



The essentials of managing Hearing loss

Key points

- Hearing loss is very common in the community. Mild-to-moderate hearing loss affects one in five adults by the age of 50 years.
- Moderate-to-severe hearing loss has a measurable impact on quality of life, employability and, in the elderly, independent living and cognitive decline.
- Hearing loss is best classified as conductive, sensory neural or mixed. This simple classification guides the clinical evaluation, range of available treatments and response to treatment.
- History, otoscopy and tuning fork tests allow most causes of hearing loss to be diagnosed, allowing a management plan to be formulated.
- Surgical treatment for conductive hearing loss is generally successful. Cochlear implants are the treatment of choice for severe sensory neural hearing loss.

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Hearing loss is a common condition that can have a great impact on a patient's quality of life. Hearing aids are an effective treatment for those with mild-to-moderate hearing loss and surgical options may be available for those with more severe loss.

Hearing, as one of the five senses, assumes an important role at all stages of life. In early childhood, near-normal hearing in at least one ear is important for speech and language development. In adulthood, advanced learning and employment capacity as well as, in later years, quality of life, independent living and preventing cognitive decline are highly dependent on hearing.¹

Although there are many causes, types of hearing loss can be conveniently classified as conductive or sensory neural depending on which part of the hearing pathway is disrupted. Using patient history, otoscopy and simple tuning fork tests, the nature of the hearing loss can be usually determined in the office setting and an appropriate management pathway devised. Effective treatments for many kinds of hearing loss now exist, including medical treatments, surgical interventions and cochlear implants for severe hearing loss. In addition, hearing aids have become smaller and more cosmetically acceptable than in the past.

HOW COMMON IS HEARING LOSS?

Hearing loss is very common in the Australian adult community.² In Australia, it has a prevalence of 17% in 50-year-olds, 48% in 60-year-olds and 64% in 70-year-olds. It is more common in men at all ages. Most of the affected population has a mild-to-moderate degree of hearing loss that is highly suitable for assistance with a hearing aid (Figure 1).

In adults of employable age, the impact of hearing loss on potential employment, quality of life and general function is much higher than has been commonly realised, and is equivalent to the disability burden of many health conditions recognised as national health priorities. For example, in terms of disability burden, mild hearing loss is comparable with mild asthma, moderate hearing loss is comparable with severe pain related to degenerative spinal disease and severe hearing loss is comparable with severe diabetes associated with visual failure.³

Severe hearing loss (greater than 70 dB)

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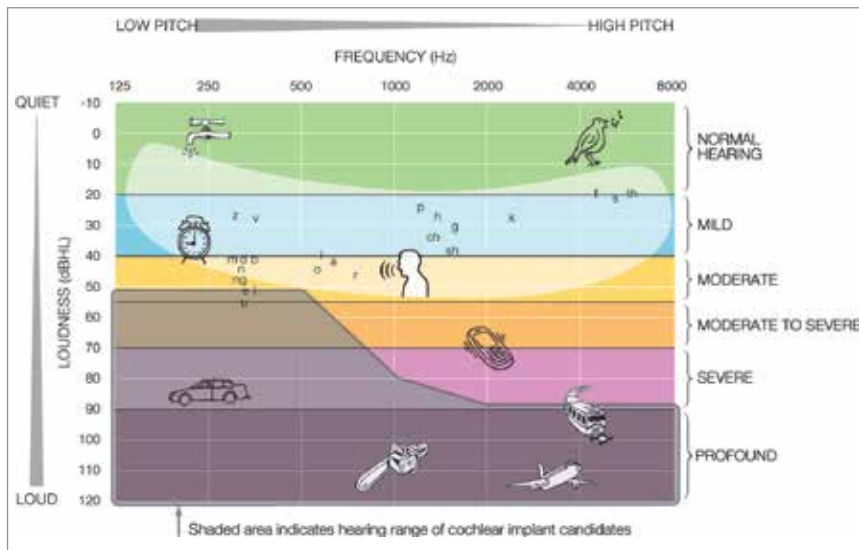


Figure 1. A schematic diagram of our hearing environment. Patients with extreme hearing losses (in the severe-to-profound range) generally fulfil the indications for cochlear implantation.

Courtesy of Cochlear Ltd.

affects a smaller proportion of the community, but carries a greater disability burden. In the elderly, hearing loss is often combined with visual failure and cognitive decline. The impact of this combination of sensory losses on quality of life and independent living can be profound.

MECHANISMS OF HEARING: THE EAR AND AUDITORY PATHWAYS

Normal hearing requires an anatomically intact and functioning auditory pathway. The pinna and external auditory canal capture sound, focusing it on the ear

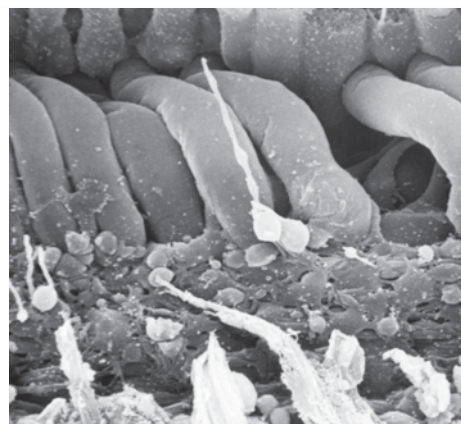
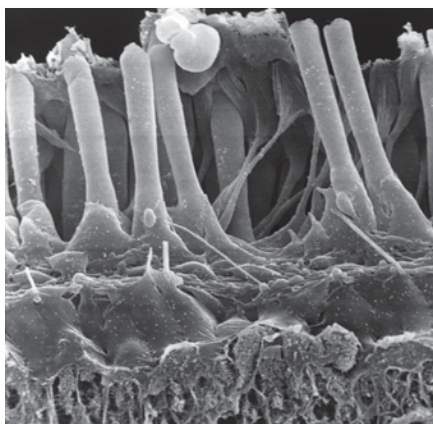
drum. This sound energy then vibrates the drum and attached middle ear bones (conductive pathways) mechanically transmitting it to the inner ear. Complex fluid and structural factors within the organ of Corti allow the vibration of sound to activate hair cells along the basilar membrane (Figures 2a and b) leading to action potentials in adjacent auditory nerve fibres (neural pathways). The electrical signals are then conducted through central pathways and networks in the brainstem and midbrain to the auditory cortex in the temporal lobe, which

recognises sound as being meaningful (higher pathways). Disturbance of any part of this auditory pathway will result in hearing loss or distortion of normal hearing (Box 1 and Figure 3).⁴

HOW WE ACQUIRE HEARING LOSS

Disruptions to the conductive hearing mechanisms (e.g. external ear canal occlusion or middle ear disruptions caused by otitis media, perforations or otosclerosis) produces hearing losses that are mild-to-moderate in degree. Such hearing losses respond well to treatment with surgery or hearing aids. In contrast, problems affecting the sensory mechanisms in the inner ear (e.g. caused by ageing, genetics, Meniere’s disease, trauma, meningitis or ototoxicity) may produce severe-to-profound hearing losses. Severe hearing loss (>70 dB) responds poorly to hearing aids and is associated with a significant reduction in quality of life.

Most causes of acquired hearing loss involve the sensorineural parts of the hearing pathway, and hearing loss occurs because of hair cell lesions within the organ of Corti in the inner ear (Figures 2a and b). The hair cells are vulnerable to a variety of toxicities. In some cases of profound hearing loss, there may be a single identifiable causative agent that leads to hair cell loss and subsequent deafness, such as exposure to ototoxic agents (e.g. gentamicin or cisplatin) or following inflammation caused by meningitis. More often there are several



Figures 2a and b. Hair cells in the organ of Corti. (a, left). The normal hearing ear has orderly arrangements of hair cells. (b, right). In most cases of sensory neural hearing loss, the hair cells are absent or in disarray, leading to poor transduction of sound-induced vibration.

© FIGURES 2A AND B: DR GORAN BREDBERG/SPL

1. THE EAR AND HEARING: THE ROLES OF THE CONDUCTIVE AND SENSORINEURAL PATHWAYS

The most useful way of approaching sound reception is to consider two pathways, as shown in Figure 3:

- the conductive pathway (outer and middle ear), and
- the sensorineural pathway (inner ear, neural pathways and auditory cortex).

The conductive pathway

The external ear

The major function of the external ear (pinna and external auditory canal) is to conduct sound to the eardrum and ossicular chain.

The curved shape of the ear canal provides protection of the tympanic membrane and bony ear canal; wax protects and moisturises the canal skin. The healthy ear is self-cleansing: squamous epithelium migrates from the tympanic membrane to the outer ear canal, carrying dead skin and wax.

The middle ear

The middle ear is made up of the tympanic membrane, ossicular chain and eustachian tube. The tympanic membrane is normally silver-grey or translucent, and shaped as a curved cone that acts as a loud speaker.

The malleus, incus and stapes mechanically amplify sound from the tympanic membrane to the stapes footplate. The area of the tympanic membrane is markedly greater than the area of the stapes footplate; this factor, in combination with the lever action of the malleus and incus, amplifies sound.

The sensorineural pathway

The inner ear

The inner ear consists of the cochlea and vestibular apparatus. The cochlea, which is shaped like a snail-shell, contains hair cells within the organ of Corti. Sound pressure waves within the endolymph move the hair cells and produce action potentials.

The vestibular system is made up of the semicircular canals, utricle and saccule. Fluid movements in the semicircular canals

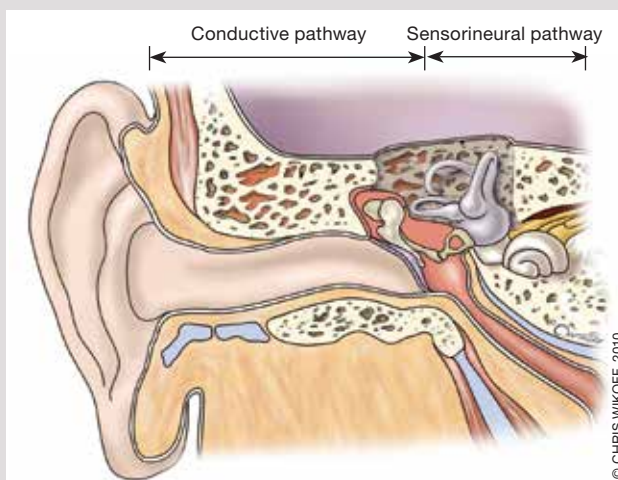


Figure 3. Causes of hearing loss may involve either the conductive or the sensorineural pathway. The external and middle ear make up the conductive pathway; the inner ear and central pathways make up the sensorineural pathway.

also cause hair cell changes and produce action potentials. The semicircular canals detect rotational movement, whereas the utricle and saccule monitor gravity and detect acceleration and deceleration.

The central pathways

The acoustic and vestibular nerves run together, through the internal acoustic meatus traversing the cerebellopontine angle, to nuclei in the brainstem. The perception of sound is relayed to the auditory cortex of the parietal lobe, where it is analysed and processed.

Adapted with permission from Atlas MD, Lowinger DSG. The GP's essential guide to hearing loss. *Med Today* 2000; 1 (6): 48-59.⁴

factors that contribute to hair cell loss and profound hearing loss or, alternatively, the deafening aetiology is unknown. One common pattern of hearing loss is the effect of ageing (presbycusis) added to a pre-existing, non-ageing factor of hearing loss, such as noise exposure, with a background genetic susceptibility leading to early hair cell dysfunction.

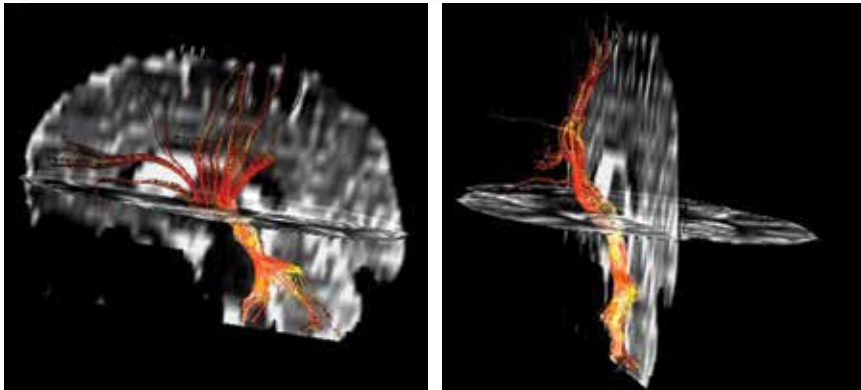
Sustained stimulation of the higher neural pathways linking the inner ear and the auditory cortex is necessary for the

development of normal sound and speech perception (Figures 4a and b). Hearing loss that occurs very early in life (usually genetically determined or as a result of in utero infections or prematurity) is associated with poor maturation of these neural pathways, particularly at a cortical level. Hearing loss acquired before the acquisition of cortical speech perception (prelingual) is usually associated with disordered higher pathways and is characterised by abnormal speech quality, which in extreme cases can be completely

unintelligible. Hearing loss acquired later in life after the acquisition of speech (postlingual) is usually associated with well-formed higher pathways, and is characterised clinically by intelligible speech quality.

CAUSES OF HEARING LOSS

Hearing loss can be broadly classified on an anatomical basis into disturbances of the conductive or sensorineural hearing pathway (Boxes 1 and 2). Mixed hearing loss involves both pathways.



Figures 4a to b. Highly processed MRI images highlighting the higher auditory pathways in a patient with normal hearing. The pathway commences with the auditory nuclei in the brainstem, ascends through the midbrain and terminates in the temporal cortex; a few fibres project to the frontal association area.

2. COMMON CAUSES OF HEARING LOSS

**Conductive
Outer ear**

- External auditory canal occlusion (wax, otitis externa, polyp, tumour)
- Stenosis (congenital, exostosis, trauma)

Middle ear

- Tympanic membrane perforations
- Cholesteatoma
- Trauma (instrumentation, skull fracture, barotrauma)
- Ossicular chain dysfunction (congenital, trauma, otosclerosis)

**Sensory neural
Inner ear**

- Infections (e.g. herpes zoster, herpes simplex)
- Presbycusis
- Noise exposure
- Genetics
- Ménière’s disease
- Ototoxic medications

Higher pathways

- Cerebellopontine angle tumours (acoustic neuroma, meningioma)
- Cerebral lesions (tumour, multiple sclerosis)

Common causes of conductive hearing loss involve blockage of the external auditory canal by wax, otitis externa, foreign bodies, polyps or stenosis, either congenital or due to exostosis. Conductive hearing loss occurs with all disruptions of the middle ear mechanism, including perforations, otitis media, cholesteatoma, ossicular chain fractures and otosclerosis. Changes in eustachian tube function with fluctuation in middle ear pressures may produce mild fluctuating hearing loss.



Figure 5. Congenital atresia. The ear canal is completely occluded, but the middle and inner ear are well formed making the hearing loss suitable for correction by the wearing of a bone conduction hearing aid.

Sensory neural hearing loss is caused by damage to the inner ear hair cells, auditory nerve or higher pathways. Common causes are ageing, noise damage, genetic hearing loss, acoustic neuroma, Ménière’s disease, temporal bone fractures or the effect of ototoxic medications. Rarely, congenital sensory neural hearing loss is caused by malformation of the inner ear or absence of the cochlear nerve.

Causes of conductive hearing loss

Ear canal stenosis. Bony or soft tissue obstruction of the ear canal may be congenital or the result of trauma, chronic infection or surgery (Figure 5).

Exostosis and osteomas are bony growths in the ear canal either occurring spontaneously or as a result of repeated exposure to cold water resulting in restriction to the ear canal with further trapping of debris and water (Figure 6). When the middle and inner ear function is normal, the resulting hearing loss and recurrent infections can be corrected by removal of the bony stenosis (canalplasty), restoring the ear canal lumen to a normal diameter and function.

Otitis media. All forms of otitis media are associated with middle ear effusion

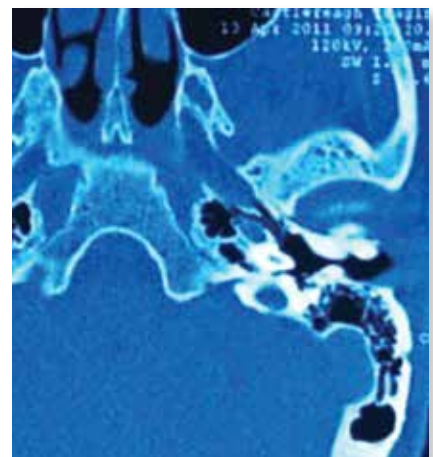


Figure 6. CAT scan showing exostoses and occlusion of the canal. Surgical widening of the ear canal is very effective in improving the hearing loss and infection control.



Figure 7. Middle ear effusions associated with otitis media. This commonly produces a conductive hearing loss by altering the resonance of the middle ear mechanism.

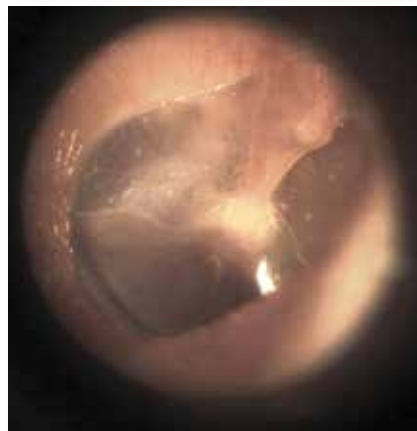


Figure 8. Congenitally high-riding jugular bulb. This middle ear mass produces a conductive type hearing loss due to splinting of the ear drum impeding the passage of sound-induced vibration through the middle ear.



Figure 9. A safe central perforation of the tympanic membrane. Hearing loss is due to loss of tympanic membrane surface area and scarring of middle ear ossicles as a result of repeated infections. Tympanoplasty surgery is often effective in restoring hearing.

(Figure 7). The fluid-filled middle ear transmits sound poorly, resulting in conductive hearing loss. The hearing can be restored by draining the middle ear effusion by medical means or insertion of a grommet. Often the otitis media resolves spontaneously. Occasionally a middle ear tumour or mass, such as a high-riding jugular bulb, may be confused with otitis media because these produce similar otoscopic appearances and conductive hearing loss (Figure 8).

Tympanic membrane perforations. It is important to distinguish between safe (central) and unsafe (peripheral) perforations. Safe perforations are most commonly the result of middle ear disease, trauma or barotrauma (Figure 9). Unsafe perforations occur in the epitympanum (posterior part of the tympanic membrane) and indicate cholesteatoma requiring surgical treatment. Both types of

perforations produce hearing loss by reducing the tympanic membrane surface area, and are often associated with dysfunction of the middle ear bones.

Cholesteatoma. Cholesteatoma is an ingrowth of keratinising squamous epithelium that usually arises from a marginal or epitympanic perforation of the tympanic membrane. The keratinising sac has the potential to expand and destroy the middle and inner ear structures, producing both conductive and sensory neural hearing loss. Further destruction of the bony coverings of the facial nerve and dura result in an increased risk of facial paralysis, meningitis and brain abscess. Surgical treatment minimises the risk of complications developing.

Trauma, skull fractures and barotrauma.

There are multiple traumatic causes of hearing loss. Slapping, blast or syringing injuries tend to cause a perforation or tear in the tympanic membrane. Many of these tears will close spontaneously within a month or two if infection is prevented by taking precautions to avoid water exposure. Fractures of the temporal bone cause bleeding into the middle ear (haemotympanum) or ossicular disruption causing a conductive hearing loss. Fractures involving the inner ear cause severe sensory neural hearing loss and vertigo and are often associated with facial nerve damage, cerebrospinal fluid leakage and brain injury (Figure 10).

Barotrauma from air pressure changes while flying or diving, usually with poor eustachian tube function, can cause middle ear effusion or haemorrhage resulting in conductive hearing loss. Rupture of inner ear membranes results in a perilymph fistula and sensory neural hearing loss.

Ossicular chain dysfunction and otosclerosis. Conductive hearing loss with a normal appearance of the tympanic membrane is likely to be due to disrupted function of the middle ear

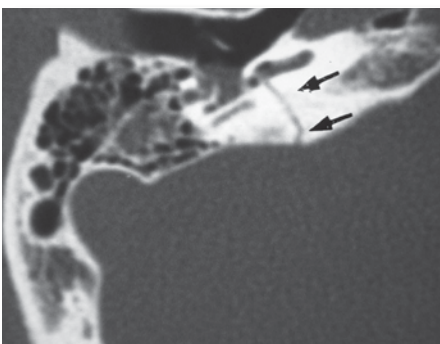


Figure 10. Extreme trauma to the temporal bone can result in a fracture through the cochlea (arrows) producing a severe sensory neural hearing loss.

© FIG 7: PROF TONY WRIGHT, INSTITUTE OF LARYNGOLOGY & OTOTOLOGY/SPL; FIG 9: CC/ISW/SPL

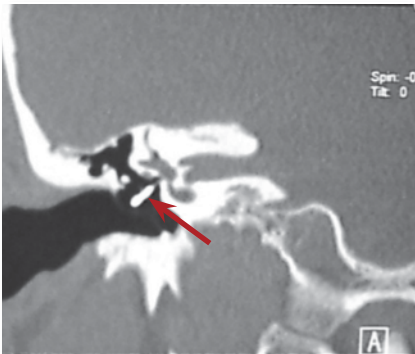


Figure 11. Stapedectomy surgery replaces part of the stapes bone with a prosthetic piston (arrow), allowing sound vibration to be transmitted to the inner ear and effectively reversing the hearing loss.

ossicles. This is most commonly due to otosclerosis, a hereditary disease in which the stapes footplate becomes increasingly stiff due to the formation of new abnormal bone. Most cases present in early to mid-adulthood, and often during pregnancy (when the high oestrogen levels accelerate the otosclerosis). Treatment with stapedectomy surgery, where part of the stapes bone is replaced by a prosthesis, is usually an effective treatment (Figure 11). Congenital fixation of the ossicles or disruption to the ossicular chain due to trauma, infection or surgery produces similar kinds of conductive hearing loss, which can be surgically improved.

Causes of sensory neural hearing loss

Presbycusis. Presbycusis is caused by age-related degeneration of the cochlear hair cells (Figure 2b). The hearing loss is usually bilateral, symmetrical, high frequency, progressive and associated with tinnitus. Speech discrimination is poor, especially in background noise.

Noise damage. Noise damage to the cochlea results from sustained exposure to high-intensity ‘industrial’ noise. Damage to the hair cells of the inner ear typically produces a hearing loss

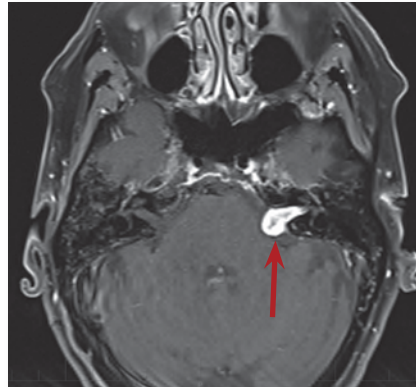


Figure 12. A contrast-enhanced MRI scan showing a small acoustic neuroma (arrow) in a patient who presented with low-tone hearing loss and a mild disturbance of balance.

maximum at 4000 Hz (Figure 1).

Viral infections. Reactivation of herpes zoster virus infection can present as Ramsay Hunt syndrome with facial nerve palsy, vesicles in the ear canal and sensory neural hearing loss. Other viral infections (e.g. herpes simplex, rubella and measles) are associated with damage to the cochlear hair cells producing a corresponding hearing loss.

Genetics. Familial hearing loss is commonly of genetic origin. It is characterised by progressive mid-tone sensory neural hearing changes, which occasionally progresses to profound hearing loss over many decades. Connexion 26, a protein that deals with ion transport within the cochlea, is abnormal in many cases.

Ménière’s disease. Ménière’s disease is a relatively common cause of unilateral fluctuating hearing loss associated with episodic disabling vertigo, tinnitus and fullness in the ear.

Ototoxic medications. Many prescribed medications have the potential to cause hearing loss. The most common groups are aminoglycoside antibiotics, salicylates, some diuretics and cytotoxic agents. Some topical ear drops have ototoxic potential in the presence of tympanic membrane perforations.

Neuromas. Acoustic neuroma is a benign

tumour arising from the Schwann cells for the vestibular nerve (Figure 12). The tumour presents with progressive unilateral hearing loss, tinnitus and mild imbalance. Microsurgical removal of the tumour is highly successful, but hearing can only occasionally be preserved. Tumours usually grow very slowly. Many small tumours are managed by observation and repeated MRI scanning each year.

Sudden sensory neural hearing loss syndrome

Sudden decline in hearing in one ear, or more rarely both ears, is a medical emergency that needs urgent referral of the patient for treatment. Usually the hearing declines from normal to severe levels over a period of hours and can be associated with vertigo and tinnitus.

An underlying cause is not usually found in unilateral cases. In cases of bilateral sudden hearing loss, a systemic autoimmune cause is sometimes found.

Recommended first-line treatment is a course of high-dose corticosteroids (e.g. prednisone 1 mg/kg for 10 to 14 days). Intra-tympanic administration of a corticosteroid (e.g. dexamethasone) is a second-line treatment that delivers a higher dose of corticosteroid to the inner ear.

CLINICAL EVALUATION OF HEARING LOSS

A simple history and examination of the ear, accompanied by tuning fork tests, will allow most hearing losses to be classified as conductive or sensory neural. The historical time course of the hearing loss and associated otological features such as discharge, bleeding, pain, vertigo, tinnitus or cranial nerve signs, allow the underlying cause of the hearing loss to be determined.

Visual inspection of the external ear, canal and drum allow many causes of conductive hearing loss to be diagnosed. An open ear canal with no element of occluding mass excludes causes of conductive hearing loss such as wax impaction, foreign bodies, tumours,

3. USING TUNING FORK TESTS TO EVALUATE HEARING LOSS

The Weber and Rinne tests are the tests most commonly used in the office setting to assess hearing loss. Results from the two tests can be used to determine the presence and severity of hearing loss.

The Weber test

The Weber test is used to determine whether a patient has symmetrical hearing. It is performed using a 512 Hz tuning fork. Place the vibrating tuning fork in firm contact with the midline of the forehead and then on the vertex of the skull (Figure 13a).

A patient who has:

- normal hearing – will hear the sound equally in both ears, ‘in the middle’ or ‘all over’
- a conductive hearing loss – will refer the sound to the affected ear
- a sensory neural hearing loss – will refer the sound to the unaffected ear.



Figure 13a. To test the symmetry of hearing between the two ears, the tuning fork is placed firmly on the forehead and then on the vertex of the skull.

The Rinne test

The Rinne test is used to compare air and bone conduction. The 512 Hz tuning fork is the most useful fork to use. Bone conduction is assessed first.

Place the fork firmly over the mastoid bone (Figure 13b). Tell the patient to listen to the sound, not to the vibration. When the patient claims not to hear the sound any longer, assess air conduction by quickly moving the fork and placing it 2 to 3 cm from the meatus of the external canal, with the tines of the fork parallel to the plane of the canal (Figure 13c).

If air conduction is heard longer than bone conduction (i.e. a Rinne positive result), the patient has either normal hearing or a sensory neural hearing loss. If bone conduction lasts longer than air conduction (i.e. a Rinne negative result), a conductive hearing loss is present.

Rinne test for bone and air conduction



Figure 13b. To assess bone conduction, the tuning fork is placed firmly over the mastoid bone.



Figure 13c. To assess air conduction, the fork is then placed 2 to 3 cm from the external canal, with the tines of the fork parallel to the plane of the canal.

Interpreting the tests

Results from the Weber and Rinne tests can be used to distinguish between conductive and sensory neural types of hearing loss. This is described in Figures 13d to f.

Combining results from the Weber and Rinne tests



Figure 13d. Symmetrical Weber test and positive Rinne test in each ear. Hearing is referred equally to each ear in the Weber test with positive Rinne results in both ears indicate symmetrical hearing with normal middle ear function.

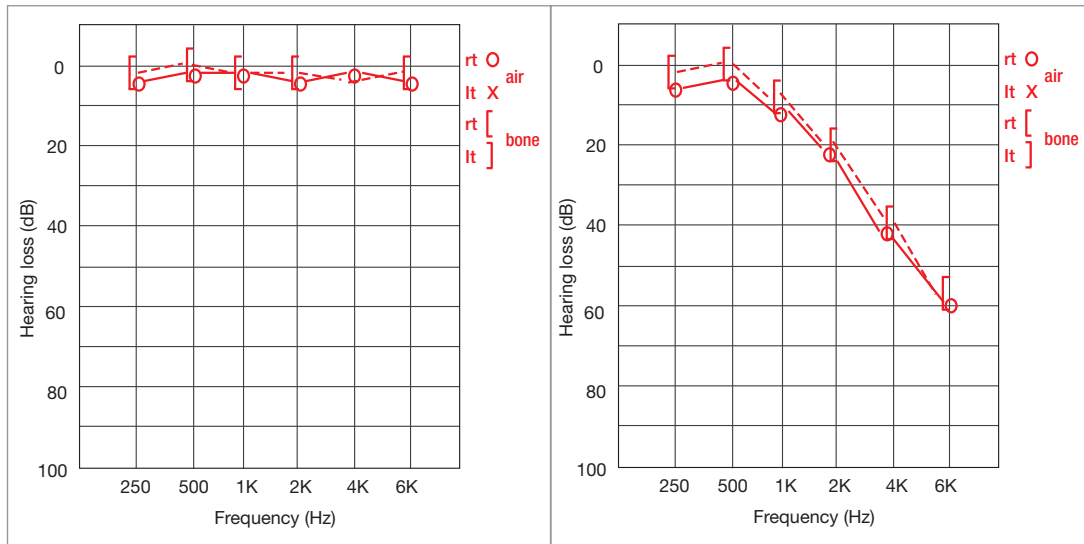


Figure 13e. Asymmetrical Weber test and unequal Rinne tests. The patient complains of right-sided hearing loss. Hearing is referred to the left ear in the Weber test, with the Rinne test negative for the right ear and positive for the left ear, indicating conductive deafness in the right ear.



Figure 13f. Asymmetrical Weber test and positive Rinne test in each ear. The patient complains of right-sided hearing loss. Hearing is referred to the left ear in the Weber test with a positive Rinne result in each ear, indicating sensory neural deafness in the right ear.

Adapted with permission from Atlas MD, Lowinger DSG. Office evaluation of hearing loss in adults. Med Today 2000; 1 (6): 117-119.⁵



Figures 14a and b. Audiometry. (a, left). Normal hearing audiogram. (b, right). Presbycusis audiogram – note the bilateral high-tone sloping hearing loss; hearing in background noise is a major problem.

exostosis and stenosis. Visualisation of a healthy intact drum that is mobile on pneumatic insufflation also excludes perforations, cholesteatoma, otitis media and middle ear effusions as causes of conductive hearing loss. Otosclerosis due to fixation of the stapes bone is a common cause of progressive conductive hearing loss where the drum appearance and mobility is normal.

In patients with sensory neural hearing loss the otoscopic examination is generally normal but the tuning fork tests indicate that the hearing changes are due to disruption of the cochlea and higher pathways. Further diagnosis of inner ear hearing losses is dependent on history, results of audiometry and imaging studies.

Tuning fork tests

The Weber and Rinne tests are most commonly used to assess hearing loss. These simple office tests are performed with a 512 Hz tuning fork by comparing symmetry in hearing between the two ears (Weber test), and the efficiency of hearing through normal-hearing middle ear mechanisms (air conduction) and direct stimulation of the cochlea through the skull bones (bone conduction; Rinne test). The technique in using the tuning fork is important and improves with practice (Box 3 and Figures 13a to f).⁵ Particular patterns of patient responses can be detected for conductive hearing losses, sensory neural hearing loss and a symmetrical hearing.

Audiometry

A pure-tone audiogram and tympanometry are part of a battery of tests that are performed by audiologists. These tests allow quantification of various aspects of a patient’s hearing and balance functions including:

- pure-tone thresholds
- speech discrimination
- middle ear function (tympanometry)
- inner ear function (otoacoustic studies).

In audiometry, air conduction thresholds are obtained using headphones and bone conduction thresholds are obtained by directly stimulating the cochlea using a vibratory stimulator that is placed on the mastoid. Conductive hearing losses

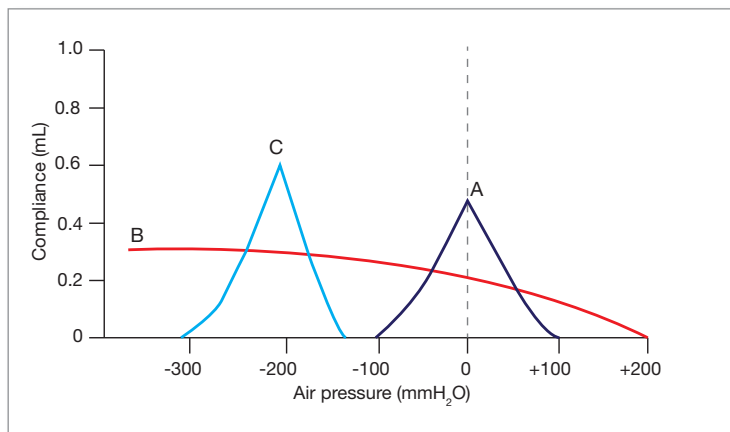


Figure 15. Tympanometry. The middle ear is most efficient in transmitting sound when the middle ear and external canal air pressures are the same (trace A). In negative middle ear pressure (the middle ear pressure is lower than the external canal pressure; trace C), most commonly seen in eustachian tube dysfunction, the sound transmission is reduced, leading to a sensation of blocked or muffled hearing. In middle ear effusion, the tympanogram is flat (trace B), sound transmission is reduced and a conductive hearing loss is suggested.

are evidenced by normal bone conduction but a reduction of air conduction thresholds. The difference between the air and bone conduction thresholds is the degree of conductive hearing loss. In sensory neural hearing loss, loss of cochlear nerve function causes both air and bone conduction thresholds to be reduced (Figures 14a and b).

Tympanometry assesses middle ear function by measuring changes in middle ear impedance at varying external canal air pressures. The various compliance curves that can be produced suggest different types of middle ear pathology including middle ear effusion, perforations or eustachian tube dysfunction (Figure 15).

TREATMENT OF HEARING LOSS

Hearing loss is a symptom that indicates an abnormality of the sound conducting mechanisms of the outer, middle or inner ear, the auditory nerve and/or higher pathways. Specific treatments for the underlying disease process often produce an improvement in hearing thresholds (Table 1). When the hearing loss is disabling, a specific medical treatment in some cases of infective or inflammatory disease can improve hearing. Many causes of conductive hearing loss respond to

TABLE 1. SURGICAL TREATMENT OF HEARING LOSS

Cause of hearing loss	Treatment
External canal	
Atresia or stenosis	Canalplasty or surgically widening the bony and cartilagenous canal
Exostosis	Drilling out the bony exotosis
Middle ear	
Tympanic membrane perforation	Tympanoplasty surgery to graft the tympanic membrane perforation
Middle ear effusions	Medical treatment with antibiotics, grommet insertion
Ossicular chain dysfunction: fracture, erosion, discontinuity	Replacement of the dysfunctional ossicle with a prosthesis
Otosclerosis	Replacement of part of the stapes bone with a prosthetic piston (stapedectomy)
Cholesteatoma	Mastoid surgery to remove the cholesteatoma and reconstruct the middle ear mechanism
Inner ear	
Acoustic neuroma	Microsurgical removal of the tumour. Hearing can sometimes be preserved
Severe to profound hearing loss (multiple causes)	Cochlear implant

TABLE 2. HEARING LOSS AND ASSOCIATED SYMPTOMS REQUIRING URGENT SPECIALIST REFERRAL AND ASSESSMENT

Severe symptoms	Assessment required
Sudden sensory neural hearing loss	A medical emergency requiring urgent assessment. Recommended first-line treatment is prednisone 1 mg/kg for 10 to 14 days
Asymmetrical sensory neural hearing loss	Can be a presentation of cerebellopontine tumour or acoustic neuroma. Needs further investigation with MRI scan
Associated cranial nerve deficits, most commonly facial nerve palsy	Can be due to an invasive inflammatory or neoplastic process
Ear canal or middle ear mass	Can be an indication of ear canal cancer or middle ear neoplasm (e.g. glomus tumour) or high-riding jugular bulb
Persistent deep ear pain	May indicate an invasive inflammatory or neoplastic process involving the temporal bone structures
Discharging ear	May indicate an invasive inflammatory or neoplastic process involving the temporal bone structures



Figure 16. A cochlear implant is the treatment of choice for severe hearing loss.

Courtesy of Cochlear Ltd.

surgical treatment. When no specific treatment is available, then a carefully fitted hearing aid can minimise the hearing disability.

Most cases of hearing loss are slowly progressive with no urgency for specialist referral or assessment. However, several presentations of hearing loss do require urgent referral (within days to a few weeks) because they may indicate more severe underlying diseases. These are listed in Table 2.

Hearing aids

Hearing aids can be effective both in patients with conductive and sensory neural hearing loss. They consist of a microphone, amplifier and speaker. Some models have multiple programmable elements, allowing for listening in varied environments. Hearing aids come in a variety of sizes and designs, but are increasingly becoming smaller and have minimal cosmetic impact on patients. Some types of hearing aids are partially implantable. There are no specific rules in selecting patients who will benefit from a hearing aid but the important feature is that the functional characteristics of the aid must match the hearing loss.

Surgery

Modern ear surgery is highly evolved, and much can be done for patients with many types of conductive hearing losses to improve hearing thresholds. Stapedectomy surgery for patients with otosclerotic hearing loss can produce sustained improvements in hearing, often at near normal thresholds.

Replacement of damaged middle ear ossicles with prosthetic replacements can also improve hearing thresholds significantly. Canalplasty, to correct external canal stenosis, and tympanoplasty, to repair perforations, often improves hearing as well as reducing infection rates.

A cochlear implant (the bionic ear; Figure 16) is an electronic device that is implanted within the cochlea to allow direct electrical stimulation of the auditory nerve endings. The surgery for cochlear implantation has now become routine.

The outcomes in postlingually deafened adults give greatly improved hearing, often sufficient to provide free communication over the telephone. In congenitally deafened children, normal speech and language development with access to mainstream educational opportunities is the common outcome.

ROLE OF THE GP

The role of the GP is to recognise that hearing loss is common in the community, and that its impact on patients and their families can be significant. Initial management consists of determining the nature of the hearing loss using clinical assessment and simple tuning fork tests, arranging audiometry to quantify the hearing loss and imaging to exclude possible conditions requiring treatment.

There is usually no urgency for specialist referral or assessment in most cases of hearing loss. More severe underlying diseases may be indicated by several presentations of hearing loss. These patients require urgent referral. **MT**

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COMPETING INTERESTS: None.

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