

# 5

## Hearing Disorders and Audiogram Interpretation

*There are many great unsolved mysteries of the world. Maybe we can solve some of them in this chapter!*

*As someone new to conducting hearing tests, you might find the process of uncovering various types of hearing disorders somewhat mysterious. You might even think that interpreting your first audiograms is like trying to uncover one of the great unsolved mysteries of the world. Although some of the hearing disorders we review might seem a little mysterious to you right now we seriously doubt that you'll get lost in the Bermuda Triangle or get carried off by Bigfoot.*

We've already solved a couple of the world's mysteries—after the previous chapter you have a good working knowledge of the normal auditory system and how to measure its function, let's turn our attention to various disorders than can affect it. The primary purpose of this chapter is to provide you with a basic understanding of hearing disorders, characteristics of the hearing loss, and, most importantly, the audiometric configuration that is most

closely associated to the disorder. The goal is not for you to memorize every possible disorder, along with the audiometric pattern, but when you are finished reading this chapter you should have a better understanding of how the results of the hearing test relate to the diagnosis of some common hearing disorders.

The first thing you need to know about hearing disorders is that it is critical to have a good understanding of when to refer a patient to a physician for a medical evaluation. In fact, before you even begin to discuss hearing aids with a patient it is imperative that you have ruled out a treatable medical problem involving the auditory system. This means that you have to recognize what a hearing disorder looks like on an audiogram. Before reviewing the various types of hearing disorders, let's discuss the difference between a symptom and an etiology.

## Symptoms Versus Etiology

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Understanding the difference between a symptom and an etiology is important. When someone walks through your door for a hearing test, they may be experiencing several symptoms related to any number of possible hearing disorders.

- A symptom is a description by the patient of what they are feeling, or an observation you make (e.g., dizziness, pain, etc.).
- An etiology is the underlying cause for the disorder. It is only through an accurate hearing test, which may lead to a diagnosis by a physician, that you may know the cause or etiology.

In some cases the etiology is never known. It is common to conduct medical tests to “rule out” pathologies that require further medical attention. A person with an unexplained unilateral loss, for example, may have an MRI to rule out a space occupying lesion. Once the MRI shows there is no obvious pathology, the patient is cleared for the fitting of a hearing aid, although the true cause for the hearing loss still remains unknown.

With accurate audiometry you often will be able to quantify the extent of the ear problem. In most cases, regardless of the etiology, once a treatable medical problem involving the ear has been ruled out, you will fit the patient with hearing aids.

Typically, the hearing disorders you will encounter will involve the inner ear. Not only are they much more common, but disorders of the cochlea usu-

ally require the use of hearing aids as part of the treatment process, and for the most part, hearing aids are the *only* treatment.

## Case History

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Before completing any diagnostic audiometry, it is important to carefully complete a case history. The case history should always be completed face-to-face with the patient, rather than having the patient complete a case history checklist or questionnaire in the waiting room. During the taking of the case history, your job is to find out if the patient has recently experienced any of the common symptoms listed below. Given that these symptoms occasionally are an indication of a more threatening medical problem, they are important to know and understand.

## Common Symptoms

The symptoms listed below are ones you will frequently encounter, and are used by physicians and audiologists on a regular basis.

### *Tinnitus*

This is the perceived sensation of ear noise, often described as a ringing or buzzing in the ear. It is not a disorder, just the sensation to hear sounds generated by the auditory system. Tinnitus, however, is often associated with hearing loss and hearing disorders. For example, most people with noise-

induced hearing loss have tinnitus. In this case, there is no medical treatment. On the other hand, someone with an acoustic nerve neuroma also may have tinnitus and, in this case, a medical workup is critical. Tinnitus can be an occasional occurrence, or it can be constant. Tinnitus is actually more common than hearing loss, as it believed that over 50 million Americans experience tinnitus to some degree. In case you're wondering, tinnitus can be pronounced either as ti-NIGHT-us or TIN-i-tus; the latter is preferred by most professionals.

### ***Vertigo and Dizziness***

True vertigo is a severe spinning sensation usually of short duration. It can be spontaneous, or associated with head movement. The patient can have the sensation the patient of spinning themselves or that the room is spinning around them. There are almost as many causes of dizziness as there are ways in which patients describe it. Recall from Chapter 3, that the balance and auditory system are located in the inner ear. Therefore, it is fairly common to encounter patients with hearing loss (especially if it is of relatively of sudden onset) who are also experiencing vertigo.

### ***Otalgia***

Simply put, this is ear pain, sometimes called an "earache." Otalgia is not always associated with hearing disorders, as it can be caused by conditions such as impacted teeth, sinus disease, and inflamed tonsils. If directly related to the ear, it may be due to middle or outer ear pathology. It's common for

there to be a generalization of pain. That is, the external ear could be painful resulting from an ear canal problem.

### **TAKE FIVE: Medical Terminology**

This chapter introduces you to many of the common terms used to describe hearing disorders and their symptoms. If you receive referrals from physicians and other medical professionals you are likely to encounter terms you don't know. One way to find out about them quickly is to use an on-line medical dictionary. One example is <http://www.medterms.com>.

### ***Aural Fullness***

The perceived sensation of a plugged ear that often accompanies vertigo and sudden hearing loss. Aural fullness can also be a symptom of a problem involving the middle ear, often related to poor eustachian tube function.

### ***Hyperacusis***

An abnormal sensitivity to sound. Hyperacusis is an internal oveamplification of environmental sounds by the auditory system. Environmental sounds of ordinary intensity that do not bother most people, really bother those suffering from hyperacusis—e.g., a sound of 65 dB SPL might be perceived like a 100 dB SPL input. This is different from people who simply are "bothered" by loud noise.

### TIPS and TRICKS: More on Tinnitus

Tinnitus is a condition that still is not completely understood. In fact, experts are still not in complete agreement the underlying causes of tinnitus and hyperacusis. To complicate matters, there is a wide range of treatment options for each that go well beyond the scope of this book. To learn more about the etiology of tinnitus, along with some of the treatment options supported by research

evidence, here are a couple of useful Web sites.

<http://www.tinnitus.org>—A British Web site devoted to a specific type of tinnitus therapy called Tinnitus Retraining Therapy (TRT)

<http://www.ata.org>—Sponsored by the American Tinnitus Association, it contains plenty of information for professionals and consumers.

### TIPS and TRICKS: Cochlear Hearing Loss, Loudness, and Recruitment

As mentioned a number of times already, the majority of your patients will be individuals with cochlear hearing loss. This population is unique regarding their loudness growth pattern; because of their hearing loss they need a loudness boost for soft sounds, but because of the way the cochlea works (see Chapter 3) they do not need a loudness boost for loud sounds. Their loudness perceptions for loud sounds are very similar to someone with normal hearing. In other words, their floor has been raised, but the ceiling has remained in the same. As a result, there is a rapid growth of loudness between the point of audibility and the point of discomfort. This abnormal growth of loudness often has been referred to as *recruitment*. Recruitment is perhaps the most common, and most commonly talked about, yet most misunderstood, symptom of cochlear

hearing loss. It is a *normal* nonpathological phenomenon associated with a damaged cochlea.

Sample Case: Your Monday morning patient has a 60 dB hearing loss, but when you did LDL testing, you found that he found things uncomfortably loud at 100 dB, the same point as many people with normal hearing, a dynamic range of only 40 dB. Does he have recruitment? Yes. Does he have “a lot of recruitment”? Well, if there were such a thing, maybe yes. Is this something to be concerned about? No. It is the *expected* finding. What *is* something to be concerned about is the patient with a 60 dB hearing loss who *doesn't* have recruitment. That would mean that the hearing loss is probably caused by a middle ear, eighth nerve, or brainstem pathology, and the patient should be referred for medical evaluation.

## Common Hearing Disorders Collapsing Ear Canal

*A close encounter is an event in which a person witnesses an unidentified flying object or makes contact with an alien.*

*According to ufologist (yes, that's a real word) J. Allen Hynek, there are four types of close encounters. Close encounters of the first and second kind are sightings of unidentified flying objects, whereas close encounters of the third and fourth type involve contact and even abduction by an alien. In a clinic the only type of encounter you are likely to find is one in which you could uncover a hearing disorder.*

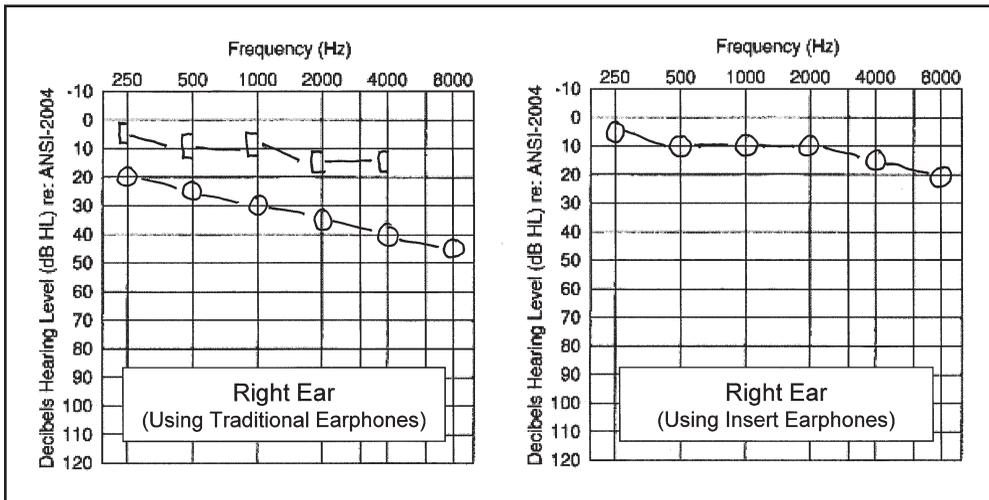
The following is a summary of some of the most common hearing disorders you will “encounter” in your daily practice—and some probably will be a mystery. The list is categorized by parts of the ear, and is not an exhaustive list. It is simply a summary of some of the most common conditions, their causes, and audiometric patterns. To make things fairly straightforward, we have organized the disorders as they relate to parts of the ear. Thanks to the World Wide Web, you can find many more examples of hearing disorders that we did not cover here. Among the Web sites devoted to hearing disorders are <http://www.merck.com.mmpe> and <http://emedicine.medscape.com/otologyngology>

### Hearing Disorders of the Outer Ear

Most disorders of the outer ear are easy to observe, respond to treatment, and usually do not cause significant hearing loss. We review five of the most common in this section.

Recall that we discussed the advantages of using insert earphones in Chapter 4; let's talk about an important reason for using them in a little more detail. Some people, especially the elderly, have ear canals that are collapsing. This means that the tissues lining the ear canal have become very soft. This is a normal condition and does not cause hearing loss in the vast majority of cases, because sound only needs a small opening to pass through. But for patients with this problem, this could change when you do a hearing test. When you place supra-aural headphones on someone with collapsing ear canals, it's possible that the pressure will totally collapse the ear canal, and you are actually causing a hearing loss. It is as though the patient is wearing an earplug. This condition results in an audiogram that has the appearance of a conductive hearing loss (usually greatest loss in the higher frequencies, as they are the easiest to attenuate). This easily can be prevented, however, by using insert earphones. Figure 5–1 gives an example of an audiogram of a patient with collapsing ear canals. The audiogram on the right is after the use of insert phones. Note how the loss returns to near normal levels (e.g., “correct” values) when the appropriate earphones are used.

Failure to recognize collapsing canals, and the resulting erroneous assumption that there is a conductive hearing loss present, is a good way to lose credibility the physicians that you refer to, as their physical examination clearly will be normal. Of course, if your scope of practice includes the use of immittance audiometry, these results will quickly alert you that the measured air-bone gap is erroneous.



**Figure 5-1.** The effects of a collapsing ear canal. The audiogram on the left shows a mild conductive loss when traditional earphones are used. The audiogram on the right shows how air conduction thresholds return to normal levels for the same ear when an insert earphone is used. High frequency conductive losses are rare, so always consider collapsed canals when this pattern is present; the routine use of insert earphones of course will mostly eliminate the problem from the onset.

## Impacted Cerumen

Cerumen (or ear wax) is a normal byproduct of a healthy ear. It lubricates the ear canal and protects the canal and tympanic membrane. As cerumen is produced by the subcutaneous glands of the ear canal, it migrates out of the ear canal by way of the tiny hairs lining the outer layer of the external ear canal.

Some people produce more cerumen than others, especially the elderly. Additionally, other people may disturb the natural cerumen excretion process by inserting Q-tips and other foreign objects into their ear canal, attempting to remove the cerumen. These objects often irritate the canal, which then results in increased cerumen production, which then results in more probing by the individual, not a good thing. Addi-

tionally, using foreign objects to attempt to remove cerumen can result in an impaction, a total blockage of an area of the ear canal.

For individuals who produce excessive cerumen, impaction sometimes also occurs because of hearing aid use. That is, the hearing aid (in the case of a custom instrument) or the earmold at the time of each insertion continues to push the cerumen to a given point (usually about 10 to 15 mm from the ear canal opening) and, eventually, a total (or near total) blockage will occur.

Impacted cerumen results in a temporary conductive hearing loss of varying degree (in severe cases, an air-bone gap as large as 30 to 40 dB will be present). Once the cerumen is removed by a qualified professional, hearing returns to pre-impact levels. A good otoscopic examination will reveal if impacted cerumen

exists. If you observe this, you may want to have the cerumen removed before conducting the hearing test, as there is little reason to conduct a test when you know a priori that the results do not represent the patient's "true" hearing.

### **TIPS and TRICKS: Cerumen Management**

Some audiologists and a few hearing instrument specialists have specialized training in the removal of cerumen from the ear canal. Known as cerumen management, your ability to conduct this service should only happen after you have checked with your state licensing board to see if you can offer it, and had additional training on it.

To find locations where cerumen management courses are offered check with the International Hearing Society or your state licensing board.

## **External Otitis**

Otitis externa is an inflammation of the outer ear and ear canal. Along with otitis media, which we address shortly, external otitis is one of two conditions commonly referred to as an "ear-ache." One common name for this condition is "swimmer's ear" because it frequently develops in people who have been swimming and have had water trapped in their ears.

External otitis is an extremely painful condition requiring treatment from a physician. Hearing tests often cannot be conducted on patients with external

otitis because the ear is too painful to allow for the placement of earphones.

Acute external otitis often occurs suddenly, rapidly worsens, and becomes extremely painful. Because the tissues lining the external ear canal are extremely thin they are easily torn or abraded by minimal force. Inflammation of the ear canal can begin when someone tries to self-clean their ear canal with a cotton swab or other small implement. Another cause of external otitis is prolonged exposure to water or extreme humidity. Regardless of the cause, external otitis occurs when active bacteria or fungus begin to infect the skin of the ear canal.

### **TIPS and TRICKS: External Otitis and Hearing Loss**

In general, we would not expect external otitis to cause a hearing loss. If the swelling was such that there was complete closure of the ear canal, then a mild conductive loss would be expected (probably greatest in the higher frequencies). In general, however, expect normal hearing with this pathology.

Some hearing care professionals have been specially trained to remove cerumen from the ear canal. Due to the thinness of the tissues of ear canals it is easy to abrade them, thus causing inflammation and possibly external otitis in some patients.

Pain that worsens on touching of the outer ear is the predominant complaint associated with external otitis. Patients may also experience discharge from the ear canal and itchiness. Swelling of

the ear canal is another symptom and when the swelling is severe enough a conductive hearing loss may occur. In advanced cases of external otitis, pain may radiate to the jaw and neck.

Because the ear is a self-cleaning system, milder cases of this condition can be addressed by simply refraining from swimming or not using implements to try and clear wax from the ear canal. Topical solutions or suspensions in the form of ear drops typically are used to treat mild and moderate cases of otitis externa. In more advanced cases a physician may have to use a binocular microscope to clean the ear canal and insert what is called an ear wick to deliver medication to the infected area. Because external otitis is so common and can be caused by the actions of even the most experienced hearing care professional during cerumen removal procedures, it's important to know the common symptoms and to immediately refer your patient to a physician for an evaluation if you suspect it.

### **Tumors of the External Ear Canal**

Both malignant and benign tumors have been found in the external ear canal. Bony tumors, called osteomas, are sometimes seen in the ears of people who have done a lot of swimming in cold water. You may not observe a tumor per se, but rather just a narrowing of the canal. Unless the bony growth or tumor closes off the entire external ear canal, they do not cause hearing loss. A detailed otoscopic exam should reveal this, and, unless this is a long-standing condition reported by the patient, a physician referral is appropriate.

### **Perforated Tympanic Membrane**

There are several ways the tympanic membrane (TM) can become perforated. A perforated eardrum is a rupture or perforation (hole) of the eardrum that can occur as a result of infection, trauma (e.g., by trying to clean the ear with sharp instruments, or even a Q-Tip), explosion, barotrauma, or surgery (accidental creation of a rupture).

Because traumatic perforations often alter otherwise normal tissue, they often heal spontaneously. One common cause of TM perforations is related to the buildup of excessive pressure in the middle ear as a result of a middle ear disorder (e.g., eustachian tube dysfunction, infection, effusion, etc.). In these cases, the excess pressure causes the TM to rupture. Because of the underlying middle ear disorder, TM perforations caused from this excessive pressure need to be managed medically.

Surgical repair of a perforated TM is called myringoplasty or tympanoplasty. In some cases, the "surgical patching" procedures are not successful, and the patient more or less will have a "permanent" perforation. Those with more severe and long-standing ruptures may need to wear an earplug to avoid water (or other liquids) making contact with the eardrum, and entering the middle ear cavity.

Perforation of the eardrum usually leads to conductive hearing loss. The amount of hearing loss caused by a perforated TM varies by both the size of the perforation and the location of the opening. Some perforations can be so small that they cannot be detected during routine otoscopy. With large perforations, it's common to see a conductive hearing loss of 30 to 40 dB. Once

the perforation heals, hearing is usually recovered fully (maybe with a slight 5- to 10-dB drop due to scarring), but chronic infection over a long period may lead to permanent hearing loss, as the structure of the TM is altered.

## Disorders of the Middle Ear

*The Bermuda Triangle is a region in the western part of the North Atlantic Ocean where a number of aircraft and surface vessels allegedly disappeared mysteriously. Popular culture has attributed these disappearances to the paranormal or activity by extraterrestrial beings. Documented evidence indicates that a significant percentage of the incidents were inaccurately reported or embellished by later authors, and numerous official agencies have stated that the number and nature of disappearances in the region is similar to that in any other area of ocean. You can think of middle ear disorders like reports of lost vessels in the Bermuda Triangle. On the surface the disorder might be unexplainable, but on further testing using tympanometry and acoustic reflexes, the disorder is no longer mysterious.*

Recall that the purpose of the middle ear is to transmit the airborne sound from the eardrum to the cochlea. This is accomplished quite effectively through the aerial ratio of the TM compared to the oval window, the through the lever action of the ossicular chain. As you would expect, anything that disrupts this flow will cause a middle ear (conductive) hearing loss. We'll describe some of the most common.

### Otosclerosis

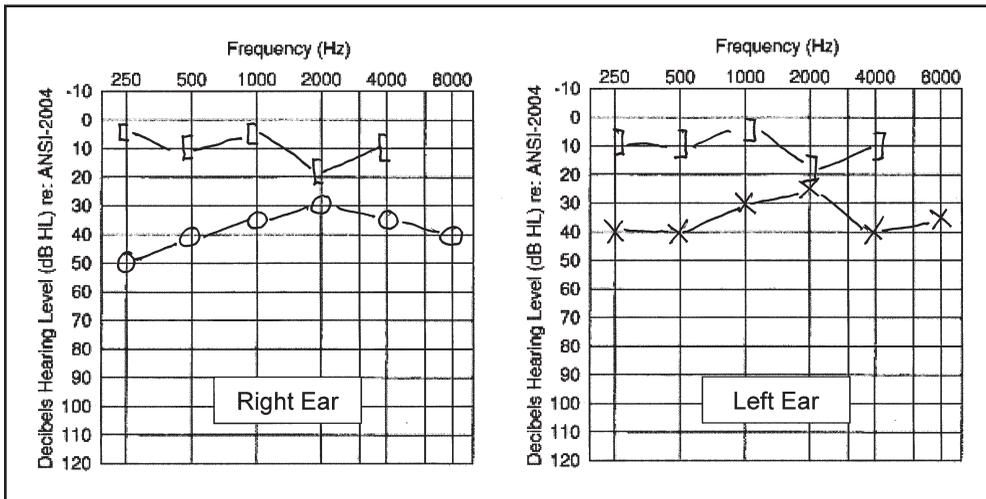
Otosclerosis is caused by two main sites of involvement of the sclerotic (or

scarlike) lesions. The best understood mechanism is fixation of the stapes footplate to the oval window of the cochlea. This greatly impairs movement of the stapes and therefore transmission of sound into the inner ear ("ossicular coupling").

Additionally, the cochlea's round window can also become sclerotic, and in a similar way impair movement of sound pressure waves through the inner ear ("acoustic coupling"). There is some documentation of sclerotic lesions that also are within the cochlea, sometimes referred to as "cochlear otosclerosis."

Treatment of otosclerosis often involves a surgical procedure called a stapedectomy. A stapedectomy consists of removing a portion of the sclerotic stapes footplate and replacing it with an implant that is secured to the incus. This procedure restores continuity of ossicular movement and allows transmission of sound waves from the eardrum to the inner ear. A modern variant of this surgery called a stapedotomy, is performed by drilling a small hole in the stapes footplate with a microdrill or a laser, and the insertion of a pistonlike prosthesis.

Otosclerosis can be hereditary, and at least in the early stages, results in a conductive hearing loss of mild to moderate-severe degree, usually with the greatest loss in the lower frequencies. In the later stages, a mixed hearing loss may be present. Figure 5-2 gives an example of otosclerosis you might see in your office or on an audiogram. While this patient certainly is a hearing aid candidate, and probably would be a successful user of hearing aids, most opt for surgical treatment. Typically, following surgery there is a significant improvement in air conduction thresholds.



**Figure 5-2.** A bilateral conductive hearing loss consistent with bilateral otosclerosis. Notice the 2000 Hz or “Carhart” notch in the bone conduction scores in both ears.

### TIPS and TRICKS: Carhart’s Notch

An audiometric characteristic of otosclerosis is something called “Carhart’s notch.” This is an apparent bone conduction loss at 2000 Hz, named after the person who first described it, audiologist Raymond Carhart, Ph.D. This finding is not a true sensorineural loss, and usually disappears following surgery.

### Negative Middle Ear Pressure and Middle Ear Effusion

As mentioned in Chapter 3, the eustachian tube equalizes the pressure between the air filled middle ear and outside air pressure. This tube is normally closed, but when healthy, opens frequently when we talk, chew, yawn, and so forth. When the eustachian tube becomes blocked or swollen from an

allergy or common cold, the air pressure outside the middle ear is greater than the air pressure within the middle ear space. Children are more prone to negative middle ear pressure and effusion, because the eustachian tube has not had the opportunity to grow to the proper angle (~45 degrees) and is much more horizontal.

Eustachian tube dysfunction causes the air trapped inside the middle ear to become absorbed by the tissues lining the middle ear space, resulting in a drop in pressure within the middle ear space. The greater pressure from the outside air causes the tympanic membrane to become retracted or pushed into the middle ear space. This condition can be observed with otoscopy, although sometimes it is quite subtle.

A specific audiologic test battery called immittance audiometry is used to measure the function of the entire middle ear system. Tympanometry, which is part of this battery, easily will reveal a retracted TM, or a middle ear system that is not moving effectively.

**TAKE FIVE: Valsalva and Toynbee**

Patients with eustachian tube dysfunction may be asked by their physician to autoinflate their eustachian tube by attempting to force air into the middle ear space while holding their nostrils shut (They actually are instructed to try to blow air out of their nose, while holding their nose shut). This is called the

Valsalva maneuver, which we discussed in Chapter 3. Divers use the Valsalva procedure to equalize pressure as they descend or surface. In a related procedure, called the Toynbee maneuver, the patient attempts to open the eustachian tube by holding his or her nose and swallowing (e.g., taking sips of water).

If negative middle ear pressure continues to develop, and is present for an extended time, the fluids normally secreted by the mucous membranes are collected in the middle ear cavity, resulting in a condition called serous effusion or middle ear effusion. When fluid partially fills the middle ear space a mild to moderate conductive hearing loss can occur. Often, when a young child has fluid in their middle ears, it is referred to by the lay person (e.g., parents) as an “ear infection.” Middle ear effusion, however, is not necessarily infectious.

The audiogram for this patient is directly related to the amount of retraction and/or the amount of fluid in the middle ear. If the patient only has a retracted TM, there probably will be little effect on hearing thresholds. If fluid begins to collect, expect thresholds, especially in the low frequencies, to drop accordingly.

**Otitis Media**

If middle ear effusion is allowed to continue unabated, otitis media can develop. Otitis media is any infection of the mucous-membrane lining of the middle ear space. Although otitis

media is thought of as a disease of childhood, it can occur at any age, and can be quite painful. When these tissues become infected they become swollen, interfering with its pressure equalization function. During this process, the tympanic membrane becomes very vascular, resulting in the TM’s red appearance.

There are two types of otitis media, called chronic and acute. As you might imagine, acute otitis media has a very rapid onset time, whereas chronic conditions of otitis media are long-standing. In some cases the fluid in the middle ear becomes thick and sticky and, hence, the nonmedical term “glue ear” sometimes has been used to describe the condition. Like many pathologies of the middle ear, the audiogram will vary with the severity of the problem. It’s reasonable to expect a conductive hearing loss of 20 to 30 dB or worse. The configuration might be similar to that shown in Figure 5–2. In severe cases, air-borne gaps of 30 dB or greater are common.

Antibiotics are used in the treatment of otitis media. If otitis media persists, however, pressure equalization (PE) tubes are inserted into the TM by an otolaryngologist. This procedure is called myringotomy with PE tubes. These

tubes are also referred to as grommets or tympanostomy tubes. If the tubes are open during audiometric testing (they sometimes become plugged), you would expect to see relatively normal hearing. If you conduct immittance testing, volume measures will quickly indicate if the tube is open or closed.

### **TIPS and TRICKS: Ear Impressions and PE Tubes**

It's common for children with PE tubes to obtain ear impressions so that they can obtain custom-fitting earplugs for swimming, showering, and so forth. Although we of course suggest that you always be *very* careful when taking ear impressions, this becomes even more critical when the patient has PE tubes. If impression material goes around the ear canal block, it easily can attach to the tube, and the tube then could be pulled out of the TM when the ear impression is removed. This is not good! It's not common that adult patients have PE tubes, but you will encounter this occasionally.

## **Cholesteatoma**

In general, cholesteatomas are the result of a long-standing middle ear condition. Cholesteatomas form a sac with concentric rings consisting of a protein called keratin; there is some evidence to classify them as low-grade tumors. In patients with TM perforations, the tissue may enter the middle ear through the perforation, producing a cholesteatoma. Cholesteatomas may also be caused by chronic episodes of otitis

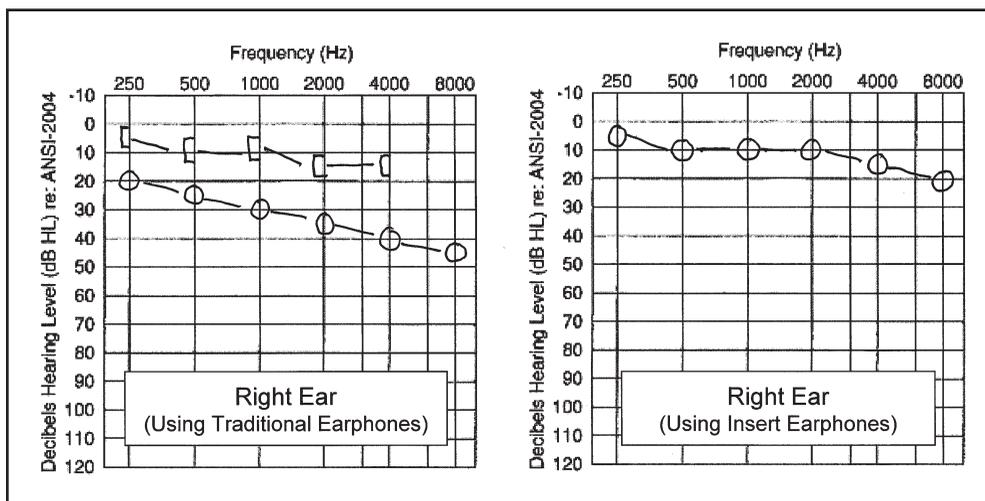
media. Cholesteatomas are dangerous because they eventually can erode the bones of the middle ear. They potentially also could damage the facial nerve, and will even invade the nose and brain cavity in rare instances. In most cases cholesteatoma are removed with surgery. As with other middle ear pathologies, the patient will have a conductive hearing loss, although the patient with a cholesteatoma will typically have a more severe loss than most other middle ear conditions, due to the extent of the disease. It's common to observe air-bone gaps of 30 to 40 dB. A sample case study is shown in Figure 5-3.

## **Tympanosclerosis**

Tympanosclerosis is characterized by white plaques on the surface of the tympanic membrane and deposits on the ossicles. It is often the result of chronic otitis media, which when untreated leaves this white residue. Tympanosclerosis can have a stiffening effect on the TM, which may result in a conductive hearing loss in the low frequencies. As mentioned earlier, PE tubes are a common treatment for otitis media. It's common for these patients (~30 to 40%) to have resulting tympanosclerosis after the tubes have fallen out, or been removed.

## **Ossicular Disarticulation**

This is also referred to as "dislocation" or "discontinuity." As the name indicates, this condition results in one of the two joints between the three ossicles being pulled apart or disarticulated (the incudostapedial juncture is the most common). It can produce a wide



**Figure 5-3.** A bilateral conductive hearing loss associated with a cholesteatoma in the right ear, and otitis media in the left.

variety of conductive hearing losses depending on the location and extent of the disarticulation. The most common causes of ossicular disarticulation are degenerative diseases and trauma to the head. In severe head trauma a TM perforation also might be observed. Interestingly, the largest hearing loss (conductive) is present when the TM is intact, not perforated. In these cases, it is possible for an ossicular disarticulation to cause up to a 50 to 60 dB conductive hearing loss. This sometimes has been referred to as “maximum” conductive loss, as the cochlea is stimulated via bone conduction for higher levels (see Figure 5-3).

### Patulous Eustachian Tube

In some cases the eustachian tube, which is ordinarily closed, is chronically open (patent). These persons often complain that their own voices sound hollow or that they hear their own breathing inside their head. Many of these patients have

an overly patent or patulous eustachian tube. One of the more common reasons for having a patulous eustachian tube is a loss of a significant amount of weight. Although a patulous eustachian tube is not a pathologic condition, it can be quite annoying. Immitance audiometry which we briefly mentioned in Chapter 4 can be used to identify patulous eustachian tubes. There is little or no accompanying hearing loss.

## Disorders of the Cochlea

*The scientific community regards the Loch Ness Monster as a modern-day myth, and explains sightings as a mix of hoaxes and wishful thinking. Despite this, it remains one of the most famous examples of cryptozoology, which is the study of animals long thought extinct. When searching for disorders of the cochlea, in most cases you don't have to search long or hard to encounter a relatively common cochlear problem causing a significant hearing loss.*

A significant number of people around the world have sensorineural hearing loss as a consequence of damage to the cochlea. For adults, sensorineural hearing loss resulting from cochlear pathology is by far the most common type of hearing impairment. In this section we spend some time reviewing the most common types of sensorineural hearing loss resulting from cochlear pathology. Because there is very limited medical or surgical treatment of cochlear hearing loss, these are the people that you will likely see for hearing aid fittings.

## Presbycusis

*Don Juan Ponce de Leon completed Spain's claim on America in 1509, and soon after was made governor of Puerto Rico. Six years later, following Indian rumors, he traveled north to the island of Bimini in search of the Fountain of Youth. Bimini turned out to be the peninsula of Florida. If you've ever been to an early-bird dinner in southern Florida, you know that people know that thousands of people are still arriving in search of that elusive fountain.*

If your patient is beyond the age of 60 years old, it's possible that the hearing sensitivity has progressively worsened over the years, and this will now be reflected in the audiogram, especially in the higher frequencies. This gradual deterioration of hearing is often a result of prebycusis (sometimes written "presbyacusis"). Simply stated, presbycusis is hearing loss caused by the cumulative effects of the aging process. This progression is somewhat more rapid for men than for women, although this partially could be due to the fact that men experience more noise exposure than women, which is difficult to separate from the aging effects on the inner ear structures.

### TIPS and TRICKS: Aging or Noise?

An intriguing question that often comes up regarding presbycusis, is whether this is indeed the result of "aging," per se, or the result of aging in a noise- and stress-filled society. Is presbycusis just a different type of noise-induced hearing loss? An often cited study related to this topic dates back to 1962, conducted with the Mabaan tribe in Sudan. Because of their isolation, there was very little noise in their lives. And guess what—there was little or no hearing loss for even the older members of the tribe (~75 years old). Interpretation of this is a little tricky, as there were also other differences (e.g., general health, diet, etc.), but it certainly is something to think about.

Presbycusis affects all parts of the ear, including neural transmissions to the brain, but the primary site of lesion is the cochlea. The outer hair cells within the cochlea are particularly sensitive to the wear and tear associated with the aging process. As a general rule, the higher the frequency, the greater effect of presbycusis (even people in their 20s and 30s experience loss of sensitivity in the >16,000-Hz range).

The classic presbycusis audiogram will show a gradually sloping downward pattern; nearly always, as the frequency becomes higher the hearing loss becomes worse (Figure 5–4). Because this is a generalized aging process, we would also expect the loss to be quite symmetric. In fact, if the loss is downward sloping, but *not* symmetric, other etiologies should be considered.

### TAKE FIVE: Taking Advantage of Presbycusis

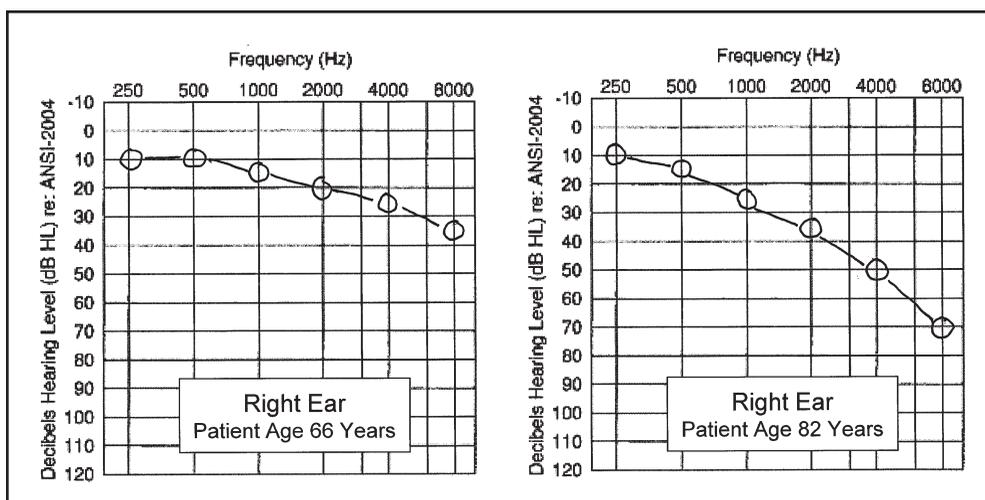
Given the known effects of presbycusis on high-frequency hearing, a cell phone ring has been developed with a center frequency around 16,000 Hz. The notion is that school children can use it to call each other during class, and their teachers won't hear it! Another technology application related to presbycusis has been to use a very loud high-pitched signal in stores where teenagers loiter. The sound is very annoying and drives them out, but the older adult customers can't hear it! Sometimes presbycusis can be a good thing.

### Noise-Induced Hearing Loss (NIHL)

Exposure to loud sounds can result in temporary or permanent hearing loss. This condition is called noise-induced hearing loss (NIHL).

Around 30 million adults in the United States are exposed to hazardous sound levels in the workplace. Among these 30 million people, it's estimated that one in four will acquire a permanent hearing loss as a result of their occupation.

The degree of hearing loss caused by NIHL depends on the intensity of the sound, duration of the exposure, frequency spectrum of the sound, individual susceptibility, along with other variables. Usually, this type of hearing loss is due to continued exposure to



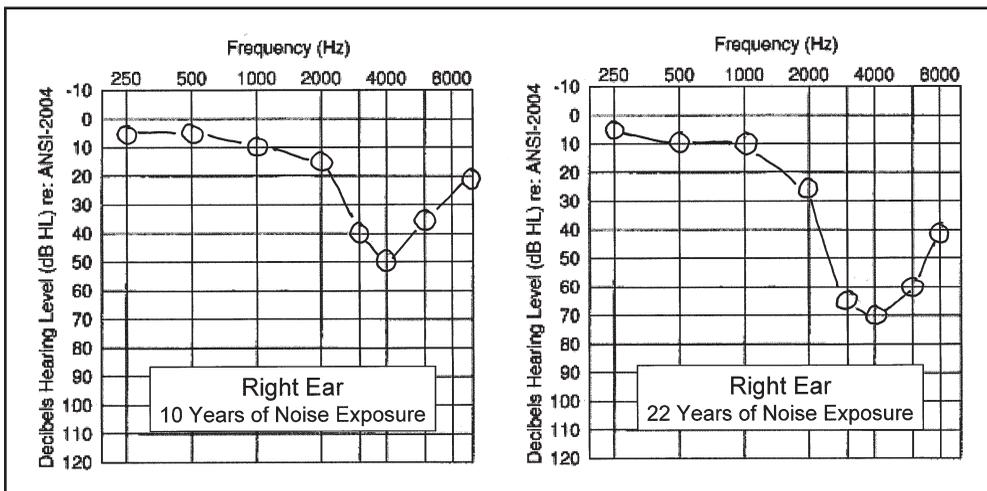
**Figure 5-4.** The progressive nature of presbycusis for an individual's left ear. The audiogram on the left is from a 66-year-old male (showing a slight noise notch with some recovery at 8000 Hz; given the relatively good threshold at 8000 Hz we would predict that presbycusis effects are minimal at this time). The audiogram on the right is for the same male patient at the age of 82. Note that the pattern now no longer looks like a noise-induced hearing loss, as the high-frequency presbycusis effects have bended into the previous 800- to 4000-Hz hearing loss. We only show the left ear thresholds, but typically a symmetrical pattern is observed.

work or recreational noise exposure that has occurred over several years. It is possible, however, for NIHL to occur for only a very short duration of exposure, or even a single blast (referred to as “acoustic trauma”). Because of the shape of the cochlea and the resonant effects of the outer ear, most cases of NIHL show a high-frequency hearing loss, with maximum loss in the 3000 to 6000-Hz range, and usually with some recovery at the highest frequencies. This pattern on the audiogram is called a “noise notch” (see Figure 5–5). NIHL can affect people of all ages.

As NIHL is a fairly common condition it is worth spending a little bit of time discussing the reason for the precipitous slope and noise notch. There are a couple of reasons why the area around 4000 KHz is most susceptible to damage. Although the noise causing

NIHL may be broadband, with roughly equal amplitude at all frequencies, the outer ear and ear canal resonances have amplified the noise in the 2000 to 4000-kHz region by the time the sound reaches the cochlea. This region, therefore, shows the greatest amount of damage from noise exposure. Another reason for NIHL causing more loss in the high frequencies compared to the lows is related to cochlear mechanics and cochlear blood flow; the positioning of the 3000 to 4000-Hz hair cell receptors along the basal turn of the cochlea. It is possible, but quite uncommon, for a noise notch to occur at lower frequencies (e.g., 500 to 1500 Hz; this is most commonly observed when the person was continuously exposed to a unique noise of a narrow bandwidth.

No matter the underlying reason, NIHL is a common etiology of cochlear



**Figure 5–5.** The effects of NIHL over time for one individual’s left ear. Thresholds were measured 12 years apart for a male patient working in a condition of intense noise (daily carpentry with skill saw). The audiogram on the right shows the progressive nature of the hearing loss consistent with the patient’s history of noise exposure. Notice how the dip at 400 Hz deepens, and other frequencies become more involved. The left ear had the same pattern but was not as severe; perhaps there was some attenuation of the noise from head shadow for this ear.

pathology. Given its prevalence, patients that are exposed to both workplace and recreational noise need to be using properly fitted hearing protection. Counseling regarding the need for hearing protection is part of all audiologic exams.

NIHL in its most common form is of gradual onset. The two audiograms below are from the same factory worker taken 8 years apart. Notice that the loss has become worse over the 10 year period. People with significant NIHL routinely are fitted with hearing aids, however, because many with NIHL have normal hearing for low-frequency sounds they sometimes are challenging to fit. Many people with the hearing loss shown in the audiogram shown in Figure 5-5 say they can *hear*, but they just can't *understand* completely. This is due to the normal low-frequency hearing, which provides them "loudness," but the missing high frequencies reduces the audibility of critical speech cues for understanding.

### TAKE FIVE: Personal Stereo Systems

In the past few years there has been a lot of discussion regarding young people obtaining noise-induced hearing loss from listening to iPods and other personal stereo systems. It probably isn't as bad as suggested by some of the articles, but there is a real problem in that many of these devices can be turned up quite loud and many people use them for several hours without giving their ears a "rest." The rest period each hour is critical (and less loud, of course, is good too).

### Permissible Levels

Our review of noise-induced hearing loss would not be complete without a discussion of permissible levels of noise exposure. There is a direct relationship between the intensity of noise, the duration of the exposure, and the degree of potential NIHL. When counseling patients about noise exposure, it's good to have a general idea of what is "safe," and when hearing protection is needed. The Occupational Health and Safety Agency (OSHA) is an arm of the federal government responsible for ensuring that workers are safely protected from dangerous amounts of noise. Table 5-1 indicates when the intensity and duration of exposure becomes dangerous for individuals. If a worker is exposed to levels of sound greater than 90 dB for 8 hours per day, they are required to wear hearing protection. Notice that as the intensity increases the exposure time needed to cause damage is reduced.

**Table 5-1.** Maximum Permissible Noise Levels

90 dB	8.0 hours
92 dB	6.0 hours
95 dB	4.0 hours
97 dB	3.0 hours
100 dB	2.0 hours
102 dB	1.5 hours
105 dB	1.0 hours
110 dB	30 minutes
115 dB	15 minutes

Source: Downloaded from [http://www.quiet-solution.com/Noise\\_Levels.pdf](http://www.quiet-solution.com/Noise_Levels.pdf)

It may be obvious to some, but workplaces are not the only conditions causing NIHL. There are plenty of recreational activities, like hunting, drag racing, and going to the disco that can cause NIHL. Even though OSHA's Permissible Noise Exposure chart wasn't created with them in mind, if you have a sound level meter, you can determine if your nightclub activities are causing some permanent hearing loss.

### Ototoxicity

There are several drugs used for therapeutic treatment of diseases that have the potential side effect of causing damage to the inner ear. Because the cochlea is such a delicate organ it is susceptible to damage from medications and chemical agents. Such drugs and agents are considered to be ototoxic or poisonous to the ears.

Ototoxic drugs have one thing in common: they cause a sensorineural hearing loss. The amount of ototoxic hearing loss depends on the exact dosage and duration of use. When you encounter a patient who has used or been exposed to an ototoxic medication or agent you should consult a physician or pharmacist. A ototoxic hearing loss can present itself in different ways, but, typically, the high frequencies are the first affected, and the hearing loss is usually downward sloping. Some facilities conduct high-frequency audiometry (10,000 to 18,000 Hz) to monitor early changes in hearing.

There are hundreds of ototoxic medications and agents. The most common ones along with their therapeutic uses are listed in Table 5-2. Also listed is whether the drug causes a permanent or reversible hearing loss. The majority of drugs cause a permanent hearing loss, but some cause reversible hearing loss.

**Table 5-2.** A Summary of Common Drug Types and Their Effects on Hearing

Type of Drug	Type of Hearing Loss	Reversible? (Y/N)
<b>1. Aminoglycoside Antibiotics</b> <ul style="list-style-type: none"> <li>• streptomycin</li> <li>• gentamycin</li> <li>• kanamycin</li> <li>• vancomycin</li> </ul>	Sensorineural	No
<b>2. Cancer Chemotherapeutics</b> <ul style="list-style-type: none"> <li>• cisplatin</li> <li>• carboplatin</li> </ul>	Sensorineural	No
<b>3. Loop Diuretics (Furosemide)</b> <ul style="list-style-type: none"> <li>• lasix</li> <li>• bumax</li> </ul>	Sensorineural	Yes
<b>4. Salicylates</b> <ul style="list-style-type: none"> <li>• aspirin</li> </ul>	Sensorineural	Yes
<b>5. Quinine</b>	Sensorineural	Yes

This list is by no means exhaustive; rather, it is designed to represent a sample of the most common ototoxic agents you will encounter. Because new medications are always being introduced onto the market it is best to consult with your local physician or pharmacist for the most current information.

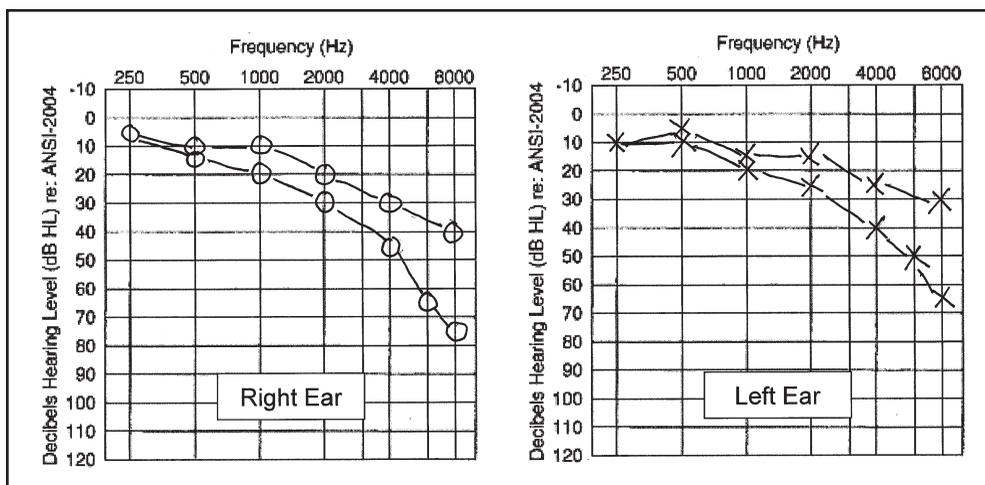
Ototoxic hearing loss is relatively common in patients receiving platinum-based chemotherapy drugs. According to several studies between 23 and 61% developed sensorineural hearing loss as a result of receiving these chemotherapy drugs. In many cases these hearing losses develop 100 to 135 days following the onset of the chemotherapy regimen. Some of the more common platinum-based agents include cisplatin, carboplatin, eloxitin, and vincristine.

Figure 5-6 shows two audiograms from a patient who has been receiving large doses of cisplatin for lung cancer. The first audiogram is 1 month after the first treatment and the second audio-

gram is 60 days later. Note the difference in the thresholds due to the treatment duration. As a dispensing professional you probably will not be directly involved in collecting these types of serial audiograms; however, it's important to note how and when various treatments may affect someone's hearing and associated hearing aid use.

### TAKE FIVE: Important Reference

In addition to causing hearing loss, prescriptive medications can cause tinnitus, hyperacusis, dizziness, and otalgia. Dr. Robert DeSogra, an audiologist in New Jersey, has created a Web site devoted to audiologic reactions to medications. By going to <http://www.earserv.com> and looking up a medication you quickly can find the side effects.



**Figure 5-6.** Audiograms for a patient taking large doses of cisplatin. The upper (better) is 30 days after the first treatment and the lower (worse) audiogram on the right is 60 days after the first treatment. Note the decline in hearing over that period of time, which can be attributed to the drug regimen. The bilateral downward-sloping pattern is common.

## Viral and Bacterial Diseases

There are several viral and bacterial infections that can result in sensorineural hearing loss. Infections, such as cytomegalovirus, can be transmitted to the child from the mother in utero. This is a condition known as prenatal. The following diseases are considered prenatal conditions that can result in a congenital hearing loss:

- Syphilis
- Rubella
- Toxoplasmosis
- Cytomegalovirus (CMV)
- Herpes simplex virus

There also are several viral and bacterial infections that occur after a child has been born that can produce sensorineural hearing loss. In most cases these postnatal infections enter the inner ear through the blood supply, which is carrying the infection. The following are some of the most common diseases acquired after birth (postnatal) causing hearing loss:

- Mumps
- Measles
- Bacterial meningitis
- Herpes zoster oticus

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## Ménière's Disease

*The Lost City of Atlantis was introduced to the West 2,400 years ago by Plato, who claimed it to be the island home of an advanced society. Legend says it was sunk by an earthquake, with later interpretations as an underwater kingdom protected by mermaids. Its whereabouts are still a mystery. Recent underwater evidence suggests it was once part of a larger ????*

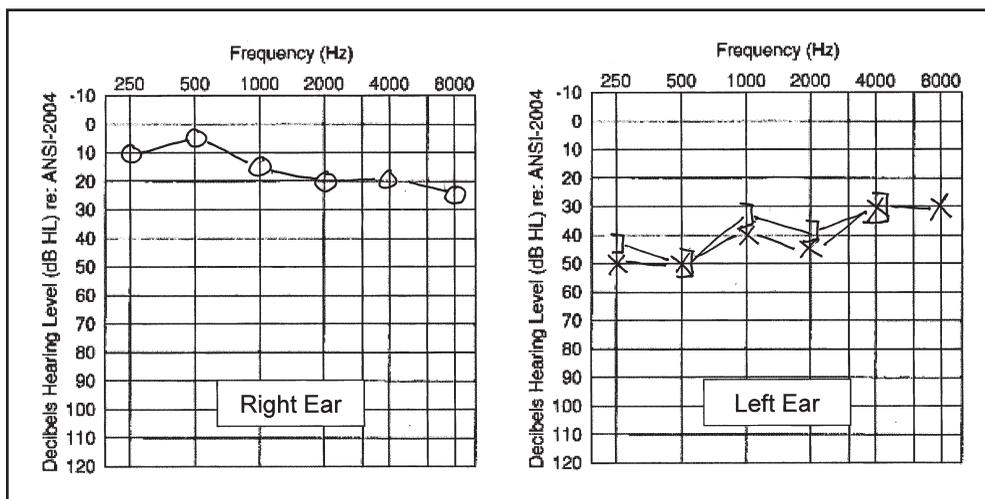
Ménière's disease is named after the French physician Prosper Ménière, who first reported that vertigo was caused by inner ear disorders in an article published in 1861. Ménière's disease, in its "classic form" is used to describe a hearing disorder with one or more of the following characteristics:

1. A hearing loss (usually in one ear) of sudden or rapid onset.
2. A fullness or pressure sensation in the ear.
3. Brief and sudden episodes of severe dizziness (vertigo)
4. A roaring (tinnitus) in the affected ear.

One or all of the symptoms require an immediate referral to a physician. There are many subcategories of Ménière's disease beyond the scope of this chapter. Some types of cochlear hearing losses of sudden onset, such as Ménière's, although they are sensorineural many cases actually return to normal levels.

The exact cause of Ménière's disease is not known, but it is believed to be related to *endolymphatic hydrops* or excess fluid in the inner ear. It is thought that endolymphatic fluid bursts from its normal channels in the ear and flows into other areas causing damage. This is called "hydrops." This may be related to swelling of the endolymphatic sac or other tissues in the vestibular system of the inner ear, which is responsible for the body's sense of balance.

There is no standard "signature" audiogram for Ménière's, but in general there tends to be more low-frequency hearing loss than observed for most other sensorineural pathologies. That is, the audiogram often appears "flat" or upward sloping rather than the more



**Figure 5-7.** Asymmetric left sensorineural hearing loss consistent with Ménière's disease.

common downward sloping pattern. Figure 5-7 shows an audiogram of a client diagnosed with Ménière's disease. Note the asymmetric (unilateral) nature of the hearing loss. After this hearing loss has stabilized, and the physician has given authorization, this person might be fit with a hearing aid in the affected ear.

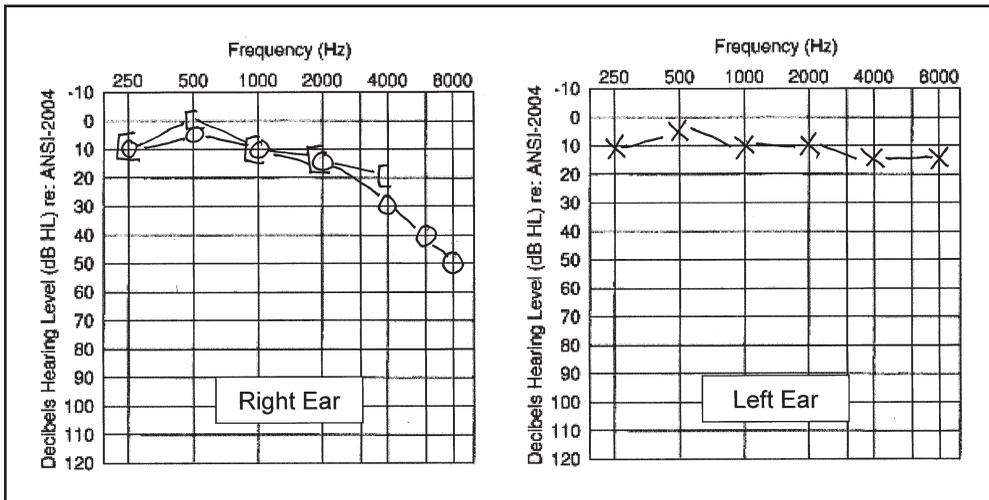
### Retrocochlear Disorders

In general terms, retrocochlear disorders or pathology refers to damage to the nerve fibers along the ascending auditory pathways, running from the internal auditory canal to the auditory cortex. In other words, we might be quite certain that the problem does not lie within the middle ear or the cochlea, and therefore, the locus must be somewhere more medial. Commonly, in audiologic practice, retrocochlear is used to refer to the eighth nerve and the

low brainstem, and auditory dysfunction at higher auditory levels is referred to as "central."

In most cases, eighth nerve retrocochlear pathologies involve tumors. Retrocochlear tumors, referred to as acoustic schwannomas, acoustic neuromas, neurinomas, or neurilemmomas, typically (but not always) produce unilateral high-frequency hearing loss in their more advanced stages. And, unlike presbycusis and many other types of *cochlear* pathology, it is unlikely that there would be uniform symmetric tumors, so there usually is asymmetry between ears in the audiogram (Figure 5-8).

The signs and symptoms of eighth nerve retrocochlear pathology are subtle and difficult to identify with conventional audiometry. In many cases, in the early stages, there is no significant hearing loss (although there may be a reduction of speech understanding for speech in noise, or other difficult speech tests). Many patients will complain of tinnitus on the affected side,



**Figure 5-8.** A mild, right asymmetric sensorineural hearing loss consistent with possible retrocochlear pathology.

vertigo or dizziness, fullness, or speech not sounding clear. In cases where retrocochlear pathology is suspected, a complete audiologic diagnostic battery and otologic referral is needed. Your job is to refer the patient to a physician or audiologist if a “red flag” for a retrocochlear pathology exists.

## Central Auditory Disorders

As mentioned earlier, technically a retrocochlear pathology would include everything medial of the cochlea, but usually we refer to pathology above the low brainstem as “central.” When thinking about auditory disorders, it’s important to remember the “subtlety principle.” That is, as the pathology becomes more central, going from the middle ear to the auditory cortex, the impact of the disorder on traditional

audiologic tests will be more subtle. For example, a cochlear pathology will nearly always cause a reduction in hearing thresholds and speech understanding. A disorder of the brainstem (e.g., multiple sclerosis, tumors, etc.) may cause no hearing loss and no loss of speech understanding (unless a difficult speech-in-noise test is conducted). The bottom line: If the patient’s history suggests a problem with the auditory or balance system, even if all the audiometric results are very normal, medical referral is still warranted.

## Nonorganic Hearing Loss

*Every few years you read reports of Bigfoot carrying someone off. In 2008 there was even a photo taken of a dead Bigfoot. We were skeptical of this finding when we heard the creature was found in the woods next to a busy highway in Georgia! A few weeks later the entire report was found to be a*

### TAKE FIVE: Pure-Tone Hearing Screenings

More than likely you will be working primarily with adults, you need to know a little about hearing screenings. When it comes to actually conducting a hearing screening, there is no acceptable standard for the identification of hearing loss in the adult population. The term “screening” implies that we are conducting some type of a “pass/fail” procedure to see if the patient needs further testing.

Some experts think that a pure-tone hearing screening should be conducted at 1, 2, and 4 kHz using a 40 dB signal. Others suggest that a 25 dB signal be used. Regardless of the cutoff criteria, the screening would be considered a “fail” if the patient does not respond to the signal at the lowest intensity level tested. Keep in

mind that during a screening we conduct testing at a designated intensity level, rather than identifying threshold. When a patient fails a screening, they usually are referred for further testing, which includes a complete auditory evaluation.

There are several tools that can be used to screen older adults. The Welch-Allyn Audioscope is a hand-held device with a built-in audiometer. There are many hand-held audiometers on the market today. There is even an application, called uHEAR, that can be downloaded onto an iPod. If you do a Google search on Unitron uHear you can download it to your favorite MP3 player. Other hearing aid manufacturers have similar screening tests.

*hoax—the dead Bigfoot body was actually made out of wax! Bigfoot might be a hoax, but we are quite sure you will have a close encounter with someone who presents a mysterious hearing loss that turns out, with careful testing, to be a hoax.*

There are cases where a hearing loss may be measured on the audiogram, but there is no organic basis to explain the impairment. Some of the terms used to describe this include nonorganic hearing loss, pseudohypocisus, and functional hearing loss. If indeed the patient knowingly is exaggerating their hearing loss, the term malingering is used.

Aside from the cases of malingering, where with adults, exaggeration of the

hearing loss usually is related to financial compensation, the reasons for non-organic hearing loss are not clearly understood. A number of signs can alert you to the possibility, however. These signs may include inconsistent test results, poor test-retest reliability, inappropriate behavior during the test (e.g., exaggerated attempt at listening or lipreading), or poor agreement between test results and real-world communication (e.g., the patient answers your questions in the waiting room, but then demonstrates a flat 70 dB HL hearing loss). In some cases, there may be an underlying hearing loss, and the patient is simply adding to it.

One reason SRTs should be conducted during routine testing is to cross check

the reliability of pure-tone thresholds. If the SRT and pure-tone average differ by more than 10 dB, the reliability of the test should be questioned. If there is a discrepancy, the SRT will nearly always be *better than* the pure-tone average. We recommend conducting the SRT before the pure-tone thresholds, as this will provide you with a general idea of where the thresholds should be falling for the speech frequencies. If there is poor agreement, there is no need to test all the other frequencies, as you would simply assume that the entire exam is invalid. Many other special tests have been developed to detect nonorganic hearing loss, including the Stenger test which is very effective when the loss is only in one ear.

## **Hereditary Hearing Loss**

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As a professional who primarily fits and dispenses hearing aids, we doubt you will spend too much time thinking about hereditary hearing loss. However, it's good to know a few important things about it. Hearing disorders can be classified into two types of groups: exogenous (outside the genes) and endogenous (within the genes).

- Exogenous hearing disorders are those caused by toxicity, noise, accident, or injury that damages the inner ear. We have already summarized many exogenous factors of hearing loss in this chapter.
- Endogenous hearing disorders originate in the genes of the individual. An endogenous hearing disorder is transmitted from the

parents to the child as an inherited trait. Hearing losses resulting from hereditary factors comprise a significant number of all hearing disorders.

It is estimated that there are over 400 different genetic syndromes in which hearing loss is either a regular or occasional feature. Unless you are regularly testing children, you are not likely to be commonly involved in the identification of hearing disorders related to genetic transmission. During a routine case history with adults, you may encounter various genetically transmitted syndromes that have hearing loss as one of their characteristics. It's also probable that you will uncover a hearing loss that is genetic that the patient was unaware of because many progress at a very slow rate. As a professional (nonaudiologist) who dispenses hearing aids you don't need to have an in-depth understanding of genetic factors as they relate to hearing disorders; however, it is useful to know a few key concepts that you may even remember from high school or college biology class.

## **Mendelian Laws**

Hereditary hearing loss is based on the Mendelian laws of inheritance. According to Mendelian law, genetic traits may be dominant, recessive, or sex-linked. Genes are located on the chromosomes and with the exception of those genes that are located on the sex chromosomes of males, chromosomes come in pairs. One member of each gene pair is inherited from each parent. Humans have 22 pairs of autosomes, or non-sex deter-

mining chromosomes, and one pair of chromosomes that determine sex. The sex chromosomes for females consist of two X-chromosomes, and for males, one X and one Y. During reproduction each egg and each sperm carries half the number of chromosomes from each parent. When the egg is fertilized, the full complement of chromosomes is restored, so that half of a child's genes are from the mother and the other half from the father.

### **Modes of Transmission**

There are three modes of transmission for hereditary hearing loss: autosomal dominant inheritance, autosomal recessive inheritance, and X-linked inheritance. The term autosomal implies that the abnormal gene is not carried on the sex chromosomes. In autosomal dominant inheritance one parent exhibits the inherited trait and this trait has a 50% chance of being transmitted to the child. Examples of autosomal dominant conditions you may encounter include Waardenburg syndrome, branchio-otorenal syndrome, and neurofibromatosis 2 (NF2).

In cases of autosomal recessive inheritance, both parents of a child with hearing loss of the autosomal recessive type are clinically normal. Appearance of the trait in the child requires that an individual possess two similar abnormal genes, one from each parent. Because the laws of probability permit this type of hearing loss to be transmitted without manifestation through several generations, the detection of the origin of autosomal recessive inheritance is very difficult. Usher syndrome and Pendred syndrome are two of the more common types of

autosomal recessive hearing disorders you may encounter in clinical practice.

Another type of genetically transmitted hearing disorders is X-linked or sex-linked inheritance. X-linked inheritance is determined by genes located on the X chromosome. About 2 to 3% of all genetic hearing loss is a result of X-linked inheritance. Alport syndrome is one type of X-linked hearing disorder.

More than 70% of hereditary hearing loss is nonsyndromic, which means the hearing loss is not associated with any other signs or symptoms. The causes of nonsyndromic deafness are complex. Researchers have identified more than 30 genes that, when mutated, may cause nonsyndromic deafness; however, some of these genes have not been fully characterized. Different mutations in the same gene can cause different types of hearing loss, and some genes are associated with both syndromic and nonsyndromic deafness. In many affected families, the gene responsible for hearing loss has not been found.

Regardless of the hereditary pattern of hearing loss, we are not even scratching the surface of this topic. To learn more go to <http://www.ncbi.nlm.nih.gov> and enter the key words genetics and hearing loss.

### **Classification of Hearing Disorders by Time of Onset**

Hearing loss is also classified by the time in which the hearing loss is acquired. One important reason for knowing when a hearing disorder is acquired is related to language development. When an infant has a hearing loss at birth

### TAKE FIVE: Hearing Loss Prevalence

Hearing loss is the most common birth defect and the most prevalent sensorineural disorder in developed countries. One of every 500 newborns has a permanent, sensorineural hearing loss. By the time a child reaches adolescence, 3.5 out of 1,000 have this condition. In the general population, the prevalence of hearing loss increases with age. This change reflects the impact between genetics and environment, as well as interactions between the environmental triggers and a

person's genetic predisposition. According to the National Institutes of Health (NIH) Web site, more than 50% of pre-lingual deafness is genetic, most often autosomal recessive and nonsyndromic. Approximately 50% of autosomal recessive nonsyndromic hearing loss can be attributed to the disorder DFNB1, caused by mutations in the GJB2 gene and the GJB6 gene. The carrier rate in the general population for a recessive deafness-causing mutation is about 1 in 33.

he or she will not develop language normally. On the other hand, a child acquiring hearing loss at say, age 12 would already have normal language development. Hearing loss caused by viral and bacterial infections are most commonly associated with the terms listed below.

**Congenital Hearing Loss:**

A hearing loss acquired at birth. Common causes include bacterial or viral infection, or ingestion of ototoxic medications.

**Prenatal:** A hearing loss that has developed before birth in which the mother has passed the hearing disorder onto the child. In other words, the hearing loss was acquired while the baby developed in utero. The most common prenatal hearing disorder are viral or bacterial infections. Many hereditary hearing disorders are acquired prenatally.

**Perinatal:** The hearing loss develops during or shortly after

birth. Many of the same conditions causing a prenatal loss can occur perinatally.

**Acquired or Postnatal:** A hearing loss that develops later in life. Prelingual hearing loss is a hearing loss acquired during the critical language development years of between birth and about 12 years of age. Postlingual hearing loss is acquired after the most critical language years.

### In Closing

During your first few months on the job, you are likely to have a close encounter with several of the hearing disorders mentioned in this chapter. When you do encounter one that seems a little mysterious, it's wise to refer that person to an audiologist or a physician specializing in disorders of the ear. Unlike hearing aid technology, which is

rapidly evolving, the subject of hearing disorders changes at a much slower pace. For this reason we recommend that you invest some of your hard-earned dollars into one or two hearing disorders textbooks. The text doesn't have to be all that current to be useful. One we like is, *Hearing Disorders* by

Jerry Northern. The third edition, which was published in 1995, is available online for a reasonable price. Another is entitled, *Audiology: The Fundamentals* by Fred Bess and Larry Humes. It has a comprehensive introductory chapter on hearing disorders that expands on this chapter.