Types and causes of hearing loss -2-
SNHL

- Refers to hearing loss resulted from the damage of the sensori-neural mechanisms.
- It results from disorders of the cochlea, 8th cranial nerve or cochlear nuclei.
• Usually, it cannot be treated medically or surgically, i.e. **Irreversible**, except in some few cases like Meniere’s disease, perilymphatic fistula or sudden hearing loss that caused by interruption of cochlear blood supply.

• Generally it’s managed through rehabilitation or habilitation includes amplification.

• It’s either **congenital** or **acquired**.
Types of SNHL based on time of occurrence

1) Congenital

- It’s either;
  - Genetic (may associated with other anomalies, syndrome)
  - Non-genetic (most of the hearing loss).
A. Non-genetic

- It’s usually caused by peri-natal infections, most are non-bacterial, or peri-natal causes like anoxia.

- Most well-known infections are toxoplasmosis (TROCHS), bacterial meningitis, Maternal rubella (German measles), Cytomegalovirus (CMV), herpes simplex and syphilis.
• **Toxoplasmosis**, causes slowly progressive SNHL.

• **Maternal rubella (German measles)**, SNHL is one of its signs.

• **Herps simplex**, no hearing loss was reported. However, studies showed that the sensory cell of the inner ear are affected.
• **Syphilis**, SNHL and vestibular disorders are of its signs.

  ➢ The SNHL is usually bilateral, sudden onset, both cochlear and neural areas are affected, and it occurs at any time from birth to 60 years-old.

• **Cyto-megalo-virus**, it’s either **symptomatic** or **asymptomatic**.
CMV

- Unilateral or bilateral SNHL is evidenced in 7-17% of asymptomatic cases.

- Unilateral or bilateral SNHL is evidenced in 30% of the symptomatic cases.

- The hearing impairment with CMV is generally progressive, and may be associated with CP and RCP and central auditory disorders.
B. Genetic

- It may occur in isolation or as part of a syndrome.

- Some examples of syndromes: Waardenburg, Klippel-Feil, Usher’s, Down, Treacher-Collins, Albinism…etc
2) Acquired

- It's the postnatal acquired hearing impairment.

- It’s either with *unknown causes (Idiopathic)* or *caused by external factors* (Trauma, drugs, noise, age, infections, systemic disease, degenerative disorders, tumors).
Idiopathic causes

Meniere’s disease

- It’s a disease that affects the membranous labyrinth.

- It’s a hydropic distention of the endolymphatic system.

- It is characterized by sudden onset of vertigo, tinnitus, vomiting and unilateral hearing loss.

- It has **three types:** classic, cochlear, and vestibular.
1. **Classic Meniere’s**

- Its main characteristics are:
  - Progressive, Fluctuating SNHL of cochlear origin
  - Pressure and aural fullness
  - Roaring tinnitus
  - And vertigo.
2. Cochlear Meniere’s

- It is classic type without vertigo.

3. Vestibular Meniere’s

- It is classic type without hearing loss.

- Cochlear and vestibular type may later develop to the classic one.
Meniere’s disease **possibly caused by**,

- Anxiety,
- Allergy,
- Trauma,
- Surgery,
- Syphilis,
- Hypothyroidism,
- Low blood sugar
• In the **early stages** of the disease, the hearing loss affects the **low frequencies** and later on it will affect the **high frequencies**.

• Recovery is possible in the early stages. However, the symptoms will persist lately.

• Sedatives, tranquilizers, and vestibular suppressant use as a **treatment** and placebo found to be effective in some cases

• The disease is **unilateral in 85%** of the cases.
Audiogram (early stages of Meniere’s disease)
Sudden Idiopathic

- It’s a sudden onset SNHL.

- The diagnosis is confirmed when all other possible known causes ruled out.

- It may be cochlear or/and 8th nerve problem and it may accompanied by tinnitus or vertigo.

- It’s usually unilateral.

- Hearing impairment recovery or improvement is possible in the majority of the cases.
Semicircular Canal Dehiscence

- It is also known as superior semicircular canal dehiscence
- It induced by intense sound or change in ME pressure
- It’s thinning or weakening of superior SCC bone
Signs and symptoms

- Dizziness
- Vertigo
- Disequilibrium
- Hearing test shows low frequency CHL
- Tympanometry normal and acoustic reflexes present
Autoimmune Inner-Ear Disease

- Inflammatory conditions when the immune system makes the body attack its own tissues
- The body fail to distinguish its tissues from bacteria, viruses or any other micro-organisms
- Mainly attacks IE

Signs and symptoms
- bilateral fluctuating and progressive SNHL
- Tinnitus
- Vertigo
- Aural fullness
Non-idiopathic causes

Peri-lymphatic fistula

- It is the rupture of the round or oval window that causes by head trauma, congenital origin, diving, physical exertion (heavy exercises, sneezing), acoustic trauma or idiopathically.

- It’s usually accompanied by sudden or progressive SNHL of cochlear origin.

- Some cases may not reveal hearing loss.
Head trauma

- Hearing loss related to head injury with notch at 3000 to 6000 Hz showed in the audiogram.

- TM damaged and ME mechanisms affected together with the IE.

- If the fracture extended to the cochlea, hearing may lost totally or severe to profound affected.

- Hearing might be also affected even if the skull intact.
Ototoxicity

- Some drugs may cause hearing loss and/or vestibular abnormalities.

- The drugs that affect the cochlear functions are **cochleotoxic**.

- The drugs that affect the vestibular functions are **vestibulotoxic**.

- Some are both cochleotoxic and vestibulotoxic.
• The severity of the Ototoxicity depends on:

- Drugs concentration in the blood stream,
- Usage duration,
- Individual susceptibility,
- Whether other ototoxic drugs have been used,
- Renal functionality
- And the presence of any noise exposure.
• Examples of ototoxic medications:

- Aminoglycoside (streptomycin, gentamycin, neomycin..etc)
- Salicylates (Alka Seltzer, anacin…)
- Cisplatinum (cancer treatment).
Cochleo-toxic drugs

- Are usually associated with generally bilateral, symmetrical SNHL of cochlear origin. Although, some unilateral, asymmetrical hearing loss has been reported.

- Tinnitus also has been reported and it’s stated to be permanent except with Salicylates.
Vestibulo-toxic drugs

- May cause disequilibrium, ataxia unsteadiness, and/or vertigo.

- The vestibular damage is generally bilateral and permanent.
Noise

- **Noise-induced hearing loss**, is a term that generally refers to the hearing impairment from **long-term, repeated** exposure to noise. Like occupational noise (military) or recreational activities (snow-mobiling).

- **Acoustic trauma** is a term that describes the hearing loss from **single exposure to a short-term noise**.
• **Initial exposure** to the harmful noise level leads to *temporary threshold shift (TTS)* i.e. threshold elevation that usually recovers after a short period (several hours to few days).

• **Continued noise exposure**, the TTS developed to *permanent threshold shift (PTS)*.

• Hearing loss from noise is bilateral, symmetrical SNHL of primarily cochlear origin.
It's audiometeric configuration shows a notch between 3000 to 6000 Hz.
Ageing

- **Presbycusis**; is the hearing loss that associated with ageing.

- It’s the result of normal degeneration of auditory system.

- The hearing loss usually bilateral and SNHL of cochlear or neural origin.

- It’s a slowly progressive hearing loss that first affecting the HF then the LF (sloping configuration).
Many studies showed sex differences (from male to female).

- It happens earlier in males than females.
- Its magnitude higher in males.
- Sloping in the audiogram greater in males.
Audiogram for typical presbycusis
Tobacco smoke

- Generally Tobacco smoke leads to many serious illnesses like cancer.
- It has been discussed that it is one of the contributors to OME and CHL.
- Nowadays, many studies showed that it is contributory factor in permanent SNHL.
- Recent works prove that it is not only affecting OHCs (SNHL) but also leads to CAPDs.
Radiation-induced hearing loss

- Radiotherapy for brain tumors and head and neck cancers, can harmfully affects the auditory system from EAM through central auditory pathway

- Acute complication to ME system occurs in 40% of the cases and it leads to no-permanent CHL

- Patients whose, radiotherapy extended to the cochlea, experiencing permanent SNHL
**Patients who are scheduled to have radiation should have:**

- Pre-radiation baseline auditory assessment
- Repeat evaluation at the end of the radiotherapy
Cochlear hearing loss following surgical complications

- 1-2% of patients with stapedectomy might suffer cochlear hearing loss

- Excessive bleeding or other complication following ME surgeries may account for cochlear hearing loss

- Some total hearing loss following ear operation without any clear cause, some of the operation were perfectly done
Auditory Nerve tumor

- Most of auditory nerve tumors are benign

- It usually arises from the sheaths that cover the vestibular branch of the 8th cranial nerve

- Some neuro-otologists believe that the term **acoustic neuroma** is the best descriptive term for such tumors as it arises from peripheral cells of the nerve
However, it’s been suggested that **vestibular schwannoma** is more useful as most of tumors arise from the schwan cells that form the sheath of the vestibular branch of the acoustic nerve.

It cannot be diagnosed from the typical audiometric configuration.

Using of a battery of tests includes Pure Tone audiometry, Speech audiometry, Acoustic Reflexes, ABR and OAE, is the proper way for the accurate audiological diagnosis.
Signs and symptoms

- Tinnitus
- Dizziness
- Speech recognition difficulties
- Progressive unilateral hearing loss
- Facial weakness
- Alteration in the sense of smell and taste
- Later, if it grows very large, speech and swallowing difficulties
Treatment

- Previously, Gamma knife
- Today, cyber knife
Mixed HL is the hearing loss that caused by damage in both conductive and sensori-neural mechanisms.

It’s caused by combination of the causes of both CHL and SNHL.
Central Auditory Disorders

- It refers to the disorders of central auditory system.

- It results from lesions of the auditory brainstem pathway or auditory cortex.

- It characterizes by auditory difficulty even with normal hearing sensitivity.
Signs and symptoms of CAD with organic lesion to the CANS and normal hearing

✓ Tinnitus.
✓ Auditory hallucination.
✓ Difficult to understand speech in noisy or reverberant environment.
✓ Difficulty in understanding complex commands.
✓ Distractibility.
✓ Localization problems.
Even if surgery can repair the organic lesion, the CAD could not be elevated. Speech-language therapy might also be required.

CAD could be **congenital** or **acquired**.
I. Congenital

- Can caused by:
  - Toxoplasmosis,
  - Hyperbilirubinemia,
  - Birth trauma,
  - Asphyxia,
  - Erytheroblastosis fetalis and
  - Any factors that cause congenital SNHL can cause CAD.
2. Acquired

- CAD is one of the signs and symptoms presented by patients with temporal lobe lesions and those with brainstem pathology.

- Intra-axial brainstem tumor and temporal lobe tumor are usually associated with normal hearing sensitivity or mild bilateral symmetrical, HF SNHL.
Multiple sclerosis, degenerative disease affects CNS, can cause SNHL of 8th nerve origin which may be bilateral or unilateral.

When the cortical or brainstem auditory pathway involved, CAD may be presented.