# Sensorineural hearing loss

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# Sensorineural hearing loss (SNHL)

- **Sensorineural hearing loss** (**SNHL**) is a type of hearing loss in which the root cause lies in the inner ear or sensory organ (cochlea and associated structures) or the vestibulocochlear nerve (cranial nerve VIII)

- *Sensory* hearing loss often occurs as a consequence of damaged or deficient cochlear hair cells

- Hair cells may be abnormal at birth or damaged during the lifetime of an individual.

# Signs and symptoms

- Since the inner ear is not directly accessible to instruments, identification is by patient report of the symptoms and audiometric testing.
- 90% report having diminished hearing,
- 57% report having a plugged feeling in ear,
- 49% report having ringing in ear (tinnitus).
- half report vestibular (vertigo) problems.

# Causes

- Sensorineural hearing loss may be genetic or acquired (i.e. as a consequence of disease, noise, trauma, etc.)
- People may have a hearing loss from birth (congenital) or the hearing loss may come on later. Many cases are related to old age (age-related).

**1. Genetic**

- Hearing loss can be inherited. More than 40 genes have been implicated in the cause of deafness.There are 300 syndromes with related hearing loss, and each syndrome may have causative genes.

# 2.Congenital

- Infections:
  - Congenital rubella syndrome, CRS, results from transplacental transmission of the rubella virus during pregnancy. CRS has been controlled by universal vaccination (MMR or MMRV vaccine).
  - Cytomegalovirus (CMV) infection is the most common cause of progressive sensorineural hearing loss in children. It is a common viral infection contracted by contact with infected bodily fluids such as saliva or urine and easily transmitted in nurseries and thus from toddlers to expectant mothers. CMV infection during pregnancy can affect the developing foetus and lead to learning difficulties as well as hearing loss.
  - Toxoplasmosis, a parasitic disease affecting 23% of the population in the U.S., can cause sensorineural deafness to the fetus in utero.

# 3.Presbycusis

- Progressive age-related loss of hearing acuity or sensitivity can start as early as age 18, primarily affecting the high frequencies, and men more than women.
- Such losses may not become apparent until much later in life.
- Presbycusis is by far the dominant cause of sensorineural hearing loss in societies.
- Hearing loss that worsens with age but is caused by factors other than normal aging, such as noise-induced hearing loss, is not presbycusis, although differentiating the individual effects of multiple causes of hearing loss can be difficult.
- One in three persons have significant hearing loss by age 65; by age 75, one in two. Age-related hearing loss is neither preventable nor reversible

# 4.Noise

- Most people living in modern society have some degree of progressive sensorineural (i.e. permanent) noise-induced hearing loss (NIHL) resulting from overloading and damaging the sensory or neural apparatus of hearing in the inner ear.
- NIHL is typically a drop-out or notch centered at 4000 Hz. Both intensity sound pressure level (SPL) and duration of exposure, and repetitive exposure to unsafe levels of noise contribute to cochlear damage that results in hearing loss.
- The louder the noise is, the shorter the safe amount of exposure is.
- NIHL can be either permanent or temporary, called a threshold shift.

- Unsafe levels of noise can be as little as 70 dB (about twice as loud as normal conversation) if there is prolonged (24-hour) or continuous exposure..

- 125 dB (a loud rock concert is ~120 dB) is the pain level; sounds above this level cause instant and permanent ear damage.

- Noise and ageing are the primary causes of presbycusis, or age-related hearing loss, the most common kind of hearing loss in industrial society.
- The dangers of environmental and occupational noise exposure are widely recognized. Numerous national and international organizations have established standards for safe levels of exposure to noise in industry, the environment, military, transportation, agriculture, and other areas. Sound intensity or sound pressure level (SPL) is measured in decibels (dB).

| db Level | Example |
| --- | --- |
| 45 dB | Ambient noise level around the home |
| 60 dB | Quiet office |
| 60–65 dB | Normal conversation |
| 70 dB | City street noise at 25'[clarification needed] or average TV audio |
| 80 dB | Noisy office |
| 95–104 dB | Nightclub dance floor |
| 120 dB | Close by thunder or a loud rock concert |
| 150–160 dB | Gunshot from a handheld gun |

- the safe daily exposure amount at 85 dB (90 dB for OSHA) is 8 hours, while the safe exposure at 94 dB(A) (nightclub level) is only 1 hour.

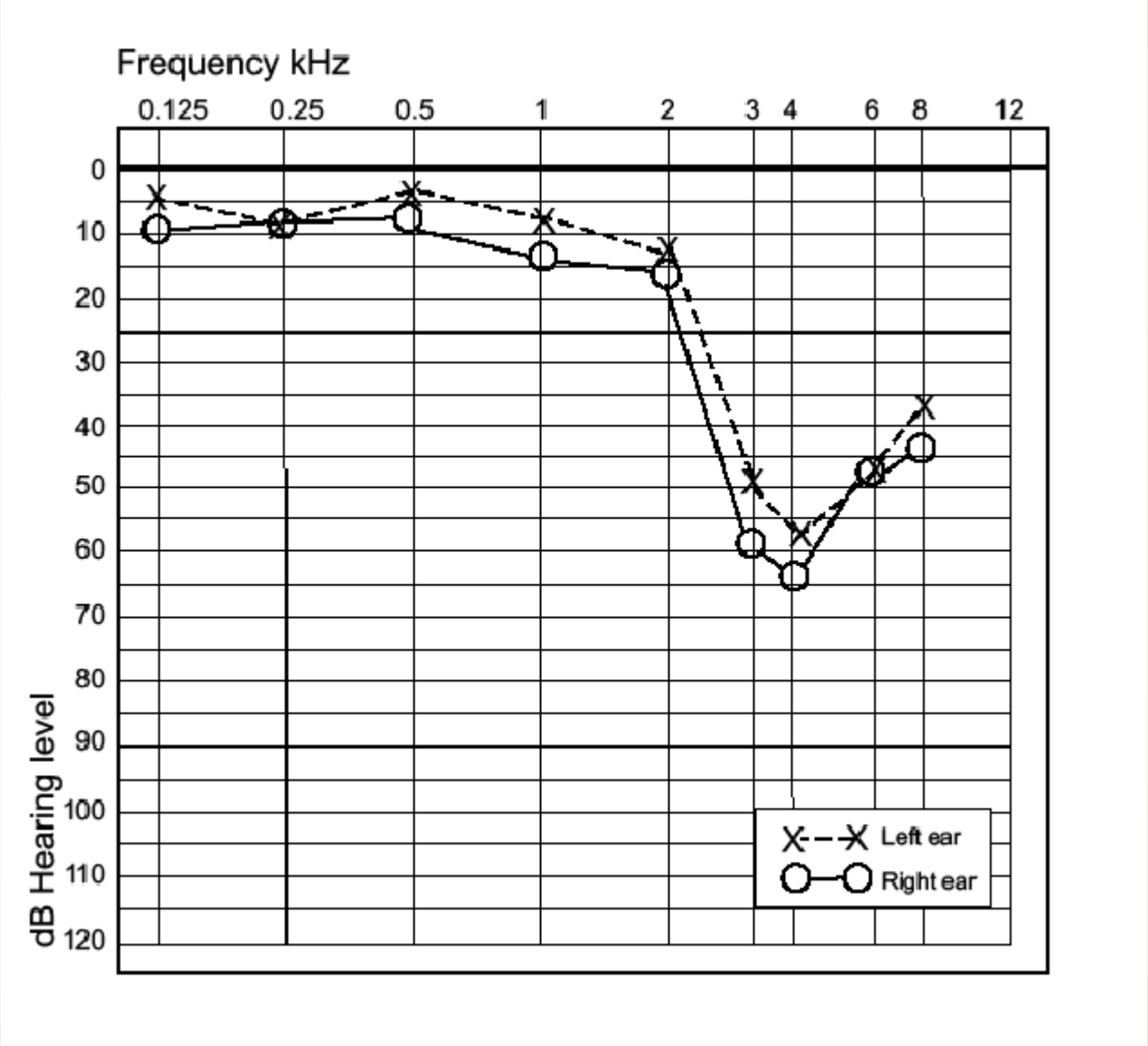
- Noise trauma can also cause a reversible hearing loss, called a temporary threshold shift. This typically occurs in individuals who are exposed to gunfire or firecrackers, and hear ringing in their ears after the event (tinnitus).

- **Ambient environmental noise**: Populations living near airports, railyards and train stations, freeways and industrial areas are exposed to levels of noise typically in the 65 to 75 dBA range. If lifestyles include significant outdoor or open window conditions, these exposures over time can degrade hearing.

- **Personal audio electronics**: Personal audio equipment such as iPods (iPods often reach 115 decibels or higher), can produce powerful enough sound to cause significant NIHL.

- **Acoustic trauma:** Exposure to a single event of extremely loud noise (such as explosions) can also cause temporary or permanent hearing loss. A typical source of acoustic trauma is a too-loud music concert.

- **Workplace noise:** Construction Industry Occupational Noise Exposure identify the level of 90 dB(A) for 8 hour exposure as the level necessary to protect workers from hearing loss.


Frequency kHz
0.125
0.25
0.5
1
2
3
4
6
8
12
0
10
20
30
40
50
60
70
80
90
100
110
120
dB Hearing level
Left ear
Right ear

# Disease or disorder

- Inflammatory
  - Suppurative labyrinthitis or otitis interna (inflammation of the inner ear)
- Diabetes mellitus
  - A recent study found that hearing loss is twice as common in people with diabetes as it is in those who don't have the disease.
  - who have prediabetes, the rate of hearing loss is 30 percent higher than in those with normal blood glucose.
  - It has not been established how diabetes is related to hearing loss. It is possible that the high blood glucose levels associated with diabetes cause damage to the small blood vessels in the inner ear, similar to the way in which diabetes can damage the eyes and the kidneys. Similar studies have shown a possible link between that hearing loss and neuropathy (nerve damage).

# Tumor

- Cerebellopontine angle tumour (junction of the pons and cerebellum) – The cerebellopontine angle is the exit site of both the facial nerve(CN7) and the vestibulocochlear nerve(CN8). Patients with these tumors often have signs and symptoms corresponding to compression of both nerves.
  - Acoustic neuroma (vestibular schwannoma) – benign neoplasm of Schwann cells affecting the vestibulocochlear nerve
  - Meningioma – benign tumour of the pia and arachnoid mater

- Ménière's disease – causes sensorineural hearing loss in the low frequency range (125 Hz to 1000 Hz). Ménière's disease is characterized by sudden attacks of vertigo, lasting minutes to hours preceded by tinnitus, aural fullness, and fluctuating hearing loss. It is relatively rare and commonly over diagnosed.

- Bacterial meningitis  may damage the cochlea – Hearing loss is one of the most common after-effects of bacterial meningitis. It has been estimated that 30% of bacterial meningitis cases result in mild to profound hearing loss. Children are most at risk: seventy percent of all bacterial meningitis occurs in young children under the age of five.

- Viral
  - AIDS patients frequently experience auditory system anomalies.
  - Mumps(epidemic parotitis) may result in profound sensorineural hearing loss (90 dB or more), unilaterally (one ear) or bilaterally (both ears).
  - Measles may result in auditory nerve damage but more commonly gives a mixed (sensorineural plus conductive) hearing loss, and can be bilaterally.
  - Ramsay Hunt syndrome type II (herpes zoster oticus)
- Bacterial
  - Syphilis is commonly transmitted from pregnant women to their fetuses, and about a third of the infected children will eventually become deaf.

- **Ototoxic and neurotoxic drugs and chemicals**

- Some over-the-counter as well as prescription drugs and certain industrial chemicals are ototoxic. Exposure to these can result in temporary or permanent hearing loss.
- Some medications cause irreversible damage to the ear, and are limited in their use for this reason. The most important group is the aminoglycosides (main member gentamicin).

# Head trauma

- There can be damage either to the ear itself or to the central auditory pathways that process the information  by the ears.
- People who sustain head injury are susceptible to hearing loss or tinnitus, either temporary or permanent.
- Contact sports like football (U.S. NFL), hockey and cricket have a notable incidence of head injuries (concussions).
- In one survey of retired  players, all of whom reported one or more concussions during their playing careers, 25% had hearing loss and 50% had tinnitus.

- **Iodine deficiency / Hypothyroidism**
  - Iodine deficiency and endemic hypothyroidism are associated with hearing loss.
- **Brain stroke**
  - Brain stroke in a region affecting auditory function such as a posterior circulation infarct has been associated with deafness.

# Pathophysiology

- Sensory hearing loss is caused by abnormal structure or function of the hair cells of the organ of Corti in the cochlea.
- Neural hearing impairments are consequent upon damage to the eighth cranial nerve (the vestibulocochlear nerve) or the auditory tracts of the brainstem.
- If higher levels of the auditory tract are affected this is known as central deafness.

- The fundamental role of the OHCs and the IHCs is to function as sensory receptors.
- The main function of the IHCs is to transmit sound information via afferent neurons. They do this by transducing mechanical movements or signals into neural activity.
- When stimulated, the stereocilia on the IHCs move, causing a flow of electric current to pass through the hair cells. This electric current creates action potentials within the connected afferent neurons.

# Hair cell damage

- SNHL is most commonly caused by damage to the OHCs and the IHCs.
- There are two methods by which they might become damaged. Firstly, the entire hair cell might die.
- Secondly, the stereocilia might become distorted or destroyed.
- Damage to the cochlea can occur in several ways, for example by viral infection, exposure to ototoxic chemicals, and intense noise exposure.
- Damage to the OHCs results in either a less effective active mechanism, or it may not function at all. OHCs contribute to providing a high sensitivity to quiet sounds at a specific range of frequencies (approximately 2–4 kHz). Thus, damage to the OHCs results in the reduction of sensitivity of the basilar membrane to weak sounds. Amplification to these sounds is therefore required, in order for the basilar membrane to respond efficiently. IHCs are less susceptible to damage in comparison to the OHCs. However, if they become damaged, this will result in an overall loss of sensitivity.

- **Vestibulocochlear nerve pathology**
- congenital deformity of the internal auditory canal,
- neoplastic and pseudo-neoplastic lesions, with special detailed emphasis on schwannoma of the eighth cranial nerve (acoustic neuroma),
- non-neoplastic Internal Auditory Canal/CerebelloPontine Angle pathology, including vascular loops

# Diagnosis[

**Case history**

- Before examination, a case history provides guidance about the context of the hearing loss.
- major concern
- pregnancy and childbirth information
- medical history
- development history
- family history
- **Otoscopy**

- **Differential testing**

# Differential testing

- Weber test, in which a tuning fork is touched to the midline of the forehead, localizes to the normal ear in people with unilateral sensorineural hearing loss.
- Rinne test, which tests air conduction *vs.* bone conduction is positive, because both bone and air conduction are reduced equally.

| *Criteria* | *Sensorineural hearing loss* | *Conductive hearing loss* |
|---|---|---|
| **Anatomical site** | Inner ear, cranial nerve VIII, or central processing centers | Middle ear (ossicular chain), tympanic membrane, or external ear |
| **Weber test** | Sound localizes to normal ear in unilateral SNHL | Sound localizes to affected ear (ear with conductive loss) in unilateral cases |
| **Rinne test** | Positive Rinne; air conduction > bone conduction (both air and bone conduction are decreased equally, but the difference between them is unchanged). | Negative Rinne; bone conduction > air conduction (bone/air gap) |

- Tympanometry
- Audiometry
- Magnetic resonance imaging

# Treatment

Treatment modalities fall into three categories: pharmacological, surgical, and management. As SNHL is a physiologic degradation and considered permanent, there are as of this time, no approved or recommended treatments.

pharmacological treatment options are very limited and clinically unproven

Profound or total hearing loss may be amenable to management by cochlear implants, which stimulate cochlear nerve endings directly.

Hearing aids are specifically tuned to the individual hearing loss to give maximum benefit