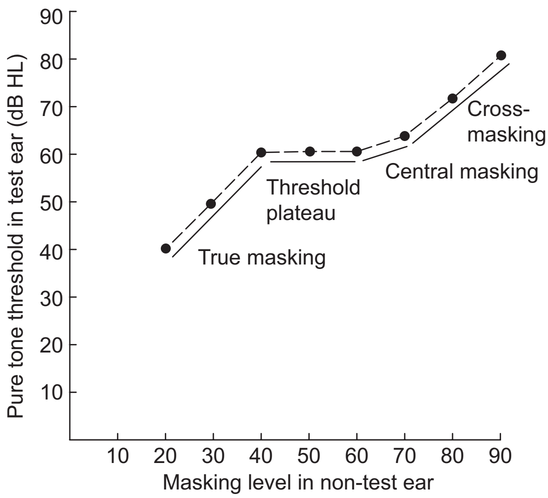


Figure 3.7 Masking chart.

In those with a bilateral conductive defect, the not-masked bone-conduction thresholds will be poorer than the air-conduction thresholds in both ears. This requires the bone-conduction thresholds to be assessed in each ear with masking of the other ear.

The magnitude of any conductive hearing defect for each ear is calculated by averaging the air and bone conduction over the speech frequencies of 0.5, 1, 2 and 4 kHz with the difference between them giving the magnitude of the air–bone gap.

General Validity of Bone-Conduction Thresholds

In testing bone conduction, because of additional test variables, including positioning and pressure of the vibrator, the test/retest variation can be considerably greater than when testing air-conduction thresholds. Hence, mean average changes of bone conduction of 10–15 dB on two test occasions should be interpreted with caution.

Definite Air–Bone Gap

Where the not-masked average bone-conduction thresholds are poorer than the air-conduction thresholds by 10 dB or more in both ears, then there must be an air–bone gap in one or both ears (**Figure 3.8**). This is because the not-masked bone-conduction thresholds apply to at least one ear, so by definition, an air–bone gap must exist in at least one ear. Masking is required to assess the bone-conduction thresholds in each ear.

Potential Air–Bone Gap

When the not-masked bone-conduction thresholds are the same as the air-conduction thresholds in one ear but not the other (**Figure 3.9**) the not-masked bone-conduction thresholds must apply to the better ear. Potentially, there could be an air–bone gap or a sensorineural impairment in the poorer ear, so masking of the better ear is required to assess the bone-conduction thresholds in the poorer ear.

Air-Conduction Masking

Having carried out any bone-conduction masking to confirm whether an air–bone gap is present or not, it is now necessary to reconsider whether any air-conduction masking is required. This may already have been considered necessary and have been carried out after the air-conduction thresholds had been determined and a difference of 40 dB or greater noted.

To understand air-conduction masking, it is necessary to realise that the air-conduction thresholds are tones assessed using headphones and not free-field. The amount of sound that escapes from these headphones is negligible but what they do is vibrate the skull. Sound can then go by bone conduction from the headphone on one side to the cochlea on the other side. Inevitably, because of the mass of the skull the sound will be attenuated to a certain degree and, for practical purposes, the minimum level is taken as 40 dB (**Figure 3.10**). There are two main situations when this might affect the air-conduction thresholds.

1. If there is a difference of more than 40 dB between the air conduction in the poorer ear and the bone conduction (which is often also the same as the air conduction) in the better ear, the air-conduction thresholds in the poorer ear could be a shadow from the better ear (*see* **Figures 3.11** and **3.12**). Repeat air-conduction testing, with masking of the better ear, will be necessary.

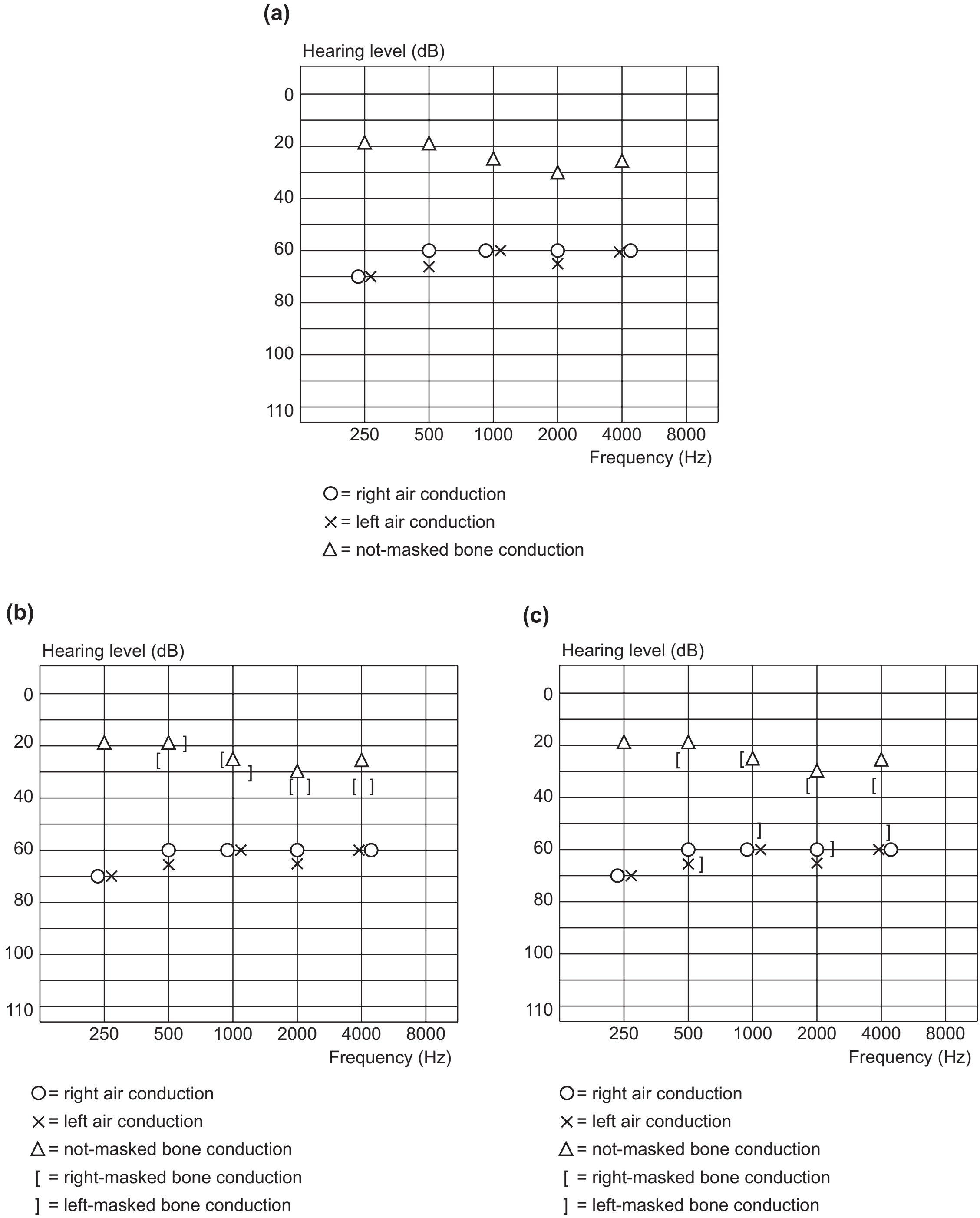


Figure 3.8 Potential air–bone gap in one or both ears. This patient in audiogram **(a)** has a bilateral moderate hearing impairment, the pure tone air-conduction FFA average in the right ear being 60 dB HL and in the left ear 63 dB HL. The not-masked bone-conduction thresholds are in ~25 dB HL and could apply to either or both ears. Masking is required to ascertain the masked bone-conduction thresholds in the right and in the left ear. The most likely outcome is a bilateral conductive impairment **(b)**. A much less likely alternative **(c)** is a conductive impairment in one ear (right in this example) and a sensorineural impairment in the other ear of the same magnitude.

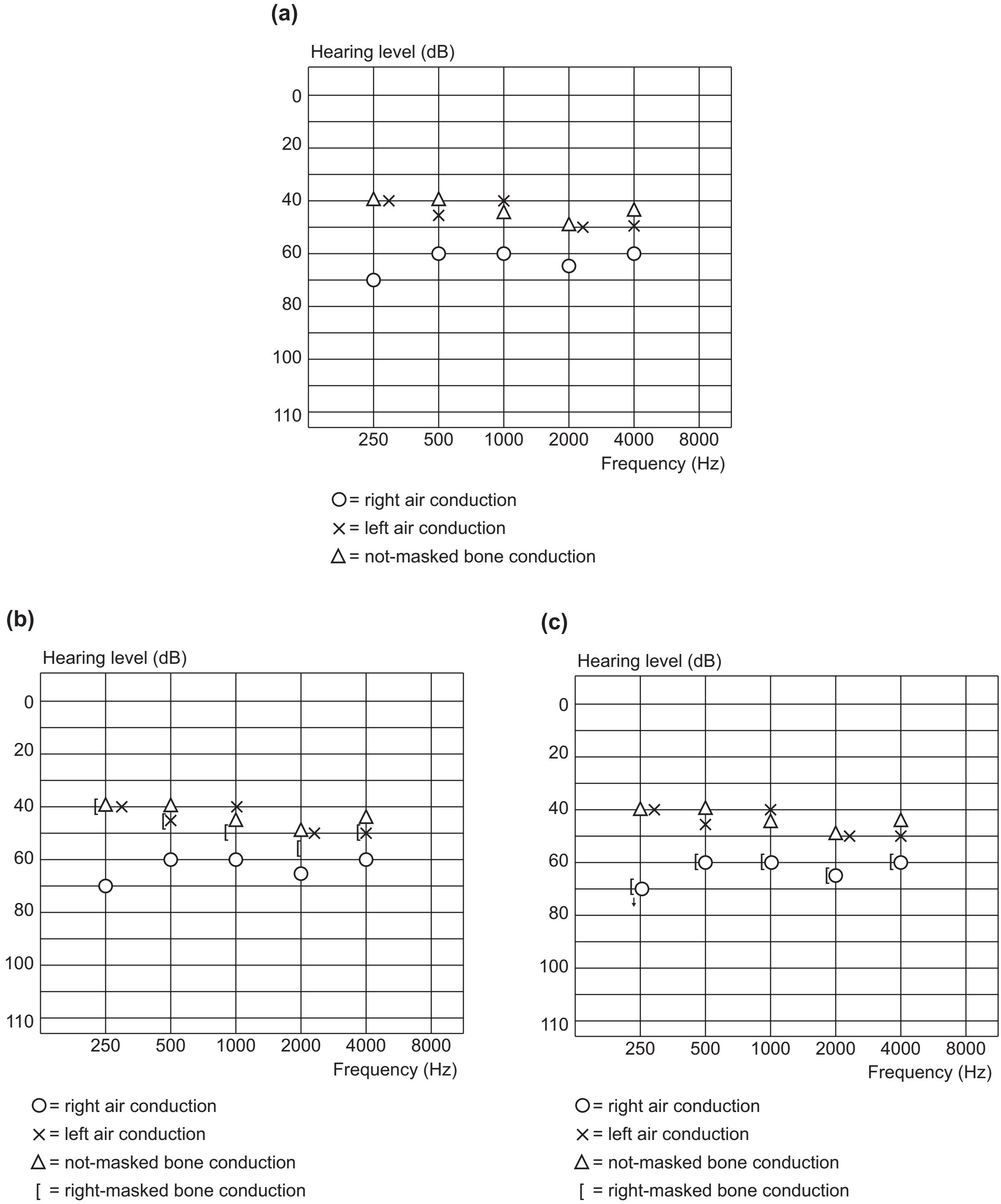


Figure 3.9 Potential air–bone gap in poorer (right) ear. **(a)** Taking the air-conduction threshold, the left ear is the better hearing ear with a mild hearing impairment with a pure tone air-conduction FFA average of 45 dB HL. As the not-masked bone conduction at all frequencies are no more than 10 dB different to the air-conduction thresholds, this is a moderate sensorineural impairment as there is no potential air–bone gap. The right ear has a 61 dB HL air conduction average, indicating a severe hearing impairment. What is not clear is whether the difference between the ears is due to an additional conduction defect. Hence, the left ear air conduction has to be masked so that the masked bone-conduction thresholds in the right ear can be assessed. The two most likely outcomes are that there is a mixed loss in the right ear **(b)** or a sensorineural loss in the right ear **(c)**.

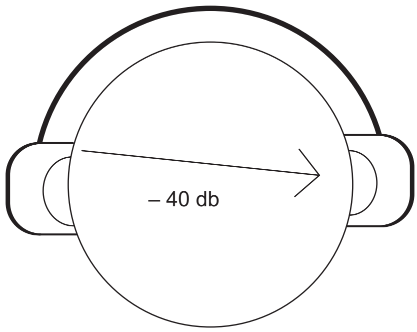


Figure 3.10 Trans-head attenuation of test 'air-conduction' sounds in pure tone audiometry. This occurs mainly by sound escaping from the head phones testing an ear by 'air conduction' and being heard by the other ear, as well as being heard, as the illustration suggests, by bone conduction. So, for practical purposes, it has to be assumed that the air-conduction sound also arrives at the cochlea on the non-test ear, attenuated by ~40 dB or greater. Thus, in audiometry, masking of the air conduction in the non-test ear is only required if the air conduction in the non-test ear is better than the test ear by ~40 dB or greater.

2. If there is a major conductive defect in the test ear, the air-conduction thresholds could be the threshold of detection of the sound from the headphones via the osseous route (**Figure 3.12**). In these circumstances, the air–bone gap would not necessarily be an accurate measure of the magnitude of the conductive defect.

Air conduction masking is again a narrowband noise in the region of the test frequency but delivered by headphones. The same shadow masking procedure is used as in bone-conduction masking.

INTERPRETING THE AUDIOGRAM

Data Required

In many situations, the audiometric results are not fully reported, and this applies particularly to many 'scientific presentation / reports papers.' The following data must be reported to allow a full evaluation of an individual's audiogram.

1. All the audiometric thresholds in both ears are required. Otherwise, the requirements for masking cannot be made if only the thresholds in one ear are reported.
2. The symbols used for the different audiometric thresholds must be 'defined'.
3. Not-masked bone-conduction thresholds must be available along with the air-conduction thresholds for each ear.
4. The non-masked bone-conduction thresholds apply only to one ear unless they are identical to the air-conduction thresholds in both ears.
5. Otherwise, bone-conduction masking is required and the masked bone-conduction thresholds recorded on the audiogram chart.
6. This is not a simple task and requires considerable training.

Method of Interpretation

The first calculation is of the average air-conduction thresholds in each ear that is usually done over 0.5, 1, 2 and 4 kHz to give a four-frequency average (FFA). For some purposes, a three-frequency average 3FA over 1, 2 and 3 kHz is used. It is then possible to categorise the severity of hearing impairment in each ear using one of the recognised WHO bandings (*see* **Table 3.2**).

A decision is also made as to whether the air-conduction thresholds are symmetric (within 10 dB of each other) or asymmetric (different by greater than 10 dB). This distinction does not always agree with a patient's report as to how symmetric the hearing is. There are many potential reasons for this but a stricter pure tone threshold criterion cannot be used because of the 10 dB test/retest error of audiometry.

In most patients, the FFA will not formally be calculated, as the degree of impairment can usually be assessed by 'eyeballing' the audiogram. Calculating an FFA is often left for specific circumstances, such as when a report is being written, for research or when the FFA is borderline between categories of impairments.

There are four types of hearing that can be identified audiometrically: sensorineural, conductive, mixed and uncertain.

Sensorineural Impairment

Here, there is no difference between the air- and bone-conduction thresholds but the FFA of the air-conduction thresholds is poorer than the level considered to be 'not hearing impaired'. Such a criterion is a pure tone FFA average over

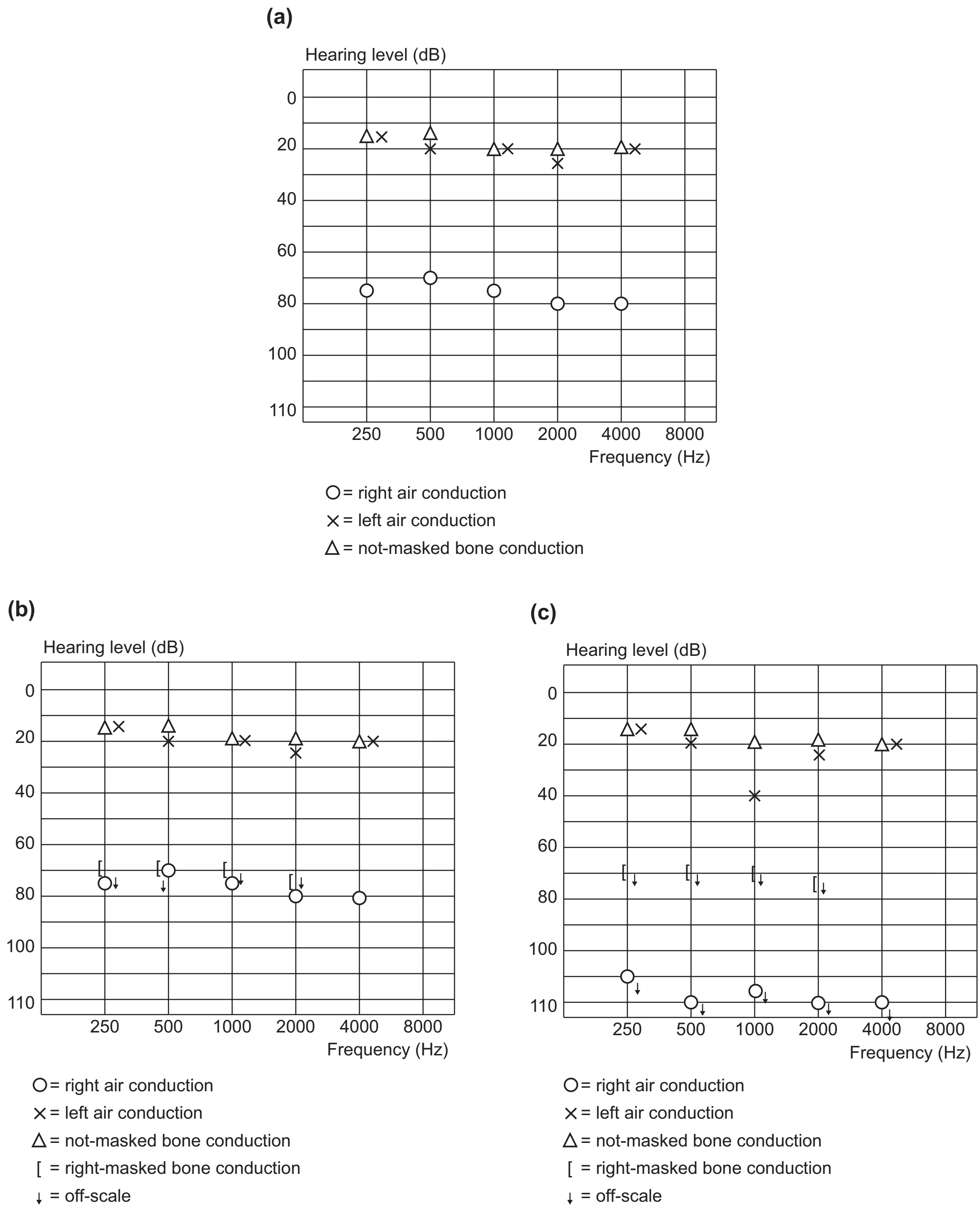


Figure 3.11 Requirement to mask the air conduction. **(a)** The pure tone average in the left ear is 21 dB HL, which is normal hearing. At all frequencies the not-masked bone-conduction thresholds are less than 10 dB different from the air-conduction thresholds and hence these must apply to the left ear. The pure tone average in the right ear is 76 dB HL. These thresholds could be a shadow from the left ear and the first requirement is to mask the air conduction in the left ear and assess the masked bone-conduction thresholds in the right ear. In this instance they are off-scale **(b)**. It is still possible that the right air-conduction thresholds might be due to transcranial hearing in the left ear because the air-conduction thresholds in the poorer ear are more than 40 dB worse than the bone conduction in the better ear. In this instance, when the air conduction in the right ear is reassessed the right ear is a 'dead' ear **(c)**.

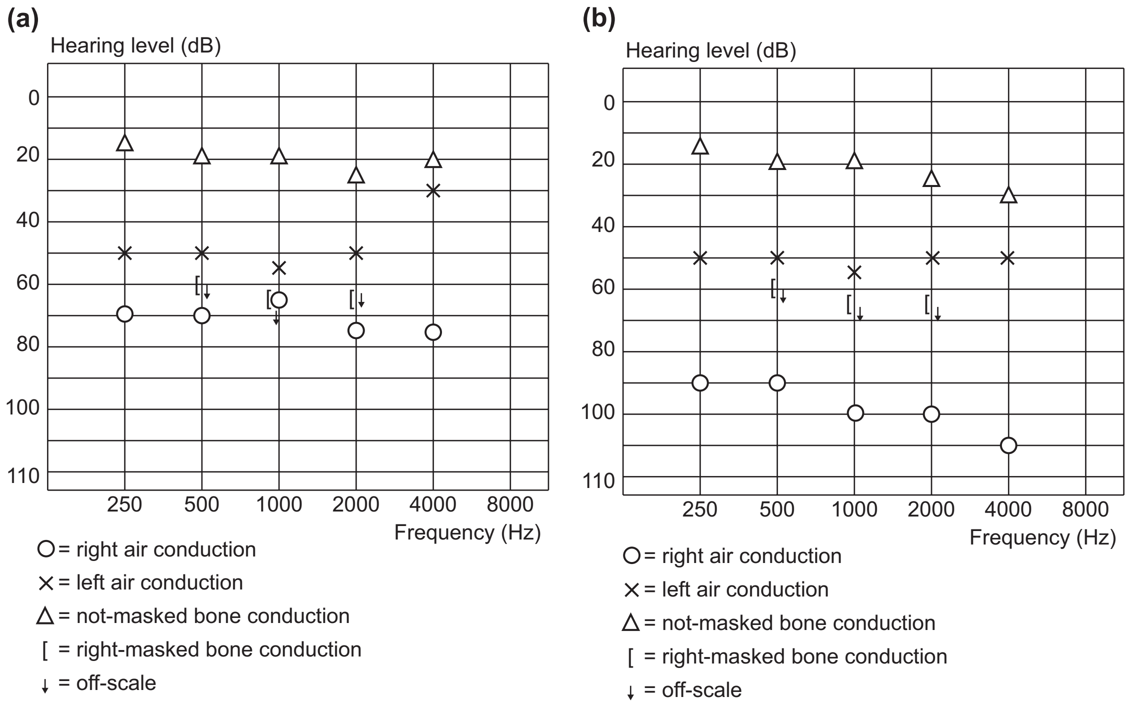


Figure 3.12 **(a)** Requirement to mask the air-conduction thresholds. The pure tone average in the left ear is 51 dB HL and in the right is 71 dB HL. The not-masked bone-conduction thresholds are likely to apply to the left ear and masking confirms this in that the masked bone-conduction thresholds in the right ear are off-scale. There is now a requirement to mask the left ear and assess the masked air-conduction thresholds because the air-conduction thresholds in the poorer (right) ear are worse than the bone conduction in the better (left) ear by greater than 40 dB. Having done this, the patient is found to have a profound rather than a severe sensorineural impairment in the right ear **(b)**.

0.5, 1, 2 and 4 kHz of 25 dB HL (*see* **Table 3.2** and **Figure 3.3**).

Criteria can also be set which categorise the patient as having poorer hearing for some frequencies. As sensorineural impairments usually affect the high frequencies rather than the low ones, this can give rise to a sloping audiogram (**Figure 3.13**) that increases with age (see **Figure 10.1**). Sometimes, there is a notch in the audiogram around 4 kHz (also visible in **Figure 3.13**) but this may not be identified unless 3 and 6 kHz are also tested (**Figure 3.14**). The presence of a notch is not unusual in ears with a mild to moderate sensorineural hearing impairment. This can occur irrespective of the presumed aetiology, though some would incorrectly suggest that it is more likely as a consequence of noise trauma (*see* **Chapter 10**).

Conduction Defect

This is considered to be present if there is a difference between the air-conduction and the bone-conduction thresholds (i.e. an air–bone gap) of greater than 10 dB, usually averaged over 0.5, 1 and 2 kHz rather than 0.5, 1, 2 and 4 kHz (**Figure 3.15**). This is because in most cases, the air–bone gap is largest at the lower frequencies for the reasons explained in **Chapter 4**.

Mixed Impairment

Here, there is both a conductive and a sensorineural impairment in an ear. The criterion for a conductive component is again an air–bone gap of greater than 10 dB and for a sensorineural component an average bone-conduction threshold worse than a certain

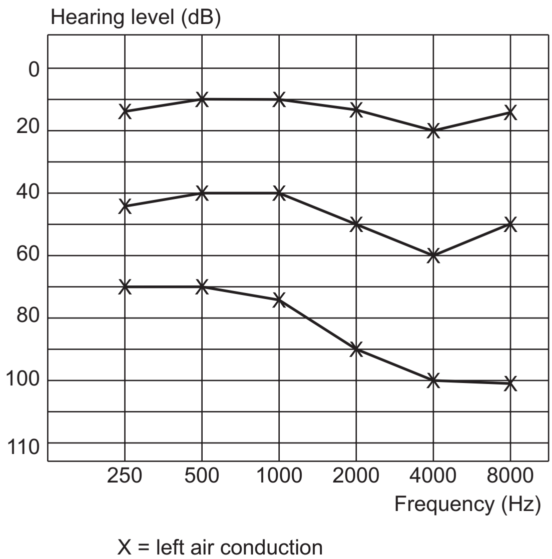


Figure 3.13 Typical progression due to ageing of a sensorineural impairment. This example shows three sequential air-conduction audiograms in the left ear. The bone-conduction thresholds are omitted for simplicity.

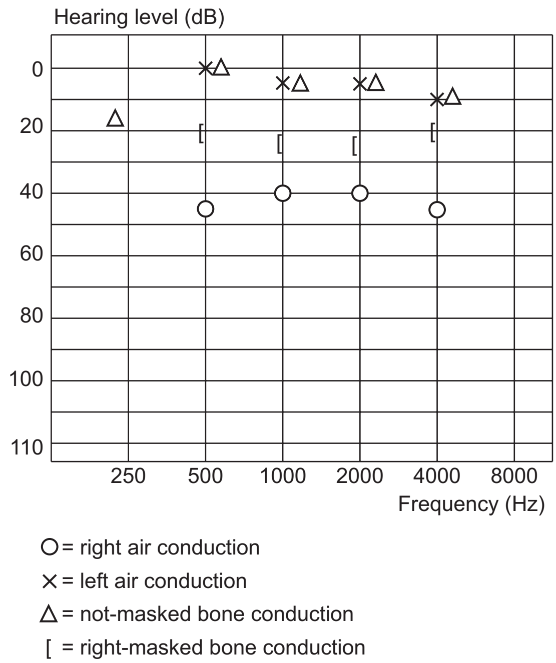


Figure 3.15 Conductive impairment in the right ear with no hearing impairment in the left ear. The not-masked bone-conduction thresholds must apply to the left ear. Masking of the air conduction in the left ear is necessary to assess the masked bone-conduction thresholds in the right ear. The pure tone FFA in the right ear is 80 + 65 + 55 + 45 ÷ 4 = 61 dB. The air–bone gap in the right ear is 55 + 45 + 30 + 25 = 155 ÷ 4 = 39 dB.

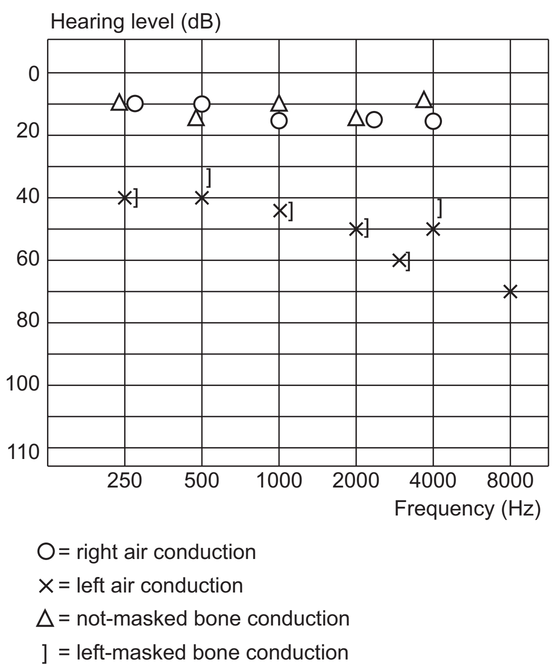


Figure 3.14 Audiogram 'notch'. This mild sensorineural hearing impairment has a notch in the audiogram at 3 kHz. This would not have been evident if the thresholds at 3 kHz had not been tested.

'normal' criterion, usually 25 dB HL. The level of bone-conduction thresholds used to define 'normal' in a mixed impairment is even more debated than with a sensorineural impairment because of the Carhart effect (*see* **Chapter 4**). It could be argued that the level should be 10 dB poorer than the 25 dB HL pure tone average usually used for a sensorineural impairment. However, in most instances, no allowance is made for the Carhart effect.

Uncertain Category

In patients with a severe or profound impairment, it is not unusual to be uncertain as to whether the impairment is sensorineural or mixed in type. This is for two reasons that apply only to this group. First, the maximum bone conduction output of most audiometers is in the region of 70 dB HL. It can be even less in portable audiometers.

Hence, if the not-masked bone-conduction thresholds are poorer than this, they cannot be assessed (**Figure 3.11b** and **c**). All ears with a profound impairment (PTA ≥ 90 dB HL) could therefore have either a sensorineural or mixed impairment if the bone-conduction thresholds are poorer than 70 dB HL (**Figure 3.16**). Secondly, masking is delivered by air-conduction and the maximum output available to mask bone conduction is usually 110 dB HL. This greatly limits the amount of masking available when the air-conduction thresholds are poorer than 70 dB HL (**Figure 3.16**). Thus, in patients with a bilateral severe (or poorer) impairment in whom some not-masked bone-conduction thresholds are available, it is not possible to get masked bone-conduction thresholds to decide whether both or only one ear has a conductive component (**Figure 3.17**).

An ear with an air–bone gap of less than 10 dB does not necessarily have a normal conductive mechanism, it is just that an impairment cannot be measured because of the test/retest error of the method (**Figure 3.18** and **Chapter 4**).

Figure 3.18 also emphasises the necessity of having an audiogram available for both ears that included the not-masked bone-conduction threshold to ensure that appropriate bone and air conduction masking has taken place. The audiogram is of the right ear and shows air and masked bone-conduction thresholds. From the air conduction thresholds, it would appear to be a mild hearing loss with an FFA of ~30 dB HL. What one would then want to know is whether it is a conductive, sensorineural or mixed impairment. Otoscopic examination would be important to assess alongside a valid audiogram.

In this case, however, the not-masked bone conduction thresholds are reported. If these were around zero, then masking would be required of the left ear to gain the right ear's masked bone conduction. If the not-masked bone conduction thresholds were similar to the right ear's air-conduction thresholds, then right ear bone-conduction thresholds would not be required to fully evaluate the right ear.

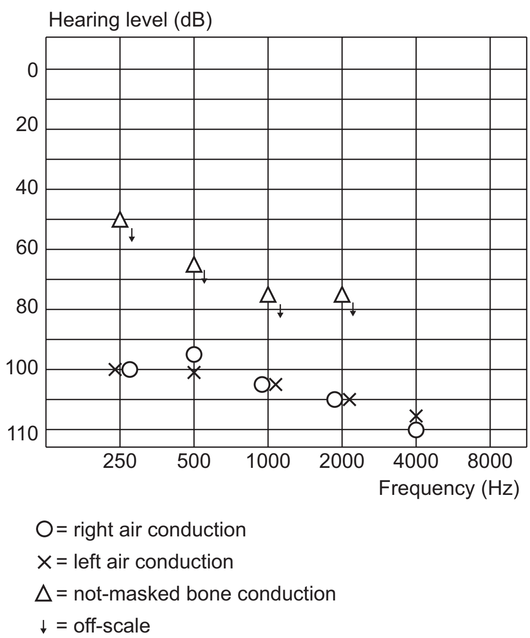


Figure 3.16 Bilateral profound impairment where it is not possible to ascertain the type of impairment because of limited presentation output of bone conduction. It could be a mixed or a pure sensorineural impairment.

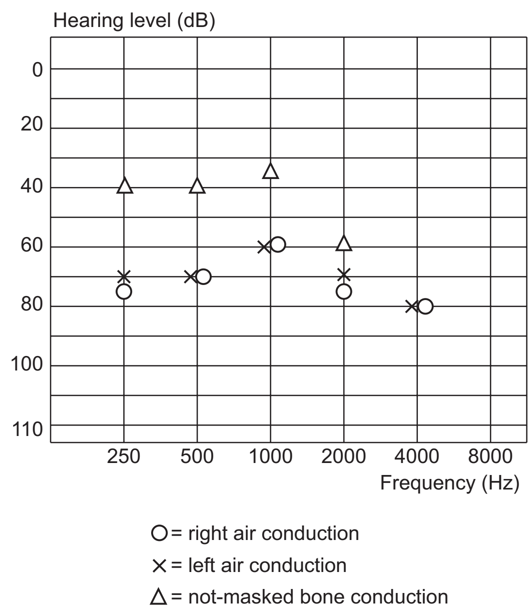


Figure 3.17 Bilateral severe impairment. One or both ears have a mixed impairment but it is uncertain which because of an inability to mask due to the limited output of masking noise; that is, 110 dB is insufficient to mask the hearing in an ear with a pure tone average of 70 dB HL.

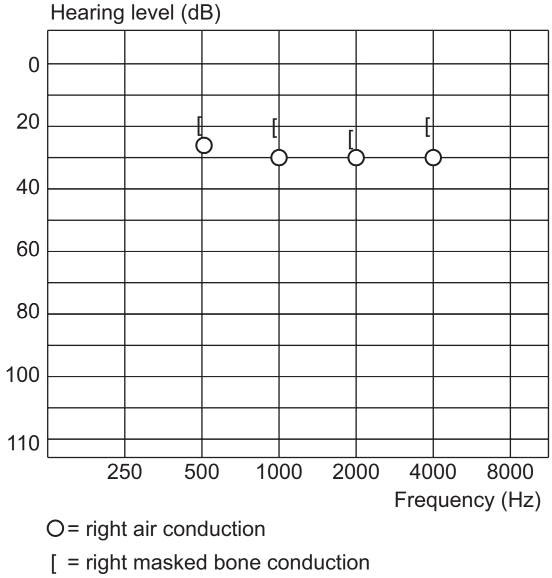


Figure 3.18 Uncertain category. Audiogram of the right ear with its air and masked bone-conduction thresholds. From the air-conduction thresholds it would appear to be a mild hearing loss with an FFA of ~30 dB HL. What one would then want to know is whether it is a conductive, sensorineural or mixed impairment. Otoscopic examination would be important to assess alongside a valid audiogram.

Many less well trained technicians, who carry out pure-tone audiometry, incorrectly use the masked bone conduction symbols to indicate which side the bone-conduction vibrator was placed and the only way someone can assess whether this is the case is having the not masked thresholds available.

So, in this case, if appropriate bone-conduction masking has been performed, an eyeball scan of the air–bone gap is no greater than 10 dB at any frequency. Hence without calculating it, the average air–bone gap over the 4 frequencies has to be 10 dB or less. This is within the test/retest error of audiometry and also of clinical relevance.

In this case then, the mild hearing impairment in the right ear is most likely a sensorineural one. However, it is not of a characteristic configuration when due to normal ageing. The threshold at 4kHZ is no poorer than those at 1 and 2 kHz and it would be interesting to have known what it was at 3 and 6 kHz.

As such the next stage to check would be the air-conduction thresholds on the opposite left side if it had been available. If it had been greater than 70 dB HL (due to a combination of cross skull attenuation of 40 dB and the apparent FFA of 30 dB) and having a conductive component the right ear could be a ‘dead’ ear.

In conclusion, for PTA to be validly interpreted both ears must be reported and include the not-masked bone-conduction thresholds.

Exaggerated Pure Tone Thresholds

In those who have a potential reason to exaggerate their hearing loss, such as having a medicolegal claim for a noise induced hearing loss or head trauma, the clinician seeing the ‘claimant’ must use a variety of evaluations to confirm that the PTA accurately measures the pure tone average. These may include just conversing with the claimant, carrying out free speech testing, and perhaps doing speech audiometry, in addition to pure tone audiometry with variations of types and ear of masking, all the while being alert for irregular or inconsistent responses. Exaggeration is most commonly attempted by responding ‘whenever they definitely hear a loud signal’ rather than pressing the button ‘whenever they think they hear a signal’, as they are instructed to do, but it is hard to do this consistently enough to be convincing to an alert audiologist.

Non-Organic Hearing Loss

This is a condition that both older children and adults can have in which a hearing impairment is reported when none can be identified by audiometry or other means. It is commonly seen, for example, in academically high-achieving teenage girls. They often return an audiogram showing a flat sensorineural hearing loss of around 50 dB, marked by the audiologist as having inconsistent responses, and out of keeping with normal responses to a quiet conversational voice during the consultation. It is often helpful to explain that hearing tests are sometimes hard to do; this test doesn’t seem to fit with the rest of the examination findings and perhaps if we do the test again in a few weeks we will get more reliable results which we expect to be normal. This gives the patient a way out without losing face. Combined with some gentle enquiries about stress and worry at school or at home, this will often point the way to the parents who will understand what is going on. Occasionally, it may be necessary to resort to objective tests such as otoacoustic emission test (OAE) and auditory brainstem response (ABR) to definitively exclude organic pathology. Referral to clinical psychology is sometimes required.

CONCLUSIONS

- Pure tone audiometry is the standard audiometric test.
- If carried out in a sound-deadened environment, with calibrated equipment, using a standard technique in a patient who responds when they 'think they can hear a tone', the results are highly reproducible.
- The results can be used to define in each ear the air-conduction thresholds for pure tones. When averaged these give an indication of the degree of hearing impairment in each ear.
- Often mathematical calculations are not required if the thresholds are fairly similar. In this situation, eyeballing the audiogram is sufficiently accurate in giving a likely average for practical purposes.
- If audiometry is carried out in a noisy environment, a false, low-frequency loss due to noise masking can occur.
- Pure tone audiometry, to be interpreted validly, requires both ears to be reported and include the not-masked bone-conduction thresholds.
- If the bone-conduction thresholds are better than the air-conduction thresholds, as evidenced by an air–bone gap of 10 dB or greater over 0.5, 1 and 2 kHz, there is a defect in the sound-conduction mechanism of the external and/or middle ear.
- Bone-conduction thresholds are initially assessed not-masked. When there is a potential air–bone gap in an ear, the bone-conduction thresholds need to be reassessed with masking of the other ear.
- A sensorineural hearing impairment is considered to be present if the bone-conduction thresholds are similar to the air-conduction thresholds and the average of the latter (usually over 0.5, 1, 2 and 4 kHz) is worse than a certain level which defines normal hearing, usually 25 or 30 dB HL.
- A mixed hearing impairment is considered to be present if the average bone-conduction thresholds satisfy the definition for a sensorineural impairment and there is in addition an air–bone gap of 10 dB or greater at the lower frequencies of 0.5, 1 and 2 kHz.
- When the air-conduction thresholds in the poorer hearing ear are poorer than the bone-conduction thresholds in the contralateral ear by 40 dB or more, the air-conduction thresholds could be a shadow of these bone-conduction thresholds and must be masked.
- The bone-conduction thresholds do not solely measure inner ear function. In a normal ear and in those with a sensorineural impairment, there is also a material air-conduction component transmitted via the external canal and ossicular chain.
- In those with a conductive impairment, the bone-conduction thresholds do not have this air-conduction component. This Carhart effect occurs at all frequencies but is greatest at 2 kHz and is of a ~10 dB magnitude.

Following successful surgery with closure of the air–bone gap, the bone-conduction thresholds improve by ~10 dB by eliminating this Carhart effect.

FURTHER READING

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4

Conductive and mixed hearing loss

IDENTIFICATION OF A CONDUCTION DEFECT

Tuning Fork Test

Tuning fork tests were the traditional clinical method of detecting a conduction defect before pure tone audiometry (PTA) became more readily available. There are even some clinicians who use tuning fork tests as a screening method for deciding whether to carry out audiometric bone conduction (BC) testing. They do this because of the difficulties in masking and interpreting bone-conduction thresholds in an audiogram.

In ears that are otoscopically normal and in which a hearing impairment is present, a Rinne negative result (BC>AC) indicates otosclerosis with an air–bone gap of 10 dB or greater with a 95% degree of confidence. On the other hand, a Rinne positive result (AC>BC) does not help, as there could still be an air–bone gap of ≥ 25 dB due to otosclerosis in at least 25% of ears (**Chapter 2, Table 2.11**). Such reliance on tuning fork tests is not justified as discussed earlier (*see* **Chapter 2** and [1]).

The Weber tuning fork test is also not helpful because of its inconsistency in results primarily because of the way the skull transmits the sounds to each side of the head with a mean variation of -6 to + 31 dB (**Figure 3.6**). Such reliance on tuning fork tests is not justified as discussed earlier (**Chapters 2** and **3**[1]).

Otoscopy

If tympanic and middle ear abnormalities are identified on otoscopy, the sound conduction mechanism will almost certainly be affected and there will be an associated conduction defect. Under these circumstances, what one wants to know is the magnitude of the air–bone gap, and the only way of ascertaining this is by pure tone audiometry with appropriate masking (**Chapter 3**).

As such, there is no point in doing Rinne or Weber tuning fork tests in ears with otoscopic tympanic membrane or middle ear pathology because it is already known that the conduction mechanism will have been affected.

Pure Tone Audiometry

Conductive defects are best identified by the presence of an air–bone gap on a PTA (**Chapter 3 Figure 3.8)** and this is the accepted 'gold standard' against which everything is compared.

The configuration of the audiometric air–bone gap should be visually examined. Because the conductive mechanism of the tympanic membrane and ossicular chain is a mechanical one, it is more efficient at some sound frequencies than others. Evolution in humans has made it more efficient at the lower audiometric frequencies which fits with the resonance properties of the ear canal which is greatest at 3 to 4 kHz (*see* **Figure 10.1**).

Correspondingly, a conductive hearing impairment due to pathology of the tympanic membrane and ossicular chain should be greater at 250 000 and 1 Khz than at 2 and 4 kHz (*see* **Figure 3.8**). If this is not evident in a suggested audiometric air–bone gap, then its accuracy must be questioned.

For management purposes, the air–bone gap is the sole method of quantifying the magnitude of a conductive defect, and this depends on

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having an accurate audiogram with appropriate masking.

Validity of a Pure Tone Audiogram in Confirming a Conduction Defect

When an audiogram is available, the first thing a clinician has to do is to assess the audiogram's validity. On occasions, PTA can be unreliable for several reasons but the majority of these can be identified by evaluating a fully completed audiogram form (**Chapter 3**).

Regrettably, such forms or similar are not routinely used and the not-masked bone-conduction thresholds are not included. The symbols for a masked bone-conduction threshold are also incorrectly used when masking has not been performed.

The machines must have been calibrated regularly, and the tests performed in a sound-deadened environment according to a protocol by a trained audiologist, whose experience should be known. The headphones should have been correctly placed over the ears, the bone-conduction vibrators placed appropriately in contact with the skull, and the subjects fully cooperative.

Pure Tone Audiometric Data Potentially Required

In many situations, the audiometric results are not fully reported, and this applies particularly to many scientific presentations / reports /papers. The following data must be reported /available to allow a full evaluation of an individual's audiogram to be performed.

1. All the audiometric thresholds in both ears are required. Otherwise, the requirements for masking cannot be made if only the thresholds in one ear are available.
2. Symbols used for the different categories of audiometric thresholds must be provided.
3. The not-masked bone-conduction thresholds must be available along with the air-conduction thresholds for each ear.
4. The not-masked bone-conduction thresholds apply only to one ear unless they are identical to the air-conduction thresholds in both ears.
5. Otherwise, bone-conduction masking is required and subsequent masked bone-conduction thresholds recorded on the audiogram chart.

This is not a simple task and requires considerable training.

Audiogram Assessment Technique

The air-conduction thresholds for each ear are first evaluated, most commonly visually, to assess the degree of impairment, if any, over 0.5, 1, 2 and 4 kHz. This should be in keeping with any free field voice testing hearing that has been performed and any rough assessment that has been evident when taking a history.

The configuration of the air-conduction thresholds can be considered with sensorineural hearing thresholds being poorer at the high frequencies and conduction defects are greater at the lower frequencies. (*see* **Figures** in **Chapter 3**).

If no bone-conduction thresholds have been recorded, which is usual when a commercial hearing aid is being provided, otoscopy is the main way that most conductive impairment might be suggested. It is also likely if the air-conduction thresholds indicate a low-frequency hearing impairment. This also often occurs if the audiometry has been carried out in a noisy situation.

When indicated by the use of masked bone-conduction symbols are reported, symbols of not-masked bone-conduction thresholds are next looked for. Some audiometricians incorrectly use the masked bone-conduction symbols to indicate which ear the bone-conduction vibrator was positioned on and this incorrect use of these masking symbols must be identified.

If the air-conduction thresholds are similar in both ears and the 'not masked' bone-conduction thresholds are within approximately 10 dB of the air-conduction thresholds, there is no conduction defect in either ear. Bone-conduction masking is therefore not required in this subject.

Masked thresholds are always required if there is a potential air–bone gap in either ear. If this is the case, then masked bone-conduction thresholds are necessary.

If masking of the bone conduction has not been performed in such a situation, then the validity of the audiogram must be questioned as they are a necessary step in attributing the bone-conduction thresholds to a particular ear.

When the air-conduction thresholds are different between ears and the not-masked bone-conduction thresholds the same as the air-conduction thresholds in one ear, then the masked bone-conduction thresholds should be ascertained in the poorer hearing ear.

Masking of the contra-lateral ear by air conduction, to confirm or refute the presence of a true air–bone gap indicative of a middle ear conduction defect in the other ear, is a vital diagnostic but often complicated procedure that requires considerable audiometric training to carry out appropriately.

When performing masking, the position of the bone vibrator on the skull is not relevant because sound can be transmitted across the skull without being attenuated. In most cases suggestive of a unilateral conductive defect, the bone vibrator will have been repositioned on the mastoid behind that ear which is understandable but not required.

The only way to mask the non-test ear is by air conduction. If there is a conduction defect in the non-test ear it may be impossible to mask it sufficiently because there is a limit to the sound output of the machine. This situation is most likely to occur when the audiogram suggests a bilateral moderate/severe conduction defect. In these circumstances, there will be uncertainty of which ear the bone conduction is being tested because of the inability to adequately mask either ear. The likelihood of one ear being a 'dead' ear is considerable.

The upper limit of the bone-conduction vibrator is around 70 dB so that when the patient has a severe or profound impairment (i.e. the air-conduction thresholds are greater than 70 dB HL) there is no way of determining whether there is a conduction component to the impairment or not. Finally, the maximum size an air–bone gap can be is around 60 dB. This is because the bone conduction vibration inevitably vibrates the air and is also heard by air conduction.

Considering all these possible difficulties with the bone-conduction thresholds, it is easy to understand why the air–bone gap does not invariably detect or accurately quantify the size of a conductive defect. The clinician has a responsibility to ensure that masking has been appropriately carried out, particularly if surgical intervention is being considered, and particularly when the provisional diagnosis of otosclerosis is being considered in an otoscopically 'normal' ear.

WHAT ARE THE BONE-CONDUCTION THRESHOLDS?

Hearing by bone conduction helps one to monitor one's own voice. Its assessment in audiometry is of value in detecting and quantifying conduction defects, but when such a defect is present the bone-conduction thresholds underestimate inner ear function.

When the skull is set in vibration by bone-conducted sounds this vibrates the inner ear by three routes (**Figure 4.1**). Firstly, the sound emanates into the external auditory canal and is heard via the middle ear (route c). Secondly, the skull vibration sets the ossicular chain in motion and this energy is transferred to the inner ear via the middle ear (route b). Thirdly, the cochlea vibrates as part of the skull, stimulating the inner ear (route a). It is difficult to ascertain the relative contribution of each of those routes, but those going via the external auditory canal and middle ear are likely to be as important as the direct route.

In ears with a conductive defect, the sound will not be as well transmitted by the first two routes, leading to poorer bone-conduction thresholds in such ears. This Carhart effect, which is more fully discussed below, occurs at all frequencies but is maximal at 2 kHz. The implication is that in an ear with a conductive impairment, the bone-conduction thresholds are not a measure of inner ear function in the same way that they are in a normal ear or one with a sensorineural impairment.

Bone Conduction as a Measure of Inner Ear Function

In patients with normal hearing or a sensorineural loss, the air-conduction thresholds are taken to represent inner ear function, and this is valid. In those with an air–bone gap, this is not the case, and the only alternative is to use the bone-conduction thresholds. However, when there is a conduction defect, the bone-conduction thresholds do not assess the same things as they normally would.

The fact that the bone conduction is not always a measure of inner ear function is best explained

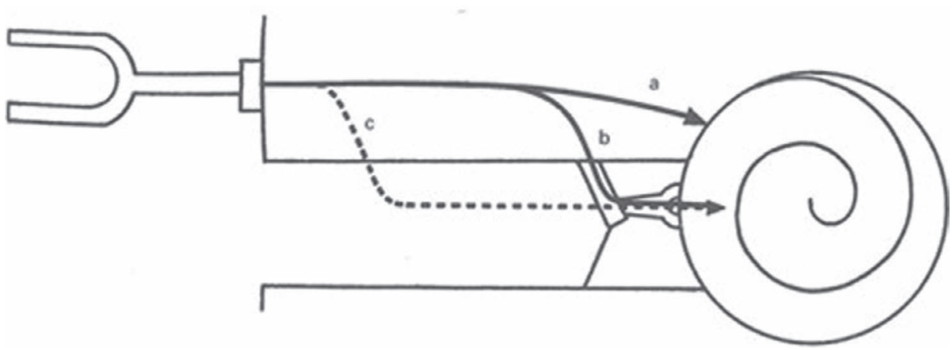

Figure 4.1 The three normal routes (a, b and c) of bone conduction to the inner ear. **(a)** Via vibrating the bony skull; **(b)** Via vibrating the ossicular chain; **(c)** Via vibrating the air in the external auditory canal.

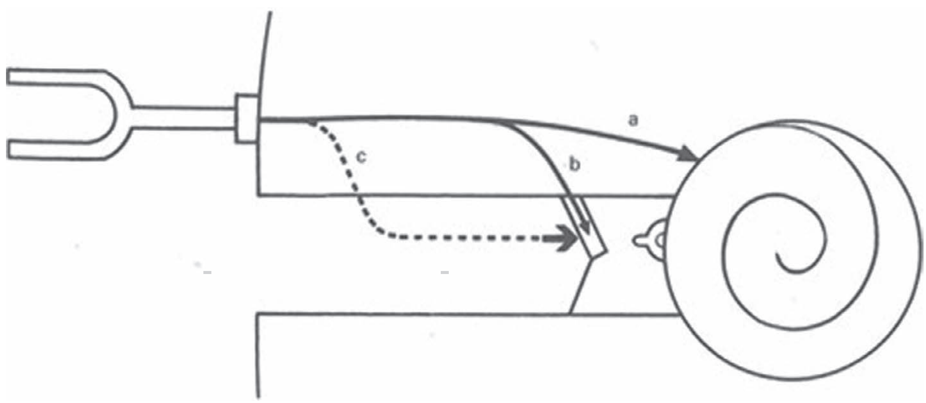

Figure 4.2 In the presence of a total middle ear conduction defect the osseous route (a) is the only route for bone conduction to stimulate the inner ear.

by reference to two diagrams, one where the conduction mechanism is normal (**Figure 4.1**) and one where it is disrupted (**Figure 4.2**). When the skull is vibrated by bone conduction, it not only vibrates the cochlea (route a) but it also vibrates the tympanic membrane and ossicular chain (route b) and the air in the external auditory canal (route c). Thus, when the ear's middle ear conduction mechanism is functioning normally, bone conduction has three components (routes a, b and c) and cannot be considered a measure of inner ear function (route [a] only).

The Carhart Effect

Routes (b) and (c) are both important as has been shown in experimental work in cats (**Figure 4.3**). When the conduction mechanism is totally disrupted (*see* **Figure 4.2**), sound can only go to the

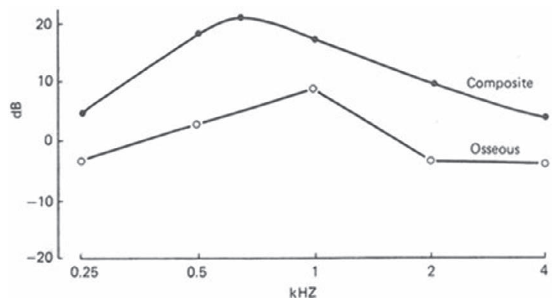

Figure 4.3 The composite cochlear microphonics at various frequencies in cats initiated by stimulating bone conduction. The osseous component is that which arrives at the inner ear via the bone (route a). The remainder is made up of the components arriving at the inner ear via the external auditory canal and the middle ear (routes b and c). Because of the different dimensions of a cat's ear, compared to the human ear, the magnitude of the effect is at 0.5 to 1 kHz (after Tonndorf [2]).

cochlea via the bone (route a). If the conduction mechanism is surgically repaired, then the components via the canal and ossicular chain (routes b and c) are added to the sound arriving at the cochlea by route (a) and the bone-conduction thresholds will improve.

That the bone-conduction thresholds can improve after successful middle ear surgery was first described by Carhart (1950) in patients undergoing fenestration of the lateral semicircular canal for otosclerosis (3). Since then, many other authors have confirmed this, not only in ears with otosclerosis, but in those with chronic otitis media and otitis media with effusion. The magnitude of the effect is greatest at 2 kHz (**Table 4.1**); hence the reason for the Carhart notch (**Figure 4.4 a** and **b**) frequently seen in otosclerosis preoperatively but also present to an extent in all middle ear pathologies causing a conduction defect. Following successful surgery to 'repair' the sound conduction mechanism, the postoperative bone-conduction thresholds will improve at all frequencies, but especially at 2 kHz with the elimination of the notch. Because otosclerotic surgery is more likely to be technically successful than surgery for chronic otitis media, the improvement in bone-conduction thresholds is therefore evident more often following stapes surgery.

There are two main practical implications of the Carhart effect. First, in ears with a conductive impairment, the bone-conduction thresholds will be falsely poorer by a mean of 10–15 dB HL. This, in many instances, will make the patient appear to have a mixed rather than a pure conduction impairment. Secondly, where surgery for otosclerosis is being considered, the postoperative air-conduction thresholds could well be better than the preoperative

Table 4.1 Magnitude of the Carhart effect (dB)

	kHz			
	0.5	1	2	4
Carhart (3)	5	10	15	5
Gunderson (6)	12	12	12	9
Ginsberg et al. (5)	15	17	18	17
Gatehouse and Browning (4)	5	8	12	5

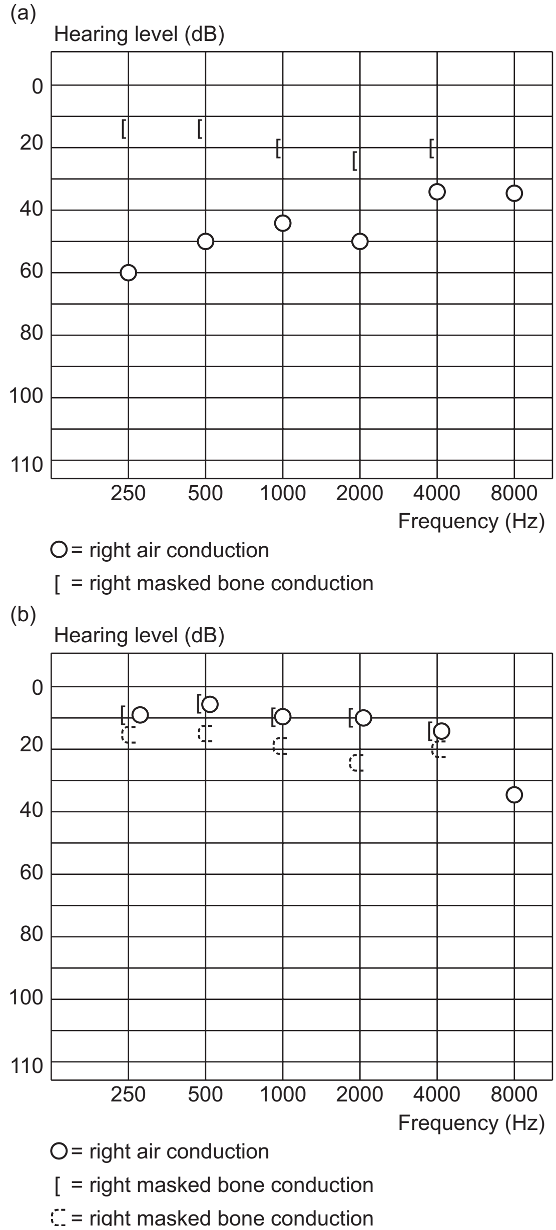

Figure 4.4 **(a)** Preoperative pure tone audiogram of an ear with a conductive impairment. There is a dip in the bone conduction around 2 kHz – the Carhart notch. Note the magnitude of the air–bone gap is greatest at the lower frequencies. **(b)** Postoperative audiogram following successful surgical repair. The air–bone gap has been over-closed with the improvement in the bone-conduction thresholds greatest at 2 kHz.

bone-conduction thresholds by 10–15 dB HL. This possibility is also present in surgery for chronic otitis media but is less frequently observed because it is less frequently technically as successful.

Mixed Hearing Loss

Individuals with a conduction defect due to otosclerosis and chronic otitis media are not immune from developing an age-related sensorineural hearing loss delete 'as they age' and may therefore have a mixed hearing impairment. The categorisation of an ear as having a 'mixed hearing loss' is thus often complicated by the Carhart effect.

The attribution of there being a mixed hearing loss is not usually done by calculating the bone conduction average but rather having an eyeball glance to see if they are poorer than 25/30 dB HL. If this is the case, there is the temptation to attribute the sensorineural impairment to the pathology responsible for the conductive defect, without considering that it might be due to natural ageing. When the conduction defect is unilateral, then there is the bone conduction in the other ear to assess the effect of ageing which is most commonly symmetrical. What is rarely considered, especially in ears with chronic otitis media, is that any associated sensorineural impairment might have been caused by ototoxic ear drops or previous surgery.

PATHOPHYSIOLOGICAL INTERPRETATION OF AN AUDIOMETRIC AIR–BONE GAP

Tympanic Membrane Pathology

Chalk patches are the initial stage and tympanosclerotic plaques the end stage of degeneration of the fibrous collagen layer of the tympanic membrane. Fortunately, chalk patches do not materially affect the sound transmission characteristics of the tympanic membrane as they do not reduce the area or affect the mobility of the tympanic membrane. A tympanosclerotic plaque, on the other hand, can sometimes reduce the membrane's freedom to move if it extends to the annulus, but this is unusual (**Figure 4.5**). It is more likely, that when there is a large plaque in the tympanic membrane, the ossicular chain will also be fixed by tympanosclerosis (**Figure 4.6**) and this, rather than the plaque, will be the main cause of the conductive defect.

Turning our attention now to a common situation in chronic otitis media, where there is a tympanic membrane defect and the ossicular chain is intact and mobile. The magnitude of the conductive defect will primarily relate to the size of the perforation. That this is a real, rather than an academic concept, has been confirmed by analysis of the air–bone gap in individuals with a tympanic membrane defect but an intact and mobile ossicular chain (**Table 4.2** and **Figure 4.7**).

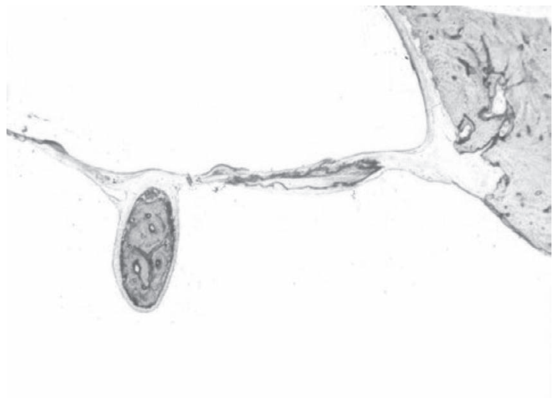

Figure 4.5 Histological section through the tympanic membrane with a tympanosclerotic plaque that does not affect its vibration (haematoxylin and eosin stain).

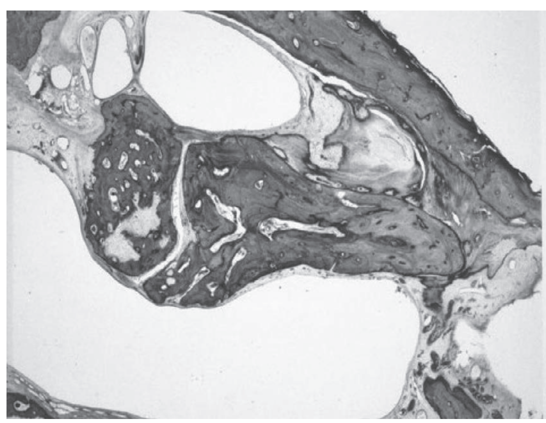

Figure 4.6 Histological section through the incus and malleus in the attic fixed by a plaque of tympanosclerosis.

Table 4.2 Correlation between size of perforation and air–bone gap

Percentage perforation	Air–bone gap (dB HL ± 1 SD)
0–25	12 ± 7.5
26–50	22 ± 8.5
51–95	28 ± 9.0

(After Austin, [7])

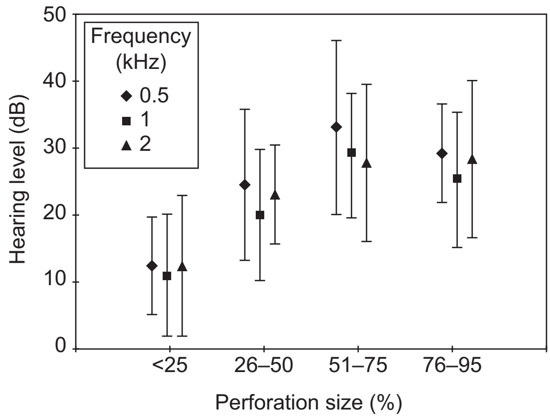

Figure 4.7 Mean air–bone gap at 0.5, 1 and 2 kHz related to the size of the perforation in ears with an intact and mobile ossicular chain. Bars indicate ± 1 SD (after Austin [7]).

The larger the perforation, the greater is the air–bone gap up to a maximum of ~30 dB. It is incorrect to consider that posterior perforations are associated with a larger conductive impairment than anterior perforations because of the loss of the round-window baffle effect. Sounds arriving in the same phase and timing at the oval and round window will dampen movement of the perilymph, no matter where the perforation is. The reason why posterior perforations are more often associated with larger air–bone gaps, than similarly sized anterior perforations, is that posterior perforations are more frequently associated with ossicular chain abnormalities.

Physiology of the Bony Ossicular Chain

The ossicular chain is held in the middle ear by its ligaments and the intratympanic muscles (**Figure 4.8**). The ligaments are not often considered to be important, but dislocation of the incus and its interposition between the stapes and the malleus in a tympanoplasty, is not as inherently stable as a normal ossicular chain (**Figure 4.9**). Hence, many surgeons simply heighten the stapes with an ossicle or cartilage to make direct contact with the tympanic membrane (**Figure 4.9**) to create a piston system. This, because of its greater stability, often closes the air–bone gap as effectively as more complicated ossiculoplasty procedures (**Table 4.3**).

Ossicular Chain Pathology

The two main pathologies that affect the ossicles are otosclerosis and chronic otitis media, In clinical

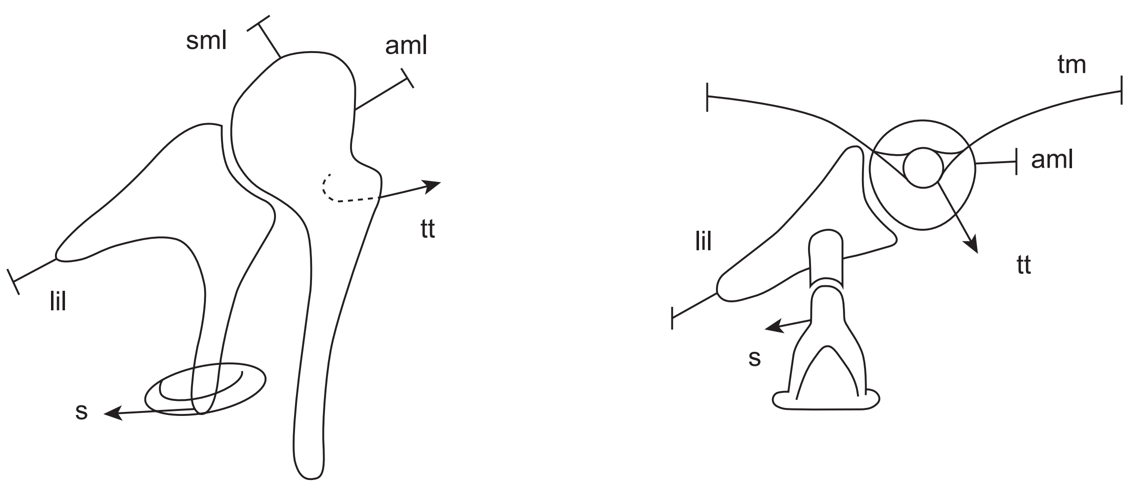

Figure 4.8 Ossicular chain suspension. aml = anterior malleolar ligament; lil = lateral incudal ligament; s = stapedius muscle; sml = superior malleolar ligament; tm = tympanic membrane; tt = tensor tympani.

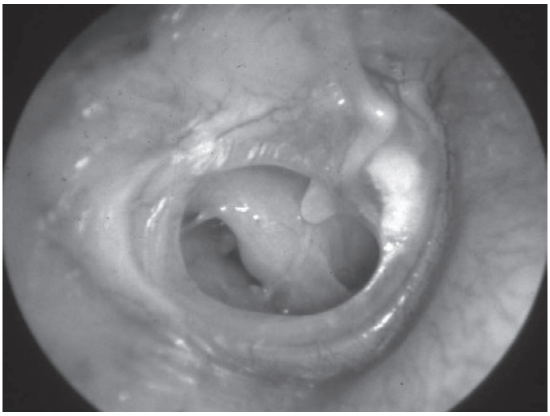

Figure 4.9 A right tympanic membrane with a 60% inferior perforation with a tympanosclerotic plaque anterior to the handle of the malleus. Mucosa seen through the perforation is inactive. If the air–bone gap is greater than 30 dB there is likely to be ossicular chain fixation in the attic as in **Figure 4.8**.

otosclerosis, the footplate can become progressively fixed by obliteration of the fibrous joint starting anteriorly. The air–bone gap can thus gradually increase up to a maximum of 60 dB. The main reason why an air–bone gap of greater than 60 dB is not possible is because the maximum output of bone conduction of most audiometers is 60 dB **(Table 4.4).**

The diagnosis of otosclerosis is a presumptive one in ears where there is an air–bone gap of 15 dB or greater over 0.5, 1 and 2 kHz and the tympanic membrane is normal and mobile. Tympanometry and acoustic reflexes are not helpful (*see* **Chapter 7**). The most modern scientific study (8), is a series of temporal bones that were scanned and then serially sectioned for histological identification of stapedial otosclerosis. In this study there was a ~10% false-negative and a ~10% false-positive diagnostic rate of otosclerosis. In clinical practice these false rates are likely to be greater. With time, and perhaps with the use of Artificial Intelligence, scanning is likely have a reduction in these false negative and false positive rates.

In chronic otitis media, apart from tympanic membrane pathology evident on otoscopy, the ossicular chain can be affected to various degrees either by tympanosclerosis or necrosis. The magnitude of the air–bone gap, along with otoscopic assessment of the ossicular chain, can give some indication of the likely pathology. A final assessment can only be made at surgery when the chain can be more accurately assessed. In many instances, there are adhesions between the middle ear structures. In addition, the tympanic membrane can be retracted on to the remaining parts of the chain, such as the long process of the incus and the stapes superstructure. Thus, when the ossicular chain is disrupted, the magnitude of the air–bone gap can vary from 0 dB up to a maximum of around 60 dB. Understandably, the size of the air–bone gap is unrelated to where the disruption in the chain is, disruption of the incudo-stapedial joint will cause as large an air–bone gap as total absence of the ossicular chain.

The general rule must be that in chronic otitis media an air–bone gap of up to 15 dB can be explained by a perforation. An air–bone gap of between 15 and 30 dB may be explained by a perforation if it affects more than 25% of the tympanic membrane, but there may also be ossicular chain problems (**Figure 4.9**). In these circumstances, it is a matter of debate whether, surgically, it is better, to leave the ossicular chain and the adhesions alone or to attempt to reconstruct the ossicular chain. If the air–bone gap is greater than 40 dB there almost certainly will be ossicular chain problems and a myringoplasty alone is unlikely to resolve the hearing problem.

Figure 4.10a illustrates the commonest ossicular chain defect in chronic otitis media with the two main techniques of its repair. Method in **Figure 4.10c** is likely to be more effective in the longer term as the reconstruction is less influenced by the middle ear muscles.

Surgical repair of such a defect entails a myringoplasty usually with a facial graft, along with dislocation of the remaining incus and its surgical modification to rebuild the ossicular chain. Its positioning between the stapes and the malleus (**Figure 4.10b**), is not as inherently 'stable' as a normal ossicular chain with its ligaments because of the ligaments suspending the malleus. Hence, many surgeons simply heighten the stapes with an ossicle or cartilage to make direct contact with the tympanic membrane to create a piston system (**Figure 4.10c**). This, because of its stability, often closes the air–bone gap as effectively as more complicated ossiculoplasty procedures.

Prevalence of Air–Bone Gaps in the Adult Population

In the UK National Study of Hearing (9) a stratified sample of 2708 British adults, aged 19–80 years, was chosen from a sample of 48 313 adults randomly selected from the electoral roll for a full

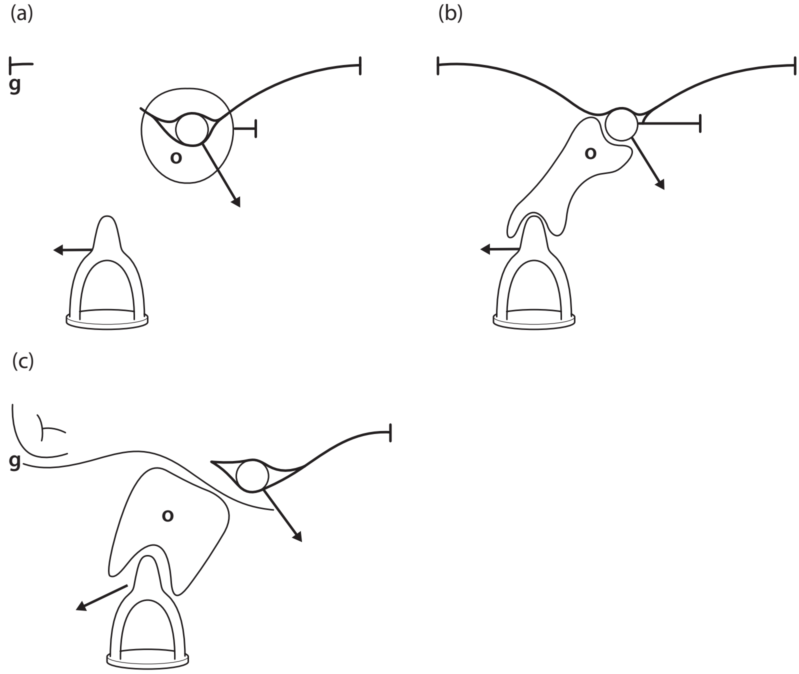

Figure 4.10 **(a)** The commonest defect in the ossicular chain due to chronic otitis media is erosion of the long process of incus and a posterior tympanic membrane perforation. **(b)** Reconstruction with a tympanic membrane graft and refashioning the body of the incus to fit between the stapes and incus handle. **(c)** Piston system created by refashioning the body of the incus and repositioning it on the stapes over which the tymmpanic graft is placed thus heightening the stapes. g = graft; o = repositioned ossicle.

otological and audiological assessment. Otoscopy was performed by consultant otologists and the audiometry and tympanometry were carried out to a strict protocol at four NHS University Hospital departments. The prevalence of middle ear disease in that population was subsequently reported separately (10) from the main analysis (9) and provides the data presented in this section.

Table 4.3 Population prevalence of a hearing impairment (FFA ≥ 25 dB HL over 0.5, 1, 2 and 4 kHz) in the poorer hearing ear with the ratio of Sensorineural / Conductive loss of ≥ 15 dB HL over 0.5, 1 and 2 kHz (10)

Prevalence poorer ear (%)		Ratio SN/CON
Overall	18	2 : 1
18–40 yrs	4	1 : 1
41–60 yrs	17	2 : 1
61–80 yrs	45	4 : 1

The age-related and overall prevalence (%) in the adult population of a hearing impairment in the poorer ear, defined as an average of the air-conduction thresholds over 0.5,1,2 and 4 kHz of 25 dB HL or greater is shown in **Table 4.3**. A conductive impairment was defined as an air–bone gap of 15 dB or greater over 0.5, 1 and 2 kHz and ears that did not have such an air–bone gap were considered to be sensorineural impairments. The ratios of sensorineural to conductive impairments in those with an impairment in their poorer ear are shown.

In the age band of 18 to 40 years, the proportion of hearing impairments that are conductive is the same as those with a sensorineural impairment. With ageing, the proportion with a sensorineural loss gradually increases due to normal ageing.

Table 4.4 shows the magnitude of the air–bone gap in the 2208 adults whose tympanic membrane could be seen past debris and wax was otoscopically categorised.

Table 4.4 Percentage of ears with an air–bone gap of various magnitudes broken down as to their otoscopic appearance

	% Normal TM	% Abnormal Intact TM	% Inactive chronic otitis media	% Active chronic otitis media
Air–bone gap (dB)				
0–14	97.3	79.6	56.4	16.3
15–29	2.2	16.9	25.7	30.9
30+	0.5	3.5	17.9	52.8

Table 4.5 Population prevalence per 100 of the presumptive diagnosis of otosclerosis, healed OM and Eustachian tube dysfunction. (the 95% confidence intervals are in parentheses)

	Otosclerosis	Healed otitis media	Eustachian tube dysfunction
Overall	2.1 (1.5–2.7)	1.7 (1.1–2.3)	0.9 (0.6–1.2)
Age (years)			
18–40	1.6 (0.6–2.6)	1.5 (0.5–2.5)	0.3 (0.1–0.5)
41–60	2.2 (1.3–3.1)	1.8 (1.5–2.5)	1.2 (0.5–1.9)
61–80	3.0 (1.7–4.3)	2.9 (1.4–4.4)	1.5 (0.6–2.4)
Sex			
Women	2.0 (1.3–2.7)	1.6 (0.8–2.4)	0.8 (0.4–1.2)
Men	2.2 (1.2–3.2)	1.9 (0.9–2.9)	1.1 (0.6–1.6)
Occupational grouping			
Non-manual	1.5 (0.8–2.2)	1.3 (o.1–2.5)	0.8 (0.2–1.4)

In those with a normal, mobile tympanic membrane ~3% had a conductive impairment of 15 dB or greater, suggesting the diagnosis of otosclerosis.

In those who had an abnormal but intact tympanic membrane because of previous otitis media, ~20% had a conductive hearing loss of 15 dB or greater. The magnitude of this conductive impairment in 'scarred tympanic membranes' is of a previously unreported frequency and magnitude. The air–bone gaps in ears with active and inactive chronic otitis are considerably greater in those that are active.

Table 4.5 is a deleted further break down of **Table 4.4** by age, sex and occupational group. Those in **Table 4.4** and an air–bone gap of greater than 14 dB with a normal tympanic membrane are assumed to have otosclerosis.

Those classified in **Table 4.4** with an abnormal intact tympanic membrane are divided into those where Eustachian tube dysfunction was evident on tympanometry and those who had normal tympanometry and are classified as having healed otitis media

THIRD WINDOW SYNDROME

Sound vibrations mobilising the stapes require the fluid in the inner ear to vibrate to stimulate hair cells on the basilar membrane. This is achieved by having a round window membrane (*see* **Figure 1.8** in **Chapter 1**) in the middle ear.

In the 1940s, fenestration of the superior semicircular canal was sometimes performed using chisels to remove the bone over the canal via a mastoidectomy in patients where their stapes was presumed to be fixed by otosclerosis. This had the objective of creating a third window to aid the hearing. Regrettably, most patients developed Tullio's syndrome (11) where loud sounds made the patient dizzy. Temporarily increasing intracranial pressure with a cough, for example, also caused dizziness.

Because of its increasing definition and therefore use of CT scanning in patients with balance problems, the presumptive diagnosis of a superior semicircular canal dehiscence and the 'third window syndrome' (12) is increasingly being made. However, histological studies (13) have shown there can still be a bony covering over the fibrous semicircular canal up to less than 1 mm thick and radiology at present would not identify bony covering of that magnitude. The diagnosis of a bony dehiscence is thus likely to be considerably over frequently made by clinicians. In addition, there will always be dura covering any true dehiscence of a thickness of ~4 mm that would minimise any effect on the balance. This minimising effect of the dura could be the reason why, in this histological study, few patients with no bony cover of the superior semicircular canal had no ear symptoms.

Many patients with balance problems and a radiologically suggestive semicircular canal dehiscence have an associated low tone hearing loss similar to a middle ear conduction defect (**Figure 4.4a**). Patients with Tullio-type balance symptoms associated with noise, along with radiological suggestions of a superior semicircular canal dehiscence, are no less likely to have a conductive hearing loss than those in the general population (**Table 4.3**).

Otoscopy, rather than tympanometry or acoustic reflexes should identify many of these but otoscopy is rarely, if ever reported, in case series. Strong reliance in case reports is also made on Weber testing. In addition, the audiometry is very poorly reported. Both ears and any masking that should have been performed are never reported. The commonest cause of an incorrect suggestion of a low-frequency hearing loss is when the audiometry has

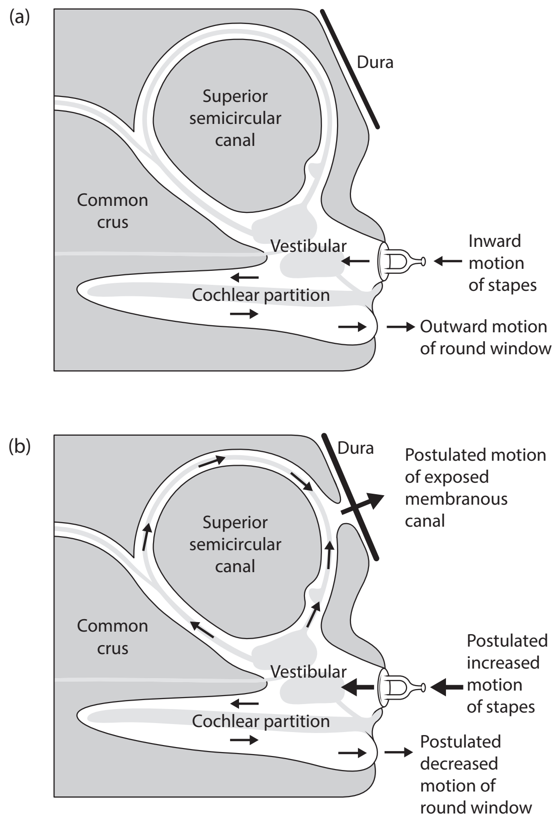

Figure 4.11 **(a)** represents the cochlear bone as it will appear on a radiological image. What would not be visualised radiologically is the dura covering the area of the superior semicircular canal in the middle cranial fossa. **(b)** represents the radiological appearance of a patient with a potential superior semicircular dehiscence, but the dura will still be covering that area and minimising any effect.

not been carried out in a sound-deadened environment (*see* **Figure 3.4**). No paper on the low tone hearing loss in association with a presumptive diagnosis of a 'third window syndrome ' has been identified that has reported the audiometric results to the standard suggested earlier in this chapter under the heading of 'Pure tone audiometric data potentially required'.

In a histological study (14) of 1000 vertically sectioned temporal bones from 596 patients, in 5 (0.5%) bones from four patients there was complete bony dehiscence over the canal (**Figure 4.12**). Medical records were available in three of these patients, one of whom had a large acoustic neuroma

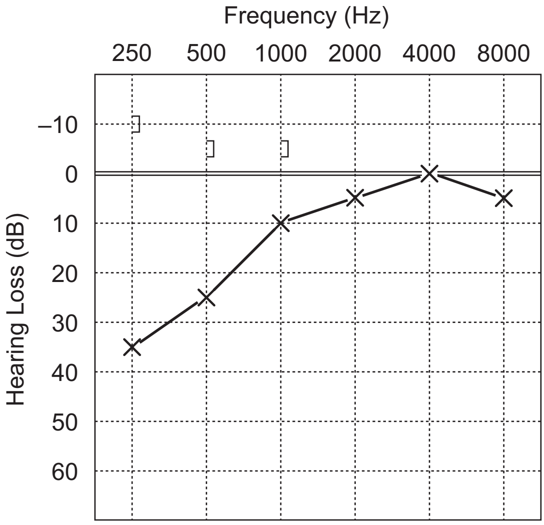

Figure 4.12 The low-frequency conductive hearing loss frequently seen in the presence of a third window phenomenon, most frequently superior semicircular canal dehiscence.

associated with imbalance. The other two patients did not have balance symptoms. In 14 (1.4%) other bones, the bone was no thicker than 1 mm. Only one of these patients had a history of vertigo but there are many other diagnoses for the causation of vertigo. It should be noted that a normal stapes footplate is only ~1 mm thickness.

Clinicians should therefore be cautious of diagnosing a 'third window syndrome' based on cross-sectional imaging, because the definition of CT is still not powerful enough to accurately determine the amount of bone between the canal and the dura covering it, from the middle cranial fossae. Radiology does not identify the fact that there is also an overlying fibrous dura on the bone. This could be the main reason why the majority of individuals with radiological otic capsule dehiscences do not manifest a clinical 'third window syndrome'.

The diagnosis of a third window syndrome is largely clinical; confirmed by bilateral audiometry in a sound-deadened environment with relevant bone-conduction masking and cross-sectional imaging.

The Tullio phenomenon represents a vestibular response to sound or sudden inner ear pressure change, and a positive Hennebert sign may be present. A suggestive clinical history and examination, including otoscopy, can be confirmed by PTA, impedance tympanometry, cervical and ocular vestibular-evoked myogenic potential, vestibular assessments, high-resolution computer tomography (CT) and magnetic resonance imaging (MRI).

In a patient with a patient with an otoscopically normal ear and a conductive hearing loss with no hyperacusis, autophony, aural fullness, pulsatile tinnitus and sound- or pressure-induced vertigo, the presumptive diagnosis would be otosclerosis.

Discovering an intact, mobile ossicular chain during exploratory tympanotomy for presumptive otosclerosis is rare, and a stapedectomy should not be performed. Following exploration of such ears, a CT scan might suggest that the cause of the conductive hearing loss could be attributed to a semicircular canal dehiscence and a 'third window syndrome' without balance symptoms. However, a much more likely explanation would be inaccurate audiometry in a non-sound deadened environment or carried out without appropriate masking.

CONCLUSIONS

- The air–bone gap averaged over 0.5, 1, 2 and 4 kHz in pure tone audiometry is the sole means of assessing the magnitude of a conduction defect. It is important to have accurate masked bone-conduction thresholds to assess this. Their interpretation also requires knowledge of what audiometry measures, as accurate masking of the non-test ear is always required.
- Tuning fork tests and acoustic reflexes have no role when there is otoscopic evidence of middle ear pathology that can cause a conductive defect. There will be a conductive hearing loss whose magnitude can only be measured by pure-tone audiometry.
- Knowledge of the physiology of sound conduction is essential to correlate the otoscopic findings with the audiogram. This is particularly important if reconstructive surgery is being considered.
- If the bone-conduction thresholds are better than the air-conduction thresholds, as evidenced by an air–bone gap of 10 dB or greater over 0.5, 1, 2 and 4 kHz, there is a potential

defect in the sound-conduction mechanism of the external and/or middle ear.

- Bone-conduction thresholds are initially assessed not-masked. When there is a potential air–bone gap in an ear, the bone-conduction thresholds need to be reassessed with masking of the other ear in all cases.
- Otoscopy will diagnose the majority of aetiologies causing a material air–bone gap, confirmed by relevant bone-conduction masking, which in the adult population are variants of otitis media.
- In the presence of an otoscopically normal and mobile tympanic membrane, with a material air–bone gap confirmed by relevant bone-conduction masking, the presumptive diagnosis is otosclerosis. No other audiometric investigation is sufficiently specific to confirm this diagnosis. Radiological scanning has currently at least a 10% false-positive and a 10% false-negative rate.
- Within the adult population, in those with a hearing impairment of at least 25 dB HL in their poorer hearing ear, one third (33%) will have and air–bone gap of at least 15 dB suggestive of a middle ear conduction defect.
- A sensorineural hearing impairment is present if the bone-conduction thresholds are similar to the air-conduction thresholds and the average of the latter (usually over 0.5, 1, 2 and 4 kHz) is poorer than a certain level which defines not impaired hearing, usually 25 dB HL.
- A mixed hearing impairment is present if the average bone-conduction thresholds satisfy the definition for a sensorineural impairment and there is in addition an air–bone gap of 10 dB or greater.
- When the air-conduction thresholds in the poorer hearing ear are worse than the bone-conduction thresholds in the contralateral ear by 40 dB or more, the air-conduction thresholds could be a shadow of these bone-conduction thresholds and must be masked.
- The bone-conduction thresholds do not solely measure inner ear function. In a normal ear and in those with a sensorineural impairment, there is also a material air conduction component transmitted via the external canal and ossicular chain.
- In those with a conductive impairment, the bone-conduction thresholds do not have this air conduction component. This Carhart effect occurs at all frequencies but is greatest at 2 kHz.
- An air–bone gap of 15 dB or greater, with an appropriately masked contralateral ear and in the presence of a normal, mobile tympanic membrane with no evidence of otosclerosis on a CT scan, can alert the clinician to the possibility of a third window phenomenon, if the scan is suggestive of the possibility of a superior semicircular canal dehiscence.

FURTHER READING

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5

Behavioural hearing testing in children

OVERVIEW

While older children can have their hearing tested by means of pure tone audiometry (PTA) in exactly the same way as adults, for young children we must tailor the test to their developmental abilities. This imposes certain fundamental limitations on what we are able to test and it is essential to understand what these limitations are. First, we must consider how the tests are done.

Age Versus Developmental Stage

The specific test that we use will depend on the child's age and development. Some tests require the child to sit unsupported and turn, others require concentration and cognitive skills that come with time. We often refer to *age-appropriate hearing testing* in children but it would be more accurate to call it *developmental stage-appropriate hearing testing*, because, of course, not all children develop at the same rate. Children with learning difficulties or motor delays may be suited to hearing tests that match their specific abilities rather than their chronological age.

Testing Environment

Ideally, children's hearing testing should be done in a large (6 m × 4 m) sound-dampened room in order to minimise background noise. If testing is to be done in an ordinary room in, for example, a community health centre then the level of background noise should be taken into consideration when interpreting the results. A substantial elevation of thresholds, particularly for lower frequencies, is commonly found with normal levels of ambient noise in such environments. Lighting should be diffuse: shadows can give visual clues to the child that can easily be mistaken for responses to sound.

Conditioning

Most tests begin with a *conditioning phase*, where the child is taught to associate performance with some behavioural response to the presence of the sound stimulus. For example, they may be taught that if they turn when they hear the sound, they will be rewarded with the appearance of a toy that lights up and moves. During this phase, the sound stimulus is presented at supra-threshold levels and the child is encouraged to perform the behavioural response every time the sound is present. Once the tester is satisfied that the child is responding consistently, the test moves on to the *testing phase*. This is where the sound is presented at much quieter levels, to determine the minimum sound intensity which produces a behavioural response. During the testing phase, it is important that the child receives no cues other than the sound stimulus such as eye gaze or parental movement. Sound stimuli are presented in a similar way to PTA: down in 10 dB increments, up in 5 dB increments, and 2 out of 3 correct responses at a certain sound intensity being taken as positive.

It is appropriate here to refer to *minimum behavioural response levels* rather than thresholds as young children will not necessarily produce a behavioural response at their true threshold (the very quietest sound that they can perceive).

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Young children tend to respond at an average of 10 dB above their true threshold. It varies according to the frequency of the sound stimulus (more like 15 dB for low-frequency sounds and 5 dB for high-frequency sounds) (1). The exact value will vary between children, and in the same child on different occasions, and it can be anywhere from 5 to 25 dB, so a young child's threshold cannot be known with any certainty on behavioural testing alone. This is the first of many fundamental limitations of behavioural testing in children that we must bear in mind. It is usual to test down to 25–30 dB and then stop: if responses are obtained at this level then the child's true threshold is likely to be in the normal range.

Test Stimuli

Most children will accept headphones for hearing testing from 4–5 years of age. Below this age, hearing testing is usually performed with sound presented by a loudspeaker in the room. The room should be adequately sound-dampened, and the sound source (the loudspeaker) should be calibrated to produce a certain dB SPL at a reference point calculated to be approximately at the centre of a child's head (with the child sitting on the parent's lap). This is referred to as *sound field* presentation.

This obviously means that the sound is being presented to both ears simultaneously, and *the sound will be heard by the better hearing ear regardless of which side the sound is presented from*. This means that we are unable to obtain responses for each ear separately and we can test only binaural hearing. This is another fundamental limitation of behavioural testing in children. To reflect this, it is necessary to plot the results on an audiogram chart using symbols which clearly show that the ears are being tested binaurally (**Figure 5.1**).

The sound used is usually a frequency-modulated pure tone (usually known as a *warble tone*) partly because it is much more pleasant to listen to, but also because pure tones are more likely to set up standing waves of reverberation, even in rooms with sound dampening.

A bone conductor can also be used as a stimulus, even in the youngest children. Masking, however, is time-consuming and cognitively demanding. For these reasons, *it is unusual to be able to obtain any masking of air- or bone-conduction thresholds until at least age 6*, and often much older. This is another very important limitation of testing in children.

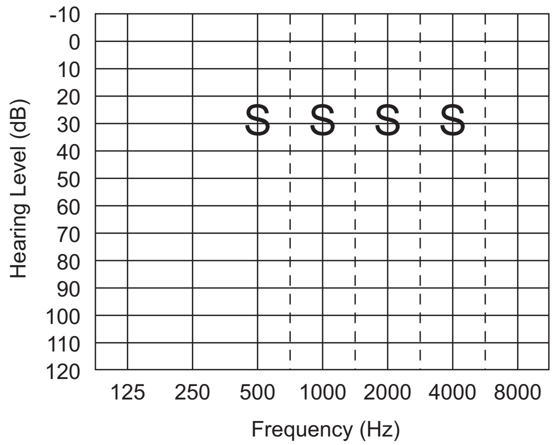

Figure 5.1 Example of a sound field visual reinforcement audiometry result. Note that responses are not plotted for each ear as the sound is presented by loudspeaker and therefore the ears cannot be tested separately. Minimal behavioural responses of 30 dB have been obtained, indicating that the child's true thresholds are likely to be within the normal range in at least one ear.

A child's attention span may be short, and it is important to gain as much useful information as possible in the limited time available.

VISUAL REINFORCEMENT AUDIOMETRY

Visual reinforcement auditor (VRA) is the most common test for children aged 8 months to 2 years. The minimum age is determined by the need for the child to be able to sit independently and turn their head. The maximum age is determined by the child being more interested in the testers and losing interest in the simple nature of the reward and therefore no longer responding reliably. The test is performed in a sound-dampened room with the child sitting on the parent's lap, slightly forward and gently supported at the waist. Two testers are required. Tester 1 is out of sight of the child and their role is to control the audiometer, present the sound, judge the child's response and present the reward. Tester 2 sits at a small table in front of the child and engages their attention with a selection of toys. Warble tones are presented from a loudspeaker at 90 degrees to one side. The child is conditioned to associate the sound with a visual reward: if they turn when they hear the sound, a toy in a cabinet next to the loudspeaker lights up and moves. Sounds are presented at up to 4 frequencies (0.5, 1, 2 and 4 kHz) until minimum behavioural responses are recorded, and responses at 30dB are taken to be normal (*see* **Figure 5.1**). Responses to bone conduction may also be obtained. A typical room set up is shown in **Figure 5.2**. Note that there are loudspeakers and reward cabinets on both sides but this is just to allow for testing at a child's preferred side. There is no value in testing a child with sounds presented from both left- and right-sided loudspeakers: this does not provide individual ear information.

PLAY AUDIOMETRY

Between 2 and 4 years of age, it is possible to condition children to perform a simple task in response to sound, such as placing a ball in a basket or a wooden peg in a board (**Figure 5.3**). Initially, this is done using loudspeakers in the room, but by age 4–5 years it should be possible to encourage the child to accept headphones at which point the test is essentially the same as a PTA. With headphones, individual ear information can be obtained. The test is usually limited by the child's attention span, so only 0.5, 1, 2 and 4 kHz are tested. Bone conduction may be used but masking is not usually possible at this stage.

DISTRACTION TESTING

The distraction test has largely been replaced with VRA, but it might occasionally be used in community settings or resource-poor environments which lack the facilities for VRA. It is suitable from age 8–18 months. The set up is shown in **Figure 5.4**. The child sits on the parent's lap as for VRA. The distractor engages the child's attention with a toy, then covers the toy at the same moment that the tester presents the sound. Sound is presented by the tester using either a calibrated hand-held warble tone generator, or both a high-frequency rattle

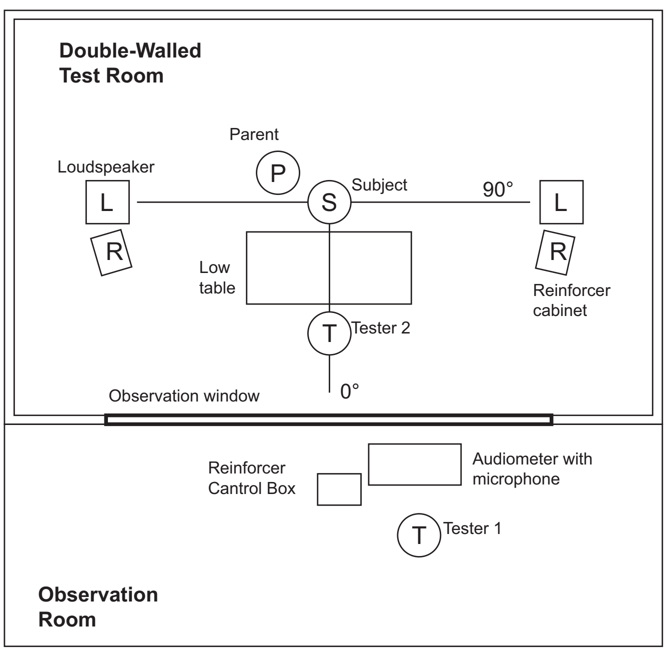

Figure 5.2 Typical room set up for visual reinforcement audiometry.

Figure 5.3 Play audiometry. The child is placing rings on a peg in response to sounds. (Photo with parental consent, courtesy of NHS Greater Glasgow and Clyde Medical Illustration Service.)

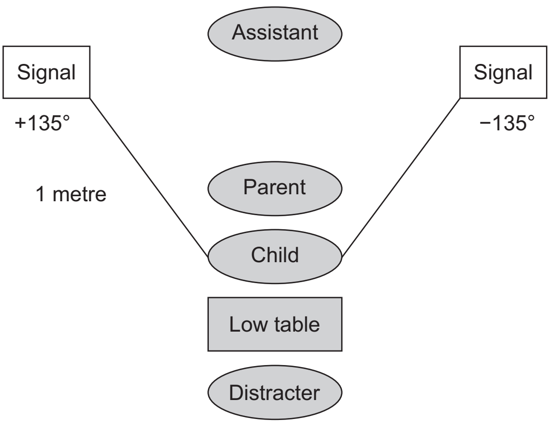

Figure 5.4 Typical set up for the distraction test.

and the tester's voice (both of which should be calibrated beforehand with a sound pressure metre). The sound is presented at the level of the child's ear, 135 degrees behind the child (so as not to be seen in the child's peripheral vision) at a distance of 1 m. A positive response is when the child turns to the sound. The distractor judges the child's response and rewards the child with praise.

COMMON PITFALLS IN BEHAVIOURAL TESTING IN CHILDREN

- Attempts to condition using sub-threshold sound intensities or failing to establish clear responses to conditioning before moving on to testing.
- Child is checking for visual reward without hearing the sound stimulus.
- Child responds to visual cues such as shadows or mirrors.
- Auditory cues (squeaky shoes, audible switches, rustling clothes, footsteps).
- Olfactory cues (tester's perfume, for example).
- Unconscious prompts from the parent holding the child.
- Child lacks sufficient head control to sit unsupported and turn.
- Child is more interested in people and environment than the toys.
- Child anticipates the sound presentation. The interval between sound presentations should be varied so as not to settle into a predictable rhythm.
- In distraction testing, the child may learn that the sound will appear when the distractor stops playing and covers the toy. No-sound controls should be performed to check for this: the distractor covers the toy but no sound is presented. This does not apply in VRA because the second tester does not stop playing with the toy when the sound is presented.
- Tester bias and expectation of normal results leading to non-definitive responses (such as checking) being classified as true responses.

UNDERSTANDING THE LIMITATIONS OF BEHAVIOURAL TESTING IN CHILDREN

Figure 5.5 shows a series of audiometric tests on the same child at different ages. In **Figure 5.5a** we see the VRA result at age 1 year. This is a normal result, but we should bear in mind what that means in the context of a test done with loudspeakers in the room. The minimal behavioural responses at 30 dB imply that there is hearing within the normal range (but we can't say exactly what the true threshold is) in at least one ear, and we can't know if that is left, right or both.

In **Figure 5.5b** we see the play audiometry done with headphones at age 4 years. This is the first time the child has given us information for each ear separately, and we can see that there is clearly a difference between the two ears. At first glance, it looks like there is a conductive hearing loss of around 55 dB and the unwary surgeon may attribute this to otitis media with effusion. However, the child is too young for the time-consuming and cognitively challenging business of masking, so the first thing to note is that, without masking, the bone-conduction thresholds are meaningless and add no information at all, given the normal hearing in the right ear. The size of the apparent air–bone gap is also larger than is usual for middle ear fluid. The air-conduction thresholds in the left ear

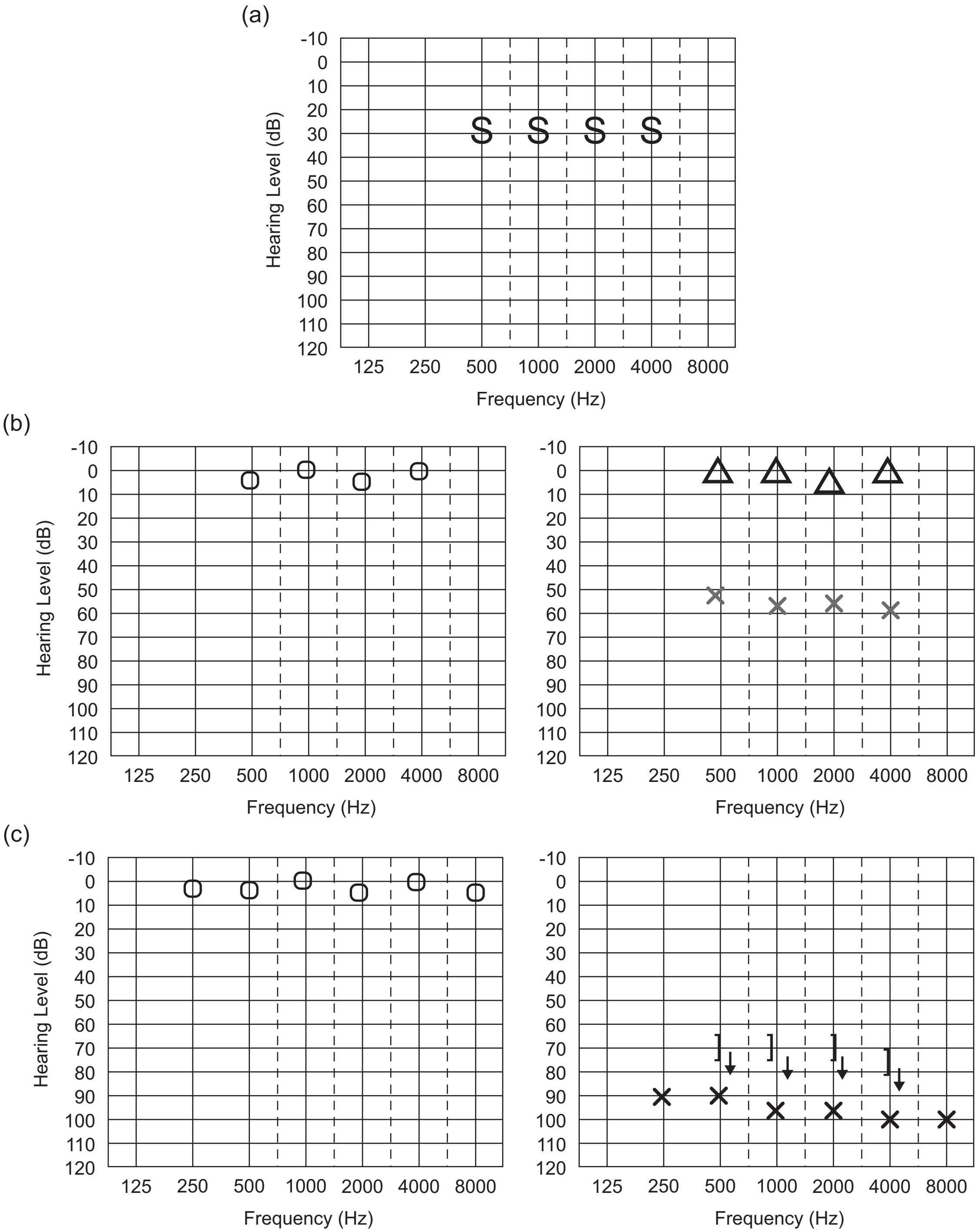

Figure 5.5 **(a)** Visual reinforcement audiogram, age 1 year. **(b)** Same child: play audiogram with headphones, age 4 years. **(c)** Same child: pure tone audiogram, age 8.

are not masked so all we know for sure is that the hearing on the right is normal and there is a hearing loss of at least 55 dB on the left. This could be conductive, sensorineural or mixed.

Figure 5.5c shows the child's PTA at age 8, when they are able to cooperate with masking for the first time. It is now clear that there is a profound left sensorineural hearing loss. MRI shows a cochlear abnormality on the left, consistent with the hearing loss having been like this from birth.

Note that at no stage was the testing at fault here, it is simply an illustration of how the fundamental limits of behavioural testing in children mean that we have to deal with imperfect and incomplete information in pre-school-age children.

CONCLUSIONS

- VRA is suitable for typically-developing children aged 8 months to 2 years.
- In VRA, sound is presented through loudspeakers so individual ear information cannot be obtained.
- Children may accept headphones from age 4 years, allowing the ears to be tested separately.
- Children may allow testing with masking from 6–8 years.

FURTHER READING

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6

Speech audiometry

OVERVIEW

The pure tone audiogram (PTA) is the basis of audiometric testing because it presents, in a very clear way, the frequency and intensity of sound which the subject can detect. It is accurate and reliable, well-standardised and relatively straightforward to interpret. However, the human auditory system did not evolve to hear beeps. The PTA does not measure the thing which really matters: the ability to hear and understand speech.

Various forms of audiometry have been devised which use speech sounds as the stimulus. They have the advantage of *ecological validity*, which is another way of saying they represent something important and useful in the real world. Pure tone thresholds do not predict the individual's ability to understand speech in the presence of background noise. It has been suggested that no assessment of hearing is truly complete until some form of speech assessment has been done. There is some evidence, for example, that speech audiometry is better than pure tone audiometry at predicting the extent to which an adult with hearing loss perceives their hearing disability (1) and their benefit from hearing aids (2), or a child's hearing disability from otitis media with effusion (3).

There are a few drawbacks to speech audiometry, however, which we need to understand.

The first is that almost all the work on developing and validating speech audiometry has used English as the test language. It is clearly inappropriate to use English language sounds to test someone for whom English is not their first language. Developing and validating speech audiometry for languages other than English is not simply a case of translation.

Many languages have very different structures in terms of the range of phonemes used and how they are emphasised. Mandarin Chinese, for example, is a tonal language so word lists for speech testing need to include a balanced range of tones as well as consonant and vowel sounds. Spanish speakers are more familiar with two-syllable words where the first syllable is stressed, and less familiar with ones where both syllables are stressed equally: this can affect speech recognition scores depending on the nature of the words used. Readers from around the world should bear in mind that the information in this chapter is based on English language testing. Even for native English speakers, accent and dialect can affect word recognition. Caution must be taken if recordings of American English are used in the UK, for example.

The second drawback is that it can be difficult to standardise the speech stimulus. Sound intensity is not so easy to define when dealing with the complex sound of a speech syllable, as compared to the simple sine wave of a pure tone.

The third drawback is that clinicians are often much less familiar with speech audiometry and how to interpret it for decision making in clinical practice. The purpose of this chapter is to address this to some degree.

TYPES OF SPEECH STIMULUS

Testing can be done in a quiet background, or with background noise (*speech-in-noise testing*). The background noise used is usually some form of incomprehensible speech babble. Speech sounds can be presented as:

- *Phonemes*, which are defined sounds equivalent to a syllable such as 'sss' or 'oo'.

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- *Spondees*, which are two-syllable words where each syllable is stressed equally, such as 'hotdog'.
- *Word lists* which have been selected for phoneme balance and reliability in scoring. Examples used commonly in the USA are the Northwest University Auditory Test 6 (NU-6) and the Central Institute for the Deaf W-22 (CID W-22) which are both monosyllabic word lists. In the UK and Australia, the Arthur Boothroyd (AB) word list is common. It consists of 15 lists of 10 words, each of which has a consonant-vowel-consonant (CVC) structure.
- *Sentence lists* are used or speech-in-noise tests. Examples include the City University of New York (CUNY) list and the Bench-Kowal-Bamford (BKB) list.
- *Long narrative passages of test* can be useful for determining comfortable listening levels (see below).

Speech stimuli are presented by headphones to test each ear in turn. Masking of the non-test ear may be required if there is more than 35 dB of asymmetry in bone-conduction thresholds.

For speech-in-noise testing, sentence lists are presented with a different speech-to-noise ratio for each sentence. Most people with normal hearing thresholds in both ears need speech to be about 2 dB louder than background noise to correctly identify 50% of words and sentences. A hearing impaired person might need, for example, 12 dB above background to get the same 50% score, and this might suggest that they would benefit from additional help such as directional microphones on their hearing aids.

Table 6.1 Word recognition score, showing that the subject got 90% of words from the NU-6 word list correct at 55dB

	WR1
Transducer	Phone
Intensity	55
Masking	
Score	90
Aided	
Wordlist	NU-6 LIST 1a

PRESENTATION OF RESULTS

The simplest tests to do are those using narrative passages of text. The tester adjusts the loudness of the voice and the test subject indicates the *most comfortable listening level* and the *uncomfortable listening level*. These can be very useful for programming of hearing aids.

The *speech recognition threshold* is measured with spondees, and is tested for each ear separately. It is the sound intensity where the subject can correctly identify 50% of the words presented. It is useful as a cross-check against pure-tone thresholds, and as a starting point for more detailed testing such as speech-in-noise.

Word recognition scores can be presented in different ways. In the USA, it is common to present a single percentage score representing the proportion of words from a defined list (usually 50 words but may be abbreviated to 25 words) which have been correctly identified at a single intensity level, usually 20–40 dB above the speech recognition threshold (**Table 6.1**). This is then graded excellent (90% or more), good (78–88%), fair (66–76%), poor (54–64%) or very poor (less than 52%). In the UK, it is more common to test across a range of intensity levels and to present the result as a graph or percentage word recognition against sound intensity known as a *speech audiogram* (**Figure 6.1**).

SPEECH TESTING IN CHILDREN

A range of tests is available depending on the age and developmental abilities of the child. The child's language skills and vocabulary are obviously very important to consider when selecting the most appropriate test, as well as motivation and concentration.

Ling sounds are used for testing children aged 6 months to 3 years. These comprise six phonemes ('ah', 'ee', 'oo', 's', 'sh' and 'um') which have been chosen to cover the frequency range of speech sounds in English. For the younger child, the sounds may be presented by sound field, with a response being a turn towards the loudspeaker. For older children, presentation may be by headphones and the response may be to copy the sound or to point to a picture that the child has been taught to associated with each sound (a snake for 'sss', for example). Testing can be done with or without

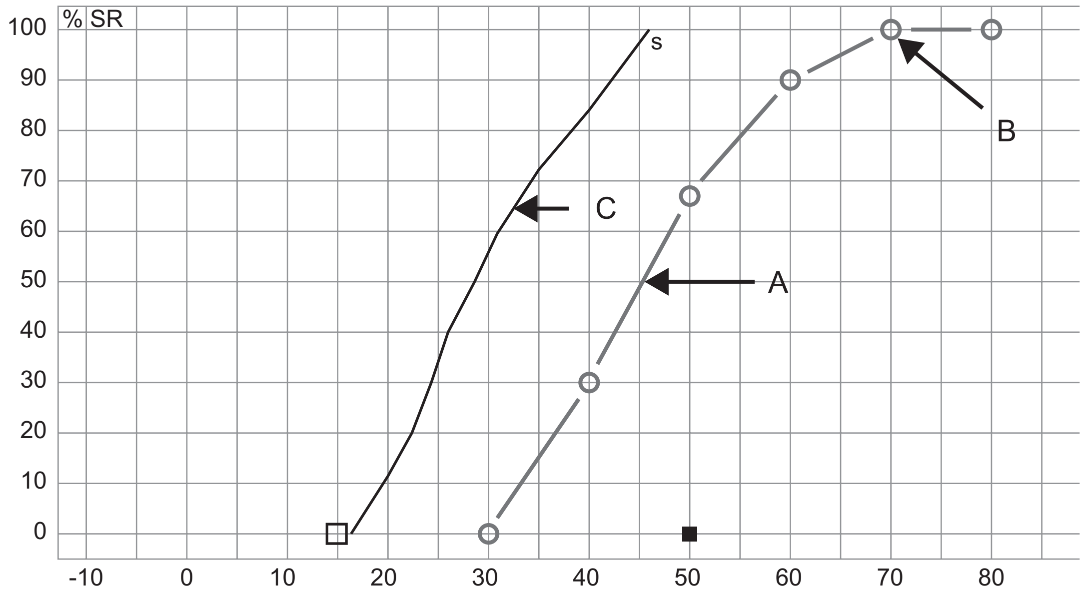

Figure 6.1 Example of a speech audiogram showing word recognition scores as a percentage on the y-axis, and sound intensity on the x-axis. Point A is equivalent to the speech recognition threshold (50% correct). Point B is the maximum word recognition score. The line marked C is a reference line showing word recognition in normal hearing individuals for the specific condition under test, which in this case is the AB word list presented via headphones. Other test conditions will produce a different reference line.

hearing aids, and sound can be presented at various intensities, although the purpose is not to establish 'thresholds' but to provide a functional assessment of what the child can and cannot hear to guide amplification.

The *CID W22 word list* can be used from age 8 years and the NU-6 word list from age 12. Speech-in-noise testing using the BKB sentence list can be done from around age 5 years.

The *McCormick Toy Test* can be used by children as young as 2 years of age, as long as they recognise (or can be taught) the names of the toys (**Figure 6.2**). There are 14 toys, comprising 7 pairs. Each toy in the pair has the same vowel sound but different consonants, giving some limited frequency information. The word pairs are tree/key, shoe/spoon, cow/house, plane/plate, horse/fork, duck/cup and man/lamb. The child must name 4 out of 5 toys correctly to pass. The child is asked, 'show me the . . .' with the tester's mouth hidden to prevent lip-reading. The tester's voice should be calibrated with a sound pressure metre at 40 dB, and a normal-hearing child should pass at this level.

The *Manchester Picture Test* is used for older children (**Figure 6.3**). It comprises six lists, each of six target words. For each target word, a card with four pictures on is presented to the child, the pictures representing the target word plus three distractors (for example, queen, three, feet and bee, with bee being the target word). The child is asked to identify the correct picture. The tester's voice should be used at 40 dB for screening, and the child must get 5 out of 6 correct to pass. The test can also be used to determine the speech

Figure 6.2 The McCormick Toy Test (with permission from Soundbyte Solutions).

Figure 6.3 Manchester Picture Test (with permission from Soundbyte Solutions).

recognition threshold by starting at 20 dB above the pure tone average and either increasing or decreasing the level until the lowest level is found at which the child correctly identifies 5 out of 6 target words.

SPEECH TESTING IN ADULTS

It is common for adults with an acquired hearing loss (e.g presbyacusis, noise-induced hearing loss, progressive idiopathic loss) to report that they can hear speech but cannot understand what is being said.

This can be explained by considering the frequency spectrum of speech, which is usefully plotted on the Speech Banana (**Figure 6.4**). When high-frequency hearing is lost and lower-frequency hearing is preserved, listeners can hear speech, but miss consonant sounds which fall into the higher frequencies, and which are essential for the recognition of speech. As mentioned above, this effect can be worsened by background noise, which serves to mask the sounds the listener needs to hear in order to understand speech (4).

Speech testing has many potential uses in adult audiology, for diagnosis and monitoring of hearing function, some of which are detailed below.

Hearing Aid Assessment

Speech recognition scores can be measured before and after hearing aid provision, to measure a listener's functional ability, and to identify the most appropriate strategy for their hearing rehabilitation (5). Speech recognition, especially in noise, can be a useful guide to the likely success of amplification and can guide decisions such as amplification adjustments (6) and the use of directional microphones. Listeners who undergo speech in noise testing as part of their hearing aid fitting and rehabilitation are more likely to have higher measures of hearing aid satisfaction (7).

Identification and Monitoring of Specific Conditions

Speech testing is an important component of the diagnostic test battery, which can help identify or monitor specific conditions.

Listeners with suspected non-organic hearing loss may produce results which are inconsistent with their pure tone audiometry, such as getting alternate words wrong. Listeners with auditory processing disorders may have unexpectedly poor speech recognition, especially in noise.

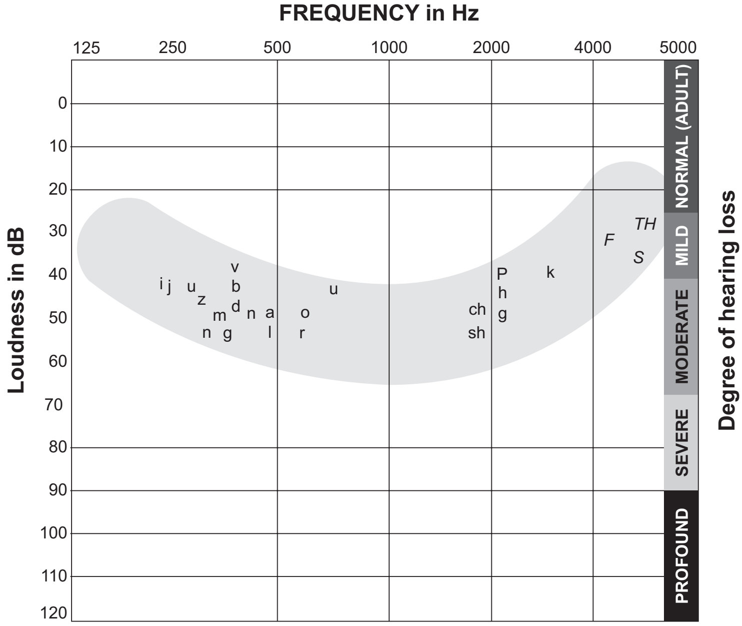

Figure 6.4 'The speech banana' showing the range of average human speech.

Other conditions where speech recognition can be useful include otosclerosis, where the 'paracusis of Willis' can cause speech recognition which is better in background noise than in quiet. Speech discrimination scores can be paradoxical in conductive hearing losses, whereas they inevitably decline when there is a sensorineural component to the loss.

Patients with vestibular schwannoma may have speech recognition scores which are worse than might be expected from their PTA. This is because their auditory nerve may have been stretched or compressed by their tumour; their intact nerve maintaining the recognition of sound, but distorting speech recognition. Speech testing can inform counselling and hearing rehabilitation in these patients, as well as surgical decision making, particularly regarding surgical approaches to tumour excision, and likely outcomes from auditory nerve preservation surgery, with or without cochlear implantation.

Cochlear Implant Candidacy

Speech testing is integral to cochlear implant (CI) candidacy assessment in the UK, though this is not always the case across the globe, where practice can vary (8). There may be several reasons for this including established practices and structure of services. However, preoperative level of speech understanding is one of the most valuable measures within the CI referral and candidacy assessment (8), and is therefore an important measure to take into account.

UK guidelines (NICE TA566) include speech discrimination thresholds as a criterion for adult CI candidacy, currently defined as a phoneme score of 50% or less on the AB word list at 70 dB in their best aided conditions (9). This was changed in 2019 from previous criteria, which mandated a score of 50% or less on the BKB sentence list. The basis for this decision was that listeners with good non-verbal communication skills are able to score well on the BKB sentence list, which renders them less likely to fall into CI candidacy. The AB word list was chosen as a more accurate measure of functional hearing.

Speech testing is central to the monitoring and rehabilitation of CI recipients after implantation in both adults and children, to assess progress, identify changes in function, and to guide approaches to auditory rehabilitation.

Outcomes Reporting

Speech recognition scores are commonly used for outcome reporting in auditory research, i.e. measuring the benefit of interventions such as hearing devices and implants, or auditory training programmes.

CONCLUSIONS

- Speech is a more 'natural' stimulus than pure tones for testing hearing in humans, but the results can be more difficult to interpret.
- Speech tests include the measurement of an individual's speech recognition thresholds in aided and/or unaided conditions; in quiet or in noise.
- Speech testing is most useful as a way to assess 'real-world' benefit from hearing aids and cochlear implants, and to monitor progress.
- Speech testing as part of a battery of audiological investigations can offer useful information regarding specific conditions, including non-organic loss, auditory processing disorders, otosclerosis and cerebellopontine angle lesions.

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7

Objective testing

OVERVIEW

So far in this book, we have considered various behavioural tests of hearing including pure tone audiometry (PTA), speech audiometry and behavioural tests in children. These are all *subjective tests of hearing* in that they all rely to some degree on the cooperation of the subject. In this chapter, we will consider *objective tests,* which do not require the cooperation of the subject.

Objective tests can be useful in many circumstances including testing of children who are too young for behavioural tests, people of all ages with learning difficulties, those who may be exaggerating their hearing loss for financial gain (e.g. in industrial noise compensation claims) and those who are feigning hearing loss for psychological reasons.

It is worth asking at this point, if objective tests exist, why do we spend time and effort on behavioural testing? The answer is that, while objective tests can give us useful information about the integrity of parts of the hearing pathway, they are not in themselves tests of hearing. Hearing is the conscious appreciation of sound and, as such, can only be tested by behavioural means. In other words, if you want to know if someone can hear something you have to ask them (or elicit some other kind of behavioural response). Objective tests have their place, especially in the diagnosis of pathology, but their results need to be interpreted alongside other sources of information to build a complete picture.

For the simpler tests described here (tympanometry, acoustic reflex and otoacoustic emissions), the testing is usually done by an audiologist with the interpretation of results being left to the referring clinician. The more complex electrophysiological tests (such as the auditory brainstem response [ABR], cortical auditory evoked potentials and auditory steady state audiometry) may be done by audiologists with particular training, or by audiological scientists and medical physicists, who will provide a report interpreting the results for the referrer.

TYMPANOMETRY

Basic Concepts

Tympanometry is a test of middle ear function which is quick and easy to perform and usually well-tolerated, even by young children. The probe consists of a loudspeaker, a microphone, an air pump and a pressure gauge, and there is a rubber tip to provide an airtight seal at the entrance to the ear canal **(Figure 7.1)**. Air is pumped in (or sucked out) to vary the pressure in the ear canal. A pure tone of 226 Hz is played into the ear canal via the loudspeaker, and the microphone detects how much of that sound is reflected back by the tympanic membrane. Of course, the tympanic membrane does not exist to reflect sound, but rather to collect the sound and transmit it onwards through the ossicles. If the pressure in the ear canal is raised, the tympanic membrane is pushed medially and stiffened, preventing it from working efficiently, leading to more sound being reflected back to the microphone. If the pressure in the canal is too low, the tympanic membrane is sucked out laterally and, again, more sound is reflected back. At the point where the pressure in the ear canal is the same as the air pressure in the middle ear, the system works at maximum efficiency, sound is collected by the tympanic membrane and less is reflected back to the microphone. This allows us to

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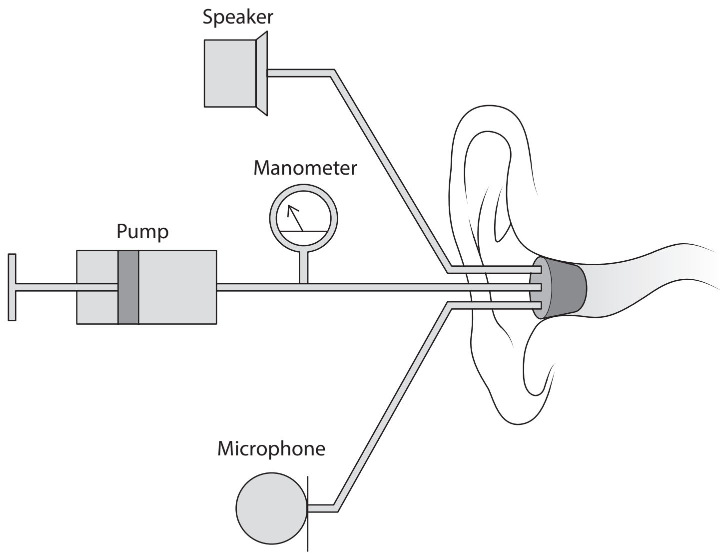

Figure 7.1 Schematic diagram of tympanometer.

indirectly measure the air pressure in the middle ear.

Classification of Tympanograms

If we plot the pressure in the ear canal against the amount of sound gathered and transmitted onwards by the tympanic membrane (the *admittance*), then there should be a peak in the plot corresponding to ambient atmospheric pressure. This is a type A tympanogram, as shown in **Figure 7.2**. If there is a peak in the trace at a negative pressure, this suggests poor aeration of the middle ear space due to dysfunction of the Eustachian tube. This is known as a type C tympanogram.

If there is no peak, just a flat trace, then this means that nothing we do to the pressure in the ear canal makes any difference to the admittance of sound. This is a type B trace. This could be because the middle ear is full of fluid (which is not compressible) or because there is a perforation in the tympanic membrane (so that no pressure change can be produced by pumping air into the canal). These two situations can be distinguished by the canal volume, which is automatically calculated by the tympanometer based on the volume of air that needs to be pumped in for a given change in pressure. A small volume (<1.1 mL) suggests an intact tympanic membrane and middle ear fluid. A large volume (>1.1 mL) suggests a perforation, with the volume measured being that of the ear canal and middle ear. A perforation may also be suggested by complete failure to achieve a seal at the ear canal despite trying various sizes of rubber tip. In this case, the instrument is measuring what is essentially an infinite volume comprising the ear canal, middle ear, an open Eustachian tube and, through it, the entire outside world.

The A-B-C classification which is commonly used across the world was described by Jerger in 1970 (1). Many modifications exist. One is to divide the type A (peaked) trace into A_S for a very shallow peak suggesting stiffness of the ossicular chain, as might occur in otosclerosis, and A_D, for a very tall peak suggesting excessive mobility, as might occur with ossicular discontinuity or a very thin, monolayer tympanic membrane. Another modification is to divide the Type C trace into C1 (–100 to –200 daPa) and C2 (–200 to −00 daPa) (2). More recently, with the development of extended low-pressure-range tympanometers, a C3 trace (–400 to −600 daPa) has been described although it adds very little to routine diagnostic assessment.

Advances in Equipment

Standard tympanometry uses a pure tone of 226 Hz. A pure tone of 1kHz gives more accurate

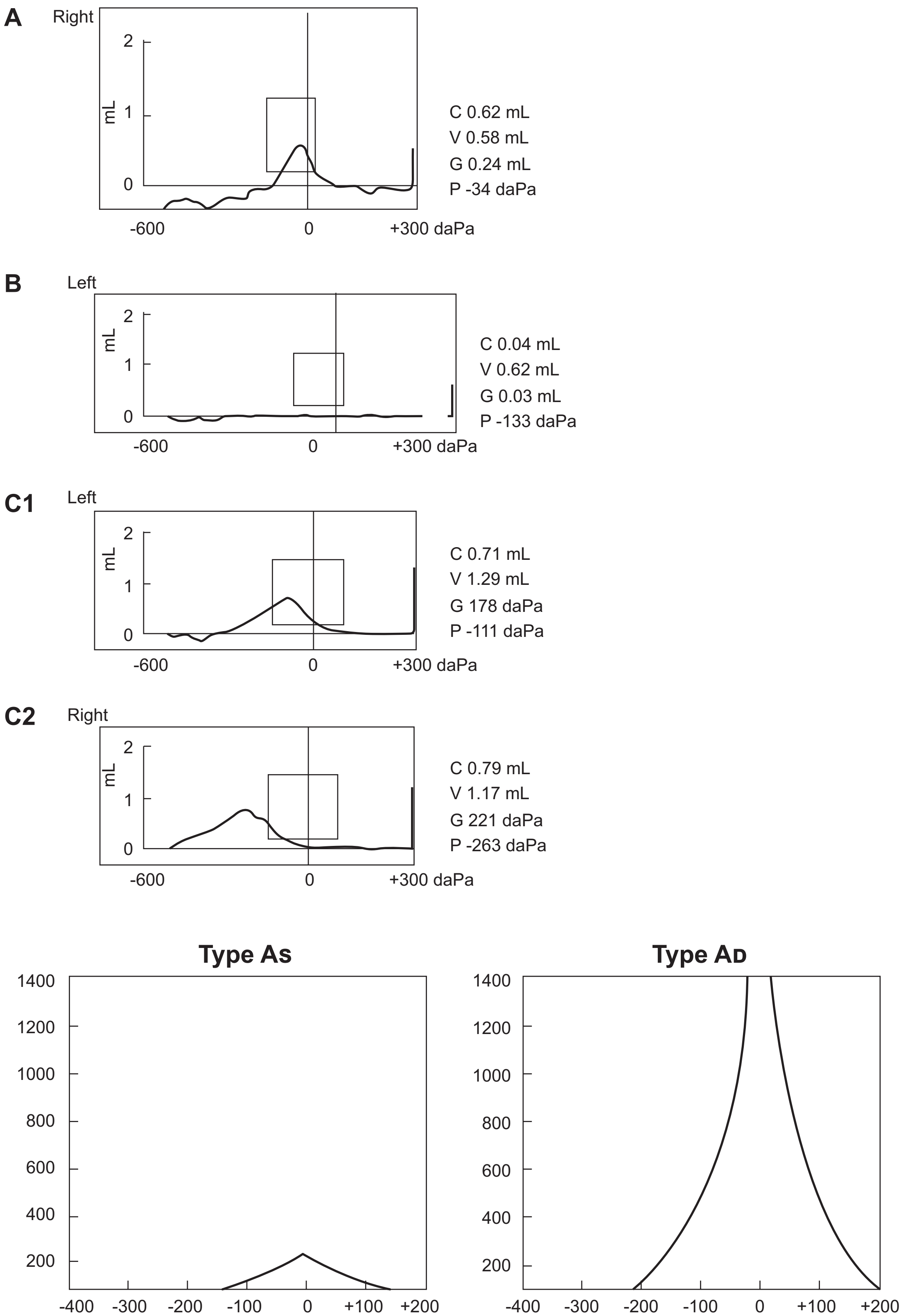

Figure 7.2 Classification of tympanometry traces. 2a Type A; 2b Type B; 2c Type C1; 2d Type C2; 2e Type A_S; 2f Type A_D.

results for infants under 6 months of age (3). The resulting traces are very different and the Jerger classification cannot be used. Broadly speaking, traces with a peak above baseline are considered normal and those with a peak below the baseline are abnormal suggesting middle ear fluid.

The use of multiple frequencies between 226 Hz and 8 kHz is known as *wideband acoustic immittance* (also as *wideband tympanometry* and *wideband absorbance*). It has not yet found widespread application, but its advocates suggest it may allow better identification of ossicular abnormalities and otosclerosis, and that it helps establish the presence or absence of middle ear fluid in children with conventional type C tympanograms.

Interpretation of Results: Otitis Media Effusion

In the outpatient clinic, a type A trace is taken to indicate a normal aerated middle ear, while a type B trace with low canal volume is taken to indicate middle ear fluid. This is a reasonable assumption in clinical practice but some diagnostic uncertainty exists. Several authors have compared the findings at myringotomy with the tympanogram type recorded immediately prior to surgery. The practical difficulty with this is that if no fluid is found, it could be claimed that it has drained down the Eustachian tube or into the mastoid air cells since the time of the examination. On the other hand, if fluid is found at operation it is unlikely to have developed since the time of examination. In children with suspected otitis media with effusion (OME), a type B trace with low canal volume has a sensitivity for fluid at myringotomy of 57–73% and a specificity of 84–93%, giving a positive predictive value of 89–96% (assuming around two-thirds of the population under study have OME) (4,5). However, the negative predictive value is only 47–64% (meaning the absence of a type B tympanogram trace does not rule out fluid any better than a coin toss) which is something of a limitation in clinical use.

Since 62% of ears with a C2 trace will have fluid at myringotomy, many research studies include B and C2 traces together to indicate the presence of fluid: this obviously increases the sensitivity at the expense of lower specificity and this goes some way to improving the low negative predictive value of the type B trace on its own.

In resource-poor settings where full behavioural hearing assessment is not possible for every child, it may be worth targeting audiometry at certain high-risk children based on tympanometry findings. Only testing the hearing of those children with bilateral type B traces will have a sensitivity of 90%, specificity of 69% and a positive predictive value of 60% for identifying children with 25 dB hearing loss or worse in their better ear (6). Including B and C2 traces together increases the sensitivity to 95%, but specificity falls to 43% and positive predictive value falls to 46%. In resource-rich settings, there is no excuse for routinely relying on tympanometry for surgical decision making and all children with suspected OME should have their hearing assessed.

Interpretation of Results: Otosclerosis and Ossicular Discontinuity

It might be expected that the compliance in ears with otosclerosis would be lower than normal because of decreased mobility of the ossicular chain (a type A_S trace), whilst the compliance in ears with ossicular discontinuity would be higher (a type A_D trace). This hypothesis has been studied by several investigators (including Jerger et al 1974) (7) **(Table 7.1)**.

Table 7.1 Tympanometric compliance in otosclerosis and ossicular discontinuity in comparison with the normal state

Pathology	n	Mean compliance	Percentiles 10th	Percentiles 90th
Otosclerosis	95	0.4	0.1	1.0
Discontinuity	19	1.9	0.8	>3.7
Normal	825	0.7	0.4	1.3

(After Jerger et al. [8]).

When compared with normal ears, those with otosclerosis have a lower mean compliance but there is considerable overlap in the levels as is evident from the 10th and 90th percentiles. Because of this, it has been calculated that in a clinic population the false-positive and false-negative rates are unacceptably high (8). If the level of compliance is taken which would correctly diagnose the majority (90%) of ears with otosclerosis, then 88% of normal ears would be incorrectly diagnosed as having the condition. Alternatively, if the level of compliance is taken which would correctly diagnose the majority (90%) of normal ears, then 62% of otosclerotic ears would be incorrectly classed as normal. Thus, tympanometry has no value in the diagnosis of otosclerosis in an individual patient.

THE ACOUSTIC REFLEX

The *acoustic reflex* helps protect the cochlea from damage due to loud sounds. It is also known as the *stapedial reflex* (only the stapedius muscle is involved in the protective reflex in humans, not the tensor tympani, whose main function seems to be stopping the tympanic membrane being blown out by sneezes and coughs). The pathway is from the inner hair cells of the cochlea via the cochlear nerve to the superior olive, from there to the facial nucleus, and then back along the facial nerve to the stapedius muscle in the middle ear. This stiffens the stapes in the oval window and reduces sound transmission to the cochlea by around 15 dB. The acoustic reflex is triggered by sounds of around 70–100 dB in people with normal hearing. This is elevated in those with conductive hearing loss. It can be measured by a standard tympanometer machine, and it may have some use in suspected non-organic hearing loss when the reflex is present at a sound intensity inconsistent with the degree of hearing loss on the audiogram. Other uses, such as locating the site of lesions within the facial nerve (proximal or distal to the nerve to stapedius) and differentiating cochlear from retrocochlear deafness in suspected vestibular schwannomas (using the acoustic reflex threshold as a marker of cochlear recruitment) are now largely historical and have been replaced by MRI scanning.

OTOACOUSTIC EMISSIONS

Basic Concepts

We saw in **Chapter 1** that the outer hair cells of the cochlea are motile and that they contract in response to sound in order to amplify the vibration of the basilar membrane. The remarkable thing about this is that *we can hear them move*. The sound of the outer hair cells contracting is called an *otoacoustic emission* (OAE, also known as the *cochlear microphonic*). About 60% of normal-hearing people produce spontaneous OAEs (SOAE). OAEs can also be evoked by sound stimuli and recorded. The OAE is measured with a hand-held probe connected to a laptop computer. The probe has a loudspeaker and a microphone, and a rubber tip to provide an acoustic seal at the entrance to the ear canal. The loudspeaker produces a stimulus noise, and the microphone measures, first, the same stimulus noise reflected by the tympanic membrane followed 3–8 milliseconds later by a new, distinctly different noise which is the OAE **(Figure 7.3)**.

If a single pure tone is used as a stimulus, the resulting OAE has the same frequency as the stimulating tone and can therefore be difficult to distinguish from it. For screening purposes, a more useful stimulus is a broad band click centred on the speech frequencies (0.5–4 kHz). This is known as a transient evoked OAE (TEOAE). Some frequency-specific information can be provided by using a pair of pure tones in a carefully calculated ratio: this produces an OAE at a different, lower frequency which can be predicted mathematically from the frequencies of the stimuli. This is known as a distortion product OAE (DPOAE) **(Figure 7.4)**. Because the DPOAE is at a different (but totally predictable) frequency from the stimulus tones it is easy to distinguish them.

Interpretation of Results

A TEOAE will be present in the majority of ears with hearing of 25 dB or better. In the sense that it gives a binary 'pass-fail' result (or, more accurately, 'pass-refer'), it is very well suited to use as a screening test for the newborn. It can also provide useful additional information in children of any age who are difficult to test behaviourally, as long as they have a normal (type A) tympanogram.

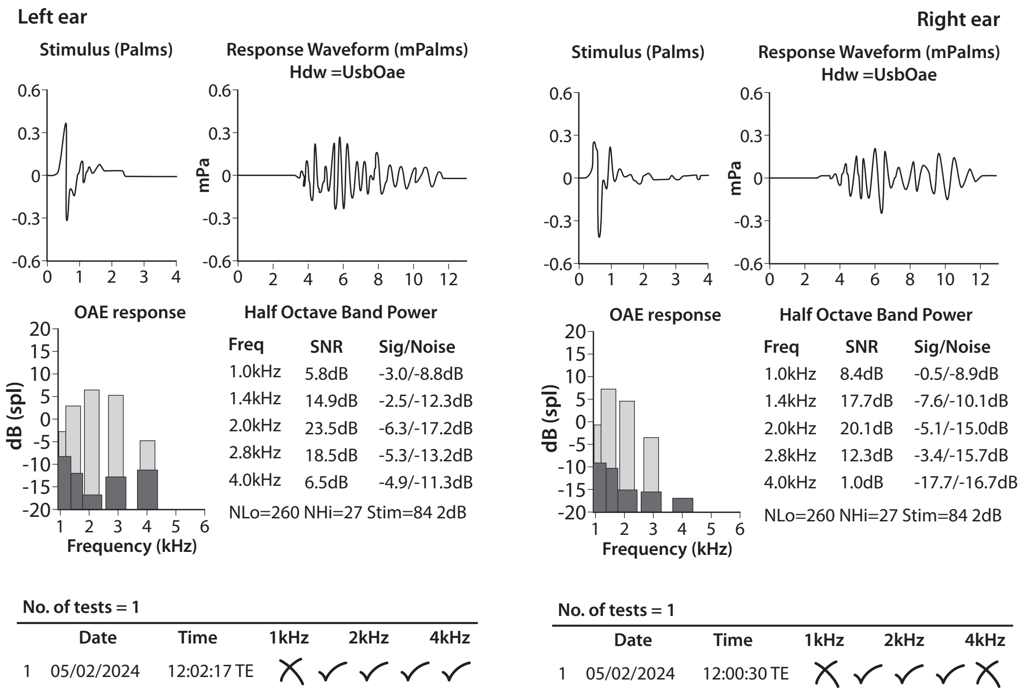

Figure 7.3 Typical TEOAE result using a broadband click stimulus of 1–4 kHz. The upper part shows the stimulus and response waveforms. The middle part shows the signal-to-noise ratio (SNR) at 5 frequencies in the 1–4 kHz range, which is key to determining whether the TEOAE can reliably be said to be present or not. An SNR of 3–6 dB is usually required. Where the SNR shows that the TEOAE is present, a tick is shown at the bottom of the chart: 4 out of 5 ticks and the result is 'pass'. Any less, and the result is 'retest required'. In this example, the left ear is 'pass' and the right is 'refer'.

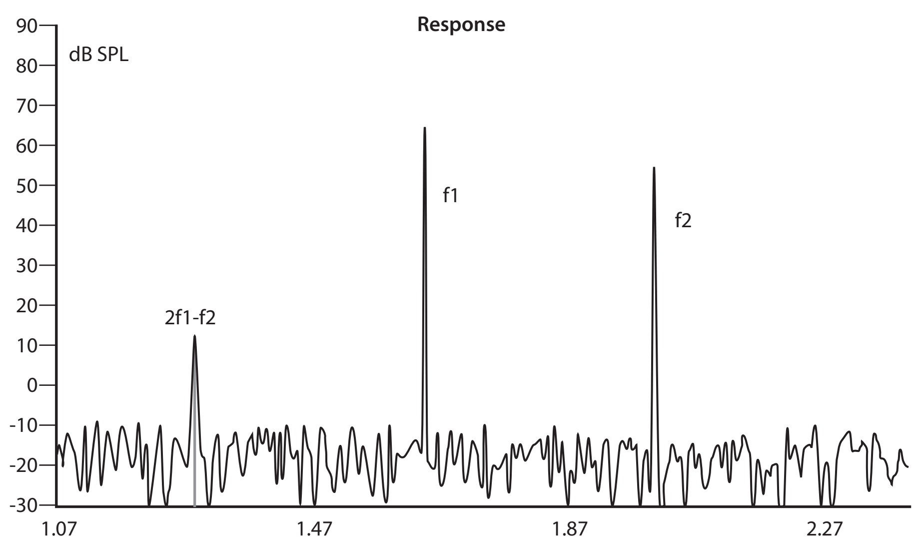

Figure 7.4 DPOAE frequency graph. The stimuli are pure tones at frequencies f1 and f2. The response is an otoacoustic emission at frequency 2f1–f2. It is generated by outer hair cells at a position on the basilar membrane that corresponds to f2, and therefore the presence of the DPOAE gives information on the function of the cochlea at this point. For example, if f1 is 1 kHz and f2 is 1.2 kHz, 2f1–f2 (the response) will be at 800 Hz. The magnitude of the response 2f1–f2 is at least 50 dB less than the f1 stimulus. The 2f1–f2 DPOAE is largest when the ratio of f1 to f2 is about 1.22 and the intensity of f1 is 65 dB SPL and f2 is 50 dB SPL.

In order to get a normal response, there needs to be an ear canal which is not occluded by anything, an intact tympanic membrane, an aerated middle ear, an intact and functional ossicular chain and a cochlea with functioning outer hair cells.

There are two important points to consider as a result of this. The first is that the most common reason for failure to record a normal OAE in the newborn baby is the presence of residual amniotic fluid in the ear canal or middle ear in the first 24 hours of life. The second is that there is no requirement for the cochlea to be connected to the brainstem by an auditory nerve, or for there to be a functioning brainstem at all. The presence of an OAE is not proof of hearing, only of cochlear outer hair cell function. We will consider the implications of these drawbacks when we discuss neonatal screening (see below).

AUDITORY BRAINSTEM RESPONSE

To record the *auditory brainstem response* (ABR, also known as *brainstem evoked response audiometry* or BSERA) three surface electrodes are placed on the skin (one on each mastoid and one on the forehead) and an electrical trace is recorded in response to sounds. The sound stimuli may be broad- or narrow-band clicks, chirps or pure tones and they can be played into each ear by means of inserting earphones or a bone conductor. The trace that is recorded has certain identifiable peaks and troughs corresponding to electrical activity in specific parts of the auditory pathway, namely the cochlear nerve (wave I), cochlear nucleus (wave II), superior olive (III), lateral leminiscus (wave IV) and inferior colliculus (wave V) (**Figure 7.5**). Wave V typically appears around 4–8 ms after the stimulus. Sound stimuli are initially presented suprathreshold to establish a clear response, and then at progressively quieter levels until the response is no longer discernible. As the sound stimulus gets quieter, the response waveform gets smaller in amplitude (so harder to distinguish from background noise) and also longer in latency. With a suitable small correction factor (specific to each brand of equipment and testing protocol) the quietest stimulus that still provokes an identifiable waveform provides a very accurate estimate of the behavioural hearing threshold at the frequency being tested. Note that electrical activity in the midbrain does not guarantee conscious appreciation of sound: ABR is not, strictly speaking, a test of hearing but in clinical practice it can be taken to be a very close approximation.

It should be obvious that the electrical activity in, say, the cochlear nucleus is very small indeed and it may seem surprising that we can detect it on the surface of the skin with electrodes. Skin

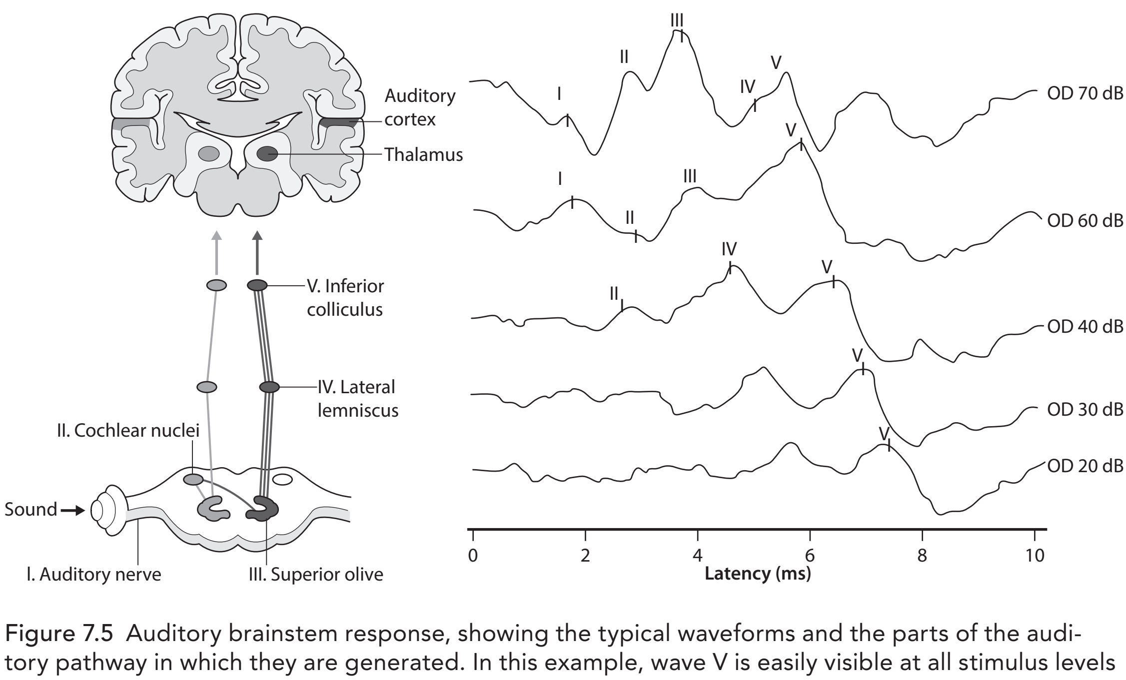

Figure 7.5 Auditory brainstem response, showing the typical waveforms and the parts of the auditory pathway in which they are generated. In this example, wave V is easily visible at all stimulus levels down to 20 dB suggesting normal hearing at the frequency under test. Note that as the stimulus level becomes quieter, the amplitude of the waveform decreases and the latency increases.

electrodes will pick up all manner of electrical activity, the vast majority of which is completely unrelated to hearing. This includes all the other brain activity (EEG), muscle activity from movement (EMG), the electrical activity of the heart (ECG), interference from electrical equipment and the mains supply and random noise. The following precautions are therefore used:

1. Testing is ideally done in a sound-dampened room. If testing is done in an operating theatre, background noise should be kept to an absolute minimum with particular attention to sources of loud, regularly-repeating noise such as pulse oximeters and other monitors.
2. The room should ideally be free from sources of electrical interference as far as possible and the ABR should be plugged directly into the mains socket, not an extension cord.
3. The test subject should be still and quiet. For adults, adequate testing may be done with them sitting in a reclining chair if they are able to settle and relax. If not, and the background EEG is too active, sedation should be considered.
4. Infants under 6 months can usually be tested under normal sleep, and this can be facilitated by feeding ('feed and sleep ABR').
5. Older children may be tested during sleep induced by melatonin, or by sedation, or under general anaesthesia.

Having minimised the effects of background noise, awake EEG, muscle activity from movement and electrical interference, we still have to find a way to identify the tiny electrical signal from the auditory pathway from the remaining EEG. This is done by means of *signal averaging*. The sound stimulus is presented around 1000 times and the EEG is recorded for the 20 ms after each stimulus. For each 20 ms sample, the signal (our ABR waveform) is the same in every sample but the background noise (the EEG which is unrelated to hearing) is randomly scattered. If these 1000 waveforms are added together, the signal adds up every time but the random noise cancels out. The SNR increases roughly in proportion to the square root of the number of measurements. In practice, as few as 800 samples may be required if the response is clear, whereas it may be advisable to record 2000 or more before declaring a response to be absent. For each frequency, the test is run twice to ensure the response recorded is genuine and repeatable.

The nature of the test stimulus depends on the clinical situation and the time available. ABR testing takes at least an hour. Adults will only sit still for a certain time, infants nap for short periods and, in the operating theatre, time is often at a premium. Decisions need to be made about priorities. In an ideal world, testing would be done initially with a broad band click to give an initial estimate of overall hearing, followed by pure tones at 0.5, 1, 2, 4 and 8 kHz with air and bone conduction for both ears and masking as required, but in reality this would take many hours and is impractical. The minimum required in most circumstances is testing with tones at 1 and 4 kHz. If these show clear responses at 20–30 dB, that is usually enough. Alternatively, a click centred on the speech frequencies (0.5–4 kHz) plus a tone at 8 kHz (to identify high-frequency sensorineural losses) gives a quick assessment of likely hearing thresholds. If only limited information can be obtained in the time available, the ABR may need to be repeated. In any event, it should be interpreted along with frequency information from DPAOEs (see above) and behavioural tests.

In adults, if the ABR results are better than expected from pure tone audiometry, non-organic hearing loss should be considered. In children, the main uses of ABR are for diagnosis, hearing aid fitting and cochlear implant assessment, as a supplement or alternative to behavioural tests, particularly for children who have limited cooperation. Caution is required when interpreting the ABR in preterm babies as the myelination of the auditory system is not complete until around 29 weeks' gestation and can sometimes be significantly delayed.

Automated ABR systems have been developed for use in neonatal hearing screening (see below). A quiet environment is still required and the electrode placement is the same, but much of the test procedure is automated such that it can be administered by someone who is not a fully qualified audiologist. The impedance of the skin electrodes is automatically calibrated to ensure they have been properly applied. The stimulus is usually a broadband click at 30–35 dB. The response waveform is identified automatically by the software and the result is presented as a simple pass/refer outcome. The whole test takes less than 5 minutes. If a clear pass is not obtained, the child will need to be referred to a paediatric audiology service for full diagnostic ABR with tone pips at various frequencies as described above.

CORTICAL AUDITORY EVOKED POTENTIALS

Cortical auditory evoked potentials (CAEP) are a measure of electrical activity in the cortex in response to sound and they can be used to estimate thresholds. Similar in principle to the ABR, recording the CAEP involves a quiet environment, insert earphones to provide sound stimuli, surface electrodes to record the resulting EEG and a computer to do the signal averaging. Unlike ABR, the test subject must be awake and alert for CAEP to be successful. Also, being further along the neurological pathway, the latency is much longer than that of the 4–10 ms of the ABR. The CAEP trace consists of two peaks separated by a trough: P1 at around 50–70 ms, N1 at 100–130 ms and P2 at 200–250 ms. The pathways concerned do not reach full maturity until adolescence so the main use of CAEP is in adults, particularly in the assessment of non-organic hearing loss.

AUDITORY STEADY STATE RESPONSE

Auditory steady state response (ASSR) testing is another electrophysiological test like ABR which can be used to estimate thresholds. A quiet environment, ear inserts, surface electrodes and a computer are required. As for ABR, the response is within the EEG. In contrast to ABR, however, the response is not measured as a waveform over time. The sound *stimulus is delivered constantly*, and the response is detected as a peak of activity in the EEG spectrum at a particular frequency. Exactly how this is determined varies between manufacturers, but it is objective and repeatable. When testing at a particular frequency (for example, 1 kHz) the 1 kHz sound wave is modulated at a second frequency (such as 80 Hz) and the neural *response is measured at the modulating frequency*, rather than the test frequency. In other words, the response in this case would be a peak of 80 Hz waves in the EEG spectrum, indicating that the 1 kHz tone has been detected. Modulation is the process by which a high-frequency wave (in our example, a 1 kHz sound wave) is varied either in amplitude or frequency in order to carry the properties of a lower frequency wave (80 Hz in our example) as shown in **Figure 7.6**. It is how TV and radio signals are broadcast. For ASSR, the test frequency is usually modulated in amplitude at around 82–106 Hz. A lesser degree of frequency modulation may also be used to enhance the response.

Multiple sound stimuli at different frequencies can be presented and tested at the same time, and both ears can be tested at once. For example, sounds may be presented at eight frequencies simultaneously (0.5, 1, 2 and 4 kHz, each to the right and left ear), as long as each test frequency is modulated slightly differently to allow the responses to be distinguished from one another. This dramatically shortens acquisition time compared to ABR.

The 90 Hz ASSR technique can be used at any age under normal sleep, melatonin sleep, sedation or general anaesthesia. In adults, the 40 Hz ASSR technique can be performed awake for suspected non-organic hearing loss.

Auditory neuropathy spectrum disorder (ANSD) is discussed in **Chapter 9**. The hallmark of ANSD is the presence of an OAE but an absent or abnormal ABR. The stapedial reflex is also absent at 95 dB. The ASSR may be present even in the absence of an ABR, so the ASSR cannot be used to estimate thresholds unless an ABR has been done first to exclude ANSD.

NEONATAL HEARING SCREENING

Prior to 2002 in the UK, there was a programme to test all babies at around 6–9 months of age using a behavioural test (the distraction test, performed, not by audiologists, but by nurses in the community). The results were poor with deaf children being diagnosed at a median age of 2 years old. Advances in technology led to rapid, reliable, objective testing becoming available towards the end of the twentieth century and universal neonatal screening became feasible.

Universal neonatal hearing screening was introduced in 2002 in the UK. The aim is to identify as soon after birth as possible all children with a permanent bilateral moderate-to-profound hearing loss. Other countries around the world have similar programmes, although the exact details may differ. In the USA, for example, the programme also aims to identify mild and unilateral losses. There are also important differences within the UK. In England, all children undergo OAE testing as a first line. If this shows a clear response in both ears, this is taken as a pass. If not, the child is tested with automated ABR as a second line. Again, a clear pass is required in both ears, otherwise the child

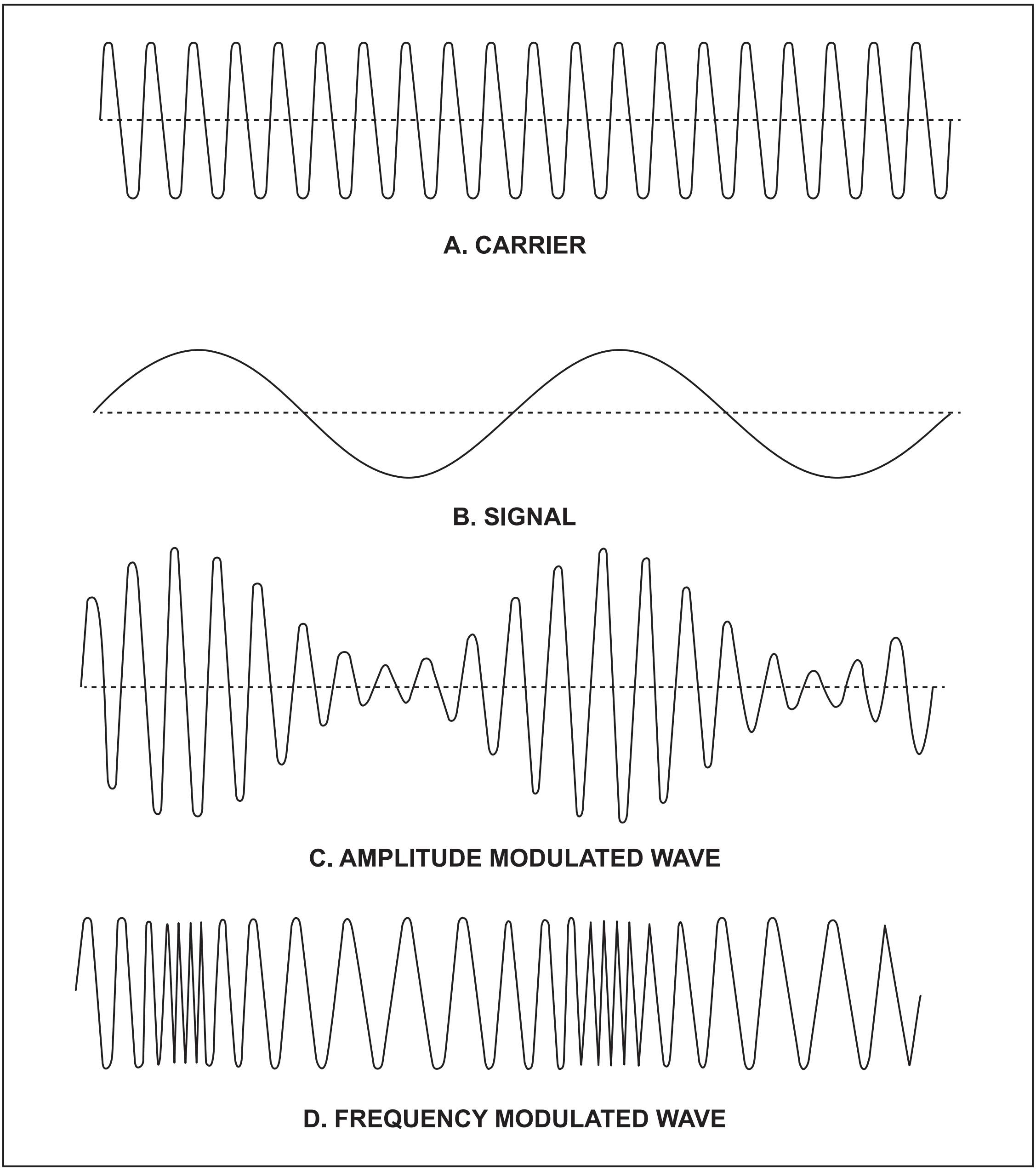

Figure 7.6 Frequency modulation and amplitude modulation, as used in auditory steady state response testing.

is referred to paediatric audiology services for full assessment including diagnostic ABR. In Scotland, automated ABR is used first and second line, and OAEs are not used in screening. Automated ABR has the advantage of detecting ANSD which might be missed by OAE screening.

Children who are deemed to be high risk for permanent hearing impairment, such as those who have spent more than 24 hours on a neonatal intensive care unit (NICO), are tested with both OAE and automated ABR at first line. Babies with microtia or meatal atresia, neonatal bacterial meningitis or meningococcal septicaemia, congenital CMV infection or programmable ventriculo-peritoneal shunts are all deemed too urgent to go through the screening pathway and are referred directly for urgent hearing assessment with diagnostic ABR.

Service models differ from place to place according to the features of the local population such as rurality and proportion of home births. In many places, first-line testing is performed by screeners in the maternity units. Referral rates are very high if OAE testing is done in the first 24 hours of life due to residual amniotic fluid in the ear canal and middle ear, but many babies are discharged home after short stays in the maternity unit so delaying testing beyond 24 hours after delivery means that many will have gone home before they can be tested. Where there are a lot of home births or where hospital stays after delivery are short, it may be more efficient to test in community settings, or to have a hybrid system with testing in both maternity units and the community. The aim is for all first-line testing to be completed ideally within 4–5 weeks of birth, and certainly within 3 months. Second-line testing is usually done in local community-based audiology facilities.

Impressive results have been obtained by the screening programme with screen coverage of well over 97% of live born children by 6 weeks and 99% by 3 months of age, a referral rate for diagnostic testing of around 2% and a median age of diagnosis of deafness of 2 months (9).

In more than 60 nations around the world, comprising more than one-third of the world's population, there is no hearing screening programme. The mean age at diagnosis of congenital deafness in such unscreened populations is 3–4 years of age (10).

CONCLUSIONS

- Objective tests are very useful for newborn hearing screening, in children too young for behavioural testing, in children and adults with learning difficulties and in those with suspected non-organic hearing loss.
- Tympanometry gives useful information on the middle ear.
- OAEs give useful information on the cochlea.
- Evoked response audiometry can give estimated hearing thresholds but it is time consuming and, in some circumstances, requires sedation or anaesthesia.
- None is truly a test of hearing.

FURTHER READING

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8

Otitis media with effusion

OTITIS MEDIA WITH EFFUSION IN CHILDREN

Background

Otitis media with effusion (OME) is perhaps the commonest disease in the western world, and yet many myths remain regarding its management (**Box 8.1**).

An effusion of fluid in the middle ear is part of normal experience: we are all familiar with having a 'blocked' sensation in the ears after an aeroplane flight or a viral upper respiratory tract infection. When a middle ear effusion persists for more than 3 months, we consider it pathological and we label it OME.

BOX 8.1: Common myths in OME in children

Myth 1: 'OME is a serious condition that needs to be treated'.

In most children, OME is a benign condition with few symptoms, which resolves spontaneously, and which is not associated with long-term speech or educational problems.

Myth 2: 'OME is caused by Eustachian tube dysfunction'.

Eustachian tube function is poorer in young children and improves with age, but at most it plays a small part in what is primarily an inflammatory disease caused by bacterial biofilms in the middle ear.

Myth 3: 'If there is fluid on tympanometry, the child should be treated'.

It is the presence of a hearing problem, not simply the presence of fluid, that should determine the need for treatment. Most OME is asymptomatic and self-limiting and does not need to be treated. Audiological testing should be routine for children with OME.

Myth 4: 'Ventilation tubes (VTs) are a treatment for OME. Hearing aids mask the problem but VTs make the OME go away'.

VTs are a way to provide short-term symptomatic relief of a hearing loss, similar in that sense to hearing aids. They do not modify the underlying tendency of the child to have OME, so when they extrude, the OME may recur. The child is not any less likely to have OME 1 or 2 years later because of the VTs. They have no effect beyond the day they extrude. Adenoidectomy is the only treatment we have which actually modifies the course of the disease over 1–2 years.

Myth 5: 'Ventilation tubes prevent tympanic membrane retraction and cholesteatoma'.

There is no evidence for this at all. In fact, children who have had VTs are slightly more likely to have tympanic membrane retractions because of the physical damage to the tympanic membrane caused by the procedure. VTs do not have any long-term protective effect.

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Aetiology of OME

Risk factors for OME are male sex, winter months, socio-economic deprivation, large number of siblings, more time in day care, bottle- rather than breastfeeding and recurrent episodes of acute otitis media: all factors which predispose to bacterial infection. OME is primarily an inflammatory condition caused by the presence of bacterial biofilms in the middle ear. Unlike acute otitis media, there is no fever, pain or purulent discharge because the bacteria are not actively dividing and multiplying. They lie dormant in a layer of glycoprotein matrix, stimulating a low-grade inflammatory response but without overt signs of infection. Young children are more prone to bacterial infection as their immune systems are still maturing, particularly their response to capsulated organisms. Eustachian tube function improves throughout early childhood and this may play some role, but the idea that OME is primarily caused by Eustachian tube dysfunction has largely been debunked. Evidence of pepsin in middle ear fluid suggests that reflux may play a role in the development of OME, although the exact role is unclear at present.

Prevalence and Natural History of OME

On any given day, approximately one out of every five 2-year-old children will have bilateral middle ear fluid on examination, and 17 out of 20 will have had bilateral middle ear fluid for at least 3 months at some point before their 6th birthday (1–3). Most of these children will have few or no symptoms and will not come to our attention. The natural history of OME is that it resolves spontaneously in the vast majority of children by their early primary school years. In a minority, OME can cause a mild-to-moderate conductive hearing impairment with educational consequences such as delay in speech and language. Mostly, children will catch up with their peers educationally once the OME resolves and the hearing returns to normal. The available evidence does not suggest any long-term detriment to education for the majority but there are subgroups of children who have been shown to have persistent deficits in speech, language and education. Unsurprisingly, it is those children with OME at an early age (under a year), with more severe or prolonged hearing loss, those exposed to multiple languages and those with the compounding effects of socio-economic deprivation or underlying medical conditions (4).

A small minority of children with OME will have disease that persists into the school years, and some will go on to develop atelectasis of the tympanic membrane, retraction pockets, chronic perforation or cholesteatoma. At present, we have no way to predict who they will be, or any treatment to prevent progression.

Assessment and Diagnosis of OME

For children presenting with suspected hearing loss, speech delay or behavioural issues, it is important to consider OME. Diagnosis is based on otoscopy (as discussed in detail in **Chapter 2**) and tympanometry (also discussed in detail in **Chapter 7**). Age-appropriate audiometric assessment is mandatory at this point as the treatment of OME is based on the presence or absence of hearing loss, not the presence or absence of fluid.

Management Options in OME: Non-Surgical

For most children with OME, the hearing is within the range of normal and the child's education and speech development are fine. Reassurance, observation and monitoring of hearing are all that is required.

No medication has been shown to be beneficial in OME. Antibiotics, steroids, antihistamines and mucolytics have all been shown in randomised controlled trials to be ineffective. They should not be used (*see* NICE Guidance NG233 [5]).

Politzerisation (blowing air up the Eustachian tube with a device such as the EarPopper, popular in the USA) and enhanced Valsalva manoeuvres (such as the use of the Otovent balloon, popular in the UK) have their advocates. Their use is promoted based on the outdated notion that OME is primarily a disease of Eustachian tube function which can be corrected by forcing air into the middle ear. Only a minority of (mostly older) children tolerate their use and persist with them long enough to see benefit. They are unlikely to do harm.

Hearing aids can be used with good effect to manage the educational and speech issues caused by hearing loss in children with OME, although there is currently no research evidence for their use in this condition. They have the obvious advantage of avoiding the need for general anaesthesia, and the complications of VTs (see below). Given the short-lived and self-limiting nature of OME for most children, and the fact that most children have normal bone-conduction thresholds, they are in some ways ideal. However, hearing aids can be uncomfortable and stigmatising, and not every child will tolerate them. Frequent visits to audiology are required if the hearing is fluctuating in severity, and to ensure that there is no danger of acoustic trauma from over-aiding once the OME resolves. Bone-conduction aids are much better than air-conduction aids in this regard as over-aiding is not an issue and frequent testing and programming are not required (*see* **Chapter 10**). They can be very expensive, however, and somewhat unsightly. Of course, the benefit of hearing aids is entirely dependent on compliance with wearing them.

Management Options in OME: Surgical

For the child with a hearing impairment, it is important to note that most middle ear effusions are transient and there is therefore no need to rush into treatment straight away. The proportion of children with bilateral middle ear fluid, confirmed on tympanometry, who will have complete resolution of their OME within a 3–6-month period depends on how long the OME has been there (which isn't always known in clinical practice) and the cause. After an episode of acute otitis media, 75–90% of middle ear effusions will resolve in 3 months (which is why we don't label it as OME until it has been present for at least 3 months and is therefore considered pathological). For OME of recent onset, around 56% will resolve in 3 months, 72% by 6 months and 87% by 12 months. If the OME is chronic then around 19% will resolve by 3 months, 25% by 6 months and 31% by 12 months (6). Either way, a reasonable proportion resolve in a relatively short time without intervention. It is therefore recommended that, in general, children should be demonstrated to have middle ear fluid with a significant hearing impairment on two occasions 3 months apart before surgical treatment should be considered. This 'watchful waiting' period is designed to prevent unnecessary intervention in those with a transient problem. There are, of course, a small number of children who are affected so badly in their day-to-day lives that earlier treatment may be considered.

Candidacy for surgical treatment, then, is based on a history of hearing concerns, education problems or speech delay; the presence of middle ear fluid, confirmed on otoscopy and tympanometry; and a hearing impairment of at least 25 dB in the better-hearing ear on two occasions 3 months apart. The mere presence of middle ear fluid is not enough reason on its own to justify intervention, being as it is a normal and commonplace feature of childhood.

Myringotomy without placement of a VT has a very short-lived effect and is, therefore, of no benefit.

The surgical procedures to consider are adenoidectomy and insertion of VTs.

It is important to note that VT insertion provides *short-term symptomatic relief from hearing loss* and in that sense is analogous to provision of hearing aids. Like hearing aids, the VT does nothing at all to modify the disease in the long term. Once the VT extrudes, its effect ceases. If the child has grown out of the tendency to develop OME, then the ear remains healthy once the VT has extruded. If the child has not grown out of the tendency to develop OME, then the OME returns. In randomised controlled trials comparing VTs to no treatment, hearing at 1 year post-VT insertion (when the vast majority of VTs have extruded) is the same in those who get VTs and those who do not (6).

The only treatment we have that modifies the underlying course of OME is adenoidectomy. By this, we mean that the child is less likely to have middle ear fluid 1 or 2 years after surgery, long after VTs have extruded (7). If adenoidectomy is performed at the time of VT insertion, the OME is around 50% less likely to recur when the VTs extrude (8–10) and repeat VT insertion is much less likely to be needed. This effect is independent of the size of the adenoid or the presence of nasal symptoms.

Adenoidectomy improves the health of the ears in the medium- to-long-term but does not have any immediate effect on hearing. VTs improve the hearing immediately but are extruded after a few months with no persistent effect on the ear after they are gone. The ideal combination for almost all children, then, is to do both adenoidectomy and VT insertion at the same time.

Many surgeons believe that VTs will prevent the development of long-term complications of OME such as retraction pockets and cholesteatoma (*see* **Box 8.1**). There is no evidence at all that this is the case. In fact, the VTs themselves cause a degree of damage to the integrity of the tympanic membrane, especially if they have been inserted on multiple occasions (11). It can be hard to sort out the effect of the disease from the effect of the treatment, particularly in observational studies, but the controlled studies that have been done suggest a small but important *increase* in the number of retractions and atrophy due to the VTs themselves (12–14). When retractions are already present, whether or not VTs are inserted makes no difference to the proportion of retractions which progress, stay the same or resolve. There is increasing evidence that tympanic membrane retractions are primarily due to middle ear inflammation rather than underventilation. The presence of tympanic membrane pathology is not, therefore, an indication for VT insertion.

Choice of Ventilation Tube

Various designs of VT are available and the choice of which to use is mostly a case of surgeon preference. They vary in size, shape, material (PTFE, silicone and titanium are common) and antibacterial coatings. Most are designed to extrude spontaneously after 8–15 months. The list of short-stay VT designs includes Shepard, Shah, Paparella, Armstrong and Reuter Bobbin. Some (such as the Goode T-tube) usually stay in place until they are removed. In all cases, there is a trade-off between duration of stay (and therefore duration of benefit) and risk of complications. For example, a short-stay tube such as a Shepard or a Shah might be associated with a persistent tympanic membrane perforation rate of around 1%, whereas for a Goode T-tube, removed after 2 years, it will be around 4%.

OME in Children with Trisomy 21 (Down Syndrome)

Children with Trisomy 21 have a much higher prevalence of OME: 90% will have bilateral middle ear fluid at age 2, compared with 20% in the general population (15). It also persists much longer, being present in 68% at age 5, and 50% at age 10 (16). It is generally assumed that we should use the same criteria for intervention, namely 25 dB in the better ear on two occasions 3 months apart. There are two major unknowns here, however. The first is that children with Trisomy 21 often have learning difficulties and visual problems in addition to hearing loss, so their ability to cope educationally with even a small degree of hearing loss might not be the same as for other children. It is possible that they may benefit from intervention at a much lower threshold. The second issue is that, since OME tends to persist much longer in children with Trisomy 21, the rationale for a 3-month watchful waiting period is less clear. So few children with Trisomy 21 will have resolution of their OME in a 3-month period that, unless the middle ear fluid is clearly a result of a recent viral upper respiratory infection, immediate treatment is probably reasonable.

VTs are not always ideal for children with Trisomy 21. Very narrow ear canals are a feature of Trisomy 21 so, in some cases, VT insertion is simply not possible. VTs also have a higher otorrhoea rate in children with Trisomy 21. This can be troublesome enough to merit VT removal in some cases. The main problem, though, is that the short duration of middle ear ventilation produced by a set of short-stay VTs is unlikely to be enough for children with Trisomy 21 whose OME is likely to persist for many years.

Hearing aids are an effective way to manage the hearing loss but are also not without their problems. Children with Trisomy 21 have pinnas which are small, cupped and soft, so earmoulds are not always easy to fit. Small dome inserts can be useful. Bone-conduction aids are also an excellent choice. Learning difficulties and behavioural issues mean that many children with Trisomy 21 refuse to wear their aids and so get no benefit. For them, VTs may be a better choice.

In reality, then, both treatments should be available and offered according to the needs of the child at the time.

OME in Children with Cleft Palate

OME is present in 90% of children with a cleft palate at age 5 (17) (the prevalence of OME in children with isolated cleft lip is the same as in the general population). The effect of the tensor veli palatini muscles in opening the Eustachian tubes on swallowing is impaired when there is a cleft palate and this contributes to the high prevalence of OME, although infection may also play a part. Repair of the palate does not seem to produce a reliable improvement in the middle ear. In the past, it was common for all children to undergo VT insertion at the time of cleft repair (6–9 months of age) but this is no longer recommended. Audiometric evaluation is recommended and VT insertion and hearing aids are only considered as options when a hearing loss is present. OME can persist for many years in these children so it is likely that multiple sets of VTs will be required.

OME in Girls with Turner's Syndrome (Monosomy X)

Turner's syndrome (TS) affects 1 in 2000 newborn girls and is therefore one of the commonest genetic disorders seen in clinical practice. It is discussed in more detail in **Chapter 9**.

Recurrent acute otitis media affects 61–88% of girls with TS, many of whom are treated for years for ear infections before their diagnosis of TS is known. OME is also very common in girls with TS, with middle ear fluid found in 55–78% of girls under 16 years of age. Conductive hearing loss is found in 39–43% of girls under 16, and it persists beyond the age of 20 in around 20%. VT insertion is required in around one-third of girls.

Tympanic membrane pathology is very common with chronic perforations noted in 10%, retraction pockets in 19% and cholesteatoma in 3–15% in large cross-sectional studies (18).

OME in Primary Ciliary Dyskinesia

Primary ciliary dyskinesia (PCD) is a rare autosomal recessive condition characterised primarily by respiratory problems including bronchiectasis and chronic rhinosinusitis. Other features include infertility and sometimes situs inversus (when it may be known as Kartagener's syndrome). More than 85% of affected children will have middle ear fluid, usually of a very thin, serous consistency and unlike the usual thick, sticky mucoid secretion of typical OME. The middle ear fluid usually resolves in the teenage years. Most have a mild hearing loss (around 20 dB) but some will need management with hearing aids. VTs should be avoided as they cause persistent otorrhoea which can be quite troublesome (19).

Follow Up after Surgery for OME

It is usual for the child to have one outpatient visit for audiological testing after surgery to demonstrate that the hearing has returned to normal with the VTs in place. This is to prove that the hearing problem was caused by the OME and to exclude an underlying sensorineural hearing loss or ossicular abnormality. If adenoidectomy is performed at the time of VT insertion then the recurrence rate of OME on extrusion of the VTs should be low, and therefore prolonged clinical follow up is not usually required.

OTITIS MEDIA WITH EFFUSION IN ADULTS

The very high incidence of OME seen in children is not reflected in adult populations. When it occurs, this is most often in response to a viral upper respiratory tract infection, in adults who are unable to aerate their middle ears.

A unilateral OME in an adult should alert the clinician to the possibility of a nasopharyngeal lesion obscuring the Eustachian tube orifice. In such patients, a flexible nasoendoscopy to examine the nasopharynx for any such lesions should be performed at the first clinic visit, with biopsy of any identified lesion subsequently carried out.

The insertion of VTs in adults with OME can be considered if conservative measures such as otoinflation and nasal decongestants have not been effective.

In an adult, VT surgery is a very simple procedure, which can be carried out in the outpatient clinic with topical local anaesthesia, a small myringotomy, and insertion of the VT. The patient will often experience immediate relief, and improved hearing.

Before proceeding, patients should be aware of the risks of VT insertion, which include bleeding, infection, persistent discharge, tube extrusion with recurrent OME, and tube extrusion with residual eardrum perforation.

A rare cause of middle ear effusion in an adult is cerebrospinal fluid (CSF) in the middle ear, secondary to a skull base defect. The clinician should be alerted to this possibility in the patient whose ear leaks clear fluid following VT insertion. A sample of the clear otorrhoea should be collected and sent for Tau Protein analysis.

VTs in adults tend to remain in place for a variable amount of time, though 6 months to 2 years is a reasonable estimate. Longer-lasting tubes such as T-tubes stay in place for longer.

The failure of an adult's conductive hearing loss to improve with a VT usually indicates another cause for their conductive hearing loss such as ossicular fixation or discontinuity, or ossicular erosion due to cholesteatoma or chronic infection.

CONCLUSIONS

- Middle ear effusion is a common feature of early childhood. If it persists beyond 3 months we consider it pathological and call it *otitis media with effusion* (OME).
- OME is usually short-lived, benign and asymptomatic. Even when symptomatic, a substantial proportion of cases resolve within 3–6 months without treatment.
- Candidacy for treatment is based on a history of hearing concerns, education problems or speech delay; the presence of middle ear fluid, confirmed on otoscopy and tympanometry; and a hearing impairment of at least 25 dB in the better-hearing ear on two occasions 3 months apart. The mere presence of middle ear fluid is not enough reason on its own to justify intervention.
- Ventilation tubes provide short-term relief of the hearing impairment but have no long-lasting effect after extrusion. Adenoidectomy is the only treatment option with long-lasting effects.
- Hearing aids are a viable treatment option where hearing and speech are the main concerns.

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9

Sensorineural hearing impairment in children

CONGENITAL SENSORINEURAL HEARING LOSS

Aetiological Investigation of Congenital SNHL

Chapter 7 contains a description of the rationale for, and results of, the universal neonatal hearing screening programme. Once babies have been identified as having a hearing loss the question arises of why. This is important as some causes of hearing loss are treatable (specifically infection with cytomegalovirus [CMV]), some have prognostic implications because they are at risk of progressive deterioration in hearing (such as Pendred) and some genetic disorders will have implications for families who wish to have more children. Parents also just want to know why their child has a hearing loss.

It is tempting simply to list all the potential conditions that cause congenital hearing impairment (and there are many hundreds) and thereby generate a list of tests that should be applied to every child. Apart from being expensive, wasteful and time-consuming, such routine application of an ill-thought-out test battery would have significant negative consequences and it is worth considering why.

Let us take as an example Jervell and Lange-Nielsen (JLN) syndrome, a very rare autosomal recessive condition causing congenital bilateral severe-to-profound hearing loss. It also causes a prolongation of the Q-T interval on ECG, leading to syncope, arrhythmia and sudden cardiac death. It may seem sensible to perform an ECG on every child with congenital hearing loss to identify this serious condition and perhaps avoid a preventable death. The problem is that the Q-T interval can be hard to measure accurately on a neonatal ECG (remember that the normal resting heart rate of a neonate is 120–170 beats per minute) and routine ECG will generate many times more false positives than true positives because the syndrome itself is so rare. Each false positive leads to further investigations, time in hospital and parental anxiety. The vast majority of cases of JLN have a positive family history for the condition, or of syncope and sudden unexplained cardiac death in close relatives. Testing is better targeted at a few babies, based on careful history taking rather than as a blanket test for every baby.

The British Association of Audiovestibular Physicians (BAAP) have produced guidance on aetiological investigation of infants with hearing loss in various scenarios (mild-to-moderate bilateral congenital loss, severe-to-profound bilateral congenital loss, progressive loss and unilateral loss). The cornerstone in each situation is careful history taking and examination. This includes a full family history and ideally pure tone audiometry for all close relatives. Most investigations of the child are recommended for use in selected cases, and there are only three which are recommended for routine use in everyone.

The first is imaging of the inner ear, which is ideally MRI scanning. In neonates and infants up to a few months of age, this can usually be achieved without general anaesthesia by giving the baby a feed, wrapping them in a blanket and keeping

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them in a quiet dark room until they drift off to sleep (often referred to as 'feed-and-wrap' or 'normal sleep' scanning). If a general anaesthetic were required, the risk-to-benefit ratio would be very different, but if the scan can be achieved under normal sleep then it can be a very worthwhile thing to do. Abnormalities of the cochlea and cochlear nerve can be seen, together with enlargement of the vestibular aqueduct suggestive of Pendred syndrome.

The second test to be recommended for all babies is investigation for congenital CMV infection. Congenital CMV affects about 1 in 200 babies, of whom 1 in 10 will have a rash or jaundice at birth, at least 75% of whom will go on to develop hearing loss. Testing can be done on saliva or urine for CMV DNA if the child is under 3 weeks of age. If the child is over 3 months of age, urine or saliva may still be tested but a positive test may indicate acquired rather than congenital CMV. Congenital CMV infection is ruled out if the mother is negative for CMV IgG. If the child's neonatal blood spot (Guthrie) test can be retrieved, tested and found to be positive then congenital CMV infection is confirmed, but a negative blood spot test does not exclude it. Even over 1 year of age, it is worth testing the child's urine for CMV DNA or blood for CMV IgG. The diagnosis is very important to make because the hearing loss is progressive without treatment, it may not be apparent until as late as 3 years of age, and it is possible that antiviral therapy might lead to improved hearing outcomes (*see* Pesch et al, 2021 for more information [1]).

The third routine 'test' is ophthalmology assessment. Partly, this is because children with hearing loss may be more reliant on vision, but mostly it is to detect associated conditions, particularly Usher syndrome. This is a rare autosomal recessive condition (or rather, set of three conditions designated types I-III). It is characterised by hearing loss with retinitis pigmentosa, making it one of the commonest causes of deaf-blindness, a situation which most people would consider devastating. It is recommended to have all neonates examined by an ophthalmologist to look for early signs of retinitis pigmentosa so that the prognosis for vision can be determined and communication strategies can be put in place before visual loss occurs. However, the diagnosis can be very difficult to establish in neonates as the ophthalmoscopic signs of retinitis pigmentosa are subtle.

Aetiological investigation of congenital deafness in the UK is changing significantly due to the introduction in 2022 of a new gene panel (the R67 monogenic hearing loss panel). Up until recently, genetic testing for children with congenital deafness was done only for a select few genes, guided by history and examination. In babies without any diagnostic features suggesting a syndrome, routine testing for *GJB2* (connexin 26) gene mutations would be reasonable given how common they are (*see* **Chapter 1**, **Figure 1.11**) but any other genetic tests would be done very selectively or not at all. The R67 panel is a result of Next Generation Sequencing where samples from multiple patients can be tested simultaneously for a large number of gene variants at reasonable cost. The panel tests for 115 gene variants, including *GJB2* (connexin 26) and a wide range of other gene mutations causing nonsyndromic deafness, the *m.1555A>G* variant associated with aminoglycoside-induced hearing loss, and mutations associated with syndromes such as Pendred, JLN, Waardenburg, Usher and many others. It is provided by NHS England and can be requested for any child with confirmed bilateral sensorineural hearing impairment, and selected cases of unilateral hearing impairment thought to have a genetic cause. Widespread use of the R67 panel will lead to many more children receiving a specific diagnosis for the cause of their deafness. Genetic counselling will be required for many families in order to properly deal with the consequences of a genetic diagnosis. The recurrence risk for future pregnancies can be discussed and targeted genetic testing of family members may be required if they are at risk of hearing loss themselves. In future, accurate genetic diagnosis will be the basis of gene therapies for deafness. This is still a few years away for most causes of deafness, but it was reported in May 2024 that near-normal hearing had been restored to a profoundly deaf child with OTOF-related deafness (a rare autosomal recessive, nonsyndromic deafness) by injecting adeno-associated virus through the round window to deliver the modified gene direct to the cochlea.

MANAGEMENT OF CONGENITAL SNHL

Once the nature and severity of the hearing loss has been established by means of ABR, early provision of amplification using hearing aids is the obvious important step. The team may take the decision to allow the family to have 2–3 months to bond at home before hearing aids are given. Parents often need time to come to terms with the unexpected news that their child has a permanent hearing impairment, and hearing aids are a very obvious reminder that things are not how they might have planned. The provision of hearing aids should not be delayed beyond this short time, however, as there is a short and very critical window for hearing development and if sound is not provided, the necessary brain pathways will not develop. Bilateral digital hearing aids are the standard. For children who have a severe-to-profound hearing loss meeting criteria for cochlear implantation, referral to the regional cochlear implant unit should be made as soon as possible. Outcomes are best when the child is implanted early, ideally under 1 year of age (*see* **Chapter 13**).

Auditory Neuropathy Spectrum Disorder

Auditory neuropathy spectrum disorder (ANSD) is a cause of sensorineural hearing loss in which the cochlea responds to sound but there is an impairment in how that is transmitted to the brain. It is described as a spectrum because of its very variable presentation and test results. The hallmark of ANSD is the presence of an OAE but an absent or abnormal ABR.

Around 40% of cases are genetic, some of which may be part of a syndrome such as Charcot-Marie-Tooth or Friedreich's Ataxia. The remaining 60% are environmental in origin, often in children who have spent time in the NICO due to neonatal jaundice, prematurity, low birth weight or hypoxia. CMV infection and meningitis may also cause ANSD. Audiometric pure tone thresholds vary from normal to profound, but the degree of difficulty understanding speech is usually much worse than might be predicted from thresholds alone, and this is always considerably worse in the presence of background noise.

These children are medically complex and may have general developmental delay. Their prognosis is very variable and hard to predict. Close audiometric follow up is essential along with family support. Communication should be encouraged by every means including lip-reading, gesture and signing if necessary. Behavioural testing provides the most useful guide to the need for amplification. Some children who fail to make good progress with amplification may be considered for cochlear implantation but this is a difficult decision to make when behavioural and objective testing produces unreliable results.

PROGRESSIVE AND ACQUIRED SENSORINEURAL HEARING LOSS IN CHILDREN AND YOUNG ADULTS

In **Chapter 1**, we saw that the prevalence of permanent bilateral hearing loss is around 1.3 per 1000 births, but this almost doubles through childhood. This increase is due to progressive hearing loss of genetic origin (particularly Pendred syndrome), congenital CMV infection and acquired causes (mostly meningitis, but also other infections, head trauma and ototoxic drugs).

Congenital CMV Infection

CMV is widely prevalent in the population, usually without any symptoms. We saw in the discussion above, however, that congenital infection with CMV can lead to progressive hearing impairment and it is in fact one of the most common causes of such a loss (**Chapter 1**, **Figure 1.11**). A blood test for CMV IgG, combined with retrieval of the neonatal blood spot card for retrospective testing, can make the diagnosis and allow for antiviral treatment.

Pendred Syndrome and Enlarged Vestibular Aqueduct

Pendred syndrome is an autosomal recessive condition which was originally described as an association between congenital sensorineural hearing loss, learning difficulties and goitre. Now that the vast majority of cases are diagnosed at birth, hypothyroidism is diagnosed and treated early so the goitre and learning difficulties are not given the chance to develop. We now know that the genetic basis of the disease is usually a mutation in the *SLC26A4* (pendrin) gene, which also causes the syndrome of enlarged vestibular aqueduct (EVA). Pendred and EVA are therefore considered to be overlapping clinical entities and different manifestations of the same disease. The hearing loss is symmetrical and progressive.

Pendred/EVA is the commonest syndromic cause of hearing loss, and one of the commoner causes of both congenital hearing loss and of progressive loss overall, affecting around 2–3 per 1000 children, as shown in **Chapter 1**, **Figure 1.11**. The diagnosis is usually made after congenital hypothyroidism is identified on the neonatal blood spot (Guthrie) test, or as a result of neonatal hearing screening followed by an MRI showing an EVA. Mondini dysplasia of the cochlea is also a common finding on MRI. The *SLC26A4* (pendrin) gene mutation is now tested for in the R67 gene panel, as are rarer mutations such as *FOXI1* and *KCNJ10* which can cause the same condition. It used to be thought that minor head injuries precipitated a step-wise deterioration in the hearing due to transmission of momentary rises in intracranial pressure to the cochlea: there is increasing evidence that this is not the case. Nonetheless, most surgeons recommend that affected individuals avoid contact sports or activities that involve large changes in barometric pressure such as scuba diving. There is no good evidence that steroids reverse the hearing loss when it occurs, although the risks of treatment are low. The same cannot be said for endolymphatic sac surgery which should be avoided.

Meningitis and Other Infections Affecting the Inner Ear

Acute bacterial meningitis, with or without septicaemia, is a very serious and life-threatening condition. It has become much less common in developed countries which routinely vaccinate children against the most usual causative bacteria (*Neisseria meningitidis, Streptococcus pneumoniae, Haemophilus influenzae*) but it remains common in the developing world. *Streptococcus pneumoniae* is the commonest cause in the UK and also the one most likely to lead to deafness as a complication. Hearing loss occurs when the infection spreads from CSF to the cochlea causing labyrinthitis. Around 5% of children will have a bilateral severe-to-profound hearing loss after bacterial meningitis in the UK, but the figure is more like 25% in developing world countries (2). Treatment with steroids alongside antibiotics reduces the incidence of neurological sequelae, including deafness, so this is recommended. Children are usually too ill at the time of infection for any hearing loss or vertigo to be noted during the acute phase. Labyrinthitis leads to progressive ossification of the cochlea which may make cochlear implantation difficult or impossible. The ossification proceeds rapidly so time is of the essence. It is very important that children are tested as soon as possible after recovery, ideally within 3–4 weeks, so that urgent cochlear implantation can be undertaken before ossification has occurred.

Infection with the measles (rubeola) virus caused around 10% of all hearing loss in the developed world in the first half of the twentieth century, but it was then essentially eradicated by childhood vaccination. It has emerged again as a problem due to widespread misinformation on the internet about vaccines, first in the wake of false claims about the MMR (measles, mumps and rubella) vaccine causing autism, and more recently due to vaccine scepticism during the Covid-19 pandemic. It is still a common cause of deafness in parts of the developing world where vaccination is not available. The deafness is usually bilateral, symmetrical and sensorineural, and it is more common in children who have suffered measles encephalitis. Mumps can also cause sensorineural deafness, usually unilateral, and it too is now occasionally seen in unvaccinated children.

Ototoxic Drugs

Ototoxicity as a cause of permanent hearing loss in children is fortunately rare and confined to certain predictable groups. Children undergoing cisplatin chemotherapy for cancer are at high risk and their hearing is regularly screened. Also screened are those on iron-chelating agents for haematological conditions such as thalassaemia and those on regular aminoglycoside antibiotics (such as some children with cystic fibrosis) or with known toxic aminoglycoside blood levels. If hearing loss occurs, it is a symmetrical, bilateral sensorineural loss which starts with the high frequencies. A mild loss may recover if the offending drug is stopped, but further dosing will lead to a permanent loss. These drugs are, of course, only used in serious clinical situations and there will always be a need to balance the risk of hearing loss against the risk of stopping the drug. Susceptibility to hearing loss from aminoglycosides is inherited down the maternal line due to a mutation in mitochondrial DNA designated *m.1555A>G*. This mutation can be tested for in order to assess risk from aminoglycoside treatment.

Trisomy 21 (Down Syndrome)

Trisomy of chromosome 21 affects around 1 in 700 newborns, making it the commonest genetic disorder in childhood. Conductive hearing loss due to OME is very common in these children, as discussed in **Chapter 8**. SNHL is also common, often described as an *early-onset presbycusis*. It is a progressive loss which starts with the high frequencies. It can start at any age but it is uncommon in the pre-school age group and typically comes on during school age or early adulthood (3). As these children also often have persistent OME, a mixed hearing loss is a common finding. One UK study found the prevalence of mixed hearing loss to be 4% at age 5 years, 7% at age 8 years and 23% at age 12 years (4). By adulthood, sensorineural hearing loss becomes the norm and is present in 36% at age 18–30 years, 46% at age 31–40 years, 64% at age 41–50 years, 74% at age 51–60 years and 100% of those 60 years old or more (5).

The provision of hearing aids can be made difficult by small, soft pinnas and narrow ear canals which accumulate wax. Bone-conduction devices and implants may have a role for some individuals.

Regular hearing screening is recommended in UK and American guidelines for all people with Trisomy 21, starting with neonatal screening followed by audiometric testing every year in childhood and every 2 years throughout adult life (6,7).

Turner's Syndrome (Monosomy X)

Turner’s syndrome (TS) is a common chromosomal disorder, affecting one in 2000 newborn girls, in which part or all of one X chromosome is missing. Characteristic features include a low hairline at the back of the neck, a wide or webbed neck, swollen hands and feet, a high arched palate, short stature, abnormalities of the aorta, hypothyroidism and coeliac disease. The features may be subtle and almost one quarter are not diagnosed until they fail to enter puberty. Most individuals with TS are of normal intelligence but may have specific learning difficulties and problems with spatial visualisation affecting tasks such as driving.

Middle ear disease is common in TS, as discussed in **Chapter 8**.

It is very common for girls and women with TS to develop a progressive, high-tone sensorineural hearing loss that resembles presbyacusis but which comes on at a much younger age and which progresses much faster (8,9). There is a characteristic audiometric pattern with a dip or notch around 1.5–2 kHz. This has been detected in girls as young as 6 years of age and is found in 58% of girls under 16 and 78% of women aged 16–34 (10,11). This progressive hearing loss seems to be more common in those with a history of recurrent ear infections earlier in life (11). More usefully, a number of large cross-sectional studies and one longitudinal study suggest that the presence of a dip or notch at 1.5–2 kHz is a good predictor of those who will go on to develop a progressive sensorineural loss (summarised in Kubba et al, 2017 [12]). Absence of a dip is a reassuring sign.

Above the age of 40, 27% of women with TS will have hearing thresholds greater than 20 dB, 27% will be wearing hearing aids and there are probably more who do not have hearing aids but would benefit (9). Regular screening for hearing loss in girls and women with TS has been recommended many times but only one such programme has been reported (13). There is no evidence at present to support either growth hormone or oestrogen as specific therapy to prevent progressive hearing impairment in girls and women with TS.

Girls and women with TS seem to have deficiencies in sound localisation compared to controls. This may result in them reporting greater hearing difficulties in day-to-day life than their pure-tone thresholds would suggest.

Auditory Processing Disorder

There is much about this condition which is controversial or unknown. It seems to be best described as a failure of *listening* rather than hearing as such. There are cognitive elements as well as audiological elements to this. The presentation is of a child with difficulty understanding and responding appropriately to speech, leading to educational concerns, problems with attention, difficulties in background noise and effects on speech and language. The degree of disability is out of all proportion to the audiogram which is usually normal. At present, there are no robust diagnostic criteria. Management involves optimising the listening environment and training the child in listening strategies.

CONCLUSIONS

- A thorough history (particularly family history) and physical examination are the basis for deciding which investigations to choose when investigating the aetiology of congenital deafness in children, but 'feed-and-wrap' MRI imaging of the inner ear, testing for congenital CMV infection and ophthalmology assessment are reasonable tests to consider in the first instance.
- The R67 monogenic hearing loss panel allows the identification of a very large number of genetic cause of hearing loss.
- 1.3 per 1000 children are born with a bilateral hearing loss of at least 40 dB HL.
- Pendred/EVA is the commonest syndromic cause of congenital deafness, and mutations in *GJB2* (connexin 26) are the commonest nonsyndromic genetic cause. Congenital CMV infection is the commonest environmental cause of congenital deafness.
- Progressive hearing loss in early childhood is most often due to congenital CMV infection or Pendred/EVA syndrome.
- Acquired hearing loss in childhood is most often due to bacterial meningitis, although ototoxic drugs, measles and head injury may occasionally be implicated.
- Early diagnosis of acquired hearing loss from meningitis is essential to allow for urgent cochlear implantation, if required, before the cochlea ossifies.
- In later childhood, progressive sensorineural hearing loss is a very common feature of Trisomy 21 (Down syndrome) and monosomy X (Turner's syndrome) and both children and adults with these conditions should have their hearing checked regularly.

FURTHER READING

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10

Sensorineural hearing impairment in adults

OVERVIEW

Once an individual has had an assessment of their hearing and an audiogram is available, those with a sensorineural impairment can be allocated to one of two categories, either *bilateral symmetric sensorineural impairment* or *unilateral/asymmetric sensorineural impairment*. These are approached in slightly different ways as the potential aetiologies for each category are different, apart from noise-induced hearing loss (NIHL), which is covered separately in the last section of this chapter.

BILATERAL SYMMETRIC SENSORINEURAL IMPAIRMENT

In 94% of patients seen with a bilateral sensorineural hearing loss, once controlled for age-related hearing loss, no additional aetiology can be identified (1). In 5%, noise exposure has an influence and in a further 1%, other aetiologies can be implicated. Correspondingly, knowledge of the effect of ageing (*see* **Chapter 1**) can be clinically helpful in communicating with patients and discussing their management.

Age-Related SNHL

By far the commonest type of hearing impairment in the population is a slowly progressive sensorineural defect that affects both ears equally. Previously, the diagnostic term 'presbycusis' was used to describe these age-related hearing impairments but this implies a pathological diagnosis. 'Age-related hearing loss' is an alternative term which removes the disease connotation. The pathological basis of age-related hearing loss is mentioned in **Chapter 1**. Surprisingly, at this stage in our knowledge, there is minimal evidence of a familial predisposition to develop a nonsyndromic age-related, sensorineural hearing impairment.

For clinical practice, what is required are accurate, prospectively obtained, data on the effect of ageing on the sensorineural thresholds, for males and females over a full range of ages from subjects that have not been exposed to potentially damaging noise. Such data are available from the UK National Study of Hearing reported by Adrian Davis in 'Hearing in Adults', which is now available online (2,3).

Background to 'Hearing in Adults'

'Hearing in Adults' (HiA) reports UK population data from the Medical Research Council National Study of Hearing (NSH) carried out in the 1980s and published in a volume in 1995 (2). Because only a limited number of books were printed, data were made available electronically online as, after peer review, they were still considered applicable to current populations in developed nations.

The strength of the NSH data is that around 3000 individuals were seen clinically, had their air- and bone-conduction thresholds evaluated over eight frequencies using defined pure-tone audiometric techniques in sound-proofed booths and then had their lifetime noise exposure estimated by means of the 'Medical Research Council Institute of Hearing Research protocol for estimate of noise exposure' (3). This enabled analysis of the average

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air-conduction thresholds with exclusion of those with a conductive defect with an air–bone gap of >10 dB HL or potentially damaging noise exposure above a defined threshold. No other national population data exist with such clinical detail, collected using a systematic protocol and controlled for noise exposure.

'Hearing in Adults': Audiogram Configuration

Figure 10.1 is an audiogram-style figure of the median thresholds for both right and left ears in men and women. It is clear that the deterioration with age is greater at the higher thresholds of 4, 6 and 8 kHz. However, for most purposes, individual

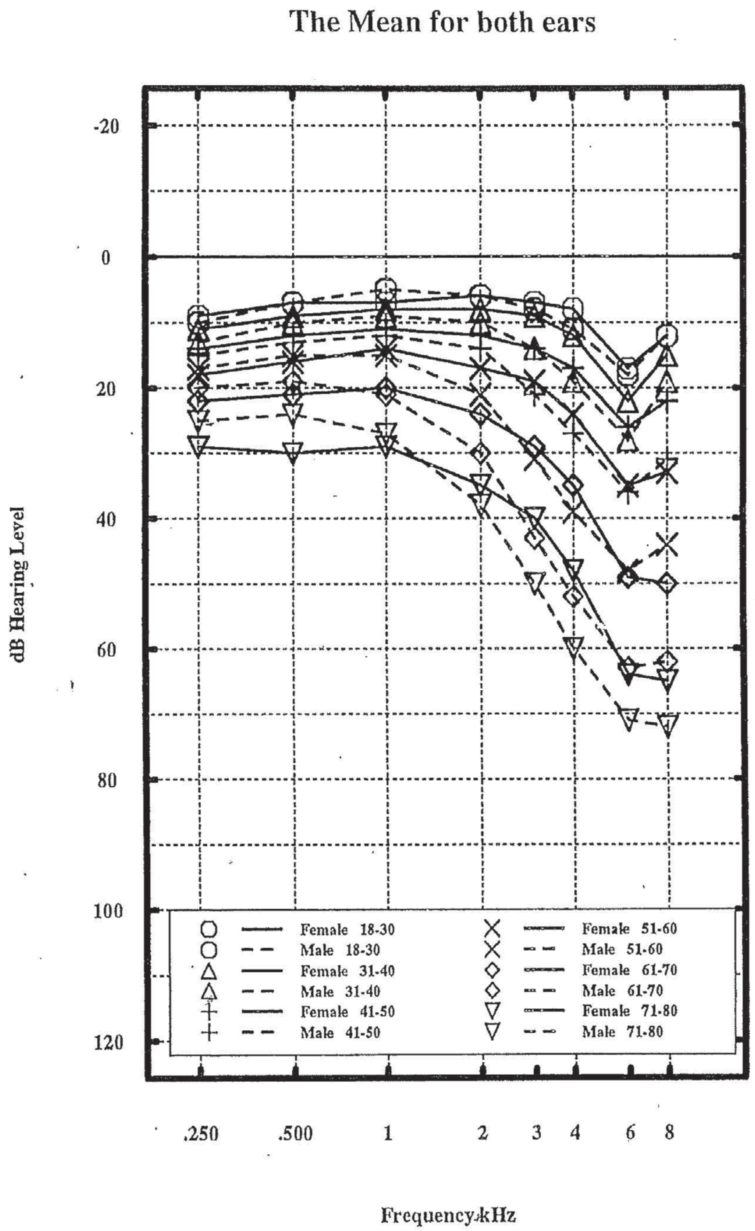

Figure 10.1 Median hearing thresholds for both right and left ears in men and women, by age.

frequency data are less useful than average data covering the speech frequencies.

'HEARING IN ADULTS': AGE-RELATED FFA THRESHOLDS

The six tables (A1.1–A1.6) in **Appendix 1** have been copied from the UK National Study of Hearing (2,3) for the Four Frequency Average (FFA) hearing thresholds on the left, right and mean of the two ears in males and females respectively. The FFA is calculated as the mean threshold over the main speech frequencies, 0.5, 1, 2, and 4 kHz. In these FFA tables, there are two population distribution data sets headed 'Screened' and 'Typical'. The 'Typical' distribution data are for a randomly selected population sample. The 'Selected' distribution data were created by removing individuals from the 'Typical' data set that had been exposed to potentially damaging noise or had a conductive defect of 10 dB or greater. As a result, the 'Screened' distribution data can used to evaluate the effect of ageing on the FFA sensorineural hearing thresholds having excluded individuals who have been exposed to potentially damaging noise. Data are presented for the various centiles of the population distribution, from the 5th to the 95th.

A clinician may therefore take an individual's FFA along with their age and sex, to identify where their FFA is on the relevant 'Screened' population distribution. If that individual has several audiograms available over a period of years, one would expect that, with 'normal' ageing, their FFA would stay in the same position in the population distribution. Taken as an example, this is aided by using Table 10.4 in this chapter. This table forms part of Table A1.1 for the left ear in men, limited to be between the ages of 38 and 55 years.

The column headed P .50 are the average thresholds at the peak of the population distribution of males of that age. Fifty per cent of the population distribution lie between the P .25 and P .75. Correspondingly, those with an FFA in the P .05 and P .10 columns have hearing thresholds better than average and those in the P .90 and P .95 columns have hearing poorer than average.

So, taking a hypothetical male patient, at the age of 38 years, if his FFA in his left ear was ~8 dB HL, this would be on the mid-point of the distribution of P .50. Then, with ageing, at the age of 55 years, one would expect his FFA to be still on the distribution of P .50 of ~13 dB HL. If the FFA was materially poorer than 13 dB HL, then a reason for its change in position in the population distribution should be sought.

It should be noted that the average data figures in these distribution data have been calculated from pure-tone averages that are in 5 dB steps. However, as the population distribution data have been mathematically calculated, the thresholds are in 1 dB steps.

In general, there appears to be minimal difference in most individuals between the FFA in the right and left ears. Having a difference between the ears that is greater than the test/retest error would suggest that there are other reasons than ageing. Correspondingly, when an asymmetric sensorineural hearing threshold is present, then consideration of diagnosable reasons is usually advised.

Asymmetry can also be present at the higher, non-speech frequencies, but this usually does not require investigation because test-retest errors are common. The presence of other symptoms, in addition to a hearing loss, such as vertigo, might alter that decision.

'Hearing in Adults': Related General Health Factors

Apart from age and sex, 'Hearing in Adults' found that socio-economic deprivation had an effect on the ageing thresholds. The exact mechanism by which it did so could not be determined but obesity, cardiovascular disease, smoking and diabetes could all play a role.

In developed countries, most older individuals will be on medication for some other age-related condition. Correspondingly, their age-related hearing losses are sometimes incorrectly attributed to these conditions or the medications they are being given for them. No material evidence exists to support these suggestions.

Ototoxic Medication

Aminoglycosides are well recognised as being potentially ototoxic, particularly for patients with the *m.1555A>G* mutation in their mitochondrial DNA. Certain chemotherapy agents (particularly cisplatin) are also ototoxic. The hearing loss is usually bilateral and symmetric, starting with the high frequencies, but can occasionally be asymmetric. As they are given in a controlled hospital setting, the hearing is monitored, and any hearing loss should be picked up early. The risk to hearing from continued use of the drug has to be weighed against the seriousness of the condition being treated and the availability of alternative treatments.

ASYMMETRIC/UNILATERAL SNHL

Definition

Clinicians tend to scan the audiogram to identify an asymmetric sensorineural hearing loss rather than calculating a difference between average air-conduction thresholds. Asymmetry of the air-conduction thresholds is most commonly due to a conductive defect in one or both ears. Correspondingly, the bone-conduction thresholds should have been appropriately masked to confirm that the hearing in the poorer hearing ear is due to a sensorineural loss rather than a conduction defect.

Having excluded a conductive component and confirming an asymmetric sensorineural loss, it would be wise to calculate the FFA in each ear to confirm that the asymmetry is greater than the test/retest error of 10/15 dB.

The question then arises as to potential aetiology. The first question to ask the patient is whether they have noticed any difference in the hearing between the ears and, if so, if they have any idea what caused the difference.

Potential Causes of Asymmetric Hearing Loss

Patients will remember if they have suffered an episode of *sudden sensorineural hearing loss*, with or without balance issues, or a *serious head injury with skull fracture*. Head injuries without fracture rarely cause sensorineural hearing loss. *Endolymphatic hydrops* will be accompanied by symptoms of vertigo and aural fullness and should therefore be apparent in the history. *Asymmetric noise exposure* will be covered in more detail below.

Vestibular Schwannoma

If all the above conditions have been considered and found to be negative, magnetic resonance imaging (MRI) is indicated to rule out a vestibular schwannoma, albeit most scans will be negative.

SUDDEN SNHL

Diagnosis

Sudden sensorineural hearing loss (SSNHL) is defined as hearing loss that develops within 72 hours, characterised by a decrease in hearing greater than 30 dB at three or more consecutive frequencies (5). A diagnosis of SSNHL is usually unilateral, warranting an MRI scan of the internal acoustic meati to exclude retrocochlear pathology.

Aetiology

It was previously believed that up to 90% of cases of SSNHL were idiopathic (6). However, recent literature is consistent in identifying that cases of SSNHL demonstrate raised inflammatory markers, suggesting an almost universal inflammatory or autoimmune process (7). Focused investigations should therefore be deployed on a case-by-case basis to identify underlying metabolic, infective and autoimmune conditions. Immune-mediated SSNHL tends to be associated with worse prognosis and irreversible hearing loss, but can be treated if recognised early, preventing additional damage to the inner ear (8). Bilateral SSNHL, fluctuations in hearing, nystagmus, visual disturbance and focal neurology are all signs that suggest autoimmune inner ear disease. Pre-existing systemic autoimmune disease is present in 30% of patients with immune-mediated SSNHL.

Treatment and Prognosis

Management of SSNHL is similar regardless of whether it is idiopathic or autoimmune. Steroid therapy is recommended as soon as possible, ideally within 2 weeks of presentation (9). This can be in the form of intratympanic and/or systemic steroid therapy. There is limited evidence for salvage therapy with steroids and hyperbaric oxygen therapy if treatment is not initiated within 2 weeks (10).

There is no consensus protocol for steroid treatment of SSNHL, with most reported regimes offering a 7–10-day course of oral prednisolone without tapering and with gastric protection, and/or a course of intratympanic steroid injections.

The American Academy of Otolaryngology–Head and Neck Surgery Foundation (AAO-HNSF) recommends follow up 6 months after presentation, with pure tone audiometry to monitor the progression of hearing loss and outcome of treatment (11). Follow up is particularly important in idiopathic cases, as up to 30% of patients may have an underlying cause diagnosed at follow up. Recovery of hearing following SSNHL varies between patients depending on its severity. Mild-to-moderate hearing loss resolves spontaneously in up to 65% of patients. Most patients display improvement in hearing, though not necessarily back to their normal baseline. Positive prognosis factors are isolated high- or low-frequency hearing loss, associated with tinnitus and the absence of vertigo (12). Severe bilateral hearing loss associated with autoimmune causes and vertigo carries the worst prognosis. Hypertension and metabolic syndrome have also been associated with poor prognosis of SSNHL due to microangiopathy of brain parenchyma (13).

Follow up is also important for the discussion of hearing rehabilitation with patients whose hearing has not recovered. Patients with serviceable hearing thresholds can be offered an air-conduction hearing aid. Those with minimal residual hearing can trial a contralateral routing of sound (CROS) aid or can consider a bone conduction device (*see* **Chapter 12**). These do not restore binaural hearing but reduce the head shadow effect by routing sound to the better hearing ear. A cochlear implant in the deafened ear is also an option, however as discussed in **Chapter 13**, does not always offer effective binaural hearing.

NOISE-INDUCED HEARING LOSS

Introduction

Arriving at a diagnosis of noise-induced hearing loss (NIHL), requires there to have been material noise exposure along with a hearing loss that is greater than one might expect from 'normal' ageing. The existence of serial occupational audiograms helps considerably in confirming NIHL and suggesting the magnitude of the effect of noise trauma in addition to the effect of ageing.

Considering, the presence of a 'notch' at 3 –4 kHz in a pure tone audiogram to be of diagnostic of NIHL is widespread but incorrect as such a notch is present in 33–53% of non-noise exposed adults (14,15). What is not generally recognised is that the literature supporting the 3–4 kHz notch as an indicator of noise trauma, excluded those with impulse noise exposure (16,17). They also never reported a control group.

Pathophysiology

Noise exposure is of two main categories that have different mechanisms of potentially causing NIHL: continuous noise and sudden impulse noise.

Continuous Noise above Approximately 85 dB A

Continuous noise can be from mechanical sources such as machinery. It can occur because of time spent in an industrial environment or by the personal use of noisy machinery such as grass-cutting equipment. Social exposure can occur from amplified music delivered through headphones or earbuds or by attendance at an entertainment event.

Such continuous noise is most frequently at low frequencies but because of the acoustic properties of the bony external auditory canal, the damage is usually greatest at 3–4 kHz and above. **Figure 10.2** illustrates the increase in sound level at the tympanic membrane of the presentation of a broadband sound at different angles to the head. This increase is due to the resonant characteristics of the bony ear canal acting as a cylinder, along with the considerably lesser effect of the pinna. This increase in sound peaks at 3–4 kHz with a gradually lesser effect at higher frequencies. Hence, the effect of excessive noise trauma is greatest at 3–6 kHz frequencies even though continuous mechanical noise is most often at lower frequencies.

It is also understandable that, when combined with the configuration of the effect of ageing being greater above 2 kHz (**Figure 10.2**), then any potential for a clearly defined notch due to continuous noise trauma can be lost.

Sudden Impulse Noise Exposure Sometimes Associated with Environmental Changes in Pressure

Blast explosions are the most damaging type of exposure in this category. This is often of a sufficient magnitude to affect the balance of subject. The stapedius muscles do not have time to contract so

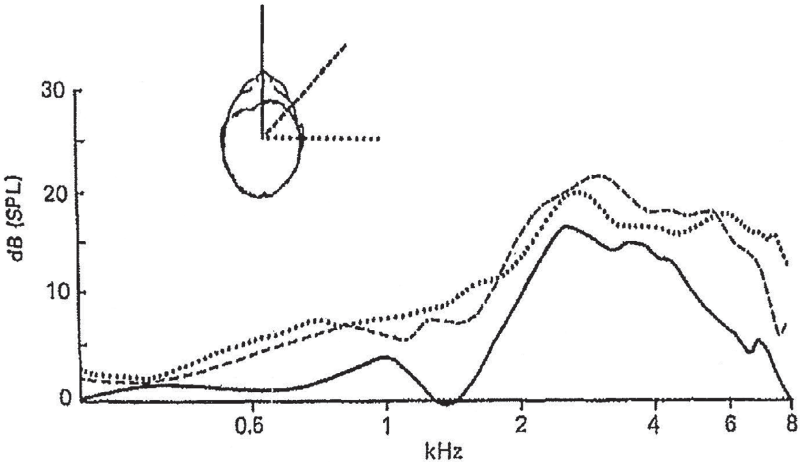

Figure 10.2 Increase in sound level at the tympanic membrane caused by resonance of the external auditory canal and the baffle system of the pinna. The effect of different angles of sound incidence is shown by the different lines (after Weiner and Ross [19]).

hearing loss is extensive, notches at 4 kHz do not occur and there can be total loss of hearing in the ear most exposed. Such blast explosions are not usually anticipated, so hearing protection is most often not worn.

Impulse noise exposure also commonly occurs from gunfire. When small arms are fired by the subject, the ear most exposed depends on how the gun is held. The commonest firearm is a rifle, which is fired from the shoulder. In such cases, the ear nearest the rifle barrel is the one that is more exposed to the greatest impulse sound levels (the left ear for a rifle held on the right shoulder). Handguns are held in front of the body when fired hence the ear most exposed will depend on the direction of fire and the stance taken by the firer.

Sudden impulse noise exposure can also occur in industrial situations such as metal hammering and riveting.

Assessment of Noise Exposure in a Subject

In taking a subject's history there are thus two types of noise exposure to assess:

- continuous noise exposure from machinery or amplified music
- sudden impulse noise exposure such as explosions and gunfire.

Convention dictates that a patient's history is taken to enquire as to whether the patient has been exposed to either of the two types of noise exposure and, if so, whether it was of a magnitude that could have caused NIHL. The thoroughness with which a noise exposure history is taken depends on whether the patient is claiming financial compensation. In developed countries, government legislation and increased awareness in the workforce has meant an increase in the number of such cases.

Increasingly, this has been balanced by the awareness in these countries of the dangers of industrial noise and the requirement for both lessening the noise exposure by the wearing of hearing protection and the lessening of the mechanical noise levels by soundproofing. If continuous noise levels of more than 85 dB A occur, workers in these occupations are required to wear hearing protection and to have their hearing monitoring by pure tone audiometry at set intervals during employment. Regrettably, the quality of these audiograms varies considerably.

With both categories of noise exposure, hearing protection might have been used. Whether this has been effective varies and should be assessed as part of the history for each of the two categories of noise exposure.

Hearing Protection

Avoidance is key. It is not to be exposed at all, or to position yourself in a quieter area. If you need to shout to communicate with others, then it is too loud and if you cannot avoid the noise, then you should have earplugs in your pocket to protect your hearing. This type of situation applies particularly to social noise exposure.

Expandable foam plugs must be compressed to allow them to be inserted into the ear canal. This can be difficult and practice is required. The earplugs may dislodge requiring reinsertion.

Pre-moulded, reusable plugs come in various materials and sizes. Trials are therefore necessary to get ones that fit well and can be comfortably inserted by pulling the ear up to straighten the ear canal with one hand and inserting the plug with the other. The plugs can be mounted on a flexible band that can be easily put on and removed.

Earmuffs are made by different manufacturers and are of varying effectiveness. These must be tried alongside any other headgear such as glasses when purchasing. Earmuffs can be used along with earplugs to enhance the degree of protection. When communication with others is necessary at the same time, then Bluetooth or a two-way radio can be built into a headset.

All the measures described above for hearing protection are passive in the sense that they work by insulating the listener from potentially damaging noise. *Active noise reduction* is the use of a microphone to detect the ambient noise which is then inverted and played through a loudspeaker. This inverted waveform cancels out the ambient sound waves to reduce the level of noise. Noise cancelling headphones are popular consumer products for commuters and aeroplane passengers, but very few are rigorously tested or rated for hearing protection in industrial environments.

Degree of Hearing Protection

Table 10.1 gives the likely attenuation of plugs and earmuffs if correctly worn. Different models of earmuffs come with their Single Number Ratings (SNR) but when worn along with earplugs the degree of attenuation is less than additive.

Employment Regulations

Understandably, regulations about controlling industrial noise exposure will vary between countries. In the UK, 'The Control of Noise at Work Regulations 2005' is the most recent legislation (18).

Table 10.1 Mean attenuation of ear defenders

Type		Attenuation (dB)
Earmuffs		24
Earplugs	Pre-moulded	21
	Foam	15

The 2005 regulations states, 'The level at which employers must provide hearing protection and hearing protection zones is 85 dB (A) (daily or weekly average exposure) and the level at which employers must assess the risk to workers' health and provide them with information and training is 80 dB (A). There is also an exposure limit value of 87 dB (A), taking account of any reduction in exposure provided by hearing protection, above which workers must not be exposed. The employer shall ensure that such employees are placed under suitable health surveillance, which shall include testing of their hearing.'

Noise exposure levels are measured in dB A, which is similar but not identical to dB HL. It is considered that noise exposure at levels of less than 85 dB A, are insignificant and can be discounted.

History of Continuous Noise Exposure

It is important to ask if there was *employment with continuous machine noise exposure.* Taking a noise exposure history in patients with a sensorineural impairment can give unexpected replies. It is surprising how often a 'dear old lady' worked in a factory without hearing protection in her youth. Individuals who have worked in similar noisy environments for the same length of time vary in their degree of potential noise trauma. The reasons for this variation in susceptibility are unknown but there could be a genetic predisposition as well as variable amounts of ear protection or exposure. It is also likely that the various aetiological factors will have a cumulative and perhaps a synergistic effect, so that in some, there will be damage due to other factors and the noise trauma simply makes the impairment more marked.

As employment with continuous machine noise exposure is more common than hobby exposure, the first question usually is, '*What was the noisiest job you have ever worked in?*' The level of noise exposure of any specific job can then be estimated from tables of the commoner noisy occupations (**Table 10.2**). This table was constructed before the UK industry realised that lessening the levels of noise was important to protect the hearing. Modifications to machinery were subsequently made and are now monitored. The noise levels in such tables are therefore approximate and should be confirmed by asking when ear protection was not being worn, how close a worker had to be and what level of voice had to be used to communicate with fellow workers. **Table 10.3** can then be used to enable a second estimation of the noise level to be made. Both these tables have been confirmed as being valid by others in recent years (20).

Table 10.2 Occupational noise exposure*

Occupation type	Noise levels (dB A)
Agricultural machinery (e.g. tractor)	93
Aviation aircrew (combat jet)	105
Bottling, bottle cleaning and canning	96
Fabrication (metal)	90
Iron and steel working	
(blast furnaces, blacksmith, heavy hammering)	99
(heavy forging)	102
(plating and press-shops)	96
Lumber work	99
Paper manufacture	90
Printing	93
Road drilling	96
Seamen (boiler room, other engines)	90
Shipbuilding (near boiler making, caulking, rust-scaling, riveting)	100
Textiles (weaving and spinning)	99
Tobacco (cigarette manufacturing machinery)	93
Transportation (diesel, heavy goods vehicles)	93
Woodworking/sawmill	90

*: These examples are a guide to equivalent continuous noise levels. The levels given are typical ones; individual exposures can be +10 to –20 dB A different.

Table 10.3 Noise exposure levels (dB A) as estimated from distance and voice levels at which conversation was possible

	Distance apart to be heard		
	4 feet	2 feet	Close to ear
Normal voice	<81	–	–
Raised voice	87	–	–
Very loud voice	93	–	–
Shouting	99	105	–
Impossible			> 110

If protection has been variably worn, the number of years of exposure is amended accordingly.

The ideal is to measure the actual noise levels but this is impracticable in many instances because the factory has often shut down or made subsequent modifications to comply with current safety standards.

History of Occupational Impact Noise Exposure

Even though a worker might only report exposure to continuous machine noise, question as to whether there were any situations when the noise was greater. This could be because they, or a co-worker, for example, hammered metal or fired rivets.

History of Social Exposure to Continuous, Potentially Damaging Noise

Though many individuals use power tools and others go to clubs, the cumulative exposure is hardly ever sufficient to cause damage. The noise levels at some clubs are likely to be at least 99 dB A, but for this to be as likely to cause damage as a job at 99 dB A would require someone to go seven times a week, for 6 hours each time for 50 weeks in the year. Few could keep this up on a social or even a professional basis. Should such a fanatic be encountered, exposure can be calculated as for employment exposure.

Was Ear Protection Used and Was It Effective?

It is essential to find what type of protection if any, was used and whether it was effective. Cotton wool plugs, even if coated with Vaseline, are useless. Commercial products can vary in their effectiveness. This is particularly the case with foam plugs, which must be collapsed by finger pressure then correctly placed in the ear canal, maintained there and found to be effective by the wearer. Some are more efficient at some frequencies than others. **Table 10.1** is a list of the mean attenuation such products give, provided they are correctly worn. These attenuation figures are then subtracted from the estimated noise exposure level. In most instances, the wearing of ear protection of any type advocated by the employer reduces the exposure levels to below 85 dB A and correspondingly that particular exposure can be discounted.

The fact that ear protection devices are provided does not mean that they are used. Earmuffs can be uncomfortable and many use them only when the task is particularly noisy. The wearing of muffs also makes it more difficult to communicate with fellow workers. Radio communication with earmuffs is an effective but costly solution and is not always available.

Where the level of sound is high and intermittent, such as in firing multiple rounds, the wearing of ear plugs along with earmuffs can be made mandatory but the effectiveness of hearing protection must be well supervised. So, in addition to asking if protection was worn, it is wise to ask if it was effective.

How Much Exposure?

The number of years spent in each noisy job is ascertained. The next question is to find out on average what proportion of the day, week and year was actually spent in the noise. It is perhaps easier, especially if ear protection has been effectively worn, to split the job into several parts and calculate the number of years out of the total that were spent at each noise level. Usually, performing such a calculation is relatively easy. Detailed calculations are only necessary when someone has been in many different jobs and is claiming compensation for one or more of them, when an apportionment has to be made.

Was There Any Temporary Threshold Shift or Temporary Tinnitus?

This is a very important question to ask as, if temporary dullness of hearing or tinnitus has occurred during or at the end of the day, it is an indication of temporary damage to the hearing due to the noise exposure.

What Magnitude of Exposure to Continuous Machinery Noise Defines Having a Potentially Damaging Effect on the Hearing?

The UK 'National Study of Hearing' categorised the reported Noise Emission rating of the subjects exposed to noise levels above 85 dB A and found that 5 or more years at levels of 90 dB A or greater would have a damaging effect (17). However, this effect is small over 4, 6 and 8 kHz and of the same magnitude [~10 dB] of 10 years of ageing (17). What is of greater value in confirming a potentially damaging effect if there is a temporary threshold shift occurs as evidenced by the report of temporary tinnitus and/or a diminution of the hearing, during or after the noise exposure event.

Audiometric Evaluation in Occupational Continuous Noise Exposure

In the UK and Europe, regulations dictate that when workers start their employment in a position where they could be exposed to noise levels of 85 dB A or greater, a base audiogram is usually obtained and repeated at intervals thereafter. So, increasingly, it is possible to calculate pure-tone averages at an individual's different ages to see whether there is a greater deterioration in these averages than expected from ageing.

The effect of ageing has already been explained, using the tables from **Appendix 1**, from 'Hearing in Adults'. So, using the table relevant to their sex, the claimant's FFA can be charted from each audiogram for the age the individual was at that time. **Table 10.4** is an example of the left ear in a male adult who, at the age of 40 years, changed his job to one that involved processing aluminium. At that time, his FFA in both ears was in the better 25–50% of non-noise exposed men. Though provided with ear protection, this was ineffective because it had parts that fell out. At the end of most days, he had ringing in his ears when he went home, indicative of temporary threshold shift. During the subsequent 10 years, his FFA in both ears deteriorated to be in the poorest 10% of the population. There was no clear notch in the audiogram at 3–4 kHz in either ear in any of the four post-noise trauma audiograms.

Table 10.4 has been used to mark in pencilled circles, where a NIHL claimant's FFAs fell in the population distribution at the age he had been when each

Table 10.4 This table is part of Table A1.1 in Appendix 1. Its age range is limited to 38–55 years and is of screened men who have not been exposed to material potentially damage noise or who have a conductive defect to their hearing thresholds.

Ear – left					Screened		Gender = male Frequency = 0.5, 1, 2 and 4 kHz		
	Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
	38	10	0	1	4	8	12	19	23
	39	10	1	1	4	8	12	19	24
	40	10	1	2	4	8	13	19	25
	41	11	1	2	4	8	13	20	25
2006	42	11	1	2	5 4	8	13	21	26
	43	11	1	2	5	9	14	21	27
	44	12	1	2	5	9	14	22	28
	45	12	2	3	5	9	15	23	28
	46	12	2	3	5	10	15	23	2+
	47	13	2	3	6	10	16	24	30
	48	13	2	3	6	10	12 17	25	30
	49	13	2	3	6	10	17	25	31
	50	14	2	4	7	11	18	26	32
	51	14	3	4	7	11	18	26 27	32
	52	14	3	4	7	11	19	27	31 33
	53	15	3	5	8	12	19	28	30 34
	54	15	4	5	8	12	20	29	34
	55	16	4	5	8	13	21	30	35

of his five work's audiograms had been obtained. His left ear has been chosen to illustrate the value of these tables; the right ear showed similar data.

His first audiogram was obtained in 2006 when he was 42 years of age. His left ear FFA then of 5 dB lay between the P.25 and P.75 columns that cover the 'normal' 50% of males of that age, P.50 being the peak of that range.

With the passage of time and normal ageing, one would expect his FFA to remain within the same position of the population data. This expectation has been confirmed in The Scottish Law courts and has been widely in Scottish legal cases.

In this individual's case, at the age of 48 years, his left ear FFA had deteriorated to 12 dB but still was within the P.25–P.75 range.

In the next available audiogram, when he was 51 years of age, his left ear FFA of 26 dB was materially poor and on the 90th percentile (P.90). This is accepted to be outwith the 'normal' range an aetiology should be looked for. In the last two audiograms, when he was 52 and 53 years of age, the FFAs of 31 dB and 30 dB fell between the P.90 and P.95 population distribution.

The only identifiable reason for his materially poorer hearing in his left was his daily, poor and ineffective hearing protection with foam ear plugs whilst working in a metal machining factory. The diagnosis of NIHL causing hearing damage, was supported by his history of temporary threshold shift and tinnitus persisting into the evening after a day at work.

Configuration of Pure Tone Air-Conduction Thresholds

Historically, when repeat occupational audiograms were not available, the presence in the audiogram of a notch in the air-conduction thresholds at 3, 4 or 6 kHz was considered diagnostic of continuous noise exposure trauma. In many studies (15, 21), such a notch has been shown to be present in a high proportion of individuals that have not been exposed to potentially damaging levels of noise. In addition, such a notch at 3, 4 or 6 Hz is only of around 10 dB in studies that have accurately investigated its magnitude following continuous noise exposure (17). This can easily be obscured by the already greater deterioration due to normal ageing of the hearing at these frequencies (15–50 dB HL between ages 41 and 80 years, *see* **Figure 10.1**).

Ascribing a Diagnosis of a Bilateral NIHL on a Single Audiogram

Having confirmed that an individual with a bilateral sensorineural hearing loss has been materially exposed to noise, a comparison should be made of their thresholds with the hearing in individuals matched for age, sex and socio-economic group who have not been exposed to noise. Such data for the UK population are available in 'Hearing in Adults' (2) and in **Appendix 1**. If the FFA of a subject of a certain age falls and is poorer than the 75th percentile [P.75] for that age then a NIHL must be a reasonable diagnosis to make, as this is the most commonly recognised factor. However, in any individual, the effect of noise is perhaps less than might be expected (17) and noise by no means affects every individual who has been exposed to it.

Sudden Impulse Noise Exposure History

Gunfire is the commonest sudden impulse noise that has potential to damage the hearing. In most countries, except perhaps the USA, gunfire exposure occurs in those in the armed forces or the uniformed police. Such officers will usually have to have regular practice of firing small arms on ranges with hearing protection. They will frequently have their hearing monitored by repeat audiometry. This makes the diagnosis of noise trauma easier to make, the most frequent reason being due to inadequate or defective hearing protection. Sportsmen also use firearms and their use of adequate hearing protection is uncontrolled.

History of Gunfire Noise Exposure

The simple question here is, 'Have you ever fired a gun and, if so, what type?' For the militarily inexperienced, the various types of guns can be confusing but it is the bore (size) of the barrel that matters. In terms of noise exposure, air guns and small-bore (.22) rifles and pistols can be discounted. Thereafter, guns can be classified as rifles (larger than .22) that can be fired from the shoulder or otherwise, and which fire single bullets or multiple automatic bursts. Finally, there are 'big guns' that are not held by the person firing.

The laterality of greatest noise exposure should be determined as this can result in asymmetric hearing trauma. The commonest example of this is damage from firing arms from the shoulder. This is most frequently the right shoulder and the left ear has the greatest exposure as that ear is nearest to the barrel.

Was Ear Protection Regularly Worn and Considered Effective When Firing?

If ear protection was worn and effective, gunfire exposure can be discounted. Whether ear protection was available depends on the era in which the exposure occurred. Protection has been provided for many years for all 'big guns' but not necessarily in active military service.

In recent years, those training the armed forces and police to use firearms have been aware of the necessity for protective headsets to be worn on the firing range. Regrettably, even though protective headsets are worn, they are often dislodged by all the other equipment that is worn at the same time, such as glasses and back packs. The use of 'double' protection by wearing earplugs in addition to a headset is often recommended.

Members of the armed forces that have been trained in firearm use require annual retraining on the firing range wearing ear protection. In addition, their hearing is monitored by having pure-tone audiograms carried out as part of their regular health assessments. These audiograms can be used in the same manner reported above **(Table 10.4)** to detect a change in their hearing that is greater than expected from ageing alone.

Again, it is only recently that many sportsmen have worn protection. On the other hand, the number of rounds that they fire is very much smaller than that fired by a soldier firing automatic repeat firearms. The number of rounds fired unprotected from a rifle requires to be between 101 and 1000 rounds. For a larger bore, it only needs to be between 11 and 100 (2).

Always ask 'After firing, were there occasions afterwards when your hearing was temporarily diminished, or you developed tinnitus?' This would indicate *temporary threshold shift*, indicating that the noise exposure has temporarily damaged the hearing.

History of Other Sudden Impulse Noise Exposure

Occupational sudden impulse noise can come from some machinery such as riveting and forging. Because of increasing awareness of the potential of hearing damage in most industrialised societies these machines are 'sound proofed', adequate hearing protection given and their usage monitored. War zones are rather different, and explosions can occur due to other causes even in civilian life. Such trauma damage to hearing is usually recognised at the time.

Audiometric Evaluation in Impulse Noise Exposure

The current FFAs for each ear should be calculated and a decision made as to whether the subject has a symmetrical or asymmetrical hearing. The next step is to compare these FFAs with the most relevant of the six population tables (**Appendix 1**) using the age of the subject on the 'screened' distribution data.

In those whose impulse noise exposure has been from occupational gunfire, then the same process is carried out on all available audiograms to help control for natural ageing over the time period covered.

Audiometric Configuration in Impulse Noise Exposure

Impulse noise trauma can affect all the pure-tone test frequencies but particularly the higher ones. As reported above, there is no evidence to support the use of any 3 or 4 kHz notch criterion in evaluating the diagnosis of NIHL due to impulse noise.

CONCLUSIONS ON BILATERAL SNHL

- Progressive, bilateral sensorineural impairments are the commonest type of hearing impairment in the adult human population.
- The majority are due to ageing and have no additional contributory aetiological factors.
- 'Presbycusis' is not a pathological diagnosis but a syndrome signifying the gradual deterioration in hearing that often, but by no means invariably, occurs with advancing years.
- Age-related hearing impairment is a better syndrome label.
- Population data from the UK 'National Study of Hearing' has electronically available tables that can be used to compare an individual's FFA in each ear to those in the population distribution of people of the same age and sex.
- There is no proven association between any 'general' human pathological disease and bilateral sensorineural hearing impairments. It is therefore incorrect to suggest that an age-related hearing loss is due to co-existing age-related conditions or their medication, such as cardiovascular disease or diabetes.
- The configuration of the PTA has no diagnostic significance in those with a bilateral sensorineural impairment.
- The main role of audiometry in this situation is to assess likely disability and to aid in management.

CONCLUSIONS ON ASYMMETRICAL SNHL

- Asymmetry in the air-conduction thresholds is most frequently caused by a conductive component in the poorer hearing ear.
- Accurate bone-conduction thresholds for the poorer hearing ear need to be obtained with appropriate masking of the better hearing ear before a diagnosis of an asymmetric sensorineural can be made.
- The commonest aetiology for an asymmetric sensorineural hearing loss is noise trauma.
- With asymmetric sensorineural losses, unless the subject has a clear reason for the asymmetry, it is conventional wisdom is to perform an MRI scan to exclude an acoustic neuroma.

CONCLUSIONS ON DIAGNOSIS OF NIHL

- Arriving at a diagnosis of noise-induced hearing loss (NIHL) requires detailed knowledge of the type of noise exposure and whether it was continuous machinery noise or sudden impact noise.

- An estimate of the continuous machinery noise level, attenuated by any hearing protection worn, is made. If greater than around 90 dB A over several years, then there could have been sufficient noise exposure to add to that individual's expected age-related hearing loss.
- Where their FFA lies on the distribution of the non-noise exposed population of the same age and sex is then determined by comparison with the relevant section of the tables in **Appendix 1**.
- If any reliable audiograms are available prior to the noise exposure, then a similar comparison is made of the FFA at the age of the individual at the time of the audiogram.
- Any change in the positioning of the FFA to a poorer percentage on the relevant table would support the diagnosis of a NIHL.
- If no audiogram is available prior to the noise exposure then, on the balance of probability, if the FFA is positioned poorer that the 75th centile, this is due to NIHL.

FURTHER READING

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11

Non-surgical management of hearing impairment

ASSESSING DISABILITY FROM A HEARING IMPAIRMENT IN ADULTS

Introduction

When assessing a patient's disability one has to consider the whole patient, their age and lifestyle. The majority of adults being seen with a hearing impairment will have an age-related sensorineural impairment. In the majority, this will be associated with other age-related health issues. Disability is best assessed by using a combination of measures to achieve the fullest picture possible of the individual in question.

Objectives of Assessment

The purpose of making a disability assessment of a patient's hearing, with or without tinnitus, is primarily to direct attention to alternative methods of alleviating their disability.

So, if a patient has a hand tremor and lived on their own, telephone amplification might be more beneficial than a behind-the-ear hearing aid.

Observation

When the patient is first seen, the clinician may already have some data available such as an audiogram. This does not negate the value of informally assessing their hearing by chatting to them and using a free-field voice test of their hearing, as a measure of speech understanding without lip-reading.

Patient Report

Patients vary considerably in their mental attitude to their symptoms and particularly others through social media and by seeing online adverts. In many, a major part of their management is based around correcting misinformation they may have.

It is important to find out about their lifestyle, how they spend their day and how their hearing loss affects their ability to participate in activities. Difficulty understanding speech in background noise is commonly reported as a major problem. Hearing aid amplification often does not help in this situation; it can be more helpful to change their position in the room further away from background chatter and focus on being able to see friends' faces, particularly their lips.

Companion Report

The importance of individualised assessment and treatment in healthcare settings is becoming increasingly apparent, and hearing healthcare is no different. Additionally, there is evidence to support the benefits of *household-centred hearing care*. This is where a family member or close friend is present during consultations to provide support, fostering a partnership between health professionals, patients and their family. Family members can have multiple roles: being a memory aid, giving emotional support, helping in decision making and being a sympathetic interpreter and transporter. Patients should always be advised that they can bring along a family member or close contact to every consultation.

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Use of Questionnaires in Assessing Disability

Just because many hearing-related questionnaires have been 'validated' does not mean that they are necessarily of any value in tailoring the management of an individual's hearing disability. However, there is one questionnaire, the Glasgow Hearing Aid Benefit Profile (1), that could be used, even by the less experienced clinician. The patient is asked to rate the degree of hearing difficulties in four pre-defined situations and four situations specifically chosen by them. The pre-defined situations are:

- Listening to the television with other family or friends when the volume is adjusted to suit other people.
- Having a conversation with one other person when there is no background noise.
- Carrying on a conversation in a busy street or shop.
- Having a conversation with several people in a group.

Questionnaires can also be used to evaluate the impact of hearing impairment on the psychological and social aspects of a person's day-to-day life, and to look for a change after an intervention, such as provision of hearing aids. These quality-of-life questionnaires are discussed further in **Chapter 14**.

PROVISION OF INFORMATION

Information given verbally during a consultation is mostly forgotten. Traditionally, verbal information has been supplemented with written pamphlets, but online information sources can be much more detailed and engaging. Examples are given in the **Further Reading** section at the end of this chapter.

In the UK, the *Royal National Institute for Deaf People* (RNID) (2) have considerable information available in leaflets and online regarding various aspects of hearing loss and its management. *Tinnitus UK* (3) have similar resources on hyperacusis as well as tinnitus. Their websites include signposts to external UK support services. Similar charitable or healthcare organisations exist in other countries. For example, the *American Speech-Language-Hearing Association* (4) has an online public portal with information on everything from the basics of hearing loss to hearing aid use and insurance.

In addition to leaflets and websites, support groups exist to give emotional and practical support as well as to train people in skills such as lip-reading. The *UK Association of Teachers of Lipreading to Adults* (5) website is a good resource, though classes are also advertised through other media.

Considerable resources exist for patients to increase their knowledge and confidence when it comes to hearing aid use. The website C2Hear (6) is a collection of videos developed to provide information on important aspects of hearing aid use, care and troubleshooting. The key aspects are understanding what hearing aids do and how they can help, being able to reliably fit the hearing aids on/in the ear, changing the batteries and ordering new batteries. The potential different functions of the hearing aid and cleaning the tubing and domes/moulds are also covered.

A key aspect of hearing aid use is habituation; getting used to wearing hearing aids and the accompanying change in sound. Patients should be encouraged to wear their new hearing aids in a variety of different situations, practise the dexterous aspects of hearing aid use and become accustomed to the different functions. Greater interaction with hearing aids in the early stages has been linked to improved outcomes and reduced levels of non-use.

CONVERSATION LISTENING TECHNIQUES

Human interaction is considerably enhanced by conversing with others, and effective conversation is as much about non-verbal (especially visual) information as it is about speech. Appropriate spectacles can do much to help conversation by allowing the observation of the speaker's face, lips and upper body. Having experienced the COVID-19 pandemic, most of us, including those with 'normal hearing', will have experienced the added difficulty of conversing when facemasks are being worn.

Conversing when there is competing noise from background music, as well as other people, can be difficult, even for those with no hearing impairment. Background music is frequently there to entertain staff rather than customers and polite requests to turn it down are usually acquiesced with. In most hospitality settings, it is possible to find quieter places which can be used for conversation. Pencil and paper can also help, provided the writing is legible.

The listener's attitude to not being able to understand what is being said is so important. One usually knows from various things when a funny story is being told. It is easy then to laugh when the others laugh without understanding why it is funny.

NON-PERSONAL AMPLIFICATION DEVICES

Having assessed an individual's hearing disability, the practitioner will have a fair idea as to whether a hearing aid will be suggested as the main method of alleviating their hearing disability. Most people do not wear their hearing aids all day and so modification of the listening environment can be helpful even when aids are being used.

Alerting Door, Telephone and Gas Alarms

Several technologies can be used to improve the effectiveness of doorbells. They can be modified to include a louder sound or to flash. Separate devices can be purchased which are situated inside the house and communicate wirelessly with the button on the door to either flash, vibrate or make a loud noise. There is also now a range of smart doorbells which can send notifications to a smartphone and can also include video feeds from an inbuilt camera.

Alerting devices should be regularly tested and updated, both to their noise levels and to position within the house to be heard, so that all in the house hear them.

The ability to hear alerting devices in the home should be considered in everyone that is hearing impaired, and it is often the house companion that identifies this is a problem. A companion dog can be helpful; if they bark, one has to find out why.

Hearing Loop Devices

It is common for hearing aids to be fitted with a loop function (also known as an induction loop system) that switches off the microphone and receives the incoming signal from a microphone or other direct sound source instead. Depending on the model of hearing aid, this can be toggled through by pressing a function button on the device or by using a smartphone app. Such hearing loops are available in a wide range of spaces, including cinemas, theatres and places of worship, and they can be invaluable in helping hearing impaired people to hear clearly in what would, otherwise, be rather difficult environments.

From a domestic perspective, a home induction loop fitted to the back of a chair can significantly enhance everyday living, from making and receiving phone calls to enjoying television.

Patients should be given information on what hearing loops are, how and when they can be of benefit, how to choose this function on their own hearing aids as well as being shown what the hearing loop symbol looks like so they know when they can use the function. There are other systems and technologies designed to interact with hearing aids, and they are discussed later in this chapter.

Wireless Microphones

One of the biggest issues with hearing aid technology is amplifying the signal of interest without amplifying background noise. Directional microphones in hearing aids can help but, in certain scenarios, a wireless microphone can provide greater benefit. The basic format of these systems is a transmitter, which includes some form of microphone to be worn or placed near the speaker or sound source, and a receiver that could be a hearing aid or other device. Frequency-modulated (FM) systems are often used in schools and they have a microphone worn or held by the teacher. Table microphones can be placed in between multiple speakers to capture conversation in a group and sound can stream via Bluetooth or similar technology to one or multiple people.

Infrared Systems

In more structured situations such as cinemas and theatres, infrared systems can be used. These systems involve people wearing a specific receiver that either contains a connected set of headphones or a neck loop allowing for magnetic field transfer to hearing aids.

Telephones

The first thing that can be done to improve the usability of telephones for people with hearing loss is to increase the volume. Most mobile phones and landlines also have hearing aid compatibility, meaning that the loop function on the hearing aid can be utilised to stream the speaker's voice directly. Mobile phones can be set to have a flashing light to indicate that someone is calling.

Those with an age-related hearing impairment frequently have poorer eyesight and are not necessarily wearing any glasses when the phone rings or if they want to use the phone in an emergency. Large buttons and large font text on mobile phones can therefore be very useful.

Subtitles and Closed-Captioned Television

Subtitling of television exists for most major platforms, both live and streaming. Automated live subtitling comes with understandable errors and delays.

When subtitling is available, speech reading becomes less necessary as most individuals who can speech read can also read text.

Speech-to-Text Applications

A variety of speech to text applications exist such as Google Live Transcribe and Ava. There are free, single fee and subscription applications for smartphones that vary in terms of accuracy, specificity and latency.

Workplace Support

The various systems and devices listed above can be used in individuals' homes, but they also have great value in the workplace. Inclusivity, equality and access for people with a hearing impairment in the workplace are important issues which are now legally supported. Workplaces in most countries have a responsibility to make reasonable adjustments to support workers with a hearing impairment and in the case of countries like the UK the purchasing and provision of assistive technology can be supported by grants from the government (e.g. Access to Work).

SIGN LANGUAGES

What Are Sign Languages?

Sign languages are languages expressed through manual articulation (hand movements) in combination with non-manual gestures such as facial expression. They convey meaning via vision rather than by hearing, and are most commonly, though not exclusively, used by Deaf populations. Deaf (with a capital D) refers to culturally deaf populations, who are mostly prelingually deaf, and for whom sign language is their primary means of communication.

The use of sign languages is well documented throughout history. First documented in the fifth century BCE by Plato, sign languages have been consistently used for communication with deaf people, as well as by those unable to speak.

It's important to understand that sign languages are complete, structured languages with their own grammar, syntax and structure. They are therefore different to non-verbal communication used by hearing populations, such as body language and gesticulation. In signed languages, specific hand shapes and movements represent specific words.

The structure of a signed language can differ significantly to that of its equivalent spoken language. For example, a British Sign Language (BSL) sentence is structured differently to the same sentence spoken in English. When individual BSL signs are used simultaneously with spoken English, this is not BSL, but Sign Supported English (SSE).

Sign language is not the same as fingerspelling. Learning a fingerspelling alphabet does not enable a person to communicate with Deaf people, any more than learning the Arabic alphabet renders one fluent in Arabic. Rather, fingerspelling is used to spell out names, and niche words or place names which do not have an allocated sign. Fingerspelling alphabets vary between sign languages, for example, the BSL alphabet uses both hands (**Figure 11.1**), whereas the American Sign Language alphabet uses only one hand (**Figure 11.2**).

Sign languages vary between countries, with over 150 recognised sign languages across the globe; some are legally recognised as official national languages. Sign languages also have regional variations, similar to regional accents.

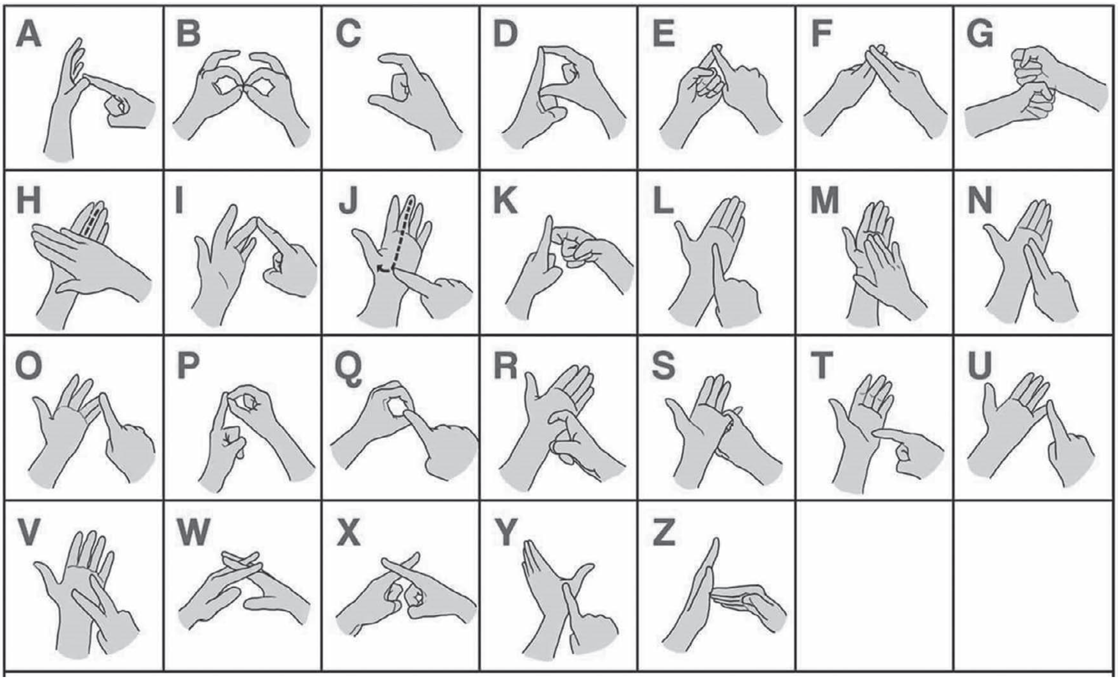

Figure 11.1 British Sign Language fingerspelling alphabet. (Credit: P Maxwell Photography, Shutterstock.)

BSL was recognised as an official language by the UK government in 2003. This has facilitated support and funding for the 150 000 people worldwide who use BSL as their first language, and increased awareness of Deaf culture.

Cultural Implications

Respect for Deaf culture is essential. Deaf culture is focused on identity, not disability. Sign languages are a complex and complete mode of communication for culturally Deaf individuals, many of whom do not consider their deafness a disability.

In the healthcare setting, this is relevant on several levels. Communication with Deaf patients and their families should be conducted with a professional interpreter, fluent in both spoken and sign language. Whilst many Deaf children and adults have a level of education which enables them to communicate in spoken or written language, it should not be assumed that communication via spoken or written language is adequate for the purpose of healthcare communication or consent for treatment. The acquisition of a basic level of sign language by healthcare professionals can be useful for establishing a connection with Deaf patients, but professional communication with Deaf patients requires a professional sign language interpreter.

In the past, cochlear implantation has been a controversial topic amongst Deaf populations. This is no longer the case. It is now largely accepted that cochlear implantation does not remove Deaf identity, but enables Deaf children to develop hearing and speech, which can enable them to enter mainstream education and employment. Deaf culture and cochlear implantation are not incompatible.

Newborn hearing screening programmes enable deaf babies to be identified, and hearing rehabilitation (which may or may not include cochlear implantation) to be offered to deaf children within their period of neuroplasticity. Babies and children from Deaf families who receive cochlear implants require exposure to spoken communication to develop hearing and speech. Auditory verbal therapy can facilitate this process. It should be noted

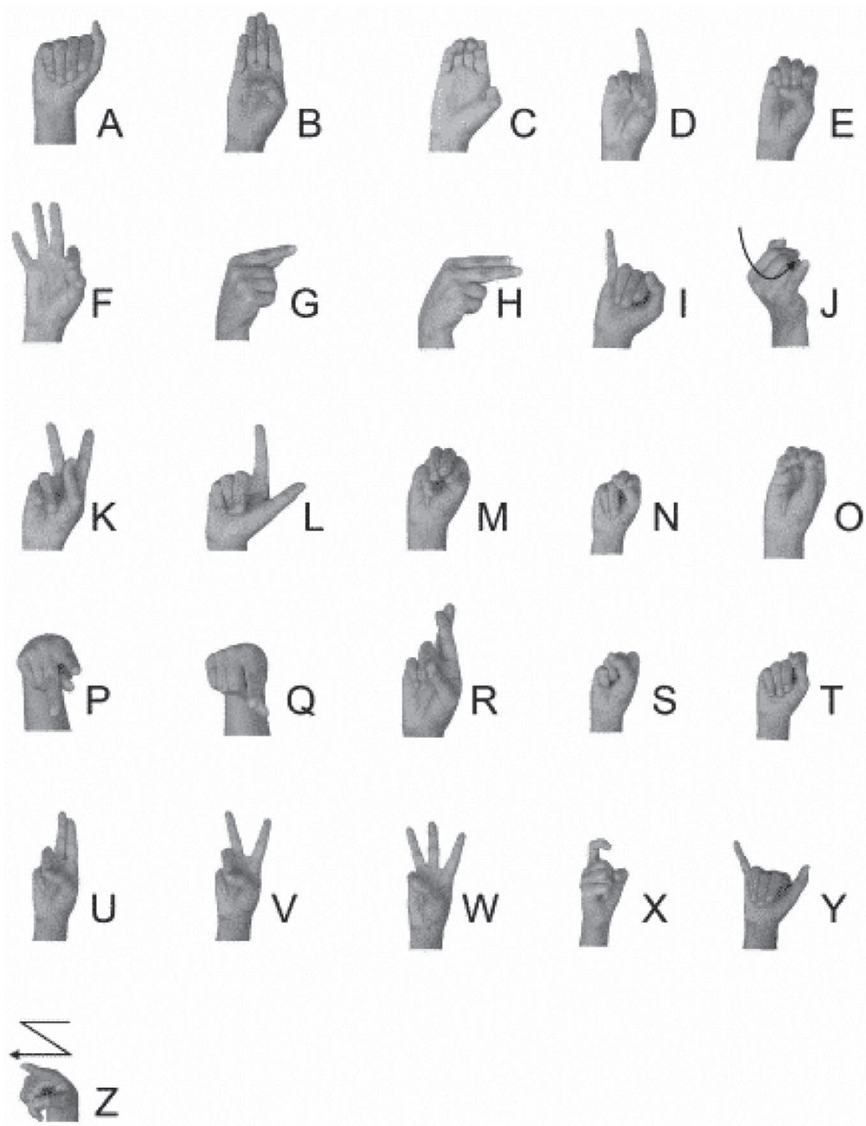

Figure 11.2 American Sign Language fingerspelling alphabet. (Credit: RRA79, Shutterstock. Copyright © 2008 StartASL.com.)

that all communication is to be encouraged, and that the use of sign language (e.g. with Deaf family members) should not be discouraged.

Makaton

Makaton is a language programme that uses a combination of signs (manual articulation), speech (spoken word) and symbols (printed images), to facilitate communication. Its scope extends beyond people with hearing loss; it can be used by people who cannot speak, or whose speech is unclear, and for children and adults with learning difficulties. Its main use, however, is in young children.

Makaton supports the development of communication skills such as attention, listening, comprehension, memory, recall and organisation of language and expression. In this sense, its focus is on communication and connection on a broader basis than hearing and speech. It is a valuable tool for the development of communication skills.

Makaton does not have its own structure and grammar. It is similar to SSE, in that signs are used alongside speech, in spoken word order, to help provide information about what that person is communicating. One of the goals of Makaton is to encourage speech, hence the importance of spoken language being used alongside the use of signs and symbols.

Research has shown that the use of Makaton encourages the development of speech and language. Learning Makaton does not impair the development of speech; children tend to stop using Makaton at their own pace, as they develop spoken language.

NON-SURGICAL AMPLIFICATION DEVICES (HEARING AIDS)

Overview

Non-surgical amplification devices, better known as hearing aids, collect sound from the outside world, and selectively amplify sounds at different frequencies to aid an individual's pattern of hearing loss and disability, via air conduction or bone conduction. Some surgical hearing solutions also work via amplification. These are described in **Chapter 12**.

The importance of non-surgical amplification devices cannot be understated. They can be lifechanging for both children and adults with hearing loss, facilitating access to sound, communication, speech, education and human connection. Whilst hearing aids are a definitive hearing solution for many patients, they also serve as a relatively risk-free intervention for those who might go on to have a surgical solution, e.g. facilitating access to sound and auditory cortex stimulation in profoundly deaf infants awaiting cochlear implant surgery, or as a trial of amplification in any patient considering a surgical intervention. They should therefore not be considered a dichotomous alternative to surgical hearing solutions, but an integral part of a holistic care pathway.

The increased visibility and acceptability of hearing devices; the availability of over-the-counter and self-fitting hearing aids, and the movement of technology companies into the 'hearables' space, have shifted the landscape.

The provision of hearing aids is traditionally the remit of audiologists and increasingly, of hearing aid dispensers who are not audiologically trained. Automated or self-directed hearing tests are increasingly used commercially, and for hearing screening prior to more focused audiological assessment. The training, expertise and acumen of audiologists cannot be replaced by technology, and Otorhinolaryngology surgeons and other clinicians must have a grasp of the principles underpinning these processes. Whilst audiologists are highly trained and experienced, not all clinics have access to audiology expertise. In clinics where ENT surgeons, audiologists and therapists/rehabilitationists work together, multidisciplinary working and joint decision making is important for optimal patient care and outcomes.

Children with hearing loss from any cause will have bilateral hearing aids fitted by trained paediatric audiologists, with the aids programmed carefully to their audiometric thresholds. They are followed up at intervals to assess benefit and to re-programme the aids as needed, guided by child and parent feedback on perceived benefit, speech testing and educational progress.

It could be argued that adults of working age should also have the same rigorous level of assessment and follow up. Many of the individuals in this age group will have an asymmetrical hearing loss, often with a conductive component. In addition, with the increasing provision of aids that can be electronically adjusted by the wearer, education in their usage is paramount. It is important for them to have regular follow up to ensure that the aids are functioning, being worn and providing benefit in domains such as education, training, employment and social participation.

Older adults with age-related hearing loss and NIHL may not be given the same level of attention. They are by far the largest group of people for whom hearing aids may be of benefit. Cost constraints in publicly-funded health systems may lead to long waiting times, short fitting appointments lacking time for adequate explanation and little or no provision for follow-up appointments. Many older adults may seek to obtain hearing aids in the private sector, even when publicly funded options are available, due to shorter waiting times and perceived (or perhaps actual) differences in cosmesis and functionality of the aids provided. Hearing aids are now heavily marketed to older adults through advertisements. Hearing aid provision in the private sector varies in terms of the level of training of the hearing aid dispensers, and the degree of after care and support for the patient. In many cases, hearing aids are provided 'over-the-counter' without being programmed to the patient's audiogram.

A randomised, double-blind, placebo-controlled trial published in 2017 (7) comparing best-practice audiology delivered hearing aids with over-the-counter hearing aids (which were not programmed to reflect the patient's audiogram) demonstrated patient benefit from both. Best-practice audiology delivered hearing aids were found to have a modest improvement in benefit compared with over-the-counter hearing aids. A systematic review on the same theme (8) concluded that self-adjustment during daily use of self-fitting hearing aids may produce equivalent or better outcomes than an audiogram-based prescription. Whilst the evidence for best fitting practice develops, one message is clear: amplification is more beneficial than no amplification, for patients with hearing loss.

Hearing Aid Candidacy

Audiometric evaluation is an integral part of determining hearing aid candidacy. This should include otoscopy, air- and bone-conduction thresholds and tympanometry as a minimum. Patients found to have hearing asymmetry, conductive or mixed hearing losses, and those who report tinnitus, pain or pressure, or vertigo should be assessed by an Otorhinolaryngology surgeon. Medical assessment should ideally be carried out prior to hearing aid provision, however, delays in medical assessment should not delay the provision of hearing aids, which can always be adjusted or reconsidered as the need arises.

The provision of a hearing aid is a multistage process that often involves more than one provider. **Table 11.1** lists the various stages in the usual order in which they are carried out along with who might perform each stage. At most stages, there are several possible providers, dependent on availability and skill mix.

Entry to this pathway can vary, with some patients being assessed medically then referred to audiology, and others having their first assessment by an audiologist. There is a NICE guideline which informs this process in the UK and which is freely available to overseas colleagues (9). This focuses on an individualised approach to patient's needs and preferences during the audiological assessment process. The guideline is aimed at primary care clinicians and specifies that referral to an ENT surgeon is needed if a patient experiences a sudden or rapid loss of hearing, or hearing loss associated with additional symptoms or signs.

Table 11.1 Stages in hearing aid provision as most commonly practised

	Professional involved				
Stages	**Medic**	**Aud Sc**	**Tech**	**HT**	**Com**
Candidacy	✓	✓	✓		✓
Medical assessment	✓				
Audiometric assessment		✓	✓		✓
Choice of ear/s	✓	✓	✓		✓
Choice of aid	✓	✓	✓		✓
Aid fitting		✓	✓		✓
Rehabilitation		✓	✓	✓	✓
Benefit assessment		✓	✓		

Abbreviations: Medic = medical personnel; Aud Sc = audiological scientist or audiologist; Tech = audiometric technician; HT = hearing therapist; Com = commercial personnel.

Aims of Medical Assessment

Medical assessment is usually provided by an Otorhinolaryngology surgeon and aims:

- To identify symptoms (tinnitus, fullness, fluctuating hearing, discharge, pain, vertigo) which might warrant investigation and management.
- To identify ear pathology such as active chronic otitis media that may merit investigation and management.
- To identify ears with a conductive impairment which could benefit from surgery to improve the hearing, following a hearing aid trial.
- To identify patients with an asymmetric sensorineural impairment who may warrant cross-sectional imaging to exclude a cerebellopontine angle lesion.
- The Otorhinolaryngology surgeon can then determine whether a patient is a suitable candidate for a hearing aid, and what their future medical, surgical and hearing needs are likely to be, within the multidisciplinary team.

Who Is Likely to Benefit from a Hearing Aid?

Any patient with a hearing impairment confirmed by audiometry who is motivated to try amplification should be provided with one or more hearing aids for a trial period.

The median threshold of individuals seeking advice about amplification is 45 dB (10) suggesting that patients tend not to present until their hearing loss is moderate. Data on hearing aid possession is surprisingly low (**Table 11.2**). With estimates of hearing aid non-use between 5% and 24% (11) and attempts to address patient motivation having little impact (12), offering hearing aids to all who might benefit from them remains an elusive goal.

Table 11.2 Possession of a hearing aid as a function of the degree of hearing impairment in the better hearing ear

Degree of impairment dB HL	% Possessing aid	% of Aid owners
< 25	< 1	7
25–34	10	8
35–44	30	18
45–54	35	21
55–64	55	18
65+	90	28

Reasons for hearing aid non-use have been explored (13) with lack of perceived benefit being the most common reason, followed by fit and comfort, maintenance and device factors. These factors indicate a need to improve the performance of hearing aids! Lesser factors for non-use included their appearance (4.6% of non-users) and complications of wearing hearing aids such as infection, tinnitus and wax impaction (1.6% of non-users).

Attempts to identify patients who have a hearing loss but do not use hearing aids have been made using UK Biobank data (14), demonstrating that patients from ethnic minority backgrounds and patients who live alone are less likely to have a hearing aid.

Monaural or Binaural Fitting?

The advantages of binaural hearing are well recognised, enabling the listener to localise sounds, and to hear in background noise. Indeed, binaural fitting is universal in children. But how does this translate to hearing aid use in adults? Are two hearing aids better than one?

A Cochrane review (15) on bilateral versus unilateral hearing aids for bilateral hearing impairment in adults concluded that published data were not sufficient to draw a conclusion regarding hearing or quality-of-life outcomes; a subsequent study of US veterans (16) demonstrated that bilateral fittings yield the best short- and long-term outcomes, and that if a single hearing aid is to be fitted, a better-ear fitting has a higher probability of long-term hearing aid use persistence than a worse-ear fitting. Factors influencing the use of binaural hearing aids include cost, personal choice, comfort, dislike of the 'blocked' feeling and lack of perceived benefit from a second hearing aid. Thus, whilst it makes sense to conclude that binaural hearing aids would be beneficial to the same extent as binaural hearing, this is not the case for every patient.

Monaural Fitting: Which Ear?

For all these reasons, most initial hearing aid prescriptions in adults are likely to be monaural rather than binaural. The question then arises as to which ear to fit and, in arriving at a decision, there are various factors to consider, the most important being whether the patient has symmetric or asymmetric hearing. An individual is usually considered to have asymmetric hearing if the pure tone thresholds in the ears are different by 10 dB or more averaged over the frequencies 0.5, 1, 2 and 4 kHz.

In asymmetric hearing losses, traditional teaching is to 'Fit the poorer hearing ear, unless the thresholds are poorer than 70 dB HL, when the better ear should be fitted'. When the hearing thresholds are poorer than 70 dB HL aids are technically more difficult to fit because they need a well-fitting mould to prevent feedback, gains may be insufficient and distortion can be a problem. The US veteran study mentioned above observed that fitting the better hearing ear led to a higher probability of long-term use. This may be because aiding the better ear tends to allow patients to achieve better speech discrimination. In a crossover study of side of use (17) 92% reported greater benefit with an aid in their poorer ear. A possible explanation for this is that their better ear is useful unaided, and a hearing aid in the poorer ear improves their ability to hear speech and discriminate in background noise.

As recommended by NICE (9), decisions regarding hearing aid provision should be individualised. It is often the case that patients have an innate preference for aiding one ear over the other, and therefore patient preference should be considered in both symmetric and asymmetric losses. Some examples of patient choice and preference include:

- A British taxi driver might wish to wear a hearing aid in his left ear, in order to hear passengers better.
- A patient who uses the telephone with their better ear may prefer to wear a hearing aid on their poorer ear, so that they continue their usual telephone use.
- A patient with dexterity problems, e.g. a previous stroke, may prefer to use a hearing aid with their more dexterous hand, or to trial two aids to see which they find most benefit and ease of use.
- A patient with active ear disease, i.e. untreated cholesteatoma or a discharging perforation, may prefer to use a hearing aid on their dry ear. Vented or skeleton moulds can also be useful in these circumstances.

Contralateral Routing of Sound (CROS) Hearing Aids

In those with a unilateral profound or complete hearing impairment and a contralateral better hearing ear, a CROS or BiCROS aid can be useful. These systems incorporate bilaterally worn devices which connect via Bluetooth technology. The device in the poorer hearing ear collects sound and delivers it to the device in the better hearing ear (or in the case of a BiCROS system, delivers amplified sound to the better hearing ear). They do not restore binaural hearing, and therefore do not improve directional hearing or hearing in background noise; they reduce the head shadow effect.

Hearing Aid Moulds

Following assessment for hearing aid candidacy, hearing aid and mould types can be discussed with the patient. Though this is the remit of the audiologist in most clinics, it is useful for Otorhinolaryngology surgeons to understand the different types, their benefits and pitfalls. Your patients and your audiologists will thank you for it!

William Hunter, the lesser-known Hunter brother, and founder of the Hunterian anatomy museum at the University of Glasgow, is thought to be the first person to describe the human pinna as a collector of sound, directing sounds into the external auditory canal. That this is beneficial is exemplified by the habit of cupping the hand behind the ear to enlarge its size to hear better. The pinna and the external auditory canal both influence the frequency spectrum of sound which finally arrives at the tympanic membrane (as discussed in **Chapter 1** and shown in **Chapter 10**, **Figure 10.2**).

The wearing of a behind-the-ear or body aid obviously alters the method of transmission of sound from the environment to the middle ear, as the sound-collecting mechanism of the pinna is not utilised. As the microphone of in-the-ear aids is in the external auditory meatus these aids theoretically are likely to be superior to other aids in sound localisation. In practice they are not, for two main reasons. Firstly, the concha is filled up by the aid and secondly, the effects of the pinna are mainly at the higher frequencies which are not amplified by standard hearing aids. Completely in-the-canal aids where the microphone is in the canal can potentially utilise the effect of the pinna at the frequencies it amplifies. All aids with an earmould inevitably have an acoustical effect, as the mould blocks and shortens the ear canal, modifying the resonance frequency of the ear (**Figure 11.3**).

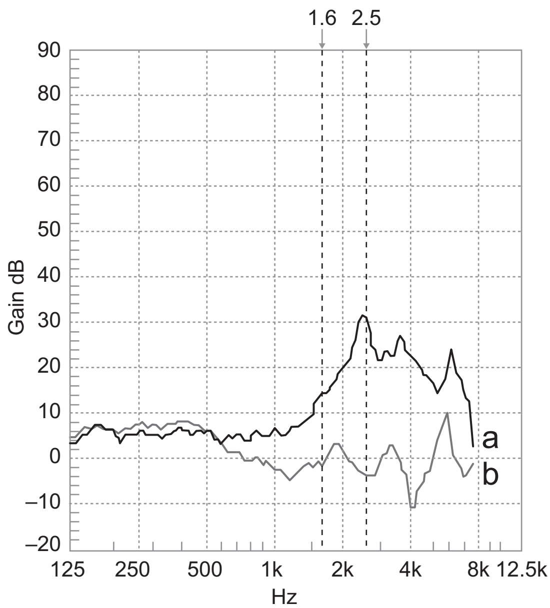

Figure 11.3 Insertion gain of a normal open ear canal **(a)** which is eliminated by wearing an earmould **(b)**.

Hearing Aid Styles

Air-conduction hearing aids come in a variety of forms (**Figure 11.4**), but all have a microphone to receive sound which is then amplified and delivered to the ear.

Because of the proximity of the microphone to the ear canal, in-the-ear (ITE) and completely-in-the-canal (CIC) aids tend to feedback at lower gains than behind-the-ear (BTE) aids and are usually only suitable for those with a mild-to-moderate hearing impairment.

Some patients express a preference for an ITE aid which they feel is less noticeable. This is not the case; modern BTE hearing aids are almost invisible in people who have hair, whilst an ITE aid can be noticeable as it obscures the natural shadows of the pinna. ITE aids may be easier to insert than BTE aids but can be harder to maintain due to moisture and wax build-up.

Power Supply

Hearing aids are battery-powered, and the smaller the battery size, the less power they produce and the more quickly they are used up. Smaller devices have less space for electronic features, however, this is compensated by the use of a separate remote, or app-based controls, which are increasingly popular since the ubiquitous use of smartphones.

Hearing aid batteries are small and need to be replaced frequently, an obvious environmental concern. In children, it is common to replace them on a time schedule (every week, for example) to ensure that they don't run out during use (which may not be reported by a young child). Adults will, of course, replace batteries when they notice the need to do so. Changing the batteries requires a degree of manual dexterity which may be an issue for some older people. Small batteries also present a risk of serious, and indeed life-threatening, injury if ingested by small children. For all these reasons, rechargeable batteries are becoming popular and will soon be ubiquitous.

Acoustical Data

Manufacturers provide acoustical data for each aid, measured on artificial ears (couplers). These are average results and are not the same as those achieved in real ears because of the varying effect of the ear canal anatomy in different patients. However, coupler data on *gain*, *frequency response* and *maximal output* are used for the initial choice of an aid.

An aid has to amplify sound sufficiently for it to be heard at a level that improves the hearing but not so loud that it is uncomfortable. The *acoustical gain* of an aid is represented as a frequency response curve (**Figure 11.5**) when the aid is set at maximal gain on the volume control with an input sound pressure level at the level of speech, usually 60 dB SPL.

Frequency response is ascertainable from the gain chart. The frequency response of most aids

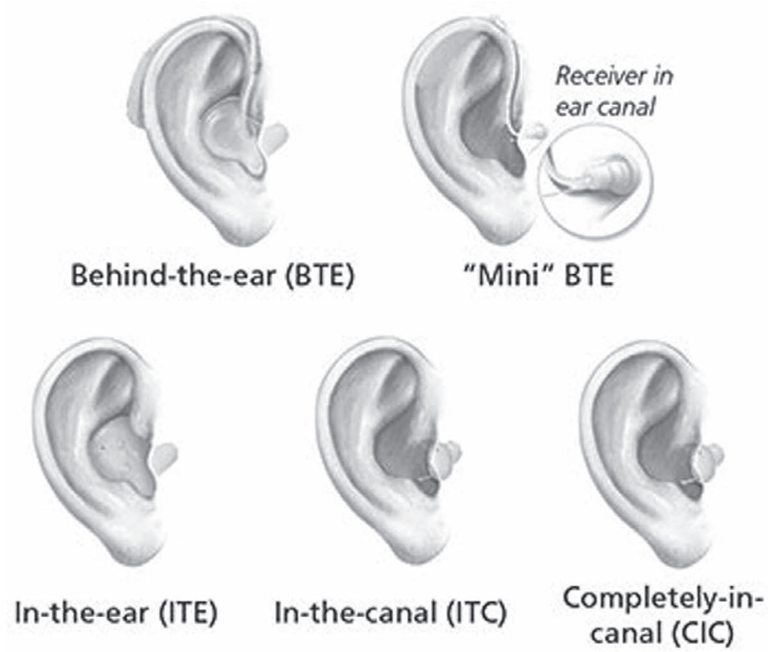

Figure 11.4 Types of air-conduction hearing aids. (Credit: NIH/NIDCD, U.S. Department of Health and Human Services.)

can be varied to give low- or high-frequency emphasis (**Figure 11.5**). Some aids have a greater range of modifications available than others. It is also possible to have a remote control, or smartphone app, that changes the frequency response for different listening situations.

Maximal output is presented as a similar response curve to that for gain, but with an input sound level of 90 dB SPL (**Figure 11.6**).

Real ear insertion gains are sound pressure levels measured with a small tube placed past the hearing aid mould into the ear canal. They can be used to guide hearing aid programming.

All modern digital hearing aids have the ability to provide more than one frequency output profile. They can automatically detect the nature of the person's listening environment and switch between different programmes chosen for each. They vary considerably between manufacturer but there are often four or five to choose from, including speech in quiet, noisy outdoor environments or listening to music. The hearing aids will automatically log total hours of use, and time spent in each defined listening environment. Although these facilities exist in some form in most modern aids, they aren't always used.

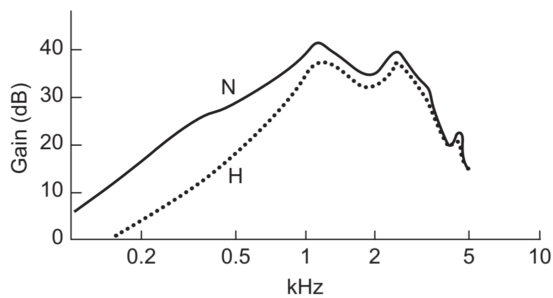

Figure 11.5 Gain characteristics of an aid represented as a frequency response curve. On this graph, the frequency response of two different settings are shown. N = normal tone setting; H = high tone setting.

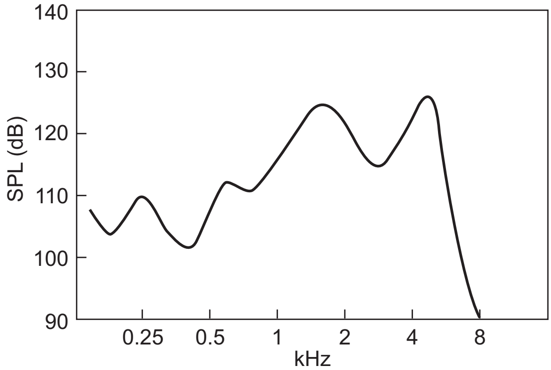

Figure 11.6 Maximal output of an aid, represented as a frequency response curve.

Directional Microphones and Background Noise Suppression

All hearing aid manufacturers have introduced innovations with the aim of making speech easier to listen to in a noisy background. Not all these modifications are successful and the apparent benefit from laboratory testing is seldom matched in real life (18). Many such aids require their output to be physically modified by the wearer for different listening situations and these changes are frequently (~35%) not made. Others make automatic modifications but then cannot deal with temporary variations such as looking sideways. The main side effect is that, by trying to separate speech from background noise, the speech signal itself can end up being slightly less distinct (19).

Tuition in Using Hearing Aids and Dealing with Problems

It is beneficial that considerable time and effort is spent explaining to the recipient how to put on the aid, use the controls, remove it and switch it off. This process should be practised several times, along with when and how to change batteries. Having a companion at these instruction sessions can be very useful in helping the subject when at home. Written information can be a useful supplement to (but not substitute for) hands-on training in practical hearing aid management, such as cleaning the aid (*see* **Appendix 2**). Commercial providers of aids have videos about how to use the aids and what to expect of them available on the internet.

Follow-Up Strategies

All recipients should be followed up by the provider around 4–6 weeks after hearing aid provision, to ensure that they are coping with the practical issues and that their disability has been improved. One study (7) found that 36% of subjects between 55 and 79 years of age had problems at follow up with inserting and managing their aids even though these had been discussed and demonstrated in detail at the time of issue.

Long-Term Usage of Hearing Aids

In the US veteran's study (16), usage was determined by the continued request for batteries to be provided up to 2 years post-aid provision. Short-term usage was more than 80% for all prescriptions. However, usage at 2 years in those provided with bilateral hearing aids for symmetrical hearing loss fell to below 60%. This was also the case of aids provided with a unilateral aid in the better hearing ear. Long-term usage was less than 40% in those provided with a unilateral aid in the poorer ear or those fitted only on one side in a symmetric impairment.

BONE-CONDUCTION HEARING AIDS

Bone-conduction hearing aids amplify sound via bone conduction, sending vibrations through the bones of the skull. The concept is the same as using a tuning fork against the skull, or testing bone-conduction thresholds during audiometry, and the same principles apply. The sound from a bone conductor bypasses conductive hearing losses in the outer or middle ear, transmitting vibrations directly to the cochlea. They do not require any part of the device to be placed in the ear canal and are, therefore, useful in patients who cannot wear conventional hearing aids due to chronic discharge, external auditory canal stenosis or congenital canal atresia. The popularity of bone-conduction headphones, which allow listeners to enjoy music transmitted via bone conduction without masking external sound, has made this technology commonplace (**Figure 11.7**).

Whilst the commonest indication for a bone conductor is a conductive hearing loss, they are also indicated for mixed losses (where they amplify bone-conduction thresholds), and for unilateral losses (where they transmit sound from the side of the deaf ear to the better hearing ear, reducing the head shadow effect).

Bone-conduction hearing devices can be surgical (an implant) or non-surgical (a type of hearing aid). Surgically implanted bone-conduction devices are discussed in **Chapter 12**. In this chapter, the role of non-surgical bone-conduction hearing aids is outlined.

Non-surgical bone-conduction hearing aids have several indications:

1. A temporary conductive hearing loss when a surgical solution is not indicated (such as in children with otitis media with effusion [OME]).
2. A trial device for patients to use whilst considering the potential risk and benefit of bone conduction, prior to proceeding with an implantable device.
3. A long-term hearing solution for people with a conductive, mixed or unilateral hearing loss who do not wish to have surgery, are not fit for surgery, or who do not wish to consider a surgically implanted device.

Bone-conduction hearing aids require the bone conductor to be applied firmly to the skull for optimal transmission of vibrations. This can be achieved

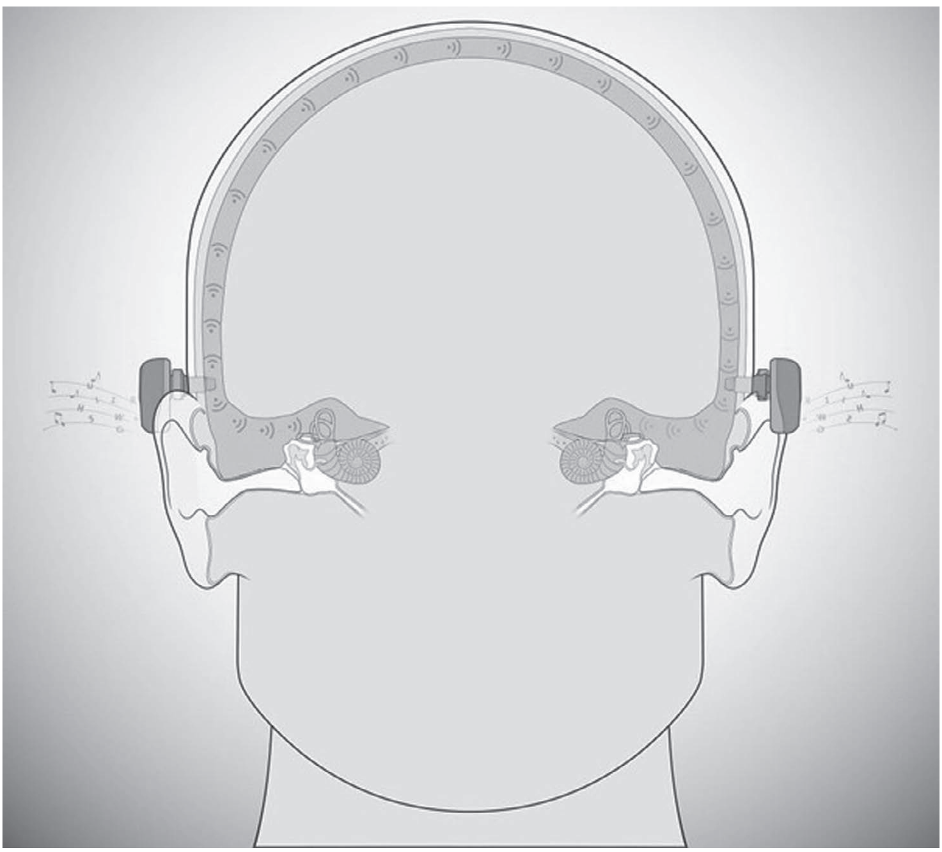

Figure 11.7 How bone-conduction hearing aids work, transmitting sound directly to the bones of the skull, bypassing the outer and middle ears. (Credit: Oticon Medical.)

by using a SoundArc (**Figure 11.8**), a soft headband (**Figure 11.9**), an adhesive device (**Figure 11.10**) or a modification of the patient's own spectacle frame (**Figure 11.11**).

The advantages of non-surgical bone-conduction hearing aids are that they are moveable and removeable, they amplify hearing thresholds, they are suitable for people with a discharging ear or an absent or stenotic ear canal and they have minimal risks or side effects.

Figure 11.8 The Cochlear BAHA® SoundArc. (Credit: Cochlear.com.)

Figure 11.9 A bone-conduction hearing aid on a soft headband. (Credit: Cochlear.com.)

Figure 11.10 The MED EL AdHear adhesive bone-conduction hearing aid. (Credit: Med-El.)

Their disadvantages include their aesthetic appearance, and the skin attenuation effect. This means their amplification potential can be dampened as the vibrations they produce need to be transmitted through hair, skin and scalp tissue, in order to reach the bone of the skull, and their fitting range is therefore more limited than an implanted bone-conduction device (**Figure 11.11**). Bone-conduction hearing aids work best in patients whose bone-conduction thresholds are normal or near-normal, whereas implanted bone-conduction implants can be fitted to patients whose bone-conduction thresholds are as low as 60 dB.

Figure 11.11 Bruckhoff glasses which contain a bone-conduction hearing aid within the frame. (Credit: Bruckhoff.com.)

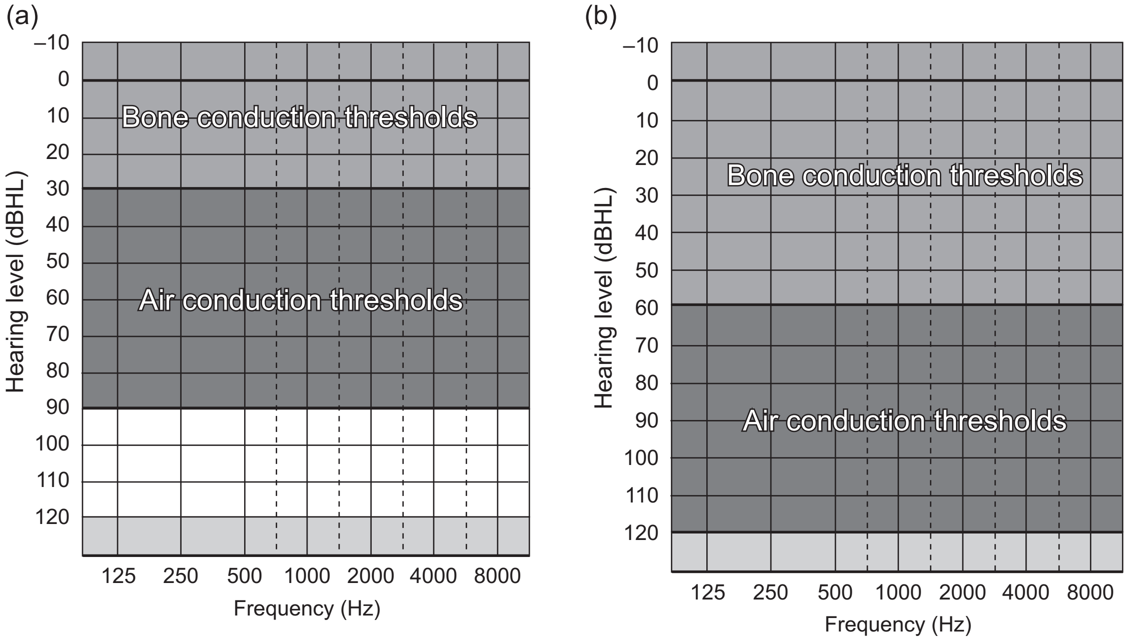

Figure 11.12 The recommended fitting range for **(a)** bone-conduction hearing aids and **(b)** implanted bone-conduction devices.

CONCLUSIONS

- Hearing impaired individuals can benefit significantly from instruction in conversation strategies, and from provision of flashing alarms, microphone systems, subtitles and other assistive devices.
- Sign languages are fully developed grammatical languages and an important part of Deaf culture. Makaton is an aid to spoken language most commonly used in young children.
- Hearing aid provision includes time spent explaining how to insert, adjust, remove and clean the aids, as well how and when to change batteries.
- All the aids currently available are digitally programmable and can be altered to 'match' the audiometric configuration of the hearing loss and the listening conditions, such as speech intelligibility in noisy situations.
- Short-term usage of hearing aids is around 80%, suggesting that people do find them beneficial. Usage of aids can fall considerably with time in older people.
- Bone-conduction aids are ideal for temporary conductive hearing loss in children, as a trial for those considering bone-conduction implants, and as a long-term option for those with meatal stenosis, meatal atresia or chronically discharging ears.

ACKNOWLEDGEMENT

Dr Jack Holman PhD is thanked for his considerable input to this chapter.

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12

Surgical management of hearing impairment

GENERAL PRINCIPLES

Sensorineural losses are managed by amplification i.e. a hearing aid, or in cases of profound sensorineural loss, cochlear implantation (**Chapter 13**). A variety of surgical options also exist for conductive and mixed losses. These aim to reverse the conductive component through reconstruction or mobilisation of middle ear mechanics, or to provide amplification of sound through bone-conduction implants, or middle ear implants.

Middle ear surgery is not always successful in improving the hearing. Surgery may fail to close the air–bone gap and carries a risk of inner ear damage. This can manifest as a sensorineural loss and sometimes a dead ear. Whilst hearing aids can be provided for both ears, most surgeons would not operate on both ears at the same time, and surgery is usually initially limited to the poorer hearing ear in the first instance.

As the magnitude of the air–bone gap influences the potential benefit of middle ear surgery, audiometry with appropriate masking of the bone-conduction thresholds is essential to study in detail in both ears (**Chapter 3**). In ears with chronic otitis media, otoscopy will identify abnormalities of the tympanic membrane and often ossicular chain problems. These findings should be compared with the magnitude of the air–bone gap with the objective of considering management options and potential surgical or non-surgical approaches.

Whilst it is wise to look for previous ear operation scars, these may not be apparent in patients who have had trans-canal or endoscopic ear surgery. Asking the patient about any previous ear surgery, and examining their previous case notes, is advisable. It is also advisable to discuss anaesthetic options. Middle ear surgery can be carried out under general anaesthesia, or under local anaesthesia with or without sedation. This choice may be influenced by the surgeon's experience and expertise, however patient choice and perspective should also be considered. General anaesthesia carries risks, and an advantage of surgery under local anaesthetic is that patients may perceive an improvement in their hearing perioperatively.

This chapter is far from a definitive guide to surgical management of hearing impairment. It is an overview of the general principles of surgical decision making, in the context of hearing. Readers wishing to learn surgical techniques will be best served not by a book, but through learning, practising and refining their surgical skills on simulation courses, and ideally completing a dedicated otology fellowship. Experienced otologists hone their techniques over many years of deliberate practice and self-audit, mindful reflection and careful decision making in partnership with their patients.

In **Chapter 4**, we considered the pathophysiological interpretation of an air–bone gap, and the option of surgical interventions to reverse conductive losses in tympanic membrane pathology and ossicular chain pathology. In this chapter, these themes are revisited with a focus on decision making in the surgical management of hearing impairment.

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The importance of consistent, ideally standardised outcome reporting after ear surgery, is discussed in **Chapter 14**. It is important to be able to predict the likely benefit from surgery. This is not necessarily the same as the audiometric outcome, or technical benefit, as will be discussed later.

MANAGEMENT OPTIONS

In those with a persistent air–bone gap of at least 20 dB due to middle ear pathology and a pure conductive or mixed hearing impairment there are multiple management options. These require to be fully discussed with the patient, and any accompanying companion, of their potential objectives and likely outcomes.

The majority of children and adults with ear symptoms are initially seen by non-specialist otolaryngologists and the audiogram should be checked to ensure that the audiogram was carried in a sound deadened environment, with the relevant masking of bone-conduction thresholds. The pathology responsible for the middle ear conductive component should have been identified by otoscopy.

In those with at least a 20 dB conductive component to their impairment in one or both ears and who have no other symptoms, and in particular an ear discharge, the clinician will then want to establish several aspects following discussion with the patient and their companion.

- Their associated hearing disability (*see* Chapter 11).
- The patient's and their companion's management objectives.
- Any previous management and their outcomes. Many will already have tried a hearing aid but this does exclude checking that it was appropriately prescribed with management follow-up.
- Their general health to identify contraindications for surgery that involves either a local or general anaesthetic.
- Their current medication, especially anticoagulants.
- Their medical 'insurance' background. This depends considerably on their country's method of provision of medical care.

The clinician can then discuss with the patient (and their companion) the most relevant management options that they, or others more skilled, could provide. In this discussion, the non-surgical management options covered in Chapter 11, must be included. In addition, the financial and practical, personal commitment to the provision and follow-up programme associated with each option, must always also be covered.

1. 'Watchful waiting' – patients may not wish to have any intervention especially if their conductive impairment is unilateral or mild. They can also have accessory aids such as louder alarm systems and text on their television screens (*see* Chapter 11).
2. Trial of a conventional ear level hearing aid.
3. Trial of a bone-conduction hearing aid.
4. Consideration of a bone-conduction hearing implant.
5. Consideration of middle ear surgery.
6. Consideration of middle ear surgery in addition to options 2, 3 or 4.
7. Consideration of parallel management of the pathological condition, in addition to options 2–6.

AUDIOMETRIC OUTCOMES OF MIDDLE EAR SURGERY TO IMPROVE HEARING

In surgery to improve hearing, the technical objective is to reduce the conductive defect as much as possible without causing a sensorineural impairment. An audiometric measure of each of these parameters is required from the pure tone audiogram (PTA). Audiologists will usually measure air-conduction thresholds at 0.5, 1, 2 and 4 kHz. Air-conduction thresholds should also be measured at 3 kHz (as per standardised Outcome Reporting Guidelines Chapter 14), and at 6 and 8 kHz to measure high frequencies, which are most frequently damaged by middle ear surgery. Bone-conduction thresholds at 0.5, 1, 2 and 4 kHz are usually measured, and should also be measured at 3 kHz, though not at 8 kHz as the calibration of bone-conduction headphones is not accurate at the highest frequencies.

Measures of Change in the Conduction Defect

The magnitude of the conduction defect is best measured by the air–bone gap, which is measured as a four-frequency average (FFA) over 0.5, 1, 2 and either 3 or 4 kHz, depending on the guidelines followed. Both air- and bone-conduction thresholds should be available pre- and postoperatively. The main debate is whether to use the preoperative, the postoperative or the better of the two bone-conduction averages, to get an accurate impression of change in the conductive deficit. If preoperative thresholds are used, the residual conductive defect is likely to be underestimated. The postoperative average can be better than the preoperative one because of the Carhart effect, or it can be poorer because of sensorineural damage. When postoperative thresholds are used, sensorineural losses associated with lessening of the conduction defect will be missed. Using the better of the two lessens both deficiencies.

This is demonstrated by considering the preoperative audiogram **Figure 12.1** with the two possible outcomes in **Figures 12.2** and **12.3**. In both, the air–bone gap has been closed but the results are very different (**Table 12.1**). So what has also to be reported is the change in bone-conduction thresholds at 4 kHz. In **Figure 12.2** the air–bone gap has been over-closed using the preoperative bone-conduction thresholds. These are better postoperatively because of the Carhart effect. In **Figure 12.3**, although the air–bone gap has been closed using the postoperative bone-conduction thresholds, these are poorer than the preoperative bone-conduction thresholds, because of inner ear damage manifesting as a sensorineural impairment. The air–bone gap in comparison to the preoperative bone-conduction threshold is of course

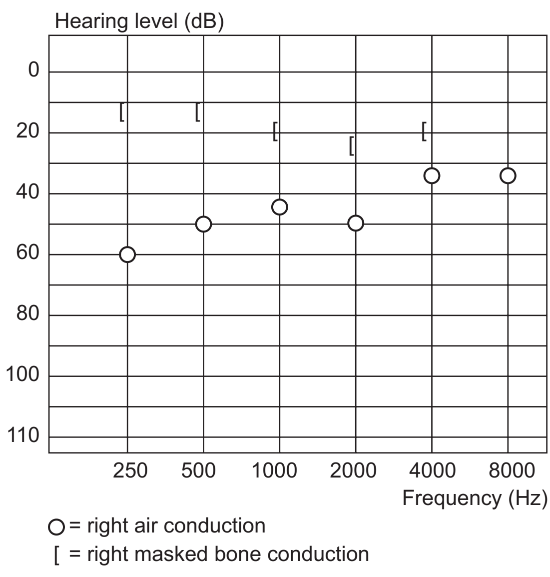

Figure 12.1 Ear with a moderate conductive impairment.

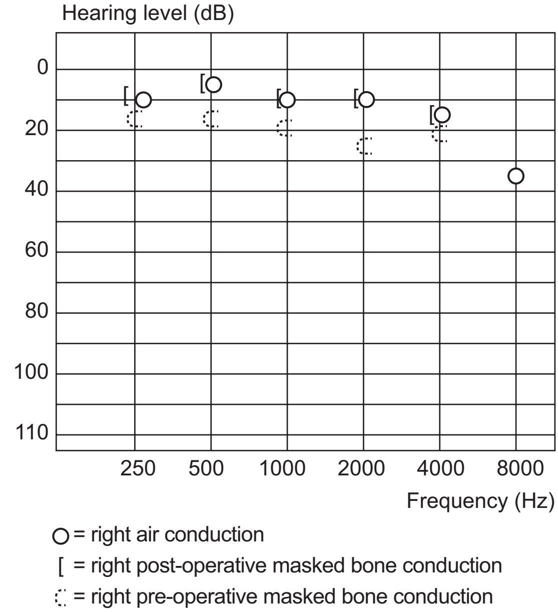

Figure 12.2 A good postoperative result from surgery on an ear with the preoperative audiogram in **Figure 12.1**.

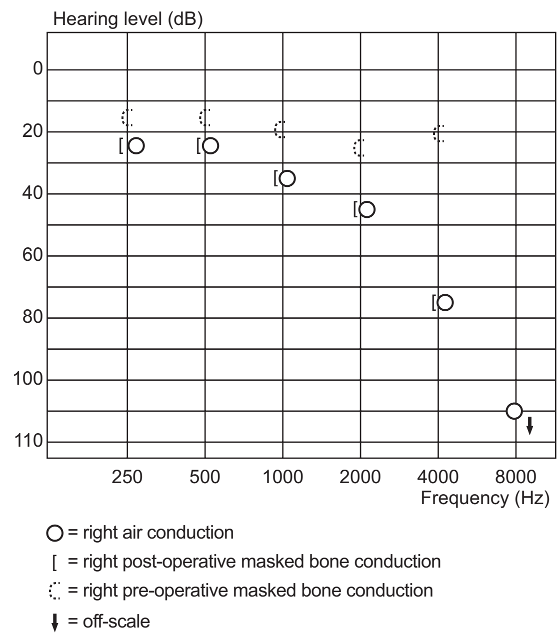

Figure 12.3 A poor postoperative result from surgery on an ear with the preoperative audiogram in **Figure 12.1**.

Table 12.1 Thresholds from **Figures 12.1**, **12.2** and **12.3**

		0.5 kHz	1 kHz	2 kHz	Total	Average
Figure 12.1, preoperative	AC	50	45	50	95	32
	BC	15	20	25	60	20
Figure 12.2, postoperative	AC	5	10	10	25	8
	BC	5	10	10	25	8
Figure 12.3, postoperative	AC	25	35	45	105	35
	BC	25	35	45	105	35
	Figure 10.2		**Figure 10.3**			
Preoperative ABG	12		12			
Postoperative ABG						
Using:						
Preoperative BC	−16		+3			
Postoperative BC	0		0			
Better BC	−16		0			

Abbreviations: AC = air conduction; BC = bone conduction; ABG = air–bone gap.

not closed but it is smaller (28 dB reduced to 15 dB). This is an example of technical results reporting (i.e. air–bone gap closure) which will not lead to benefit for the patient.

Measures of Damage to the Hearing

Surgical damage to the inner ear is more likely at the higher frequencies and hence worsening of the air-conduction threshold at 8 kHz would be the most sensitive measure. Damage localised to this frequency is not as detrimental to the hearing of speech as damage at 4 kHz. Hence worsening of the hearing at 4 kHz by air conduction is a better clinical measure, though it will include patients whose hearing is worse due to an increase in the conductive defect as well as those suffering sensorineural damage. To distinguish between such cases is not clinically important, what matters is the overall number. In the example shown (**Figures 12.1** and **12.3**) the 4 kHz air-conduction thresholds are poorer by 40 dB postoperatively. As well as reporting at 4 kHz, the lower frequencies should also be reported. As explained in **Chapter 4**, the bone-conduction thresholds are affected by a conductive defect. They may improve with surgical correction of the conductive defect. If they get poorer it is almost certainly because of inner ear damage. It is reasonable to report any change for the worse in bone-conduction thresholds as a measure of sensorineural damage. Thus, in the previous example (**Figures 12.1** and **12.3**) there is a worsening of the bone-conduction average by 15 dB.

PATIENT BENEFIT

Benefit is an essential theme to consider when consenting patients for surgery to improve their hearing. During discussion and consent (**Chapter 14**), clinicians must explain to patients not only the aims and risks of surgery, but also the likely outcome in terms of benefit the patient will perceive, or experience. The main reason why patient benefit is not equitable to technical results is that a patient has two ears, and surgery at any one time is on one ear only. Frequently, this involves the poorer hearing ear, for the very valid reason that, in the event of the hearing being damaged, the patient will consequently be less disabled. Unfortunately, the contrary is also the case, that if the hearing is improved, the benefit will be less than if the same improvement had occurred in the better hearing ear.

To explore this further, we are going to consider three patients with the same moderate, conductive hearing impairment in the right ear as in **Figure 12.1**. The pathology responsible is otosclerosis and their bone-conduction thresholds are ≤ 30 dB HL. They have no other otological symptoms such as tinnitus that could affect their management.

They are all going to have technically successful surgery with closure of the air–bone gap but with no change in the preoperative bone-conduction thresholds.

The difference between these three patients is the hearing in their left ear.

Patient A **(Figure 12.4 [a] prior to surgery)** has no hearing impairment in the left ear.
Patient B **(Figure 12.5 [a] prior to surgery)** has a mild conductive impairment in the left ear due to otosclerosis.
Patient C **(Figure 12.6 [a] prior to surgery)** has a bilateral, moderate impairment in both ears due to otosclerosis.

Following technically successful surgery in their right ears.

Patient A **(Figure 12.4 [b] post-surgery)** has no impairment in either ear.
Patient B **(Figure 12.5 [b] post-surgery)** has no impairment in the right ear but still a mild conductive impairment in the left ear.
Patient C **(Figure 12.6 [b] post-surgery)** has no impairment in the right ear but still a moderate impairment in the left ear.

Comparative benefits between Patients A, B and C.

Because all three patients have a similar preoperative conductive impairment with bone-conduction thresholds ≥ 30 dB HL, following surgery their right ears are no longer impaired.

From a Delphi poll of specialist otological surgeons, there is agreement that the greatest benefit that can be achieved following surgery, is that there is no longer a hearing impairment in the operated ear (1). This has been achieved by all three patients, but their postoperative position is different and, in particular, as to whether subsequent surgery in their other ear might then be beneficial.

Another benefit criterion that has been suggested is in the 'Belfast rule of thumb' (2). This suggests, that as well as improving the FFA to be ≤ 30 dB HL, the difference between the FFA of two ears should also aimed to be no greater than 15 dB.

This part of the 'Belfast rule' was not supported by the Delphi surgeon's subsequent poll and is no longer used as an important objective (1) but it still

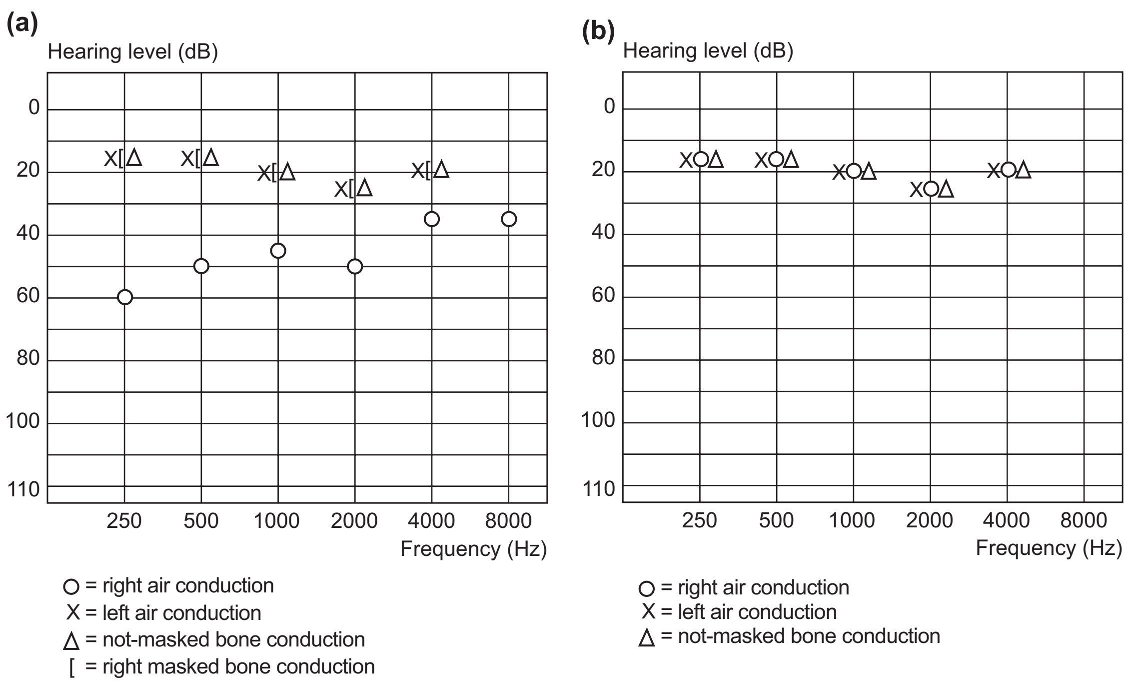

Figure 12.4 **(a)** Patient A, preoperative. **(b)** Patient A, postoperative.

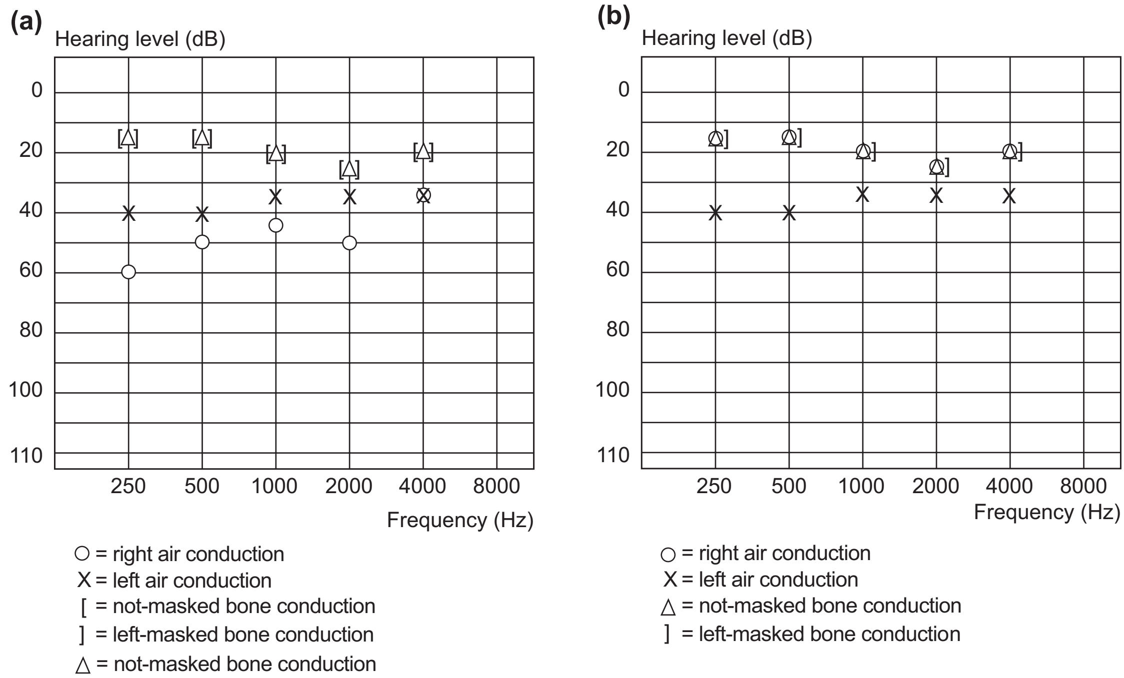

Figure 12.5 **(a)** Patient B, preoperative. **(b)** Patient B, postoperative.

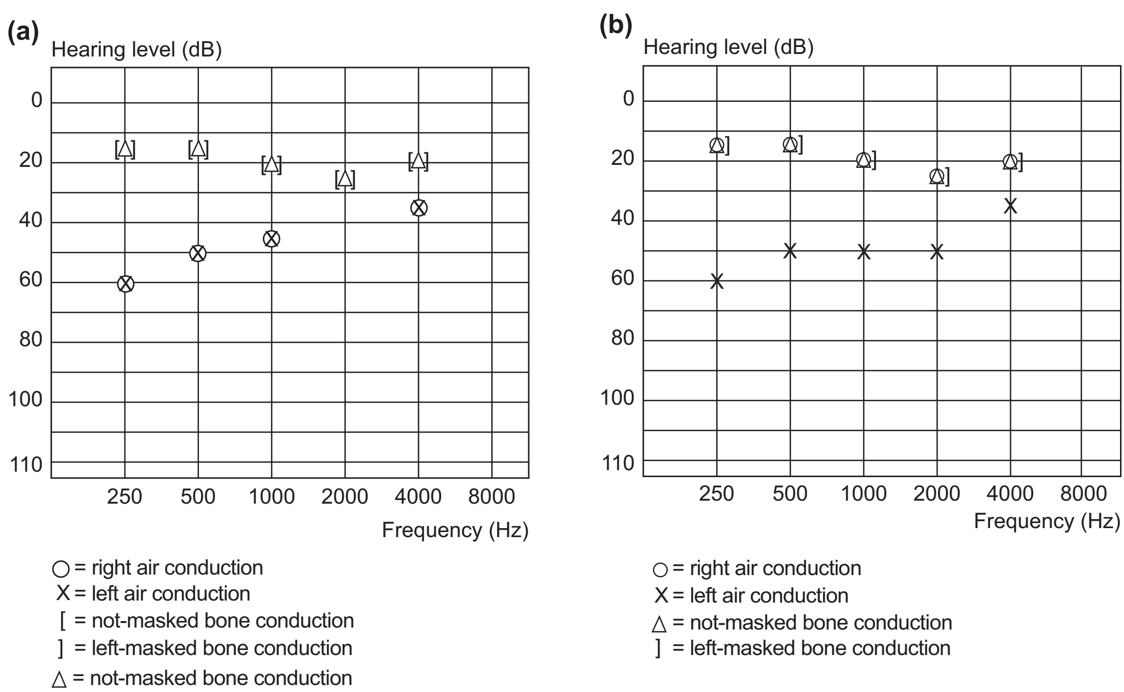

Figure 12.6 **(a)** Patient C, preoperative. **(b)** Patient C, postoperative.

does need consideration as an outcome in the overall management of a hearing-impaired individual.

In Patient C (**Figure 12.6a**), prior to surgery satisfied this 'Belfast rule', as there was no (0 dB) difference in the FFA between each ear. However, following surgery, the difference became 26 dB. However, they might be so delighted with no longer having any impairment in the right ear, they might request the surgeon to operate on their left ear. They would be hoping that the operation on the left ear would result in them no longer having an impairment in that ear rather than making in within 15 dB of the other ear. The idea of wearing a hearing aid in their left ear would not be appealing.

Experienced surgeons who have received such a request to have binaural surgery, regularly advise that the benefit of the second operation would be less and the risk of developing a sensorineural loss had to be remembered. Those involved with providing hearing aids for bilateral conductive impairments are well aware that a high proportion of them prefer to mainly use a unilateral rather than bilateral aids.

The position of Patient B (**Figure 12.5**) is slightly different. Preoperative, the difference between the ears was less than 15 dB and, following surgery it was about the same. The non-operated ear has a mild hearing impairment and their overall disability following surgery will be less having had the operated ear changed from a moderate impairment to having no impairment. In this patient, surgery on the other ear is unlikely to be requested, mainly because the difference in FFA between the ears is less than 15 dB. Nor is it likely that they would wear a hearing aid in that ear.

GLASGOW BENEFIT PLOT

Because of the clinical requirement of having to advise many patients with a conductive or mixed hearing impairment of the potential benefits of middle ear surgery, the Glasgow Benefit Plot was developed to help discuss the potential benefit of middle ear surgery as well as reporting the outcomes of a case series (1).

The value of a Benefit Plot was evaluated before and after otosclerosis surgery (1), otosclerosis having been chosen as the condition where closure of the air–bone gap is frequently achieved by fully trained surgeons.

Figure 12.7 from that paper shows the potential changes from a specific patient's preoperative impairment category to postoperative impairment category in the 41 Glasgow patients where the audiometric data was also available, at least 6 months following surgery.

The current Glasgow Benefit Plot (**Figure 12.8**) was developed from this paper which also includes several different pre-and post-surgery patient categories.

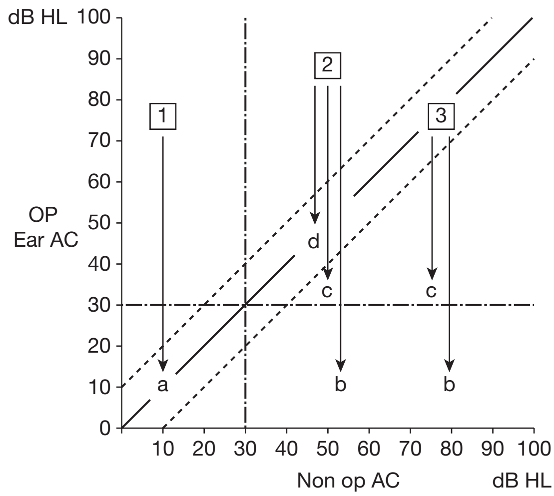

Figure 12.7 Potential changes from preoperative impairment group to postoperative impairment category, representing different typles of benefit (1).

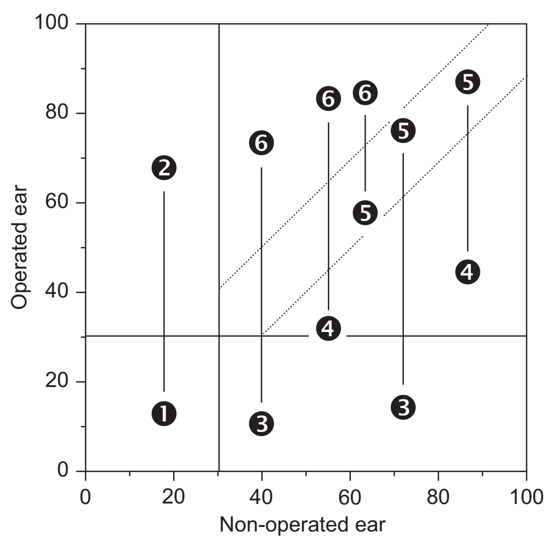

Figure 12.8 Glasgow plot (3).

The four-frequency average (FFA) of the ear being considered for surgery is positioned on the vertical axis of the Plot and the FFA of the other (usually better) ear on the horizontal axis.

The preoperative categories in those with a bilateral impairment will usually start above the plot's 30 dB HL horizontal line in areas 4, 5 and 6, and area 2 when the impairment is unilateral.

This figure is now mainly used pre-operatively to help evaluate the likely outcome of surgery, taking into consideration the hearing in the other ear. In a case series, Plots can also be recorded alongside an audiologcal analysis to help predict likely outcome in a specific patient. It can also help predict the potential benefit of an additional hearing aid following surgery.

An example of reporting the pre-/ postoperative use of the Glasgow Benefit Plot of patients A, B and C is illustrated in **Figure 12.9.**

A parallel, audiometric analysis was also carried out on the 41 patients reported following stapes surgery in Browning et al 1991 (1).

Overall 45% (18/41) of the patients had no impairment post-surgery. One of the main reasons for this percentage not being higher, is that 19 of the patients had a mixed impairment, as evidenced by having a severe impairment prior to surgery. In them, the mean improvement in the air-conduction

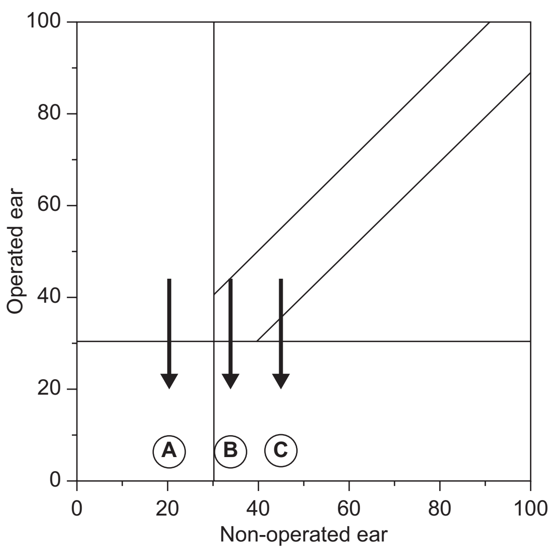

Figure 12.9 Glasgow Benefit Plot used for the three patients (A, B, and C) presented earlier in the chapter.

was 28 dB which resulted in them having a moderate impairment rather than a severe impairment. This would give them considerable benefit even though they might be provided with a hearing aid. The benefit from a hearing aid is easier to obtain in those with a moderate impairment rather than those with a severe impairment.

The conclusion is that when reporting the results in a case series, the magnitude of an improvement in the FFA in the operated ear should also be a reported outcome alongside any change in WHO category of impairment. The magnitude of the mean improvement for similar operations can also be used when assessing the likely change in the Glasgow Plot in patients who are being evaluated as to the likely benefit from surgery. So, at Glasgow Royal Infirmary in 1991, the mean improvement data of otosclerosis surgery improving the FFA by a mean of 28 dB could be used, along with their pre-surgery positioning of both ears on a Glasgow Benefit Plot, to predict their likely position post-operatively and the clinical benefit thereof. This numerical data cannot, of course be used by others including those at Glasgow Royal Infirmary in 2025.

SPECIFIC MIDDLE EAR PATHOLOGICAL CONDITIONS THAT CAN CAUSE A CONDUCTIVE HEARING DEFECT

This section considers some more specific aspects of the three main conditions that can cause a conductive hearing impairment due to middle ear pathology; otosclerosis, otitis media with effusion and chronic otitis media. The adult population frequency of these three conditions are detailed in Chapter 4.

Otosclerosis is essentially an adult condition, whilst otitis media with effusion and chronic otitis media can affect both children and adults.

Otosclerosis

Otosclerosis is a pathological condition that affects the temporal bone. Its histological extent varies considerably, but the main area that causes stapes fixation is around the cochlea, giving a resultant middle ear conductive hearing impairment without any otoscopic abnormalities.

The clinical prevalence in the adult population is 2% (CI 1.5–2.7) and it is as frequent in men as in women. The prevalence increases with age but is more clinically evident in those aged 41–60 years, in whom an associated age-related sensorineural impairment is infrequent. Thereafter, in those aged over 60 years, a mixed impairment understandably becomes more frequent.

The diagnosis of otosclerosis is not particularly aided by radiology or tympanometry with reflexes (see Chapter 4).

The usual presentation is of a unilateral conductive hearing impairment with no obvious otoscopic abnormality. Subsequent stapes fixation in both ears is common but, to date, cannot be prevented by medical therapy including fluoride (4).

The surgical technique to improve the hearing has dramatically changed since the 1970s. At that time, removing the stapes footplate and replacing it with a blob of fat onto which the stapes crus was then placed, was correctly described as a stapedectomy. However, a true stapedectomy is now rarely performed.

Currently, with microscopic vision and fine ear instruments, a stapedotomy is performed by drilling the footplate, and inserting a prosthesis attached to the incus **(Figure 12.11)** after partial removal of the stapes crus. Variations in the materials and design of the prosthesis are numerous, as are the techniques used to create the stapedotomy, including laser surgery.

Such surgery requires considerable expertise with the appropriate instruments, that can only be obtained by having had considerable previous experience in middle ear surgery. In **Figure 12.10**, it looks as if a partial stapedectomy has occurred with removal of one crura with part of the footplate, necessitating the resultant gap having to be filled with a fat plug. Suction in the middle ear with this gap could have resulted in a sensorineural hearing loss.

The non-surgical options, covered in Chapter 11, can of course be tried before deciding about surgery. Indeed, it is many clinicians' opinion that in those with otosclerosis, there should have been a formal trial of hearing aid/s before considering surgery. Otosclerosis surgery should only be carried out by those who have had a sub-specialist ear surgery training because of the dangers of causing a sensorineural hearing loss, as illustrated earlier in this chapter (**Figure 12.3**). In many cases, this might mean that the patient should be referred to a more experienced surgeon who has actually analysed their audiometric outcomes, rather than giving an enthusiastic guess.

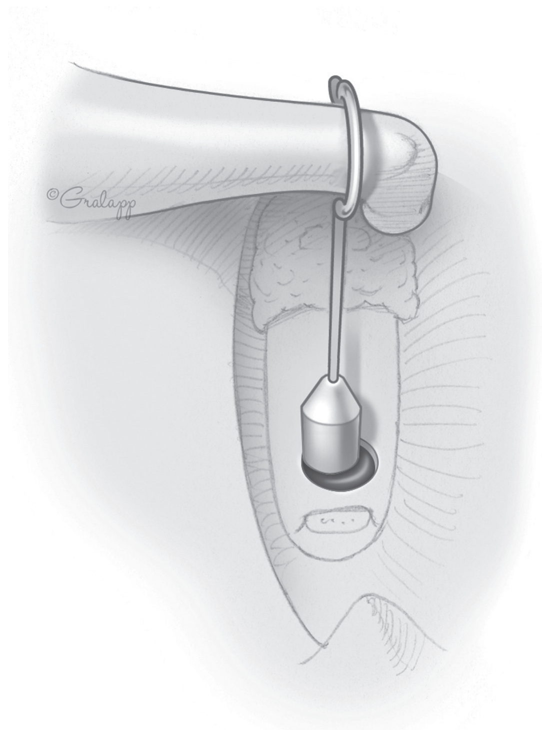

Figure 12.10 Stapes piston prosthesis *in situ* following a stapedotomy operation. (Credit: Chris Gralapp.)

Otitis Media with Effusion

It is incorrectly considered that otitis media with effusion (OME) is primarily a condition of childhood whose management options have already been covered in Chapter 8 and include adenoidectomy and sometimes tonsillectomy.

In children, the condition is most frequently bilateral, so ear surgery is usually caried out bilaterally, rather than just in the poorer hearing ear. This is because the insertion of a ventilation tube (VT) through an anterior/inferior myringotomy slit is unlikely to damage the ossicular chain or cause a sensorineural hearing loss.

What has not been emphasised sufficiently is that children managed with VTs might benefit from more frequent postoperative follow-up assessments. This is because of the high percentage of blockage (~ 20%) and infection (15%) rates (6) in the 6-month period before normal extrusion of the grommet. In this study, otoscopy was less reliable than tympanometry in diagnosing blockage that is usually accompanied by a return of a conductive impairment. How best to deal with blocked tubes has yet to be answered.

Extensive clinical practice in aiding those with an age-related hearing loss, has made the frequency in them of OME evident by them having problems of self-management by performing Valsalva manoeuvres.

The MRC National Study of Hearing did not study adults over the age of 80 years but the percentage of adults with eustachian tube dysfunction on tympanometry (*see* **Table 4.5**) in those between the ages of 61–80 years is 1.5 %. In them, VTs to improve the hearing is not usually advised as it does not overcome the disability due to their age-related hearing impairment. Better to encourage auto-inflation that most will have already used frequently on holiday, aeroplane flights.

Chronic Otitis Media

The classification of ears with chronic otitis media is presented in otoscopic photographs (*see* **Figures 2.6–2.9**). Many patients with active mucosal or active squamous disease will merit surgery to eradicate the active diseases and, in them, if the ossicular chain and tympanic membrane is involved, then surgical reconstruction of the middle ear conduction system is advised along the lines indicated in Chapter 2 for myringoplasty and ossiculoplasty.

Many otolaryngologists will fairly routinely request CT imaging before middle surgery to repair a perforated ear drum and/or a damaged ossicular chain, but that will not diagnose tympanosclerosis nor reliably confirm the ossicular degree of erosion. It cannot also assess the degree of fixation of the ossicles.

Regarding myringoplasty, the current surgical debate is which material is best to use for the reconstruction, with fascia and cartilage being the main competitors.

Regarding ossiculoplasty, the commercial trade promotes many different prostheses, but the general belief is that using patient material, such as residual ossicles or cortical bone, is superior. The only technical difficulty in reconstructing the ossicular chain is when there is no stapes superstructure. **Figure 12.11** illustrates metal prosthesis that can overcome this problem. It is useful to have these routinely in the theatre store cupboard along with stapedotomy pistons.

For those who report their results of ossiculoplasty, such as when comparing visualisation with

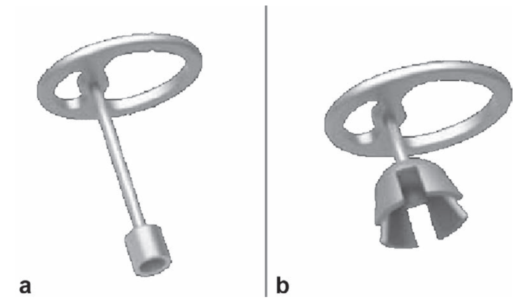

Figure 12.11 Total **(a)** and Partial **(b)** ossicular replacement prostheses.

a microscope or an otoscope, showed no difference in the audiometric results in 157 cases, but the overall mean lessening of the air–bone gap in this case series was only 4 dB (5).

CONCLUSIONS ON MIDDLE EAR SURGERY FOR MANAGING HEARING IMPAIRMENTS WITH A CONDUCTIVE COMPONENT

- All management options should be discussed with the patient, and any companion, of their likely benefit and risks. Patients should be given the option of a trial of conventional hearing aids and other devices, before opting for middle ear surgery. This carries risks and its benefit is dependent on the pathology determined at surgery and the surgeons' recorded experience of performing the procedure proposed.
- Patients have two ears and, in many of the patients seen for management of their hearing, will have a bilateral impairment. If there is a conductive component to their hearing impairment in the audiometrically poorer ear, then middle ear surgery should be considered, generally initially in this poorer hearing ear.
- This does not mean that a 'better', but impaired hearing ear should also not be non-surgically managed, perhaps with a hearing aid as discussed in Chapter 11.
- Technically successful middle ear surgery of a conductive or mixed impairment, due to chronic otitis media, OME and otosclerosis, can improve the FFA by lessening the magnitude of the air–bone gap in the poorer ear.
- This will give both clinical and practical benefit as would a hearing aid.

- The benefit is greatest in those with a conductive impairment with the bone-conduction thresholds of ≥ 30 dB as in them, there is the potential to eliminate the hearing impairment.
- Following such beneficial middle ear surgery, the hearing is improved throughout the day and does not require the use of 'small knobs' and a battery that needs changed/recharged.
- Patients with a mixed hearing impairment, whose preoperative bone-conduction thresholds are poorer than 30 dB HL, can also benefit from middle ear surgery but with different objectives.
- Even if their air–bone gap is surgically closed, they would still have the sensorineural component to their impairment and would still gain benefit from a hearing aid. However, this aid benefit would be greater, as well being easier to prescribe, than if they still had their severe or moderate, mixed impairment.
- Middle ear surgeons who operate on ears, and particularly those who perform otosclerosis surgery, should have been rigorously surgically trained and have the appropriate facilities and instruments.
- Such surgeons should be analysing their post-surgery outcomes, and particularly their mean improvement in their FFA outcomes. They can then predict the likely FFA following surgery and can relate that to the FFA in the other ear.
- The Glasgow Benefit Plot can be useful in doing this as it considers the FFA in both ears.
- Overall from the literature, in experienced hands, middle ear surgery for ears with otosclerosis gives greater improvements in the air-conduction thresholds than for ears with chronic otitis media.
- However, the patient's general health and medications for the required associated general or local anaesthetic, must be considered along with the risk of causing inner ear trauma and a sensorineural impairment.
- In patients with a bilateral hearing impairment with a conductive component in both ears, middle ear surgery on the second ear can be considered following beneficial surgery in the first ear, but the benefit of this can vary considerably as illustrated in the three patients (A, B &C) in this chapter.

BONE-CONDUCTION HEARING IMPLANTS

The principles of bone conduction are outlined in **Chapter 11** and are therefore not repeated here. A bone-conduction implant is a surgically implanted device which amplifies sound via bone conduction. Because they osseointegrate with the bone of the skull, they do not have the skin attenuation effect that limits the use of non-surgical bone conductors and can be used for patients with conductive and mixed hearing losses, with bone-conduction thresholds as low as 60 dB. They can also be used to reduce the head shadow effect in single-sided hearing loss, though it should be noted that they do not confer binaural or directional hearing.

Because they do not require any part of the device to be placed in the ear canal, bone-conduction hearing implants are indicated for patients with microtia or external auditory canal atresia or stenosis. They can also be useful for patients with acquired canal atresias, or for patients who are unable to wear a conventional hearing aid due to active chronic ear discharge.

Bone-conduction implants can be percutaneous (**Figure 12.12**) or transcutaneous (*see* **Figure 12.13**). They can also be classified as active or passive, depending on how sound is processed and transmitted to the implanted component. Percutaneous devices require a minor procedure to tap a titanium abutment into the skull cortex, ideally at the level of the external auditory canal. An external processor is clipped to the abutment, delivering sound directly to the inner ear via bone conduction. Transcutaneous devices involve an implanted component which connects via a magnet to an external processor and are associated with fewer skin complications.

Because bone-conduction implant surgery does not involve surgery to the middle ear or inner ear, these structures are not at risk. A bone-conduction hearing implant is therefore an option for patients

Figure 12.12 Passive percutaneous bone-conduction hearing implant. (Credit: Cochlear.)

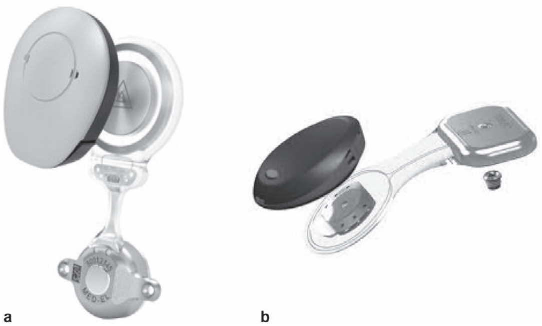

Figure 12.13 Types of active transcutaneous bone-conduction implant. **(a)** MED EL Bonebridge. **(b)** Cochlear Osia. (Credit: Cochlear and Med-El.)

who do not wish to accept the risks associated with middle ear and in particular stapes surgery.

MIDDLE EAR IMPLANTS

Middle ear implants are surgically implanted hearing aids. They can mechanically vibrate the ossicular chain when coupled to an ossicle, and in the absence of mobile ossicles, can be coupled to the round window or a third window of the cochlea. They are suitable for conductive, sensorineural and mixed hearing losses.

They are indicated in patients where the use of conventional hearing aids or bone-conduction devices is not feasible. Eligible patients might include those who:

- Find the use of conventional hearing aids difficult due to ear allergy, eczema, recurrent otitis or discharge.
- Are born with microtia or canal atresia.
- Are unable to wear a bone-conduction hearing aid due to skin problems or previous loss of an abutment.
- Have a stable middle ear (i.e. no infections) and stable bone-conduction thresholds but cannot wear a conventional hearing aid.

Several middle ear implants have been introduced; the most widely used is the MED EL Vibrant Soundbridge, which is semi-implantable and has both internal and external components, which attach through the scalp via magnets (*see* **Figure 12.14a**). The internal component has a floating mass transducer which can be coupled to the ossicular chain or the cochlea (*see* **Figure 12.14b**) using specifically

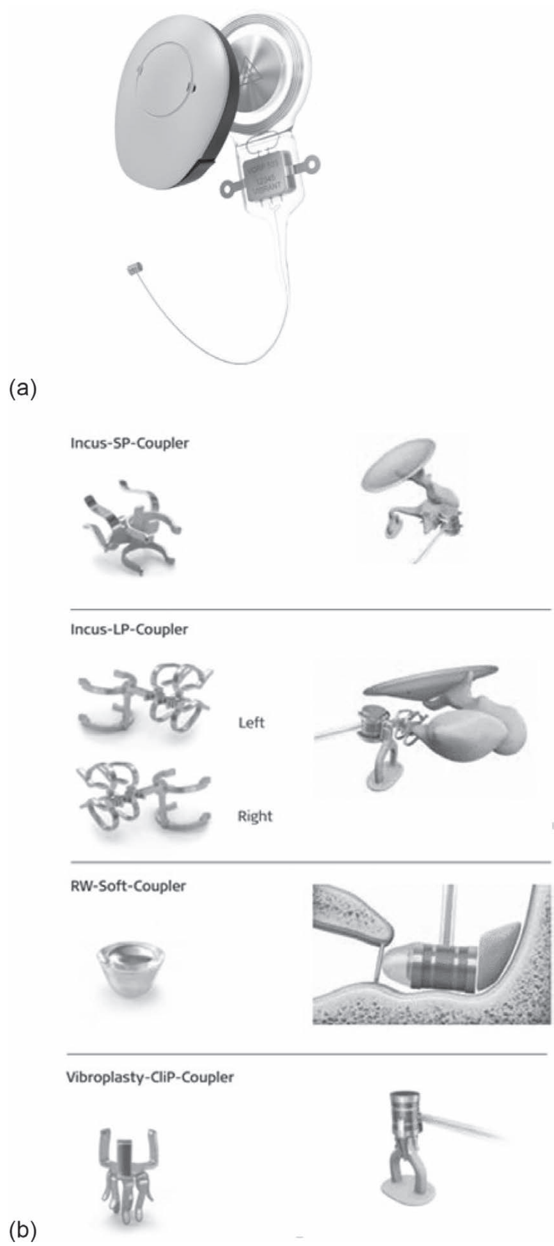

Figure 12.14 **(a)** Middle ear implant (Vibrant Soundbridge, MED EL). Processor and implant. **(b)** Couplers to attach floating mass transducer to the ossicular chain. (Credit: Med-El.)

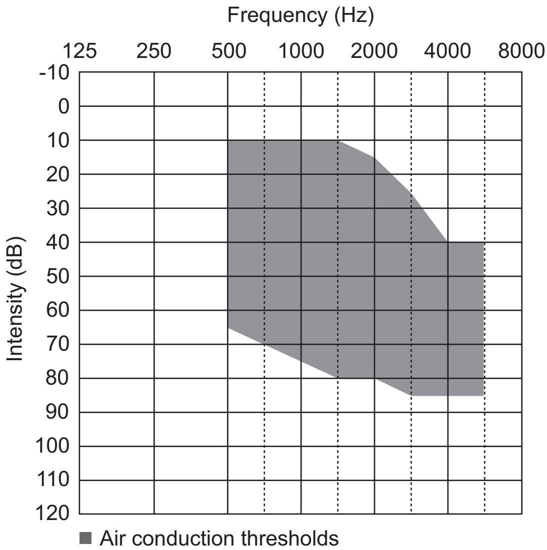

Figure 12.15 Candidacy guideline for the MED EL Vibrant Soundbridge Implant.

designed coupler mechanisms, to transmit sound vibrations to the inner ear.

Fully implantable devices have the benefit of being invisible, as well as allowing the user to hear at night, and whilst swimming and showering. However, they have several inherent design challenges:

1. *Microphone* – in a semi-implantable device, the microphone in the external component, which processes sound and delivers it to the internal component. Fully implantable devices require an internal microphone, these are less effective, and tend to pick up body sounds, which patients can find intrusive.
2. *Battery* – semi-implantable devices have a battery in the external component which can be replaced or recharged. Fully implantable devices have internal batteries that require charging via an external connector and require a surgical procedure for battery replacement.
3. *Hardware* – semi-implantable devices have minimal internal hardware, as the microphone, battery and sound processor are all within the external component. Fully implantable devices have an increased amount of internal hardware.

Surgery to implant a middle ear implant is fairly straightforward, but carries risks including bleeding, infection, dislodgement or failure of the implant, dizziness, tinnitus, taste change, deterioration of hearing or a dead ear and a small risk of injury to the facial nerve.

Middle ear implant surgery, like cochlear implant surgery (*see* **Chapter 13**) requires a multidisciplinary approach, with a thorough pre-operative audiometric assessment to ensure the patient's audiometric thresholds are stable and within recommended candidacy guidelines (*see* **Figure 12.15**). Patients with deteriorating audiometric thresholds should not proceed with middle ear implant surgery, as their hearing may drop below the level at which they will benefit from the middle implant, and head towards cochlear implant candidacy.

SPECIFIC CONCLUSIONS ON BONE CONDUCTION AND MIDDLE EAR IMPLANTS

- Bone-anchored hearing aids are acoustically superior to conventional bone-conduction aids. Minimum bone-conduction thresholds of 60 dB must be present for aiding to be beneficial.
- Bone-conduction aids are a suitable option for patients with bilateral congenital or acquired atresias of the external auditory canal.
- Bone conduction may also be suitable for patients unable to wear a conventional hearing aid due to discharge, allergy or recurrent infection.
- Middle ear implants mechanically vibrate the ossicular chain or the round window via a floating mass transducer. They are suitable for patients with sensorineural conductive or mixed hearing losses, who are unable to wear a conventional or bone-conduction hearing aid.

FURTHER READING

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13

Cochlear implants

Cochlear implants are electronic devices which can restore a sense of hearing in profoundly deaf recipients.

Hearing aids, middle ear implants and bone conduction devices all deliver amplified sound signals to the middle or inner ear, stimulating the ossicular chain and/or the cochlear hair cells to deliver signals to the brain, where they are interpreted as sound. Cochlear implants work differently, bypassing the ear almost entirely, and delivering electrical signals directly to the cochlear nerve (**Figure 13.1**).

This difference in the way cochlear implants send sound signals to the brain has implications for the way cochlear implant recipients process and perceive sound, and for the outcomes of cochlear implantation.

AUDITORY NEUROPLASTICITY

Neuroplasticity of the human brain is a key theme in understanding how and why cochlear implants work, and influences the timing of cochlear implantation, especially in children. Neuroplasticity is the brain's ability to learn from experience. For the human brain to interpret signals from the auditory nerve as meaningful sound, the brain must be either within its period of neuroplasticity, or the recipient must have auditory memory, i.e. previous experience of hearing.

The period of maximal auditory neuroplasticity is within the first years of life. There is a dramatic decrease in auditory neuroplasticity around the age of 3.5 years and a significant loss of auditory neuroplasticity after 7 years of age (1,2). The

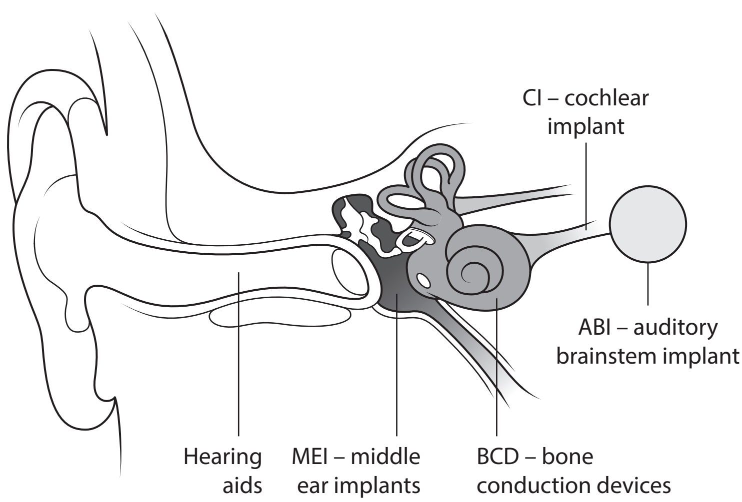

Figure 13.1 Different hearing devices stimulate a variety of parts of the auditory system.

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implications of this are that congenitally deaf children who receive cochlear implants within the first 3.5 years of life have the potential to develop sound and speech recognition which enables them to have a mainstream education, provided their development is otherwise normal. The optimal age for cochlear implantation is around 1 year. Cochlear implantation of congenitally deaf children after the age of 3.5 years is likely to lead to suboptimal hearing, speech and educational outcomes, and after the age of 7 years is likely to offer environmental sound recognition only, with expectations for speech recognition and speech development being unrealistic.

Older children and adults with profound deafness can be suitable cochlear implant candidates if they have auditory memory. This means their auditory cortex has had previous experience of sound and can therefore be stimulated to perceive sound again, through cochlear implantation. The development of speech is a useful indicator of previous auditory cortex stimulation. Children's language development can be measured using the Manchester spoken language development scale as part of pre-implant assessment and an outcome measure of post-implant rehabilitation (3).

In adults, the Speech Intelligibility Rating can be used to gauge an adult's speech development, as a proxy for their previous experience of hearing speech, and their likelihood of developing meaningful sound and speech perception with a cochlear implant. The rating varies from intelligible speech, through various categories of deaf intonation, to unintelligible speech (4). The concept of deaf intonation can be understood by considering the speech banana (**Chapter 6**, *see* **Figure 6.5**). People whose hearing falls below the speech banana and have not been rehabilitated to within the speech banana, cannot produce sounds they have never heard. This is usually higher frequency sounds, and leads to the development of deaf speech intonation, where higher frequency sounds (e.g. f/s/th) are not produced.

Profoundly deaf patients therefore do not neatly fall into categories of congenital deafness (i.e. no previous experience of sound) and acquired deafness (normal speech development with subsequent loss of hearing). They exist on a spectrum, with many patients having had some experience of sound, and in the case of deaf adults, often a long period of sound deprivation. This has implications for their likelihood of benefitting from cochlear implantation. The multidisciplinary team (MDT) is therefore essential

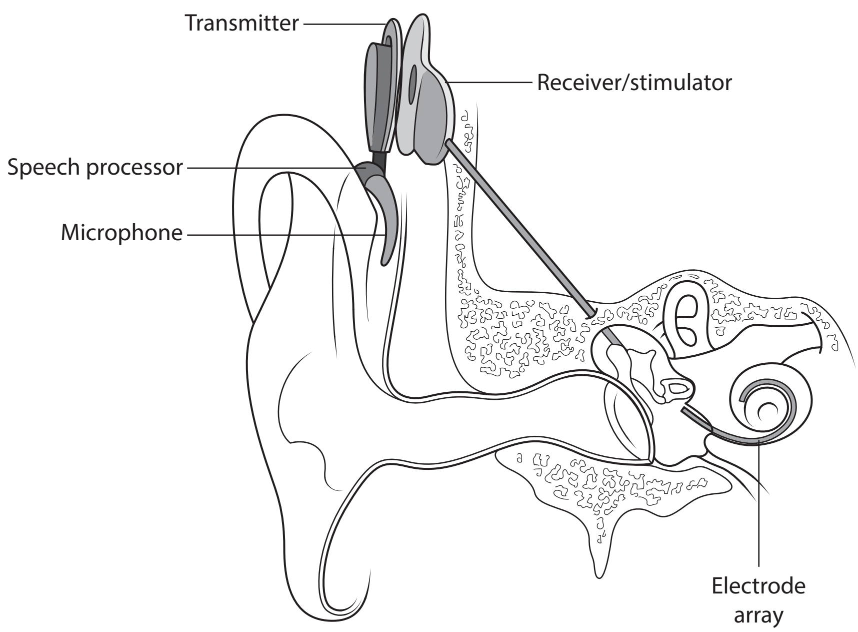


Figure 13.2 The internal and external components of a cochlear implant.

in pre-implantation assessment, post-implantation rehabilitation, and in the communication of expectations regarding the likely outcomes of cochlear implantation, on an individual patient basis.

An MDT within a cochlear implant programme might include otolaryngologists (surgeons), audiologists, rehabilitationists, speech and language therapists, teachers of the deaf, clinical scientists and psychologists.

WHAT IS A COCHLEAR IMPLANT AND HOW DOES IT WORK?

Cochlear implants consist of internal and external components (**Figure 13.2**). The external component comprises a microphone and speech processor which collect and synthesise sound; a coil transmitter which delivers sound signals to the internal component; and a magnet for connection to the internal component through the scalp. The internal component is surgically implanted and comprises a receiver/stimulator package which sits deep to the scalp and converts sound signal to electrical impulses, and an electrode array which is inserted into the cochlea, delivering electrical impulses to the cochlear nerve. The electrode is usually placed in the scala tympani via the round window, but insertions can vary according to surgical challenges and electrode types.

The electrode array of a cochlear implant has between 12 and 22 channels, depending on the manufacturer of the implant. Cochlear implant recipients have similar hearing outcomes, despite this variation in electrode number. Notably, the human cochlea has around 3500 'channels'. The ability of the human brain to learn to perceive detailed sound with a fraction of this number of channels reflects auditory neuroplasticity. It also reminds us that cochlear implants do not restore natural hearing. Rather, they restore a sense of sound recognition, which enables the recipient to make sense of their auditory environment.

The tonotopic arrangement of the cochlea and cochlear nerve (*see* **Chapter 1**) means that cochlear implants can be programmed such that the electrodes are mapped to deliver frequencies of sound which match those represented by the corresponding auditory nerve and auditory cortex (**Figure 13.3**).

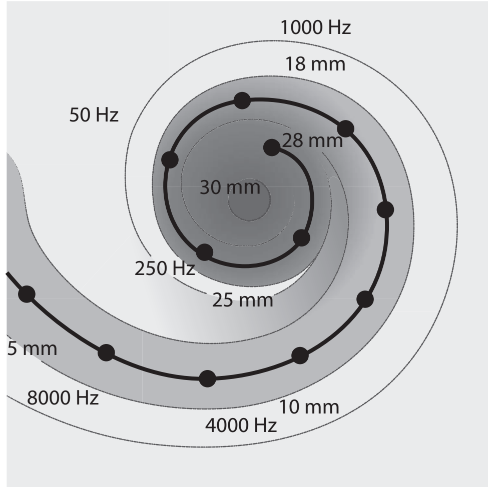


Figure 13.3 The tonotopic arrangement of the cochlea.

COCHLEAR IMPLANT CANDIDACY

Guidelines for cochlear implant candidacy vary throughout the world (5). However, the underpinning principles are consistent. In the UK, cochlear implant candidacy is governed by the National Institute for Health and Care Excellence (6), which recommends:

- Unilateral cochlear implantation for people with severe-to-profound deafness who do not receive adequate benefit from acoustic hearing aids.
- Simultaneous bilateral cochlear implantation is recommended for children, and for adults who are blind or who have other disabilities that increase their reliance on auditory stimuli as a primary sensory mechanism for spatial awareness.
- Severe-to-profound deafness is defined as hearing worse than 80 dB at two or more frequencies (500 Hz, 1000 Hz, 2000 Hz, 3000 Hz and 4000 Hz) bilaterally, without acoustic hearing aids.
- Adequate benefit from acoustic hearing aids is defined as a phoneme score of 50% or less on the Arthur Boothroyd word list (for adults).
- Adequate benefit from acoustic hearing aids is defined as speech, language and listening skills appropriate to age, developmental stage and cognitive ability (for children).

- Cochlear implantation should be considered only after an assessment by an MDT, and after a valid trial of an acoustic hearing aid for at least 3 months, unless contraindicated or inappropriate.

This guideline is broadly represented by **Figure 13.4**. The requirement for only two frequencies to be worse than 80 dB means that patients with good low-frequency hearing and poor high-frequency hearing can fall within cochlear implant candidacy, provided their speech recognition scores are also within the candidacy guidelines (**Figure 13.5**).

Equity of access is recommended, with MDTs advised to remain mindful of disabilities such as cognitive impairments, linguistic or other communication difficulties, and administration of tests in a language in which the patient is fluent.

Although not recommended by the UK's NICE guideline, some health systems offer cochlear implantation bilaterally in deaf adults, and unilateral cochlear implantation for unilateral deafness. As described in previous chapters, binaural hearing offers advantages in terms of directional hearing and hearing in background noise. However, there is not yet sufficient evidence to support the efficacy or cost effectiveness of bilateral cochlear implantation in adults. Cochlear implantation for unilateral hearing loss has been shown to deliver some benefits but the mismatch between neural

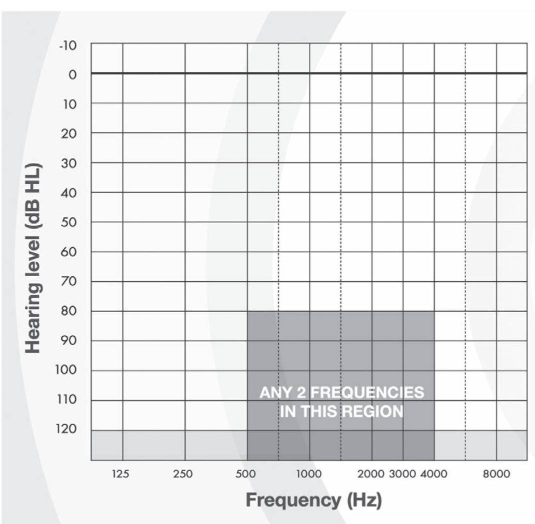


Figure 13.4 Cochlear implant candidacy guidelines. (With permission from BCIG.)

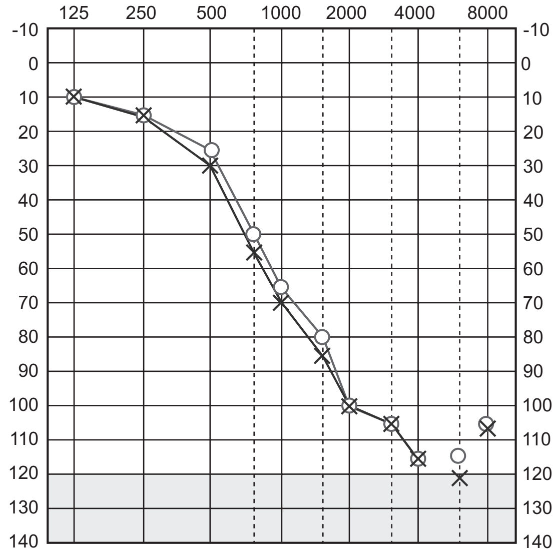


Figure 13.5 An example audiogram of a cochlear implant candidate with good low-frequency hearing thresholds.

signals from natural hearing in one ear, and a cochlear implant in the contralateral ear, means that the true benefits of binaural hearing are not achieved (7).

PATIENT PATHWAY AND AFTERCARE

Patients are recommended for cochlear implantation following audiometric assessment, rehabilitation assessment, cross-sectional imaging and MDT discussion. The British Cochlear Implant Group Quality Standard Guidelines outline a recommended patient journey (8). This outlines the recommended process for a high resource setting and is not necessarily applicable to cochlear implant programmes in low- and middle-income countries, where resource limitations may prohibit its full implementation.

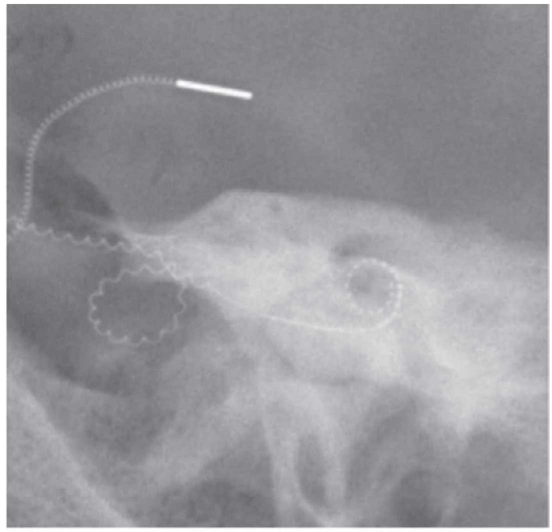
The goal of cross-sectional imaging is to confirm the patient's eligibility for cochlear implantation. An MRI scan would normally be carried out, to ensure the patient has intact cochlear nerves, implantable cochleas and no retrocochlear lesions. CT imaging can also be useful, especially in patients with a previous otological history.

Patients who are considered suitable for cochlear implant surgery are offered an appointment with a cochlear implant surgeon to discuss surgical risks and to consent for surgery. Cochlear implant surgery can be carried out under general anaesthesia; or local anaesthesia, with or without conscious sedation. The avoidance of general anaesthesia is traditionally reserved for frail patients for whom general anaesthesia is a risk, but some surgeons offer local anaesthetic surgery to all eligible adult patients, and a discussion between surgeon and patient regarding anaesthetic choice is essential. Local anaesthetic cochlear implant surgery is not appropriate for children.

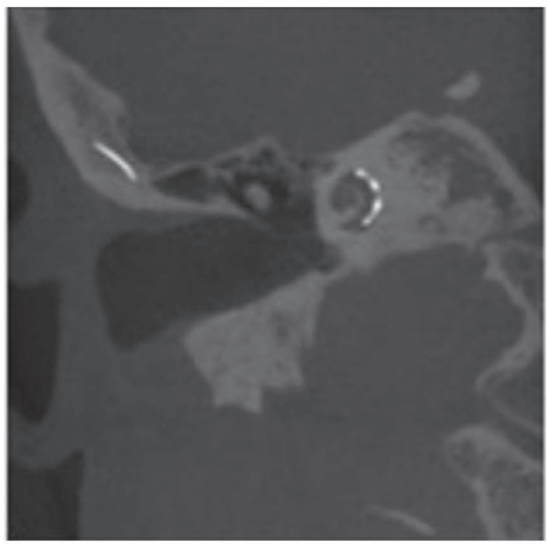
Following cochlear implant surgery, the position of the cochlear implant electrode is confirmed by radiological imaging. i.e. X-ray or cone beam CT (**Figure 13.6**) or by impedance measurements taken intraoperatively. Cone beam CT is increasing in popularity due to its low radiation dose, detailed imaging and lack of metal artefact.

It is common practice to allow between 2 and 4 weeks after surgery for the patient's wound to heal, and any scalp swelling to settle, before device activation. At device activation, an audiologist or clinical scientist performs a PTA to check residual hearing thresholds, and programmes the cochlear implant. In adults, the device is programmed according to the patient's hearing thresholds and uncomfortable loudness levels. For babies and young children, behavioural responses are often used. Hearing with a cochlear implant is plotted using soundfield thresholds (**Figure 13.7**). Following initial programming, the cochlear implant is activated, and the patient can hear spoken voices, often for the first time in many years for long-deafened adult patients. At device activation, patients frequently comment that voices sound 'different' or 'electronic'. This is inevitable considering the relatively small number of channels on a cochlear implant electrode, however sound quality improves as the brain becomes used to hearing sound again. Cochlear implant recipients who have had hearing recently tend to adapt to their cochlear implant more quicky and effectively than long-deafened patients, as their auditory cortex has been more recently active.

Following initial device activation, onward contact with audiology and rehabilitation is essential. Patients benefit from reassessment of their cochlear implant mapping to make any necessary adjustments. Rehabilitation therapy can guide them regarding useful techniques to practise using



(a)



(b)

Figure 13.6 Plain X-ray **(a)** and cone beam CT **(b)** of cochlear implant electrodes.

their cochlear implant. This is especially the case for adult cochlear implant recipients who live alone and are not exposed to conversation in their daily life, and for children from Deaf families, where spoken language may not be used at home. Auditory verbal therapy is essential for children who require additional support in speech and language development. Signposting to support groups and social groups can also be useful.

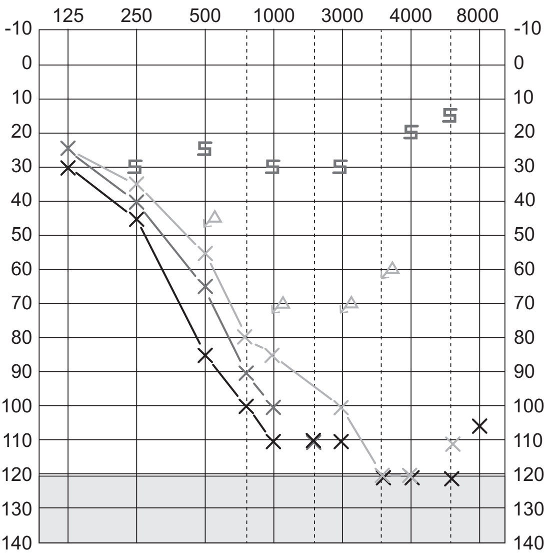


Figure 13.7 Soundfield audiogram demonstrating thresholds heard through the cochlear implant (marked S) in a patient with preserved low-frequency hearing.

Having a point of contact with a cochlear implant centre is important, as devices require repairs, upgrades, battery changes and troubleshooting. There are also online support options offered by the cochlear implant manufacturers.

STRUCTURE PRESERVATION SURGERY

It was previously believed that the insertion of an electrode into the cochlea would destroy any residual hearing. In fact, soft surgical techniques allow residual hearing to be preserved at the time of surgery. These techniques include avoidance of trauma to the ossicular chain, avoidance of intracochlear suction or turbulence, and a slow, steady electrode insertion. The use of dexamethasone over the round window is widely used.

Cochlear implant recipients whose low-frequency hearing is preserved at the time of surgery can use a hybrid external processor which delivers amplification to their preserved lower frequencies, and cochlear implant stimulation for hearing higher frequencies. This is known as electro-acoustic stimulation. Throughout the literature, a trend for preserved hearing to diminish over the subsequent months or years has been observed and the use of electro-acoustic stimulation is often short-lived.

Soft surgical techniques continue to be recommended, however, for several reasons. The initial preservation of natural hearing is useful to patients, particularly between surgery and device activation. Soft surgical techniques are also more likely to preserve vestibular function, avoiding postoperative dizziness, nausea and balance loss. Structure preservation surgery is also recommended in terms of avoiding the loss of any structure or function which may be useful in future when new technology becomes available.

SUDDEN PROFOUND HEARING LOSS

Sudden profound hearing losses require special mention due to their urgency, as the events which lead to sudden catastrophic loss of hearing can predispose patients to cochlear obliteration which, when advanced, renders implantation of the cochlea impossible.

The most frequent causes of sudden catastrophic loss of hearing are infection (e.g. bacterial meningitis) which is commoner in children, and autoimmune causes which are commoner in adults. In both cases, urgent cross-sectional imaging is indicated. Loss of fluid signal in the cochlea and labyrinth on MRI is an early indicator of cochlear obliteration. Abnormalities on CT imaging appear later and are suggestive of more advanced, bony obliteration.

In the early stages of obliteration, the basal turn of the cochlea is often filled with soft tissue which can be bypassed using a stiff cochlear implant electrode. In some cases, a scala vestibuli insertion is possible, and can offer good hearing outcomes. However, it is rarely feasible to implant a cochlea which has progressed to bony obliteration. Any sudden, catastrophic sensorineural hearing loss with signs of cochlear obliteration on MRI, warrants urgent referral to a cochlear implant programme.

AUDITORY BRAINSTEM IMPLANTS

Auditory brainstem implants are implantable devices which aim to restore a sense of sound recognition in patients who are profoundly deaf, in whom cochlear implantation is not feasible. This includes patients born without cochleas or without cochlear nerves, and patients whose cochlear nerve has been surgically removed, for example, during cerebellopontine angle tumour excision. They bypass the peripheral auditory system and cochlear nerves, aiming to deliver sound perception directly to the brain.

Auditory brainstem implants are placed on the auditory nucleus of the brainstem, which lies in the Foramen of Luschka. Unlike the cochlea and cochlear nerve, the cochlear nucleus is not tonotopically arranged. This means hearing outcomes are inferior to those achieved with cochlear implantation. Auditory brainstem implants can offer environmental sound recognition, but they rarely impart hearing of sufficient quality to develop useful speech recognition.

CONCLUSIONS

- Cochlear implants are electronic devices which can restore a sense of hearing in profoundly deaf recipients.
- They bypass the middle and inner ear structures, stimulating the auditory nerve directly.
- An understanding of auditory neuroplasticity is essential in understanding how and why cochlear implants work.
- The development of comprehensible speech is an indicator of auditory memory in profoundly deaf patients. Patients with incomprehensible speech are unlikely to develop speech recognition with a cochlear implant.
- Multidisciplinary care is important both before and after cochlear implant surgery, particularly auditory verbal rehabilitation in children.
- Auditory brainstem implants are implanted devices which aim to restore a sense of hearing in profoundly deaf patients who cannot have a cochlear implant.

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14

Outcomes, audit and consent

OVERVIEW

Whilst not a definitive guide, this chapter aims to give a broad overview of the relevance of outcomes, audit and consent, in the context of audiology and ear surgery.

Ear surgery is nearly always carried out for benign conditions. The purposes of otological surgery fall into several overarching categories: surgery to halt or prevent the progression of disease (such as cholesteatoma); surgery to improve quality of life (such as repairing an eardrum to stop persistent otorrhoea) or surgery to improve hearing, as described in **Chapter 12**.

This is in contrast to, for example, surgery for malignant disease, where the goal of treatment is to increase the patient's 5-year survival. When offering interventions for a patient's quality of life, rather than for their likelihood of survival, a different approach is required regarding consideration of risks and expected outcomes.

This is not to say that hearing loss or otorrhoea are any less important than malignant disease. In terms of years affected, and impact on education, social development and communication in work, family and social environments, their impact is considerable. The length of time patients live with their ear and hearing symptoms consolidates the importance of making treatment decisions carefully, responsibly and in partnership with the patient.

But if non-surgical management, such as a hearing aid improves quality of life, under what circumstances would we consider performing an operation which carries greater risks of complications **(Box 14.1)** including poorer or complete loss of hearing?

For this reason, personal audit, consistent outcome reporting and robust consent are essential for any surgeon who carries out operations for hearing. A strong understanding of, and engagement with, these concepts will enable the clinician to monitor their own practice and to communicate risk and benefit to their patient accurately as part of an informed consent process.

BOX 14.1: Potential complications of ear surgery

- Risks of anaesthesia and intubation
- Bleeding
- Infection
- Allergy to medication or packing
- Failure to improve hearing
- Increased conductive hearing impairment
- Sensorineural hearing loss
- Complete catastrophic loss of hearing (dead ear)
- Acute or chronic vertigo
- Chronic imbalance
- Tinnitus
- Taste change (due to chorda tympani injury)
- Facial palsy
- CSF leak
- Injury to venous sinus
- Recurrence of ear disease

OUTCOME REPORTING

Standardisation of outcome reporting is increasingly encouraged by institutions such as COMET, which supports the development and application of Core Outcome Sets (COS). These represent the

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minimum data set that should be measured and reported in all clinical trials of a specific condition.

Standardised outcome reporting is also essential for data synthesis. As all authors of systematic reviews and meta-analyses will recognise, heterogeneity of the method of data reporting often renders it impossible to draw a meaningful conclusion. COS are not limited to clinical trial outcomes reporting: they are also suitable for use in routine care, clinical audit and research other than randomised trials. The use of a COS to report outcomes does not limit researchers to reporting only the outcomes recommended by the COS.

Stakeholder involvement is a key theme in outcomes research. Patient and public involvement and engagement is encouraged and falls into two categories: the development of Patient Reported Outcome Measures (PROMs), and the involvement of patients as stakeholders in designing and conducting research. The involvement of patients in the development of COS is a gold standard for stakeholder engagement and recognised as best practice. Patient engagement is also relevant when considering our personal surgical outcomes. Patient-centred outcomes should be gathered and reported in addition to outcomes that are important to clinicians. For example, reporting audiometric outcomes from stapes surgery but neglecting to report patient reports of taste disturbance, pain, vertigo, or other outcomes, represents suboptimal outcome reporting.

In the absence of a validated PROM, patient-reported outcomes can be reported qualitatively, or quantitatively using questionnaires based on recognised grading scores.

The American Academy of Otolaryngology, Head and Neck Surgery (AAOHNS) has compiled a database of research outcome tools (*see* **Further Reading**).

The 2012 AAOHNS paper 'A New Standardized Format for Reporting Hearing Outcome in Clinical Trials' recommends the use of a 4-frequency PTA using 0.5, 1, 2 and 3 kHz as a minimal data set to facilitate interstudy comparability (1). This is in keeping with a similar previous guideline published 33 years earlier (2). It is interesting to note that 3 kHz thresholds are not universally assessed in many surgical series, with many researchers instead reporting a calculated average of 2 kHz and 4 kHz thresholds. This is not always an accurate representation of hearing thresholds at 3 kHz and most surgeons use 4 kHz instead.

AUDIT

Personal and Departmental Clinical Audit

Audit of the outcomes of clinical intervention, especially surgical, should be an essential part of everyday clinical practice. As such, audit should be prospectively planned and starts when an intervention is being proposed, the outcome objectives having been agreed when obtaining informed consent (*see* 'Consent').

When consenting a patient for surgical intervention, it is best practice for clinicians to have assessed their own outcomes of surgery, and risks of complications having audited their own or departmental case series.

This is particularly important, because most ear operations worldwide are performed by general otolaryngologists rather than sub-specialist otologists who have had a professionally recognised otological training programme. Even within this later group, there can be those who are particularly experienced in paediatric, otosclerosis or cochlear implant surgery. It is these otologists who most frequently report their outcomes and it is naive to expect less experienced surgeons to have the same outcomes.

The easiest way for a patient to assess a surgeon's experience is to ask how often they currently perform the operation being suggested. Asking how many they have done will most often be answered by a very enthusiastic, incorrect guess.

Some institutions host databases which can be used for logging personal outcome data and/or contributing to larger data sets. These should be encouraged, and the outcomes regularly reported to institute meetings.

Follow-Up Process of Managing Hearing Impairment and Any Associated Tinnitus

Clinical follow up will be required of all medical and surgical interventions in the management of hearing issues with the purpose of monitoring hearing outcomes, any complications of surgery, and any associated tinnitus. Such a follow-up process may already be in place for surgical interventions, for routine postoperative care, pack removal, etc. It should also be part of the management process of providing an individual with a hearing aid, to ensure that they can put it in their ear, use the various controls and change the batteries.

There is considerable benefit to be had in routinely reviewing all patients at 6 months after surgery. This allows the patient and surgeon to discuss the outcome of surgery in relation to the patient's expectations, and the routine collection of personal and departmental audit data for comparison with published studies.

Objective and Subjective Outcomes

Objective hearing outcomes are mostly used when reporting outcomes of ear surgery where the objective was to improve the hearing, and include:

- Magnitude reduction in the air–bone gap over 0.5, 1, 2 and 3 kHz.
- Effect on symmetry of hearing; Glasgow Benefit plot (3).
- Change in sensorineural hearing thresholds, especially at the higher frequencies of 4, 6 and 8 kHz.
- Change in ear or tympanic membrane structure. This can be record pre- and post-surgery using endoscopic photography with or without AI analysis.

The improvement in hearing with amplification can also be measured in the laboratory with audiometric tasks such as speech in noise, with or without visual input. The weakness with such laboratory hearing assessments is that the benefit is not necessarily transferable to non-laboratory situations.

Regarding subjective hearing outcomes, it is possible to use generic outcome measures, such as the SF 36 and the HUI. These are designed to allow economic cost-benefit calculations based on metrics such as cost per quality-adjusted life-year and to allow comparison to be made between outcomes in different conditions. However, their major drawback is that the generic nature of their questions means that many of the questions, such as mobility, are not relevant to patients with hearing problems. They may, therefore, fail to capture many of the issues that ear disease cause. Ear-specific questionnaires will capture much more detail but cannot be used for economic analyses and comparisons with non-ear conditions.

Subjective hearing and tinnitus problems can be assessed by the completion of the same questionnaire before and after intervention. Some of these questionnaires might have been completed at an initial interview to help the clinician guide the patient's management. This is particularly applicable with tinnitus where symptoms are all subjective.

A more common method of assessing an outcome is a single questionnaire, completed after intervention, which asks about subjective changes in symptoms/difficulties.

GLASGOW BENEFIT INVENTORY

This 15-question outcome measure has been validated through a national Otorhinolaryngology outpatient study of 4543 adults seen and subsequently managed for various diagnoses (4). It reports a total score and five factors (*Support, General health, Quality of life, Self-confidence, Social involvement*) that contribute to that score (**Figure 14.1**).

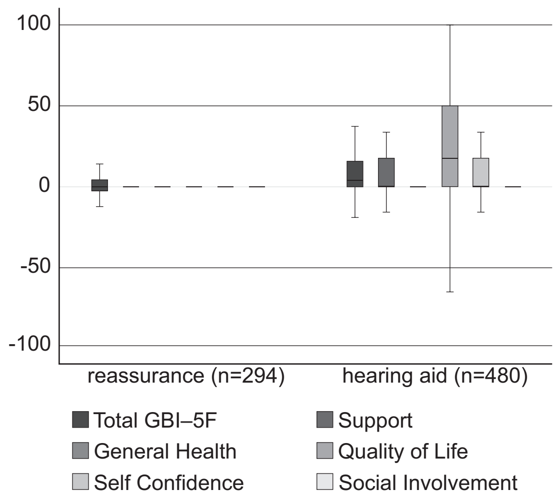

Figure 14.1 This Glasgow Benefit Inventory (GBI) figure is taken from Kubba et al 2023 (4) of a national study of 774 Otorhinolaryngology outpatients who had a sensorineural hearing loss. Of them 480 were provided with hearing aids and 294 declined aids but were given reassurance and advice. Hearing aids gave greater benefit in all five factors *(General health, Support, Quality of Life, Self-confidence and Social involvement)* but particularly in Self-confidence.

Glasgow Children's Benefit Inventory

Analogous to the GBI in adults, the GCBI was developed for use in children to measure benefit after intervention. It has 24 questions and can be scored as a total score with 4 factor scores (Psycho-social, Physical, Behaviour, Vitality). It has been used after a wide range of interventions in many surgical disciplines.

Hearing-Specific Outcomes for Non-Surgical Interventions Including Hearing Aids

These have been well summarised and appraised in the British Society of Audiology Patient reported outcomes in Audiology 2023 (PROETA toolkit 2003) (5). In this document, authors have categorised the multiple outcomes that apply to non-surgical intervention as follows:

1. *Laboratory assessment*. This is used mainly as a research tool, rather than a clinical outcome, where variations of digitally altered hearing aid systems worn by subjects are measured on tests such as speech in noise. Subsequent benefits from such aids in clinical practice are seldom translated to clinical usage.
2. *Patient report*. There are many questionnaires of patient subjective reports of their experiences of using their hearing aid prescription. These cover a variety of listening situations and methods of answering the question. These questionnaires are of minimal value for audit purposes.
3. *International Outcome Inventory for hearing aids (IOI-HA)*. This Inventory is available in 30 languages, the authors being multiple, internationally recognised participants attending a workshop to create it (6). There are seven domains, in order: Daily use in hours, benefit, residual activity limitations, satisfaction, residual participation restrictions, impact on others and quality of life. The first domain is perhaps the most relevant of these domains and is automatically recorded and reported by most modern digital hearing aids.

Tinnitus Questionnaires

Several tinnitus questionnaires exist, including mini-TQ (7), Tinnitus Handicap Inventory (8,9), and the Tinnitus Functional index (10).

The Tinnitus Functional Index is considered the most useful audit tool; it covers the impact but also more practical aspects such as the strength of the tinnitus, percentage time aware of the tinnitus, control of the tinnitus and coping with the tinnitus. It also has a 'past week' timescale for response. A tinnitus grading system was published in 1999 (11).

All tinnitus measurement tools are most informative when used at intervals to measure change and are less useful as a snapshot view of symptom severity due to differential patient reporting.

CONSENT

Consent for surgical interventions should be carried out by the clinician proposing the intervention, and not delegated to others. Consent is a process, and not a single timepoint at which a consent form is signed (*see* **Further Reading**).

Guidelines for best practice consent include the option of a 'supporter' being present and involved in giving informed consent. This can be particularly relevant when the patient requires personal support. It should be noted, however, that for consent to be valid, it must be informed, voluntary, and the patient must have capacity to consent. A supporter cannot consent on their patient's behalf, unless they are a parent, guardian, or have legal Power of Attorney. Details of the content process can be found in **Table 14.2**.

A full record should be made of the consent discussion, including the aims, risks and likely outcomes. A landmark legal case in the UK (*Montgomery vs Lanarkshire 2015*) established the importance of patient autonomy in the consent process, with the patient as partner in decision-making (12). Montgomery also introduced the concept that consenting surgeons must counsel patients regarding all risks which may be material to the patient, no matter how rare. They must also counsel patients regarding the likely outcomes, and risks associated with not going ahead with surgery. This record should be shared with the patient, who should then be offered a period to consider their options, before signing consent for surgery.

Although the *Montgomery case* refers to UK law, its themes are relevant to consent across the world, especially regarding surgery for hearing, where the risks associated with not going ahead with a surgical intervention are unlikely to be life threatening.

Table 14.2 Informed Consent as suggested by the Royal College of Surgeons of England

- Informed decision making takes time, patience and clarity
- It is not an anaesthetic room process
- Establish if there is a partner or supporter can be involved
- Establish patient has mental capacity to give consent
- Consent is obtained by the surgeon who is providing the treatment
- Surgeon suitably trained to provide treatment in question
- Anaesthetist involved if operation is under general anaesthetic
- Initial discussion /information given ideally at least 2 weeks before proposed date of surgery, often at a 'pre-operative clinic'
- Information in given in diagnosis and prognosis
- Personal/department audit of outcomes and complications given on purpose, benefit and likelihood of success. Inherent risks, side effects and complications. Alternative treatments available. Details of follow -up process
- Pamphlets and advice available electronically are given.
- Sufficient time given to make an informed decision. Suggested to be at least 2 weeks
- Clinicians must record in writing, details of the consent discussion
- An opportunity to further discuss the proposals should be available

Adapted from www.rcseng.ac.uk/standardsandguidance.

CONCLUSIONS

- Standardised outcome reporting facilitates data synthesis, as well as comparison of personal outcomes with published data.
- Outcome reporting should include patient reported outcomes, and outcomes research should include patient and public involvement and engagement.
- Personal audit is essential for clinician development, and clinical consent.
- Consent for surgical intervention must be informed, voluntary, and the patient must have capacity to consent.
- Consenting surgeons should respect patient autonomy, with the patient as partner in the decision-making process.
- Consenting surgeons must inform patients of all risks which may be material to them, no matter how rare, and of the risks of not going ahead with surgery.

FURTHER READING

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Appendix 1: UK population distribution of FFA by age and sex (1,2)

Tables A1.1 to **A1.6** show data for the four frequency average (FFA) thresholds at various ages for males and females, for left and right ears and for the mean of the two ears. The FFA is calculated as the mean threshold over the main speech frequencies, 0.5, 1 ,2 & 4 kHz. In the following FFA tables, there are two population distribution data sets headed 'Screened' and 'Typical'. The 'Typical' distribution data are for a randomly selected population sample. The 'Selected' distribution data were created by removing individuals from the 'Typical' data set who had been exposed to potentially damaging noise or had a conductive defect of 10 dB or greater. As a result, the 'Screened' distribution data can used to evaluate the effect of aging on the FFA sensorineural hearing thresholds having excluded individuals that have been exposed to potentially damaging noise. Data are presented for the various centiles of the population distribution, from the 5th to the 95th.

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Table A1.1 Male left ear. Distribution of hearing levels for screened and typical population as a function of age using a smoothing regression function on the 7−year running average

Ear = left								Frequency = 0.5, 1, 2 and 4 kHz								Gender = Male
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
22	7	−2	−1	1	5	10	14	17	7	−2	0	2	5	10	15	20
23	7	−2	−1	1	5	10	15	18	7	−1	0	2	5	10	16	21
24	7	−1	−1	2	5	10	15	18	8	−1	0	2	6	11	17	22
25	7	−1	−1	2	5	10	15	18	8	−1	0	3	6	11	18	24
26	7	−1	0	2	5	10	15	19	9	−1	0	3	6	12	18	25
27	8	−1	0	2	6	10	16	19	9	0	0	3	7	12	19	26
28	8	−1	0	2	6	10	16	20	9	0	0	3	7	13	20	27
29	8	0	0	2	6	11	16	20	10	0	0	3	7	13	21	28
30	8	0	0	2	6	11	16	20	10	0	1	4	8	14	22	30
31	8	0	0	3	6	11	17	21	11	0	1	4	8	14	22	31
32	8	0	0	3	6	11	17	21	11	0	1	4	8	15	23	32
33	9	0	0	3	7	11	17	21	12	0	1	4	8	15	24	33
34	9	0	0	3	7	11	17	22	12	0	2	4	9	16	25	35
35	9	0	1	3	7	11	18	22	13	0	2	5	9	16	26	36
36	9	0	1	3	7	12	18	23	13	1	2	5	10	17	27	37
37	9	0	1	3	7	12	18	23	13	1	2	5	10	18	27	38
38	10	0	1	4	8	12	19	23	14	1	3	5	10	18	28	39
39	10	1	1	4	8	12	19	24	14	1	3	6	11	19	29	40
40	10	1	2	4	8	13	19	25	15	2	3	6	11	19	30	42
41	11	1	2	4	8	13	20	25	15	2	3	6	12	20	31	43
42	11	1	2	4	8	13	21	26	16	2	4	7	12	21	32	44
43	11	1	2	5	9	14	21	27	16	2	4	7	12	21	33	45
44	12	1	2	5	9	14	22	28	17	3	4	7	13	22	34	46
45	12	2	3	5	9	15	23	28	17	3	4	8	13	22	35	47
46	12	2	3	5	10	15	23	29	18	3	5	8	14	23	35	48
47	13	2	3	6	10	16	24	30	18	3	5	8	14	24	36	49
48	13	2	3	6	10	17	25	30	19	3	5	9	15	24	37	50
49	13	2	3	6	10	17	25	31	20	4	5	9	15	25	38	51
50	14	2	4	7	11	18	26	32	20	4	6	9	16	26	39	52
51	14	3	4	7	11	18	27	32	21	4	6	10	16	27	40	53
52	14	3	4	7	11	19	27	33	21	5	6	10	17	28	41	54
53	15	3	5	8	12	19	28	34	22	5	7	11	18	28	42	55
54	15	4	5	8	12	20	29	34	23	5	7	11	18	29	43	56
55	16	4	5	8	13	21	30	35	23	6	8	12	19	30	44	57
56	17	4	6	9	14	21	30	36	24	6	8	12	20	31	45	58
57	17	4	6	9	14	22	31	37	25	6	8	13	20	32	46	60
58	18	5	7	10	15	23	32	38	25	6	9	13	21	32	47	61
59	19	5	7	11	16	23	33	39	26	7	9	14	22	33	48	62
60	19	6	8	11	16	24	34	40	27	7	10	14	22	34	48	62
61	20	6	8	12	17	25	35	41	27	7	10	15	23	35	49	63
62	21	7	9	12	18	26	36	43	28	8	11	16	24	36	50	64
63	21	7	9	13	18	27	37	44	29	8	11	16	25	37	51	65
64	22	8	10	14	19	27	37	44	29	9	11	17	25	38	52	65
65	23	8	10	15	20	28	38	45	30	9	12	18	26	38	52	66

(Continued)

(Continued)

Ear = left				Frequency = 0.5, 1, 2 and 4 kHz											Gender = Male	
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
66	24	9	11	15	21	29	39	46	31	9	12	18	27	39	53	67
67	24	9	11	16	21	30	40	47	31	10	13	19	28	40	54	67
68	25	10	12	17	22	30	41	48	32	10	13	19	28	41	55	68
69	26	11	13	17	23	31	41	49	33	11	14	20	29	42	56	69
70	26	11	13	18	24	32	42	50	33	11	14	21	30	43	56	69
71	27	12	14	19	25	33	43	51	34	12	15	21	30	44	57	70
72	28	12	14	19	25	33	44	52	35	12	15	22	31	45	58	71
73	28	13	15	20	26	34	45	53	36	12	16	23	32	45	59	71
74	29	13	15	21	27	35	45	54	36	13	16	23	33	46	59	72
75	30	14	16	21	28	35	46	54	37	13	17	24	33	47	60	73
76	30	14	17	22	28	36	47	55	38	14	17	24	34	48	61	74

Table A1.2 Male right ear. Distribution of hearing levels for screened and typical population as a function of age using a smoothing regression function on the 7–year running average

Ear = right				Frequency = 0.5, 1, 2 and 4 kHz											Gender = Male	
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.5	P.10	P.25	P.50	P.75	P.90	P.95
22	5	−3	−2	0	4	6	13	18	7	−2	0	1	5	9	14	23
23	5	−2	−2	0	4	6	13	18	7	−2	0	1	5	9	15	24
24	5	−2	−2	1	4	6	13	18	8	−2	0	1	5	10	15	25
25	6	−2	−1	1	5	7	14	18	8	−1	0	2	6	10	16	26
26	6	−2	−1	1	5	7	14	19	8	−1	0	2	6	11	17	27
27	6	−2	−1	1	5	7	14	19	9	−1	0	2	6	11	18	28
28	6	−2	−1	1	5	7	14	19	9	−1	0	2	6	12	19	29
29	6	−2	0	1	5	8	15	19	10	−1	0	3	7	12	19	30
30	7	−2	0	2	5	8	15	19	10	0	0	3	7	13	20	31
31	7	−1	0	2	6	8	15	20	10	0	0	3	7	13	21	32
32	7	−1	0	2	6	8	15	20	11	0	1	3	8	14	22	33
33	7	−1	0	2	6	9	16	20	11	0	1	4	8	14	23	34
34	7	−1	0	2	6	9	16	20	12	0	1	4	8	14	23	35
35	8	−1	0	3	6	9	16	20	12	0	1	4	9	15	24	36
36	8	−1	0	3	6	10	16	21	13	0	1	5	9	15	25	37
37	8	−1	0	3	7	10	17	21	13	0	2	5	9	16	26	38
38	8	0	1	3	7	10	17	22	13	0	2	5	10	17	27	39
39	9	0	1	4	7	10	17	22	14	1	2	5	10	17	28	40
40	9	0	1	4	7	11	18	23	14	1	2	6	10	18	29	42
41	9	0	1	4	7	11	18	24	15	1	3	6	11	18	30	43
42	10	0	1	4	8	12	19	24	15	1	3	6	11	19	31	44
43	10	0	2	5	8	12	19	25	16	1	3	7	11	19	32	45
44	11	0	2	5	8	13	20	26	16	2	3	7	12	20	33	46
45	11	0	2	5	8	13	21	27	17	2	4	7	12	21	34	48
46	11	0	2	5	9	14	21	27	17	2	4	7	13	21	35	49
47	12	0	3	6	9	14	22	28	18	2	4	8	13	22	36	50
48	12	1	3	6	10	15	23	29	18	3	4	8	14	23	37	51
49	12	1	3	6	10	16	23	30	19	3	5	8	14	23	38	53
50	13	1	4	7	10	16	24	31	20	3	5	9	15	24	39	54
51	13	1	4	7	11	17	24	31	20	4	5	9	15	25	40	55
52	14	2	4	7	11	18	25	32	21	4	6	10	16	25	41	56
53	14	2	5	8	12	18	25	33	21	4	6	10	17	26	42	57
54	15	2	5	8	12	19	26	34	22	4	6	11	17	27	43	58
55	15	3	5	8	13	20	27	35	23	5	7	11	18	27	44	59
56	16	3	6	9	14	20	27	36	23	5	7	11	19	28	45	60
57	16	4	6	9	14	21	28	37	24	5	7	12	19	29	46	61
58	17	4	6	10	15	22	29	38	25	6	8	13	20	30	47	62
59	18	5	7	10	15	23	30	39	25	6	8	13	21	30	48	63
60	18	5	7	11	16	23	30	41	26	6	9	14	21	31	49	64
61	19	6	8	11	17	24	31	42	27	7	9	14	22	32	50	65
62	20	6	8	12	17	25	32	43	27	7	10	15	23	33	51	65
63	20	7	9	12	18	26	33	44	28	7	10	15	24	34	52	66
64	21	7	9	13	19	27	34	45	29	8	10	16	24	34	53	67
65	22	8	10	13	19	27	34	46	29	8	11	17	25	35	53	68

(Continued)

(Continued)

Ear = right				Frequency = 0.5, 1, 2 and 4 kHz												Gender = Male
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.5	P.10	P.25	P.50	P.75	P.90	P.95
66	22	8	10	14	20	28	35	47	30	9	11	17	26	36	54	69
67	23	9	11	14	21	29	36	48	31	9	12	18	27	37	55	69
68	24	10	11	15	22	30	36	48	31	9	12	18	27	37	56	70
69	24	10	12	15	22	30	37	49	32	10	13	19	28	38	56	71
70	25	11	12	16	23	31	38	50	33	10	13	20	29	39	57	72
71	26	11	13	16	24	32	39	51	33	10	14	20	30	40	58	72
72	26	12	13	17	24	33	39	52	34	11	14	21	31	40	58	73
73	27	12	14	17	25	33	40	53	35	11	15	21	31	41	59	74
74	28	13	14	18	26	34	41	54	36	12	15	22	32	42	60	74
75	28	14	15	18	27	35	42	55	36	12	15	23	33	43	60	75
76	29	14	15	19	27	36	42	56	37	12	16	23	34	43	61	76

Table A1.3 Male average ear. Distribution of hearing levels for screened and typical population as a function of age using a smoothing regression function on the 7−year running average

Ear = Average				Frequency = 0.5, 1, 2 and 4 kHz												Gender = Male
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
22	6	−1	−1	1	4	8	14	17	7	−1	0	2	5	10	15	18
23	6	−1	−1	1	4	8	14	17	7	−1	0	2	5	10	16	19
24	6	−1	−1	1	4	8	14	18	7	−1	0	2	5	11	16	20
25	6	−1	0	1	4	8	14	18	8	0	0	2	6	11	17	21
26	7	−1	0	1	5	8	15	18	8	0	0	3	6	12	18	22
27	7	0	0	2	5	9	15	18	9	0	0	3	6	12	18	23
28	7	0	0	2	5	9	15	19	9	0	0	3	7	13	19	24
29	7	0	0	2	5	9	15	19	10	0	0	3	7	13	20	25
30	7	0	0	2	5	9	16	19	10	0	1	4	7	14	21	26
31	8	0	0	2	6	9	16	20	11	0	1	4	8	14	21	27
32	8	0	0	2	6	10	16	20	11	0	1	4	8	15	22	28
33	8	0	0	3	6	10	16	20	11	0	1	4	8	15	23	29
34	8	0	1	3	6	10	16	20	12	0	2	5	9	16	24	30
35	8	0	1	3	6	10	17	21	12	1	2	5	9	16	24	31
36	9	0	1	3	6	10	17	21	13	1	2	5	9	17	25	32
37	9	1	1	3	7	11	17	22	13	1	2	5	10	17	26	33
38	9	1	2	4	7	11	18	22	14	1	2	6	10	18	27	34
39	9	1	2	4	7	11	18	23	14	2	3	6	11	19	28	35
40	10	1	2	4	7	11	18	24	15	2	3	6	11	19	29	36
41	10	1	2	4	8	12	19	25	15	2	3	7	11	20	30	38
42	10	2	2	4	8	12	20	25	16	2	4	7	12	20	30	39
43	11	2	3	5	8	13	20	26	16	2	4	7	12	21	31	40
44	11	2	3	5	9	13	21	27	17	3	4	7	13	22	32	41
45	11	2	3	5	9	14	21	28	17	3	4	8	13	22	33	42
46	12	2	3	5	9	14	22	29	18	3	5	8	13	23	34	44
47	12	2	4	6	9	15	23	30	18	3	5	8	14	23	35	45
48	12	2	4	6	10	15	23	30	19	4	5	9	14	24	36	46
49	13	3	4	6	10	16	24	31	19	4	5	9	15	25	37	47
50	13	3	4	7	10	17	25	32	20	4	6	9	16	26	38	48
51	14	3	5	7	11	17	25	32	20	4	6	10	16	26	39	50
52	14	3	5	7	11	18	26	33	21	5	7	10	17	27	40	51
53	15	4	5	8	12	18	27	34	22	5	7	11	17	28	41	52
54	15	4	5	8	12	19	27	34	22	5	7	11	18	29	42	53
55	16	4	6	9	13	20	28	35	23	6	8	12	19	29	43	54
56	16	5	6	9	13	20	29	36	24	6	8	12	19	30	44	56
57	17	5	7	10	14	21	30	37	24	6	9	13	20	31	45	57
58	18	5	7	10	15	22	30	39	25	7	9	13	21	32	46	58
59	18	6	8	11	15	23	31	40	26	7	9	14	21	33	47	59
60	19	6	8	11	16	23	32	41	26	7	10	14	22	33	48	60
61	20	7	8	12	17	24	33	42	27	8	10	15	23	34	49	61
62	20	7	9	12	17	25	34	43	28	8	11	16	24	35	50	62
63	21	8	9	13	18	26	35	44	28	8	11	16	24	36	51	63
64	22	8	10	14	19	26	35	45	29	9	12	17	25	37	52	63
65	22	9	10	14	20	27	36	46	30	9	12	18	26	38	53	64

(Continued)

(Continued)

Ear = Average				Frequency = 0.5, 1, 2 and 4 kHz												Gender = Male
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
66	23	9	11	15	20	28	37	46	30	10	13	18	27	38	54	65
67	24	10	11	16	21	28	38	47	31	10	13	19	27	39	54	66
68	24	10	12	16	22	29	38	48	32	10	14	19	28	40	55	67
69	25	11	12	17	23	30	39	49	32	11	14	20	29	41	56	67
70	26	11	13	18	23	30	40	50	33	11	15	21	30	42	57	68
71	26	12	13	18	24	31	41	51	34	12	15	21	31	42	57	69
72	27	12	14	19	25	32	42	51	34	12	16	22	31	43	58	70
73	28	13	14	20	26	33	42	52	35	12	17	23	32	44	59	71
74	28	13	15	20	26	33	43	53	36	13	17	23	33	45	60	71
75	29	14	15	21	27	34	44	54	36	13	18	24	34	46	60	72
76	30	14	16	22	28	35	45	55	37	13	18	24	34	47	61	73

Table A1.4 Female right ear. Distribution of hearing levels for screened and typical population as a function of age using a smoothing regression function on the 7−year running average

Ear = right				Frequency = 0.5, 1, 2 and 4 kHz												Gender = Female
	SCREENED								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
22	4	−1	−1	1	4	6	9	13	5	−2	−1	1	4	8	13	21
23	4	−2	−1	1	4	7	9	14	6	−2	0	1	4	8	13	22
24	4	−2	−1	1	4	7	10	14	6	−2	0	2	4	8	14	23
25	5	−2	−1	1	4	7	10	14	7	−2	0	2	5	9	15	24
26	5	−2	−1	1	4	7	10	15	7	−2	0	2	5	9	16	25
27	5	−2	0	2	4	8	11	15	7	−1	0	2	5	9	16	26
28	5	−1	0	2	5	8	11	16	8	−1	0	2	5	9	17	27
29	6	−1	0	2	5	8	11	16	8	−1	0	3	6	10	18	27
30	6	−1	0	2	5	8	12	17	8	−1	0	3	6	10	19	28
31	6	−1	0	2	5	9	12	17	9	−1	0	3	6	11	19	29
32	6	−1	0	2	5	9	13	18	9	−1	0	3	7	11	20	30
33	7	−1	0	2	5	9	13	18	9	0	0	3	7	11	21	31
34	7	−1	0	3	5	9	13	19	10	0	1	3	7	12	22	32
35	7	0	0	3	6	10	14	19	10	0	1	4	7	12	22	33
36	7	0	0	3	6	10	14	20	11	0	1	4	8	13	23	34
37	8	0	0	3	6	10	14	20	11	0	1	4	8	13	24	35
38	8	0	0	3	6	11	15	21	11	0	1	4	8	13	24	36
39	8	0	1	3	6	11	15	21	12	0	2	4	8	14	25	36
40	9	0	1	4	7	11	15	22	12	0	2	5	9	14	26	37
41	9	0	1	4	7	11	16	22	12	0	2	5	9	15	27	38
42	9	0	1	4	7	12	16	23	13	0	2	5	9	15	27	39
43	9	0	1	4	7	12	17	23	13	0	2	5	10	16	28	39
44	10	0	2	4	8	12	17	24	13	0	3	5	10	16	29	40
45	10	0	2	5	8	13	18	25	14	0	3	6	10	17	30	41
46	10	0	2	5	8	13	18	25	14	1	3	6	11	17	30	42
47	11	0	2	5	9	14	19	26	15	1	3	6	11	18	31	43
48	11	0	2	5	9	14	19	27	15	1	3	7	11	18	32	44
49	12	1	3	6	9	14	20	28	16	1	4	7	12	19	33	45
50	12	1	3	6	10	15	20	28	16	1	4	7	12	20	34	46
51	12	1	3	6	10	15	21	29	17	2	4	8	13	20	35	48
52	13	1	3	7	10	16	22	30	17	2	5	8	13	21	36	49
53	13	1	4	7	11	16	23	30	18	2	5	8	14	21	37	50
54	14	1	4	7	11	17	24	31	18	3	5	9	14	22	37	51
55	14	2	4	7	12	17	25	32	19	3	6	9	15	23	38	52
56	15	2	5	8	12	18	26	33	19	3	6	9	15	24	39	53
57	15	2	5	8	13	19	27	34	20	4	6	10	16	24	40	54
58	16	3	5	9	13	19	28	35	21	4	6	10	16	25	41	55
59	17	3	6	9	14	20	29	37	21	4	7	11	17	26	42	56
60	17	3	6	10	15	21	30	38	22	4	7	11	18	27	43	57
61	18	3	6	10	15	22	31	39	23	5	7	12	18	28	44	58
62	19	4	6	11	16	23	32	41	23	5	8	12	19	30	46	59
63	19	4	7	11	17	24	33	42	24	5	8	13	20	31	47	61
64	20	4	7	12	17	25	34	43	25	6	8	13	20	32	48	62
65	21	5	7	12	18	26	36	44	26	6	9	14	21	33	49	63

(Continued)

(Continued)

Ear = right				Frequency = 0.5, 1, 2 and 4 kHz												Gender = Female
	SCREENED								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
66	21	5	8	13	19	27	37	46	26	6	9	14	22	34	50	64
67	22	5	8	13	19	28	38	47	27	7	9	15	22	35	51	66
68	23	6	8	14	20	29	39	48	28	7	10	15	23	36	53	67
69	23	6	9	14	21	30	40	49	29	8	10	16	24	37	54	68
70	24	6	9	15	21	31	42	51	29	8	10	16	24	38	55	69
71	25	7	9	15	22	31	43	52	30	8	11	17	25	39	56	71
72	26	7	10	16	22	32	44	53	31	9	11	17	26	40	57	72
73	26	7	10	16	23	33	45	54	31	9	11	18	27	41	58	73
74	27	7	10	17	24	34	47	55	32	10	12	18	27	42	60	75
75	28	8	11	17	24	35	48	57	33	10	12	19	28	43	61	76
76	28	8	11	18	25	36	49	58	34	10	12	19	29	44	62	77

Table A1.5 Female left ear. Distribution of hearing levels for screened and typical population as a function of age using a smoothing regression function on the 7−year running average

Ear = left				Frequency = 0.5, 1, 2 and 4 kHz												Gender = Female
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
22	4	−1	−1	0	4	7	11	14	6	0	0	1	5	8	14	24
23	5	−1	−1	1	4	8	11	14	6	0	0	2	5	9	15	25
24	5	−1	−1	1	4	8	12	15	7	−1	0	2	5	9	15	26
25	5	−1	0	1	4	8	12	15	7	−1	0	2	5	9	16	26
26	5	−1	0	1	5	8	12	15	7	−1	0	2	5	9	16	27
27	5	−1	0	1	5	8	12	15	8	−1	0	2	6	10	17	28
28	6	−1	0	2	5	9	12	16	8	−1	0	2	6	10	17	28
29	6	−1	0	2	5	9	13	16	8	−1	0	3	6	10	18	29
30	6	−1	0	2	5	9	13	16	8	0	0	3	6	11	18	30
31	6	−1	0	2	5	9	13	16	9	0	0	3	6	11	19	31
32	6	−1	0	2	6	10	13	17	9	0	0	3	6	11	19	31
33	7	−1	0	2	6	10	13	17	9	0	0	3	7	12	19	32
34	7	−1	0	3	6	10	14	17	10	0	1	3	7	12	20	33
35	7	0	0	3	6	10	14	17	10	0	1	4	7	12	20	33
36	7	0	0	3	6	10	14	18	10	0	1	4	7	12	21	34
37	8	0	1	3	7	11	14	18	10	0	1	4	8	13	21	34
38	8	0	1	3	7	11	15	18	11	0	1	4	8	13	22	35
39	8	0	1	3	7	11	15	19	11	0	2	4	8	13	22	35
40	8	0	1	4	7	11	15	19	11	0	2	5	8	14	23	36
41	9	0	1	4	7	12	15	19	12	1	2	5	9	14	23	37
42	9	0	2	4	8	12	16	20	12	1	2	5	9	14	24	38
43	9	0	2	4	8	12	16	20	12	1	2	5	9	15	25	39
44	9	0	2	5	8	13	17	21	13	1	3	6	9	15	25	40
45	10	0	2	5	8	13	17	21	13	1	3	6	10	16	26	40
46	10	0	2	5	9	13	17	22	13	2	3	6	10	16	26	41
47	10	1	3	5	9	14	18	23	14	2	3	6	10	17	27	42
48	11	1	3	6	9	14	18	23	14	2	4	7	11	17	28	42
49	11	1	3	6	9	14	19	24	15	2	4	7	11	18	29	43
50	11	1	3	6	10	15	19	25	15	3	4	7	11	18	29	43
51	12	2	4	6	10	15	20	25	16	3	5	7	12	19	30	44
52	12	2	4	7	10	16	21	26	16	3	5	8	12	19	31	45
53	13	2	4	7	11	16	21	27	17	3	5	8	13	20	32	45
54	13	3	5	7	11	16	22	28	17	4	5	9	13	21	33	46
55	14	3	5	8	12	17	23	29	18	4	6	9	13	22	34	47
56	14	3	5	8	12	17	24	30	18	4	6	9	14	22	35	47
57	15	4	5	8	12	18	25	31	19	5	6	10	14	23	36	48
58	15	4	6	9	13	19	26	32	19	5	7	10	15	24	37	49
59	16	4	6	9	13	19	27	33	20	5	7	10	16	25	38	50
60	16	5	6	10	14	20	28	35	21	5	7	11	16	26	39	50
61	17	5	6	10	14	21	29	36	21	6	8	11	17	27	40	51
62	18	5	7	10	15	22	30	37	22	6	8	12	18	28	41	52
63	18	5	7	11	16	23	31	38	23	6	8	12	18	29	42	53
64	19	6	7	11	16	24	32	40	23	7	9	13	19	30	43	54
65	20	6	8	12	17	25	34	41	24	7	9	13	19	31	45	55

(Continued)

(Continued)

Ear = left						Frequency = 0.5, 1, 2 and 4 kHz									Gender = Female	
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
66	20	6	8	12	17	25	35	42	25	7	9	14	20	32	46	56
67	21	7	8	13	18	26	36	43	26	7	10	14	21	33	47	58
68	22	7	8	13	18	27	37	45	26	8	10	14	21	34	48	59
69	22	7	9	13	19	28	38	46	27	8	10	15	22	35	49	60
70	23	7	9	14	20	29	39	47	28	8	10	15	23	36	50	61
71	23	8	9	14	20	29	40	48	28	8	11	16	23	37	51	62
72	24	8	10	15	21	30	42	49	29	9	11	16	24	38	52	63
73	25	8	10	15	21	31	43	51	30	9	11	17	25	39	54	64
74	25	9	10	16	22	32	44	52	31	9	12	17	25	40	55	65
75	26	9	10	16	22	33	45	53	31	9	12	18	26	41	56	66
76	27	9	11	16	23	34	46	54	32	10	12	18	26	42	57	67

Table A1.6 Female average ear. Distribution of hearing levels for screened and typical population as a function of age using a smoothing regression function on the 7−year running average

Ear = Average							Frequency = 0.5, 1, 2 and 4 kHz									Gender = Female
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
22	4	−2	−1	1	3	6	10	13	7	0	0	2	5	7	13	21
23	4	−2	0	1	3	6	10	13	7	0	0	2	5	7	13	22
24	5	−1	0	1	4	7	10	14	7	0	0	2	5	8	14	22
25	5	−1	0	1	4	7	10	14	7	0	0	2	5	8	15	23
26	5	−1	0	1	4	7	11	14	8	0	0	2	5	9	15	24
27	5	−1	0	2	4	7	11	15	8	0	0	3	6	9	16	25
28	6	−1	0	2	4	8	11	15	8	0	0	3	6	9	16	26
29	6	−1	0	2	5	8	12	16	8	0	0	3	6	10	17	27
30	6	−1	0	2	5	8	12	16	8	0	0	3	6	10	18	27
31	6	0	0	2	5	8	12	16	9	0	0	3	6	10	18	28
32	6	0	0	2	5	9	12	17	9	0	1	3	7	11	19	29
33	7	0	0	3	5	9	13	17	9	0	1	4	7	11	20	30
34	7	0	1	3	6	9	13	18	10	0	1	4	7	11	20	31
35	7	0	1	3	6	9	13	18	10	0	1	4	7	12	21	32
36	7	0	1	3	6	10	14	18	10	0	1	4	7	12	22	32
37	8	0	1	3	6	10	14	19	11	0	2	4	8	13	22	33
38	8	0	1	3	6	10	14	19	11	1	2	4	8	13	23	34
39	8	0	2	4	7	10	15	20	11	1	2	5	8	13	23	35
40	8	0	2	4	7	11	15	20	12	1	2	5	8	14	24	35
41	9	0	2	4	7	11	15	21	12	1	2	5	9	14	25	36
42	9	0	2	4	7	11	16	21	12	1	3	5	9	15	25	37
43	9	0	2	5	8	12	16	22	13	1	3	6	9	15	26	38
44	10	1	2	5	8	12	17	23	13	2	3	6	10	16	27	38
45	10	1	3	5	8	12	17	23	13	2	3	6	10	16	27	39
46	10	1	3	5	8	13	18	24	14	2	3	6	10	17	28	40
47	11	1	3	6	9	13	18	24	14	2	4	7	11	17	29	41
48	11	1	3	6	9	14	19	25	15	2	4	7	11	18	30	42
49	11	1	3	6	9	14	19	25	15	2	4	7	11	18	31	43
50	12	2	3	6	10	14	20	26	16	3	4	8	12	19	32	44
51	12	2	4	7	10	15	20	27	16	3	5	8	12	19	32	45
52	13	2	4	7	10	15	21	27	17	3	5	8	13	20	33	46
53	13	2	4	7	11	16	21	28	17	3	5	9	13	21	34	46
54	13	3	5	8	11	16	22	29	18	4	6	9	14	22	35	47
55	14	3	5	8	12	17	23	30	18	4	6	9	14	22	36	48
56	14	3	5	8	12	18	24	31	19	4	6	10	15	23	36	49
57	15	4	5	9	13	18	25	32	19	5	6	10	15	24	37	49
58	16	4	6	9	13	19	26	33	20	5	7	11	16	25	38	50
59	16	4	6	9	14	20	27	34	21	5	7	11	16	26	39	51
60	17	4	6	10	14	21	28	36	21	5	7	11	17	27	40	52
61	17	5	6	10	15	21	29	37	22	6	8	12	18	28	41	52
62	18	5	7	11	15	22	30	38	23	6	8	12	18	29	43	53
63	19	5	7	11	16	23	31	39	23	6	8	13	19	30	44	54
64	19	6	7	12	17	24	32	41	24	6	9	13	20	31	45	55
65	20	6	8	12	17	25	34	42	25	7	9	14	20	32	46	56

(Continued)

(Continued)

Ear = Average				Frequency = 0.5, 1, 2 and 4 kHz												Gender = Female
	Screened								Typical							
Age	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95	μ	P.05	P.10	P.25	P.50	P.75	P.90	P.95
66	21	6	8	13	18	26	35	43	25	7	10	14	21	33	47	56
67	21	6	8	13	19	27	36	44	26	7	10	15	22	34	48	57
68	22	7	8	14	19	28	37	45	27	8	10	15	23	35	49	58
69	23	7	9	14	20	29	38	46	27	8	11	16	23	36	50	59
70	23	7	9	15	20	29	39	48	28	8	11	16	24	37	51	60
71	24	7	9	15	21	30	40	49	29	8	11	17	25	38	52	61
72	25	8	9	16	22	31	41	50	30	9	12	17	25	39	53	62
73	25	8	10	16	22	32	42	51	30	9	12	18	26	40	55	62
74	26	8	10	16	23	33	43	52	31	9	12	18	27	41	56	63
75	27	8	10	17	24	34	45	54	32	10	13	19	27	42	57	64
76	27	9	11	17	24	35	46	55	32	10	13	19	28	43	58	65

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Appendix 2: Example of patient information regarding hearing aids

What follows is an example of written patient information regarding care and maintenance of hearing aids (reproduced with the kind permission of the audiology department at Queen's Medical Centre, Nottingham).

WHAT TO EXPECT WITH YOUR HEARING AID(S)

Hearing aids amplify sound to make it louder. This can help to overcome a hearing loss and improve communication. It also means that sounds around you are easier to hear.

To begin with, you will hear lots of sounds that you may not have heard for a while. Your own voice may sound strange at first, but this will quickly begin to sound more natural.

Remember, the world is a noisy place. Don't worry, you won't be hearing anything that you wouldn't hear with normal hearing, but it may take some time for you to get used to these new sounds.

Some people like to wear their aids straight away, for others it takes a little time to be able to build up to wearing them all day. Whichever you do, just remember that the more you wear the aids, the more you will get used to them and the more they will help you.

Advantages

Hearing aids give you a better awareness of sounds around you and help with conversation in certain situations (e.g. quieter environments, listening to people close by or the television or telephone). However, they have some limitations, and how much they help will depend on your hearing loss.

Hearing aids are less effective:

- In very noisy environments.
- In large groups.
- Listening to someone in another room.
- Speech from more than six feet away.

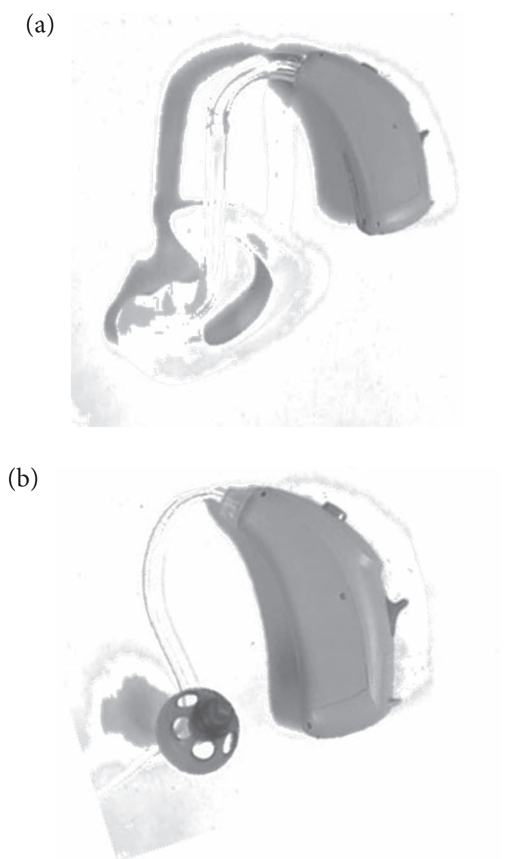


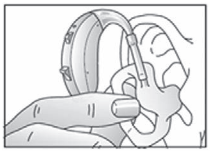
Figure A2.1 Types of hearing aids. **(a)** Hearing aid and earmould. **(b)** Open fit hearing aid.

Your hearing aid may be on an earmould or a slim tube and dome fitting. This is dependent on your individual needs. This will have been discussed with you at your initial appointment.

PUTTING YOUR HEARING AID IN YOUR EAR

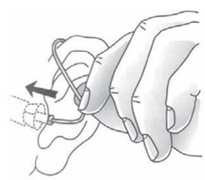
Earmould Insertion

1. Hold the edge of the earmould between your thumb and index finger.
2. Take the earmould backwards, pointing it towards your ear canal.
3. Slide the earmould all the way into your ear with a gentle, twisting movement.
4. Tuck the top part of the earmould behind the fold of skin above your ear canal.
5. Finally, hook the hearing aid behind the ear.



Thin Tube Insertion

1. Hook the hearing aid over the top of your ear first.
2. Hold the thin tube where it bends and gently push the dome into the ear canal.
3. Push the dome far enough into the ear canal so that the thin tube lies flush with the side of your face (checking in the mirror can be helpful).



DAILY CLEANING

CLEANING EARMOULDS

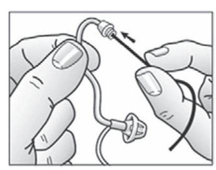
1. Detach the tubing from the hearing aid before cleaning. **Don't pull the tube out of the earmould.**
2. Use mild soap and warm water to clean the earmould and tube.
3. Dry the earmould thoroughly ensuring no water is left in the tubing.
4. Reattach to the hearing aid.

If water is blocking the tubing – give the mould a shake, blow through the tube and leave on the side to fully dry.

Alternatively, you can purchase a cleaning kit from us which includes an earmould blower to remove any excess water.

CLEANING THIN TUBES

1. Unscrew the thin tube carefully from the hearing aid.
2. Wipe down the thin tubes and domes with a damp cloth.
3. Push the cleaning wire right through the thin tube, beginning at the end opposite the dome to push out any wax or debris blocking the tube.
4. Screw the tube back onto the hearing aid.



HEARING AID MAINTENANCE

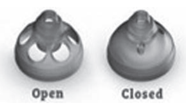
Batteries

Battery life is dependent on battery size, hearing aid type and usage:

Typical battery life:
Size 675 – 2–4 weeks
Size 13 – 7–10 days
Size 312 – 3–6 days

Thin Tubing and Domes

Open fit parts have a limited lifespan. Domes should be replaced every 6–12 months and thin tubes every 12 months. If you remove a dome, it is advisable to attach a new dome to replace it.



Earmould Tubing

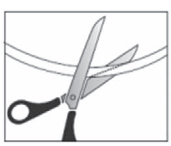
Your earmould tubing will need to be changed every 4–6 months.

If you are confident with tools and small objects, we can supply spare tubing so you can change the tubing yourself. For how to do this, see the next page. Ask your audiologist for advice.

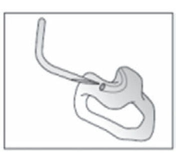
How to Change Earmould Tubing

Detach the mould from the hearing device.
Pull out the old tube and keep for now.

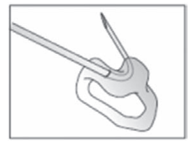
Cut the last 2” / 5 cm of a pre-bent piece of tubing to a long tapered end.



Thread the long tapered end through the mould until it comes out of the other end.

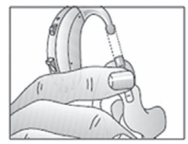


Pull the pointed end of the tubing through the mould until the other end of the tubing is vertical (90 degree angle).



Cut off the pointed end as close to the mould as possible. Ensure there are no rough edges of the tube left sticking out.

Place the old piece of tubing, which you removed earlier, alongside the newly fitted tube measure and cut to length. Then reattach to aid.



Additional Information

You may find it useful to watch short interactive videos on maintaining and making the most out of your hearing aids at www.C2Hearonline.com.

C2Hear
Hearing well, together

INDUCTION LOOP

An induction loop (**loop / telecoil / T setting**) enables the hearing aid user to pick up sound more clearly at a distance or across a counter window where a loop system is installed. When using the Loop / T setting, sound is transferred directly into the hearing aid from the person speaking, reducing most background noise.

Some examples of places you may find induction loops:

- Theatres.
- Cinemas.
- Conference halls.

- Churches.
- Banks.
- Post offices.
- Supermarkets.

If you would like to use the loop system, you need an additional setting added to your hearing aid; ask your audiologist to add this. They will then show you how to use it.

OTHER ASSISTIVE LISTENING DEVICES

Specialist equipment designed for those with a hearing loss include:

- TV listening devices.
- Doorbells.
- Smoke alarms.
- Amplified telephones.
- Alarm clocks.
- Remote microphones.

If you have a problem with your hearing aid, you may be able to solve the problem easily yourself.

Here are a few common problems, their causes and a possible remedy.

COMMUNICATION TACTICS

Hearing aids can help a lot with communication, however they can't do everything. There are things you and other people can do to improve your chances of hearing the conversation. This will also be less stressful. The following logos are useful communication visual aide memoires.

Table A2.1 Troubleshooting guide

Problem	Cause	Possible remedy
No sound	• Hearing aid is turned off The battery is dead / out-of-date • The tube is blocked/broken • Hearing aid on loop system setting	• Switch on by closing battery drawer • Insert new battery • Clean or replace the tube • Switch to normal setting
Feedback 'whistling'	• Earmould or thin tube and dome not in the ear properly • The tube is split or broken • Are you holding an object too close to a hearing aid? • There may be excess wax in your ear	• Insert earmould or dome correctly into your ear • Replace tube • Move the object away from the hearing aid • See a specialist to remove the wax
Sounds distorted/quiet	• Is the tube blocked/broken? • Hearing aid has become wet or there is condensation in the tube	• Clean or replace the tube • Use the drying crystals (purchase from Ropewalk)
Earmould uncomfortable	• Earmould is inserted incorrectly in the ear? • Are there any sharp or bulky areas that are causing discomfort? • Tubing is too short?	• Re-insert correctly, making sure it tucked in at the top • See audiologist to adjust the shape slightly • Change tube and cut tubing at correct length

Get my attention
Before you start to speak you must ensure you are in the same room as the person and you have their full attention.

Don't speak too fast
If someone doesn't understand what you are saying, you need to try and slow down your speech.

Face them
Always turn and face the person as it helps them pick up any visual clues you may give them.

Get to the point
Use plain language and don't waffle.

State the topic
If the context is known, it is easier for someone to understand what you are saying. State the topic before moving from one conversation to another.

Rephrase
Rephrase what you have said with different words if the person has not understood what you have said even after you have repeated it.

Confirm details
Do not assume that someone has understood what you have said to them, confirm important details.

Time
Give plenty of time for the person to understand what you have said.
Say what needs to be said slowly and pause between phrases.

Don't shout
You should keep your voice at a normal level. It is uncomfortable for a hearing aid user if you shout and it looks aggressive.

Write things down
If someone is still struggling to understand you, write down what you are trying to say.

Index

Note: page numbers in *italic* refer to tables or figures in the text

A

acoustic neuroma see vestibular schwannoma
acoustic reflex 78
acute otitis media, diagnosis *15*
adenoidectomy for OME 88
age-related hearing loss 8, 9, 100
  co-existing conditions/medications 102, 111
  prevalence *15*
air–bone gap definition 14, 55–57
  identification by Rinne test 26, *26*, 28
  pathophysiology in chronic otitis media 53–5
  and potential benefit of surgery 131
  prevalence in adult population 55–7, *56–7*
air-conduction masking in pure-tone audiometry 21–2, *22*
  methods 21–2, *21*, *22*
air-conduction thresholds
  asymmetry 103
  postulated configuration due to noise exposure 109–10
alerting door, telephone and gas devices 116
American Academy of Otolaryngology, Head and Neck Surgery (AAOHNS) 103, 153–4
American Sign Language, fingerspelling alphabet *119*
aminoglycoside antibiotics 96–7, 102
amplification devices, non-personal 116–17
anaesthesia, cochlear implantation 149
assistive listening devices 116, 176
audit 153–4
auditory brainstem implants 150–1
auditory brainstem response (ABR) 80–1, *80*
auditory memory 146
auditory neuropathy spectrum disorder (ANSD) 95
auditory neuroplasticity 145–7
auditory pathway 12, *12*
auditory processing disorder 97
auditory steady state response (ASSR) 82, *83*
auditory system, development 8
auditory verbal therapy 118–19
autoimmune disease in sudden hearing loss 104

B

background noise; effect on audiometry 32–3, *33*
  hearing aid suppression 126
bacterial biofilms in OME 87
bacterial meningitis in children 96
balance issues in superior semicircular canal dehiscence 58–9
  in testing 25, *25*
Barany box 22, *22*, 25, 27–8
batteries
  hearing aids 124, 175
  middle ear implants 142
behavioural testing, limitations 65–7, *66*
behind-the-ear (BTE), hearing aid 124, *125*
blast explosions 104–5
bone conduction audiometry *see* pure-tone audiometry 50
  as measure of inner ear function 50–1, *51*
  testing 25
  transcranial attenuation 36, *36*
bone-conduction hearing aids 126–8, *128–9*
  audiometric fitting range vs implanted bone-conduction devices *129*
  Bruckhoff glasses *128*
  Cochlear BAHA SoundArc *128*
  MED EL AdHear adhesive *128*
bone-conduction hearing implants 141, *141*
  transcutaneous 142, *142*
bone-conduction clinical masking 25–6, *25*
bone-conduction thresholds, child behavioural testing 66
brainstem-evoked response audiometry, *see* auditory brainstem response
British Sign Language, fingerspelling alphabet *118*
Bruckhoff glasses *128*

C

C2Hear videos 115, 175
Carhart effect 51–5, *52*, 132
cerumen, *see* wax
chalk patches, tympanic membrane 53
Charcot-Marie-Tooth 95
chemotherapy, ototoxicity 96, 102
child hearing testing 62–7
  conditioning 62–3
  and developmental stage 62
  distraction testing 64–5, *65*
  environment 62
  limits of behavioural testing 65–7
  pitfalls in behavioural testing in children 65
  play audiometry 64, 65, *65*
  speech testing 69–71, *70–1*
  test stimuli 63, *63*
  visual reinforcement audiometry 63–4, *64*, *66*
children
  cochlear implants 146
  congenital hearing loss, *see* congenital hearing loss
  hearing aids 120
cholesteatoma 19–20, *19*
  complications *19*, 20
  inability to prevent following OME 89
  prevention in OME 89
  radiology 20
  surgical management 138
chronic otitis media 18–21
  active 19–20, *19*
  active mucosal 19, *19*
  active squamous 19, *19*
  diagnosis *15*, 18
  healed 16, 18, *18*, *57*
  inactive 18–19, *19*
  ossicular chain pathology 55, *56*
  pathophysiology of air–bone gap 53–55, 57
  prevalence *17*
cleft palate OME 89–90
clinical evaluation
  hearing thresholds 21–2
  history taking 14–16, *15*
  otoscopy 16–21
  tuning fork tests 24–7, *25–6*, 28
  voice tests 22–4, *24*, 27–8
  close-captioned/subtitled television 117
cochlea
  anatomy and function 7, *7*, *8*
  tonotopic arrangement 147, *147*
Cochlear BAHA SoundArc *128*
cochlear implant candidacy
  NICE guidelines 147–8, *148*
  speech testing 72–3
  bilateral 147
  internal and external components *146*, 147
  likelihood of benefit 146–7
  optimal age for implantation 146
  patient aftercare 149–50, *149–50*
  patient pathway 148–9
  principles 145, *145*
  structure preservation 150
  sudden profound hearing loss 150
  unilateral 147
cochlear nerve 7
cochlear nucleus 7
Cochlear Osia *141*
COMET and Core Outcomes Set 152, 156
communication tactics 176–7
completely-in-the-canal (CIC), hearing aid 124, *125*
computed tomography (CT)
  cochlear implantation 148, 149, *149*
  conditioning phase in children before hearing testing 62–3
conduction defects 12
  otoscopy 48
  pure tone audiometry 42, 48–50
  tuning fork tests 48
congenital sensorineural hearing loss in children 8–9, *9*, *10*
  aetiological investigation 93–4
  causes 8–9, *9*, *10*, 94
  cochlear implantation 145–6
  epidemiology 9, *9*
  management 94–5
  permanent at birth 8–9, *9*
  progressive 95–7
consent: informed 153, 155, *156*
contralateral routing of sound (CROS) hearing aids 104, 123
Control of Noise at Work 2005 Employment Regulations 106
conversation listening techniques 115–16
Core Outcome Sets (COS) 152–3
cortical auditory evoked potentials (CAEP) 82
cytomegalovirus (CMV) infection 9, *9*, 93, 94, 95

D

data required interpretation
  conduction defects 42, 48–50
  exaggerated hearing loss 45
  mixed impairment 42–3
  non-organic hearing loss 45–6
  sensorineural impairment 40–2
  uncertain category 43–5
deaf culture 118–19
deaf intonation *72*, 146
deafness, severe-to-profound 147
decibel (dB) scales 1–2, *2*
decibel (dB) A 5
  dB SPL (sound pressure level) 32
  dB HL (hearing level) 13, 31
  dB A (noise level) 104–7
  levels of detection of human ear 31–2, *32*
developing world, hearing loss 9
distraction testing 64–5, *65*
doorbell alerting alarms 116
Down Syndrome (trisomy 21) 89, 97

E

ear defenders 106, *106*
  ear muffs 106, *106*, 108
  ear plugs; cotton wool, expandable foam, pre-moulded 107–8
ear protection
  gunfire noise exposure 110–11
  history taking 107–8
  types and efficacy 105–6, *106*

earmoulds 123–4, *124*, *173*
cleaning 174, *174*
insertion 174, *174*
tubing *174–5*
Eustachian tube dysfunction 75, *76*
in OME 86, 87
population prevalence *57*
external auditory canal, acoustic properties 104, *105*

F

FFA, *see* four-frequency average (FFA) thresholds
fingerspelling 117
American Sign Language alphabet *119*
British Sign Language alphabet *118*
floating mass transducer 141, *142*
follow-up of hearing management 153–4
four-frequency average (FFA) thresholds
by age and sex 160–71
female 166–71
male 160–5
occupational noise exposure 108–9, *109*
free-field voice tests 22–4, *24*
comparison with audiometry 24
grading of severity 24, *24*
method 23
value of 22–3

G

gene therapies 94
gene panel; R67 monogenic hearing loss 94, 96
genetic counselling 94
genetics, hearing loss 9, *9*, *10*
GJB2 (connexin 26) gene mutations *9*, *10*, 94
Glasgow Benefit Inventory 154, *154*
Glasgow Children's Benefit Inventory 155
Glasgow Hearing Aid Benefit Profile 115
Glasgow Benefit plot *135–7*, 136–7, 143
gunfire noise exposure 110–11
ear protection 106, *106*, 110
evaluation 110–11

H

hair cells; inner and outer 7
head injuries 103
Hearing in Adults 100–3
age-related FFA thresholds 102
audiogram configuration 101–2, *101*
related general health factors 102
hearing aid assessment in children, speech testing 71
hearing aids 120–6
acoustical data 124–5, *126*
advantages 173
background noise suppression 126
best-practice audiology delivered 121
bone-anchored 141–3
bone-conduction 126–8, *128–9*
candidacy 121, *121*
in child with trisomy 21 89
children 120
communication tactics 176–7
contralateral routing of sound (CROS) 104, 123
daily cleaning *174*
domes 175
ear moulds 123–4, *124*, *173*, *174*
earmoulds *173–5*
follow-up strategies 126
habituation 115
long-term usage 126
maintenance 175, *175*
medical assessment 122
monoaural, choice of ear 122–3
monoaural vs binaural fitting 122
open fit *173*, 175
in otitis media with effusion 87–8
outcome reporting 155
overview 120–1
patient information 115, 173–7
power supply 124, 175
situations where less effective 173
styles 124, *125*
thin tubing *174–5*, 175
troubleshooting guide *176*
tuition in use 126
Hearing in animals
fish 3
snakes 4, *4*
hearing impairment 13
severity classification *13*
hearing loop devices 116, 175–6
hearing loss
age-related, *see* age-related hearing loss
congenital, *see* congenital sensorineural hearing loss
cultural implications 118–19
epidemiology 8–9
exaggerated 45
non-organic 45–6, 71
hearing protection from noise *see* ear defenders
history taking 107–8
types and efficacy 105–6, *106*
Hearing specific outcomes for non-surgical interventions including hearing aids 155
Hennebert sign 59
history taking in making a diagnosis 12, 14–15, 14–16, *15*
in noise exposure 106–8
human hearing
normal sound pressure levels (dB SP) 31–2, *32*

I

in-the-ear (ITE), hearing aid 124, *125*
incudo-mallleolar joint 5
induction loop 116, 175–6
informed consent 153, 155, *156*
infrared aiding systems 116
inner ear
anatomy 5–8
bone conduction measure 50–1, *51*
infections affecting 96
surgical damage 133–4
International Outcome Inventory for hearing aids (IOI-HA) 155

J

Jervell and Lange-Nielsen (JLN) syndrome 93

K

KCNJ10 mutations 96

L

ling sounds 69
listening, failure of 97
listening techniques/devices 115–16, 176

M

m.1555A>G mutation 94, 96–7, 102
McCormick Toy Test 70, *70*
magnetic resonance imaging (MRI)
  cochlear implantation 148
  congenital sensorineural hearing loss 93–4
Makaton 119–20
Manchester Picture Test 70–1, *71*
Masking see Pure-tone audiometry 36–40
  air-conduction 37–40
  in child hearing testing 63, 66–7
  conduction defects identification 50
mastoid surgery 138, *138*
measles, mumps and rubella (MMR) vaccine 96
MED EL AdHear adhesive bone-conduction hearing aid *128*
MED EL Bonebridge *142*
MED EL Vibrant Soundbridge 142, *142*
  candidacy guidelines 141
meningitis 96
middle ear, anatomy and function 2–5
middle ear disease
  prevalence 16
  see also chronic otitis media; otitis media with effusion
middle ear implants 141–3
  candidacy guidelines 143, *143*
  design challenges 141
middle ear surgery 131–2
  and bone-conduction thresholds 52–3, *52*
  myringoplasty 140
  technical objectives 132, *132*
minimum behavioural response levels 62–3
mixed hearing loss 53
  pure tone audiometry 42–3
monosomy X (Turner's syndrome) 90, 97
Montgomery vs Lanarkshire 2015 155
MRC National Study of Hearing 13, *13*, 16, *16*, 55–7, *55*, *56*, 100–2, *101*, 159–71, *159–71*
multidisciplinary team (MDT), cochlear implantation 146–7
mumps 96
myringoplasty 137–8
myringotomy 88

N

nasopharyngeal lesions in OME 90
National Institute for Health and Care Excellence (NICE) guidance
  cochlear implantation 72, 147–8
  hearing aid candidacy 121
neonatal hearing screening 81, 82–4, 93, 118–19
neonatal intensive care (NICO) 95
neuroplasticity; AUDITORY 145–7
next generation sequencing 94
noise exposure
  assessment 105
  by occupation type *107*
  estimation from voice levels/distance *107*
  hearing protection 105–6, *106*
  social 107
noise reduction, active 106
noise-induced hearing loss 104–12
  audiometric evaluation 108–10, *109*
  continuous noise 104
  history taking 106–8
  pathophysiology 104
  sudden impulse noise exposure 104–5, 110–11
non-organic hearing loss 45–6, 71
Northwest University Auditory Test 6 (NU-6) 69, *69*

O

objective testing 74–85
occupations, noise exposure levels *107*
OME, *see* otitis media with effusion
ophthalmology assessment in congenital hearing loss 94
ossicles 4, *4*, 5, *6*
ossicular chain 54
  fixed 53, *53*
  pathologies 54–5, *56*
  physiology 54
ossicular chain disruption 15, 21
  diagnosis 21
  management options 138
  tympanometry 77–8, *77*
ossiculoplasty 138–9
otitis externa 17
  otitis media, *see* chronic otitis media; otitis media with effusion
otitis media with effusion (OME) 17–18, *18*, 86
  in adults 90–1, 140
  aetiology 87
  assessment and diagnosis 87
  in children 86–90
    common myths 86
  children with trisomy 21 (Down syndrome) 89
  in cleft palate 89–90
  Complications
    infection and blockage 139–40
  diagnosis *15*
  Eustachian tube dysfunction 86, 87
  follow up after surgery 90
  nasopharyngeal examination 90
  non-surgical management 87–8

prevalence and natural history 87
in primary ciliary dyskinesia 90
surgical management 88–9
in Turner's syndrome 90
tympanometry 77
otoacoustic emissions 78–80
distortion product (DPOAE) 78, *79*
transient evoked (TEOAE) 78, *79*
otosclerosis 27, 55, 138
diagnosis *15*, 20–1, *20*, 138
management options 138
prevalence *17*, 57
radiology false diagnosis rates 55
semicircular canal fenestration 58, *58*
speech testing 72
stapes surgery 138–9, *139*
tympanometry 77–8, *77*
otoscopy 16–21, 27, 48
conductive defects 48
methods 16
middle ear pathology 16
wax (cerumen) 16–17, *17*
ototoxicity 96–7, 102
Otovent balloon 87
outcomes
hearing-specific for non-surgical interventions 155
objective 154
subjective 154
outcomes reporting 152–3
speech audiometry 73

P

paracusis of Willis 72
patient information
hearing aids 173–7
hearing loss 115
Patient Reported Outcome Measures (PROMs) 153
patient 'supporter' 155
Pendred syndrome 9, *9*, *10*, 93, 95–6
perilymph 6
phonemes 68
pinna 2
pitch 1
play audiometry 64, 65, *65*
politzerisation in OME 87
presbycusis, *see* age-related hearing loss
primary auditory cortex 7–8
profound deafness, and cochlear implantation 146–7
pure tone audiometry (PTA) 30–47
assessment of bone-conduction without masking 34
automated screening/monitoring 30
BSE pure-tone audiogram 30–32, *31*
BSA recommended procedure *33*
calibration 33
child testing *66*, 67
manual diagnostic 30
masking techniques 36–40
masking to determine bone-conduction thresholds 37–40, *38–9*
reproducibility of air-conduction thresholds 33
standard diagnostic chart 30–2, *31*
test environment 32–3
threshold determination 33
validity of air-conduction thresholds 33–4

Q

questionnaires
Glasgow benefit inventory 154
Glasgow children's benefit inventory 154
tinnitus 155

R

retinitis pigmentosa in congenital hearing loss 94
retraction pocket, prevention in OME 89
Rinne tuning fork test 24–7, *25*, 28
clinical role 26–7
identification conductive defects 48
validity 25–6, *26*
round window 5–6, *6*, 57–8
Royal College of Surgeons of England (RCSEng), consent guidance *156*
Royal National Institute for Deaf People (RNID) leaflets 115

S

scala media 6, *6*, *7*
scala tympani 5, *6*
scala vestibuli 5, *6*
semicircular canal *6*
dehiscence 58–9, *58–9*
fenestration in otosclerosis 58, *58*
sensorineural hearing loss (SNHL)
age-related 100
asymmetric/unilateral 103, 111
congenital, *see* congenital sensorineural hearing loss
progressive and acquired 95–7
sudden 103–4
sentence lists 69
sign languages 117–20
definition 117
SLC26A4 (pendrin) gene mutations 95–6
socio-economic factors in age-related hearing loss 102
sound field presentation in test environment 63, *63*
sound localisation 2, *3*
soundfield audiogram, cochlear implant 149, *150*
soundproof booth 30
speech audiometry 68
in adults 71, *72*
in children 69–71, *70–1*
cochlear implant candidacy 72–3
drawbacks of 68
hearing aid assessment 71
outcomes reporting 73
presentation of results 69, *69–70*
in specific conditions 71–2
types of speech stimulus 68–9
validity 68
'speech banana' 71, *72*, 146
speech development, and cochlear implantation 146
Speech Intelligibility Rating 146
speech recognition threshold 69

speech-in-noise testing 68, 69
speech-to-text applications 117
spondees 69
stakeholder involvement in outcomes 153
stapedius muscle 5, 104–5
stapes piston prosthesis 139, *139*
stapes surgery 139–40, *139*
subtitles on televisions 117
sudden impulse noise exposure
- audiometric evaluation 111
- ear protection 106, *106*, 110–11
- history taking 110–11

sudden profound hearing loss 150
sudden sensorineural hearing loss (SSNHL) 103–4
superior semicircular canal dehiscence syndrome 58
surgeon, assessing their experience 153
surgical management outcomes
- measures of change in conduction defect 132–3, *132–3*
- measures of damage to hearing 134
- ossiculoplasty 138–9
- patient benefit 134–6, *134–5*
- potential complications 152, *152*
- stapes surgery 139–40, *139*
- technical objectives 132

T

tectorial membrane 7, *7*
telephones and television aids 117
tensor tympani muscle 5
test environment
- adult audiometry 32–3
- child hearing testing 62

third window syndrome 57–9
tinnitus 12, 139
- questionnaires 155
- temporary 108

Tinnitus Functional Index 155
tragal rubbing 22, *22*
transcranial attenuation 14, *14*, 36, *36*
trisomy 21 (Down syndrome) 89, 97
tubing (earmould) 175
- changing *175*

Tullio's syndrome 58
tuning fork tests 24–7, 48
- choice of forks 24–5
- Rinne test 24–7, *25–6*
- sensitivity and specificity 25–6, *26*
- Weber test 27, 48

Turner's syndrome (monosomy X) 90, 97
tympanic membrane 3–4, *4*
- chalk patches 53
- movement 4–5, *5*
- otoscopy 16
- structure 4, *5*
- tympanosclerotic plaque 53, *53*

tympanic membrane perforation 53–4, *54*
- management options 140
- myringoplasty 140
- tympanometry 75, *76*

tympanometry 74–8
- advances in equipment 75, 77
- classification of tympanograms 75, *76*
- interpretation in OME 77
- interpretation in otosclerosis/ossicular chain discontinuity 77–8, *77*

U

UK Association of Teachers of Lipreading to Adults 115
UK National Study of Hearing *see* MRC National Study of Hearing 55–6, *56*
Usher syndrome 94

V

Valsalva manoeuvre 87
ventilation tubes (VTs) 88–9
- choice of 89
- common myths 86
- insertion in adult OME 90–1

vertigo and hearing loss 58–9
vestibular schwannoma 103
- diagnosis *15*
- speech testing 72

visual reinforcement audiometry (VRA) 63–4, *64*, *66*
voice tests
- criticism 23
- free-field 22–4, *24*
- screening with whispered voice *21*, 23–4, *24*

W

warble tone in testing children 63
wax (cerumen) 27
- diagnosis *15*
- otoscopy 16–17, *17*

Weber tuning fork test 27, 48
whispered voice screening *21*, 23–4, *24*
wideband acoustic immitance 77
wireless microphones 116
word lists 69
word recognition scores 69, *69*
workplace
- facilities for workers with hearing impairment 117
- noise exposure levels *107*
- regulation noise levels *107*
- support to workers with hearing impairment 117
- World Health Organization (WHO), classification of hearing impairments 13, *13*, *35*