Hearing loss

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Also known as hearing impairment, is a partial or total inability to hear

Normal human being can hears sounds of intensity lower than 15 dB

Disabling hearing loss refers to hearing loss greater than 40 dB in the better hearing ear in adults (15 years or older) and greater than 30 dB in the better hearing ear in children (0 to 14 years).

Types of hearing Loss

1. Conductive hearing loss : defect In the external ear , tympanic membrane or the middle ear
2. Sensori-neural hearing loss : defect in the cochlea or its neural connection
   Sensori – related to the cochlea
   Neural – related to the cochlear nerve or its neural connections
3. Mixed hearing loss : both conductive and sensori-neural elements are present

Degree of hearing loss

Hearing loss classified according to the severity to

Slight, mild moderate, moderately severe, severe and profound hearing loss, below table showing thresholds of hearing of each class of hearing loss and its impact on the children

class	Hearing threshold	Impact
Normal hearing	-10 to 15 dB	Child can hear sounds and speech even if they are very soft
Slight hearing loss	16 to 25 dB	Child may have trouble hearing soft speech, soft sounds, or speech spoken from a distance
Mild hearing loss	26 to 40 dB	Child will consistently miss some speech sounds.
Moderate hearing loss	41 to 55 dB	Child just barely hears speech at a conversational level in a quiet environment and may or may not be able to understand what is being said.
Moderately severe hearing loss	56 to 70 dB	Child may hear some speech sounds, but will be unable to understand speech without hearing aids.
Severe hearing loss	71 to 90 dB	Child may detect loud sounds in the environment, but will not hear normal conversational speech
Profound hearing loss	> 91 dB	Child likely responds more to vibrations than to sounds.

Causes of conductive hearing loss:

External ear canal Wax

Congenital anomaly of the outer and middle ear

Foreign bodies

Stenosing otitis externa

Exostoses and osteomas

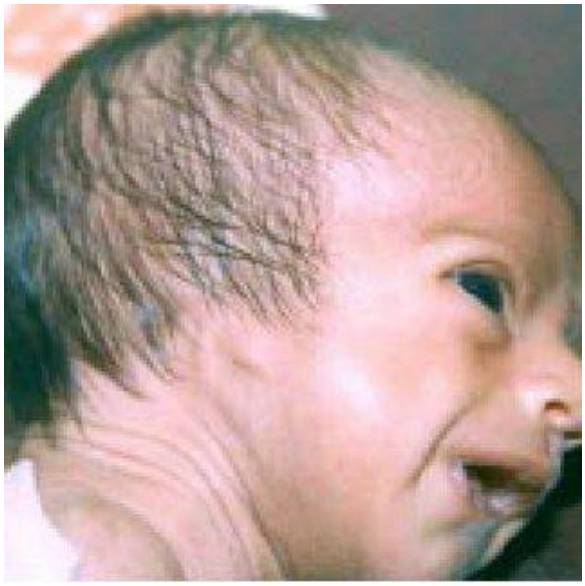
Tympanic membrane Perforation

Ossicular chain Fixation, e.g., otosclerosis

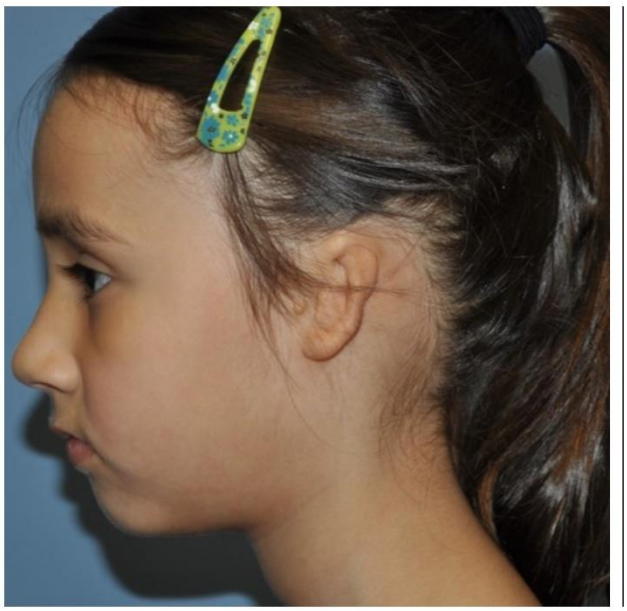
Erosion, e.g., cholesteatoma

Subluxation and fracture, e.g., trauma

Anotia: congenital absence of the external ear



Microtia: congenital small auricle with no external canal



Causes of sensori-neural hearing loss:

Idiopathic

Hereditary

Degenerative: Presbycusis

Infection: Ramsay Hunt syndrome, syphilis, meningitis

Ischaemia: Cerebrovascular accident

sickle cell disease

Inflammation: Ménière's disease, Labyrinthitis

Autoimmune disease, e.g rheumatoid arthritis, sarcoidosis

Neoplastic: Vestibular schwannoma

Trauma: Noise

temporal bone fracture

Iatrogenic Surgical,

ototoxicity

Neurological: Multiple sclerosis

Common drugs that lead to hearing loss

- Aspirin, when large doses taken.
- Non-steroidal anti-inflammatory drugs (NSAIDs), such as ibuprofen and naproxen.
- Certain antibiotics, especially aminoglycosides (such as gentamicin, streptomycin, and neomycin).
- Loop diuretics , such as furosemide or bumetanide.
- Medicines used to treat cancer, including cyclophosphamide, cisplatin, and bleomycin.

Mixed Hearing Loss

Pathology affecting the middle and the inner ear, a typical example is cholesteatoma, advanced stage of otosclerosis also leads to mixed hearing loss

Presbycusis

With increasing age, there is loss of inner and outer hair cells within the cochlea as well as loss of cochlear nerve and central auditory neurons. This typically results in a high-frequency sensorineural hearing loss that progresses slowly over time. The age of onset and degree of hearing loss are variable between individuals.

Otosclerosis

Otosclerosis results in the formation of new, abnormal bone at the fissula ante fenestram, a small connective tissue filled cleft between the oval window and cochleariform process. The new bone fixes the stapes and prevents the conduction of sound through the ossicular chain. In most cases, it is an autosomal dominant condition with variable penetrance. As with other forms of conductive hearing loss, rehabilitation options include traditional hearing aids. Stapedectomy is, however, a very successful surgical solution that, in experienced hands, can close the air-bone gap to less than 10 dB in 90% of cases. It involves removal of the arch of the stapes, formation of a small fenestrum in the footplate of the stapes, and placement of a piston through the fenestrum that is hooked around the long process of the incus proximally. There is a 1 to 2% risk of dead ear with stapedectomy.

Ossicular Fixation

Tympanosclerosis and fibrous adhesions resulting from inactive chronic otitis media may result in fixation of the ossicular chain. In many cases tympanosclerotic plaques can be removed although great care needs to be taken around the stapes superstructure as excessive manipulation can

result in sensorineural hearing loss or even a dead ear. Fibrous dhesions can be carefully divided and the laser can be extremely helpful in this setting. Traditional hearing aids may also be offered as an alternative to middle ear surgery.

Stenosing otitis externa

results in progressive inflammatory narrowing of the external auditory canal with eventual formation of a deep ear canal fibrous plug. The underlying tympanic membrane is usually normal. Once the otitis externa has burnt out, surgical correction may be considered in order to restore hearing although stenosis recurs in up to 60% of cases.

The fibrous plug is removed down to the healthy tympanic membrane. A bony canalplasty is usually carried out and the deep ear canal can then be grafted using a split-skin graft. Alternatively, a number of different implantable bone conduction hearing aids can be used and these provide excellent hearing outcomes

Exostoses

result from repeated prolonged exposure to cold water. This causes an hyperostotic reaction resulting in formation of several bony swellings in the deep ear canal. It is most commonly seen in surfers. When small, they do not cause any problems, but large ones can completely occlude the external auditory canal.

Progression can be stopped through appropriate ear plugging during exposure to cold water. This may avoid the need for intervention. Large ones can be removed by carrying out a bony canalplasty

Osteomas

are benign neoplasms of the bony external auditory canal. They are solitary, have a pedunculated base, and usually arise at the bony margin of the tympanic ring. These features differentiate them from exostoses. In the same way as exostoses, they can occlude the external auditory canal when large. It is usually a straight forward matter to fracture the osteoma at its base and remove it although formal canalplasty may be required in some cases.

Ossicular Erosion

Chronic otitis media with or without cholesteatoma can result in erosion of the ossicular chain, particularly the long process of the incus. As a result, the ossicular chain becomes discontinuous and can no longer transfer sound to the inner ear. Again, traditional hearing aids may be used to rehabilitate hearing but surgical reconstruction may also be considered. Localized erosion of the long process of incus can be repaired surgically. Some surgeons may also remove, modify, and then transpose the incus. Success rates are variable with the most important factor being the status of the stapes arch. If present, the reconstruction is more stable and successful closure to within 15 dB air-bone gap closure can be achieved in 70%

Noise-Induced Hearing Loss

Short-term exposure to excess noise results in a reversible reduction in cochlear function termed temporary threshold shift. If such exposure is very prolonged (it takes 10 years of exposure for 8 hours a day at 90 dB or more to cause significant noise-induced hearing loss [NIHL] in an averagely sensitive individual), free radical formation results in loss of cochlear hair cells. The damage is most severe at the 4-kHz portion of the basilar membrane and this typically manifests as a notch at 4 kHz on pure-tone audiometry.

Vestibular Schwannomas

Vestibular schwannomas (often, inaccurately, called acoustic neuromas) are benign tumours originating from the Schwann cells lining the vestibular nerve as it passes from the brainstem, across the cerebellopontine angle (CPA) to the internal auditory meatus. They are the commonest type of tumour to occur in the CPA. Ninety-five percent of tumours are associated with hearing loss. Eighty percent of tumours do not grow after diagnosis and do not require active treatment.

They are usually monitored with serial imaging. Those that grow may be treated with stereotactic radiosurgery or with surgical resection. Any

patient with a significant asymmetry in his or her sensorineural hearing thresholds should be screened for a vestibular schwannoma using MRI.

Sudden sensorineural deafness

The incidence of idiopathic sudden sensorineural hearing loss is around 10 per 100,000 per year. It is usually unilateral. There are a number of theories as to how this type of hearing loss arises.

This includes direct injury from a viral infection (mumps and herpes viruses are most often implicated), a vascular event, an autoimmune response and an abnormal cytokine-mediated response to certain stimuli such as a viral infection or physical or mental stress. Such hearing losses can spontaneously recover but recovery may be facilitated by the use of either oral or intra-tympanic steroids.

Endolymphatic Hydrops

The classic triad of episodic vertigo associated with fluctuating sensorineural hearing loss and tinnitus is the hallmark of Ménière's disease. The underlying pathology is an excess of endolymph resulting in expansion of the endolymphatic compartment and stretching of the neuroepithelial membranes of the inner ear. The pathophysiology is not fully understood but is likely to involve disordered endolymph metabolism that, in some, may be autoimmune mediated. There are many treatments available for Ménière's disease including intra-tympanic steroids or gentamicin, saccus surgery (drainage, duct clipping or duct avulsion), labyrinthectomy and vestibular nerve section. These treatments aim to improve the vertiginous element of symptoms and to not affect hearing or cause or increase tinnitus. Hearing loss is usually treated with appropriate aiding or, if profound, with cochlear implantation. Betahistine has recently been shown to be ineffective in Ménière's disease.

Labyrinthitis

Inflammation or infection of the inner ear structures is termed labyrinthitis. It results in acute failure of the vestibulocochlear system and presents with acute vertigo and profound sensorineural hearing loss. It should be differentiated from vestibular neuronitis in which only the vestibular system is affected. It may be of viral or bacterial aetiology. The most common viruses to be implicated are rubella, cytomegalovirus, mumps, measles and varicella (Ramsay Hunt syndrome). Bacterial labyrinthitis usually results from chronic otitis media or meningitis. There is no identifiable cause in some cases. Fibrosis of the cochlear duct can occur, especially with bacterial labyrinthitis.