SENSORY NEURAL HEARING LOSS

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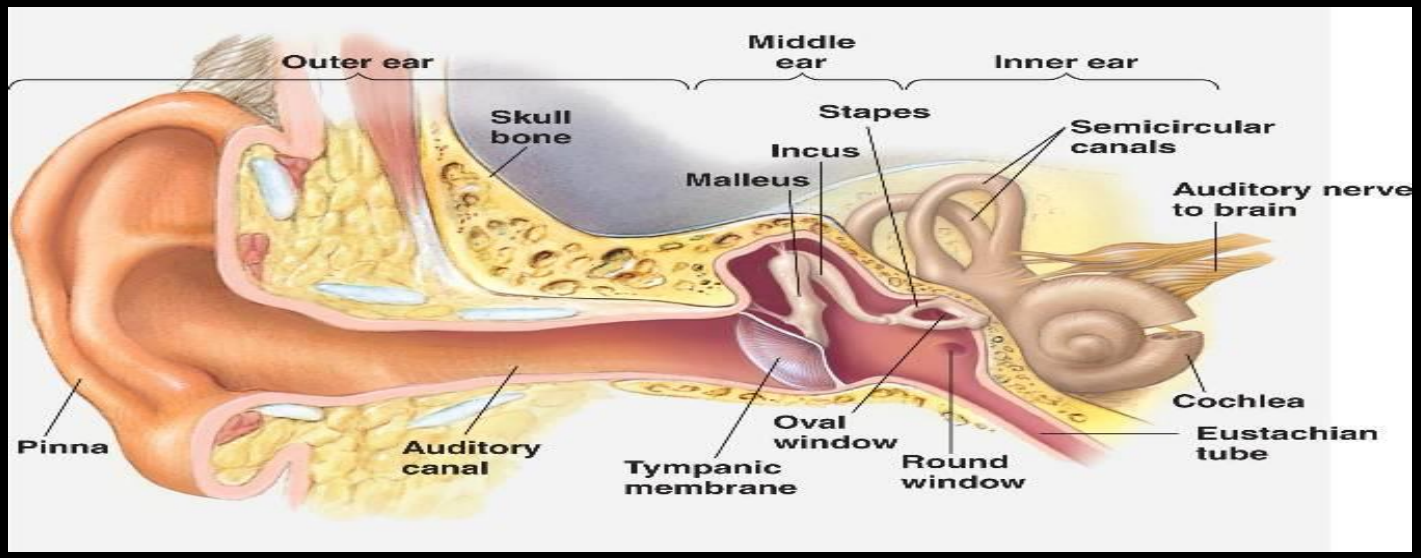
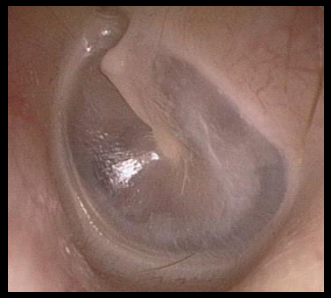
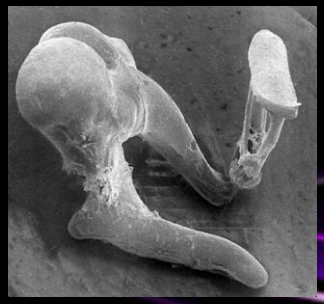
The Auditory System

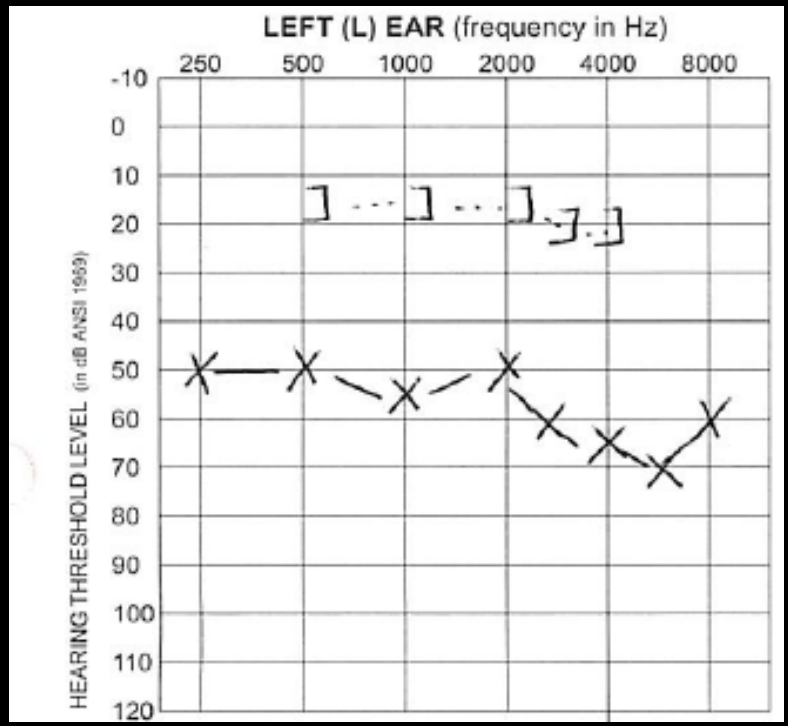






Types of Hearing Loss

Conductive Hearing Loss: caused by an abnormality of the conductive system.



Pathology of the EXTERNAL EAR

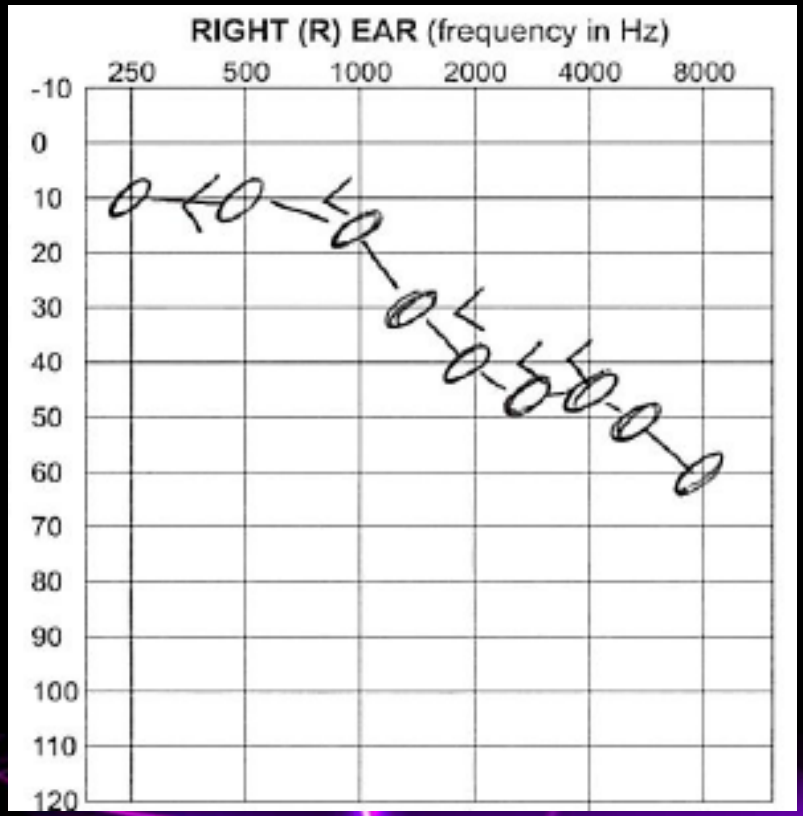
- External ear canal deformity (*atresia)*
- Blockage from *cerumen or foreign object*
- Perforated tympanic membrane

Pathology in the MIDDLE EAR

- Serous and Purulent Otitis media
- Otosclerosis
- Cholesteatoma

Sensorineural Hearing Loss: caused by **hair cell damage** or abnormality within the **cochlea** (inner ear).

- Noise exposure
- Ototoxic medication
- Meniere's disease
- Infections
- Idiopathic
- Congenital
- Aging (presbycusis)



Retro-cochlear Pathology: Hearing loss due to a misconnection between the inner ear and auditory centre of the brain.

Disorders include:

- Auditory Neuropathy
- Central Auditory Processing Disorder
- Acoustic Neuroma

Sudden Sensorineural Hearing Loss

- sudden SNHL as a decrease in hearing of at least *30 dB* at *3 contiguous frequencies* occurring within a *72-hour* period.
- develops hearing loss rapidly, awakening with it in the morning or developing a progressive loss over 72 hours or less.
- The incidence of sudden SNHL has been estimated to range from 5 to 20 per 100,000 persons per year
- peak incidence seems to be in the *6th decade*

- The distribution is essentially equal.
- Bilateral involvement is rare, and simultaneous bilateral involvement is very rare.

The most common presentation is a patient noticing a unilateral hearing loss on awakening.

Occasionally, patients note a fluctuating hearing loss.

A sensation of aural fullness in the affected ear is common and frequently is the only complaint.

Tinnitus is present in the ear to a variable degree

hearing loss sometimes is preceded by the onset of tinnitus.

Vertigo or dysequilibrium is present to a variable degree in approximately 40% of patients

- Without treatment of any kind, a significant proportion (30% to 65%) of patients experience complete or partial recovery.
- Four variables seem to affect the prognosis of idiopathic sudden SNHL:
- *(1) severity of loss,*
- *(2)audiogram shape,*
- *(3) presence of vertigo,*
- *(4) age*.

- The more severe the loss, the lower the prognosis for recovery, and profound losses have an exceptionally poor prognosis.
- Up sloping and midfrequency losses recover more frequently than downsloping and flat losses.
- The presence of vertigo, particularly with a down sloping loss, is a poor prognostic indicator.
- Reduced speech discrimination carries a poor prognosis.
- children and adults older than 40 years have a poorer prognosis than others.

- Most recovery occurs within the first 2 weeks after onset; the prognosis for recovery decreases the longer the loss persists. Those who have not meaningfully recovered by 3 months are unlikely to do so.

Etiology of Sudden Sensorineural Hearing Loss

Infectious Disorders

- Viral Infection.
- SNHL can complicate clinically evident infections with ***mumps, measles, herpes zoster, and infectious mononucleosis***, and with ***congenital rubella*** and ***CMV***.
- Of patients who come to medical attention with sudden SNHL, 28% report a viral-like upper respiratory infection within 1 month before the onset of their hearing loss

- *Bacterial Infections.*

➢ *Bacterial meningitis* is a well-recognized etiology of acquired severe to profound SNHL.

➢ *Serous or suppurative labyrinthitis* can also precipitate sudden SNHL.

➢ the incidence of *syphilis* in patients with sudden SNHL is 2% or less. Syphilitic hearing loss may manifest at any stage of the disease, and it may be associated with other manifestations of syphilis.

➢ *Lyme disease* is a well-established etiology of acute facial paralysis, and also cause of sudden SNHL

➢ Sudden SNHL may be associated with *HIV infection*.

In the presence of HIV infection, sudden SNHL may occur with or without the presence of opportunistic infection, and it may occur without clinical evidence of AIDS.

Neoplasms

- **Acoustic Neuroma.**
- It is common for sudden SNHL to be the initial manifestation of a vestibular schwannoma.
- Approximately 10% to 20% of acoustic neuromas initially manifest with sudden SNHL.
- The prevalence of acoustic neuroma among patients with sudden SNHL is less clear, although estimates range from 3% to 10% of those who undergo MRI.
- The presence of tinnitus in the ipsilateral ear before sudden SNHL is suggestive, but it is not present in most cases.
- In addition, midfrequency and high-frequency hearing loss are more commonly associated with acoustic neuroma .
- electronystagmography abnormalities are more common with acoustic neuroma.

- Responsiveness of the hearing loss to treatment with steroids *does not rule out a retrocochlear lesion*.

- The clinician should have a high level of suspicion for acoustic neuroma in any patient with sudden SNHL.

- As recommended , patients with sudden SNHL should undergo an evaluation for retrocochlear pathology.

- The *most sensitive* means of detecting a structural lesion is with *gadolinium-enhanced MRI*.

- ABR has been used as an initial screening measure. However, ABR may be falsely negative in patients with small acoustic neuromas. If a patient is unable to undergo MRI , a fine-cut temporal bone CT may be used to detect larger lesions (>1.5 cm).

Other Neoplasms.

- Neoplasms of the CPA or internal auditory canal other than acoustic neuromas have also been associated with SNHL.
- These include meningioma, epidermoid (cholesteatoma), hemangioma,arachnoid cyst, metastatic neoplasms, and other skull base neoplasms that erode into the inner ear.

Trauma and Membrane Ruptures

- **Head Injury.** Sensorineural hearing loss of any degree can occur after closed or open head injury.
- The mechanism of injury in such patients has been shown pathologically to vary from mild loss of outer or inner hair cells or cochlear membrane breaks to fracture across the labyrinth or intralabyrinthine hemorrhage.
- Many of these injuries are pathologically indistinguishable from injuries of acoustic trauma.
- Some patients experience a variable degree of recovery from head injury-induced hearing loss.

- *Perilymphatic Fistula.*
- Round or oval window fistulae can occur congenitally, after stapedectomy, or after barotrauma.
- Some investigators theorize that these fistulae can occur after heavy lifting or straining or even spontaneously. Patients with such fistulae can have sudden or fluctuating SNHL and varying degrees of vestibular symptoms. No test is reliable for detecting the presence of such a fistula, and even surgical exploration is subject to error.Except in post-stapedectomy patients, it is doubtful that perilymph fistula is a significant cause of SNHL.

Pharmacologic Toxicity

- ototoxic causes of SNHL may result in the relatively sudden onset of hearing loss. In addition to these drugs, others have been associated with sudden SNHL. *Interferon* has been associated with SNHL that has been reversible in most patients.

- The insecticides *malathion* and *methoxychlor* have been associated with bilateral SNHL.

Ototoxic Drugs

Drug	*Vestibulotoxicity*	*Hearing Toxicity*	*Toxic Level*
Erythromycin		yes	High IV doses only
Gentamicin	8.6%	minor	Usually 2 weeks
Streptomycin	very toxic	minor	
dihydrostreptomicin	minor toxic	very toxic	
Tobramycin	Yes	minor in 6%	Less toxic than Gentamicin
Netilmicin		2.4%	
Amikacin	not toxic	13.9%	
Neomycin	minor	very toxic	In topical ear drops
Kanamycin	minor	very toxic	
Etiomycin	moderate		
Vancomycin	nontoxic	none to moderate	synergistic with gentamicin
Metronidizole	toxic (rarely)	unknown	
Capreomycin		yes	

Immunologic Disorders

- The finding that many patients with SNHL seem to benefit from glucocorticoid therapy and the finding of cross-reacting circulating antibodies in many patients with sudden and rapidly progressive SNHL suggest that at least a subset of SNHL cases are caused by inner ear autoimmunity. In addition, many well-known autoimmune diseases
- have been associated with SNHL, including *Cogan syndrome,systemic lupus erythematosus, temporal arteritis,* and *polyarteritis*
- *nodosa.*.

Vascular Disorders

- Sudden hearing loss can occur with occlusion of the cochlear blood supply.
- Because of the abruptness of onset of SNHL and the fact that the cochlea depends on a *single terminal* branch of the posterior cerebral circulation, vascular occlusion has been thought to be an attractive hypothetical etiology for idiopathic sudden hearing losses.

- *Migraine, hemoglobin sickle cell disease*, and *macroglobulinemia* have been documented to be associated with sudden SNHL. Rare cases of thromboangiitis obliterans (*Buerger disease*) have been associated with sudden SNHL.
- *Small cerebellar infarctions* may mimic labyrinthine lesions, including sudden onset of hearing loss.
- *Cardiopulmonary bypass* and non cardiac surgery have been associated with an increased risk of sudden SNHL. Sudden SNHL has also been reported *after spinal manipulation*, with probable injury to the vertebrobasilar arterial system.
- It has long been believed that patients with *diabetes* have a higher incidence of idiopathic sudden SNHL.

Idiopathic Disorders

- *Meniere Disease.*
- Some patients seen with typical sudden SNHL ultimately may develop a history more suggestive of endolymphatic hydrops or even frank Meniere disease; this probably constitutes only 5% of all patients with sudden SNHL

Multiple Sclerosis.

MS is a demyelinating disorder of the CNS manifested by differing neurologic lesions separated by space and time. *Sudden SNHL is a rare initial manifestation of MS*.

Among patients with MS, auditory abnormalities are common.

Treatment

Treatment of sudden SNHL should be based on its etiology.

Because of the poor understanding of idiopathic sudden SNHL, controversy surrounds its treatment,.

Because of the dictum "*First, do no harm*," new or unconventional treatment protocols should be well reasoned and carefully applied.

- *Steroids* in *moderate doses* have become the most widely accepted treatment option for idiopathic sudden SNHL.
- A common approach is to prescribe a *10- to 14-day* course of prednisone, approximately *1 mg/kg per day* (not to exceed 60 mg/day) in a single undivided dose, with a slow taper.
- If a partial recovery is noted at the end of the 10 days, one may consider extending the full dose another 10 days and repeating the cycle until no further improvement is noted.

Local delivery of steroids to the inner ear via IT instillation has seen increasing use. IT steroid treatment has the potential advantage of achieving very high steroid concentrations within the inner ear without the associated systemic side effects.

As a result, this approach is potentially more suitable for patients with diabetes, glaucoma, and cataracts.

The most commonly administered IT steroids are *dexamethasone* and *methylprednisolone.*

Dosing concentrations and regimens range widely, with some injecting weekly or on several consecutive days, and others performing continuous infusions.

- *Vasodilators* have been tried extensively in the treatment of sudden SNHL.

Any proposed vasodilator would have to cross the blood-brain barrier and have an effect on intracranial circulation.

- *IV histamine* infusion, *oral papaverine*, and oral *nicotinic acid* have been used most frequently.
- Other agents proposed to improve cochlear blood flow include low-molecularweight dextran, mannitol, pentoxifylline, and heparin.

- Still other treatment ideas are directed at other presumed etiologies.
- Because endolymphatic hydrops is a common final pathology for many inner ear injuries and may be associated with some cases of sudden SNHL, some authors have advocated treatment with a *sodium-restricted diet* and a *diuretic*.
- Because of the evidence for a viral etiology and, specifically, evidence of herpes virus, treatment with oral antiviral medications, such as valacyclovir, has been proposed.

- *No study showed a significant difference in recovery between those receiving antivirals with steroids versus steroids alone*.

Presbycusis

- **Hearing loss associated with aging**
- **Most common cause of SN HL – and *most common cause of HL overall***
- **Presbycusis begins in *adolescence*. Sad but true.**

- Symmetrical sensorineural hearing loss

•over 50 years of age.

•Approximately one-third of persons over the age of 65 have significant hearing loss, averaging 35 dB or more on pure-tone audiometry.

Pure-tone hearing is often better than the hearing for speech, and hearing for syllables often better than that for sentences *(schizacusis).*

Other symptoms include noises in the ears and psychological disturbances

Four types of presbyacusis can be distinguished on the basis of morphologic degenerative lesions:

PRESBYCUSIS TYPES

1. *Sensory presbyacusis due to hair cell degeneration. The* audiogram shows high tone loss.

2. *Neural presbyacusis: A large proportion of the population* of cochlear neurons is lost, so that the predominant symptom is loss of discrimination for speech.

3. *Strial presbyacusis, due to degeneration of the stria vascularis.*
This causes abnormalities in endolymph production and secretion, with repercussions on the energy metabolism of the hair cells.

Audiography shows a flat curve of pancochlear hearing loss with retained discrimination for speech

4. *Conductive cochlear presbyacusis: An age-related degenerative* process in the cochlear duct causes lesions in the structure of the basilar membrane.

This affects stimulus transport in the cochlea and is demonstrated on audiography by bilateral symmetrical sensorineural deafness causing a characteristic sloping curve, with a linear increase in hearing loss above 1000 Hz.

Noise-Induced Hearing Loss

Exposure to high levels of noise can damage HCs and cause SN HL. Two types:

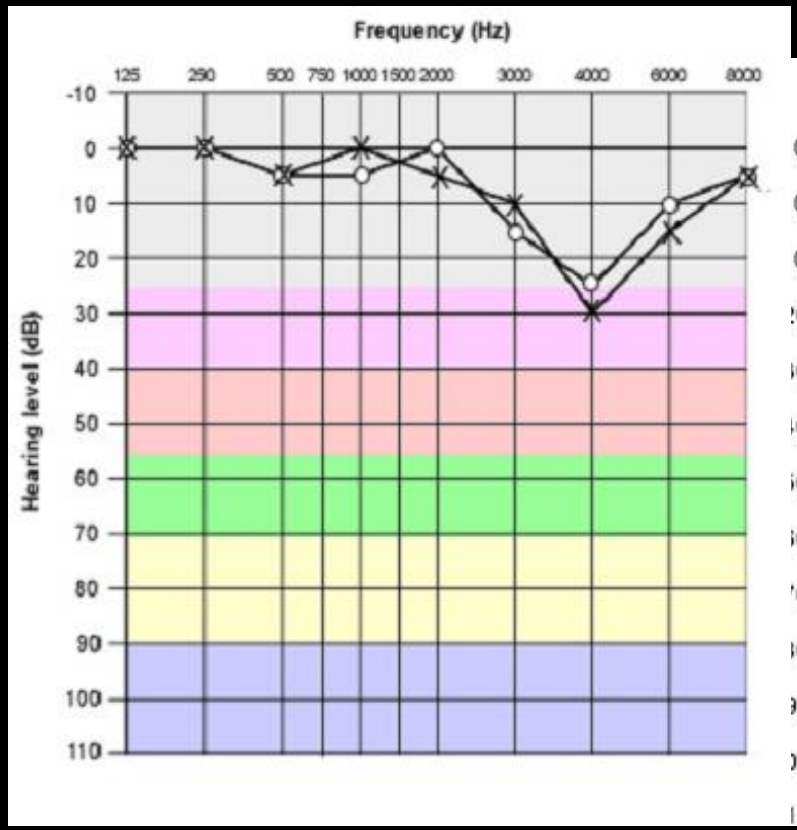
***Acoustic trauma*:**

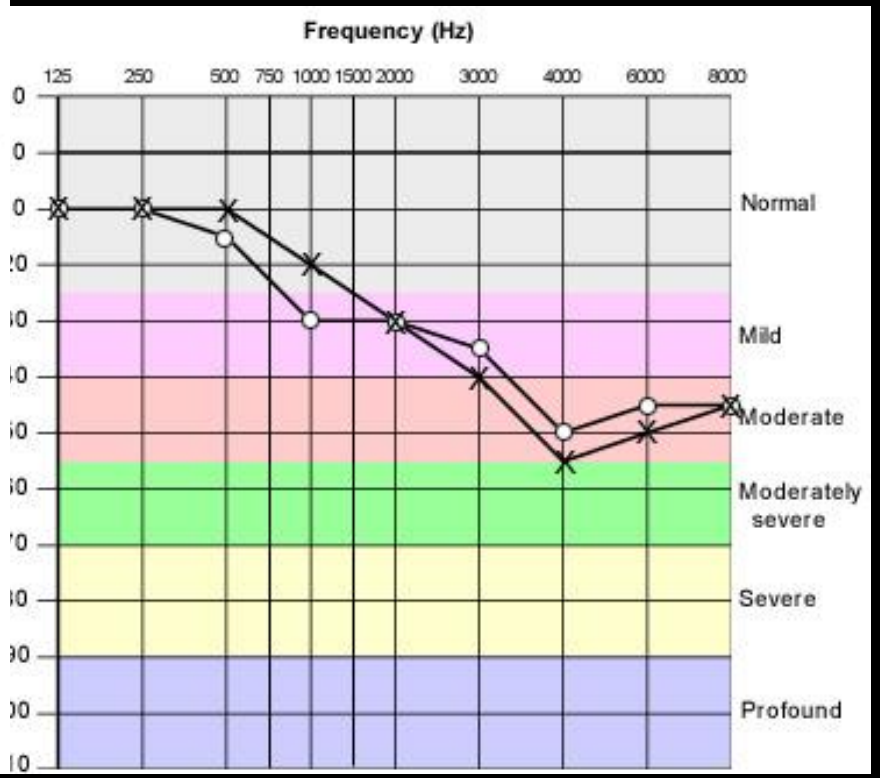
- **Injury due to brief exposure to very intense sounds such as gun shots, artillery fire, explosions, etc.**
- HL *may* be severe and permanent, but substantial recovery is common.

Long-term noise exposure (more common):

- Damage results from long-term exposure **to high levels of noise.**
- **Common in some occupational settings – *heavy manufacturing and agriculture* being the most common.**
- **Amount of inner-ear damage depends on the combination of:**
  - **Intensity of the noise**
  - **Length of exposure**

Audiometric Pattern is distinctive (audiogram on right shows more advanced progression than left)





Note:

(1) Dip or "notch" at ~3-6 kHz

(2) Typical progression shows the notch broadening (especially on the high frequency side) and deepening

(3) High frequencies more affected than lows

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Capreomycin		yes	

d. *Meniere's Disease*

Serious, often debilitating disease of hearing and balance of uncertain cause.

MD affects a single ear in about 75% of cases.

major symptoms:

(1) Periodic episodes of *rotary vertigo* (the sensation of spinning) or dizziness (the *"Meniere's attack"*)

(2) Fluctuating, progressive, low-frequency *hearing loss*

8th N Tumors (*acoustic neuroma*)

- Benign (i.e., nonmalignant) tumor that exerts pressure on 8th N
- Almost always slow growing
- Most common symptom: hearing loss (mild initially), often accompanied by tinnitus
- Vestibular problems may also occur
- Cause is unknown
- Continued tumor growth can be life threatening

8th N Tumors (*acoustic neuroma*)

- **Treatment: Surgical removal or radiation**
- **Early detection is really important: Small tumors can be removed with less risk of destroying the 8th N (and sometimes the 7th N as well).**
- **But, early detection is difficult – *early-stage symptoms are not dramatic.***
- **Acoustic neuromas sometimes run in families (case in point to follow).**