

## CLINICAL CORRELATIONS RELATED TO THE AUDITORY SYSTEM

### HEARING TESTS

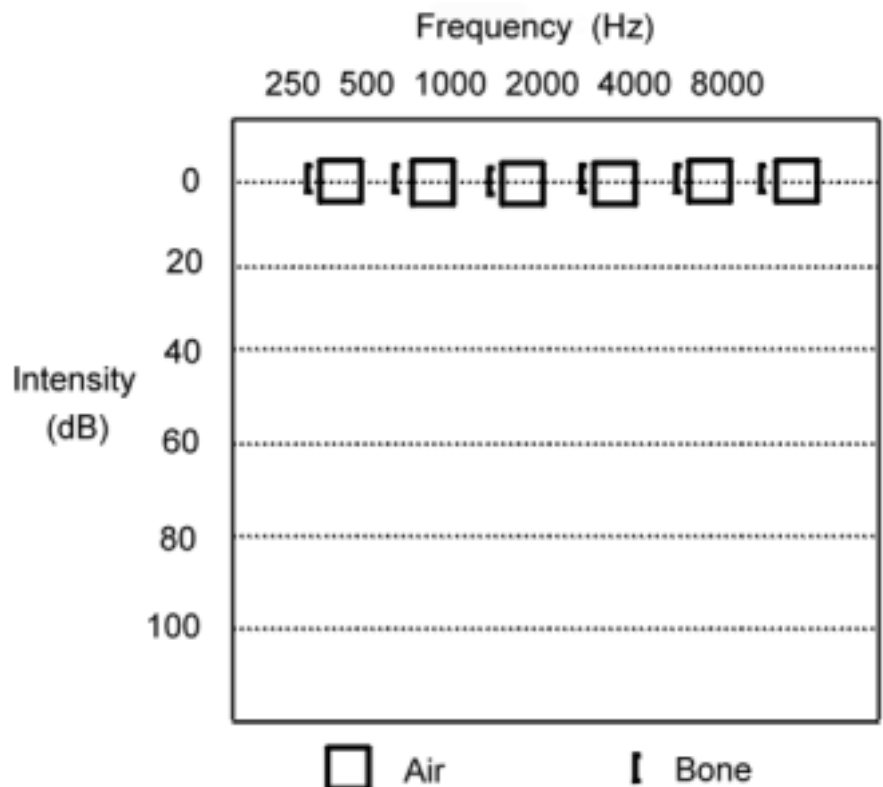
#### The Audiogram - a basic clinical test of hearing sensitivity

The auditory system can be stimulated via sound energy that is sent through air to the ear drum (air conduction) or by placing a bone vibrator against the skull (bone conduction). Sound sent through air tests all parts of the auditory system—the outer ear, middle ear, inner ear and central auditory pathways. In contrast, sound conducted through bone bypasses the outer and middle ear. It directly sets up a traveling wave in the cochlea and stimulates the cochlea and central auditory pathways. By comparing the auditory thresholds using these two methods, we can determine the site of hearing loss.

An audiogram is a graph showing hearing threshold as a function of frequency. There are three variables to know whenever an audiogram is performed: the frequency of sound that is being presented (Hz), the intensity of sound that is being presented (dB HL), and the method of sound presentation (air conduction {head set} or bone conduction). Sound frequency has already been described, but the latter two variables require some additional discussion.

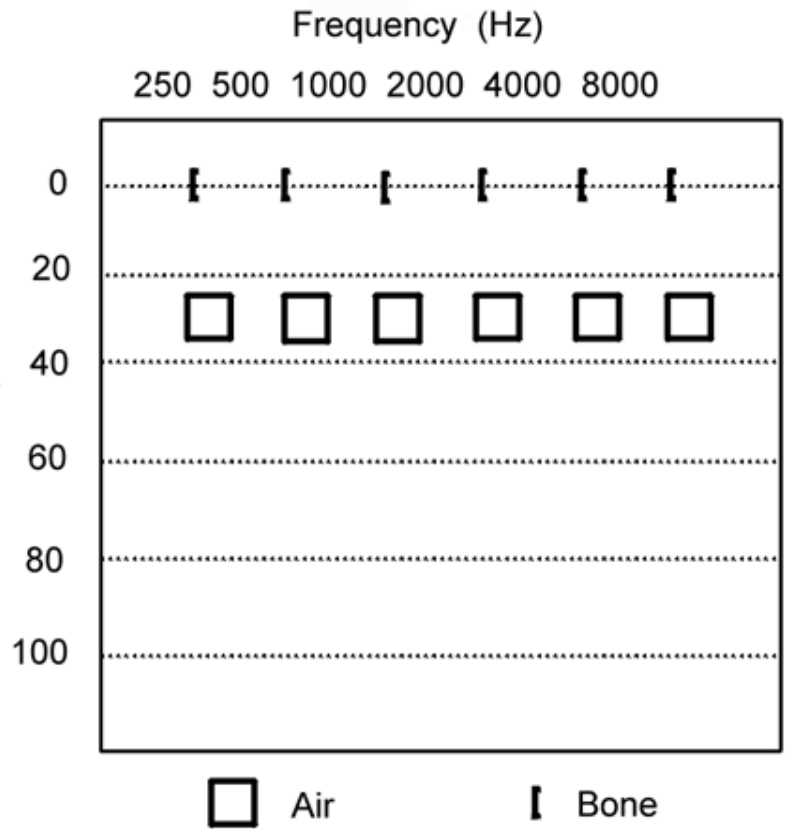
As you recall, the auditory threshold (measured in deciBels) depends upon the frequency of the sound that is presented. If we “recalibrate” the sound intensity scale such that the auditory threshold for humans is set to a value of zero at each frequency, the resulting scale is called the dB Hearing Level (dB HL) scale. The dB HL scale is used in clinical practice. If there is a hearing loss, the amount of loss is directly related to the value of the auditory threshold. Thus, a threshold of 30 dB HL at any frequency reflects a 30 dB hearing loss, and a threshold of 0 dB at any frequency is consistent with normal hearing.

The adjacent figure shows the **normal** auditory thresholds (in dB HL) from 250 to 8000 Hz. Both air conduction and bone conduction thresholds are represented. Normal hearing, on the dB HL scale, is 0 dB.

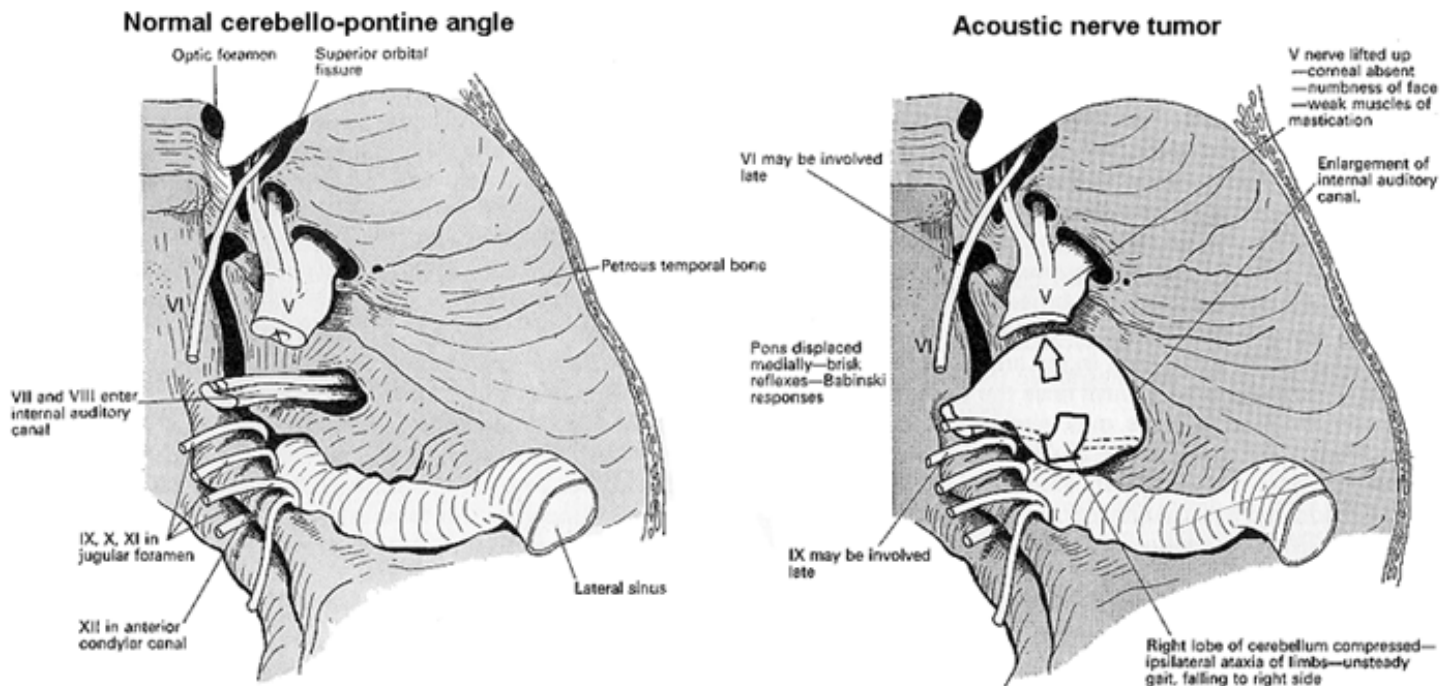


### Conductive vs. Sensorineural Hearing Loss

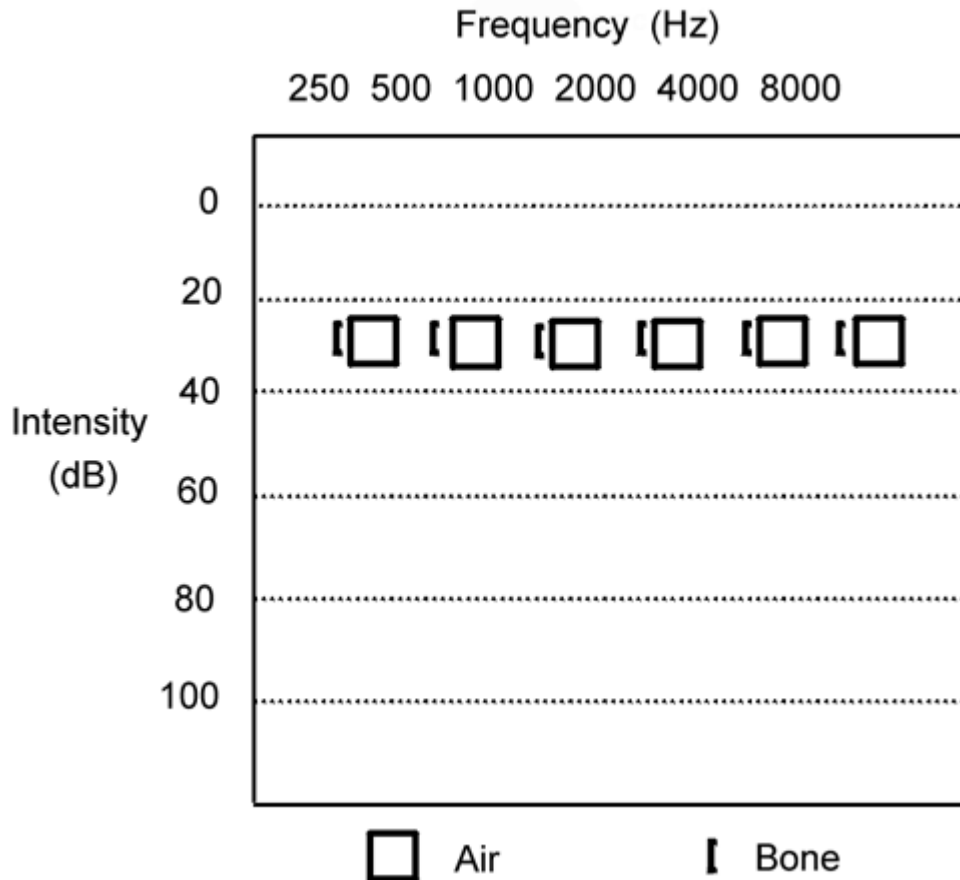
If a hearing loss is detected when hearing is tested via air conduction, but **not** by bone conduction, it infers that the inner ear and central auditory pathways are normal and that the site of hearing loss is localized to the **outer** ear or **middle** ear. Diseases affecting the outer or middle ear cause interruption of effective sound conduction — and the resulting deficit is termed a **conductive** hearing loss. The difference between air conduction threshold and bone conduction threshold on the audiogram is called an “air-bone gap”.



The audiogram shows the audiometric pattern of a conductive hearing loss. When sound transmission in the outer or middle ear is decreased, auditory thresholds to sound transmitted through air are poorer. However, bone stimulation (which directly stimulates the inner ear and bypasses the outer and middle ear) shows normal hearing. In this example, there is a 30 dB conductive hearing loss that affects all frequencies equally.



If a hearing loss is detected by **both** air and bone conduction methods, one can conclude that the cause of hearing loss is in the **inner ear** or **central auditory pathways**. Diseases affecting the inner ear or central auditory pathway result in a *sensorineural* hearing loss. Additional tests can determine whether the site of lesion is the inner ear (a sensory loss) or central auditory pathway (a neural loss) but these are beyond the scope of our discussion.



The figure above shows the audiometric pattern of a sensorineural hearing loss. When function of the inner ear or central auditory pathway is affected, thresholds are poorer as tested both by air conduction and bone conduction. In this example, there is a 30 dB sensorineural hearing loss that affects all frequencies equally.

The table below summarizes possible results from audiometric testing and how tests results are analyzed.

#### Bone Conduction Threshold

		Bone Conduction Threshold	
		Normal	Abnormal
Air Conduction Threshold	Normal	Normal hearing	Not possible
	Abnormal	Conductive hearing loss	Sensorineural hearing loss

For instance, if bone conduction is normal and air conduction is normal, hearing is normal. If bone conduction is normal and air conduction is abnormal, there is a conductive hearing loss. Finally, in a sensorineural hearing loss, both bone and air conduction will be abnormal.

### **Tympanometry**

A tympanogram assesses the mobility or compliance of the tympanic membrane and thereby provides important information about the function of the *middle ear* including the tympanic membrane, ossicles, and Eustachian tube. When a tympanogram is performed, a sound is introduced into the ear canal. A microphone in the ear canal measures the intensity of the sound as it reflects off of the eardrum. If the eardrum is functioning normally, more of the sound energy will be ‘absorbed’ and little will be ‘reflected’ (i.e., there will be little impedance to sound transmission and this will be reflected in the tympanogram). If the eardrum is not functioning normally, more of the sound energy will be reflected and little will be absorbed i.e., there will be great impedance to sound transmission. For instance if there is a middle ear infection most of the sound is reflected back and the tympanogram is flat (low compliance). If a part of the tympanic membrane is flaccid or the ossicles are broken there is even greater compliance and the tympanogram will display an abnormal peak.

### **Otoacoustic Emissions**

Otoacoustic emission testing assesses the integrity and function of outer hair cells in the *inner ear*. When a very sensitive microphone is placed in the ear canal, sounds can be detected that are caused by traveling waves in the basilar membrane of the inner ear. The traveling waves (which are usually thought of as a response to sound stimulation) are conducted in ‘reverse’ through the ossicles and, in turn, vibrate the eardrum. The traveling waves are set into motion by the movement of outer hair cells. You will remember from the basic science lectures that these cells are controlled via efferents from the brain stem. If normal otoacoustic emissions are detected, it gives evidence of a normally functioning set of outer hair cells in the inner ear.

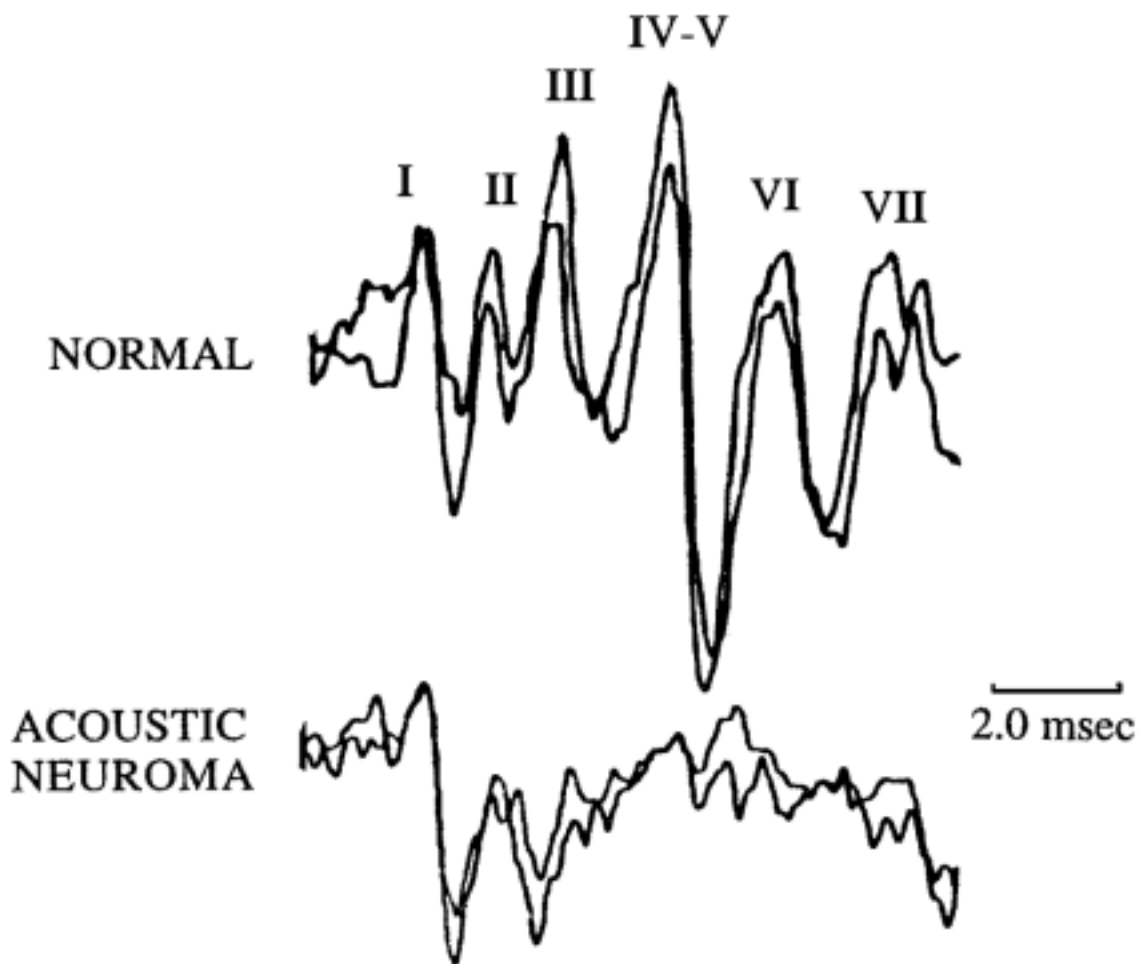
### **Evoked potentials- Auditory Brainstem Response**

Auditory brainstem response (ABR) testing is used to measure the function of the *central auditory pathways*.

Recording electrodes taped to the skull record the electrical activity of the brain (EEG). When a brief acoustic stimulus (e.g., a click or short tone burst) is presented to the ear there is a synchronized burst of action potentials generated in the auditory nerve which spreads up the central auditory pathway. Because of its very low amplitude (in the microvolt range) this wave of activity is generally buried in the EEG and can only be recovered using computerized signal-averaging techniques. When such methods are employed the complex waveform recorded is called the *auditory evoked potential* and it includes contributions from many sites that are activated sequentially in time along the auditory pathway. Remember from the brain stem and basic science auditory lectures that the auditory nerve projects to the cochlear nuclei. Then, the information heads toward the auditory cortex via the lateral lemniscus, superior olive, inferior colliculus, and medial geniculate body. An averaged waveform has multiple peaks and valleys stretched out over a period of several hundred milliseconds after the presentation of the acoustic stimulus.

The time period most commonly studied covers the first 10 msec after the stimulus is presented to the ear and represents **the electrical activity evoked in neurons in the auditory nerve and brain stem**. This is referred to in the experimental and clinical literature as the auditory brainstem response (ABR). This technique is very useful in studying hearing loss of central auditory origin, as may be caused by a lesion affecting the brainstem (e.g., acoustic neuroma or multiple sclerosis). It is also helpful in documenting the hearing loss in infants who cannot cooperate with a behavioral-based audiometric exam.

## The Auditory Brain Response (ABR)



The positive-going waves of the ABR are numbered I-VII in the figure above and are thought to reflect activity of the auditory nerve (I) and activity of cells in the auditory nuclei listed earlier. A hearing loss, whose etiology is a disease affecting the auditory nerve or brain stem, will degrade, destroy or prolong time intervals between the peaks of the waves. The figure shows a highly distorted ABR resulting from an acoustic neuroma. Since the neuroma affects the auditory nerve, the rest of the ABR is abnormal. So, what is the take home message to be **BOLDED**? Well, it is that the ABR can test the integrity of the of central auditory nuclei. Such a tool is especially important when dealing with infants who are too young for other tests.

## PATHOPHYSIOLOGY OF EXTERNAL EAR

From the standpoint of hearing and hearing disorders, abnormalities of the pinna and external ear canal that result from any condition can cause a *blockade to sound conductance*. As described above, they create a *conductive hearing loss*. Naturally, hearing loss may be only one of the considerations for intervention in an external ear disorder. There are a number of conditions that may present themselves, some of which are unrelated to hearing disorders. The presence of foreign bodies in the ear canal and infection of the skin of the external auditory canal (otitis externa) are conditions most commonly associated with conductive hearing loss in the external ear.

### Foreign bodies

Almost anything can become lodged in the external auditory canal. Even naturally occurring cerumen may become the impacted material which results in a noticeable conductive hearing loss. Probing the ear canal with an instrument (e.g., a Q-tip) is dangerous for it can force the impacted material further into the canal and can perforate the tympanic membrane resulting in damage to the middle ear structures.

### Otitis externa

Infection of the skin of the external auditory canal results in otalgia (ear pain), otorrhea (ear drainage) and a conductive hearing loss. It is usually caused by a break in the skin of the ear canal and prolonged water exposure—conditions that are favorable for bacterial growth. Otitis externa is often called “swimmer’s ear.”

### Congenital malformations

Congenital malformations of the pinna and external ear canal are related to developmental defects of the first and second branchial arches and the branchial groove which joins the first pharyngeal pouch to form the external ear canal. Malformation of the external ear canal results in an atresia, which is a conductive blockade of connective tissue or bone. Maldevelopment of the first pharyngeal pouch, leads to abnormalities in Eustachian tube, middle ear, and mastoid differentiation. These malformation may occur singly or in combination.

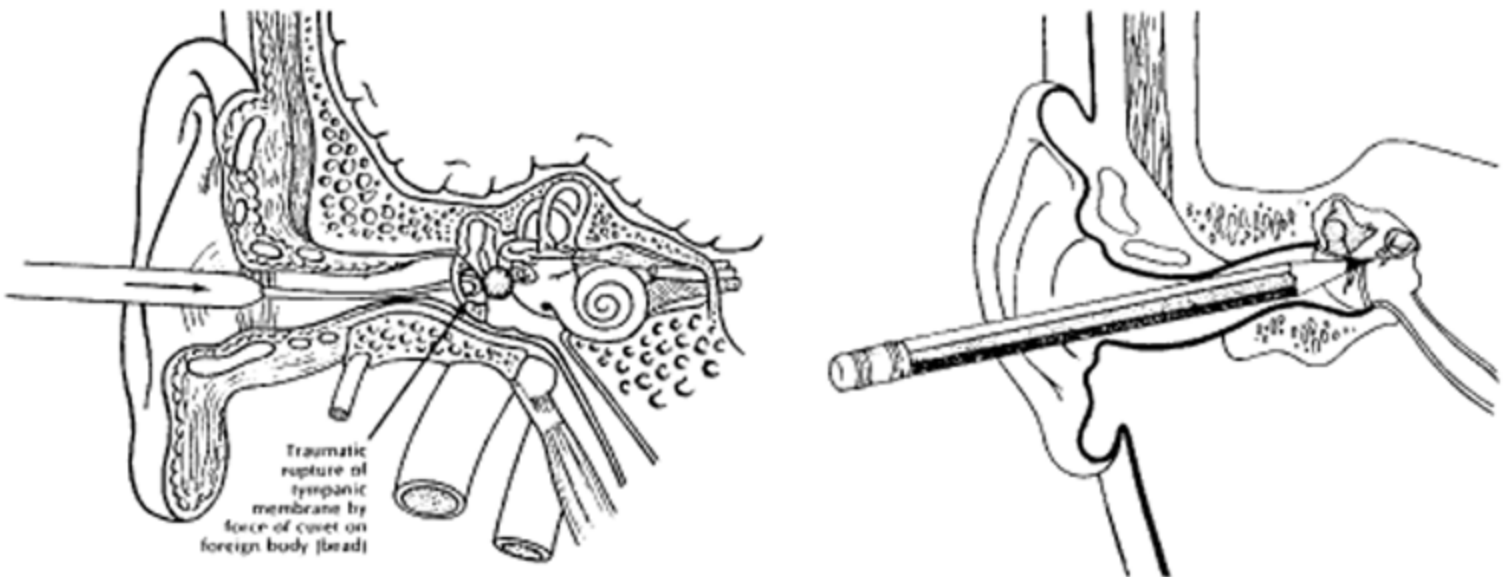
## PATHOPHYSIOLOGY OF THE MIDDLE EAR

Disorders of the middle ear and mastoid arise from maldevelopment, inflammatory and degenerative processes, trauma, or neoplastic disease. From the point of view of hearing, these disorders may result directly in a *conductive hearing loss*. Some of them (e.g., inflammation and neoplastic disease) can become serious medical problems if not treated, with involvement of the inner ear and systems beyond.

## Tympanic membrane perforations

Perforation of the tympanic membrane is a common serious ear injury that may result from a variety of causes including projectiles or probes (e.g. Q-tips, pencils, paper clips, etc.), concussion from an explosion or a blow to the ear, rapid pressure change (barotrauma), temporal bone fractures, and middle ear infections. Perforations may be associated with damage to the ossicles. Examples are shown below.

Hearing loss accompanying tympanic membrane perforation is conductive in nature. There may be two mechanisms at play that contribute to this hearing disorder. First, the normal structure, and hence action, of the tympanic membrane is altered. Sound pressure on either side of the tympanum is quickly equalized. The degree of conductive hearing loss is directly related to the size of the perforation. Second, sound waves that enter the middle ear space reach both the round and oval windows and do so nearly in phase.



Recall that under normal conditions inward motion of the stapes footplate in the oval window results in an outward movement of the round window, and vice versa. A “leaky” tympanic membrane means that the normal “push-pull” action of these two membranes is, to some extent at least, circumvented and as a result the sound energy entering the inner ear is reduced.

## Ossicular chain injuries

The various injurious mechanisms associated with the tympanic membrane apply to the ossicles as well, occasionally even without rupture to the membrane itself. **Closed-head injuries, especially if associated with a temporal bone fracture, are common causes of ossicular chain disruption.** A major conductive hearing loss (30-60dB) may result which does not improve after tympanic membrane repair. The most common traumatic ossicular chain lesion is a incudostapedial joint dislocation with or without a fracture of the long process of the incus. However, just about any imaginable fracture or displacement can be found. Ossicular dislocation interrupts the normal

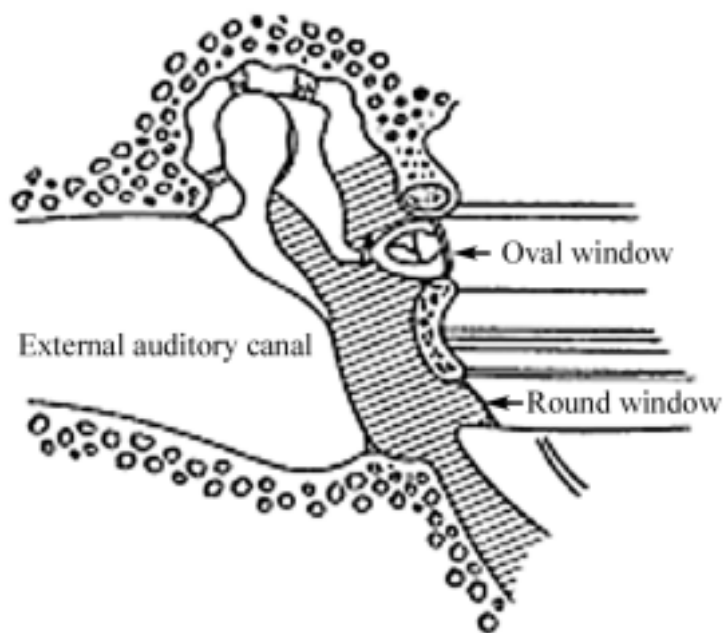
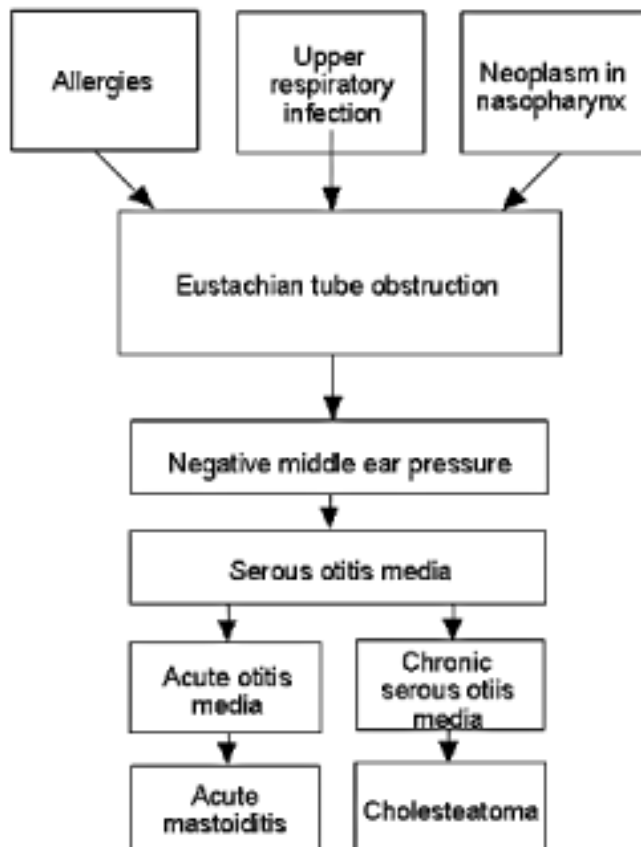
transmission of sound energy from the tympanic membrane to the fluid of the middle ear. Hence, under these conditions the impedance matching mechanism, which alone overcomes the nearly 30 dB loss of energy when sound waves in air meet a fluid boundary, is lost.

### Inflammatory processes in the middle ear - Otitis media

Inflammatory diseases of the middle ear are related and the occurrence of one often leads to the other. The pathogenesis of otitis media is shown in the following. The underlying cause of otitis media is Eustachian tube dysfunction.

Otitis media evolves from the common cold, allergies, cigarette smoke exposure, or anything that can cause obstruction of the Eustachian tube. For instance, loss of ciliary action, hyperemic swelling, and increased production of mucus associated with an upper respiratory infection leads to temporary closing of the Eustachian tube and, as a result, *negative pressure develops within the middle ear* and the tympanic membrane bulges in (toward the middle ear). This has two consequences: One involves pressure and pain as the result of retraction of the tympanic membrane innervated primarily by the trigeminal nerve. The other is a mild conductive hearing loss due to added **stiffness** of the middle ear transmission mechanism. An increase in the stiffness of the ear ossicle means that lower frequency sounds are especially affected.

Negative pressure within the middle ear, if left unrelieved, can lead to fluid accumulation in the normally air-filled middle ear space. This condition is referred to as *serous (or secretory) otitis media*. As noted above, it may have predisposing factors including lymphatic engorgement, cleft palate, hypertrophic adenoids, allergic rhinitis, and neoplasms of the nasopharynx and, thus, may develop in the absence of infection. The middle ear is filled with an amber-colored serous transudate. The hearing loss is now further complicated by the presence of a fluid-air boundary at the tympanic membrane and mass-friction loading of the ossicles. The patient has a worsening conductive hearing loss with an immobile tympanic membrane. The degree of hearing loss will vary depending on the amount and viscosity of the transudate and tympanic membrane edema. It may be as low as 5-10 dB (and not too noticeable) to 30-40 dB (where it is disabling).





The disease can develop rapidly into an *acute otitis media* as organisms migrate to the middle ear from the nasopharynx. The fluid changes rapidly from serous to sero-purulent and finally to the purulent stage. Clinically, it is usually characterized by ear pain, fever and signs of systemic illness. Acute otitis media is a potentially serious disease. Because of the relationships between the middle ear cavity and surrounding structures, there is a wide range of possible complications that involve areas outside of the middle ear itself. The infection may break through the confines of the middle ear and lead to intracranial complications including meningitis, brain abscesses, or lateral sinus thrombophlebitis. The infection can lead to facial paralysis (affecting the facial nerve as it runs through the middle ear) or can spread to the labyrinth (labyrinthitis). Another important possible sequelae is acute mastoiditis. Here there is bony destruction and coalescence of the mastoid air cells.

**Take home message - middle ear problems = conductive hearing loss**

## DISORDERS OF THE INNER EAR

**Impairment in the cochlear transduction mechanisms, in auditory nerve transmission, or in both, results in a *sensorineural hearing loss***

Because both the auditory and vestibular structures of the inner ear have a similar embryological origin, and because they share the same fluid environment, a disorder of one frequently includes a disorder of the other, resulting in a complex of symptoms.

The inner ear is vulnerable to damage or destruction from a variety of sources. Malformations of the labyrinth may be inherited or acquired. Inflammatory and metabolic processes may disrupt permanently normal vestibular and auditory function at the level of the end organ. Drugs and other substances have teratogenic effects on the inner ears of the fetus and destructive effects on the cochlea and vestibular organs in young and adult individuals. Trauma, either physical or acoustic, can cause hearing loss and vestibular damage. Viral infections may destroy the receptor organs, especially in utero.

**Sensorineural disorders of hearing fall into two categories: congenital and acquired**

### **Congenital disorders**

Hereditary syndromes include labyrinthine disorders that are associated with no other abnormalities, and those that are associated with external ear malformations, integument disease, ophthalmic lesions, CNS lesions, skeletal malformations, renal disease, and miscellaneous defects. One example where there is bilateral inner ear deformation is Usher's Syndrome. This disease accounts for about 10% of all hereditary deafness; there is no vestibular involvement and it is associated with retinosis pigmentosa (degeneration of rods in the retina). Another cochlear deformation is associated with Waardenburg's Syndrome. This accounts for 2-3% of all cases of congenital deafness in the U.S. It is associated with a white forelock and widened intercanthal distance.

## **Acquired disorders**

### **Traumatic Lesions**

Noise induced hearing loss - Excessive noise can cause permanent damage to the cochlea. It may occur as the result of a sudden blast (e.g. gun shot) or it may come because of lengthy exposure to high intensity sound (e.g. factory noise). Even relatively brief exposure to a high-noise environment is potentially hazardous to the health of the organ of Corti as evidenced by the studies done at a 4-hour Bruce Springsteen concert in St. Louis and during the 1987 Twins-Cardinals World Series games in domed stadiums (see following journal abstracts).

TEMPORARY THRESHOLD SHIFTS FROM ATTENDANCE AT A ROCK CONCERT. W. Clark and B. A. Bohne, Central Institute for the Deaf and Dept. of Otolaryngology, Washington University School of Medicine, St. Louis, MO 63110). From J. Acoust. Soc. Am., 79:548, 1986.

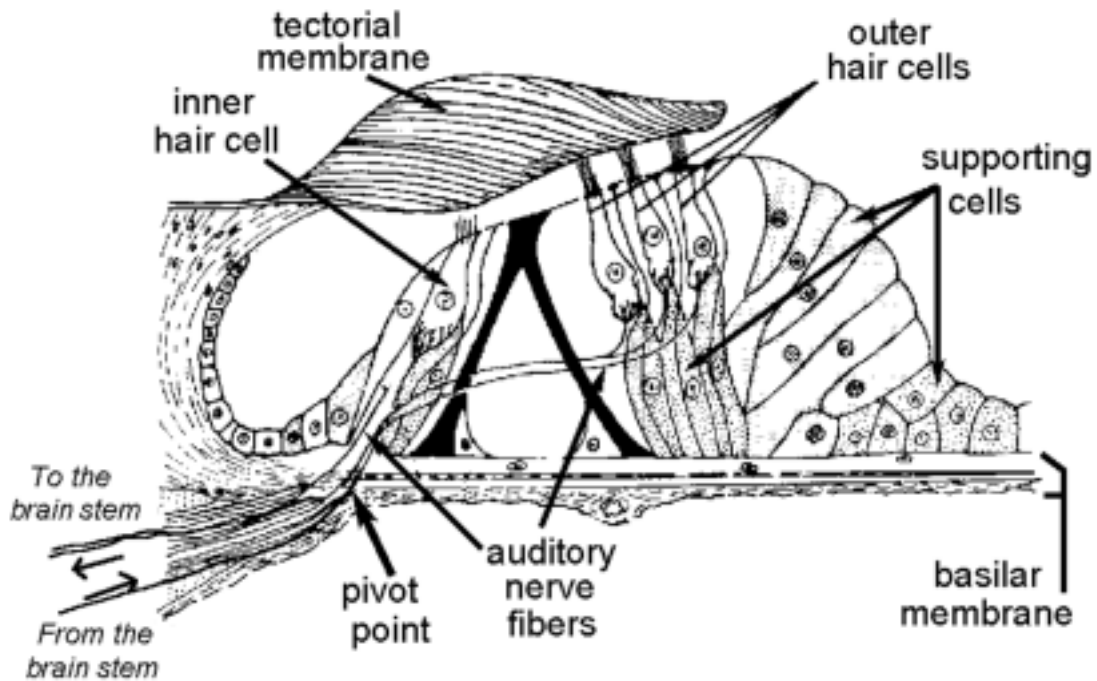
The relation between exposure level and hearing loss in rock concert attendees was studied. Six volunteer subjects, ages 16-44, participated. All except the 44 year old had normal hearing sensitivity as revealed by audiometric evaluations made immediately before the concert. They attended a Bruce Springsteen concert at the St. Louis Arena and returned to CID for another hearing test within 30 min. following the concert. Noise exposure was assessed by having two subjects seated at different locations in the arena, wear calibrated dosimeters during the event. Sixteen hours after the concert all subjects returned for a final audiometric evaluation. Results indicated the average exposure level was 100-100.6 dBA during the 4 1/2 hr concert. Five of the six attendees had significant threshold shifts (<50 dB) predominately in the 4-Khz region. Measures made 16h after the concert and thereafter indicated that hearing returned to normal in all subjects. Although no PTS was observed, comparison of these data with studies of hearing loss and cochlear damage in animal models suggests that these subjects may have sustained some sensory cell loss from this exposure. (Work supported by NIOSH and NINCDS.)

The loss is of cochlear origin and is most pronounced in the vicinity of 4 kHz. This frequency corresponds to the frequency region of enhanced sensitivity due to the resonance properties of the external ear. The consequence of exposure to intense sound is a temporary or a permanent hearing loss. Whether one or the other condition prevails depends on a number of variables including the intensity, frequency, and duration of the sound exposure. It is believed that the structural damage to the inner ear that accompanies a permanent hearing loss arises from the interplay of mechanical and metabolic processes.

### **There are several mechanisms that underlie peripheral hearing disorders**

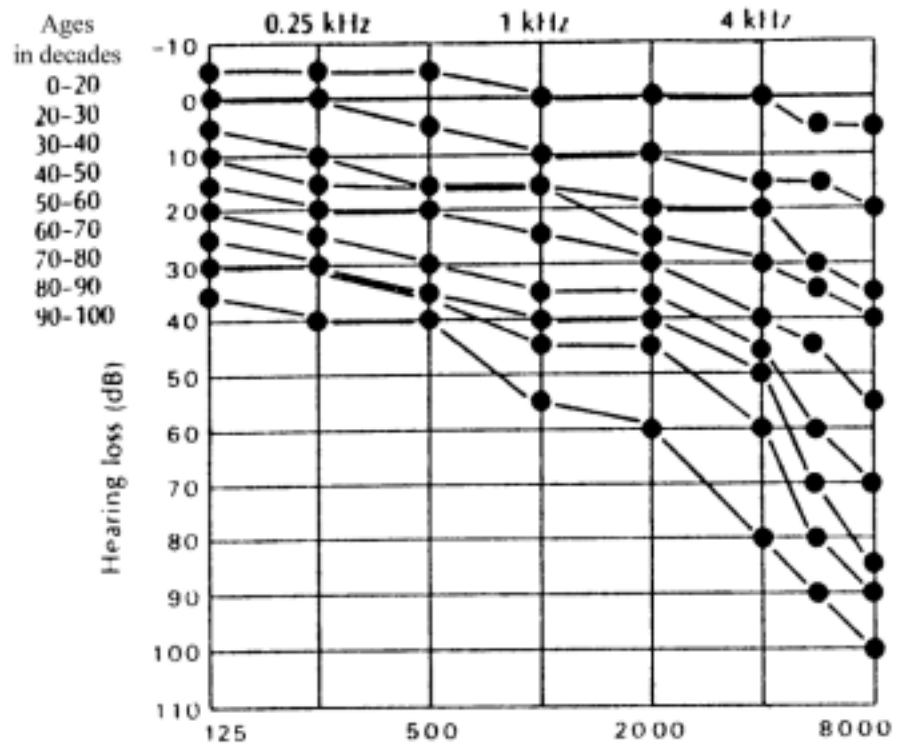
#### **Inner vs. Outer Hair Cells**

Outer hair cells (OHCs) are more susceptible than inner hair cells (IHCs) to acoustic overstimulation. One reason may be that OHCs, because of their greater distance from the fulcrum (pivot point) of the basilar membrane, undergo greater velocity of motion and hence are at greater risk of mechanical damage. Second, the direct mechanical linkage of OHC stereocilia with the tectorial membrane may enhance this cell's susceptibility. Thirdly, the difference may be metabolic, reflecting the differences in internal organelle structure of the IHCs and OHCs. It is noted that OHCs are also more susceptible to ototoxins. Also, the first row of OHCs seems to be at greatest risk.



### Presbycusis

The term “presbycusis” has been traditionally applied to the **hearing loss that normally accompanies aging**. Although it commonly refers to hearing loss resulting from degenerative changes in the cochlea alone, it is now clear that the aging process affects the whole auditory system and that hearing loss of old age probably involves changes in the **middle ear, inner ear and central auditory pathways**. Although there is no clear relationship between age-related changes in the middle ear and the audiographic findings, there are documented cases of ossicular fixation and arthritic changes in ossicular joints with fibrous and calcific changes. The correlation between the types and patterns of cochlear lesions and the patterns of hearing loss has long been recognized. While there may be wide variations in these patterns, in general, the common finding is a high frequency sensorineural hearing loss associated with degeneration of the organ of Corti in the base (high frequency representation) of the cochlea. Hearing loss may progress over time to involve the apex i.e., lower frequencies. The above figure shows audiograms taken at decade intervals. Note here the gradual and progressive loss of sensitivity at high frequencies. Presbycusic individuals may also have central nervous system involvement in their hearing disorder. Within the cochlear nuclei, for example, the injury may

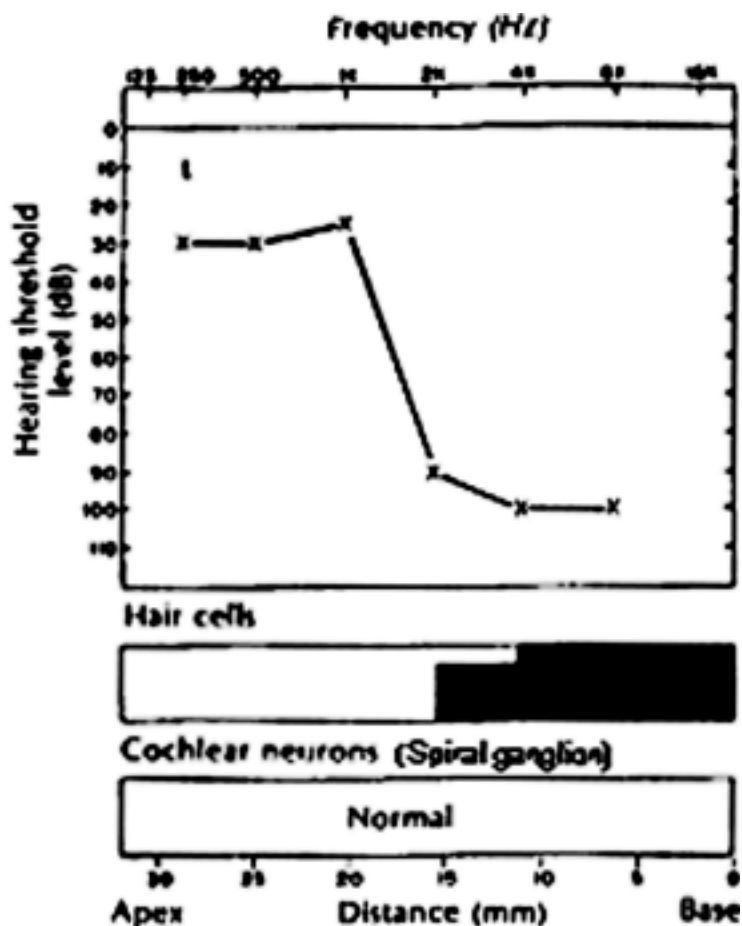
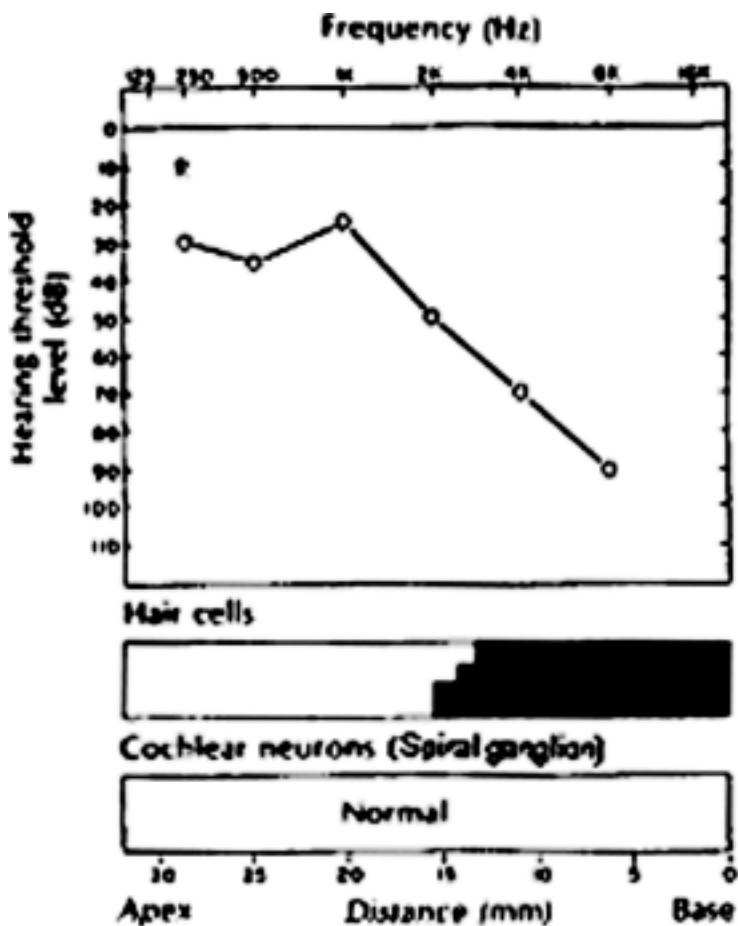


range from little or no alteration in cellular structure to complete destruction of cells. Whether this occurs independently of a cochlear lesion is currently not known.

### Ototoxicity

It has long been known that certain drugs and chemicals can have strong effects on the auditory and vestibular receptors of the inner ear. The clinical signs of ototoxicity are variable but include one or more of the following symptoms: sensorineural hearing loss, tinnitus, and “dizziness” of one description or another. Over the past 40 years, there has been a steady accumulation of data, from both the clinic and the laboratory, on the mechanisms of action of various ototoxins. With current understanding of the normal cellular-molecular mechanisms of receptor cell action, we are on the threshold of understanding the mechanisms of many of the disorders that affect hair cells. A brief description is given a few of some of the more common agents and their actions.

**Aminoglycoside antibiotics** - Most of the antibiotics recognized as having ototoxic properties belong to the family of aminoglycosides. The primary ones that require respect are **streptomycin**, dihydrostreptomycin, neomycin, gentamicin and tobramycin. The figure below shows the audiograms from the left and right ears of an individual treated with kanamycin. Below the audiogram are plots of hair cell and spiral ganglion cell loss in each of the ears taken after postmortem histological preparation of the temporal bones. Note the correspondence between hair cell loss in the basal half of the cochlea and the high frequency hearing loss which is typical of aminoglycoside ototoxicity.



Clinical and experimental evidence collected from human and animal studies over many years has given a picture of the pathophysiological mechanisms which underlie the damage inflicted by these agents. First, the toxic substances must reach the labyrinthine fluid either via the blood stream or, when applied topically to the middle ear, by direct penetration of the oval and/or round windows. Second, primary damage is to the hair cell; auditory nerve fibers may degenerate secondary to sensory cell degeneration. Both kanamycin and neomycin affect first the *outer hair cells* of the cochlea base; over time the lesion progresses to the cochlear apex. Inner hair cells seem less vulnerable to these agents. Third, at the cellular-molecular level, the action of aminoglycosides seems to alter plasma membrane permeability for there is microscopic evidence for the swelling of sensory hairs with the deformation of the cell surface. This may involve several processes upon which cellular integrity depends. Two of them are the cellular metabolic and protein synthesizing machinery, for there is also evidence that mitochondria and ribosomes are damaged. Another is that the ionic channels which are responsible for mechano-electric transduction to occur may be blocked or otherwise affected.

**Diuretics** - Animal studies have shown that intravenous injection of ethacrynic acid or furosemide produces within seconds depression of the cochlear microphonic potential (hair cell receptor potential) and auditory nerve action potentials and a decrease in endocochlear potential which is necessary for normal transduction and transmission in the inner ear receptor organs. Anatomical changes include *outer hair cell* degeneration in basal and middle turns of the cochlea. In those cells that survive there may be distortion of the stereociliary bundle. Moreover, there are marked changes in the *stria vascularis*, with intra- and extracellular edema and destruction of the intermediate cell layer. Thus, it would appear that diuretic ototoxicity involves changes in the transduction and transmission properties of the hair cells and a breakdown in the intra-labyrinthine secretory mechanisms of the stria vascularis.

**Salicylates** - High doses of salicylates predictably produce a bilaterally symmetric, flat hearing loss up to about 40 dB HL. The magnitude of the hearing loss is directly related to the serum levels of the substance. The hearing loss and accompanying tinnitus are completely reversible within 24-72 hours after the drug is discontinued. There is no consistent morphological change observed in the inner ears of humans or animals subjected to high doses of salicylates. While biochemical changes of the perilymph and endolymph have been noted along with consistently reduced electrical activity of the cochlea and auditory nerve, the precise mechanisms of this form of ototoxicity are not known.

## CENTRAL CAUSES OF HEARING LOSS

### **Neonatal hyperbilirubinemia - bilirubin encephalopathy**

Bilirubin, a yellow pigment, is the major end product of hemoglobin metabolism. It has long been known that, in human neonates, there is a close association between elevated blood bilirubin levels and disorders of the central nervous system. The most extreme neurological consequence of hyperbilirubinemia is referred to as “kernicterus” - a condition that may include hearing impairment, choreoathetosis, spasticity, oculomotor problems, cognitive dysfunction, and mild forms of mental retardation. Classical kernicterus in term infants, resulting from Rh incompatibility, has been in many places nearly eliminated by prophylaxis and the use of early exchange transfusion. With the decrease in the incidence of classical kernicterus induced by Rh incompatibility, attention has shifted

to the occurrence of this disorder in premature and gravely ill infants. The hearing loss that accompanies hyperbilirubinemia is of the **sensorineural type**. In studies of temporal bones of humans and animals with this condition there has been no clear-cut evidence of damage to the inner ear structures. Rather, the damage appears to occur in the auditory nuclei of the brainstem; neurons in the cochlear nuclei, in particular are severely damaged or destroyed.

**Tumor of the VIIIth nerve** - Lesions of the eighth nerve are characterized by tinnitus, sensorineural hearing loss, mild vertigo, and in some patients, other cranial nerve signs. The classic lesion is the so-called “acoustic neuroma”, a benign tumor that is usually not of auditory nerve origin nor is it a neuroma (tumor of a neuron). The tumor is a schwannoma typically arising from the vestibular nerve within the internal auditory canal—so a more accurate term is vestibular schwannoma. The growth of the tumor in the vestibular nerve does not typically produce vestibular signs and it is not until the tumor compresses the auditory nerve that it is noticed. The most common first symptom is unilateral tinnitus (ringing in the ear). This may be followed by a progressive (and unilateral) sensorineural hearing loss as shown on an audiogram. The mechanism for the hearing disorder probably involves the disruption of normal transmission of action potentials in the fibers of the auditory nerve due to compression by the tumor. Pressure from the growing tumor may eventually involve cranial nerves VII, VI and V. An ABR will show abnormalities of waveforms in cases of central auditory pathology such as an acoustic neuroma.

## HEARING LOSS AND ITS EFFECTS ON COMMUNICATION

Hearing loss may be categorized by degree. This table below does not take into account some important variables, including age of the individual which, as we will see later impacts critically on language development.

### **25-40 dB**

Misses hearing many consonants, difficulty in auditory learning, mild speech - language problems

### **40-65 dB**

Speech - language retardation, learning disability, hears little or no speech at normal conversational levels

### **65-90 dB**

Voice pathology, aural language seriously compromised, severe learning problems

### **>90 dB**

Profound hearing loss (deaf), voice-speech characteristic of deaf, severe learning disabilities

## PRACTICE QUESTIONS

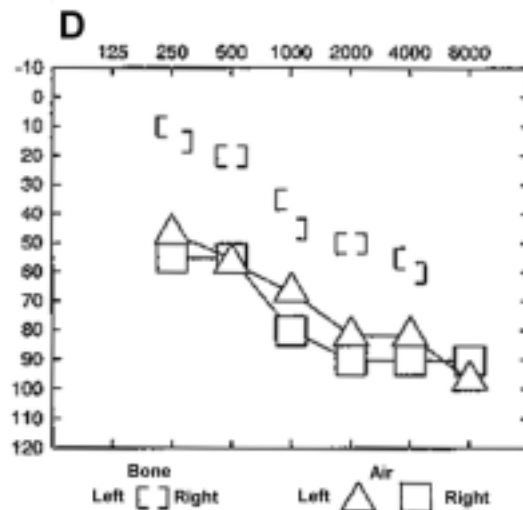
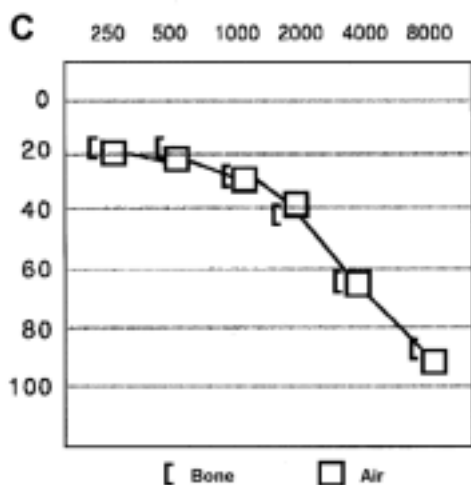
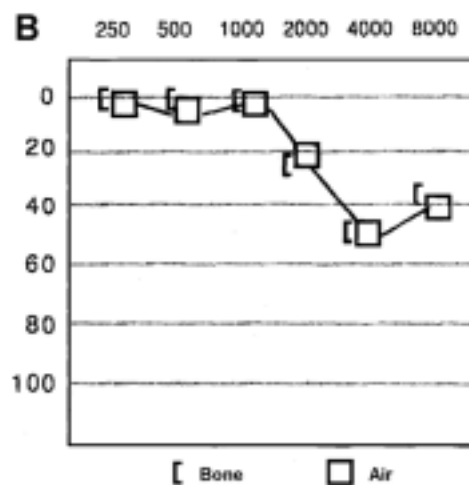
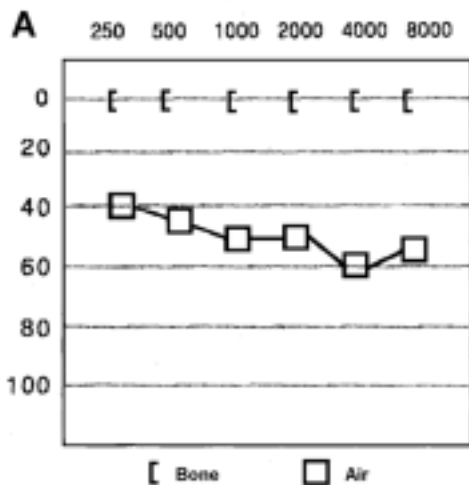
1. Acute otitis media is associated with:
  - A. a conductive hearing loss
  - B. a sensorineural hearing loss
  - C. a combined conductive and sensorineural hearing loss
  - D. a normal tympanogram
  - E. a normal ABR
  
2. Acute otitis externa is associated with:
  - A. a conductive hearing loss
  - B. a sensorineural hearing loss
  - C. a combined conductive and sensorineural hearing loss
  - D. a normal ABR
  - E. none of the above
  
3. Excessive noise exposure is associated with:
  - A. a conductive hearing loss
  - B. a sensorineural hearing loss
  - C. a combined conductive and sensorineural hearing loss
  - D. damage mainly to inner hair cells
  - E. abnormal tympanogram
  
4. An audiogram from a patient with multiple sclerosis would show:
  - A. a conductive hearing loss
  - B. a sensorineural hearing loss
  - C. a combined conductive and sensorineural hearing loss
  - D. a normal ABR
  - E. an abnormal tympanogram
  
5. Otitis media results from:
  - A. obstruction of the round window
  - B. obstruction of the Eustachian tube
  - C. obstruction of the oval window
  - D. obstruction of ossicular motion
  - E. none of the above
  
6. Thresholds to bone conducted sound stimuli measure the function of:
  - A. the outer and middle ear
  - B. the middle and inner ear
  - C. the inner ear and central auditory pathways
  - D. the entire auditory pathway
  - E. none of the above

7. Thresholds to air conducted sound stimuli measure:
- A. the outer and middle ear
  - B. the middle and inner ear
  - C. the inner ear and central auditory pathways
  - D. the entire auditory pathway
  - E. none of the above
8. Serous otitis media causes a hearing loss by:
- A. damaging hair cells
  - B. reducing the effective ratio between the tympanic membrane and oval window
  - C. reducing the lever ratio of the incus and malleus
  - D. causing an air-fluid interface at the tympanic membrane
  - E. none of the above
9. A 65 year old man developed a unilateral *conductive* hearing loss. This may be caused by:
- A. an acoustic neuroma
  - B. noise exposure
  - C. nasopharyngeal cancer
  - D. aspirin
  - E. none of the above
10. A 75 year old man developed a unilateral sensorineural hearing loss. This may be caused by:
- A. a unilateral acoustic neuroma
  - B. noise exposure from a rock concert
  - C. nasopharyngeal cancer
  - D. aspirin
  - E. none of the above
11. A child with acute bilateral otitis media:
- A. will have a sensory hearing loss
  - B. can develop meningitis
  - C. will have an abnormal auditory thresholds to bone-conducted sound stimuli
  - D. will hear low frequency sounds better early on (increase stiffness of ossicles)
  - E. will hear high frequency sounds better early on when there is a lot of junk in the middle ear (increased mass)
12. Which of the following statements is false?
- A. a tympanogram would reveal a punctured eardrum
  - B. damage to the cochlear hair cells can be revealed by studying the otoacoustic emission
  - C. outer hair cells are damaged by streptomycin
  - D. there is one row of outer hair cells
  - E. when descending in an airplane, your ear ossicles become stiffer (true due to negative pressure in the middle ear)



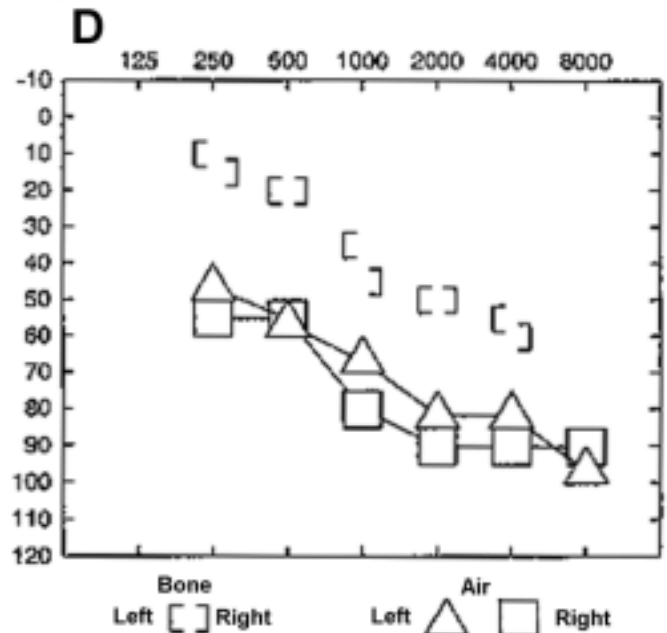
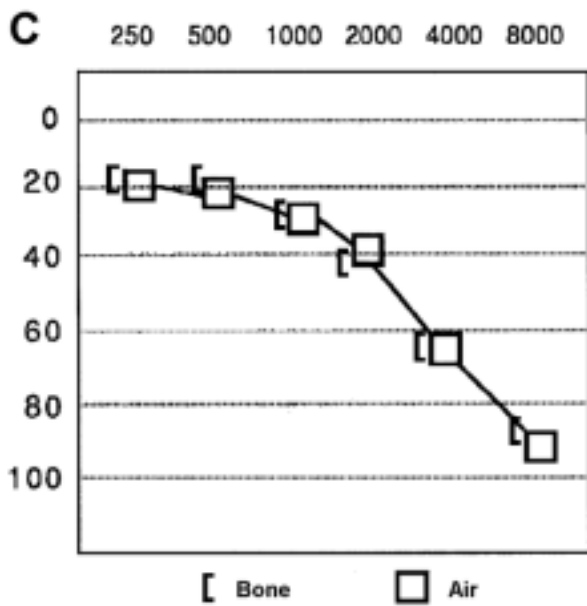
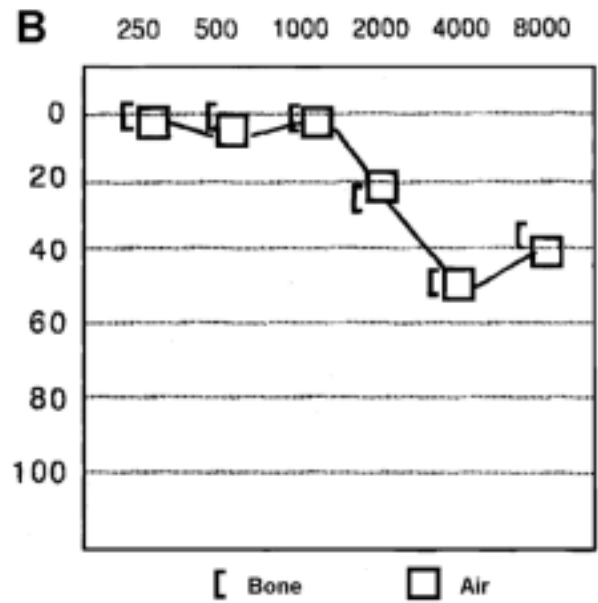
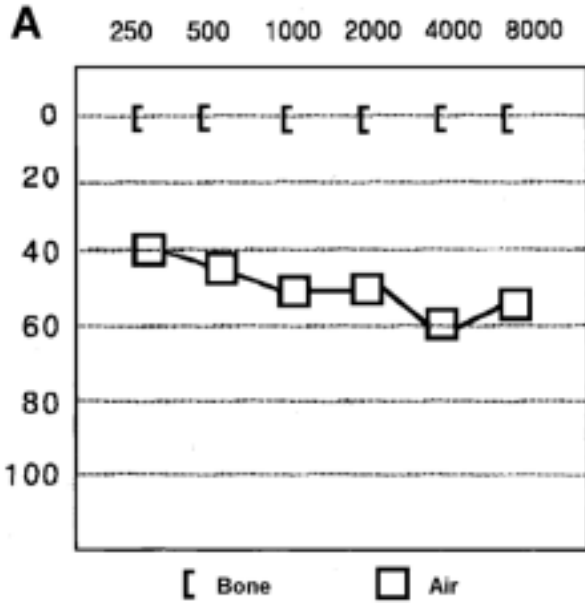
**Match the clinical scenario to the most likely audiometric pattern. KNOW THIS!!**

1. An 84 year-old woman presents to her primary care physician with slowly worsening hearing over the past several years. She notes difficulty hearing when there is a high level of background noise.
2. A 45 year old woman sees her primary care physician with a chief complaint of hearing loss in the left ear. She had been scuba diving the week before and had sudden severe left-sided ear pain while descending. Although the ear pain went away the next day, she has noted decreased hearing ever since.
3. A 43 year old woman sees her primary care physician with a chief complaint of hearing loss in the left ear. She had had an upper respiratory infection two weeks prior that has since improved. However, the hearing loss has not improved.
4. A 26 year old man received high dose intravenous gentamicin to treat a pneumonia caused by *Pseudomonas* sp. Following recovery from the pneumonia, he noted a hearing loss and a high pitch ringing in both ears.
5. A 47 year old farmer has had daily exposure to high intensity noise including a tractor, chain saw and other powered equipment for the past 20 years. He also fires a rifle without hearing protection when he goes out to hunt deer, duck and squirrels.



6. Which of the following is true regarding the audiogram shown below?

- A. the patient could have otosclerosis
- B. the patient would have normal otoacoustic emissions
- C. there is an air bone gap
- D. the patient may have dirty (real dirty) ears
- E. three of the above are true



## Answers to practice questions

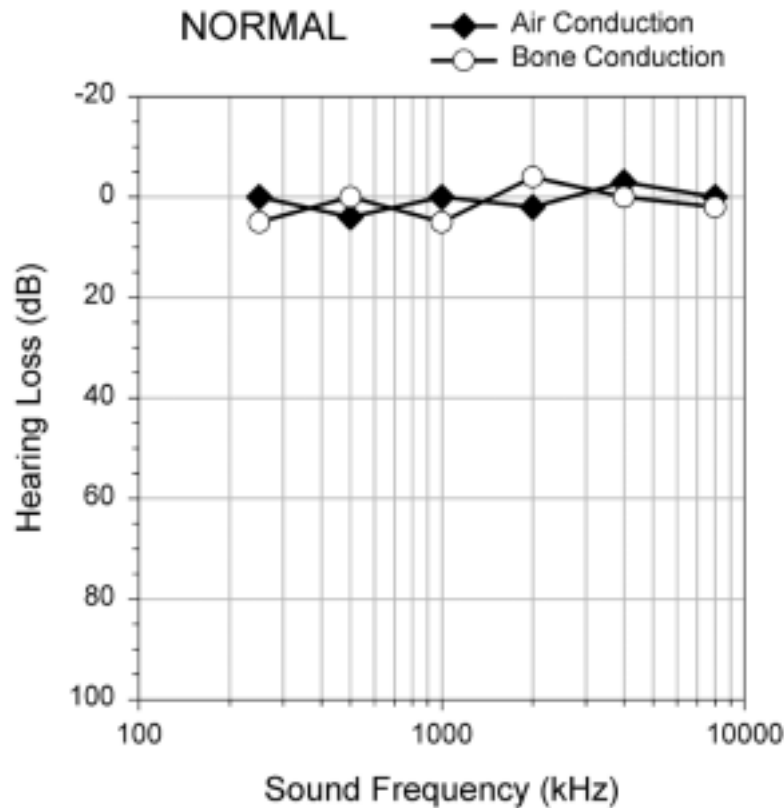
### Multiple choice

1. A
2. A
3. B
4. B
5. B
6. C
7. D
8. D
9. C
10. A
11. B
12. D

### Audiograms

1. C (Audiogram showing presbycusis exhibits a gradual slope that includes the highest frequencies. Background is bothersome since the patient has trouble hearing the conversation (higher frequency) and the lower frequencies are bombarding away.)
2. A
3. A
4. C
5. B
6. A TRUE this is a mixed hearing loss. Anytime time bone is better than air = a conductive loss and such a loss could result from otosclerosis  
B. FALSE there is a sensorineural loss. So otacoustic emissions would be abnormal if there is hair cell damage. If there is only CN VIII nerve damage (sensorineural deficit) the OAEs would also be abnormal as the efferents to the OHCs are damaged  
C. TRUE  
D. TRUE  
E. TRUE (A, C and D)

## Auditory System Case Histories



### CASE 1:

A 14-year old girl is brought to the emergency room by her mother who tells you that she thinks her daughter “poked something in her ear.” The girl states that while she was cleaning her ears with a Q-tip, her brother pushed her arm and the Q-tip went deep into the left ear canal. She is in pain and cannot hear well from that ear.

You examine her ears otoscopically.

Describe the otoscopic findings.

What type of hearing loss is this person experiencing?

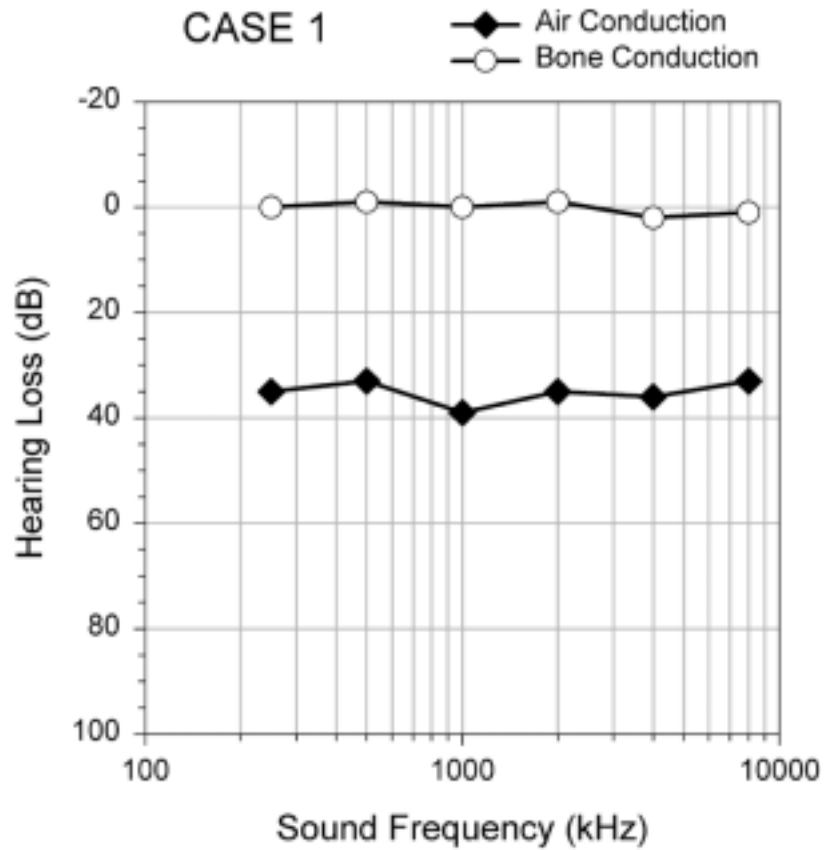
What do you base your conclusions on?

What are the pathophysiological mechanisms that underlie this hearing impairment?

After several months, the girl is still experiencing a hearing loss in that ear. You again examine the ear otoscopically and order an audiometric workup.

Describe the otoscopic findings at this stage.

Describe the audiogram.



What type of hearing loss is this person experiencing?

What do you base your conclusions on?

What are the pathophysiological mechanisms that underlie this hearing impairment?

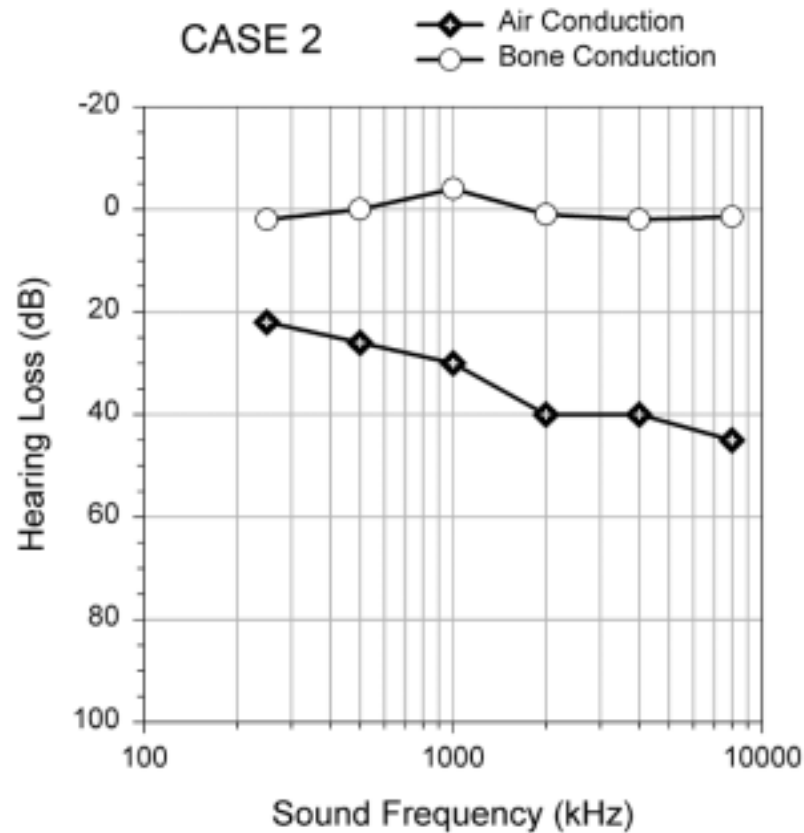
**CASE 2:**

A three year old child is brought to you by his mother because he has started to complain of an earache. He is also having trouble hearing. The child has had a cold for several days. You also decide to have a hearing test done (because this is a good learning experience for medical students).

You examine the child’s ear otoscopically.

Describe the physical findings.

Describe the audiometric findings you would expect to see early and later during the course of this condition.



What type of hearing loss is this child experiencing?

What do you base your conclusion on?

What are the pathophysiological mechanisms that underlie this hearing loss

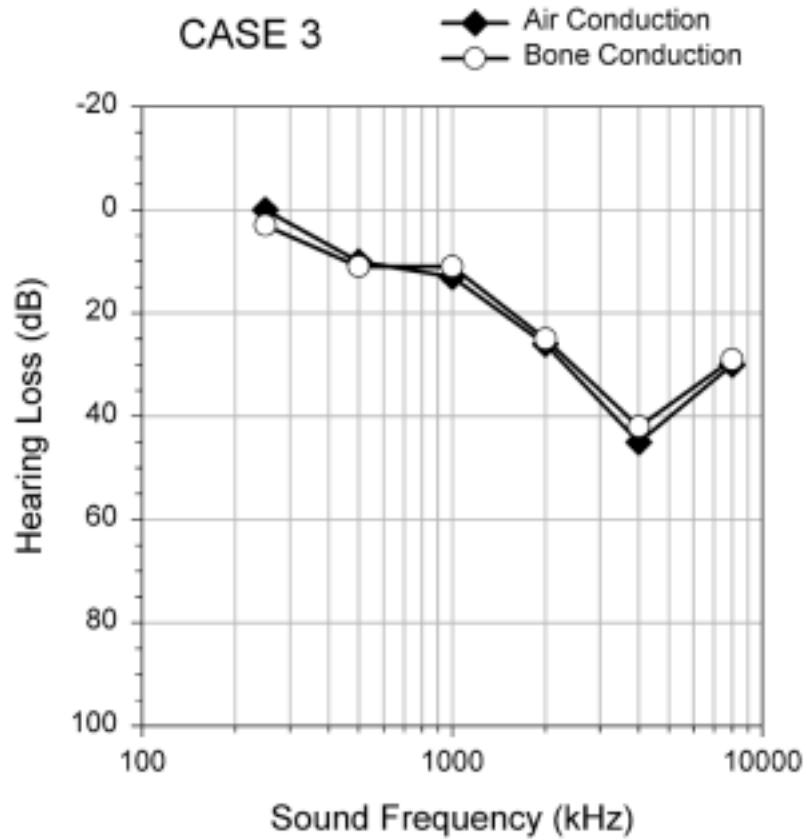
### CASE 3:

A 24-year old man complains that he is becoming 'hard of hearing'. He noticed this while he was serving with the U.S. Army during the Gulf War. His duty there was with an artillery company and, for a short but intense period of time, he was firing heavy shells across the desert skies. After returning home, he has been having greater and greater difficulty understanding everyday conversation. In a quiet room he has little difficulty, especially if he can concentrate on the speaker's face. Where he has problems is when there is any kind of background noise.

You examine his ears otoscopically. You also refer him to an audiologist for further evaluation of his hearing.

Describe the otoscopic findings.

Describe the audiometric findings.



What type of hearing loss is this person now experiencing?

What do you base your conclusion on?

What are the physiological mechanisms that underlie this hearing loss?

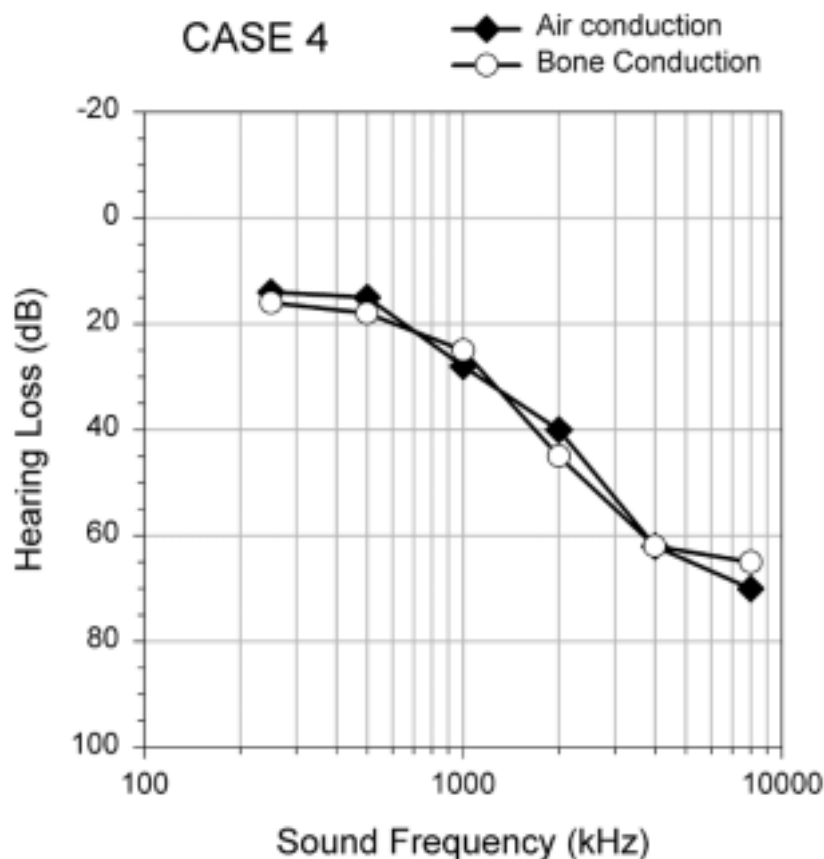
**CASE 4:**

During the course of a routine physical exam, a 76 year old man states that over the past 10 years it has been increasingly difficult for him to hear what others are saying. He especially notes difficulty in social situations when there are multiple competing sounds and he is trying to pay particular attention to one of them—he can tell that someone is talking, but cannot reliably tell what they are saying. He also has noted a high-pitch ringing in his ears—especially at night when it is quiet. He does not note a difference in hearing between ears. He does not have ear pain, drainage from the ear, or dizziness.

You examine his ears with an otoscope and order an audiometric examination.

Describe the otoscopic findings.

Describe the audiometric findings.



What type of hearing loss is this person now experiencing?

What do you base your conclusion on?

What are the physiological mechanisms that underlie this hearing loss?

### **CASE 5:**

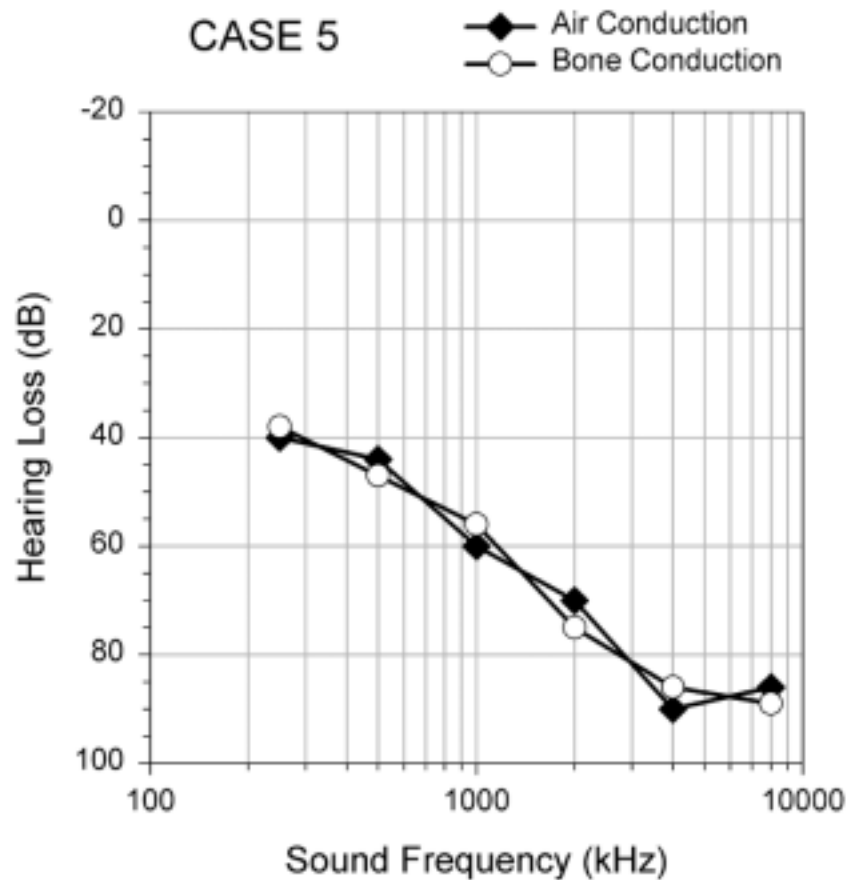
A mother brings her 3-year old child to see you because she suspects the child may be 'hard of hearing'. The child has not begun to speak and has made relatively few sounds since the babbling stage. The mother notices that even loud sounds, such as a banging door, fails to startle the child. During the interview, you discover that the mother had an undiagnosed illness, accompanied by a rash, during the early states of pregnancy. Otherwise, the pregnancy was uneventful. The child has no history of illness.

You examine the ears otoscopically. You also request consultation with an audiologist trained to test the hearing of young children.

Describe the otoscopic findings.

Describe the audiometric findings.





What type of hearing loss is this person now experiencing?

What do you base your conclusion on?

What are the physiological mechanisms that underlie this hearing loss?

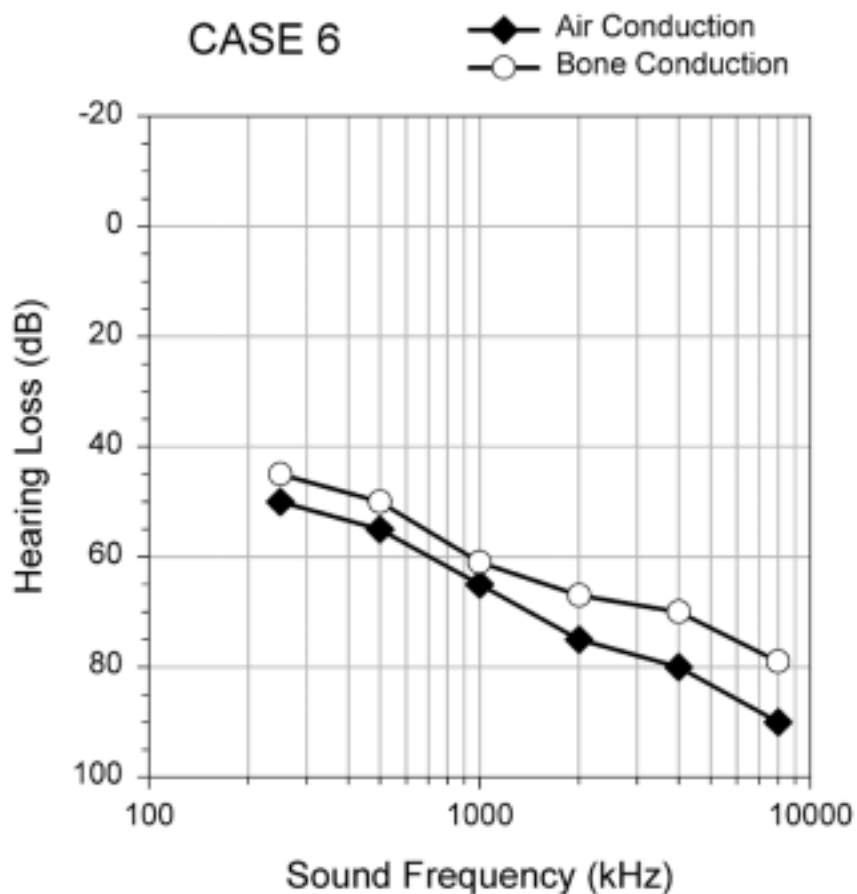
### CASE 6:

During a baseball game, a 16-year old boy was accidentally struck in the back of the head with a baseball bat. After regaining consciousness, the boy exhibited facial weakness on the right side. He was dizzy and remained so for days ahead. He complained of a severe hearing loss in his right ear, which did not improve.

You examined the ear canals and tympanic membranes otoscopically. You also ordered an audiometric examination.

Describe the otoscopic findings.

Describe the audiometric findings.



What type of hearing loss is this person now experiencing?

What do you base your conclusion on?

What are the physiological mechanisms that underlie this hearing loss?

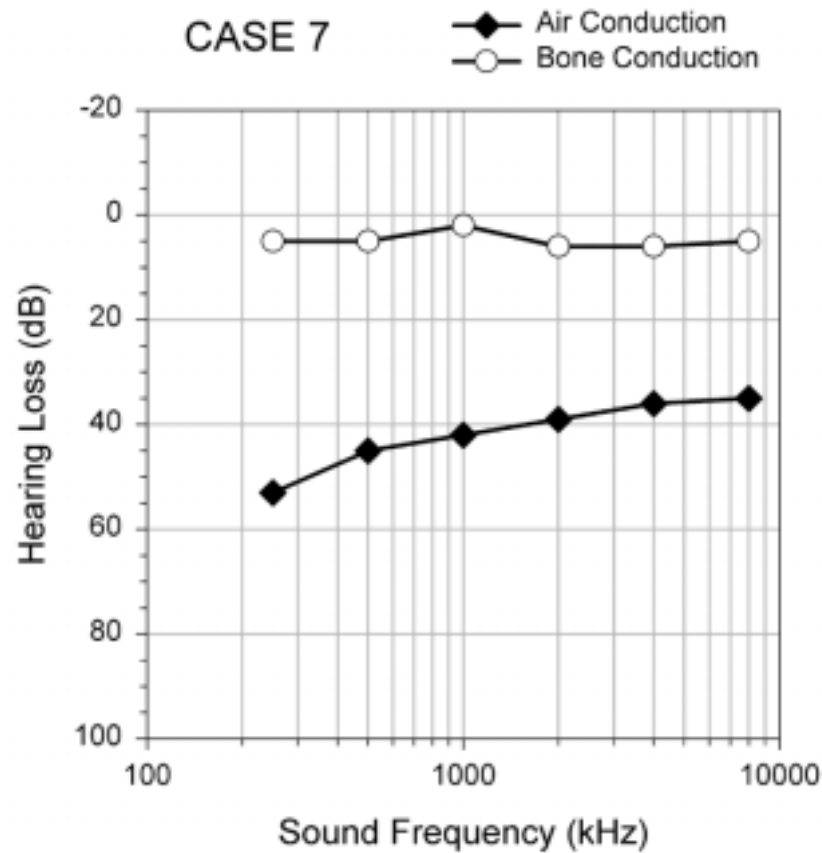
### CASE 7:

A 40 year old woman comes to you because she believes she is losing her hearing. This has been gradually building up over several years and now is to the point where she is having difficulty in hearing normal conversation. Even her friends and family members comment on it.

You examine the ears otoscopically. You then refer her to an audiologist for a more complete evaluation of her hearing.

Describe the physical findings.

Describe the audiometric findings.



What type of hearing loss is this person now experiencing?

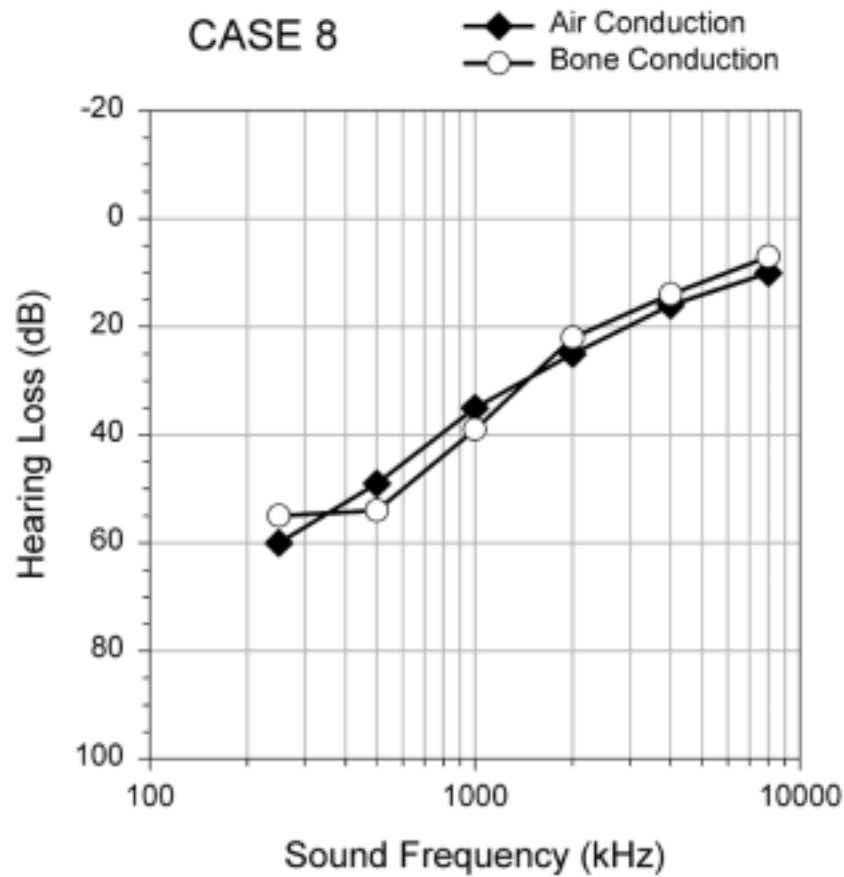
What do you base your conclusion on?

What are the physiological mechanisms that underlie this hearing loss?

### CASE 8:

A 43-year old accountant suddenly began experiencing severe episodes of dizziness accompanied by nausea and vomiting. He also experienced a hearing loss in his right ear, with a feeling of fullness in that ear. This was accompanied by a loud roaring sound. Each attack lasted for days and returned 4-6 months later. The attacks were so severe that he often was confined to his bed. Neurological examination revealed spontaneous and positional nystagmus during these episodes.

Otoscopic examination of the ears was carried out. A thorough audiologic examination was also conducted.



Describe the physical findings.

What type of hearing loss is this person now experiencing?

What do you base your conclusion on?

What are the pathophysiological mechanisms that underlie this hearing loss?

What are the pathophysiological mechanisms involved in the non-hearing symptoms?