Disorders of the Inner Ear

Inner ear disorders

- Disorders of hearing produced by abnormality or disease of the cochlea probably constitute the largest group of hearing losses called sensory/ neural.
- Much of cochlear hearing loss originates with pathology at the level of the hair cells.

Dysacusis

- When damage or abnormality occurs in the cochlea, loss of hearing sensitivity is not the only symptom.

– A common complaint of patients with sensory or neural hearing loss is not so much that they cannot hear, but that they have difficulty understanding speech.

- This speech- recognition problem has been called dysacusis
- Patients with greater cochlear hearing losses have more dysacusis..

Disorders in the inner ear

- Can be:
  - Congenital, examples:
    - Inherited hearing loss
    - Prematurity, lack of oxygen at birth, or other birth traumas
    - Damage to the unborn baby due to a virus, such as German measles (rubella) or CMV (cytomegalovirus)
    - Jaundice, particularly when serious enough to require blood transfusion.

Disorders in the inner ear

– Acquired, examples:

- Excessive noise exposure
- Viruses, such as mumps and measles
- Drugs which can damage the hearing system
- Head injuries
- The ageing process
- Diseases such as meningitis and Meniere's disease

Prenatal Genetic factors (hereditary)

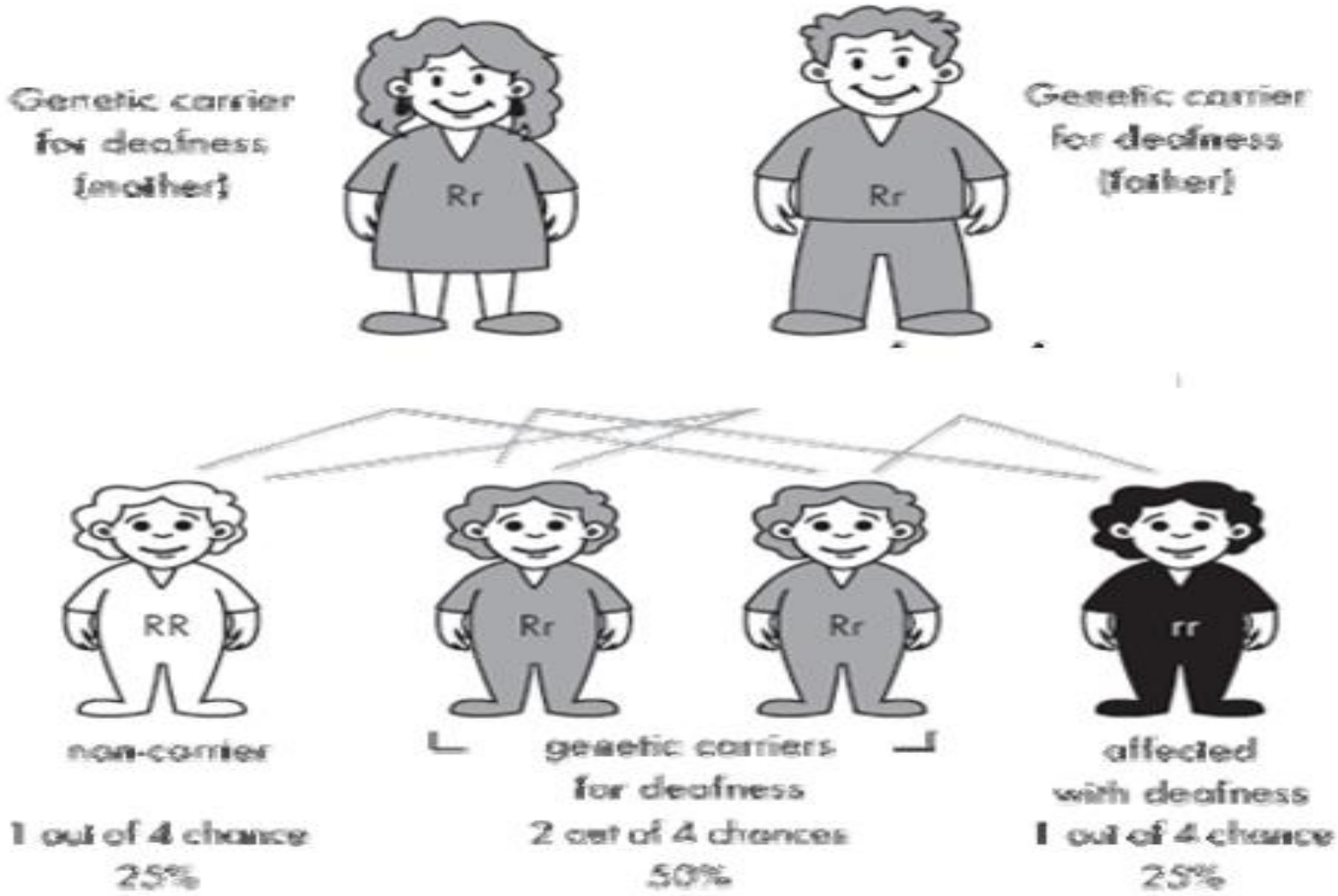
- Are thought to cause more than 50% of all hearing loss.
- Hearing loss from genetic defects can be present at birth or develop later on in life.
- Most genetic hearing loss can be described as autosomal recessive or autosomal dominant.

Genetic factors (hereditary)

- – In **autosomal recessive hearing loss**, both parents carry the recessive gene and pass it along to the child. Parents are often surprised to discover their child has a hearing loss because they are not aware that they are carrying a defective gene. This type of inheritance pattern accounts for about 70% of all genetic hearing loss.

Hereditary disorders

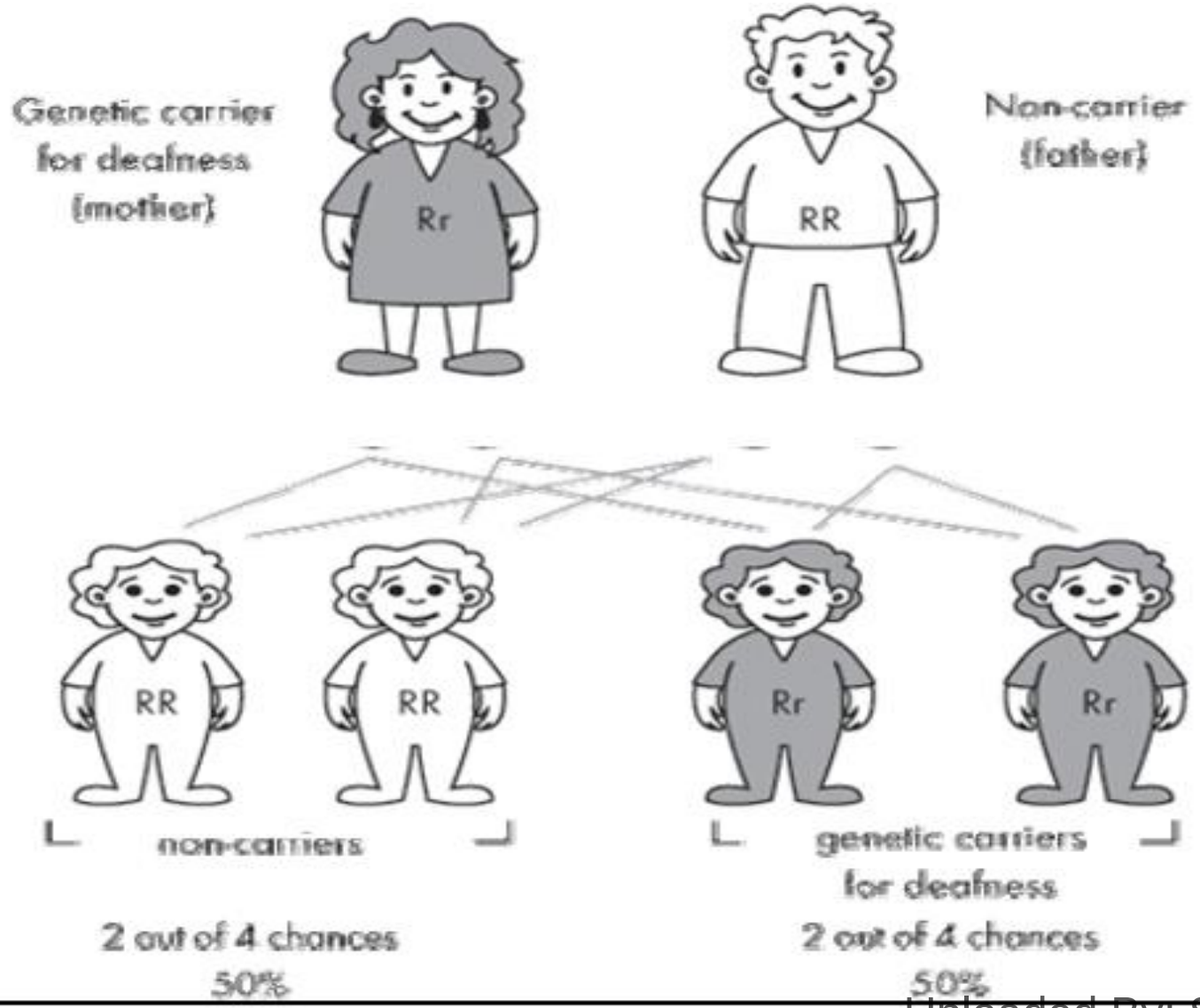
r= faulty gene copy
R= working gene copy



Genetic factors (hereditary)

- – An **autosomal dominant hearing loss** occurs when an abnormal gene from one parent is able to cause hearing loss even though the matching gene from the other parent is normal. The parent who is carrying the dominant gene may also have hearing loss as well as other signs and symptoms that make up a genetic syndrome. The autosomal dominant pattern accounts for 15% of all genetic hearing loss cases.

Hereditary disorders



Prenatal causes

- Syndromes e.g.: Asher syndrome
- Rh incompatibility e.g.: resulted in cerebral palsy
- Toxic drugs e.g.: thalidomide (tranquilizing drug)
- Rubella
- Cytomegalovirus (CMV)

Perinatal causes

- Prenatal causes of hearing loss are those that **occur during the process of birth itself**. Such causes frequently produce multiple handicaps.
- Anoxia: deprivation of oxygen to important cells, which alters their metabolism and results in damage or destruction.
- Accumulations of toxic substances in the mother's bloodstream may reduce the passage of oxygen across the placenta, which also results in anoxia. The fetus may also suffer from damage produced by the toxic substances themselves.
- Prematurity: infants less than 1500 grams (3.5 pounds) at birth, they are considered premature. – Prematurity is often associated with multiple births, and both are associated with sensory hearing loss.
- Trauma to the fetal head during delivery.

Postnatal causes

- Postnatal causes of cochlear hearing loss are any factors occurring after birth. E.g.

– Otitis media: the toxins from the bacteria in the middle ear may enter the inner ear by way of the round or oval window, or pus may enter the labyrinth from the middle ear or from the meninges, the protective covers of the brain and spinal cord. Bacterial meningitis, inflammation of the meninges, may cause total deafness.

– Viral infections have been identified as the causative factors in cochlear hearing loss. These infections include measles, mumps, chickenpox, influenza, and viral pneumonia.

– Infections of the kidneys may result in the deposit of toxic substances in the inner ear. Kidney disease may prevent medications from being excreted, thereby raising their levels in the blood abnormally high and producing ototoxicity.

Toxic Causes of Cochlear Hearing Loss

- **Side effects of antibiotics**

– Most noted among the drugs that are:

• Cochleotoxic (i. e., cause hearing loss) are viomycin, neomycin, kanamycin, and dihydrostreptomycin.

– Because hearing losses ranging from mild to profound may result from the use of these drugs, it is hoped that they are not prescribed unless it is fairly certain that other drugs, with fewer or less severe side effects, will not be just as effective.

• Vestibulotoxic drugs (those that are known to affect the vestibular organs) include streptomycin and gentamycin.

Toxic Causes of Cochlear Hearing Loss

- Other drugs used to treat certain medical conditions. E.g. *quinine* is a drug that has long been used to combat malaria and to fight fever and reduce the pain of the common cold.

– Many patients who have taken this drug have complained of annoying tinnitus and hearing loss. Although with less frequency, quinine is still prescribed for certain disorders.

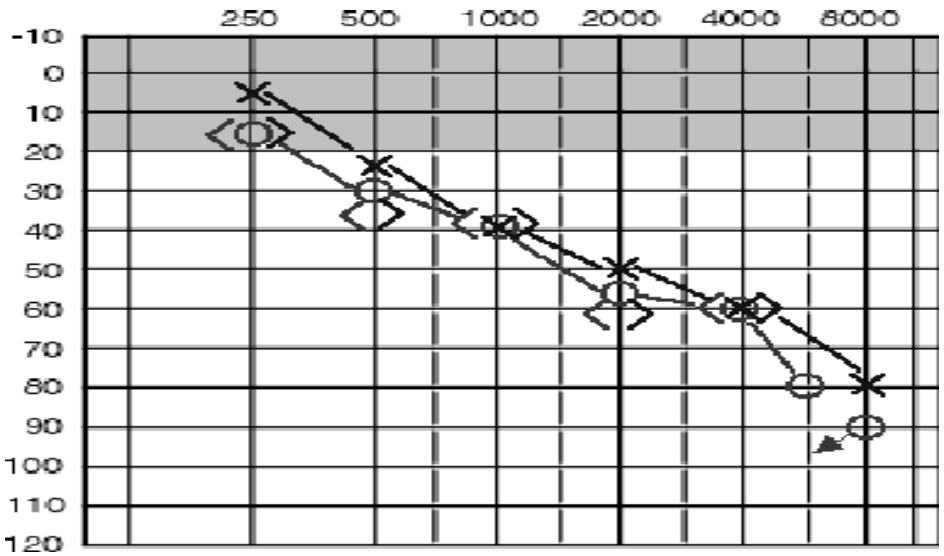
– Other drugs that have been associated with hearing loss include aspirin, certain diuretics, nicotine, and alcohol

- Using extended **high- frequency audiometry** can assist in detecting early hearing loss.

Audiological profile

- Tinnitus or ringing in one ear or both ears.
- Dizziness, loss of balance.
- Hearing loss usually occurs in both ears (first occurs at high frequencies –don't affect speech or language-, but if this impairment is not detected, the hearing loss will affect all frequencies and speech recognition will be affected.
- It can lead to deafness.
- Generally, the degree of

hearing loss is similar on both sides.



Radiation- Induced Hearing Loss

- The use of radiotherapy for the treatment of brain tumors and head and neck cancers can adversely affect any of the auditory structures.

– Nearly one- third of patients whose radiotherapy extends to the cochlea experience permanent sensory hearing loss.

- Given the delayed onset of radiation- induced hearing loss and its documented progression, regular monitoring of hearing status is essential.

Noise- Induced Hearing Loss

- Exposure to high- level sounds:

– Temporary threshold shift: cases in which hearing thresholds improve after an initial impairment following noise.

– Permanent threshold shift: irreversible losses.

- A number of agents may interact with noise to increase the danger to hearing sensitivity. E.g. aspirin, which has been known to produce reversible hearing loss after ingestion, synergizes with noise to produce a greater temporary threshold shift than would otherwise be observed.

Noise- Induced Hearing Loss

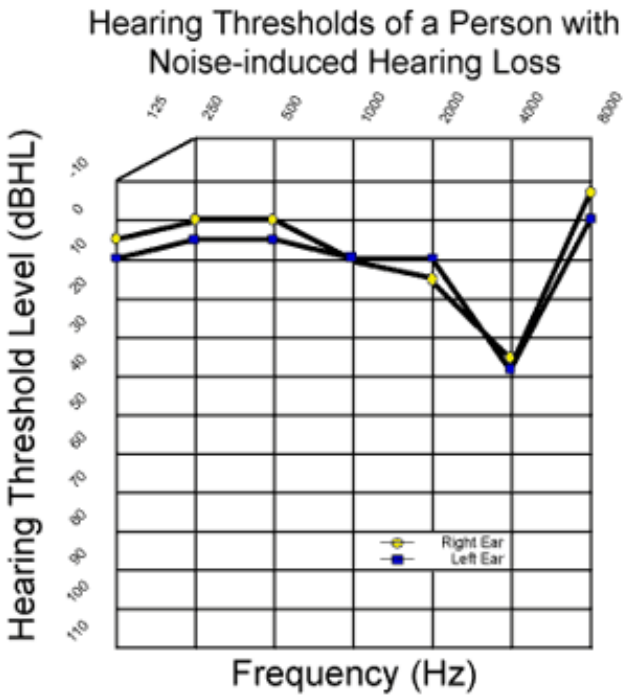
- the Occupational Safety and Health Administration (OSHA) (1983) has recommended a scale on which the time that a worker may be safely exposed to intense sounds is decreased as the intensity of the noise is increased.

  – Under this rule, the maximum exposure level before hearing conservation measures must be implemented is 85 dBA for an eight-hour work day. For every 5 dB increase in noise above 90 dBA, the allowable exposure time is cut in half.

Audiological profile

- Deficiency to hear high frequency sounds, like bird sounds.
- Muffled or distorted speech, they hear the sounds softer than they used to be.
- Tinnitus.
- Feeling fullness or pressure in the ear.
- Hypersensitivity to certain sounds (certain sounds are very bothersome or create pain)
- Hyperacuses: when we hear the sounds louder than they should be.
- Dizziness and vertigo, if the damage reaches the vestibular system.
- Diplacuses (double hearing), hearing eco-like sounds, although this is rare.
- High frequency notch that could be at 3 – 6 kHz, usually at 4 kHz.

Typical Audiogram of Noise Induced Hearing Loss



Sudden idiopathic/neural hearing loss

- A hearing loss that may develop over the course of few days or occur seemingly instantaneously with many patients claiming that they awoke from sleep in the morning to find that their hearing had changed.
- Usually unilateral loss
- More frequent in adult patients

Sudden idiopathic/neural hearing loss

- Causes\ Suggestions
  - Autoimmune diseases
  - Viral or other infections
  - Vascular disorders
- Therapy most often vasodilation.

Autoimmune inner ear disease

- Is an inflammatory conditions that occur when the immune system causes the body to attack its own tissues that are mistaken for bacteria, viruses, or cells from other organisms.

– Specifically attacks the inner ear.

– Rare disease probably accounting for less than 1% of all cases of hearing impairment or dizziness.

– Manifested as bilateral fluctuating and progressive sensory hearing loss which may occur over several months.

- Patients may feel fullness in the ear (aural fullness) , Vertigo and Tinnitus

- Treatment :Steroids can be administered

Presbycusis

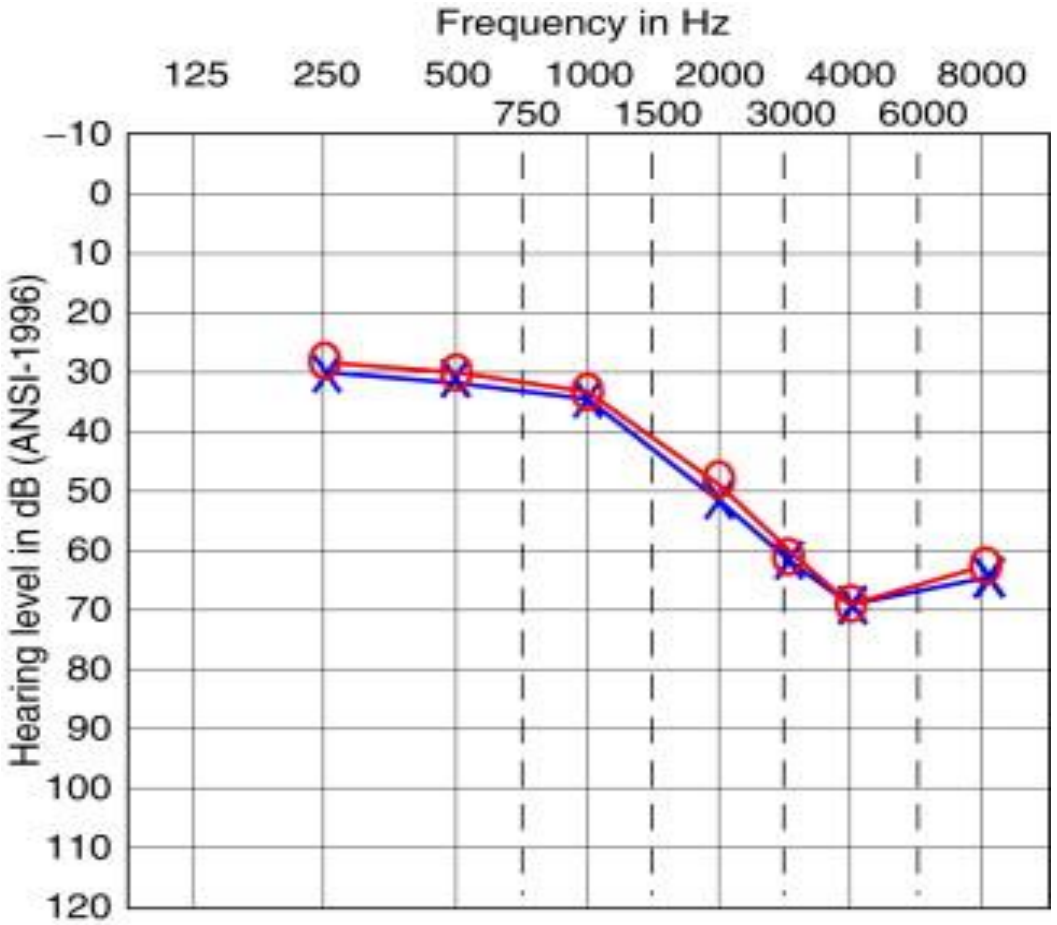
- Presbycusis is hearing loss due to age
- Expected in men by the early 60s and women by the late 60s
- Patients have difficulty in speech recognition

Presbycusis

- Physiologic effects of aging
  - Sensory Presbycusis
    - Degeneration of hair cells & cochlear fibers
    - Sloping, slowly progressive high-freq hearing loss (HL)
  - Neural Presbycusis
    - Loss of cochlear neurons
    - High-freq HL with poor word recognition abilities
  - Strial (metabolic) Presbycusis
    - Degeneration of the stria vascularis
    - Flat HL with good speech recognition
  - Mechanical Presbycusis
    - Alterations to the cochlear mechanics caused by thickening & stiffening of basilar membrane
    - Gradually sloping, high-freq HL with average speech recognition

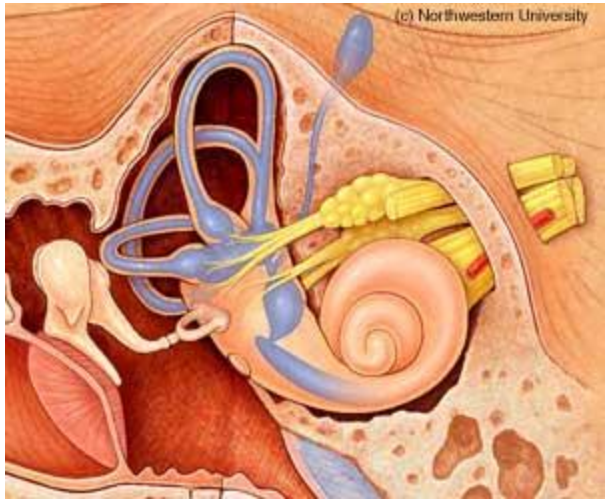
Symptoms

- Difficulty hearing conversations especially in noisy environments.
- Trouble understanding speech, the perception that others are mumbling
- Increased volume settings on electronic devices
- A decreased ability to hear high-frequency sounds like the ringing of a phone or doorbell.
- May complain of tinnitus or ringing in ears – not all
- May exhibit of vertigo – not all
- Even though it is SNHL, but middle and outer ear may be affected (impacted cerumen)

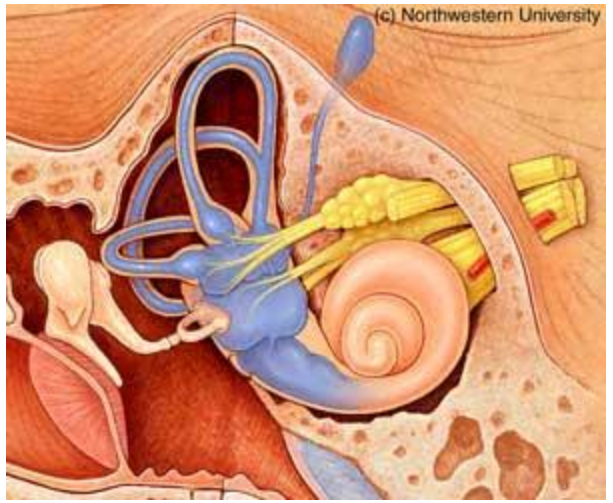


Menieres Disease

- Causes:
- Abnormal accumulation of fluids in the inner ear.
- Overproduction (by stria vascularis) or under absorption (by endolymphatic sac) of endolymph



normal



hydrops

Menieres Disease

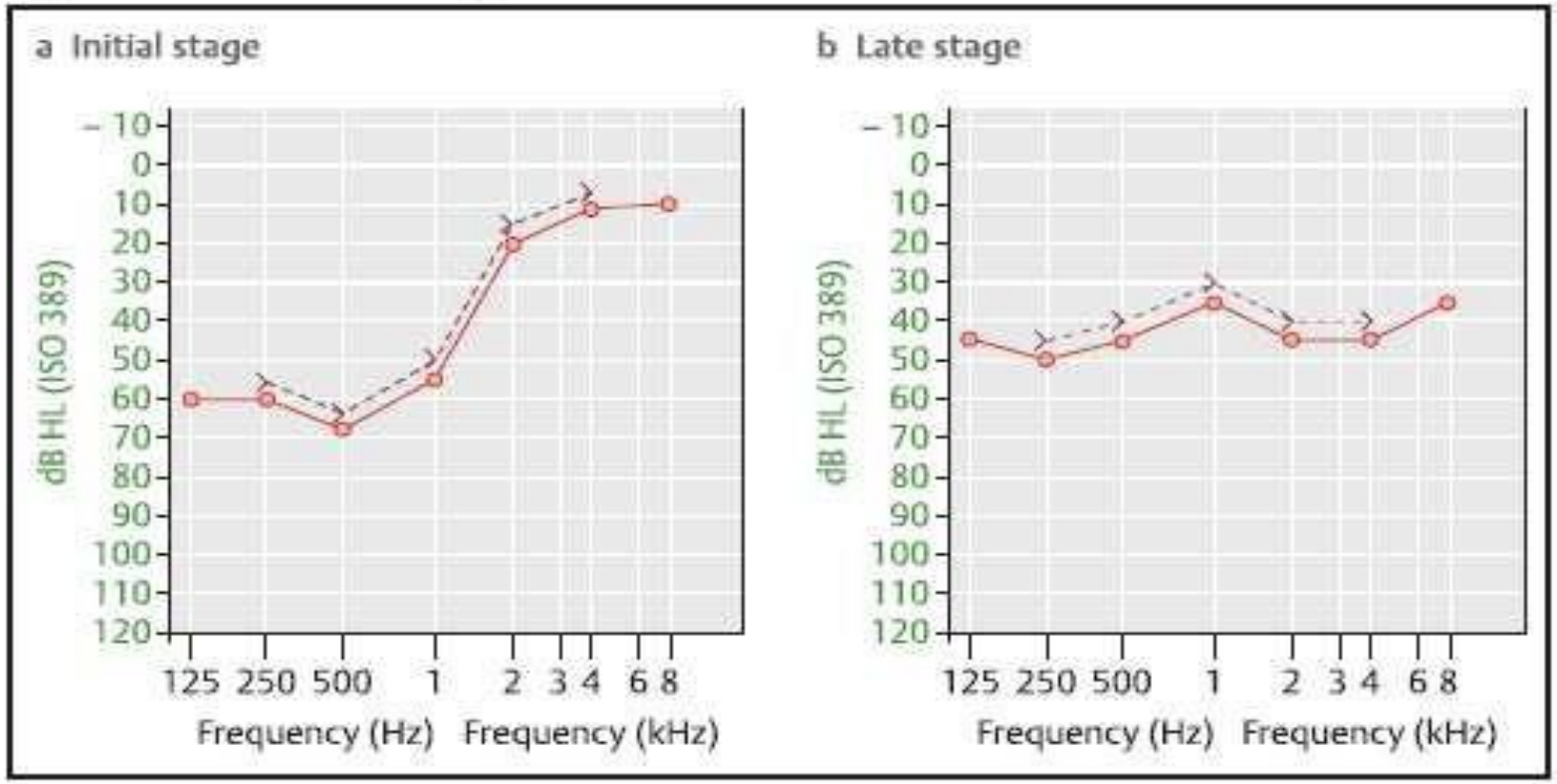
- Symptoms:

- Fluctuating hearing loss and dysacusis.
- Tinnitus, which is continuous but of varying intensity and low frequency (roaring).
- Sensation of aural fullness.
- Attacks of rotary vertigo, nausea, and vomiting, which last for hours.
- Unsteadiness and dizziness may be present shortly before and long after the attacks.
- The tinnitus and hearing loss may change before or during the attack.
- Most commonly, the tinnitus becomes louder, while hearing becomes poorer.
- Hearing generally improves again following the attack.

Treatment :

Diet change , medications or surgeries

Fig. 13.15 Pure-tone audiograms in Ménière's disease



Acoustic Neuroma

- Acoustic neuromas (AN), known as vestibular schwannomas, cerebellopontine angle tumors, are regularly benign, slow-growing tumor that develops from the balance and hearing nerves supplying the inner ear (vestibulocochlear nerve).
- In this case there is an overproduction of Schwann cells, leading to the formation of a mass or growth called a Schwannoma or neurofibroma.
- If the tumor is located inside the internal auditory canal, it is referred to as intracanalicular, while if it is outside the canal, it is called extra canalicular.
- Their incidence is around 1 in 100,000 individuals.
- Should be considered for assessment in case of any **unilateral** Otologic/ vestibular symptoms.

Signs and symptoms

- Asymmetric hearing loss. (usually unilateral).
- Distorted speech perception
- Aural fullness
- Impaired balance (in some cases)
- Facial pulsy (in some cases)
- Tinnitus in the impacted ear.

Treatment

- First = Observation

Regular hearing tests and MRI to monitor the size of tumor

- Depending on the size and effects of vestibular schwannoma on
- Hearing, and balance one of the following intervention can be chosen:

- Micro-surgery

- Radiation therapy (reduce size and limit growth)

- The removal of tumors affecting the hearing, balance, or facial nerves can sometimes make the patient's symptoms worse because these nerves may be injured during tumor removal.

The End