# Hearing loss

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## ABSTRACT

Hearing loss affects one in six people in the UK and is a significant disease burden. In addition to communication problems, there is also an association with depression and dementia. Clinical assessment with targeted history and examination can identify the characteristics and cause of hearing loss, and complementary audiological testing can confirm its type and severity. Retrocochlear screening is recommended for sudden, rapidly progressive or asymmetric sensorineural hearing loss. Medical or surgical therapies may be indicated in cases of conductive hearing loss, while hearing assistive devices and hearing aids are the mainstay of rehabilitation for sensorineural hearing loss.

## INTRODUCTION

According to the Global Burden of Disease Study, hearing loss is the fourth leading cause of disability around the world.[1] In the UK, hearing loss affects 11 million people, or 1 in 6, with 900 000 of them having severe or profound hearing loss.[2] The prevalence of hearing loss increases sharply with age, affecting 42% of people aged over 50 years, rising to 71% of people aged over 70 years.[3]

Hearing loss can lead to loss of productivity, impaired communication, social withdrawal and reduced quality of life. Furthermore, as compared with age-matched adults with unimpaired hearing, older persons with hearing loss have higher rates of depression, hospitalisation, falls and death and hearing loss can increase the risk of dementia by up to five times[3]. The use of hearing aids may reduce these risks. Screening for adult hearing loss is currently not routine, so it is essential that all clinicians increase their awareness in this field to enable early, appropriate care.

## ANATOMY AND PHYSIOLOGY OF HEARING

The function of the ear is to conduct and convert sound, in the form of mechanical vibration, into an electric signal that is processed by central auditory pathways. The neuroanatomy of the peripheral and central auditory systems is organised to extract specific information from the complex waveforms of speech and music.

### The external and middle ear

The pinna, as a part of the external ear, collects sound and directs it into the external auditory canal. It is angled so that it catches sounds more from in front than behind and so contributes to localisation of sound at higher frequencies through interaural differences in sound amplitude. In lower frequencies, sound localisation depends more on the interaural time difference.

The middle ear is an air-filled space, separated from the external auditory canal by the conical-shaped tympanic membrane. The middle ear contains the ossicles (malleus, incus and stapes), which conduct sound from the tympanic membrane to the inner ear. Amplification is provided by the relative difference in size of the surface area of the tympanic membrane and the stapes footplate and by the ossicular chain acting as a system of mechanical levers. The stapes footplate abuts the oval window membrane of the vestibule at the point of communication between middle and inner ears.

### The inner ear

The inner ear transduces the mechanical energy of sound into an electrical signal that stimulates the auditory nerve and also produces a frequency (pitch) and intensity (volume) analysis of the sound. The osseous cochlea within the petrous apex of the temporal bone houses the membranous cochlear duct; its core is the modiolus, which allows the passage of auditory nerve fibres from the internal auditory meatus to the hair cell synapse. The cochlea is divided into three compartments in cross-section. The upper chamber (scala vestibuli) and lower chamber (scala tympani) contain perilymph; the scala media contains endolymph and is separated from the scala tympani by the basilar membrane.



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The organ of Corti is the principal sensory structure of the inner ear and runs along the length of the basilar membrane. It contains four rows of receptor hair cells, together with supporting cells—a single row of medial inner hair cells and three rows of lateral outer hair cells. Vibration of the stapes footplate in response to sound generates a compressional wave in the perilymph that causes vibration of the basilar membrane and organ of Corti. This in turn leads to movement of the hair cells and deflection of their stereocilia, triggering stretch-sensitive cationic channels to open and causing hair cell depolarisation. The afferent innervation of cochlear hair cells consists of 30 000 auditory nerve fibres, with their cell bodies lying in the spiral ganglion within the modiolus.

To respond to variable frequencies of sound, the cochlea has a tonotopic representation, where the basilar membrane responds at each point to a characteristic frequency—the highest frequencies at the basal end nearest the oval window and progressively lower frequencies towards the apical end. The travelling wave generated by sound reaches its corresponding place of resonance along the membrane and travels no further. The response to intensity of sound is determined by the action of the outer hair cells, which contract to amplify the travelling wave at the point of maximal stimulation.

#### Central auditory pathways

The central auditory pathways involve all neuronal projections between the cochlear nerve, brainstem, midbrain, thalamus and cerebral cortex. The tonotopic organisation of the cochlea is reiterated throughout each component of the central auditory system.

1. The cochlear nerve is a trunk of the vestibulocochlear nerve, containing afferent fibres from the cochlear hair cells. The nerve travels through the internal auditory canal and cerebellopontine angle to reach the cochlear nucleus.
2. The cochlear nucleus lies in the pontomedullary junction of the dorsolateral brainstem and is divided into ventral and dorsal subdivisions. The dorsal nucleus probably contributes to sound localisation and is implicated pathologically in tinnitus. The ventral nucleus is responsible for encoding sound frequency, spectral shape and intensity. The cochlear nucleus neurones project to other brainstem regions, including the contralateral cochlear nucleus and inferior colliculus, via acoustic stria.
3. The superior olivary complex lies in the caudal aspect of the pons and is the first central auditory centre to receive binaural innervation. Its ascending pathways have a major role in sound localisation, hearing in noisy environments and in complex sound processing, and its efferent limb provides inhibitory feedback to the cochlea. The efferent neurones of the olivocochlear bundle travel with the inferior division of the vestibular nerve and reach the outer hair cells of the both ipsilateral and contralateral cochleae via the vestibulocochlear anastomosis of Oort and into the spiral ganglion. The inhibition provided by the olivocochlear bundle helps to protect the cochlea from acoustic trauma.
4. The lateral lemniscus is the principal pathway by which auditory signals from the cochlear nuclei and superior olivary complex reach the inferior colliculus.
5. The inferior colliculus is located in the midbrain and is a synaptic centre for all ascending and descending pathways between the brainstem and cerebral cortex. Its main functions are frequency determination and integration of auditory and non-auditory somatosensory and visual systems.
6. The medial geniculate body of the thalamus contains relay connections between the auditory cortex, vestibular nuclei and spinal cord, with a role in arousal and attention to acoustic stimuli.
7. The auditory cortex is deep within the sylvian fissure on the superior surface of the temporal lobe and includes the primary and secondary auditory cortices and four auditory fields. The primary auditory cortex AI is Brodmann area 41, and the secondary cortex AII is Brodmann area 42, on the posterior aspect of the superior temporal gyrus. The adjacent Wernicke area, or Brodmann area 22, is associated with receptive language. The arcuate fasciculus connects auditory association areas in the inferior parietal lobe with the pars triangularis of the frontal operculum, where the Broca area, or Brodmann areas 44 and 45, deals with expressive language and music perception.

## GENETICS

The genetics of hearing loss are complex, involving over 110 causative genes and over 6000 genetic variants. Congenital hearing loss has a genetic cause in 80% of cases. In contrast, acquired hearing loss in adults is most often attributed to environmental factors, but more likely reflects interaction between environmental and genetic factors, the most frequent of which are age-related (estimated to be 45% genetic) and noise-induced hearing loss.

Hereditary hearing loss can be classified into syndromic or non-syndromic and by pattern of inheritance including autosomal-dominant,

autosomal-recessive, X linked and mitochondrial. Non-syndromic hearing loss may be referred to by the causative gene or by its genetic locus. Deafness loci are designated DFN (for DeaFNess), mode of inheritance (A for autosomal-dominant, B for autosomal-recessive, X for X linked) and a number that indicates the order of gene discovery.

Over 75% of non-syndromic hearing loss is inherited in an autosomal-recessive pattern, which is more likely to result in prelingual hearing loss or deafness prior to speech development. Conversely, non-syndromic autosomal-dominant hearing loss is more likely to cause postlingual and progressive hearing loss.

The most common genetic cause of congenital sensorineural hearing loss is the Connexin 26 mutation, affecting the *GJB2* gene and DFNB1 locus. This is inherited in an autosomal-recessive manner and usually presents as prelingual severe-to-profound hearing loss.

X linked and mitochondrial genetic mutations contribute to less than 2% of cases of non-syndromic inherited hearing loss. However, a cause of hearing loss that is particularly pertinent to medical care is the A1555G mutation in the mitochondrial *MT-RNR1* gene, which encodes for the 12S ribosomal RNA. In people with this genetic variant, hearing loss may be induced by appropriate doses of aminoglycosides.

Over 400 genetic syndromes are associated with hearing loss. The most common causes of syndromic hearing loss are:

1. Autosomal-dominant:
   Waardenburg syndrome—hearing loss with pigmentation anomalies of the skin, hair and eyes, branchio-oto-renal syndrome—abnormalities of the second branchial arch structures, ears and kidneys.
   Stickler syndrome—disorder of types II and XI collagen leading to facial dysmorphism, ocular anomalies, hearing loss and joint problems.
2. Autosomal-recessive:
   Pendred syndrome—hearing loss and thyroid goitre due to abnormal iodide/chloride transporter.
   Usher syndrome—hearing loss with retinitis pigmentosa).
   Jervell and Lange-Nielsen syndrome—hearing loss with long QT syndrome.
3. X linked:
   Alport syndrome—disorder of type IV collagen leading to hearing loss and glomerulonephritis.
   Mohr-Tranebjaerg syndrome—neurodegenerative disorder characterised by hearing loss, adolescent dystonia or ataxis and optic neuronopathy and early-onset dementia.

For the purposes of genetic counselling, the risk of hearing loss in family members of people with genetic hearing loss depends on the pattern of inheritance and the variability of expressivity. If a specific diagnosis or mode of inheritance cannot be established, for a normal-hearing non-consanguineous couple with one deaf child, and otherwise negative family history of hearing loss, there is an 18% empirical risk of deafness in future children.

## TYPES OF HEARING LOSS

Hearing losses are divided into those that prevent sound reaching the otherwise normal inner ear (conductive hearing loss), those that have problems in the inner ear/auditory nerve (sensorineural) and combinations of both (mixed). Figure 1 shows where the sites of hearing loss may occur.

### Conductive hearing loss

Conductive hearing loss is generally caused by conditions affecting the outer or middle ear. Common conditions include: cerumen impaction or foreign body, otitis externa, tympanic membrane perforation, otitis media (acute and chronic), ossicular disruption or fixation and otosclerosis.

Conductive hearing loss is mild to moderate in severity and the medical or surgical treatment of most causes results in improvement in hearing.

### Sensorineural hearing loss

Sensorineural hearing loss is caused by dysfunction in the cochlea, primarily of the sensory hair cells or the spiral ganglia neurones of the auditory nerve.

Common conditions include: presbycusis, noise-induced hearing loss and ototoxicity.

This type of hearing loss is generally permanent and so its management is primarily with hearing rehabilitation using hearing aids or cochlear implants for bilateral severe–profound loss.

### Mixed hearing loss

Mixed hearing loss occurs when the patient has both conductive and sensorineural hearing loss.

## SCREENING

One per 1000 babies is born with permanent congenital hearing loss, and newborn screening allows early detection and intervention to prevent delay in language acquisition. The UK National Health Service (NHS) newborn hearing screening programme is offered to babies born or resident in the UK, ideally within 4 weeks of birth. Babies who have an already-known risk of hearing loss, including those born with atresia or having bacterial meningitis can be referred for full assessment without screening. The aim is to diagnose hearing loss by aged 60 days and to fit required hearing aids by the age of 90 days.

In the adult population, routine auditory screening is not currently recommended for asymptomatic persons. The UK National Screening Committee and US Preventive Services Task Force[4] reviewed the evidence for screening for hearing loss in older adults using clinical screening (testing ability to hear whispered voice, finger rub or tick of a watch), a screening questionnaire (Hearing Handicap Inventory for the

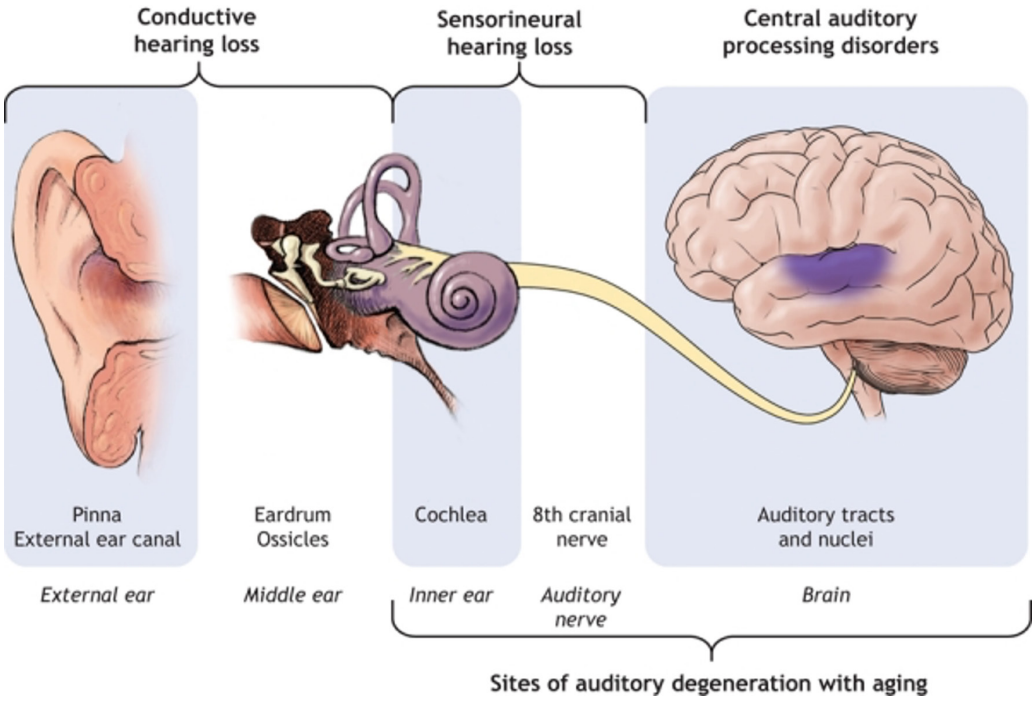


**Figure 1** Types of hearing loss by site of pathology. Previously published in: Bance, M. Hearing and ageing. CMAJ. 2007 March; 176(7): 925–927. Reproduced with permission.

Elderly-Screening) and the AudioScope (otoscope with built in audiometer). They concluded that the tests did help and could easily be performed in the primary care clinic, but that as yet no test with sufficient reliability had been fully validated. The use of screening for patients with regular exposure to loud or occupational noise is being evaluated.

Under the NHS Direct Access Audiology scheme, general practitioners can refer all patients aged over 50 years who have concerns about hearing loss for free audiological testing and hearing services.

## HISTORY

For patients presenting with hearing loss, it is important to identify its characteristics:

1. Duration and progression of hearing loss.
2. Unilateral or bilateral.
3. Associated symptoms, such as otalgia, otorrhoea, tinnitus or vertigo.

Patients should also be asked about a history of chronic or recurrent ear disease or previous otologic surgery. History can also reveal the cause of hearing loss including:

1. Exposure to acoustic trauma.
2. Use of ototoxic medications, such as loop diuretics, salicylates, aminoglycoside antibiotics and platinum chemotherapeutic agents.
3. Family history of hearing loss.
4. History of head trauma or barotrauma.
5. Chronic medical conditions, such as cardiovascular or cerebrovascular disease, diabetes mellitus, autoimmune disease and renal disease.

## PHYSICAL EXAMINATION

Clinical examination for hearing loss includes examination of the external ear and otoscopy, followed by tuning-fork testing. Cranial nerve and vestibular examination may also be indicated in certain conditions.

The external auditory canal should be examined for obstruction by cerumen, foreign body or discharge, structural stenosis or mass and for evidence of inflammation of the canal skin. The tympanic membrane should be examined for perforation, inflammation, opacity or retraction. It may be possible to visualise the middle ear structures and the presence of fluid, mass or ossicular erosion.

The Weber and Rinne tuning fork tests (typically using a 512 Hz tuning fork) should be used to help differentiate between conductive and sensorineural hearing loss. The Weber test is performed by placing a vibrating tuning fork on the patient's vertex, in middle of the forehead or over the top teeth equidistant from the patient's ears. The patient is asked to report where they hear the sound or in which ear the sound is louder. With symmetrical hearing, the sound is heard as central. The Weber will lateralise to the affected ear in case of conductive hearing loss or to the better ear in sensorineural hearing loss. It is far more sensitive for asymmetric conductive hearing loss than for asymmetric sensorineural hearing loss.

The Rinne test is performed by placing a vibrating tuning fork adjacent to the outer ear and then pressing the footplate of the tuning fork firmly on the mastoid. The patient is asked to report which sound is louder. A positive Rinne is when the sound

is heard louder outside the ear (air conduction). A negative Rinne is when sound is heard louder on the mastoid (bone conduction), which indicates conductive hearing loss. Conductive hearing loss needs to be large, in the order of 20–25 dB, to be detected reliably.

## AUDIOMETRY

Hearing is measured in decibels (dB) and the auditory threshold is the softest sound a patient can hear at a particular frequency at least 50% of the time. The pure-tone audiogram plots the patient's thresholds across the range of frequencies, for air-conducted and bone-conducted stimuli. Sound frequency or pitch is recorded on the horizontal axis, and sound intensity is recorded on the vertical axis. By convention, right ear thresholds are recorded as a red circle and left ear thresholds are recorded as a blue X.

From this, the clinician can determine the type, severity and pattern of hearing loss. The difference in hearing thresholds between air-conducted and bone-conducted stimuli is known as the air–bone gap and indicates the presence of conductive or mixed hearing loss. Equal air and bone conduction thresholds above 25 dB indicate a purely sensorineural hearing loss.

Other audiological tests include:

1. Tympanometry. This is a test of middle ear function using pressure and compliance; it can indicate the presence of a middle ear effusion, tympanic membrane perforation or Eustachian tube dysfunction.
2. Acoustic reflex. This is a test of integrity of the stapedius reflex arc; it is useful if suspecting otosclerosis, on the basis of the pure-tone audiogram suggesting conductive hearing loss, but with normal otoscopic examination and tympanometry.
3. Speech discrimination. This measures a person's ability to detect and understand spoken words at particular hearing thresholds; it can provide information regarding central auditory function and is a major determinant in assessing whether a hearing aid will help or whether someone is a candidate for cochlear implantation.
4. Otoacoustic emissions. These test outer hair cell function due to the production of audio-frequency signals by the cochlea in response to sound, as measured by insertion of a probe with miniature loudspeaker and microphone into the sealed external auditory canal.
5. Auditory brainstem responses. These use surface electrodes to test neural activity in the auditory nerve and auditory brainstem nuclei. Waves I and II of the auditory brainstem responses reflect activation of the cochlear nerve; waves III and IV reflect activation of the cochlear nucleus and superior olivary complex and wave V is associated with activation of the lateral lemniscus and inferior colliculus.

Figures 2–5 are examith varying pathologies.

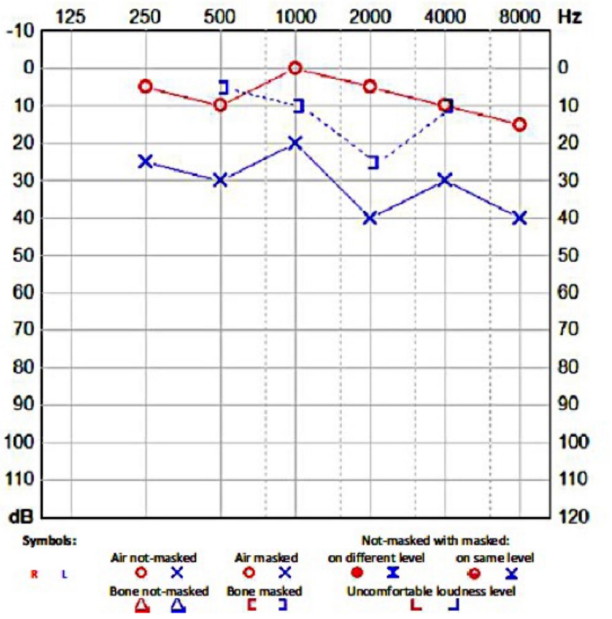


**Figure 2** Audiogram showing left mild conductive hearing loss in a patient with a tympanic membrane perforation.

## SPECIAL TYPES OF HEARING LOSS

### Presbycusis

Age-related hearing loss is due to a combination of degenerative effects on the cochlear hair cells and spiral ganglia neurones, decreased functioning of the stria vascularis which metabolically supports the hair cells and cumulative effects of noise and ototoxic exposure. Presbycusis is usually sensorineural, bilateral and symmetrical, gradual and progressive and worse at higher frequencies. A prominent feature is reduction in speech perception.

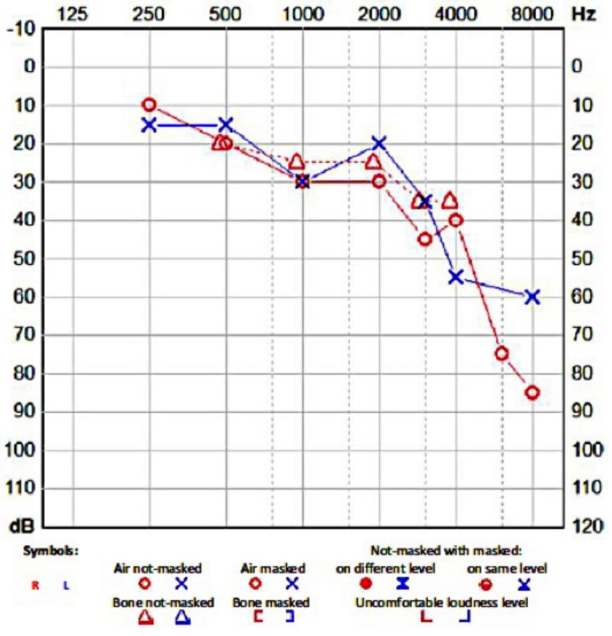


**Figure 3** Audiogram showing bilateral downsloping sensorineural hearing loss, typical of presbycusis.



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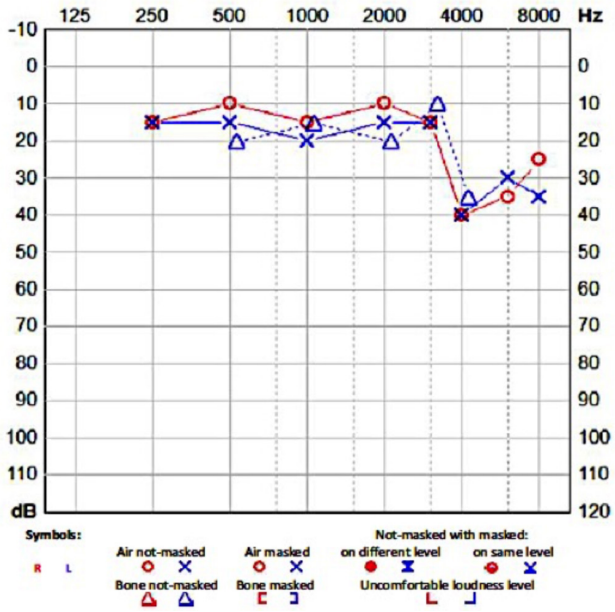


**Figure 4** Audiogram for a patient with noise-induced hearing loss, characterised by symmetrical high frequency sensorineural hearing loss with a 'notch' at 4 kHz.

### Noise-induced hearing loss

Loud noise exposure can occur with occupations such as factory workers or military personnel or with recreational activities involving loud music, power sports or firearm use. If acoustic trauma is severe, prolonged or repetitive, it can cause death of hair cells and spiral ganglion neurones leading to permanent hearing loss. Audiological features of noise-induced hearing loss are hearing loss primarily between 3 and 6 kHz with a 'noise notch' at 4 kHz on pure-tone audiogram and loss of otoacoustic emissions.

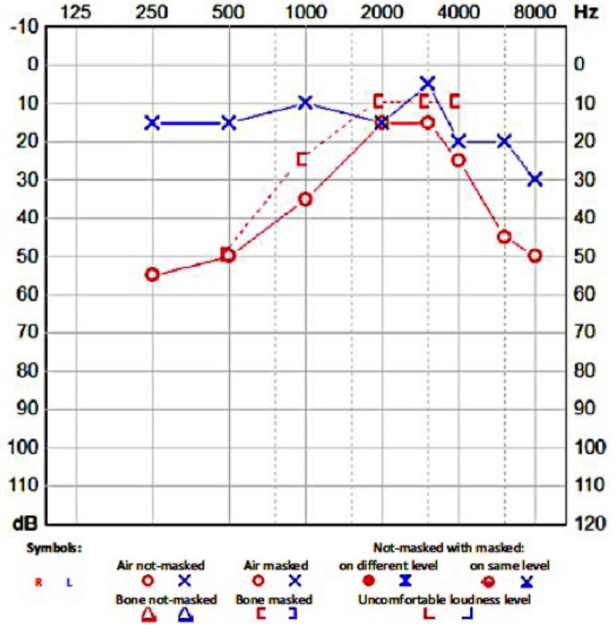


**Figure 5** Audiogram showing left sensorineural hearing loss, primarily affecting the low frequencies. This is typical of Ménière's disease.

### Autoimmune disease and hearing loss

Autoimmune ear disease accounts for less than 1% of hearing impairment or vertigo. Hearing loss may be associated with rheumatoid arthritis, systemic lupus erythematosus, Cogan's syndrome, sarcoidosis, granulomatosis with polyangiitis and other autoimmune disorders. Female preponderance is apparent and symptoms often occur between ages 20 and 50 years. The clinical hallmark of autoimmune ear disease is bilateral sensorineural hearing loss that is rapidly progressive (over weeks to months) or fluctuating, with up to 80% of patients also having vestibular dysfunction. Granulomatosis with polyangiitis may also feature middle ear inflammation leading to effusion, along with systemic manifestations of the underlying autoimmune disease. The main differential diagnoses are Ménière's disease, otosyphilis and retrocochlear neoplasm. The diagnosis of autoimmune ear disease is primarily clinical, as no laboratory test can definitively confirm it. Recommended investigations include blood count and white cell differential, erythrocyte sedimentation rate, serum C reactive protein, rheumatoid factor, antinuclear antibodies, antineutrophil cytoplasmic antibodies, anti-double-stranded DNA antibodies, anti-SSA/B antibodies, antiphospholipid antibodies, serum complement, thyroid function tests and syphilis serology. MR scan of the internal auditory meatus with gadolinium is used to exclude a neoplastic process. Patients with suspected autoimmune ear disease should be managed in a multidisciplinary team approach including otolaryngology, audiology and immunology. Recommended empirical treatment is with corticosteroids, provided there are no contraindications. Other evaluated treatments include cyclophosphamide, methotrexate and etanercept (tumour-necrosis factor antibody).

### Sudden sensorineural hearing loss

Sudden sensorineural hearing loss is defined as onset of hearing loss of at least 30 dB, affecting at least three frequencies, over 72 hours or less and is considered an otological emergency. The cause is presumed to be viral, vascular, autoimmune or neoplastic. Urgent referral to ENT and audiology services can establish that the hearing loss is sensorineural, and oral corticosteroids are recommended and give statistically significant improvement in hearing recovery compared with no treatment. Intratympanic corticosteroids are considered in patients who have not improved with oral corticosteroids or in whom systemic corticosteroids are contraindicated.

### Single-sided deafness

Patients may develop unilateral hearing loss from trauma, infection, neoplasm or conditions such as Ménière's disease. There are approximately 9000 new

cases of single-sided deafness in the UK per year. Due to loss of binaural processing, patients may have difficulties with:

1. Sound localisation.
2. Understanding speech in background noise.
3. Cognitive load: listening while performing other tasks.
4. Head shadow effect: inability to hear sounds directed to the affected ear.

## IMAGING, RETROCOCHLEAR SCREENING AND ADDITIONAL INVESTIGATIONS

The investigation of choice for conductive hearing loss is a CT scan of the temporal bones. Gadolinium-enhanced MR imaging is the investigation of choice for retrocochlear pathology. The primary indications for MR scanning in hearing loss are:

1. History of sudden sensorineural hearing loss: less than 1% of patients with sudden hearing loss have a vestibular schwannoma but this can be the presenting complaint in 10% of tumours.
2. Asymmetric hearing loss: this is defined as a 30 dB threshold difference at a single frequency, 15 dB threshold difference at two contiguous frequencies or a 10 dB threshold difference at three contiguous frequencies.

Additional investigation of sensorineural hearing loss may be indicated if:

1. There is associated episodic vertigo or progressive ataxia, to rule out inner ear diseases such as Ménière's disease or autoimmune inner ear disease.
2. The hearing loss is rapidly progressive: consider ototoxicity, autoimmune inner ear disease or systemic diseases such as syphilis.

## HEARING REHABILITATION

The management of hearing loss depends on the cause and type of hearing loss and may entail medical therapy, surgery, amplification or hearing implants. Medical therapies are used for hearing loss that results from infection or systemic causes, which may require topical or oral antibiotics or systemic corticosteroids. Surgery may be offered for most causes of conductive hearing loss, such as myringotomy for middle ear effusion, tympanoplasty for tympanic membrane perforation, ossiculoplasty for ossicular disruption or stapedotomy for otosclerosis.

Hearing-assistive technologies and hearing aids are the mainstay for management of sensorineural hearing loss. Assistive devices include amplified telephones, television captioning, video conferencing and visual or vibrotactile alerts. Hearing aids provide amplification and are based on prescriptive algorithms using the patient's audiological tests and personal preferences. People with single-sided deafness may be fitted with a contralateral routing of sound aid, which transmits sounds to a receiver worn on the better hearing ear. Deterrents to the use of hearing aids include financial cost, stigma, perceived ineffectiveness, discomfort and cosmetic appearance.

A bone-anchored hearing aid is a surgically implanted osseo-integrated titanium screw that transmits sounds directly through the skull to the cochlea, bypassing the outer and middle ear. They can be considered in patients with conductive hearing loss who cannot use traditional hearing aids or for people with unilateral hearing loss. Middle ear implants are directly attached to the ossicles or round window and are an option for people with conductive or mixed hearing loss.

Patients with severe–profound sensorineural hearing loss (greater than 90 dB hearing loss at 2 and 4 kHz without aids), who have not adequately benefitted from hearing aids,[5] may be candidates for cochlear implantation. A cochlear implant uses an electrode array that bypasses the cochlear hair cells and directly stimulates the spiral ganglion neurones. An external processor receives sounds and converts it into electrical impulses that are sent to a receiver and routed to the electrode array. Cochlear implants permit partial restoration of hearing and improved speech perception. Current evidence shows that elderly people benefit from cochlear implants from improved cognition, psychological functioning and quality of life, and advanced age is not a contraindication for implantation.

### Key points

- Hearing loss affects a significant proportion of the population, particularly the elderly and can lead to disability and reduced quality of life.
- Hearing loss may be conductive, sensorineural or mixed.
- Assessment of hearing loss relies on accurate history and examination to determine its characteristics and cause and is complemented by audiological testing to determine severity and type.
- Asymmetric or sudden sensorineural hearing loss can indicate retrocochlear pathology; retrocochlear screening with MR imaging is recommended.
- Hearing rehabilitation options include hearing-assistive devices, hearing aids and hearing implants.

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