SENSORINEURAL HEARING LOSS

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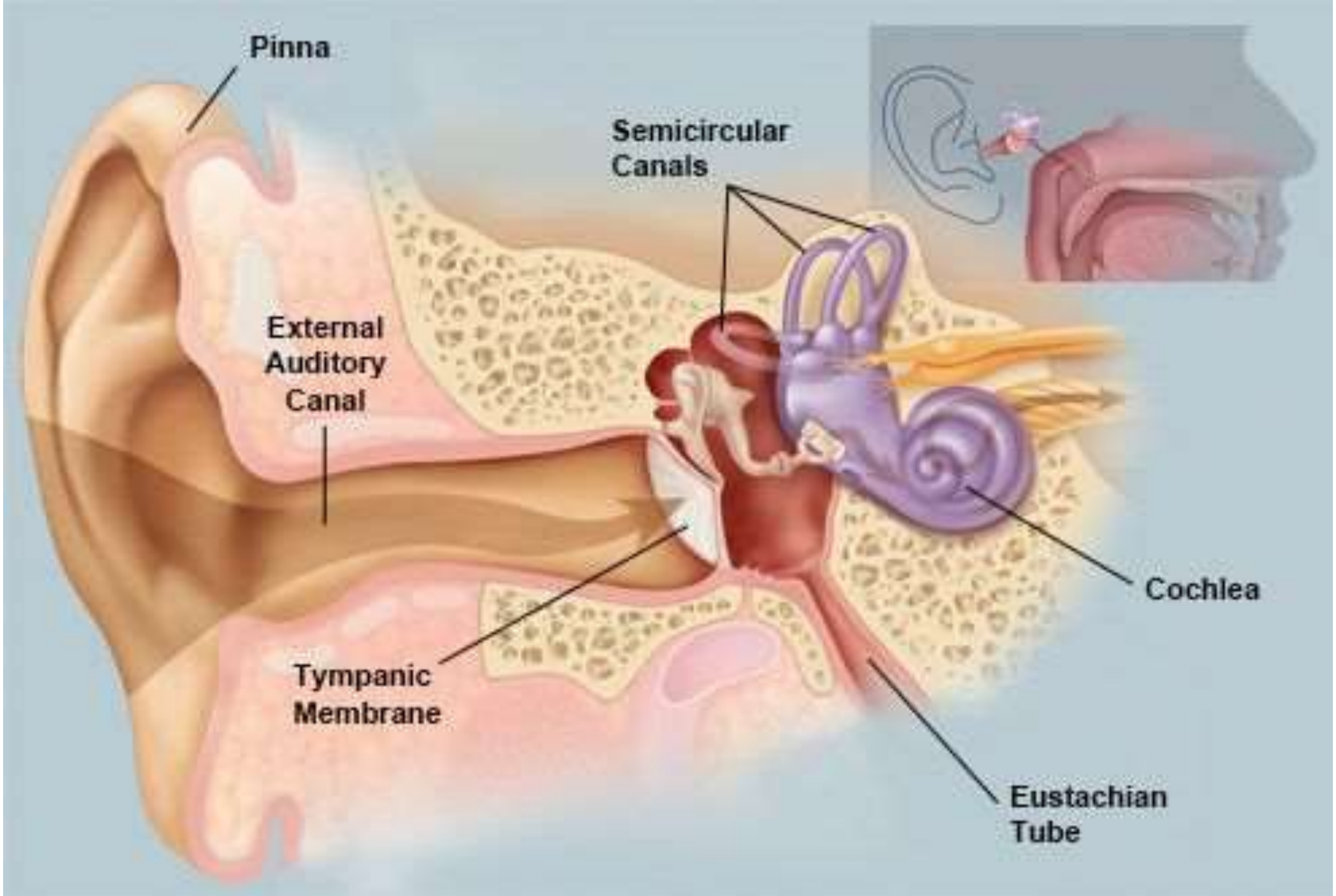
NMCH.


Pinna
Semicircular Canals
External Auditory Canal
Tympanic Membrane
Cochlea
Eustachian Tube

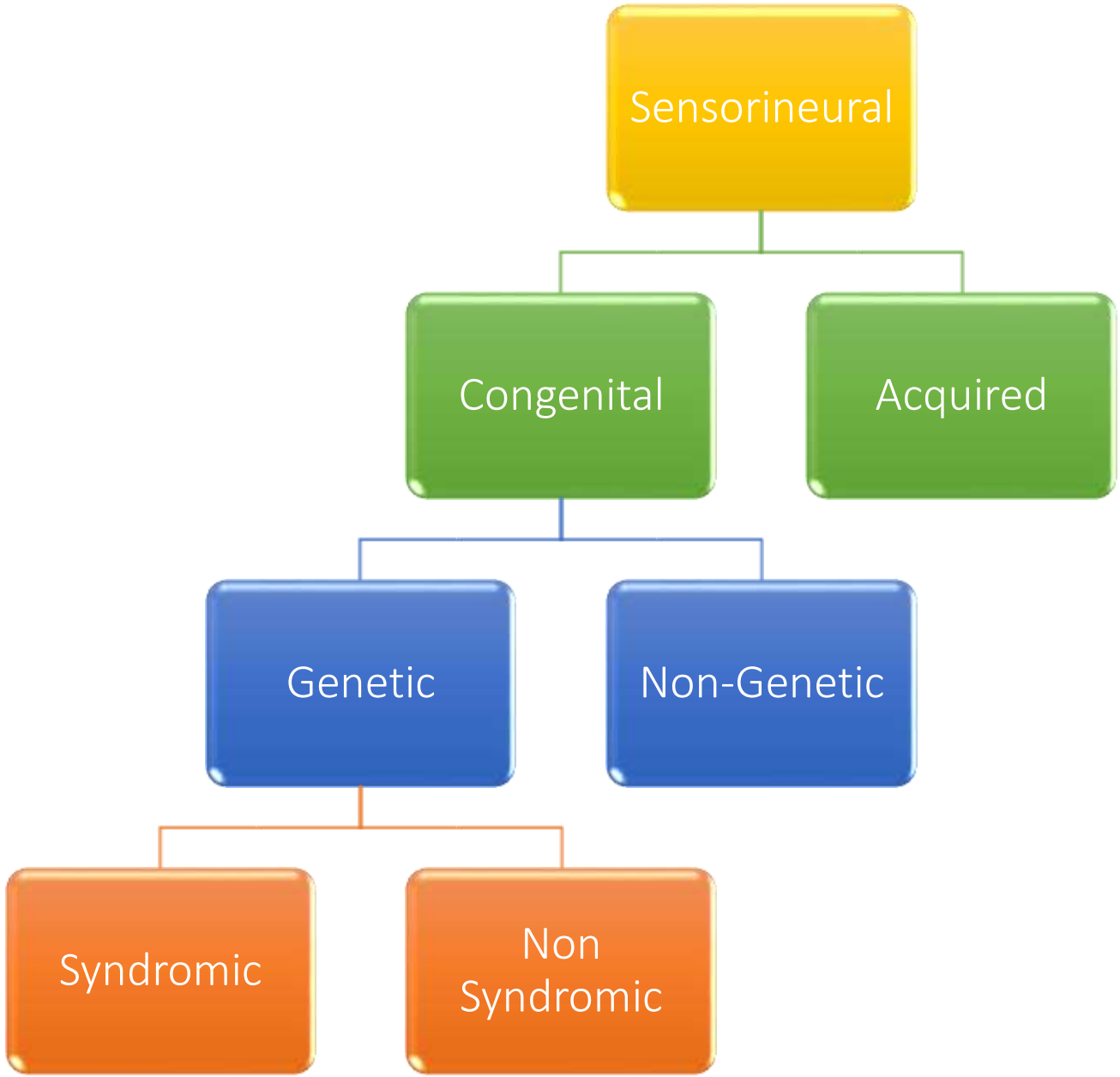
INTRODUCTION -

Sensorineural hearing loss (SNHL) is caused by damage to the structures in the inner ear or auditory nerve along the auditory pathway.

Sensorineural hearing loss (SNHL) results from lesions of the

- cochlea,
- VIII th nerve or central auditory pathways.
- Speech discrimination is poor.

CLASSIFICATION



CAUSES of SNHL

CONGENITAL- SYNDROMIC

1. Usher syndrome -• Retinitis pigmentosa
• Night blindness

2. Jarvell and Lange Nielson syndrome -• Repeated syncopal attacks
• Prolonged QT interval in ECG

3. Pendred syndrome -• Goiter (nontoxic) usually evident before puberty along with hypothyroidism

4. Alports syndrome- with nephritis

CONGENITAL-NONSYNDROMIC causes of SNHL

- **Michael's aplasia**: There is *complete absence of bony and membranous labyrinth*. Even the petrous apex is absent but external and middle ears may be completely unaffected.
- **Mondine's dysplasia**: incomplete development of bony and membranous labyrinth
- **Scheibe's** : cochleosaccular aplasia with normal bony labyrinth
- **Bing Sieben Mann dysplasia**: malformation of membranous labyrinth

CONGENITAL-NONGENETIC

Infections

Rubella

CMV

Measles

Toxoplasma

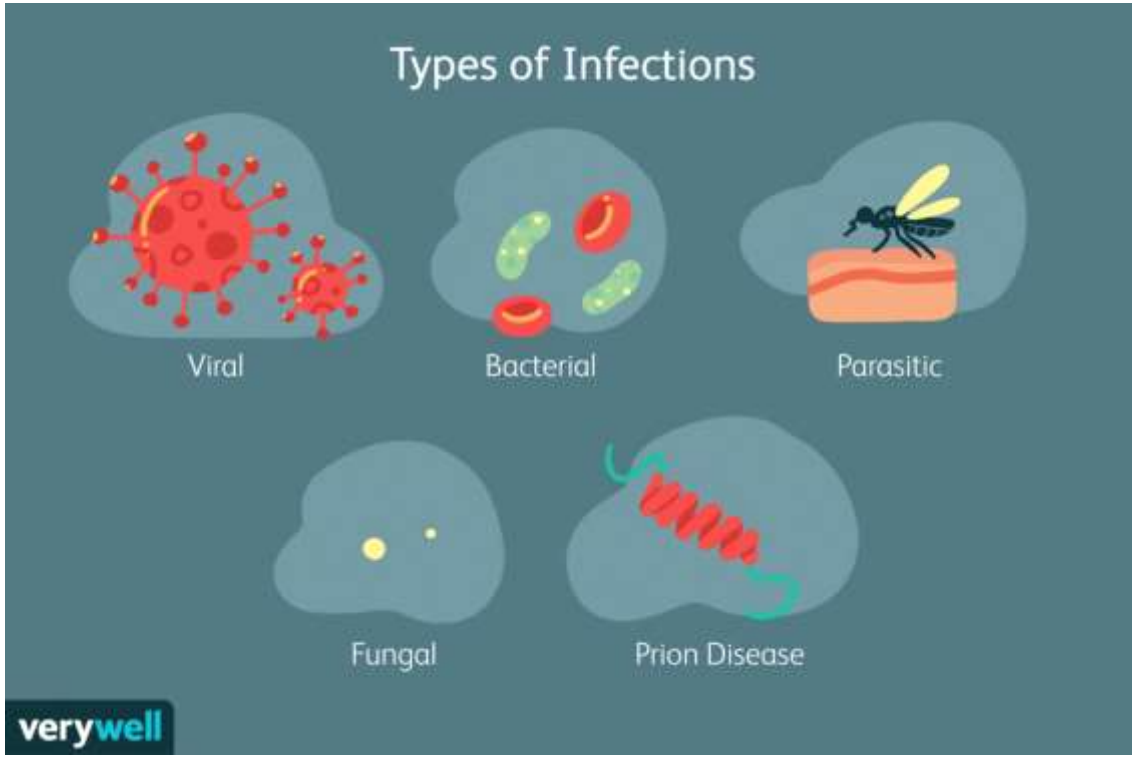
Perinatal

Anoxia

Prematurity and low birth weight

Birth injuries

Neonatal jaundice and meningitis



ACQUIRED HEARING LOSS

- Idiopathic SNHL
- Labyrinthitis - viral or bacterial infection
- Ototoxicity
- Trauma
- Presbycusis
- Endolymphatic hydrops
- Acoustic neuroma
- Noise induced hearing loss

Ototoxicity

TABLE 5.4 OTOTOXIC DRUGS

A. ***Aminoglycoside antibiotics***
- Streptomycin
- Dihydrostreptomycin
- Gentamicin
- Tobramycin
- Neomycin
- Kanamycin
- Amikacin
- Netilmycin
- Sisomycin

B. ***Diuretics***
- Furosemide
- Ethacrynic acid
- Bumetanide

C. ***Antimalarials***
- Quinine
- Chloroquine
- Hydroxychloroquine

D. ***Cytotoxic drugs***
- Nitrogen mustard (Mechlorethamine)
- Cisplatin
- Carboplatin

E. ***Analgesics***
- Salicylates
- Indomethacin
- Phenylbutazone
- Ibuprofen

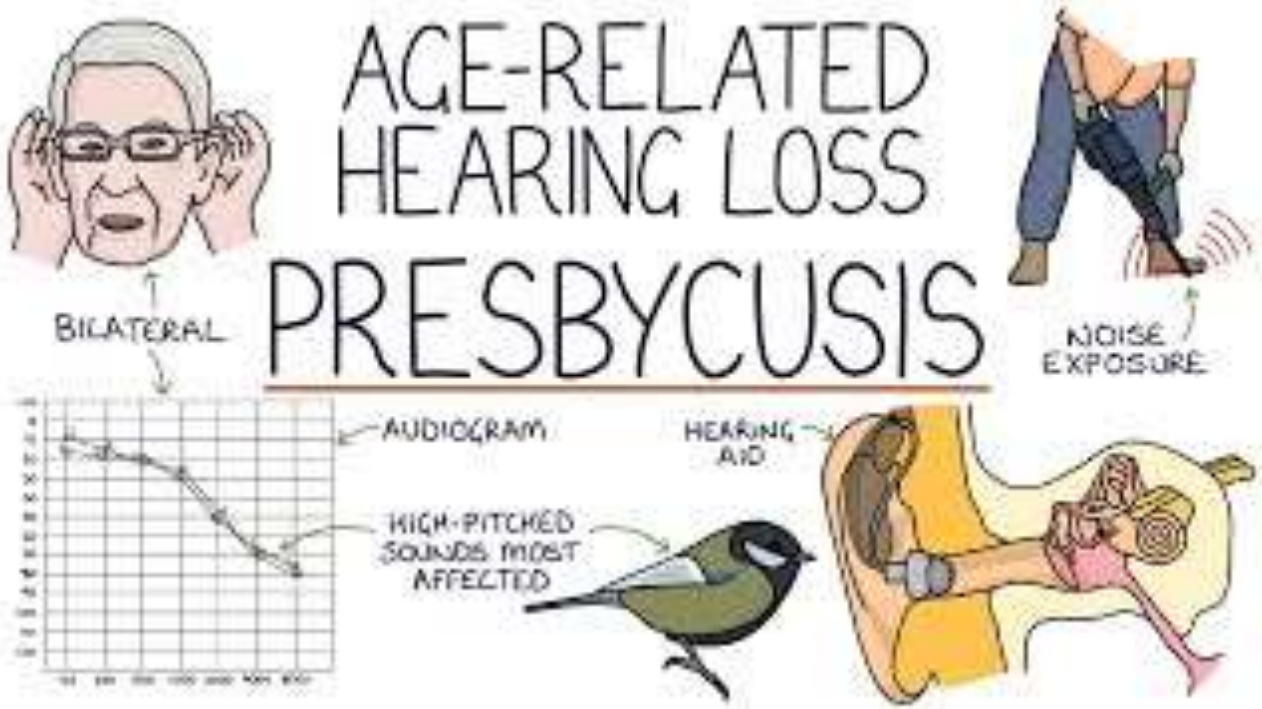
F. ***Chemicals***
- Alcohol
- Tobacco
- Marijuana
- Carbon monoxide poisoning

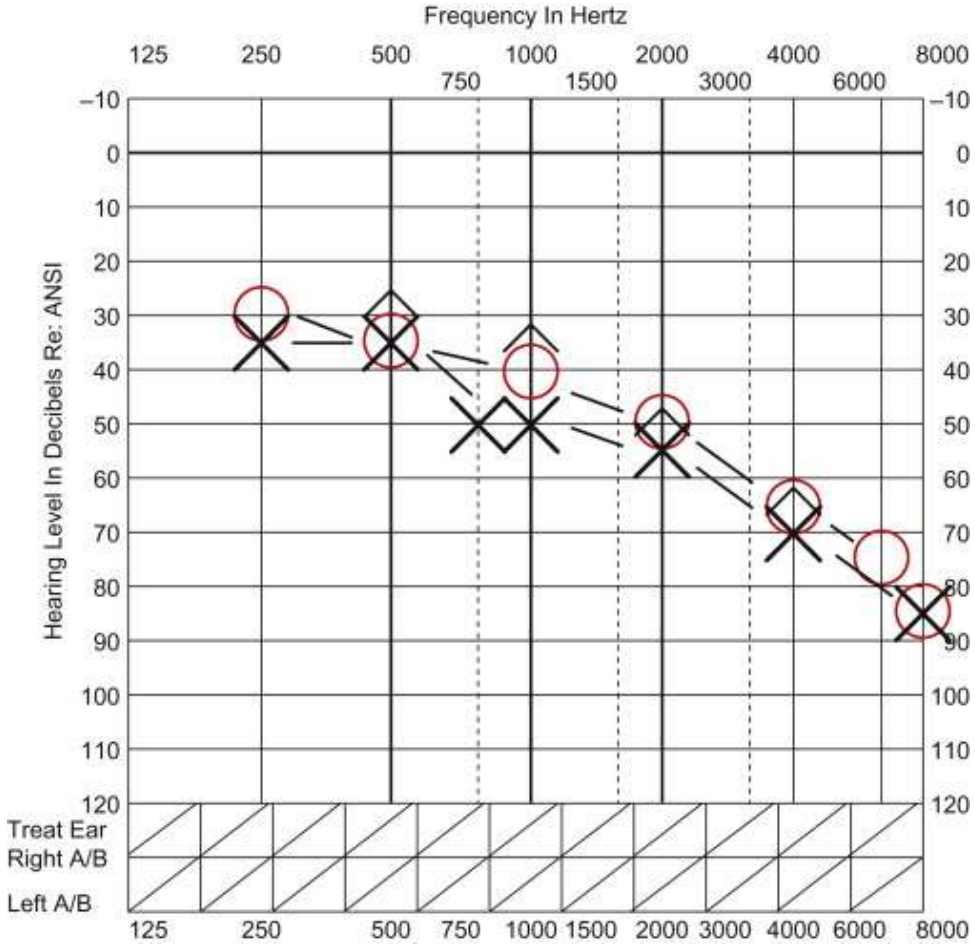
G. ***Miscellaneous***
- Erythromycin
- Ampicillin
- Propranolol
- Propylthiouracil
- Deferoxamine

Presbycusis

- Ageing.
- ***Around 65 y.***
- ***Clinical features***
- ***Deafness*****:** gradual and progressive, difficult to understand speech(high frequency hearing loss)
- Tinnitus
- **Investigations**
- Tuning fork tests
- Audiogram

PRESBYCUSIS



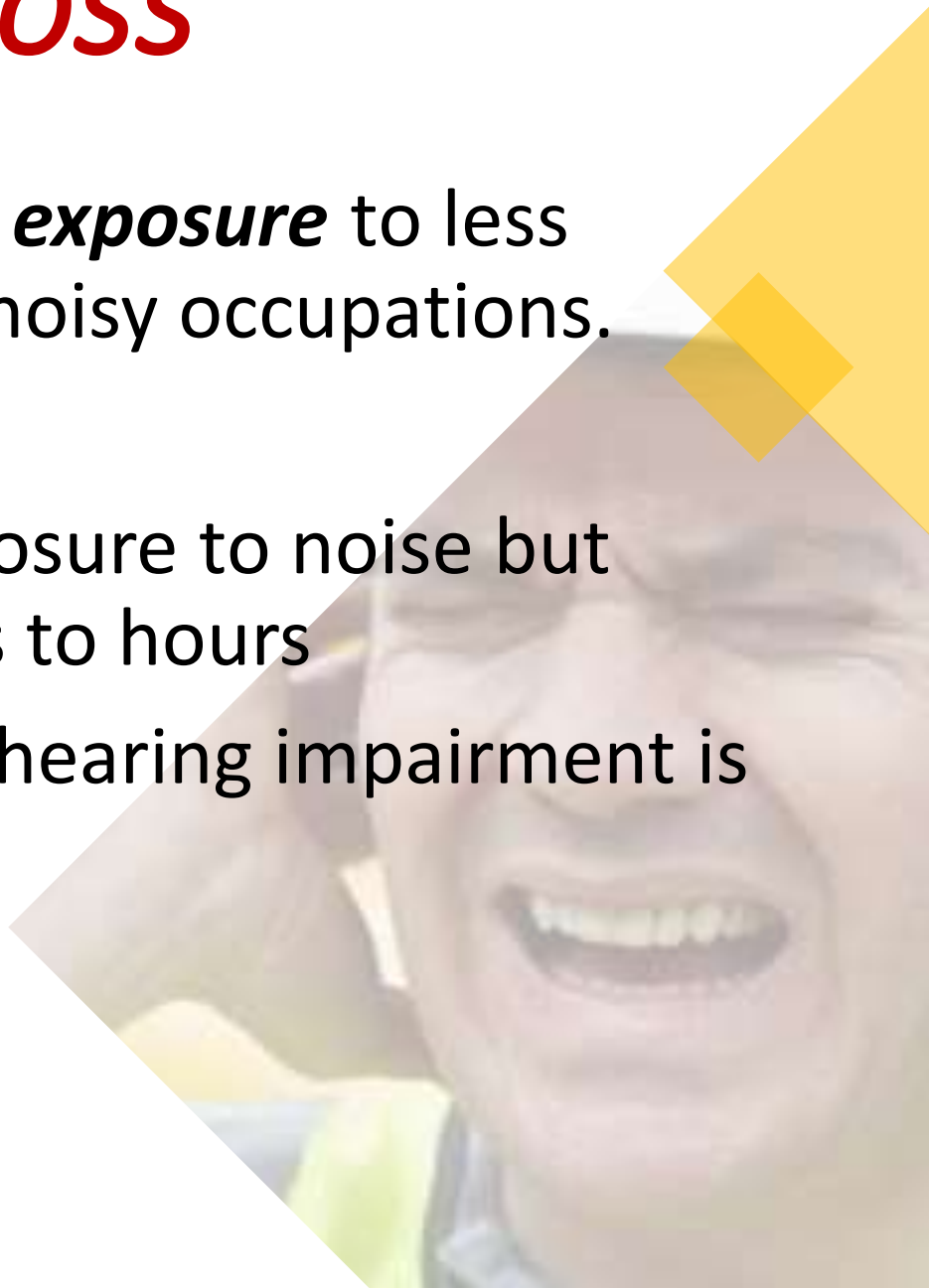


Noise induced hearing loss

- Hearing loss, in this case, follows ***chronic exposure*** to less intense sounds and is mainly a hazard of noisy occupations.
- ***(a) Temporary threshold shift (TTS)***

Hearing impairment is immediate after exposure to noise but recovers after an interval of a few minutes to hours

- ***(b) Permanent threshold shift (PTS)***. The hearing impairment is permanent and does not recover at all.



Acoustic Trauma.

single brief exposure to very intense sound

More than ***140 db.***

WHAT CAUSES NOISE INDUCED HEARING LOSS

A noisy work environment causes industrial deafness either by a sudden, damaging noise or prolonged exposure to harmful noise levels. More high-risk occupations include:

- Long-term exposure to 80dB of noise can damage your hearing
- A drill can make 100-110 dB of noise
- A quiet office generates 40-50 dB of noise

Idiopathic sudden sensorineural hearing loss

- ***30 db*** or more sensorineural loss at ***3 consecutive frequencies*** occuring in less than ***3 days***
- Multiple factors are implicated in the etiopathogenesis of SSNHL.
- 90% cases are idiopathic.
- 10% of the cases have specific causes.

Assessment of hearing

Assessment of hearing

<table>
<tr><td colspan="2">▪ Initial Otoscopic examination</td></tr>
<tr><td>▪ Speech test
○ Loud
○ Whisper</td><td>▪ Tympanometry</td></tr>
<tr><td rowspan="2">▪ Tuning fork test
○ Weber
○ Rinne
○ Schwabach</td><td>▪ BERA</td></tr>
<tr><td>▪ EChocG</td></tr>
<tr><td>▪ Audiometry
○ Speech audiometry
○ Pure Tone Audiometry</td><td>▪ OAE (Otoacoustic Emission)</td></tr>
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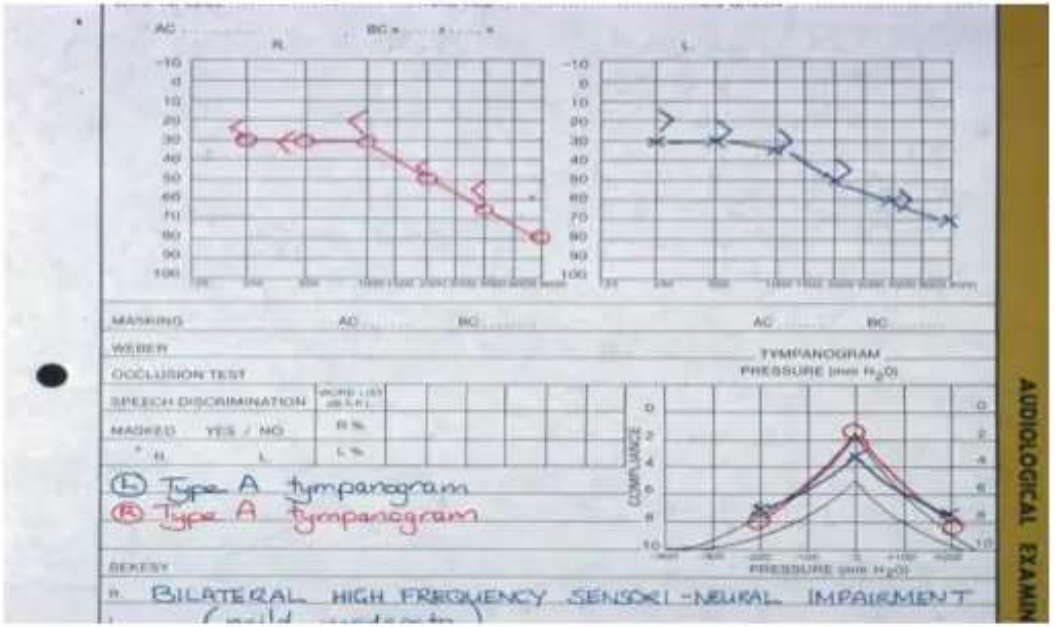
AUDIOGRAM - SNHL

-Both air and bone conduction threshold below normal .

-Gap between AC and BC less 10 db (no air bone gap).

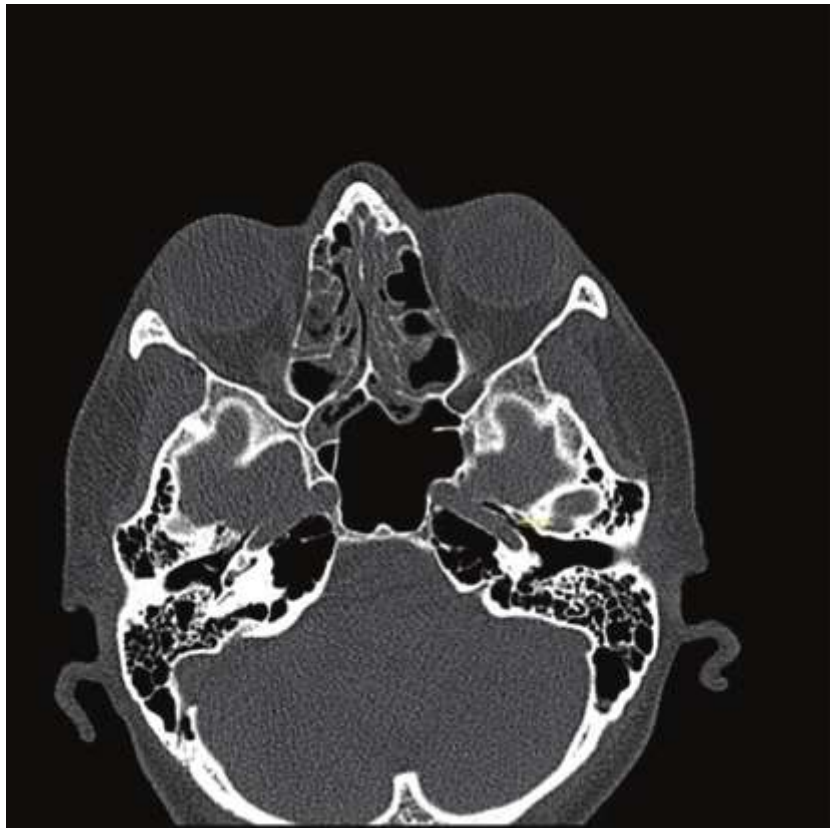


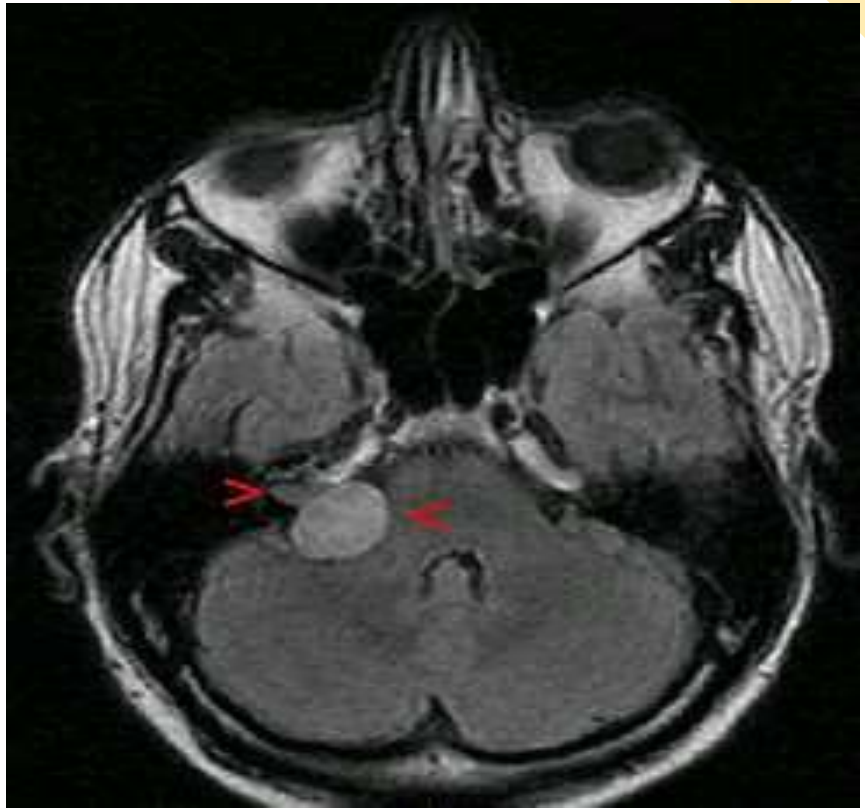
AUDIOGRAM

- 0 dB - 20 dB ..... normal hearing
- 20 dB - 40 dB ..... mild hearing loss
- 40 dB - 55 dB ..... moderate hearing loss
- 55 dB - 70 dB ..... moderately severe hearing loss
- 70 dB - 90 dB ..... severe hearing loss
- >90 dB..... profound hearing loss

Radiological investigations

- **An MRI scan** with gadolinium enhancement should be performed to exclude an **acoustic neuroma** but is also useful in evaluating multiple sclerosis and cerebrovascular accidents.
- **CT scan**



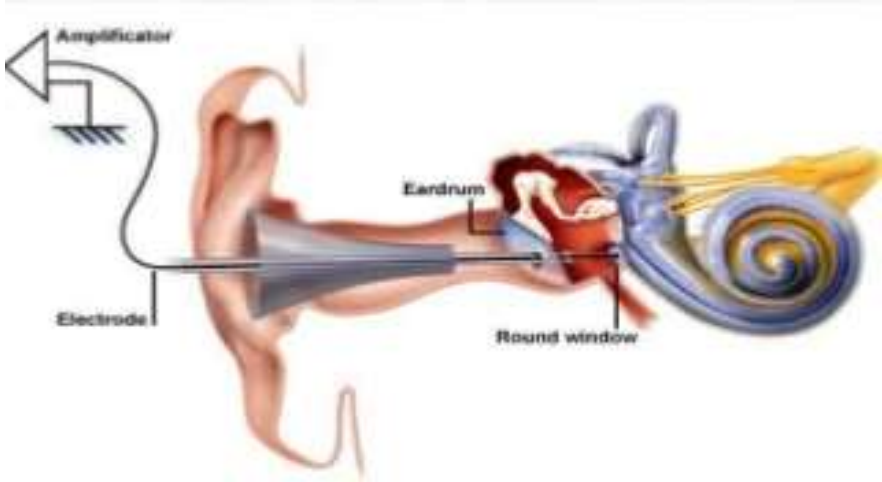


Investigations - Otoacoustic emissions(OAE)

- This is used to measure cochlear function by recording signals produced by hair cells
- Can be elicited by very sensitive microphone placed in EAC.
- **Uses**
- Used as a screening tests in neonates
- Differentiate between cochlear and retro cochlear pathologies
- To test hearing in mentally challenged and uncooperative patients after sedation.

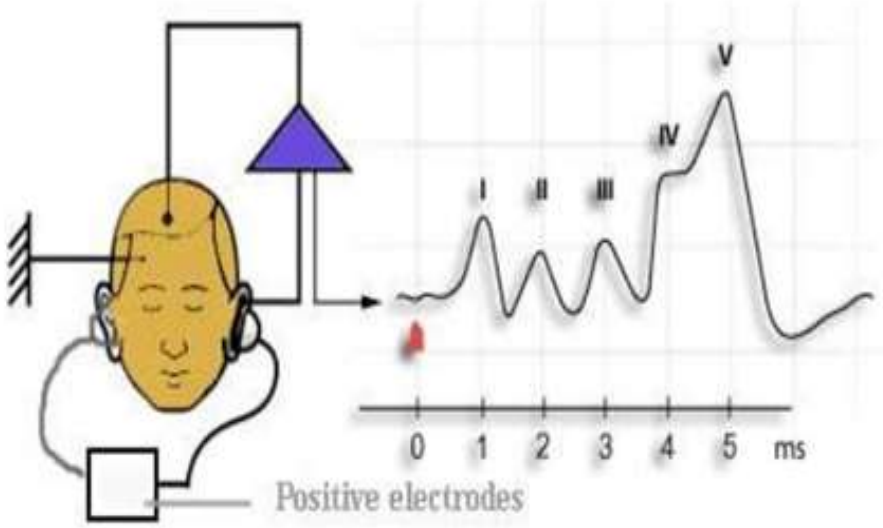
Investigations - Electrocochleography

- It measures electrical potentials arising in the cochlea and VIIIth nerve in response to auditory stimuli within first 5 millisec.
- ***Uses***
- To find threshold of hearing in infants and young children
- To differentiate lesions of cochlea from that of VIIIth nerve

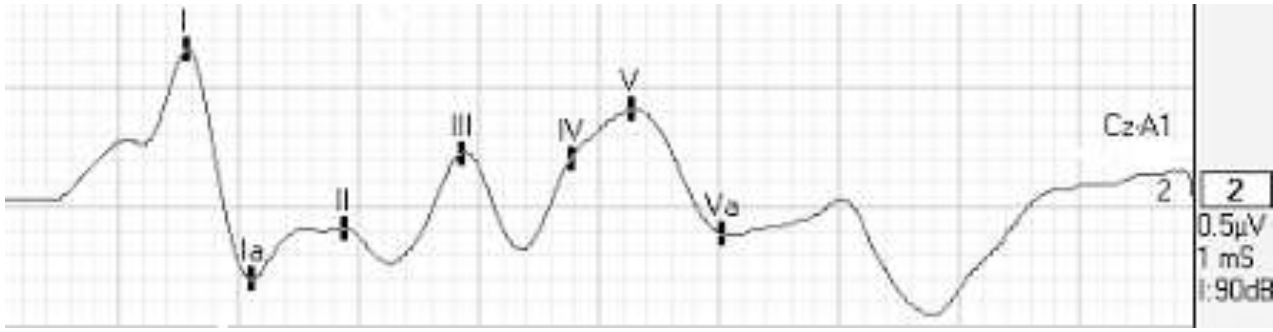


BERA- brainstem evoked response audiometry

- Objective way of eliciting brainstem potentials in response to audiological stimuli.
- Recorded by electrodes placed on scalp.
- ***Uses of BERA:***
- As a screening procedure for infants
- To diagnose retrocochlear pathology
- To diagnose brainstem pathology.
- To monitor cranial nerve VIII intraoperatively



BERA wave forms



- Wave I - Distal part of CN VIII
- Wave II - Proximal part of CN VIII near the brainstem
- Wave III- Cochlear nucleus
- Wave IV - Superior olivary complex
- Wave V - Lateral lemniscus
- Waves VI and VII - Inferior colliculus

- **Neonatal screening procedures**
- ABR/OAEs
- Arousal test
- Auditory response cradle
- Behavior observation audiometry
- Moro’s reflex
- Cochleopalpebral reflex
- Distraction techniques (6–18 months)
- Conditioning techniques (7 months – 2 years)
- Visual reinforcement audiometry
- Play audiometry (2–5 years)
- **Objective tests**
- ABR
- Otoacoustic emissions
- Impedance audiometry

Treatment of SNHL

Initial treatment

-Treatment for reversible hearing loss depends on its cause. It is often treated successfully.

Hearing loss caused by:

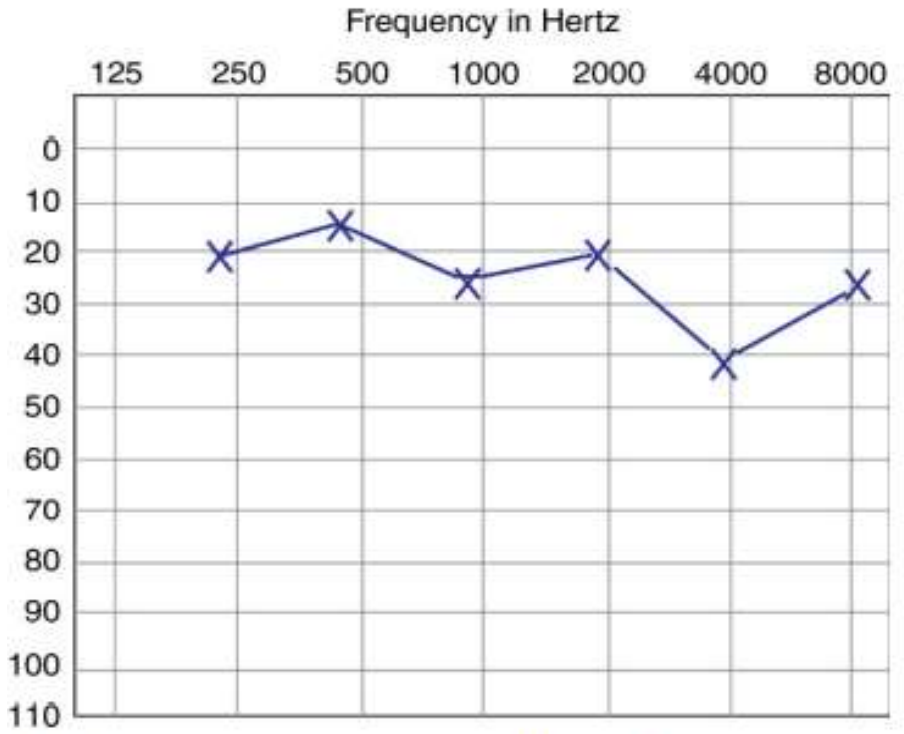
✓ Ototoxic medicines (such as aspirin or ibuprofen) often improves after you stop taking the medicine.

✓An ear infection, such as a middle ear infection, often clears up on its own, but you may need antibiotics.

✓An injury to the ear or head you may need surgery.

Treatment for Noise Induced hearing loss

- Prevention is better than cure
- Reduction of noise level at work place
- Use of hearing protection devices

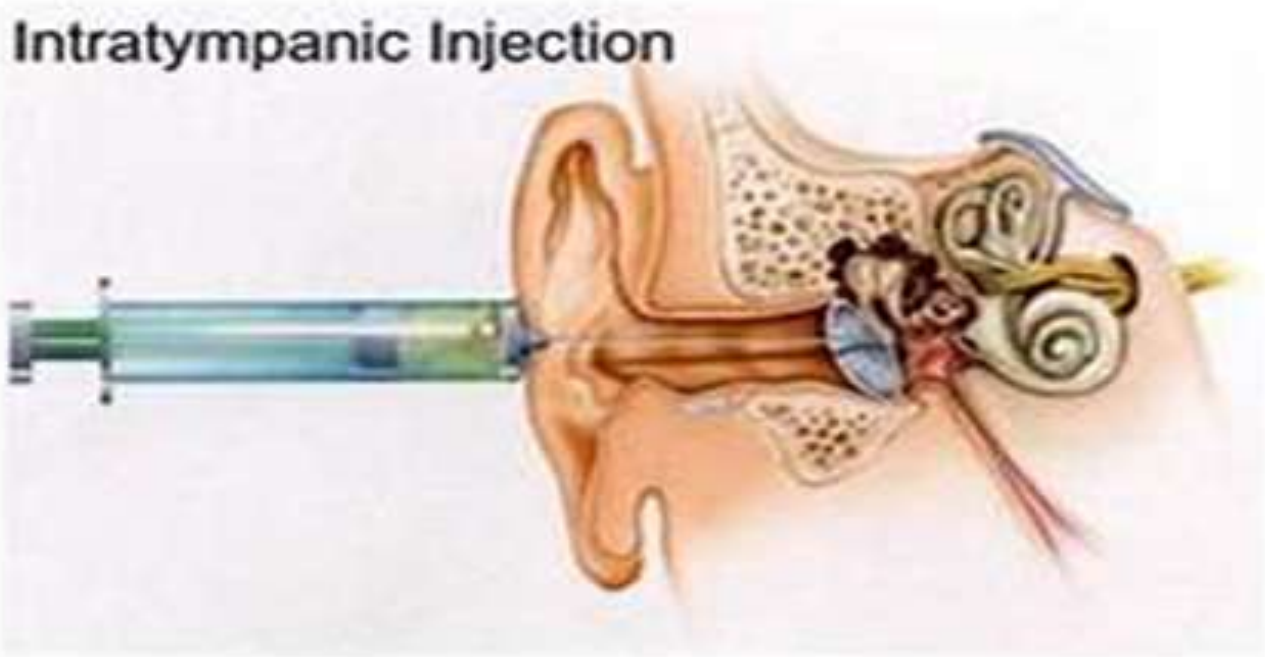


e 5.7. Early case of noise-induced hearing loss. Note dip at Hz.

Treatment for SSNHL

- ***STEROIDS***
- Spontaneous improvement in hearing is most likely to occur during the first 2 weeks; late recovery has been reported but is rare.
- Early corticosteroid treatment within the first 2 weeks is associated with the greatest hearing recovery, with minimal benefit after 4–6 weeks.
- **Dose**
- The best treatment outcomes are associated with a single dose of oral prednisolone of 1mg/kg/day, up to a maximum of 60mg daily, for 10–14 days.

- Dexamethazone preferably in concentration of 10 mg/cc is used. 1ml of it is mixed with 0.1 ml of 1% xylocaine. About 0.5 ml of this solution is injected into the middle ear cavity. Injections are administered twice a week for a period of 6 weeks. Introduction of Silverstein microwick has made the job little bit simpler.



Treatment for Presbycusis

- Prophylaxis
- Psychological support
- Hearing aids
- Drugs

ASSISTIVE TECHNOLOGY

HEARING AID

CIC
Completely in canal

ITC
In the canal

ITE
Inside the ear

BTE
Behind the ear

Cochlear implants

- A cochlear implant is an electronic device that can provide hearing and improved communication abilities for persons who have severe to profound sensorineural hearing loss and who cannot benefit from hearing aids.

Selection criteria-children

- At birth the cochlea is fully formed but the auditory pathway is not fully developed.
- The auditory pathway is *dependent on stimulation for its maturation*
- This stimulation is vital for developing speech and language skill.

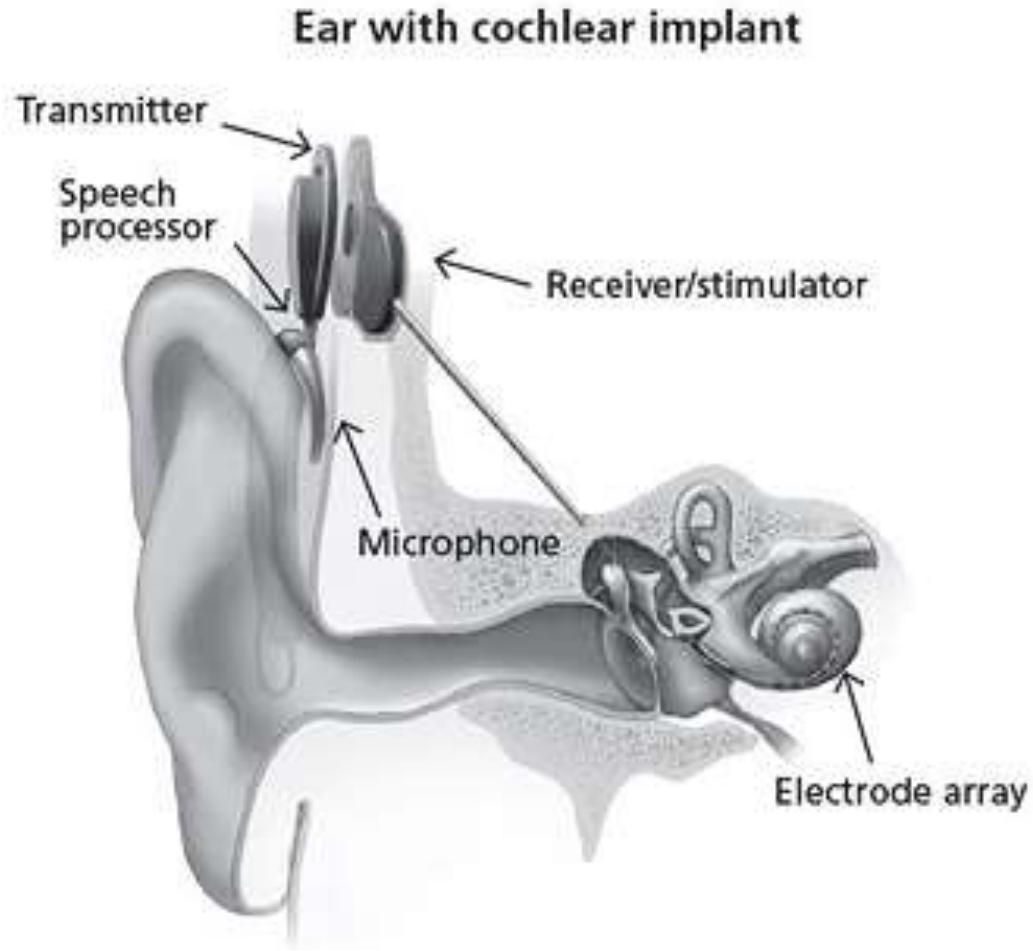
- Child above 12 months below 7yrs in pre- lingually deaf children
- Degree of deafness- child with profound hearing loss >90 dB and with good cochlear reserve.

Cochlear implants

- A cochlear implant works by producing electrical stimulation of the auditory nerve where degeneration of the hair cells in the cochlea has progressed to a point such that amplification provided by hearing aids is no longer effective

Components and Functioning of a Cochlear Implant

- A cochlear implant has an external and internal component.
- **1. External component.** It consists of an *external speech processor and a transmitter.* The speech processor may be body worn or behind the ear type; the latter being preferred



- **2. Internal component**.
- It is surgically implanted and comprises the *receiver/stimulator package with an electrode array.*

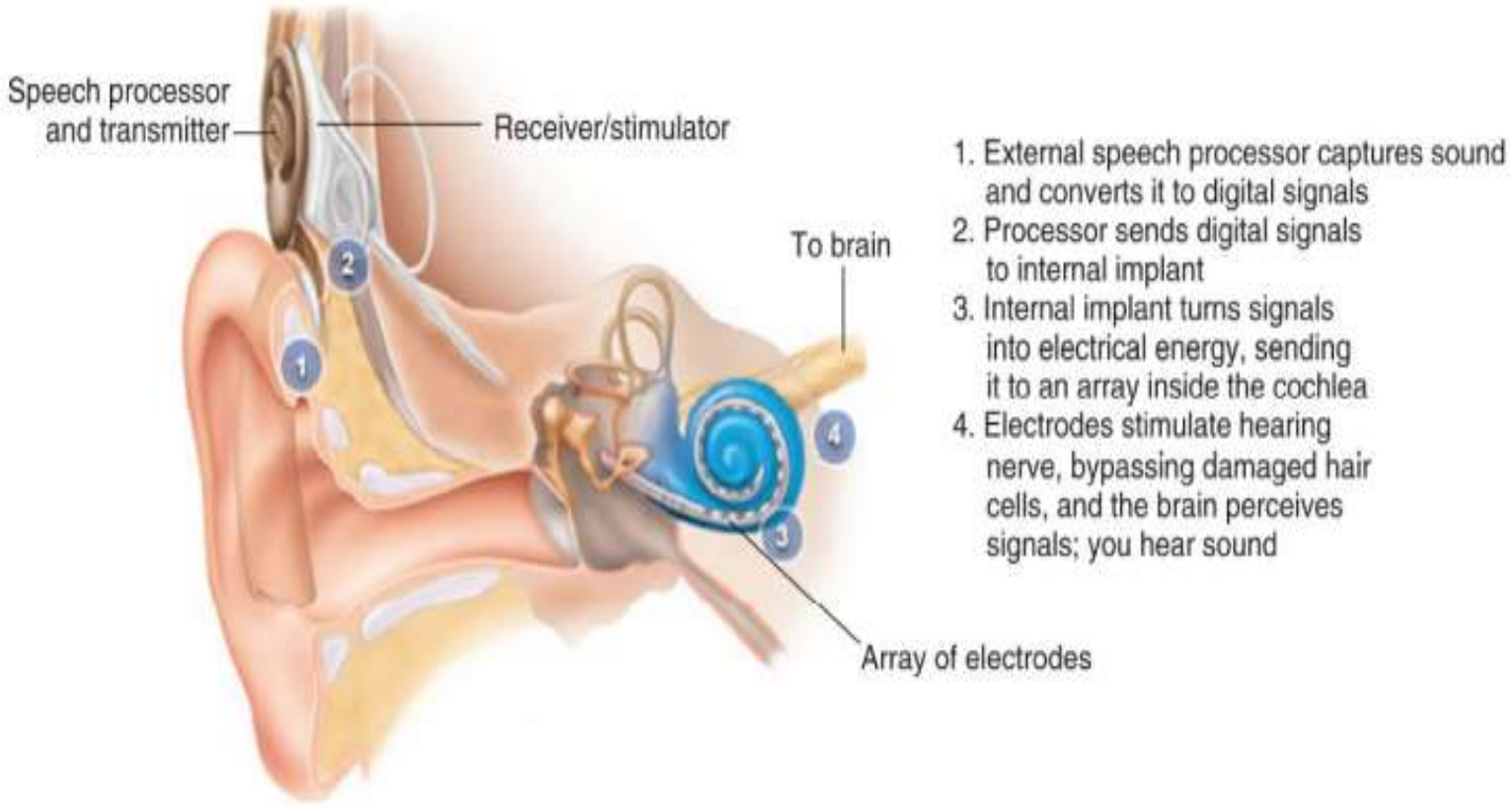


Figure 20.8. Advanced bionics cochlear implant system.

Mechanism of cochlear implants

Sound is picked up by the microphone in the speech processor. The speech processor analyses and codes sounds into electrical pulses

The electrical impulses are sent from the processor to the transmitting coil which in turn sends the signal to the surgically implanted receiver/stimulator via radiofrequency.

The receiver/stimulator decodes the signal and transmits it to the electrode array.

The electrode array which has been placed in the scala tympani of the cochlea stimulates the spiral ganglion cells.

The auditory nerve is thus stimulated and sends these electrical pulses to the brain which are finally interpreted as sound.

Protective measures to prevent hearing loss

- Prevention of congenital deafness
  - Genetic counseling and prenatal diagnosis
  - Pregnant women are prohibited to use ototoxic drugs
  - To avoid viral infection when pregnant
- Prevention of viral and bacterial infection
  - Measles
  - Varicella
  - Parotitis
  - Epidemic meningitis

To avoid long term exposure to noise

To avoid ototoxic deafness.

THANK YOU