Hearing loss

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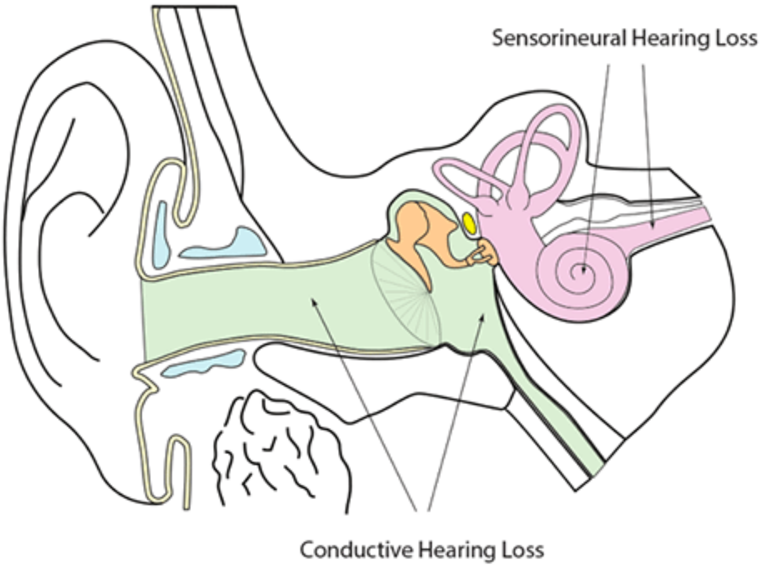
Remit

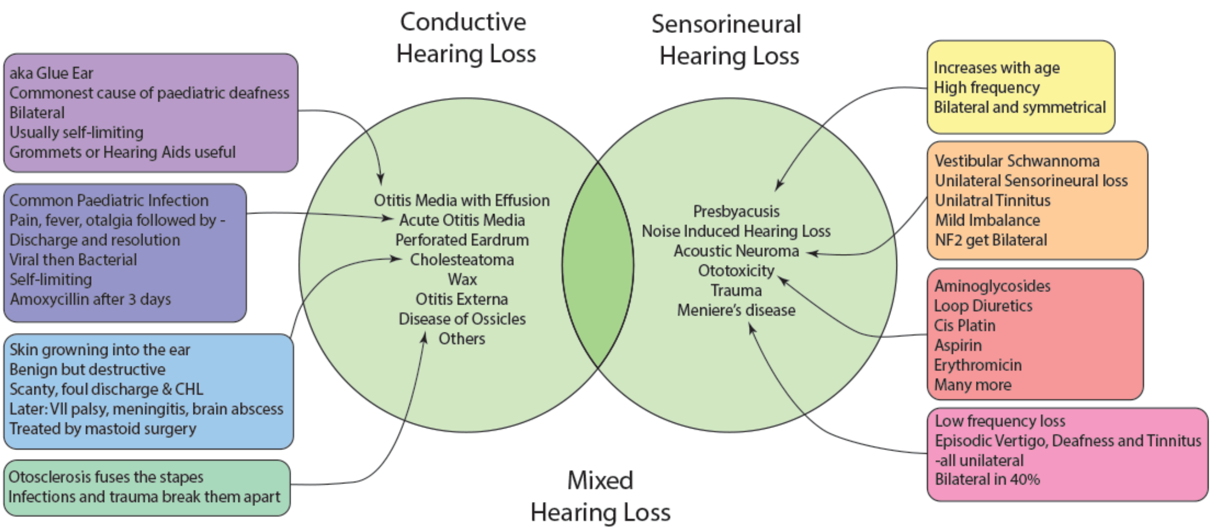
This tutorial examines the different types of hearing loss and their causes.

After the introduction it is divided into three sections: Sensorineural hearing loss, Conductive hearing loss (including mixed losses) and presbyacusis which is the commonest sensorineural loss and deserves a separate discussion.



Introduction

Broadly speaking deafness may be sensorineural, conductive or mixed. The type of hearing loss depends on the site of disease. Diseases that affect the outer or middle ear cause conductive loss and those that affect the cochlea or cochlear nerve cause sensorineural losses. There are a few diseases that can cause both types of deafness (e.g. otosclerosis, cholesteatoma and acute suppurative otitis media).



Hearing loss is discussed at length in this tutorial. The image above summarises hearing loss with the underlying aetiology shown.

Causes

The following diseases cause sensorineural hearing loss. It would be prudent to know a little about all of them but only the ones highlighted are covered here. Note that they are not in order of frequency. Presbyacusis is by far the commonest cause of sensorineural hearing loss.

1. Presbyacusis
2. Drug related
3. Traumatic
4. Congenital
5. Acoustic neuroma
6. Meniere's Disease (and other vestibular syndromes)
7. Noise induced hearing loss

Drug related deafness

The drugs that can cause hearing loss are many and only the common ones are listed here together with examples in the class and whether the deafness is usually reversible or not. The hearing loss may be unilateral or bilateral and typically affects the high frequencies first. Drugs that cause hearing loss are termed ototoxic drugs.

1. Aminoglycosides (gentamicin, neomycin) - irreversible
2. Loop diuretics (furosemide, bumetamide) - largely reversible
3. Chemotherapeutic agents (cisplatin, carboplatin) - irreversible
4. Aspirin – reversible

The first aminoglycoside was streptomicin and it was used for the treatment of TB. It became apparent that it caused hearing loss and it is now known that other drugs in the same family cause the same problem.

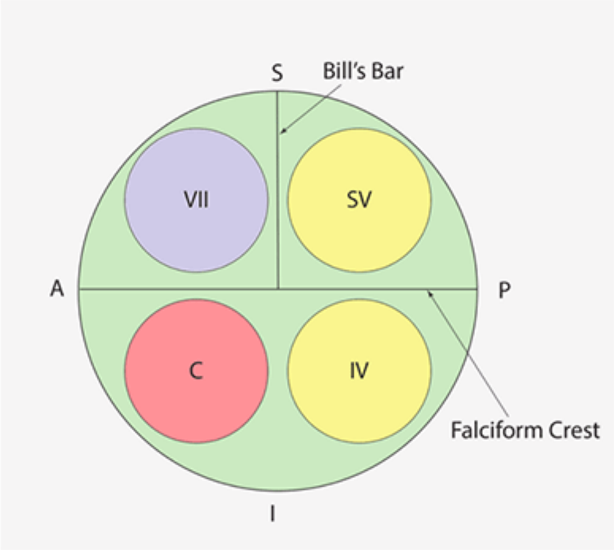
These days it is gentamicin that is most commonly quoted as an ototoxic drug but other aminoglycosides such as neomycin and kanamycin are also important. In general ototoxicity occurs when the serum level of the drug is high and this may occur in patients on systemic therapy who also have impaired renal function (as this is the principle route of excretion).

Ototoxic drugs can be subdivided into those that predominantly cause deafness (cochleotoxic) and those that cause vestibular damage (vestibulotoxic). Remember that predominantly cochleotoxic drugs also cause some vestibulotoxicity and vice versa.

Interestingly the ototoxic nature of gentamicin can be used for therapeutic benefit in patients with Meniere's disease. Gentamicin is predominantly vestibulotoxic (with a small ototoxic potential) and can be instilled into the middle ear in order to poison the vestibular apparatus on that side. See the Meniere's tutorial for more.

Acoustic Neuroma

This is a badly named tumour as it is in reality a schwannoma and seldom occurs on the cochlea nerve (usually it arises on the superior vestibular nerve). So really it should be called a vestibular schwannoma.



The figure shows the anatomy of the right Internal Acoustic Meatus as seen from the brainstem. SV is the superior vestibular nerve and IV the inferior and C the cochlear nerve. VII is the facial nerve.

The tumour is benign but is situated in the bony internal auditory meatus and as it expands it grows towards the brain stem. Hence, although it is benign, it can be extremely dangerous by virtue of its position and causes death by brain stem compression.

The characteristic of the ear symptoms caused by an acoustic neuroma are that they are unilateral. For example, the patient may suffer with unilateral sensorineural hearing loss or unilateral tinnitus. Interestingly, although the tumour usually arises from the vestibular nerve, vertigo is uncommon. Patients may notice a mild disequilibrium only. This is due to the slow growing nature of the tumour and the ability of the brain to compensate for balance disturbance.

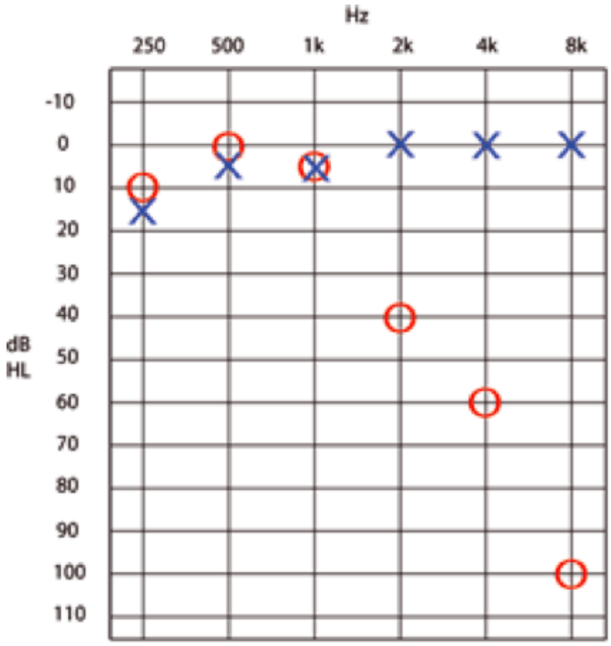
Although the facial nerve is also in the IAM a neuroma causes facial weakness relatively late on in the disease. This is because motor nerve fibres are relatively resistant to pressure. This is in contrast to sensory fibres (hearing) as these are thin and easily damaged.

Investigation of acoustic neuromas is relatively simple - MRI. No other modality has the sensitivity, specificity or convenience. Audiograms are done to confirm sensory asymmetry but more arcane audiological techniques are seldom used now. CT scanning can be used but is less sensitive.

On the right is the audiogram of a patient with an acoustic neuroma in their right IAM. Note how the left side hearing is normal and that there is a drop-off in hearing thresholds on the right.

Almost all patients with asymmetric sensorineural hearing loss have an MRI. Asymmetry is variously defined but the Northern Regional Guidelines state that asymmetry is a difference in at least 20dBHL in two adjacent frequencies.

In this audiogram there is greater than 20dBHL asymmetry at 2,4 and 8kHz and an MRI would normally be ordered.



Acoustic neuromas are treated in a number of possible ways:

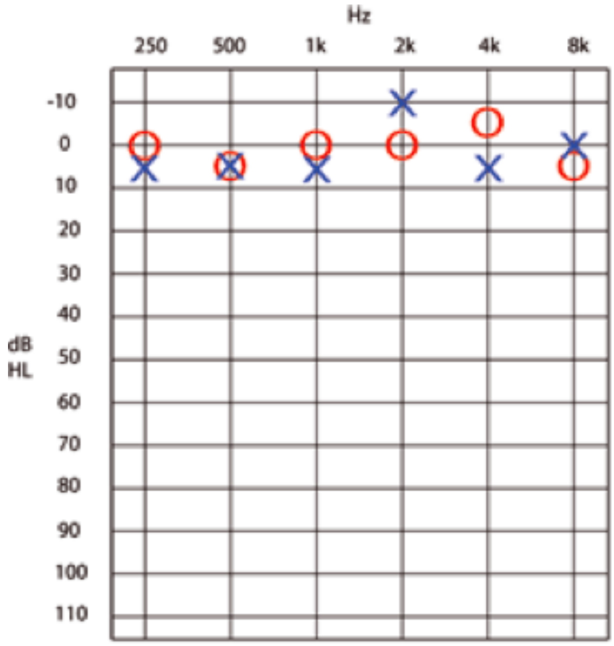
1. Conservative. The tumour is slow growing usually. Small tumours can be safely watched for many years without risk to the patient. This is done by serial scanning
2. Surgery. This requires a major neurosurgical procedure with it's attendant risks
3. Radiosurgery. This is relatively new modality but is a non-invasive treatment.

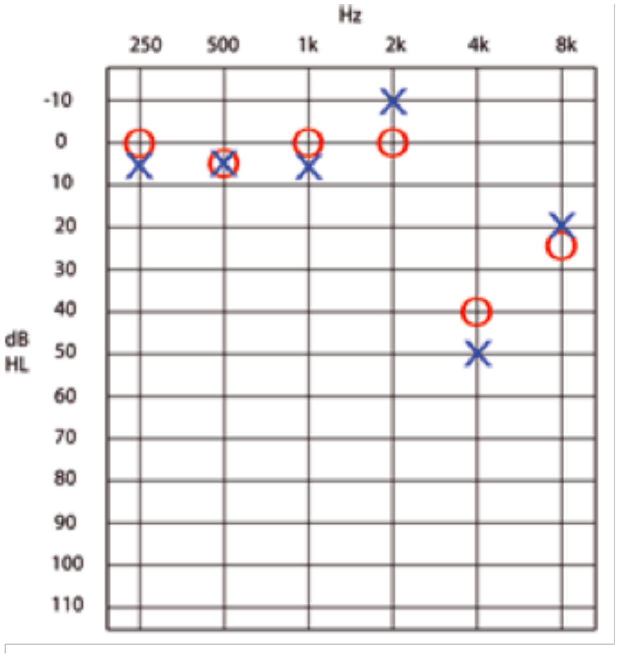
Noise Induced Hearing Loss

Daily exposure to high intensity noise over a prolonged period will lead to NIHL. This is seen in people who have worked in the mining industry, steel works, panel beating etc. It can also happen to musicians and commonly occurred in members of the armed forces from gunfire.

These days companies must provide hearing protection to their workforce if they work in a loud environment for long periods and NIHL is becoming progressively less common.

The hearing loss is characteristically sensorineural and at its maximum at 4kHz. It is almost always present in both ears. Since the damage is irreversible only hearing aids offer any help to the severely affected.





The audiogram on the left shows normal hearing thresholds on the right (red circles) and on the left (blue crosses). The one on the right is typical of a patient with noise induced hearing loss. Note that at 4kHz there is a marked increase in the threshold of hearing on both sides. The 8kHz threshold has suffered a little as well.

Congenital Hearing Loss

This is deafness present at birth and is distinct from hereditary deafness (where a deafness may or may not be present at birth).

In the UK 1:1000 babies are born with hearing loss. This may be in isolation or as part of a variety of other abnormalities. The deafness may be sensorineural or conductive.

It is essential that these babies are identified as early as possible. The sooner they get appropriate hearing help the better. Normal hearing helps us with language acquisition but also with social, emotional and intellectual development.

Here is a brief list of the causes of congenital hearing loss. Some of these will be familiar to you if you have done your Obs & Gynae blocks.

1. Toxoplasmosis
2. Rubella
3. CMV
4. Herpes
5. Syphilis
6. Neonatal hypoxia
7. Rhesus incompatibility

Babies that have been admitted to Special Care Baby Unit (or equivalent) have 10 times the risk of deafness than their peer group.

Presbyacusis

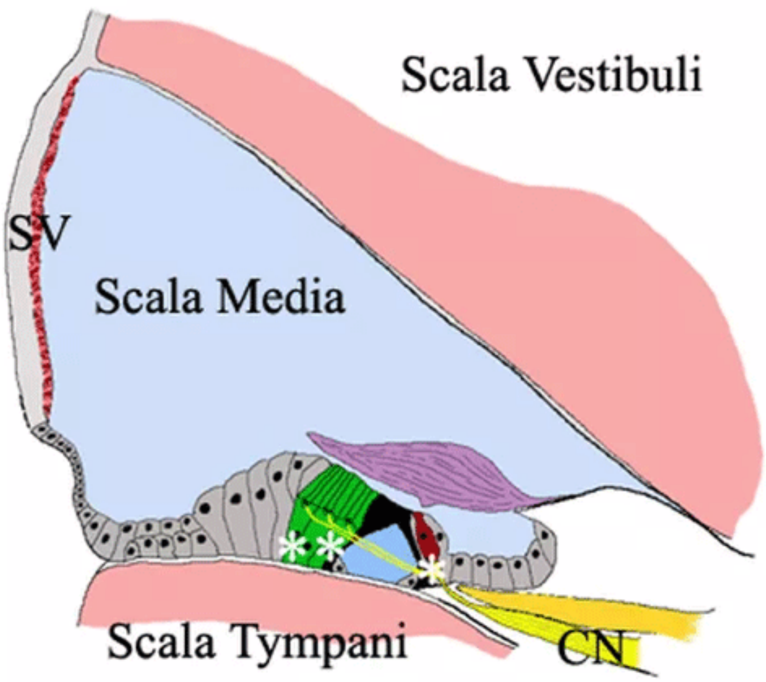
Presbyacusis is the decline in hearing thresholds that is associated with age. Note that age does not cause it.

It is estimated that 15% of those 55-64 years of age, 30% of those 65-74 years of age, and 40% of those over 75 years of age in the UK have a hearing loss that affects communication.

Pathophysiology

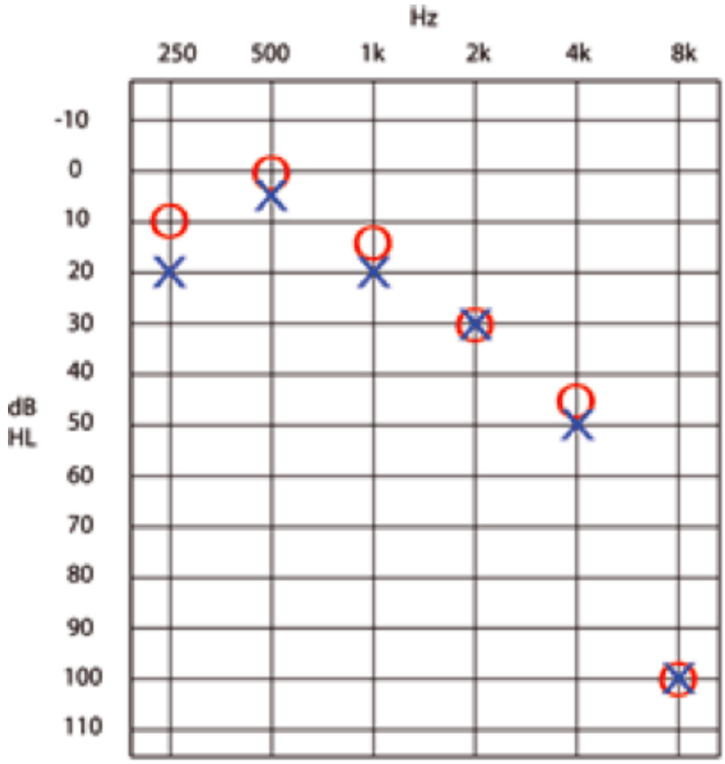
Presbyacusis is a process that affects both the cochlea and the nerves that run from it into the brainstem.

Below is a cross section of the cochlea zoomed in on the scala media. It is in the scala media that the organ of Corti is found. This organ is responsible for the transduction of vibration into neural activity.



Key to diagram of cochlea:
* inner hair cell
** outer hair cell
CN Cochlear Nerve
SV Stria Vascularis.

In the cochlea both the outer and inner hair cells of the organ of Corti are affected. This is especially so at the basal turn of the cochlea where high frequency sound is detected. This means that the patients get a high frequency hearing loss. As time passes the hair cells further up the cochlea are affected and progressively lower frequencies are lost.

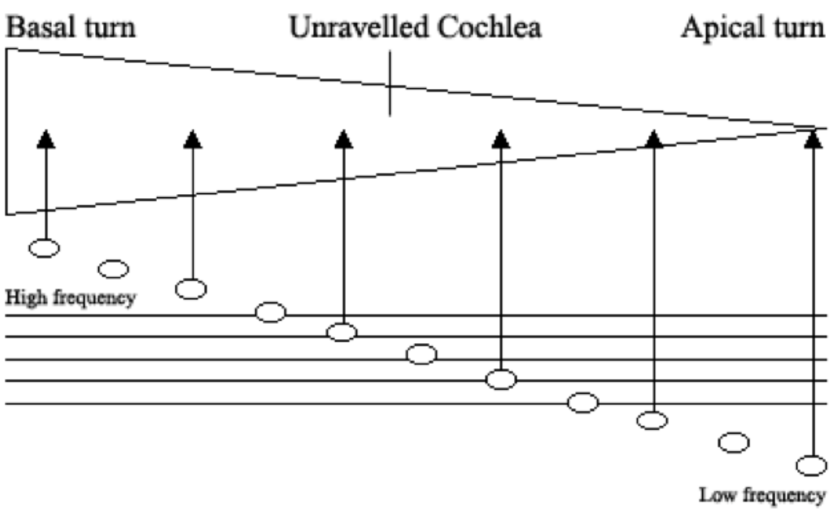


The audiogram on the left is typical of presbyacusis. Note that the thresholds for hearing in the high frequencies are elevated. At 2kHz this patient can only hear when the sound is louder than 30dBHL. At 4 and 8kHz the sound must be 50 and 100dBHL before the patient can hear it.

Note also that presbyacusis is a bilateral process.

With loss of hair cells is associated loss of neural cells in the spiral ganglion and hence in the cochlear nerve.

In the diagram below the cochlea is 'unravelled' and the tonotopic organisation shown. Musicians should note that the scale shown below represents only part of the range a human can hear. In fact we can hear frequencies between 64 and 23,000Hz. Look at the diagram carefully and note that high frequency sound is detected in the basal turn and low frequencies at the thin apex of the cochlea (this is the opposite way round to the strings in a piano where the high notes come from short wires and the low notes from long ones).



Loss of high-frequency hearing is a particular problem when listening to speech.

Speech consists of consonants and vowels. Consonant sounds tend to be higher frequency sounds. Try saying the following letters aloud: 'F', 'S', 'Ch', 'Sh' and you will see.

Vowel sounds are low frequency. Try saying: 'Oo', 'ee', “m”

The patient with presbyacusis can hear (low-frequency) vowel sounds quite well but not (high-frequency) consonants. The problem is that consonants provide a lot more information about the content of a sentence than vowels do. Read the following two sentences. In the first the consonants have been removed and just the vowels remain, in the second the vowels have been removed. The sentences have nine words.

'e ui o o u oe e a o'

'Th qck brwn fx jmps vr th lzy dg'

Understanding the first is very difficult but the second is easily recognisable. In fact, both sentences are the same:

'The quick brown fox jumps over the lazy dog'

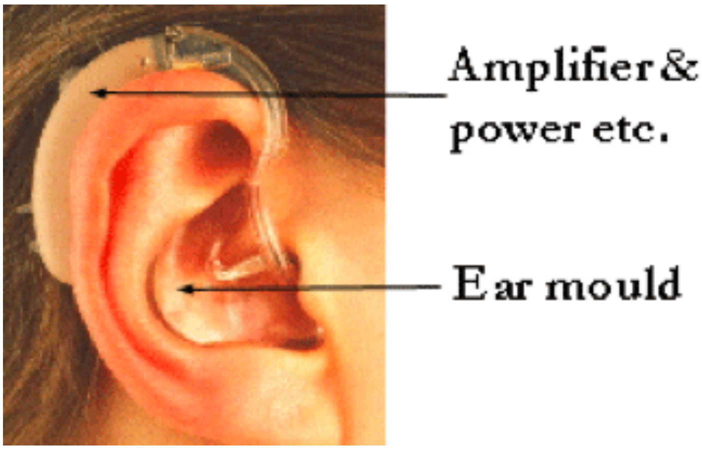
This illustrates the importance of high-frequency hearing. Our environment has continual noise. It tends to be low-frequency background noise and this poses another problem for the patient with high-frequency hearing loss - the frequencies in speech that they can hear are drowned in background noise.

Management

Sensorineural hearing loss of this type is irreversible so the patient requires assessment as to the degree of disability and provision of a hearing aid and hearing rehabilitation.

Hearing aids

All hearing aids have the same basic design: a receiver, a processor/amplifier and a speaker. There are different models: some are hidden in the ear canal while others fill the conchal bowl of the pinna. A full treatment of hearing aid design and technology is not within the scope of this article.



This aid is a 'behind the ear' (BTE) aid. The amplifier, power source, switches and volume control sit behind the pinna. The sound output passes down the clear plastic tubing, through the plastic insert and down the ear canal to the ear drum.

Other types of hearing aid are: Bone Anchored Hearing Aid (BAHA) and Cochlear Implant.

Rehabilitation

Lip reading, enhancing the environment to favour the patient and counselling about hearing aid use are sometimes needed for the patient.