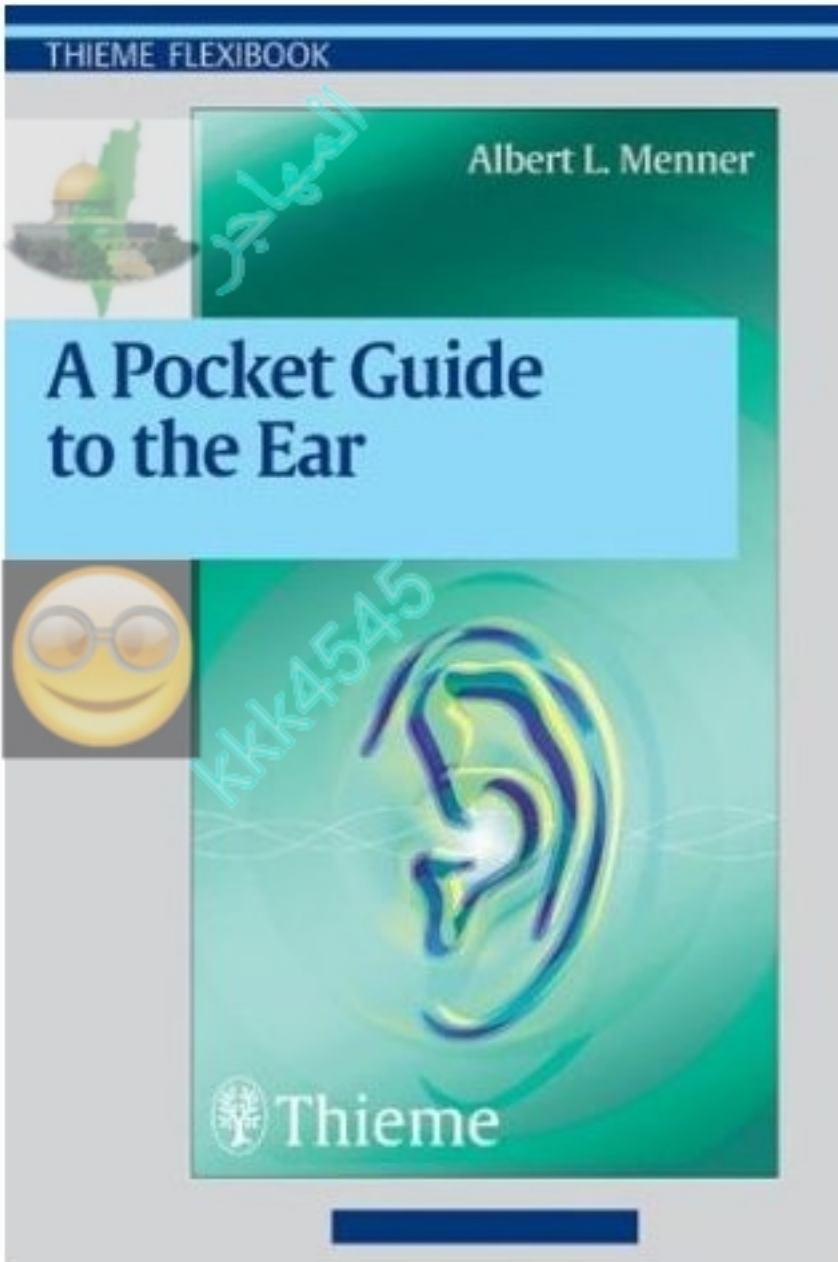

A Pocket Guide to the Ear

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Author's Note

A sincere effort has been made for this textbook to contain up-to-date, accepted guidelines for diagnosing and treating ear problems. However, medical knowledge is ever changing, and treatment choices may also vary from geographic region to region. Astute practitioners should be aware of even the most recent changes in medical information. Furthermore, the medical professional should exercise great care in performing any procedure or prescribing any medication. For example, seemingly straightforward ear cleaning may bring about unexpected complications. Regarding the prescription of drugs and their dosages, one should refer to the manufacturer's leaflet accompanying the product and be aware of the patient's known drug intolerances and allergies.

If there is any doubt about a course of action, one should obtain appropriate consultation with another physician. Even then, a patient may experience an adverse or unexpected outcome. As caregivers, we must all take personal responsibility for our actions. The author and publisher cannot be held responsible for patient responses to procedures and treatments described in this book.

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Foreword

A major trend in American medicine in recent years has been to promote primary care medicine. Well-trained primary care physicians are essential to making high-quality health care available to all. For a surgical subspecialty like otolaryngology–head and neck surgery, a valuable contribution to this effort consists of making the knowledge and experience of this field accessible to primary care practitioners. This is particularly helpful because diseases of the ear make up a large part of primary care practice. Doctor Menner's book is a good example of such a contribution, making the wisdom and insights of a career in this specialty available in a short, concise, easy-to-read guide to diseases of the ear.

Doctor Menner graduated cum laude from Johns Hopkins University, received his M.D. degree from the University of Virginia School of Medicine in 1970, and completed his residency in otolaryngology–head and neck surgery at the University of Maryland in 1975. He has been in private practice in Elmira, New York for more than a quarter century. He has distilled his knowledge into this practical and useful guide to otology.

Whether the book is read cover to cover or used as a reference as needed, it offers sound advice on the common and rare conditions of the ear that will be seen by a primary care physician.

William C. Gray, M.D.
Associate Professor, Division of Otolaryngology
University of Maryland School of Medicine

Acknowledgement

Writing this book has been an entertaining task for the past several years. I am grateful to a number of individuals for their help. My daughter, son, and wife were the initial contributors. Stephanie encouraged me to start this project, Mike instructed me in the necessary computer skills, and Donna has been an ongoing advisor and loving supporter.

Numerous professional friends from my geographic area have helped. Linda and Dr. Frank Gudas smoothed out an early manuscript. They also directed me to Joy Perry, an excellent medical editor who greatly improved the text and helped in so many other ways. Physicians in various specialties read it and gave advice and/or encouragement, most especially family practitioner Dr. Tom Mitchell. Sincere thanks are also due to pathologist Dr. Terry Lenhardt, pediatrician Dr. Jon Homuth, psychologist Dr. Joel Grace, dermatologist Dr. Bill Clack, pulmonologist Dr. Bill McCauley, surgeon Dr. Ben Willwerth, otolaryngologists Dr. Nils Peterson and Dr. John Dooley, and my father-in-law, general practitioner Dr. Donald Coon. Much help came from audiologists Sheila Giovannini and Mary Ann Bullett. Kathi Menner and Ann Rose assisted with graphics, and Scott Moore, with legal advice.

Thanks also to Thieme Medical Publishers for making this such an attractive book. Brian Scanlan, the president of Thieme, Dr. Cliff Bergman, and Dr. Liane Platt-Rohloff were so good as to believe in this type of presentation and to work with me to polish it to its present form, especially with the excellent graphics.

My former fellow ENT residents from the University of Maryland School of Medicine made major contributions. Early on, Dr. Jack Biedlingmaier advised me regarding teaching enhancements, and Dr. Hubert Léveque carefully helped with the factual content. More recently Dr. Bill Gray, a knowledgeable and dedicated educator at the ENT program, collaborated to make this edition as up-to-date and accurate as possible; I also thank him for writing the foreword. Finally, I especially wish to honor the memory of Dr. Cyrus L. Blanchard, the late Professor of Otolaryngology who trained Jack, Hubert, Bill, myself, and countless other ENT residents for the longest span of years of any ENT residency program in the United States. We miss you, dear Dr. B.

Albert L. Menner, M.D.
Elmira, N.Y.

Contents

Introduction

As an ENT specialist who has been in private practice for years, I obviously see many new patients with ear problems. Often, a patient will give a history similar to the following one of a patient whom we will refer to as Pat. Her case is presented here as an example of the types of ear complaints that clinicians often encounter in everyday practice.

"Doctor, I asked for a referral to you because I'm tired of these ear infections. There has been pressure, on and off, in my right ear for months. It feels full and aches a little bit, even down my neck. My own doctor saw fluid in it once, and the antibiotics he prescribed cleared things up for a while. However, the problem keeps coming back. Sometimes both ears bother me, and I also get dizzy from time to time. I personally think it's an inner ear infection. Nasal sprays and decongestant pills don't help. For the last few days, I feel it more than ever in the right ear again, and I'm glad I got in to see you now. I probably need another course of antibiotics, but is there anything different you can do for me?"

At this point in the interview, I inwardly made a tentative guess at the diagnosis. There are certain chronic ailments, some related to tension and stress, that might refer so many symptoms to one or both ears. Nonetheless, a more complete evaluation was justified. Pat needed careful questioning, along with a good ENT and hearing assessment.

The diagnosis was unexpected. It was determined with a brief but thorough examination, one that a family practitioner could carry out in the office. I will return to Pat's ear troubles later in the book, in the Epilogue. This short volume is intended to provide you with all the necessary knowledge and tools to evaluate patients like Pat and to reach the same diagnosis as myself.

Intended for all those who treat ears and hearing, this book can be an aid to nurse practitioners, physicians' assistants, family practitioners, internists, ER physicians, pediatricians, audiologists, and even ENT residents. The medical student or resident rotating through ENT will find it especially useful. Though the text is intentionally short, it deals with almost everything that can go wrong with ears, based on careful study and observation during a busy ENT practice for more than 25 years. In one way, it is a brief clinical "encyclopedia" of ear disease. In another way, it is an entertaining text that can provide the reader with considerable expertise on ears after a few hours' reading. The presentation cuts through to important truths

about ear symptoms, diagnosis, and treatment—subjects that are not always taught in medical training programs.

Some features have been added to aid in the book's use as a pocket reference after it has been read. **Bold print** is used to stress important concepts of **diagnosis** and **treatment**. Highlighting is used in summary paragraphs. Recommendations for primary treatment and specialty follow-up are especially emphasized in most summary paragraphs.

The first three chapters deal with the relevant basics—audiometry, ear examination, and functional anatomy. Occupying 24 pages, they require the close attention of readers not familiar with ears and hearing, but provide the basis for a good understanding of the clinical chapters that follow. The remaining chapters journey through all the disorders of the ear, from outer, through middle, to inner ear. A short chapter on facial nerve problems, which are often related to the ear, is included.

The book closes with a "Top Ten" list of the most common ear–related oversights that I have seen made by clinicians over the years. If curious, you might skip to this part now to assess your own familiarity with ear disorders.

1 Hearing and Basic Audiometric Concepts

The essentials of hearing evaluation and measurement are certainly an appropriate starting point for this chapter. Sound perception is the primary reason for the existence of the complex sensory organs on each side of the head. Awareness of the environment and the ability to communicate are key elements to survival. Of course, balance and orientation are significant parts of ear function as well—these will be discussed in more detail later. The contents of this brief chapter might seem very technical to readers who do not specialize in ears, but familiarity with the basics of hearing is necessary for an understanding of ear complaints. Audiometric testing, represented in graphic form, is the easiest way to "visualize" a patient's hearing, and this will be discussed first. Tuning-fork evaluation will be covered in the next chapter.

Pure Tone Audiometry

Audiometry is the precise method of hearing assessment. It is performed in a soundproof compartment by an audiologist, who uses an audiometer to introduce measured sound intensities of selected tones to the listener, usually through earphones. **Pure tone thresholds** are the minimal intensities of given tones (frequencies) that can be heard by the person being tested. These thresholds are usually recorded on a grid, like the one shown in Figure 1.**1**, to create an **audiogram**. This one has no responses recorded on it and is presented purely to give an idea of the frequencies and loudness of common sounds.

On this graph, the vertical axis plots hearing level (HL) in decibels (dB), tiny units of loudness. These two abbreviations are usually used together (dB HL) to report a patient's test results or to refer to the level of loudness of a given tone. Zero dB HL is near the top of the graph; this level is barely audible to a human with "perfect hearing." The louder the sound needed for a test response, the greater a patient's hearing loss. Thus, the scale is inverted—a lower number means better hearing. Above 0, there is a line for –10 dB HL, indicating "supranormal" hearing, something like 20:15 vision. At the other extreme, some very loud sounds can be 110 dB HL or even greater, such as a nearby jet engine (roughly 125 dB HL). Most audiometers only go to 110 dB HL. For all practical purposes, an individual

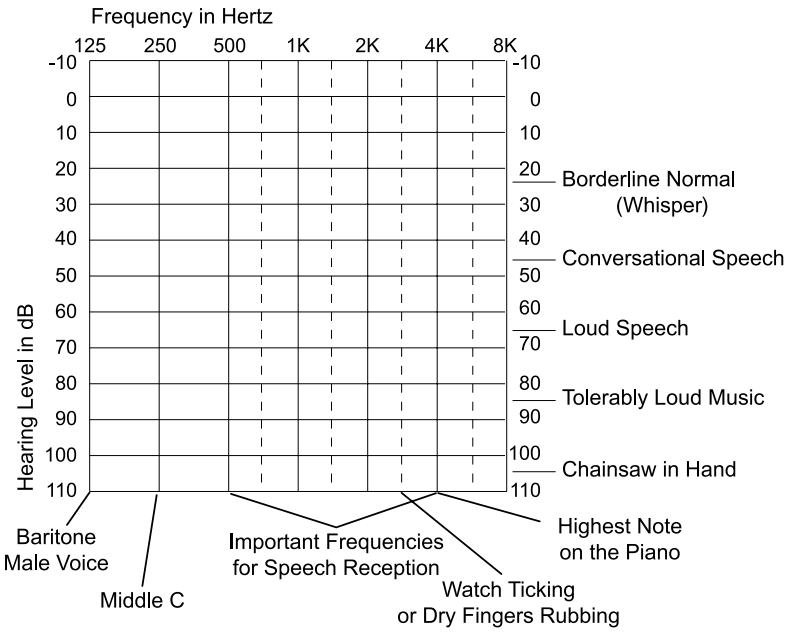

Fig. 1.1 Audiogram grid showing recognizable frequency and loudness levels.

who cannot hear noises or tones at 100 dB is considered profoundly deaf.

Between these extremes, shown on the right side of Figure 1.1, are examples of everyday sounds at their approximate decibel levels. For instance, conversational speech is roughly at 45 dB HL. The *borderline normal* range of hearing is considered to be 25 dB HL or less for adults and 20 dB HL or less for children. Most normal subjects will test close to the 0 line. Hearing loss that falls between 25 and 45 dB HL is considered *mild* loss; between 45 and 70 dB HL, *moderate* loss; between 70 and 90 dB HL, *severe* loss; and greater than 90 dB HL, *profound* loss. Often, to patients, we refer to decibels of hearing loss as "percentage" loss, which is not completely accurate but a convenient approximation.

Frequencies are displayed along the horizontal axis. The lowest frequency measured by the audiometer is usually 125 Hz, or cycles per second. Actually, this is not a very low tone—it corresponds to the note C one octave below middle C on the piano. The fundamental vocal tone of an adult male, when producing vowel sounds, is about here. Each dou-

bling of this number in Hertz corresponds to a frequency one octave higher. Thus 4000 Hz, or 4 KHz, is the highest C on the piano.*

The human ear perceives a vast range of frequencies, from 20 to 20 000 Hz or more, but as the graph shows, the 125–8000-Hz range is what we usually test audiometrically. The frequencies most important for hearing and understanding human speech are from between 500 and 4000 Hz. Vowel sounds are at the low end of this range or even lower (as previously stated, a deep-voiced male actually phonates at 125 Hz). Consonant sounds tend toward the higher frequencies, especially the non-voiced ones like "s," "f," "sh," and "th."

Speech Audiometry

An audiologist is not only able to test hearing levels at selected frequencies (i.e., pure tones), but can also measure the ability to perceive speech. One method is to recite familiar two-syllable combinations, such as "hot dog" or "baseball," into the headphones at calibrated loudness and then measure responses. This gives us the patient's **speech reception threshold (SRT)**, a key measurement. The SRT usually corresponds to the average of one's pure tone thresholds between 500 Hz and 2 KHz, and may be considered to represent the overall hearing ability of an individual.**

A different type of measurement, **speech discrimination**, is the percentage of a long list of one-syllable words that the subject can repeat correctly when these words are presented at a moderately low intensity. Discrimination of 90 % or better is considered acceptable. Hearing loss in the higher frequencies, where consonant sounds are heard, often reduces speech discrimination scores.

Different types and causes of hearing loss can affect different frequencies. Certain recognizable patterns of loss show up repeatedly. Examples include the sloping high-frequency loss of presbyacusis (hearing loss associated with aging), the "notched" high-frequency loss of noise damage, or the low-frequency loss of early Méniere's disease. These problems will be discussed further in later chapters.

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* The frequencies mentioned are rounded off slightly. A middle C tuning fork is exactly 256 Hz, one octave higher 512 Hz, and so on.

** The abbreviation used to follow a patient's speech reception threshold number is *dB HTL (decibels hearing threshold level)*. Briefly, this technicality refers to an adjustment in the actual sound pressure used in the audiometer when presenting words rather than tones.

Conductive vs. Sensorineural Hearing Loss

There are two major categories of hearing loss that are key concepts for the clinician to understand. The first, **conductive hearing loss**, is due to an *outer* or *middle ear* problem—a problem "conducting" sound waves through the ear canal to the eardrum and then through the middle ear apparatus toward the inner ear.

Causes of conductive loss might include obstruction of the ear canal by cerumen (wax), impairment of middle ear function by fluid, or fixation of the middle ear ossicles by disease. With conductive loss, sounds coming from within, such as one's own voice, are perceived as *louder* because of reduced competing ambient noise. Plug your right ear with your finger, creating a conductive loss, and note how your own voice sounds louder on this side. This phenomenon is known as *autophony*. A patient with a conductive loss often feels like he or she is talking "in a barrel," or "under water."

Sensorineural hearing loss is due to a malfunction somewhere in the *inner ear*, from the cochlea inward through the auditory nerve. This is often termed "nerve deafness" and with this type of loss even one's own voice does not sound loud.

The distinction between these two types of loss is obviously important for determining the cause of a patient's hearing complaint. Tuning-fork

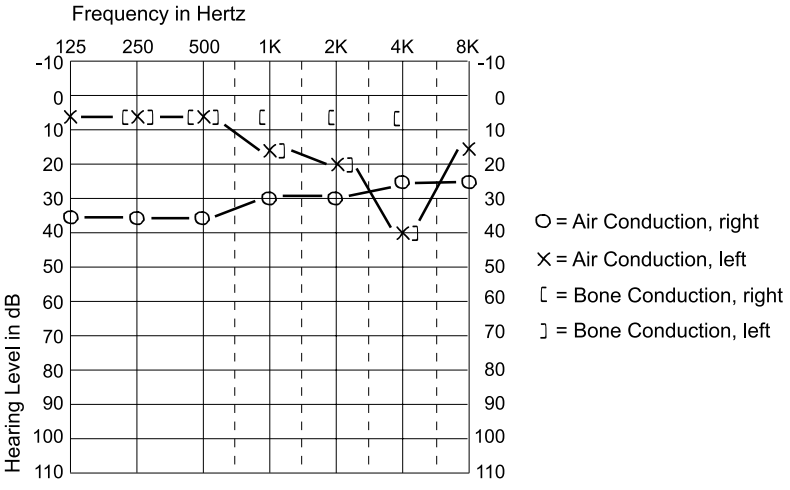


Fig. 1.**2** Audiogram of a patient with a right cerumen impaction and a left noise-induced cochlear loss.

evaluation can differentiate between the two; this will be discussed in the next chapter. Audiometric testing can do the same, more precisely. Figure 1.**2** shows an audiogram of a hypothetical patient with a conductive loss in the right ear and a sensorineural loss in the left ear. This is a representation of a complete, and somewhat complex, pure tone audiogram. If you can understand it, with the help of the following explanation, you have gained a good working comprehension of hearing loss and its assessment.

The audiometer distinguishes between conductive and sensorineural hearing loss in the following way. When earphones are placed on the ears for testing, **air conduction** is measured. This is the way we normally hear; sound waves go from outer through middle to inner ear. The audiometer can also measure **bone conduction**. Here, a bone-conducting transducer is placed on the subject's skull behind the ear and sound is transmitted by vibration directly into the inner ear, bypassing the outer and middle ear. Bone conduction measures how well the inner ear (cochlea and auditory nerve) is working, regardless of what troubles may impair the outer or middle ear.

The audiogram in Figure 1.**2** depicts measurements of air and bone conduction in the ears of a hypothetical patient who has a cerumen impaction in the right ear and cochlear noise damage in the left ear. Appropriate symbols denote which type of conduction, and which ear, is being tested. Brackets indicate bone conduction thresholds.*** O's or X's indicate air conduction thresholds of the right or left ear, respectively.

The cerumen impaction in this patient's right ear is causing a *conductive* loss. Note that the O's (denoting air conduction thresholds in the right ear) show a mild loss of around 30 dB HL. Bone conduction thresholds for this ear (denoted by brackets opening to the right) are all quite normal at about 5 dB HL. In audiometry, this difference is called an **air–bone gap**, the hallmark of a conductive hearing loss.

In the left ear, the patient has noise damage to the cochlea, but the outer and middle ears are normal. Note that the brackets and X's for the left ear are at the same threshold levels, even where there is a dip in the higher frequencies. This high-frequency *sensorineural* loss shows no air–bone gap.

Incidentally, if we tested the SRT on each ear of this patient, we would expect it to be just over 30 dB HTL in the right ear and just under 15 dB HTL in the left. Do you remember why? One more point: a patient may have

***The brackets shown in this audiogram actually symbolize masked bone conduction, which is often needed. The scope of this text precludes further discussion of this.

both conductive and sensorineural loss in the same ear. This would be referred to as a *mixed loss*.

The above discussion deals with formal audiometric testing, which is seldom readily available in the clinician's office. In most cases, however, tuning forks, along with other basic clinical assessments, can give us a good idea about patients' hearing in the office. We will further discuss these tools, as well as the Audioscope, in Chapter 2.

Summary

If there is any doubt about a patient's hearing, a thorough audiogram is the best way to assess it. In normal individuals, barely perceptible sounds are near 0 dB, and the loudest sounds are 100 dB or more. Most everyday sounds fall between these extremes. It is essential to differentiate between a conductive hearing loss and a sensorineural one. The former results from an outer or middle ear problem, and the latter from an inner ear problem. Air conduction is decreased with conductive hearing losses, while bone conduction remains normal. Both air and bone conduction are decreased when there is a sensorineural loss. Most types of sensorineural hearing losses, such as presbyacusis and noise-induced loss, affect the high frequencies predominantly. Consonants are voiced in the high frequencies, and thus these losses tend to decrease speech discrimination. Overall reception of speech, as measured with two-syllable combinations presented through the earphones, is represented by the SRT. The SRT usually matches an average of the patient's mid-frequency pure tones and is considered to be the most important number reflecting one's hearing ability.

Recommended Reading

Feldman AS, Grimes CT. Audiology. In: Ballenger JJ, ed. *Diseases of the Nose, Throat, Ear, Head, & Neck*. 14th ed. Philadelphia: Lea & Febiger; 1991:1029–1022.

Gelfand SA. *Essentials of Audiology*. 2nd ed. New York, NY: Thieme Medical Publishers; 2001.

2 Equipment and Ear Examination Methods

In a text designed for all medical personnel who treat the ear, we should discuss several levels of equipment needed for the office. To illustrate, a family practitioner from Big Moose, New York is more likely to deal aggressively with ear problems than an internist in a New York City multispecialty clinic. The nearest ENT doctor is a long drive from Big Moose, whereas in the city the specialist might be just down the hall. Thus, rural practitioners will probably desire more equipment and capabilities than their counterparts in the city.

Generally, in the medical field, a well-trained physician is likely to make a correct diagnosis 90% of the time after taking a patient's history and conducting a minimal examination. With ear complaints, however, this is not necessarily the case. Obstacles in the external canal, including cerumen or other debris, can make it impossible to carry out a proper ear examination. Anecdotally, I have seen a number of patients referred for eardrum perforations who simply had strange-looking exudates in the canal and no perforation. A range of equipment and different levels of skill are required to remove these impediments. Then, once the canal is cleared, there is the difficulty of inspecting the eardrum and judging its mobility. Furthermore, how well can one assess a patient's hearing in the office? Diagnostic equipment makes all the difference.

Basic Equipment

Essential equipment for ear examination includes the following items, which will enable the astute clinician to diagnose a majority of problems:

1. An otoscope with a magnifying head, a pneumatic bulb, and different-sized specula
2. Tuning forks
3. An irrigating device, such as a large ear syringe or a WaterPik (be careful!)
4. A bottle of wax softener.

Otoscope

The hand-held **otoscope** with magnification offers a fairly good view of the canal and tympanic membrane (TM). Using an appropriate-sized speculum and **pneumatic bulb**, one can get an idea of TM mobility. This is extremely important for assessing middle ear aeration or lack thereof, due to fluid or partial vacuum. A normal drum shoud move inward and outward when applying positive and negative pressure with the pneumatic bulb. A retracted drum (concave due to negative middle ear pressure) should only move outward with bulb vacuum. A *very* retracted drum, or a fluid-filled middle ear containing no air, will barely show mobility. A perforated TM is completely immobile.

Sometimes it is impossible to achieve a good "air seal" due to the elliptical shape of many patients' outer canals. A poor seal can cause a normal TM to appear immobile. Here, trial and error with different-sized specula, and just plain experience, helps. One other tip: be careful not to exert too much pressure on the canal wall or this could be very painful!

Tuning Forks

The 512-Hz **tuning fork** is the most accepted frequency for assessing hearing using the Weber and Rinne tests. The **Weber test** consists of placing the stem of a vibrating fork on the center of the patient's forehead, crown, or nasal bridge. Sound is transmitted equally into each cochlea by bone conduction. While the fork is vibrating, ask the patient in which ear the sound is louder. A patient with a one-sided sensorineural loss will hear the fork louder in the opposite, normal ear. A patient with a conductive loss will hear it louder on the problem side, due to the autophony phenomenon (see Chapter 1). Remember, however, that the test only applies to the frequency used, in this case 512 Hz. For example, a patient with a one-sided sensorineural loss in the higher frequencies will still have a normal midline Weber test with a 512-Hz fork if there is no damage at this frequency.

The **Rinne test**, modified here for simplicity, is a quick way to screen one ear for conductive (outer or middle ear) hearing loss. Place the stem of the vibrating tuning fork on the patient's mastoid bone for a few seconds, then hold the still-vibrating forked portion about 10 cm from the patient's external ear. Ask, "Is it louder on the bone or out here in the air?" A patient with normal hearing (or a sensorineural loss) will note louder hearing "out in the air," rather than "on the bone." On the other hand, a patient with a significant conductive loss (25 dB HL or more) will hear the fork louder on the bone than in the air. If you have a tuning fork available, try this on your own right ear with the canal open; the Rinne should be "normal." Now occlude your canal tightly with the left index finger, creating a conductive loss, and test again; the Rinne should be "abnormal" or "reversed." Using

the terms "positive" and "negative" is often recommended, but these can be ambiguous. Documenting "air conduction is greater than bone" (or vice-versa) leaves the least room for doubt.

Tuning forks of 256, 1024, and 2048 Hz are also available and can be useful in many ways. For example, most conductive losses are severest in the lower frequencies. The 256-Hz fork may pick up abnormalities that the 512-Hz one would miss; thus it is the most sensitive one for conductive losses, especially when using the Rinne test. All the frequencies can be used to compare a patient's right ear to his/her left ear, or to the examiner's ear, for rough assessment. Individuals with advanced high-frequency loss from either noise damage or aging may show impairment at 1024 or 2048 Hz.

Other simple assessments can be made. The light rubbing of dry fingers is in the 3000 Hz range, as is the ticking of a watch**.** The sound intensity, when near the ear, is probably 25 dB HL or less. These are rudimentary but valuable screenings for high-frequency loss.

Cerumen Removal Devices

Irrigation of the external canal for cerumen removal can be done with any of several devices that can introduce a stream of water under moderate pressure. I mention the WaterPik as a good option, although it is not medically intended for this purpose. It is commercially available at a reasonable price, but it can be cumbersome and messy. In addition, it can generate huge pressure; be careful to use the lowest settings. Ear syringes are also available.

Here is some general advice for irrigation. Use body-temperature water to avoid caloric-induced vertigo. Clean tap water is fine; adding peroxide may help the loosening process but is probably not necessary. Have the patient hold a receptacle beneath the ear to catch the water. Pull the auricle posteriorly with the free hand to straighten the canal. To avoid trauma, aim the stream posterosuperiorly at the canal wall and not directly at the TM; this tends to force the water around and out, without applying severe pressure on the drum. Even with 20 years of experience, I perforated a patient's thin, scarred drum several years ago using proper technique. A good rule would be to never irrigate an ear when the patient has a history of perforation or scarring. Also, one should stop irrigating if the patient complains of pain.

A frequently encountered problem is firm or gummy cerumen that will not budge. In this case, postpone further clearing efforts and use a wax softener, such as **Debrox** or **Cerumenex**. Some physicians use a stool softener, **docusate sodium**, and claim it works effectively. Softening could be done during the office visit by having the patient lie down for 15 minutes

with an earful of softener, after which the ear would be re-irrigated. The patient could also be sent home with instructions to use the drops tid for a few days before the next visit. Usually, the ear can then be cleared. If not, a referral would be appropriate.

Advanced Equipment

The purchase of two more pieces of equipment may be considered to further expand a clinician's office diagnostic abilities:

1. An Audioscope
2. A tympanometer

The **Audioscope** is a hand-held device used to screen hearing. It resembles an otoscope in size and shape. The one I have, patented by Welch Allyn, can test hearing at 500 Hz and 1, 2, and 4 KHz, at levels of 20, 25, or 40 dB HL. It requires a charger and periodic returns to the manufacturer for calibration, but it is a most valuable screening device. One can look through its otoscope speculum to examine the ear as well, though the orifice is quite small, and its main use is to make sure you are aiming the device at the TM rather than the canal wall. One could consider having a more elaborate and expensive **audiometer** in the office, but if an audiologist is available, a referral is probably more appropriate.

Fig. 2.**1** Equipment for ear evaluation. Clockwise, from upper left: WaterPik, Audioscope, tuning forks, otoscope with pneumatic bulb, specula, wax softener.

A **tympanometer** is a device that assess compliance of the TM and middle ear pressure. It plots a curve that indicates middle ear fluid, pressure abnormalities denoting eustachian tube dysfunction, or abnormal mobility of the drum due to scarring or ossicular chain problems. It is an especially useful item for screening the pediatric population, the little ones who are difficult to examine or test audiometrically. A pressure probe is placed just like a speculum in the outer canal, and compliance readings are taken in a short period of time. These assess TM mobility in a much more sensitive way than the pneumatic otoscope. As a specialist, I enjoy matching my impressions from the office examination with the "tympanogram." Drawbacks of the device include air-seal difficulties, errors in probe placement, and false readings due to cerumen impactions. Tympanometry curves will be discussed in Chapter 5.

Finally, ENT specialists have equipment for direct ear examination that expands their capablities beyond most nonspecialists. By using a light source on the head, they are able to use *both hands* to examine an ear—or nose and throat, for that matter. The illumination can be from an electrical headlight or a traditional head mirror with an indirect light source on a stand. With the use of bright, direct light from the vantage point of the eyeball, one is able to look "down the pipe" into narrow, deep spaces. Head-mounted lighting enables the specialist to use a magnifier with an ear speculum in the nondominant hand, and to work with instruments in the dominant hand. Half-circle "operating" lenses allow one to use various curettes, loops, or hooks to remove cerumen or foreign bodies under magnification. "Alligator" forceps and suctions may also be used to remove debris or secretions. Irrigation with pinpoint spray devices, or insufflation of medications, can also be done. These require more equipment, both small and large. The electrical pumps and their associated apparatus are usually contained in a console, which takes up space and is expensive. Hardware of this type is available through various ENT equipment companies.

Summary
Emphasis must be placed on the *pneumatic otoscope* for diagnosis. The *mobility* of the drum is a key factor in diagnosing TM retraction, middle ear fluid, and TM perforation. Tuning forks, using the Weber and Rinne tests, can provide a good idea about conductive (outer or middle ear) hearing loss versus sensorineural (inner ear) loss. Tuning forks of varied frequencies (along with a ticking watch or rubbing of fingers) can provide additional information about hearing, especially when used to compare a patient's hearing to the examiner's. Cerumen removal is often necessary and can be facilitated by commercial wax softeners or stool softeners. Debris of any type in the canal can impair diagnosis, thus necessitating more sophisticated equipment, or perhaps even ENT referral. The Audioscope and the tympanometer are two other more elaborate pieces of equipment that one might have in the office to screen hearing and middle ear pressure.

3 Useful Anatomy and Function

This chapter describes the structure of the ear and discusses its pertinent physiology and innervation. I have limited the coverage to what is most important to the practitioner, while making points of clinical interest along the way. The anatomy and physiology of the ear are quite complex for such a small structure of the body, but reading through these pages will provide a good background for the clinical chapters. In this and the following chapters on diagnosis and treatment, we progress anatomically from the outside inward.

The External Ear

The Auricle

The external ear consists of the outer portion, the *auricle* (or *pinna*), and the *external auditory canal,* leading to, but not including, the tympanic membrane (TM). Figure 3.**1** shows an auricle with its landmarks labeled.

The auricle is composed of skin overlying an irregular framework of fibroelastic cartilage; it "gathers" sound and directs it into the canal. With the external canal, it actually acts as a resonator to slightly amplify lower frequency sounds. It also helps to localize sounds, especially in conjunction with the other ear. Innervation of the auricle occurs through the greater and lesser auricular nerves (from cervical roots C3 and C2) pos-

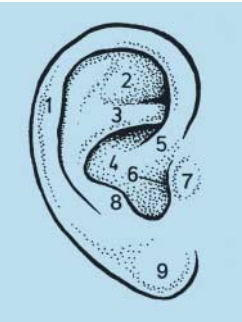


Fig. 3.**1** The auricle and its landmarks:
1. Helix; 2. Scapha; 3. Antihelix; 4. Concha;
5. Crus of helix; 6. External auditory meatus;
7. Tragus; 8. Antitragus; 9. Lobule.
(Source: Becker W, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994)

teriorly, and the auriculotemporal branch of cranial nerve V anteriorly. The skin and vascular channels on the outer surface are somewhat adherent and "brittle," making the auricle susceptible to traumatic hematoma formation that might lead to "cauliflower ear" if not treated. The innervation is not particularly sensitive, especially in the lobule. To prove this, pinch yourself anywhere on the auricle; then compare the pain with the same insult to other body sites. This insensitivity facilitates ear piercing, but unfortunately masks frostbite pain.

The External Auditory Canal

The external auditory canal (EAC) consists of a cartilaginous outer one-third and a bony inner two-thirds. Figure 3.**2** shows its relationship with deeper structures. The entire length of the canal is a little over 3 cm from the external meatus to the TM. From the outside in, the outer third is directed slightly posteriorly, whereas the inner two-thirds is directed anteroinferiorly and has an anterior bony hump that occludes a view of the very front portion of the TM. Because of this curvature, the canal can best be straightened, as for eardrop insertion, by pulling the auricle backward. The skin of the external canal changes greatly from the outside in. In the outer cartilaginous segment, it is thick and contains hairs and numer-

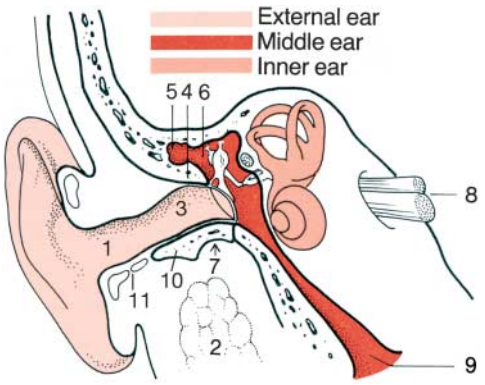


Fig. 3.**2** The external auditory canal and its relationships:
l. Cartilaginous part; 2. Parotid gland; 3. Bony part; 4. Lateral wall of epitympanum (attic); 5. Mastoid antrum; 6. Attic (epitympanum); 7. Temporomandibular joint fossa; 8. Facial, vestibular, and auditory nerves (VII and VIII); 9. Eustachian tube; 10. Bone of tympanic ring; 11. Fissure of Santorini in cartilage of external canal. (Source: Becker W, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994)

ous glands, both sebaceous and cerumen-producing. These three elements exist together as "apopilosebaceous" units. The skin lining the inner bony two-thirds is very thin, with little hair and no glands, but it is well-innervated and very sensitive to touch, in contrast to the skin of the auricle. The bony portion of the external canal is also known as the *tympanic ring*.

The "fissures of Santorini" (discontinuities in the cartilage of the outer canal) enable infection or neoplasm to easily spread down and forward into the adjacent parotid gland. Hence, a patient with a severe external otitis can develop cellulitis and parotitis adjacent to the ear. The outer third of the canal can be very narrow in width in some patients, limiting good access to the inner two-thirds and the TM. However, these individuals usually have normal-caliber bony canals further in. In general, the external canal is about 9 mm in height and 7 mm in width (due to the anterior bony hump).

The EAC is innervated by branches from four nerves: cranial nerves V (anteriorly), VII, IX, and X. The last three, innervating most of the canal, send their branches into the brainstem via the nervus intermedius and Arnold's nerve. The relays of Arnold's nerve near the nucleus ambiguus explain why stimulation of the canal during cleaning will often produce a cough reflex.

It is significant that other areas innervated by these four cranial nerves can transmit *referred pain* to the ear. Examples include post-tonsillectomy otalgia (ear pain) via cranial nerve IX or, more ominously, otalgia from malignancies in the tonsil, hypopharynx, or supraglottic larynx via cranial nerves IX and X. Several years ago I was referred a patient whose *only* complaint was a left earache, not even during swallowing. On laryngoscopy, he had a massive squamous cell carcinoma of the left hypopharynx! Finally, be advised that otalgia may even be referred from organs in the chest—disease of the heart, lungs, great vessels, and esophagus may all cause ear pain via the vagus nerve (X).

The Middle Ear

The Tympanic Membrane and its Landmarks

The middle ear begins with the *TM*. This is the most important structure for the clinician diagnostically, because its appearance, and what can be seen through it, are by and large the only anatomic clues to what goes on inside (Figs. 3.**3** and 3.**4**). The most visible landmark beneath the TM is the *manubrium*, or handle, of the *malleus*, the lower portion of this first ossicle. The upper portion of the manubrium has a visible prominence, seen near the top of the TM, called the short process. The bottom end of the handle, at the

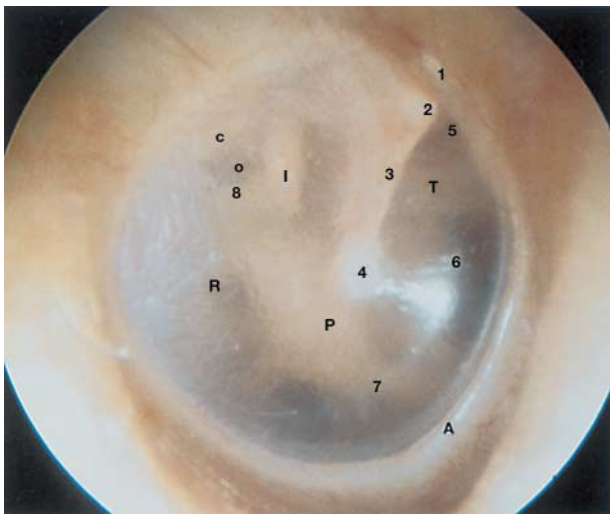


Fig. 3.**3** Normal tympanic membrane of right ear:
1. Pars flaccida; 2. Short process of the malleus; 3. Handle (manubrium) of the malleus; 4. Umbo; 5. Supratubal recess; 6. Eustachian tube orifice (just to the right of the light reflex); 7. Hypotympanic air cells; 8. Stapedius tendon; c. Chorda tympani; I. Incus; P. Promontory; o. Oval window; R. Round window; T. Tensor tympani; A. Annulus. (Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

center of the TM, ends in the *umbo*, the "apex" of the drum (the TM is actually somewhat conical in shape, from the periphery toward the center). Thus, the umbo, the deepest central depression, has a circular appearance.

The examining light usually reflects back most brightly from the TM in a triangular shape, located anteroinferiorly to the umbo, the *light reflex*. When a TM is dulled by acute or chronic infection, or thickening for other reasons, the light reflex may be decreased or absent, but this is not an infallible diagnostic criterion. A light reflex can still be seen in many abnormal middle ears, especially those with clear fluid effusions.

Most of the TM (the *pars tensa*) has three layers: outer squamous epithelium, middle fibrous tissue, and inner cuboidal epithelium. Above the short process of the malleus is a variable and irregular small portion of the drum called the *pars flaccida*, which contains no fibrous middle layer, only an outer squamous and inner mucosal layer. The pars flaccida is clinically significant as a site for possible *cholesteatoma* formation. The same applies for the marginal posterosuperior portion of the pars tensa, which may also be

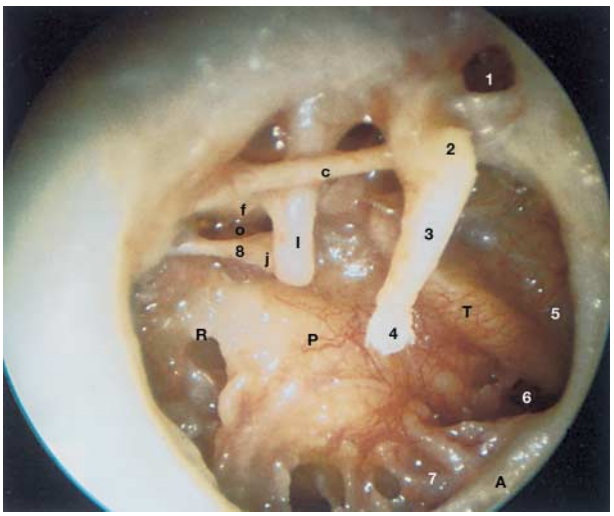


Fig. 3.**4** Structures of the right ear after removal of the tympanic membrane: f. Facial nerve bulge; j. incudostapedial joint. See also key to Fig. 3.**3**. (Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

deficient in the middle fibrous layer in some disease states. Cholesteatoma, a squamous ingrowth, is discussed in more detail later.

Posterosuperiorly, the long process of the *incus* is frequently visible, deep to the drum, as it inserts into the stapes head. Going backward, the *stapedius* tendon may be seen as well. The vertical long process of the incus and the horizontal stapedius tendon can often be seen as a faint white backward-tilted "V." Overlying and above them, just adjacent to the margin of the drum, the white band of the *chorda tympani nerve* may also be seen through the drum. This arises from the facial nerve (cranial nerve VII) and courses forward between the malleus and incus to eventually innervate the anterior tongue, receiving taste sensation. Not well seen through the TM, the *stapes*, the smallest bone in the body, transmits sound from the incus into the oval window via its footplate. It is shaped like a stirrup and is directed medially toward the inner ear. The *round window niche* and the bony bulge of the *promontory* are structures of the medial wall of the middle ear. The visibility of all these landmarks varies with individual anatomy and scarring of the drum.

The periphery of the pars tensa, bordering the innermost external canal, contains a dense white fibrous ring called the *annulus*. Variably present is

the *vascular strip* of vessels, not visible in Figure 3.**3**, but very prominent in Figure 5.5. This is seen on the upper drum just posterior to the manubrium, giving off vessels that course down the TM just behind the manubrium, as well as posteriorly, running along the peripheral annulus. These vessels may become bright red and inflamed in the early stages of acute otitis media. However, they may also be somewhat prominent in the normal individual.

The Ossicles and their Function

Here the ossicles and contents of the middle ear will be discussed in more detail. Figure 3.**5** depicts the position of middle ear and related structures. The TM, the bone of the tympanic ring, and outer (cortical) mastoid bone have been removed in this depiction.

The *ossicles*—the malleus, incus, and stapes—transmit sound vibrations from the large tympanic membrane into the small oval window, at a huge mechanical advantage. This system is really an "impedance-matching" one that enables sound waves in the air, at relatively low energy, to transfer sound waves into the fluid-filled medium of the inner ear, which offers higher resistance to sound flow. A fish, for example, has no need for a mid-

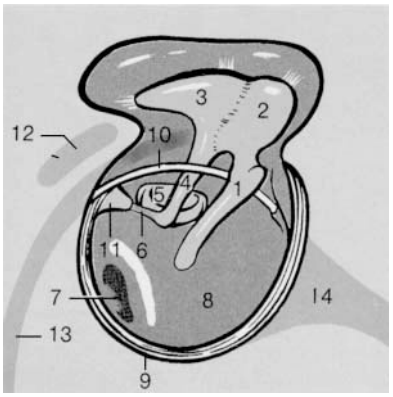


Fig. 3.**5** Middle ear and related structures:
1. Manubrium of malleus, near the short process above; 2. Head of malleus; 3. Body of incus; 4. Long process of incus; 5. Stapes footplate; 6. Stapedius tendon; 7. Round window; 8. Promontory; 9. Fibrous annulus of the TM; 10. Chorda tympani nerve; 11. Pyramidal eminence leading to stapedius muscle and tendon; 12. Location of lateral semicircular canal bulging within the mastoid antrum (this cavity is covered over here); 13. Vertical portion of facial nerve; 14. Eustachian tube.
(Source: Becker W, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994)

dle ear—sound waves already come in from a fluid medium. Thus, there is only a membrane between the ocean and its fluid-filled inner ear. Take this fish out of water, and it will have about a 30-dB hearing loss for air-transmitted sounds. If a human had no middle ear mechanism, he too would have about a 30-dB conductive loss. This situation is actually simulated with a fluid-filled middle ear cavity, or, alternatively, a tympanic perforation in combination with a disruption of the ossicles. In either situation, we are trying to transmit unenhanced sound waves from air directly into a fluid medium. The large ratio of TM/oval window surface area, further strengthened by the mechanical lever action of the ossicles, overcomes this potential loss.

Conductive hearing losses of varying degrees result from different impairments of the middle ear system. Examples include perforations, scarring, negative pressure, fluid effusions, ossicular fixation, ossicular disruption, and, of course, external canal blockages. Contrary to popular belief, a moderate-sized perforation of the TM does not cause a very large hearing loss—there is still ample drum left, along with the mechanical lever action of the ossicles. The tiny perforation of a ventilating tube that is placed to correct middle ear fluid causes no measurable loss, and of course, may correct a sizable loss related to the middle ear trouble that it remedies. The worst conductive losses are on the order of 50–60 dB HL. These might be seen with a congenital atresia of the canal and middle ear, with dense outer obstruction as well as associated deformity of the ossicles.

Related and Adjacent Middle Ear Structures

Continue to refer to Figure 3.**5** as we discuss the anatomy and function of other structures in or near to the middle ear. As shown, the largest parts of the ossicular mass, namely the heads of the malleus and incus, are located in the *epitympanum,* or *attic,* above the middle ear cavity. Thus, they cannot be seen through the TM. As mentioned earlier, the chorda tympani courses forward horizontally from the facial nerve between the malleus and incus. It is not always seen through the TM.

There are two tiny muscles in the middle ear, the *stapedius* and the *tensor tympani*. The stapedius, innervated by a branch from the facial (VII) nerve, contracts reflexively to loud sounds, protecting the ear from noise damage by bracing the stapes to immobilize it. The tensor tympani, not shown in Figure 3.**5** but in Figure 3.**4**, inserts into the manubrium of the malleus. It is innervated by the trigeminal (V) nerve and its function is probably similar to the stapedius for sound protection. It also contracts with certain pharyngeal muscles, suggesting a role in bracing the ossicles when swallowing.

The *facial (VII) nerve* has a complex course through the temporal bone. It courses laterally from the brainstem through the internal auditory canal

(IAC) toward the anterosuperior middle ear wall, then turns sharply posteriorly, just deep to the malleus, still in the otic capsule of the inner ear. From here, it courses horizontally backward above the stapes in the oval window and gradually turns vertically downward behind the middle ear to exit anterior to the mastoid tip, after giving off branches to the chorda tympani and stapedius. Figure 3.**5** shows only the horizontal and vertical portions.

The facial nerve may have bony dehiscenses in its course just deep to the middle ear, making it susceptible to surgical injury. Very rarely, a patient with acute otitis media may present with a facial paralysis due to infection through a dehiscence. Also, the nerve's usual *encasement* in bone may aggravate Bell's palsy. This viral or postviral neuropathy causes inflammation and edema of the nerve, which then has no place to expand! In times past, surgical facial nerve decompressions were done for this disease with some reports of success.

Continuing on its course, the nerve enters the parotid gland anteriorly after leaving the stylomastoid foramen. Once again, it is exposed to possible compression by a parotid tumor, or to surgical injury while removing that tumor. Obviously, this nerve poses a great challenge and threat to the ENT physician.

The *mastoid antrum*, a cavity posterior to the epitympanum (covered over in this diagram), contains the bulge of the *lateral semicircular canal* beneath the tip of the short process of the incus. Surrounding this, the honeycomb of the rest of the mastoid air cells pneumatizes a large area of the bone (also not shown), providing an "air cushion" to ameliorate pressure changes in the middle ear. The development of the air cell system occurs in childhood and varies in amount at maturity. Healthy, aerated middle ears develop well-pneumatized mastoids. Adults who have weathered many eustachian tube problems during childhood often have underdeveloped or nonaerated mastoid bones. These changes, seen on scans or conventional X-rays, are sometimes incorrectly read by radiologists as "mastoiditis"—a better term would be an "underdeveloped" or "sclerotic" mastoid. Patients with these findings are susceptible, however, to chronic middle ear and mastoid problems and may well develop mastoiditis.

Return again to Figure 3.**5** and note that most of the medial wall of the middle ear is taken up by the *promontory*, a convex bony bulge overlying the basal turn of the cochlea. Posteroinferiorly, there is a niche for the *round window*, the termination of the scala tympani of the cochlea (discussed later). Jacobson's plexus (cranial nerve IX) (not shown) innervates the lining of the middle ear, branching over the promontory.

Anteriorly, the respiratory mucosa of the middle ear becomes thicker, with increased mucous glands, at the mouth of the *eustachian tube*.

This gives the potential for the thick mucoid effusions we see with chronic blockage of the eustachian tube. The tube itself heads inferomedially to terminate in the nasopharynx and is usually not patent. Muscular actions, mainly by the tensors of the palate, intermittently open the tube. This occurs during speech, yawning, and swallowing, as well as with other unconscious movements of the pharnyx. Opening the tube allows air, which is otherwise steadily passively absorbed from the middle ear, to be replenished to atmospheric pressure.

Just beneath the drum, the *jugular bulb* resides in the *hypotympanum* (not shown in Fig. 3.**5**). It receives blood from the sigmoid sinus, which courses down from the posterior mastoid. A *glomus jugulare* vascular tumor may rarely arise from the jugular bulb, presenting with pulsatile tinnitus as a usual early complaint. Some are visible as red masses deep to the TM, but others may be hidden from view.

The Inner Ear

The interconnected sensory organs of the inner ear are the cochlea (the auditory apparatus) and the vestibular apparatus, composed of the utricle, saccule, and semicircular canals. These are surrounded by the petrous por-

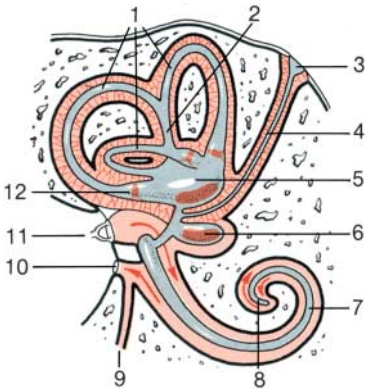
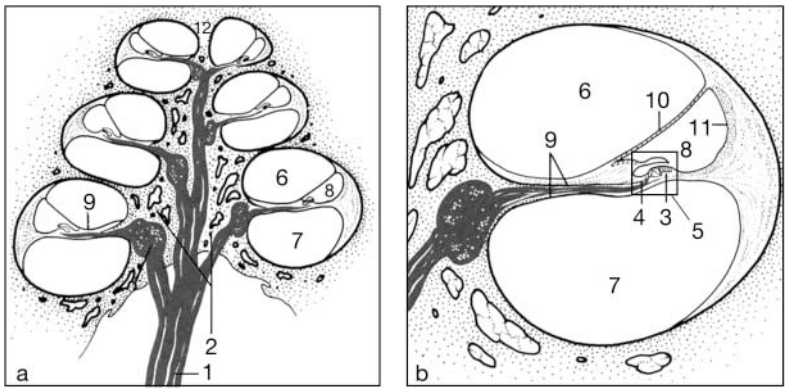


Fig. 3.**6** Diagram of the inner ear:
1. Membranous semicircular canals (lateral, superior, and posterior); 2. Common crus of posterior and superior canals; 3. Endolymphatic sac; 4. Endolymphatic duct; 5. Utricle; 6. Saccule; 7.Cochlear duct; 8. Apex of cochlea; 9. Perilymphatic duct; 10. Round window; 11. Oval window; 12. Ampulla of posterior semicircular canal.
(Source: Becker W, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994)

tion of the temporal bone known as the *otic capsule*. Figure 3.**6** diagramatically shows the structural relationships of a right inner ear, as seen from an anterosuperior angle. The middle ear lies to the left of the diagram and nerve structures are not depicted.

The Cochlea

The *cochlea* is a complex membranous organ within a cavity whose bony outer surface approximates the spiral shape of a snail shell. The wide part of this spiral, containing the basal turn, is posterior and medial. The narrow apex points anterolaterally. There are about two and two-thirds turns through the spiral, from base to apex. The fibers of the auditory nerve run through its bony center, or modiolus, eventually to join the vestibular nerve divisions to form cranial nerve VIII (the vestibulocochlear nerve) in the IAC, which opens into the posterior cranial fossa. The overall shape is also similar to a Christmas tree—the auditory nerve, within the modiolus, makes up the stem (Fig. 3.**7a**). The tube of this spiral, wound around the "stem," actually contains three compartments. An enlarged cross-section is diagrammed in Figure 3.**7b**.



Figs. 3.**7a,** b Axial cross-section of the cochlea (**a**) and enlarged section of a single turn (**b**).
l. Auditory nerve; 2. Modiolus (bony center); 3. Inner and outer hair cells; 4. Basilar membrane; 5. Organ of Corti (outlined by box); 6. Scala vestibuli (containing perilymph); 7. Scala tympani (containing perilymph); 8. Scala media (containing endolymph); 9. Spiral lamina; 10. Reissner's membrane; 11. Stria vascularis; 12. Apex of cochlea.
(Source: Becker W, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994)

The organ of Corti is the key sensory area within the scala media. Here, inner and outer hair cells are stimulated, via bending of their stereocilia, by sound waves. These waves are transmitted from the vibrating stapes footplate into perilymphatic fluid to displace the basilar membrane, on which the hair cells rest. This membrane is narrower near the oval window, where higher frequencies are perceived, and gradually widens toward the apex of the cochlea, where lower frequencies are perceived. The hair cells send their impulses through nerve fibers in the spiral lamina toward the auditory nerve, converting mechanical energy into electrical energy.

Sound waves from the vibrating stapes footplate in the oval window enter the scala vestibuli at the posterolateral end of the basal turn to circulate through the cochlear fluids. The round window and its membrane, located beneath the oval window, also connect the middle and inner ear cavities by way of the scala tympani. The scalae vestibuli and tympani communicate at the cochlear apex. Thus, the round window can actually receive sound waves and transmit them retrograde in some middle ear conditions where the stapes footplate is fixed and does not vibrate.

The cavities of the scalae vestibuli and tympani contain high-sodium, low-potassium perilymph, which communicates with cerobrospinal fluid (CSF) via the cochlear aqueduct. Endolymph, within the scala media, or cochlear duct, is high in potassium and low in sodium. Without going into great detail, there are other ducts and sacs (depicted in Fig. 3.**6**) that maintain and stabilize this difference, which is vital to normal cochlear function. In *Méniere's disease*, or *endolymphatic hydrops*, there is a malfunction of secretion/absorption that results in electrolyte imbalance here, causing swelling of the scala media and bulging of Reissner's membrane. The result is a sense of ear fullness, tinnitus, and hearing loss. Vertigo develops as well, from swelling of the vestibular endolymphatic cavities.

Over 15 000 hair cells are arranged in three to four outer rows, and an inner single row, along the organ of Corti. "Tuning" of the cochlea seems to be present, with highest frequencies perceived at the basal turn near the oval window, and lowest frequencies near the apex, medially. Whether this explains the predominance of high-frequency loss from noise over-exposure is controversial.

The physiology of the inner ear is complex. Years of research and publications have been devoted to the subject and are available elsewhere to the interested reader.

The Vestibule

Regarding the *vestibular* (balance) *apparatus*, the *utricle* and *saccule* make up the portion that perceives linear acceleration and position sense. They are located just deep and posterior to the end of the basal turn of the cochlea (Fig. 3.**6**). These structures, like the cochlea, also contain hair cell sensors, but they receive stimulation from positional shifts rather than sound waves. "Otoliths" are microscopic crystals embedded in a gel above the hair cell receptors. These dense little "weights" shift with position change, thus bending and stimulating the hair cells, sending electrical information into the brainstem via the vestibular portion of cranial nerve VIII.

More posteriorly and superiorly, the *semicircular canals*—lateral, superior, and posterior—comprise the other portion of the vestibular system. They are oriented in three planes, each perpendicular to the other. These sense "rotational" head motion, or angular acceleration, interacting with the neck and eyes to maintain orientation during turning motions. If they are artificially stimulated calorically by cold or warm water in the ear, classic vertigo is produced, just as it would if the head rotated repeatedly and then suddenly stopped. Diseases of the labyrinth produce vertigo, such as with the endolymphatic swelling of Méniere's disease. Another common condition of recurrent vertigo, benign positional vertigo, is believed to result from abnormal displacement of otolithic debris (cause uncertain) into the posterior semicircular canal. More information about these disorders, including a discussion of nystagmus, will follow in Chapter 8.

The *vestibular nerve*, receiving information from the balance organs, has a superior and inferior division. These divisions converge with the *auditory nerve* from the cochlea to form cranial nerve VIII. Cranial nerve VIII is immediately adjacent to the facial nerve (cranial nerve VII) as the two enter the brainstem via the IAC and its internal auditory meatus. In the brainstem, nerve fibers from the utricle and saccule, perceiving linear acceleration, interconnect mostly with nerves to the anti-gravity muscles of the back and limbs. Fibers from the rotation-perceiving semicircular canals, on the other hand, have connections with the ocular muscles and the muscles of the neck. The IAC and its meatus are the site of the often-discussed "acoustic neuroma." This rare benign tumor indolently grows over years to create hearing loss, dysequilibrium, and sometimes facial nerve paralysis.

A single source of blood supply to the inner ear is a nonanastamosing end artery, the internal auditory artery. Its smaller branches terminate in the stria vascularis, which supports the circulation and probably aids in the fluid balance of the organ of Corti. In the past, an occlusion of the internal auditory artery was often seen as the cause of the phenomenon of "sudden sensorineural hearing loss." This view no longer accepted,

although vascular compromise of its branches does play a part in the disorder.

Recommended Reading

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4 Disorders of the External Ear

This chapter begins our discussion of ear diseases. In this and the following chapters we will progress anatomically from the outer ear inward. At the outset, let us emphasize that the ear is fairly *compartmentalized* with respect to most of its disorders. That is to say, the outer, middle, and inner ear have their own individual problems, without much overlap between these anatomic compartments, although a number of exceptions exist.

Unfortunately, some medical and lay persons with a limited knowledge of ears tend to lump the symptoms and diagnoses of ear disorders together in one large basket. The educated clinician, on the other hand, knows that certain ear symptoms, even without the benefit of an examination, point toward the involved portion of the ear. Disorders of the *external ear* are likely to cause one or more of the following specific complaints: itching, pain, tenderness, swelling, blockage of hearing (autophony), and drainage. Secretions in the canal may also cause noises during chewing or manipulation of the ear.

The external ear includes the auricle and the external auditory canal (EAC), but some disorders *outside* the external ear cause referred aural symptoms. These will be discussed first, and we will then proceed from the auricle inward.

Temporomandibular Joint Syndrome

Temporomandibular joint (TMJ) syndrome (or disorder) was first known as Costen's syndrome, having been described in l934 by an otolaryngologist, Dr. James B. Costen. The name refers to the discomfort caused by a malfunction and/or inflammation of the TMJ, where the condyle of the mandible sits in a fossa of the temporal bone to form a hinge joint.

With a cartilage disk intervening, the mandibular condyle slides down and forward as the jaw opens, and then back and upward as it closes. A number of factors can subject the joint to wear and tear. Since it borders on the external ear canal and shares innervation with it via the auriculotemporal branch of the trigeminal nerve, symptoms of joint inflammation or derangement are often referred to the ear. The disorder has a relative, **myofascial pain syndrome.** This term refers more to facial pain symptoms

from tension and spasm in the muscles around the joint. The two disorders often coexist.

TMJ syndrome is the cause of a large number of ear complaints. Symptoms and findings vary greatly from patient to patient. The actual pathologic changes range from nothing at all to mild arthritic change to extreme mechanical disruption of the joint. Thus, there can be an anatomically normal, but painful, joint or advanced degeneration of the disk and joint surfaces, with severe clicking or even locking of the joint. The cause is most often clenching or grinding of the teeth (**bruxism**), either unconsciously in sleep or as a habit when awake. However, dental or bite abnormalities, previous trauma to the mandible, and other factors (possibly even genetic) may be responsible.

Those afflicted have variable complaints. Some patients report a feeling of intermittent or chronic fullness in the ear, with a perceived need to "clear" it. Others may complain of pain, sometimes sharp and momentary, especially when chewing, or sometimes dull and chronic. The pain or ache in the ear may also radiate in any direction, but most often inferiorly down the neck, possibly secondary to muscle spasm. A sense of hearing loss, tinnitus, or even vertigo may be present. The cause of these latter symptoms is unclear, although sensitizing of the ear by way of neurologic referral is a convenient explanation. The patient is often convinced there is an ear blockage or infection, but may occasionally point directly to the TMJ as the source of the discomfort. Although the mandible works as a single unit, the symptoms are often worse on one side or completely unilateral.

Careful questioning by the examiner may uncover more historical information, which might be valuable in formulating the cause of the problem. Here are some examples:

1. The symptoms are worse overnight or on awakening.
2. A spouse, or even the patient, notes audible or visible bruxism during sleep.
3. Bruxism occurs while the patient is awake.
4. The patient chews gum or other objects (e.g., pencils) constantly.
5. The patient has been subjected to undue tension and stress lately.
6. A number of posterior molars have been extracted in the past.
7. Chewing food hurts.

Physical findings should support your suspicions. Place a finger over the TMJ on both sides, or actually in the external canal, pressing gently forward on the joints. Have the patient open and close the mouth, and ask if either joint is tender. If so, you can feel strongly about the diagnosis. You may even feel or hear **crepitance** (a crackling sensation) or clicking if the disk is damaged. On oral examination, abnormal wear of the teeth from

the bruxism may be present. Look for gross bite misalignment or multiple missing molars, findings that can cause stress on the joint.

Of course, the ears themselves should be examined and they should show no evidence of real disease. TMJ syndrome and eustachian tube dysfunction have similar symptoms and are often confused. In a typical scenario, the practitioner listens to the patient's symptoms, sees a scarred (but functionally normal) tympanic membrane (TM), and puts these findings together to conclude that there is a middle ear problem. Some patients are thus treated for long periods with antihistamines, decongestants, steroid nasal sprays, and even antibiotics. To avoid this error, an accurate ear examination is necessary, with good assessment of hearing and TM mobility. One equivocation, though, is that eustachian tube and TMJ disorders may coexist in the same patient! Here is where good diagnostic capability with ears makes all the difference.

Once middle ear or other problems are ruled out and TMJ syndrome is diagnosed, treatment begins with a caring and interactive discussion. A patient's insights and acceptance are needed to pursue remedies. Often, muscle tension and spasm aggravate the joint pain. Stress reduction, if possible, with conscious avoidance of bruxism while awake, will help. Gum chewing should be avoided, as well as very chewy foods. **Anti-inflammatory medications** are a great help. Rehabilitative jaw exercises can be recommended.

Referral to a dentist or dental subspecialist should be an early consideration. Oral appliances called **night guards** can be made. They are worn at night and serve to separate the posterior upper and lower teeth, relieving stress on the joint. Joint surgery is available for extreme cases. The treatment of this disorder is more in the realm of the dental specialist than the general physician or otolaryngologist, but correct *diagnosis* is imperative.

Temporomandibular Joint Dislocation

One distressing TMJ problem is occasionally seen in the emergency room. A patient may present with the jaw *locked open*, holding a towel to catch saliva. This usually occurs after a wide opening of the mouth, such as with yawning. Individuals with significant disk derangement or laxity in the joint are prone to it. The treatment is manual reduction of the dislocation by rotating the mandible with the thumbs pressing downward on the mandibular molars inside the mouth. This will often require someone with experience in this procedure, such as an oral surgeon or ENT specialist. Muscle relaxants are often necessary before the reduction.

Summary
The key point to make about TMJ syndrome is that it is the most common cause of multiple ear symptoms in patients with normal ears, and that it is often misdiagnosed as a eustachian tube or middle ear disorder. The patient usually presents with unilateral or bilateral fullness in or near the ear, and ache may be present. Thorough evaluation of the TM, its mobility, and hearing can rule out ear disease. A positive history of bruxism, coupled with findings of joint tenderness or crepitance, supports the diagnosis. Patient counseling and anti-inflammatory medicines, along with possible dental consultation and nighttime appliance fitting, are the mainstays of treatment. The primary practitioner can diagnose and initiate treatment for TMJ syndrome, with elective referral to an ENT or dental specialist for confirmation of the diagnosis and further options.

Neuralgias Involving the Ear

Webster defines "neuralgia" as a severe pain along the course of a nerve or in its distribution. Obviously, this is a concise and accurate description. In diagnosing neuralgia, there are few physical findings to support one's impression. Clinical suspicion is aroused mostly by the patient's history—the description of the type of pain and its location. Typically, neuralgia pains are severe and lancinating, lasting only a few seconds to a half-minute, and are variable in frequency. Often, there is tenderness to light touch. The duration of the disorder is also variable. Some neuralgias are short-lived, accompanying a viral illness, and others are chronic and disabling, lasting months to years. Notably, almost all are unilateral.

The cause of most neuralgias is uncertain, although viral or postviral neuropathy may play a role. It has recently come to light that sometimes direct nerve compression is involved. Trigeminal neuralgia, for example, has been cured by surgically alleviating a vessel's compression of the nerve root near the brainstem. In addition, ablation procedures, by nerve section or toxic injection, have been done successfully for years. These surgical successes certainly support an anatomic basis for the disorder.

As discussed in Chapter 3, innervation of the external and middle ear arises from multiple roots, namely C2 and C3, and cranial nerves V, VII, IX, and X. Thus, several neuralgia syndromes include ear pain. Diagnosis of the following neuralgias is often by exclusion—one must rule out other reasons for the pain. However, the severity of the pain, sensitivity in the area of nerve distribution, and unilaterality are all contributing considerations.

Trigeminal neuralgia, also known as *tic douloureux*, is the most well-known. It involves one or more of the three divisions of cranial nerve V, usually the lower two. The lancinating pains are typically triggered by light touch, or even the wind! The pain distribution may include the ear, presumably via the auriculotemporal branch. **Occipital neuralgia** occurs in the distribution of this branch of C3 in the posterior scalp and mastoid regions. Often the patient even complains of "hair" tenderness in these areas.

Extremely rarely, **glossopharyngeal neuralgia**, arising from cranial nerve IX, is experienced as pain in the posterior oropharynx, tonsil, or base of tongue, with radiation to the ear (via Jacobson's nerve). Also rare, **sphenopalatine neuralgia** arises from the ganglion of the same name in the fossa behind the maxillary sinus, with nerve relays from V and VII. It manifests itself as a unilateral pain in the maxillary, orbital, and temporal regions, with ear pain as well.

Treatment for all these disorders depends on the chronicity of the symptoms and the certainty of the diagnosis. Acutely, one should consider the usual pain medications, if all other causes are ruled out. More chronically, **carbamazepine** (Tegretol) is effective, although the side effects of drowsiness and rare severe reactions (bone marrow depression) warrant great caution. Monitoring the complete blood count, before and during use, is indicated. **Oxcarbamazepine** (Trileptal) is a newer compound without the above side effects, although neuralgia is not yet listed in its indications.

Summary

Most importantly, neuralgias are severe pains, often lancinating and accompanied by tenderness to light touch. They may be acute or chronic, and an exact cause is seldom known. To make the diagnosis, all other causes of the pain should be ruled out. We must emphasize here again that when unexplained ear pain is a symptom, *malignant tumors elsewhere in the respiratory tract, especially in the pharynx and larynx, may cause referred ear pain*. These should be ruled out by a full ENT examination, if at all suspected. Traditionally, Tegretol is a popular treatment, to be prescribed with care. Trileptal is proving to be a safe and effective replacement. The primary practitioner can diagnose and treat these disorders, with referral to an ENT specialist or neurologist if there is any doubt.

Hematoma of the Auricle

This problem occurs most often in young athletes, particularly wrestlers or football players who practice without their headgear. However, any severe blunt trauma to the auricle can cause a hematoma at any age. The vascular anastamoses of the auricle make subperichondrial accumulation of blood, with recurrences and lack of reabsorption, a likelihood. Usually, the hematoma occurs on the superolateral surface, centered over the scapha and upper concha (Fig. 4.**1**). If left untreated, fibrosis and even calcification can develop in time, causing the classic "cauliflower ear" deformity.

Incision and drainage should be done aseptically to avoid the dreaded complication of perichondritis, which will be discussed next. Antipseudomonal antibiotics should be prescribed. Evacuation of the hematoma may be carried out by making an incision, or two parallel incisions, and inserting a rubber drain. Pressure dressings are applied and the drain removed several days later. A follow-up visit to rule out recurrence should also be made.

Another method of treatment is aseptic **needle aspiration**, using an 18-gauge needle after numbing the skin with a tiny-needle lidocaine injection. After aspiration, a cotton wad soaked with collodion is form-fitted over the area and held in place until it dries. Over it, an additional small

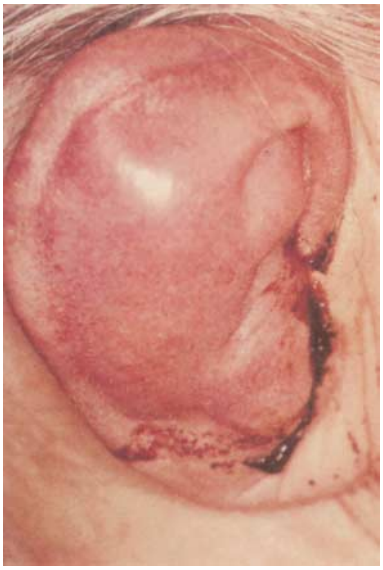


Fig. 4.**1** Hematoma of the auricle. (Source: Becker W, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994)

pressure dressing is taped into place on the auricle. The patient is instructed to hold it there firmly for 20 minutes or so and to keep the dressing on for a few days. With either technique, there may be recurrences that need repeat drainage procedures, but success usually eventuates with careful treatment, observation, and avoidance of the activity that brought on the hematoma.

The primary practitioner or emergency physician, if comfortable, may perform the second of these two procedures on an initial encounter, but ENT follow-up is recommended as recurrences are likely.

Perichondritis of the Auricle

This devastating infection occurs most often as a result of trauma, with penetration of the skin and a contaminated wound. Another possible cause is iatrogenic injury, i.e., ear surgery. The auricle becomes hot, red, swollen, and tender after the contaminating injury (Fig. 4.**2**).

When perichondritis is suspected, aggressive treatment is necessary. The organism is usually *Pseudomonas aeruginosa*, although *Staphylococcus aureus* may be involved. If there is evidence of fluctuance from pus, drainage should be carried out, of course with a culture. Appropriate antibiotics (antipseudomonal, if not cultured otherwise) should be administered,

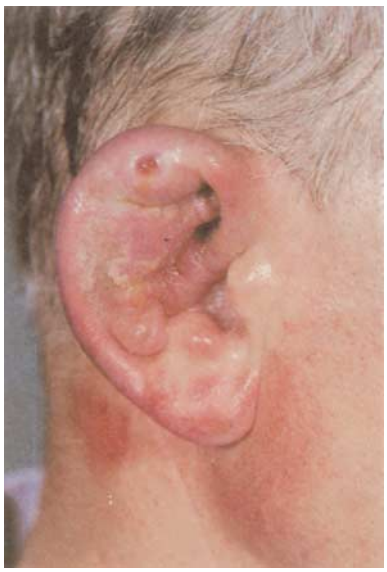


Fig. 4.**2** Acute perichondritis of the auricle.
(Source: Becker W, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994)

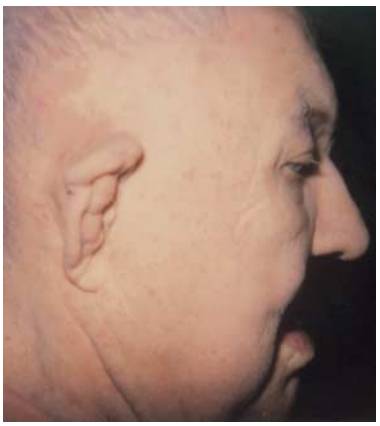


Fig. 4.**3** Poor end result after treatment of perichondritis of the auricle.

perhaps even intravenously in the hospital, with close observation and warm moist dressings. The quinolones, as well as the aminoglycosides, such as tobramycin, are effective against *Pseudomonas* and staph. A severe infection, which begins and stays localized under the perichondrium, often results in necrosis of the cartilage and eventual fibrosis with a permanent severe auricular deformity (Fig. 4.**3**).

A condition simulating perichondritis can be encountered—the allergic insect bite reaction. Typically, the patient is seen during the summer months with rapid onset of a swollen, warm, itching, pink auricle. The culprits are often gnats, and an obvious insect bite is not necessarily seen. The absence of gross skin injury and predominance of itching, rather than pain, favors this diagnosis rather than perichondritis. In these cases, antihistamines and topical steroids are the indicated treatments. If there is any doubt about the diagnosis, precautionary antipseudomonal antibiotics should be used.

If the primary physician suspects full-blown perichondritis with its characterisitc red, swollen, tender auricle, hospitalization for IV antibiotics and early ENT consultation are indicated.

Congenital Disorders of the External Ear

Microtia

The scope of this text will not include embryology nor a detailed discussion of all of the types of external ear deformities that can occur. The most severe deformity is **microtia**, which is immediately noticeable at birth.

Here, there is nothing more than a few deformed nubbins of tissue where the auricle should be. Less severe deformities of the auricle may also be encountered. These external defects are often accompanied by **stenosis** or **atresia** of the external canal, as well as by middle ear anomalies.Occasionally, they accompany syndromes with other craniofacial defects. With complete occlusion of the canal, the conductive hearing loss is very large, on the order of 50–60 dB HL.

Sometimes the other ear is normal and there is no urgency for treatment of the deformed ear, although a hearing aid should be placed at a young age for the sake of good bilateral hearing. A child with deformities of *both ears* should have a **hearing aid** placed as soon after birth as possible to gain speech input. The specialist should get involved early. CT scans will show the extent of the defect, and surgery can be done on the more correctable ear before the school years. The procedure is extremely specialized, with risk for complications and failure. Regarding the cosmetic deformity of the auricle, multiple procedures, or simply a prosthesis, may be indicated. Obviously, referral to a "super-specialist," who deals with such cases often, is recommended.

Preauricular Cyst

A less severe congenital condition, the **preauricular cyst** and/or sinus tract, may occur just anterior to a normally formed external ear (Fig. 4.**4**). This

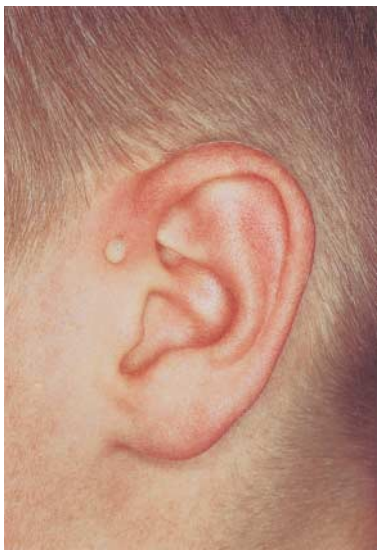


Fig. 4.**4** Infected preauricular cyst. (Source: Hughes GB, Pensak ML. Clinical Otology. New York: Thieme; 1997)

usually presents as a small fistula in the skin anterior to the helix at the upper tragus. A number of people have only a punctum here as an embryonic remnant with no clinical problems. However, the associated sinus tract can develop a dilated cyst with repeated infection and abscess formation. An acute abscess should be treated by **drainage** through an incision as close to the punctum as possible. In problem cases, **surgical excision,** with complete removal of the tract, is the answer. Care must be taken to avoid the upper branches of the facial nerve.

First Branchial Cleft Cyst

These cysts occur just beneath the lobule of the auricle and may be mistaken for parotid gland tumors. They often have a sinus tract and tiny fistula that empties into the floor of the EAC. Thus, when infected, they may present with swelling below the ear and drainage into the canal. Even though these are "benign cysts," a skilled specialist should do the surgery. The tract is closely associated with the facial nerve, which may be injured as a surgical complication.

Noncongenital Cysts and Keloids of the Auricle

Epidermal Cysts

Two cystic conditions may be encountered externally, each in a different location. **Epidermal inclusion cysts**, traditionally known as *sebaceous cysts*, are usually located low in the postauricular crease. They represent backed-up oil glands and occur in individuals with oily skin and acne. These patients tend to have them behind both auricles and in other facial areas as well. Their usual content is cheesy sebum, but at times they may swell up and abscess, often infected with staph. If they are infected, the treatment is an **antistaphylococcal antibiotic**. Often the infection will resolve, but **incision and drainage** may be necessary. Troublesome recurrences can be surgically excised, when not infected, taking care to remove the entire cyst lining. Even then, they may reappear.

Epidermal cysts of the lobule can occur within the epithelialized tract of an ear-piercing site. They present with swelling, weeping, and repeated infection. If conservative treatment with antibiotics and cleansing fails, **surgical excision**, with removal of the entire epithelialized lining, may be necessary. The defects in the lateral and medial skin of the lobe are sutured and the patient is doomed to use conventional earrings here and find another site for body piercing.

Keloids

At the same piercing site on the lobe, **keloids** (nodular hypertrophic scars) may develop in keloid-prone individuals, who are usually dark-skinned. These can be an extremely difficult problem, with growth to incredible size. They can recur even when excised completely. **Triple therapy**—excision, postoperative steroid injections, and irradiation—may be necessary.

Skin Disorders of the External Meatus

The three major "dermatoses" of the external ear are **seborrheic dermatitis**, **eczema**, and **psoriasis**. They have some overlapping characteristics and often affect the same areas, namely, the external canal, its meatus, and the concha. Sometimes adjacent regions, such as the lobule and postauricular areas, are affected. They seldom extend deeper than the outer one-third of the canal. Dermatologists refer to all three as the *papulosquamous* disorders. Patients afflicted with these disorders complain of itching and weeping of the external canal. Occasionally, there is pain if inflammation or superinfection is present.

Seborrheic Dermatitis

This is the most prevalent of the three dermatoses affecting the external ear. It presents as a diffuse scaliness, with a pink or orange discoloration of the skin, in and around the external meatus. Often the involved skin is greasy, but other times it is simply dry and flaky. The lesion may be seen behind the auricle as well, along with other locations on the face, especially on the forehead between the eyes and lateral to the nose. It occurs more often in the older adult population. Dandruff (seborrheic dermatitis of the scalp) often accompanies it. Treatment centers around mild **topical steroids**, as well as **selenium sulfide shampoo**. When the latter is applied to the scalp for the dandruff, it may be applied to the ears as well. Sometimes yeast accompanies it, which responds to topical **ketoconazole** cream or shampoo.

Eczematoid Dermatitis

Eczema of the meatus and surrounding structures may affect any age group. It may be "familial atopic dermatitis," "acquired-irritant," or "allergic" eczema. The lesions usually start as small blisters, which itch intensely and are scratched away, leaving skin that becomes "lichenified" with exaggerated striations and scales. Weeping of sticky clear fluid is often present. **Allergy** to topical irritants, such as fabrics, soap, hair coloring, or hair spray, as well as other environmental allergens, may be causative. Neomycin allergy from eardrops can cause an acute eczema (Fig. 4.**5**), aggravating

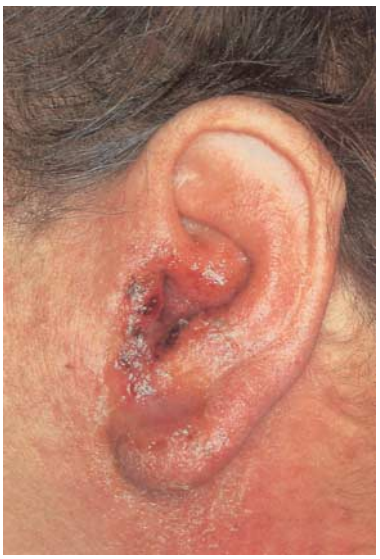


Fig. 4.**5** Eczematoid neomycin reaction.
(Source: Hughes GB, Pensak ML. Clinical Otology. New York: Thieme; 1997)

a condition you may be already treating! In addition, food allergy has been implicated—eggs, milk, cheese, chocolate, and nuts head the list.

Unfortunately, many times there is no identifiable allergen. Bacterial superinfection, especially with staph**,** may complicate the picture. A culture will help if infection is suspected. Treatment also hinges on topical **steroids**, oral or topical **antihistamines** for the itching, antibiotics if indicated by culture, and of course, avoidance of the allergen, if known.

Psoriasis

This affliction of the external ear has some similarities to seborrheic dermatitis but tends to be more localized and patchy. It is also thicker, with a superficial white "micaceous" scale. Underneath, the skin is often deep red and tends to bleed if the scales are peeled off. Patients with psoriasis of the ears usually have the lesions elsewhere and have probably already been diagnosed. The other favorite body sites for the lesions are the extensor surfaces of the extremities. **Warm water soaks**, which soften the scales, are an easy remedy for accessible lesions, as well as the topical steroids or **vitamin D ointment**. Dermatologists are more qualified to treat this and should be involved.

Summary
The dermatoses of the external ear usually present with itching and weeping. Topical steroids are a mainstay of treatment in these scaly skin disorders. Dermatologists often advise the use of *weak* steroid preparations to avoid thinning or ulceration of the skin. A culture for both bacteria and fungus might grow out a pathogen, which may be treated. The primary practitioner can recognize and treat these disorders. ENT or dermatology referral can be made for persistent cases, especially if psoriasis is suspected.

Otitis Externa

We will now discuss several forms of external otitis. At this point, it is important to introduce two key clinical concepts. The first is that with an acute earache the presence of *tenderness* helps to distinguish between otitis externa and otitis media. If you are called on the phone at an inconvenient time by a mother whose child is screaming with an earache, ask her to pull backward on the auricle or press on the tragus. This will hurt if the problem is external, but will not if only the middle ear is infected. At least you can get a feel for the cause, and perhaps the treatment.

This leads to the second point. The bacterial organisms causing *Otitis externa* are usually *Pseudomonas*, *Staphylococcus*, *Proteus*, *Enterobacter*, or other Gram-negatives. On the other hand, the bacterial offenders for *acute otitis media* are usually *Pneumococcus*, *Haemophilus influenza*, and *Moraxella catarrhalis*, the ones that are often seen in acute sinusitis or other bacterial respiratory infections. This generalization is quite reliable and implies a different treatment for each entity.

Acute Diffuse Otitis Externa

This condition, a well-known painful infection of the canal, is otherwise known as swimmer's ear. Water immersion is not always the cause, but the disease occurs most often in warm, humid conditions. Moisture in the ear, even from perspiration, plays a causative role. Local trauma to the canal is also a precipitating factor. Abrading a wet, macerated canal with a cotton swab to clean it or scratch an itch is often the initiating insult, implanting bacteria under the epithelium. The darkness of the canal, its warmth, high pH, and moisture all promote microbial growth. *Pseudomonas* causes this acute infection almost exclusively, although staph and others may rarely be involved. The bacteria go on to infiltrate, growing beneath the epithelium; then more itching ensues, progressing to soreness.

In the full-blown florid stage, the patient presents with a swollen, draining, tender canal. Touching the tragus or pulling the auricle backward elicits severe pain. Swelling narrows the lumen of the canal, sometimes to a pinpoint. In addition, the infection may spread through the fissures in the anterior cartilage to the parotid gland and adjacent skin, causing parotid cellulitis and localized adenopathy.

Treatment of this condition has traditionally centered on topical therapy with eardrops, namely, combinations of **neomycin, polymyxin, and hydrocortisone** (Cortisporin, or its generic substitute). In recent years, additional tools have become available. Often, the canal is so swollen shut that drops will not penetrate. The new **Pope ear wicks** are easily inserted without too much trouble and are then soaked with the topical preparation. They soften and expand when moistened and stay in place so that the medication can work "around the clock." The drops are applied several times a day, and the wick may be removed a few days later.

Quinolone eardrop preparations containing Floxin or Cipro have now become available, although the traditional Cortisporin is still effective. In addition, the *Pseudomonas*-killing quinolones may be administered orally in severe cases if the patient is old enough. **Analgesics** should not be forgotten—this infection, when severe, ranks with kidney stones and acute gout for pain intensity. The patient should keep the head elevated at home and expect two or three more days of hard times, even with good treatment.

Anecdotally, I have seen a number of patients in the past with this disease who had been treated for days with only oral amoxicillin or cephalosporins. It should be emphasized that *Pseudomonas* is the vastly predominating pathogen and that it will not respond to these antibiotics.

Acute Localized Otitis Externa

This disorder, a different disease from the diffuse type, also presents with a very painful ear. It is otherwise known as a **furuncle** of the canal. The infection is localized in an obstructed sebaceous gland or hair follicle out near the meatus. A tender red, raised pustule is readily seen occluding the meatus (Fig. 4.**6**). *Staphylococcus aureus* is the usual offender here, and appropriate **oral antibiotics** such as cephalosporins or amoxicillin/clavulanate, as well as topical neomycin preparations, are indicated. When bulging and soft, incision and drainage at the most fluctuant point with a #11 blade will benefit.

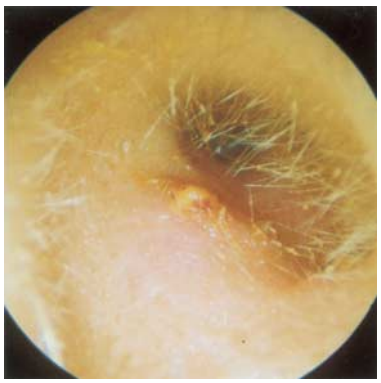


Fig. 4.**6** Furuncle of the external meatus.
(Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

Malignant Otitis Externa

Malignant otitis externa is also known as *necrotizing otitis externa* or *skull base osteomyelitis* in its full-blown form. Obviously, the second name is the gentlest one, but the other two imply its ominous characteristics. This type of infection is typically seen in elderly diabetics or immunocompromised patients. It can spread from the external canal to cause osteomyelitis in the temporal bone with potentially fatal complications. Characterisically, the patient presents with an external earache similar to other forms of external otitis. However, examination of the ear shows something different. The canal may be swollen and tender, but a small area of red granulation tissue is seen posteroinferiorly in the canal at the junction of cartilage with bone, one-third inward. This finding, plus the type of patient described, points to the diagnosis.

When suspected, ENT consultation for aggressive treatment should ensue. The organism is almost always *Pseudomonas*, but a culture should be done. Biopsy of the granulation can rule out neoplasm. Treatments include appropriate **topical** and **systemic antibiotics** and aggressive debridement. Medical management of the diabetes and/or immunodeficiency can also improve the prognosis. Conventional CT scans and bone scans can image the areas of involvement. Even with appropriate aggressive treatment, devastating bone infection and death can occur.

Summary of Acute External Infections
Acute diffuse otitis externa ("swimmer's ear") is much more common than the localized furuncle, and should be recognized by its diffuse swelling and tenderness. Some clinicians err in prescribing oral antibiotics that will not work on *Pseudomonas*, the vastly predominating organism. Topical antibiotic drops, with wick insertion if the canal is swollen shut, are the mainstay of treatment. Oral quinolones, if the patient is old enough, may be used in severe cases. A *culture* should be done if there is any doubt about what you are dealing with, and pain medication should not be forgotten. An obvious localized *furuncle* has staph as its cause. Treatment with topical drops and antistaphylococcal oral antibiotics is indicated.
The primary practitioner can diagnose and treat these infections, even with the insertion of a wick if the canal is swollen. However, granulation tissue in the canal of an elderly or diabetic patient portends *malignant otitis externa* and warrants early ENT consultation.

Mycotic Otitis Externa

This disorder is also known as *fungal otitis externa* or *chronic diffuse otitis externa*. It differs from the previously mentioned infections in that it is not quite so painful, but more indolent, yet persistent. The usual complaints are of blockage, thick drainage, dull pain, and itching. These infections occur more often than most clinicians expect, and they are often treated inappropriately with antibiotic drops. The most notable finding on ear examination is the presence of "matter"—thick moist debris—in the canal (Fig. 4.**7**).

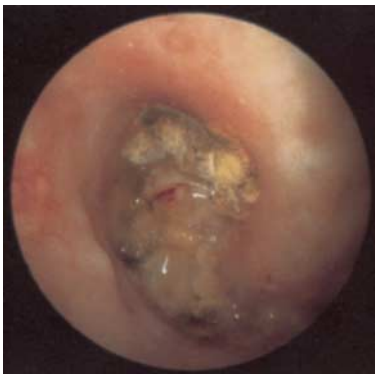


Fig. 4.**7** Fungal otitis externa. (Source: Hughes GB, Pensak ML. Clinical Otology. New York: Thieme; 1977)

Aspergillus niger is the most common infection, and its exudate appears as part black, part light-gray "wet blotter paper" in the deeper portion of the canal, even against the TM. Other species of *Aspergillus* appear tan or yellow-orange and also locate themselves deeply. *Aspergillus* is more apt to cause pain than itching. When the canal is cleared of the exudate, the underlying skin is red and raw. This organism is a mold that may be picked up from the environment, wherever molds may grow.

Candida albicans and other *Candida* species are also frequent pathogens. Their exudates tend to be flocculent and white or creamy in appearance, and itching is a notable complaint, in addition to blockage. It is often seen in antibiotic-treated or immunocompromised (including diabetic) individuals. Other fungi, such as actimomyces and phycomycetes, may also rarely occur.

One diagnostic point—the fungi mentioned have a mild musty odor, or none at all, whereas *Pseudomonas* has a characteristic sweet, musty smell, and *Staph. aureus* and *Proteus* are downright putrid. In any external infection, a culture for both bacteria and fungi should be done if there is doubt about the organism. Sometimes bacteria and fungi coexist, especially *Pseudomonas* with *Aspergillus*.

Treatment of these fungal infections keys on **complete removal**, so that no spores are present for regrowth. If there is no perforation of the TM, gentle irrigation and suction may be the best way to clear the canal. (Caution: irrigating an infected ear is not usually recommended, but with fungal infections it has not caused problems in my experience, as long as the canal is suctioned dry.) The ear can then be insufflated with a large amount of **nystatin** (Mycostatin) powder, which comes in containers that facilitate this. **Clotrimazole** (Lotrimin) drops are another recommended therapy. Antibiotics or steroids do not help and may even promote growth, especially with *Candida*. In fact, fungal otitis may be a complication of overtreatment with Cortisporin-type preparations. Oral antifungals, such as **fluconazole** (Diflucan) can be considered in refractory cases.

Summary
Mycotic otitis externa, synonymous with *chronic diffuse otitis externa*, is characterized by milder pain than the acute infections. Patients complain of chronic moisture, blockage, thick drainage, itching, and mild discomfort. A characteristic finding is the presence of thick matter in the canal, without severe swelling or tenderness. *Aspergillus* and *Candida* are the usual culprits, but a *culture for all organisms* should be done if there is any doubt. Successful treatment hinges on *complete removal* of the debris, followed by the topicals mentioned in the text.
The primary practitioner can make the diagnosis, but usually referral to ENT is needed for thorough cleansing. Even then, there is a tendency for recurrence and persistence, and repeat cleanings and topical applications may be necessary. Oral systemic antifungals, such as Diflucan, might also be considered in refractory cases.

Other Chronic External Ear Disorders

Some patients suffer from another disorder, **chronic stenosing otitis externa.** These individuals have repeated infections; sometimes cultures are positive for bacteria or fungi, and sometimes there is no identifiable pathogen. The dermatoses may be involved. The external canal itches, drains repeatedly, and becomes chronically swollen, with progressively severe narrowing of the lumen. This problem responds temporarily to office cleansing and wick/eardrop insertions, but it is often relentless. Severe cases may eventually need surgery to widen the canal. Canalplasty with skin grafting, or even limited mastoidectomy, can be performed to open the canal and regain the hearing.

Other individuals have problems with ongoing or **recurrent acute otitis externa** without the complication of stenosis. These patients often create their own problems and should be cautioned regarding the cause and prevention of external otitis. Many individuals feel the need to dowse their ears daily with water in the shower, and then vigorously clean with applicators. The old adage about "nothing smaller in the ear than your elbow" is not bad advice. Gentle removal of cosmetically visible cerumen in the meatus is all that should be done. In the population at large, the vast majority of ears are self-cleaning—only 5% or so have problematic cerumen buildups.

Prophylaxis for external otitis in swimmers, however, is a valid consideration. Insertion of alcohol/acetic acid drops before or after swimming may prevent infection. In fact, an effective, nontoxic, "all-purpose" remedy for all forms of external otitis is the **propylene glycol/acetic acid eardrop** (e.g., Vosol). The desired effect of drying and lowering pH can also be com-

bined with steroids, if indicated, in certain preparations (e.g., Vosol HC). The steroids are effective for itching, but may aggravate fungal infections.

Cerumen Accumulations and Keratoses

Cerumen exists for several reasons. It protects the skin of the EAC from water penetration; its low pH discourages microbial growth; and it traps foreign material, carrying it outward by migration. It is produced in the outer third of the canal, where the gland/hair units are located.

In normal individuals, the skin of the entire canal migrates very slowly and steadily from the inside outward. Studies with ink dots have shown that epithelial migration actually starts near the center of the TM and proceeds all the way out, at a rate of about 2 mm a month! In the outer third, cerumen migrates together with the epithelium and eventually sloughs, carrying foreign material with it.

In individuals with repeated cerumen impactions (about 5% of the population), analysis of the cerumen shows high amounts of keratin. Apparently, migration of the skin and its sloughing pattern are abnormal—the epithelium and cerumen tend to roll up in a ball. This is readily appreciated when cleaning ears in the office—many times one will see desquamating segments of epithelium that still cling to the midcanal after most of the cerumen has been removed.

Irrigation, as described in Chapter 2, is the simplest way for most clinicians to clean problem ears. Wax softeners may help the process. Sometimes, suction or curetting is needed to remove troublesome buildups and/or epithelial debris, and this is where specialized equipment and skills come into play.

Patients with recurrent accumulations often ask what they can do to clean their own ears and avoid periodic office visits (which may be needed as much as two or three times a year in some individuals). First, these patients should be advised not to use cotton swabs in their ears, as they will often aggravate a buildup by blindly packing it in. Wax softeners like Debrox or Ceruminex are often helpful for the impaction-prone individual, to be used in each ear once or twice a week. Self-irrigating kits are available at pharmacies as well. These aids may help some patients, but failure of removal or infection may be complications.

A rarer buildup in the ear is **keratosis obturans**. This is a mass of squamous epithelium accumulating in large whorls that are difficult to remove. It can erode through the skin of the bony canal and then erode bone itself, causing pain and draining infections. It is tenacious, and removal often requires the "headlight and two-hands" approach, using hooks, curettes, and alligator forceps. Thus, an ENT referral might be necessary. Individuals

with this problem should be seen at frequent intervals, perhaps every six months, for cleaning. Chronic, untreated cases of this disorder may show up with huge excavations into the bone of the canal wall, usually inferiorly or posteriorly.

A related entity is **cholesteatoma of the external canal**. This entity differs in that the epithelial accumulation tends to be deeper, near the TM. It is seen more often in older individuals and is usually unilateral.

Discussing cerumen problems raises an important point. Occasionally a clinician will see a patient (usually a child) with a fever and an earache and will not be able to see the drum due to cerumen or other debris. If this cannot be removed at the time, it is reasonable to go ahead and treat empirically for an ear infection. However, if the problem recurs, and the blockage cannot be removed, ENT referral is indicated.

Trauma and Foreign Bodies

Abrasions and Lacerations of the External Canal

This type of trauma to the external canal is most often self-inflicted by an individual with a cotton applicator or a hairpin while attempting to remove wax. The patient may feel pain or notice blood welling up, and thus present to the office. The examiner sees an ear full of blood and the question arises, "Is there a perforation of the drum?" Suctioning out the blood is the only way to get a good look, but often it is too copious and tenaciously clotted, even for a specialist with good equipment. In these cases, it is best to leave things alone and treat the patient for a week with antibiotic drops to prevent infection and loosen the blood. Avoidance of water in the ear should be advised.

A follow-up visit will then give more information. Most of the time, only an abrasion or laceration of the canal is present—these bleed very easily, but usually heal with no complications. It is important to see the patient in follow-up until things are completely healed, however. The cotton-tip swab may have abraded a small carcinoma of the canal to cause the bleeding! If a perforation has occured, continued water avoidance and initial observation are indicated. Perforations are discussed more fully in Chapter 5.

Incidentally, this raises the question of whether neomycin-polymyxin eardrops should be used if a tympanic perforation is present. Much has been said in recent years about their potential ototoxicity. Be advised that most otolaryngologists have been using these drops routinely for years, even in middle ear surgery. Less than a handful of cases of ototoxicity (assuming entry of the drops into the inner ear via a hole in the oval or

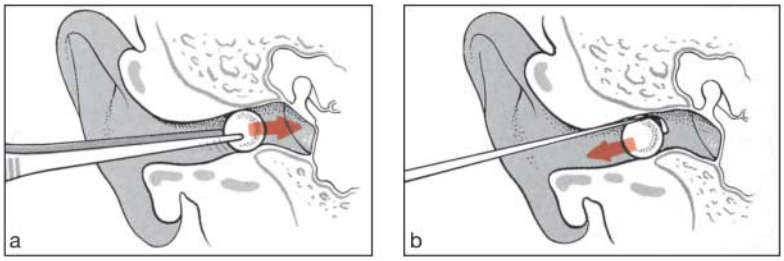
round window) have been reported in literature. However, now that other nontoxic antipseudomonal drops such as Cipro and Floxin are available, these can be used to play it safe, keeping in mind the much greater cost.

A bloody traumatized canal can be initially evaluated and treated with antibiotic drops by the primary physician. ENT follow-up within a few days would be appropriate.

Foreign Bodies

Objects lodged in the canal occur most often in young children, mentally handicapped adults, or temporarily mentally handicapped adults. If the foreign bodies are soft, such as the end of a cotton swab or furniture stuffing, removal with alligator forceps is usually not difficult. More often, however, they are solid round or oval objects, such as beads. These are more difficult. Do not attempt instrumental removal unless you feel you can "get around behind" the object to pull it out (Figs. 4.**8a, b**). Many times, a well-meaning attempt with a suction tip or forceps will lodge the object deeper in, beyond the bony hump of the anterior canal wall. A general anesthetic may then be necessary for removal, especially with an uncooperative young child. With cooperative patients, the best instrument for removal of solid objects, is the Day hook, a straight, thin, metal probe with a right-angle turn of about 2 mm on the end. The end is inserted just past the object and turned 90° to retrieve it. Also, irrigation, as with cerumen impactions, may be successful in some cases.

Special mention must be made of insects. Live ones in the ear can be terrifying to the patient because the fluttering and crawling is relatively loud, and skin of the bony canal is very sensitive to light touch. Mineral oil, wax softener, or dish detergent, all being mild and viscous, can be in-



Figs. 4.**8a,** b Removal of rigid foreign body from the external canal. An attempt to remove a foreign body with simple forceps (**a**) usually displaces the object deeper and may cause middle ear damage. Using a hook (**b**) is the effective way.
(Source: Becker W, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994)

stilled in the canal to kill the insect quickly. Then, removal can be done with alligator forceps or possibly irrigation. A few days of antibiotic drops are then a good idea.

Primary and emergency physicians should exercise great discretion when removing foreign bodies. If there is any doubt about one's ability to extract a foreign body, an ENT specialist should be consulted. The inert ones may wait a day or two. Live insects should be killed immediately, as described above.

Auricular Trauma

Sharp and blunt trauma to the external ear might result in contusions, hematomas, lacerations, or even disruption of the cartilaginous framework. Appropriate surgical intervention is certainly indicated, and antibiotic coverage for *Pseudomonas* and staph is recommended to prevent perichondritis. Hematoma and perichondritis have been discussed earlier in this chapter.

Temporal Bone Fracture

Also termed basal skull fracture, this injury occurs with severe head trauma and characteristically presents with blood oozing from the external meatus. Hearing loss, vertigo, and facial paralysis may be present. Patients with this type of trauma are usually severely injured in other ways as well, and are hospitalized by the neurosurgeon after presenting to the emergency room. Occasionally however, a stoic individual will not seek acute care. Sometimes, a cerebrospinal fluid (CSF) leak is present, with steady dripping of clear fluid from the ear.

Initial treatment is bed rest, with the head elevated, and observation. Prophylactic antibiotics should be given, even if there is no apparent CSF leak. CT imaging will identify the fracture. Two types—longitudinal and transverse fractures—may occur, each with a different pattern of damage. (A more detailed discussion can be found elsewhere.) Permanent hearing and vestibular damage can result. Facial paralysis, if immediate and not delayed, warrants surgical exploration with decompression and/or repair.

Frostbite of the Auricle

This type of damage from prolonged exposure to cold is likely to affect the ear first. The auricle is quite vulnerable due to its exposed location, superficial blood supply near the skin's surface, and lack of sensitivity. At first, the involved skin is pale and numb. As warming occurs, the affected areas become hyperemic and painful and may even blister.

Treatment should be with **gradual warming** in a cool room. Direct heat, massage, or snow application is not recommended, as it will simply aggravate tissue trauma. "Hands-off" observation is the rule, although prophylactic antipseudomonal antibiotics are indicated for severe injuries. Eventual necrosis and loss of tissue may occur, but even then, delineation by sloughing gives a better result than premature surgery, unless gross infection is present.

Regarding the last three categories of trauma, the physician involved depends on the severity of the injury. Obviously, temporal bone fractures and severe frostbite require ENT consultation in a timely fashion, whereas minor auricular trauma can be repaired by the capable family or ER physician, taking care to prescribe antibiotics to prevent perichondritis.

Tumors of the External Ear

Bony Tumors

The most frequently seen tumors of the EAC, at least in northern climates, are the bony ones—exostoses and osteomas. **Exostoses** are sessile rounded bony projections in the inner two-thirds, often seen on the floor of the canal anteriorly and posteriorly, although they may be based superiorly as well. The inferior ones tend to be more external, flatter, and broader, whereas the superior ones are often deeper, smaller, and more rounded (Fig. 4.**9**). Their epithelial covering is normally smooth and unremarkable in appearance. The cause is usually cold-water swimming over several years.

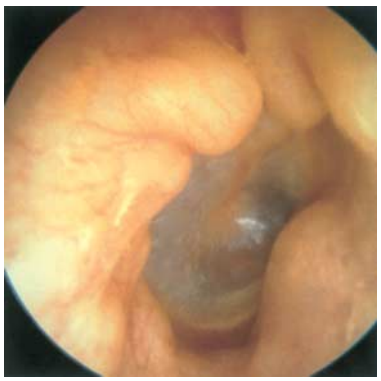


Fig. 4.**9** Multiple exostoses of the external canal.
(Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

Many individuals are not aware of their exostoses, and one can impress these patients by taking a look in their ears and discussing their swimming history. Very gentle palpation with an instrument will confirm that these projections are in fact bony and not soft-tissue lesions. Often they present no problem, but larger ones may trap cerumen or debris and cause chronic infection or hearing loss.

Osteomas are histologically different from exostoses and appear more completely rounded—almost pedunculated. Either entity can be surgically removed if there are problems with obstruction, but the surgery itself can lead to complications. Facial nerve injury has resulted in seemingly simple cases.

Squamous Papillomas

These lesions are not extremely rare. Usually occurring in ears with recurrent external otitis, they are often located at or near the meatus and resemble warts seen elsewhere. They are associated with human papillomavirus (HPV) and tend to recur when excised. Cauterization of the base, after excision, may improve the chance for success.

Other Neoplasms

Numerous other tumors can arise on the auricle or in the canal and they are mentioned only briefly. The point to make is that any questionable lesion deserves a biopsy. A benign-appearing granulation in the canal may actually be a squamous cell carcinoma. The auricle, with its sun-exposed surface, can be the site of basal cell carcinomas, squamous cell carcinomas, and malignant melanomas. These malignant epidermal tumors, along with adenoid cystic carcinoma and rhabdomyosarcomas, can occur inside the canal, although much less frequently than on the auricle. Angiomas, ceruminomas, adenomas, nevi, and myxomas are rare tumors of the external ear. The temporal bone may be the site of fibrous dysplasia or eosinophilic granuloma (histiocytosis X).

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5 Disorders of the Middle Ear

This chapter discusses diseases of the middle ear, also known as the tympanic cavity. Its lateral boundary is the tympanic membrane (TM), and its medial bony wall, containing the oval and round windows, separates it from the inner ear. Earlier, we spoke of the ear as having separate "compartments" with separate problems. Disorders of the middle ear may cause symptoms of pain, pressure, decreased hearing with autophony, certain types of tinnitus, and drainage (if there is a TM perforation). Occasionally, deep itching can be experienced secondary to allergic tubotympanitis. Our discussion will begin with a most vital appendage of the middle ear cavity, the eustachian tube. Diseases involving other adjoining structures—the epitympanum and mastoid—will also be discussed in this chapter.

The Eustachian Tube

The eustachian tube ensures that air pressure inside the drum is equalized with atmospheric pressure lateral to the drum. Unfortunately, in combination with the cumbersome middle ear and related structures, it is the source of many problems. If we were fish, we would need only an external membrane over an inner ear. Evolution of life in the earth's atmosphere has mandated the development of the drum, the ossicles, and a mucosa-lined middle ear cavity. Thus, the eustachian tube exists—an essential conduit for equalizing pressure and clearing secretions.

This tube exits the middle ear cavity to course inward toward the nasopharynx. It is lined with respiratory mucosa and contains numerous mucous glands near the middle ear. The proximal one-third is surrounded by bone, and the distal two-thirds (toward the nasopharynx) by cartilage. The narrowest part of the tube is at the bone–cartilage junction, and the internal carotid artery is just medial to the eustachian tube in this area. (Many years ago, a well-meaning ear surgeon experimentally tried to surgically ream out the tube to improve a patient's middle ear problems. The result was a disastrous carotid artery rupture.)

In the lateral nasopharynx, the tube terminates with a cartilaginous hook, the *torus tubarius*. This remains closed unless it is opened by contraction of the muscles of the palate. In normal individuals, yawning, swal-

lowing, and other motions of the palate open the tube to aerate the middle ear, which otherwise absorbs air at a slow and steady rate.

Before discussing the numerous factors that cause eustachian tube problems, we should note that poor aeration by the eustachian tube is not the *only* cause of middle ear disease. Problems with the mucosal lining of the tympanic cavity itself can cause effusions and infections. Allergic and mucociliary disorders often affect the entire respiratory tract, including this chamber of the ear. Immunoglobulins and leukocytes may be found in thick middle ear fluid, supporting immunologic phenomena as a cause for problems.

However, empirical evidence shows that when the cavity is aerated surgically with a small ventilating tube in the drum (an artificial eustachian tube!), most chronic middle ear effusions reverse themselves, as long as the tube is present and patent. Exceptions exist, such as when acute infections spread from the nasopharynx to cause middle ear suppuration that can drain out the ventilating tube.

This brings us to a discussion of the epidemiology of the numerous eustachian tube and middle ear maladies that can occur. Ear infection and fluid occur more often in early life. Immature eustachian tubes have structural characteristics that predispose to spread of infection as well as poor aeration. These tend to be outgrown in the early school years. In addition, there is a genetic tendency toward eustachian tube and middle ear problems—ear trouble runs in families. Over the years, I have seen a number of parents and offspring who share identical TM retraction pockets in the same unilateral ear!

The above-discussed factors, **youth** and **heredity**, relate to the mechanical structure of the tube. Another major factor is mucosal disease of the eustachian tube. Middle ear problems predominate in **cold weather**. New Yorkers have more problems than Floridians, and Alaskans more than New Yorkers. In the winter, dry heated indoor air, with associated dust and mold, predisposes to eustachian tube obstruction and middle ear disease. A huge causative factor, in addition, is the presence of any **respiratory infection**. Of course, these infections (viruses, sinusitis, adenoiditis, and the like) are seen more often in the cold seasons. **Day care** settings also place children at risk for increased exposure to all microbes.

Respiratory allergens other than dust and mold may cause swelling and abnormal ciliary clearance of the eustachian tube and middle ear. There has been some suggestion that food allergy may play a part, although this is hard to prove, given the complexity of avoidance trials and the labile nature of middle ear effusions. In addition to infection and allergy, the airway irritation of **cigarette smoke** is a proven factor. Numerous studies show that the children of smokers have an increased incidence of middle ear disease.

Finally, aberrant nasal anatomy, such as with deviated septum and structural turbinate deformities, may cause abnormal airflow and predispose to eustachian tube problems. **Cleft palate** may be included here; those afflicted have mechanical impairment of tubal opening. Physical obstructions in the nasopharynx may block the tube. Swollen, infected **adenoids** are a factor in children. **Tumors** here, benign or malignant, might also cause tubal obstruction. Beware of unilateral middle ear fluid occurring in an adult with no record of previous middle ear problems, especially if there is cervical adenopathy on the same side. *Carcinoma of the nasopharynx* could be the cause.

Summary
In perspective, there are three classifications of factors causing eustachian tube malfunction. *Structural abnormalities* of the tube are related to genetics and immaturity, and often improve with age. We have little control over these factors, except to artificially ventilate the middle ear. *Mucosal swelling* of the tube may be caused by infection, allergy, and irritants, which in turn may be aggravated by underlying problems in sinonasal or palate anatomy. Here, we may be able to help, medically or surgically. *Physical blockage* of the tube orifice is the third factor. This may be from large adenoids, or possibly, a tumor in the nasopharynx. Accurate diagnosis and appropriate intervention are indicated here.

Myringitis

The tympanic membrane is the outermost structure of the middle ear and our "window" to middle ear diseases, both past and present. There are two inflammatory disorders that are limited to the drum, and these will be discussed first.

Nonspecific myringitis is a rare disorder of the TM. It tends to be chronic or intermittently recurrent, and is seen more often in warm, humid conditions. Those afflicted complain of moisture and blockage in the ear. The TM shows tiny, weeping, **polypoid granulations** on the surface, with a moist but clear external canal. The drum has good mobility and the hearing is near normal except in cases of heavy secretions or advanced scarring of the drum. The middle ear is clear. Use of **antibiotic–steroid eardrops** is an effective treatment, although the problem may persist and recur. An ear culture might provide further useful information. Chronic cases can build up large granulations on the drum that lead to dense scarring and significant hearing loss. Surgery may be necessary in some cases.

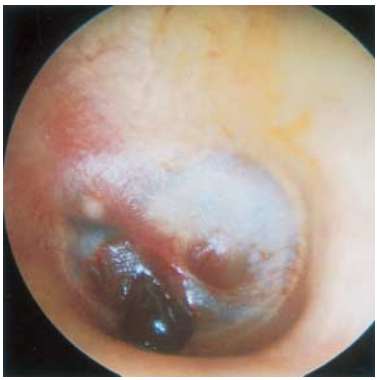

Fig. 5.**1** Bullous myringitis with hemorrhagic blebs, left ear. (Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

Bullous myringitis is an acute disorder that presents with rapid onset of a severe earache. One or more blebs, resembling the bulge of an inner tube through a worn tire, are seen on the drum (Fig. 5.**1**). They may be clear, or red and hemorrhagic. There is great sensitivity when the pneumatic otoscope is used. The disease is a self-limited viral one and thus **analgesics**, both systemic and topical (**antipyrene/benzocaine drops**), are the recommended treatment.

These disorders can be diagnosed, treated, and followed by the primary practitioner who is able to use the pneumatic otoscope. Elective ENT referral can be made for refractory cases.

Tympanic Membrane Perforations

Traumatic Perforations

The TM can be injured in several different ways. The scenario involving a person cleaning the ear with an applicator or hairpin, which we described in the last chapter, is the first way—a direct, **penetrating injury**. Frequently, a second person inadvertently hits the elbow of our unfortunate ear hygienist to cause the injury. Another cause is an **implosion** of the drum by a striking force, such as a slap or fist to the ear. This type of perforation is usually anterior and inferior. An abusive family member may be involved, and sometimes, pathetically, the victim will try to hide any detail of the incident when presenting to the office. Diving and water skiing accidents may also implode the drum. Rarely, a forceful **explosion** near the ear can also implode the drum, usually causing acoustic damage to the inner ear as well. Finally, a hot **slag particle**, as with welding, may

penetrate the TM, cauterizing the edges as it goes through into the middle ear. In this case, spontaneous healing is less likely and recurrent infection and drainage may ensue.

Traumatic perforations vary in their size and location. Some may be difficult to see on examination. They may be small and hidden behind exudates or blood clots or may also be obscured by the bony hump of the anterior canal wall. If the examiner can see part of the drum, the pneumatic otoscope, with an adequate air seal in the canal, is the key to diagnosis. A totally immobile TM will be seen with any perforation. (An extremely scarred TM or glue ear may also show immobility). Conversely, if the drum is mobile, there is no perforation.

In all traumatic perforations, middle ear **ossicle damage**, even with oval or round window rupture, may occur. Look for inordinately large hearing loss (>35 dB HL) or the presence of vertigo as a clue. The Weber and Rinne tests are helpful here. Most traumatic perforations (probably 90%) heal spontaneously. **Avoidance of water** and **observation** are the only initial treatments needed. **Topical antibiotic eardrops** may be indicated if drainage and infection are present. Very large traumatic perforations and those from slag are less likely to heal. These will require surgery if they show no signs of closing after observation for a few months.

Perforations from Acute Infections

The most frequently occurring perforations are, thankfully, the most short-lived. These are the ones resulting from **acute otitis media**. Here, the TM is so red, wet, and distorted that the small opening is not always seen. Almost all of these heal within days, assuming that antibiotics are given. An exception occurs with the rare, aggressive**, acute necrotizing otitis media**. This is usually caused by **beta streptococcus** in conjunction with a severe viral infection like measles. In other countries, scarlet fever is still a cause. In these cases, a large permanent perforation is created. Necrosis of the central TM typically leaves a large horseshoe-shaped hole in the drum surrounding the manubrium. In the pre-antibiotic era this was one of the leading causes of chronic perforations.

Chronic Perforations

Long-standing perforations may be seen in patients who have experienced years of eustachian tube problems and intervening infections. Ventilating tubes may have been inserted repeatedly. The surrounding TM is often thick and scarred (Fig. 5.**2**). Affected individuals have conductive hearing loss and may be plagued with recurrent drainage through the perforation. These episodes of drainage (**otorrhea**) are often initiated by water in the ear or upper respiratory infections.

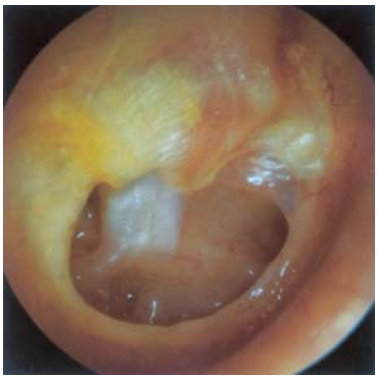

Fig. 5.**2** Chronic otitis media with perforation and sclerosis of upper TM. (Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

The exudates usually culture out the same organisms as seen in external otitis, namely *Pseudomonas*, *Staphylococcus*, *Proteus*, and *Enterobacter*. Incidentally, otorrhea from *any* middle ear infection may initiate external otitis, an exception to our statement that most ear problems involve only one "compartment."

Persistant or recurrent otorrhea through a perforation is known as **chronic suppurative otitis media**. Topical antibiotic/steroid eardrops can clean up the drainage. **Tympanoplasty**, surgical reconstruction of the TM (and eroded ossicles, if needed), may be performed if and when no infection is present. Often there is **chronic mastoiditis** in the adjacent cavity, and **mastoidectomy** may accompany the procedure.

With the emergence of **AIDS** in recent decades, **tuberculous otitis media** deserves mention. This very rare disorder usually starts with painless thickening of the TM followed by **multiple perforations**, with clear discharge. The hearing loss is inordinately large due to inner ear involvement with the bacillus. These findings should alert suspicion, and a positive culture for acid-fast organisms will confirm the diagnosis.

Summary
Regarding perforations in general, the *cause*, as one can ascertain from the patient's history, determines the treatment and prognosis. Those of acute otitis media, if not the streptococcal necrotizing type, will heal, especially if the infection is cleared up with oral antibiotics. In fact, early antibiotics may help the necrotizing ones. Traumatic ones will also heal most of the time. Conservative observation, with water avoidance, is the usual initial treatment. If there is moist or purulent drainage from any perforation, antibiotic/steroid eardrops, preferably following a culture, will help. Large or chronic perforations, and those from slag burns, will probably need surgical repair. Do not forget to evaluate the hearing. A substantial loss (>35 dB HL) may indicate traumatic ossicular damage; this also will need surgical attention. Finally, multiple perforations may indicate tuberculosis, especially in the presence of AIDS.
The primary practitioner may initiate treatment for all the types of perforations discussed so far, although elective ENT follow-up is recommended for all but the responsive acute otitis media. One should note that the perforations discussed so far occur in the "safe" central or anteroinferior part of the TM. There is a "dangerous" area for perforations at the posterior and superior margins of the drum. Here, there is a predisposition for the development of cholesteatoma (discussed later in this chapter), and early ENT referral, within a week, should be made.

Scarring and Tympanosclerosis

A normal TM is often described as "pearly" in appearance. In fact, it is almost transparent. Scarring, which occurs in the middle fibrous layer, destroys this lucency. Scarring may result from repeated effusions or

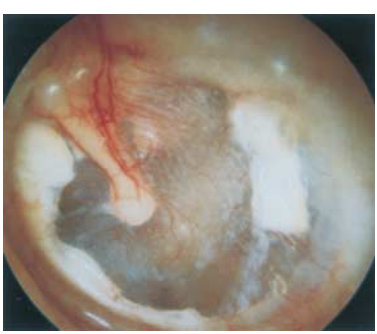

Fig. 5.**3** Peripheral tympanosclerosis with central neomembrane, left ear. (Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

infections, and may have many different patterns. Simple aging often causes a diffusely hazy TM. In patients with a history of earlier ear disease, thickening and opacity may occur in reticular streaks or patches. Incidentally, these changes may be clinically misinterpreted as ear fluid; the pneumatic otoscope and an experienced eye help to distinguish the difference.

In some patients, there may be peripheral scarring with thin central areas of the drum. These thin areas lack the middle fibrous layer and usually represent healed perforations.

They are known as **neomembranes** (Fig. 5.**3**), and might actually be mistaken for a perforation if the pneumatic otoscope is not used.

Tympanosclerosis is a severe form of scarring—a dense white plaque of hyaline substance (again, Fig. 5.**3**), perhaps even thickly calcified. These deposits are located in the middle layer of the drum and probably result from chronic, thick mucoid middle ear effusion ("glue ear") that has finally dried up.

I have repeatedly witnessed a stage in the formation of tympanosclerosis, and briefly publish my findings here for the first time. On three occasions and in three separate patients, years apart in time, tube insertions were planned. At preoperative examination, these children had the findings of shiny, immobile TMs, typical of gluey mucoid effusion, but also showed pinkish, oval plaque-like deposits under the anteroinferior drum. Except for the pinkness, these deposits had the size and location of typical tympanosclerotic plaques. At surgery, I observed the pink deposits and made the myringotomy incisions directly over them. These globules, all roughly 2 mm in diameter, were semisolid and even paler in color than they appeared through the drum. They were removed separately from the remaining thick middle ear mucus and sent to the pathology department The microscopic diagnoses were all reported as "granulation tissue."

Thus, granulation tissue on the inner drum, originating from inspissating mucus, is probably an intermediate stage between glue ear and tympanosclerosis. Surgeons would be advised to suction out as much mucus as possible during tube insertion to prevent this complication.

Summary

It is important for the primary physician to *recognize* scarring and tympanosclerosis. They are sometimes mistaken for fluid, infection, perforation, or even cholesteatoma. Careful examination with the pneumatic otoscope, coupled with experience, will help.

Acute Otitis Media

Acute infections of the middle ear are usually bacterial. Some cultures grow out no organisms and are presumed to be viral. The usual bacterial offenders are *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Moraxella catarrhalis*. Beta streptococcus, staphylococcus, and others may rarely be involved. Often, an upper respiratory infection precedes the ear involvement and spreads up the eustachian tube.

Acute otitis media is classically described in several stages. The first is that of inflammation. Here, the patient complains of earache and fullness, and the drum shows red injection along the most vascular areas—behind the manubrium of the malleus, and at the annular periphery. This proceeds quickly to the stage of exudation, where the middle ear fills with pus. Pain increases, with a decrease in hearing. The reddened drum appears thick, opaque, and bulging.

The stage of suppuration may soon follow, rupturing the drum and spilling bloody exudate into the canal. With the release of middle ear pressure, the pain usually decreases.This whole sequence may occur within a 12-hour period with a virulent infection or be spread out over a few days in milder ones. Of course, early antibiotic therapy alters the course of events. When treated, these acute infections, even after rupture and drainage, usually resolve. The draining perforation heals, except with the rare necrotizing beta streptotococcus infections. Middle ear effusion may persist, however, and require further treatment, to be discussed later in this chapter.

There is some controversy regarding the antibiotics recommended to treat acute otitis media. A number of the organisms mentioned above have developed resistance to the traditional antibiotics. However, in the interest of cost-saving, many sources still recommend **amoxicillin** as the first-line treatment for newly diagnosed, uncomplicated infections. Penicillin-allergic individuals may be given **trimethoprim/sulfa** combinations or the **macrolides** (e.g. Biaxin), although there are more resistant bacteria than to amoxicillin. However, with persistent or frequently recurring cases, **amoxicillin/clavulanate** (Augmentin) offers much broader and more reliable coverage. **Newer generation cephalosporins** (e.g. Ceftin and Vantin) and **clindamycin** are also more effective against resistant bacteria, as is **IM ceftriaxone** (Rocephin).

Acute Mastoiditis

Now let us assume we have a severe acute infection and no antibiotic treatment. This scenario is extremely rare these days. The suppuration in the middle ear and adjacent mastoid cavity may continue. There is usually

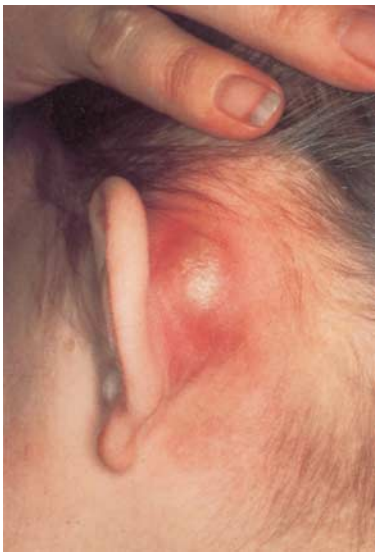

Fig. 5.**4** Mastoiditis with subperiosteal abscess.
(Source: Hughes GB, Pensak MP. Clinical Otology. New York: Thieme; 1997)

little pain because the middle ear continues to decompress through the draining perforation.

Two or three weeks may elapse before the fourth stage of mastoiditis develops. Pockets of pus continue to brew in the middle ear and adjacent mastoid cavity, breaking down the bony septal walls. Physical findings may now include tenderness over the mastoid process, sagging of the posterior external canal wall, persistent drainage through a "nipple" in the TM, and possibly, a subperiostial abscess over the mastoid, bulging behind the ear (Fig. 5.**4**). This bulge usually causes the auricle to protrude abnormally.

This condition is classically termed **acute coalescent mastoiditis** and can be demonstrated radiographically. Incidentally, with any middle ear fluid, even sterile and serous, the mastoid air cells will contain fluid that shows on routine mastoid X-rays or CT scan. Radiologists sometimes over-read any opacification of the mastoid cells as "mastoiditis." The difference with the true disease is the apparent breakdown of bony air cell septa, or "coalescence."

Aggressive treatment is required, with hospitalization and broad-spectrum **IV antibiotics**, based on a **culture** of the secretions. Surgical **mastoidectomy** should be performed. If these measures are not taken, or are unsuccessful, a fifth stage of complication may occur, due to spread of infection beyond the mastoid. Here, the patient may have complications

including meningitis, epidural or brain abscess, and sigmoid sinus thrombosis, all potentially fatal. Look for severe headache, high fever, obtundation, and possibly a stiff neck. One rare complication is Gradenigo's syndrome, caused by localized mastoiditis deep in the cells of the petrous apex of the temporal bone. It is symptomatic with persistent ear discharge, deep eye pain, and diplopia due to paresis of cranial nerve VI.

Rarely, unoperated cases of acute coalescent mastoiditis may resolve with aggressive antibiotics and no surgery, but a sequestrum of chronic osteomyelitis may form in the mastoid cavity. This will cause ongoing purulent drainage and **chronic mastoiditis**, which will also eventually require surgery.

Inflammatory postauricular adenopathy deserves mention here. One or more lymph nodes reside under the skin behind the auricle overlying the mastoid bone. These drain adjacent areas of the scalp and may become enlarged for any reason, sometimes with tenderness. They may be mistaken for mastoiditis; the important distinguishing point is that no infection is seen in the middle ear, and mastoid X-rays or CT (if done) should be clear.

Summary

An individual who has rapid-onset earache following a respiratory infection, and a red or bulging drum, probably has acute otitis media! Antibiotic choices are discussed in the text. The extremely rare complication of acute mastoiditis may be recognized by persistent drainage through a nipple-like perforation in the drum for several weeks following an untreated acute infection, with mastoid swelling and tenderness. The primary clinician should follow up on a treated middle ear infection until it is resolved. A CT scan and ENT consultation should be obtained if mastoiditis is suspected, or if middle ear fluid persists.

Retraction of the Tympanic Membrane

This and the next few sections discuss changes in the middle ear that result from obstruction of the eustachian tube. Retraction of the drum is the earliest visible change. Studies have shown that if a eustachian tube is experimentally obstructed, visible retraction of the TM will occur in a matter of 20 minutes. The affected individual usually complains of a sense of blockage or pressure, even though the symptoms are caused by a vacuum. Popping and crackling sounds may be heard if the middle ear aerates intermittently. Conductive hearing loss, with autophony, occurs. The patient might say, "I feel as though I'm talking in a barrel or under water." The loss

is usually mild and in the low frequencies. The individual may also feel the desire to "clear" the ear with yawning or jaw motion.

Findings with a retracted membrane may be quite subtle. ENT texts describe "foreshortening of the malleus" and spreading or dulling of the light reflex, but this is not always easily seen. Once again, the pneumatic otoscope is the most reliable tool. The drum, being retracted, will not move inward with positive pressure, but will move outward with negative pressure. Experience in doing and seeing this helps. In addition, tuning forks will show the Weber test lateralizing to the involved ear. The Rinne test may not be reversed with a mild loss, although it is more apt to be when using the 256-Hz fork because the lower frequencies are the ones most affected at first.

The **modified Valsalva maneuver** can reverse a retracted TM. The patient is instructed to hold the nose and mouth shut and simulate blowing the nose, forcing air up the eustachian tube. This can be observed in the office with the otoscope; the retracted drum will inflate to a normal position with a successful maneuver. This can be repeated by the patient at frequent intervals to reverse the problem, although caution should be exercised because an infection in the nasopharynx, if present, may spread up the tube.

The cause of the obstruction, as discussed in the section on the eustachian tube, should be addressed. One should look for infection, allergy, or obstructing lesions in the nasopharynx. Treatments addressing the eustachian tube have often included antihistamines, decongestants, and nasal steroids. Unfortunately, there is not much objective evidence in the literature that these are effective, but they make empirical sense and help associated nasal symptoms. Of course, bacterial sinonasal infection can be treated with antibiotics.

Middle Ear Effusions

If the experimental model of eustachian tube obstruction continues for hours or days, the negative pressure will eventually lead to transudation of serous fluid into the middle ear cavity. This is known as **serous otitis media**. The patient will now complain of increased hearing loss, depending on the remaining air content in the middle ear. High frequencies will also be affected as fluid replaces air. Also, a sensation of moisture in the ear may be noted, with bubbling sounds. This can occur with position changes of the head, if some air is still present in the middle ear. If tinnitus is present, it is usually a low-frequency humming or roaring sound. The patient may also have pulsatile tinnitus, hearing his own small arteries pulsate near the ear due to the conductive loss.

Experience has shown that some very sensitive patients may experience mild dizziness, although there is no good physiologic reason for this; the inner ear should not be affected by middle ear fluid. Also, in the absence of infection, ear pain is not a usual complaint unless there are rapid pressure changes.

If a serous effusion continues for weeks, the fluid will likely thicken. The mucous glands of the middle ear and eustachian tube tend to proliferate and secrete more actively. The fluid can progressively thicken to a syrupy consistency and eventually become, in ENT jargon, "glue" (gelatinous mucus). These secretions may contain eosinophils and immune globulins, suggesting an allergic component. The conditions described here are known progressively as **mucoserous otitis media, mucous otitis media**, and **glue ear**. Various other names and abbreviations for these effusions have been used in the literature. **OME** (otitis media with effusion) is a good general term.

Findings through the otoscope vary. In early stages, clear fluid with bubbles or air–fluid levels might be seen (Fig. 5.**5**). As fluid increases and thickens, with loss of any air content, the drum may look darker, thick, or dull (Fig. 5.**6**). Notably, the serous and mucous ear effusions are usually sterile and do not cause the diffuse thick redness of acute bacterial infections. Sometimes the findings are subtle; the drum may remain fairly bright, with a good light reflex. The most reliable finding is **decreased TM mobility**. Little or no motion at all can be elicited when there is thick fluid and no air content. Audiometry will document **conductive hearing loss.** It may range from a slight (15 or 20 dB HL) loss in the low frequencies, to as much as 45 dB HL "across the board," depending on the type and

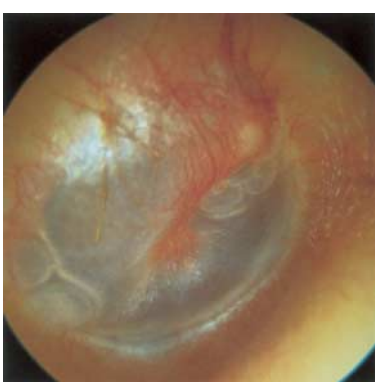

Fig. 5.**5** Serous otitis media with air bubbles and prominent vascularity. (Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

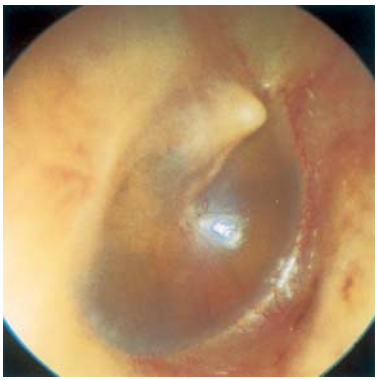

Fig. 5.**6** Mucous otitis media. (Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

amount of fluid. Tympanometry, discussed in the next section, will be abnormal.

A rare condition that probably represents an advanced sequela of thick effusion is known as **cholesterol granuloma**. This is a solid mass of granulation tissue in the middle ear containing clefts of cholesterol within it. Here the drum appears blue as if there is old blood behind it, although blood is not actually present.

Treatment Summary

The family physician is certainly qualified to diagnose and treat middle ear fluid unless it fails to improve in the long run. If the modified Valsalva maneuver has failed, and appropriate antibiotics to clear sinonasal or adenoid infection have not helped, then an ENT referral should be considered. Myringotomy and aspiration of the fluid can be done, although the drainage site will close very soon. A **ventilating tube** is the definitive treatment, to be decided upon when all other options have failed. Incidentally, with cholesterol granuloma, myringotomy and tube insertion does not work; more aggressive surgery is an option, albeit with limited success.

Ventilating Tubes and their Problems

Under local or general anesthesia, the ENT surgeon makes a small incision in the anteroinferior TM, away from the ossicles. Fluid is aspirated and a tube is inserted to keep the perforation open. Hearing loss is usually immediately corrected. Various types of tubes can be used, with different types of inner flanges and different diameter lumens. The smaller grom-

met-shaped ones usually self-extrude in a year or less. The larger ones, such as "T tubes," tend to stay in much longer but are more apt to leave a permanent perforation when they extrude. However, a small perforation is usually a great improvement over the original middle ear fluid. The nuisance with tubes or perforations is the need for earplugs to keep water out. A perforation from a tube can be surgically repaired. With children, the ear surgeon should wait until the eustachian tube problem has been outgown.

Tubes may have other complications besides persistent perforation. Recurrent otorrhea through a patent tube can occur. The organism is often *Pseudomonas*, and the quinolone eardrops are an effective treatment. Extremely rarely, a localized cholesteatoma can arise from the border of insertion (discussed later in this chapter).

Ear Effusions in the Young—Special Considerations

Infants and toddlers are afflicted most often with middle ear effusions, and they are not about to tell us they have conductive hearing loss or autophony. If their ears hurt, they will cry and fuss, especially when recumbent at night. Certainly, pain is present with acute or smoldering infection. However, many little ones have ongoing thick sterile effusions and offer very few complaints. Their mothers may or may not suspect a hearing loss. We have little history to go on.

Physical diagnosis may also be difficult, with subtle findings. During a well-baby visit, an ear examiner is likely to overlook a problem in a squirmy child whose canal is 4 mm wide and half full of wax. The narrowness of the developing canal lets in much less light from the otoscope and sometimes darkens the appearance of a normal TM. Conversely, thick mucoid effusions can be present behind a noninflamed drum and cause litttle change in color or light reflex.

What is the answer? There is a saying, "What you don't know won't hurt you," but the fact is that many studies show that even chronic mild losses, as small as 20 dB HL, may impair speech awareness and development. The extra effort of a careful examination with the pneumatic otoscope may pick up a problem. In addition to this, and audiometry, there is another very useful tool—tympanometry. Tympanometers are used increasingly frequently to screen the ears of young children.

The tympanometer measures the compliance, or elasticity, of the drum at various pressure settings. When a TM has some mobility, a "peak" in the compliance curve (pressure versus compliance) will be seen. If the peak is properly tall and occurs at 0 pressure, we have a normal middle ear. A peak in negative territory, below -150 or so, indicates significantly negative middle ear pressure, a retracted drum. As we progress toward

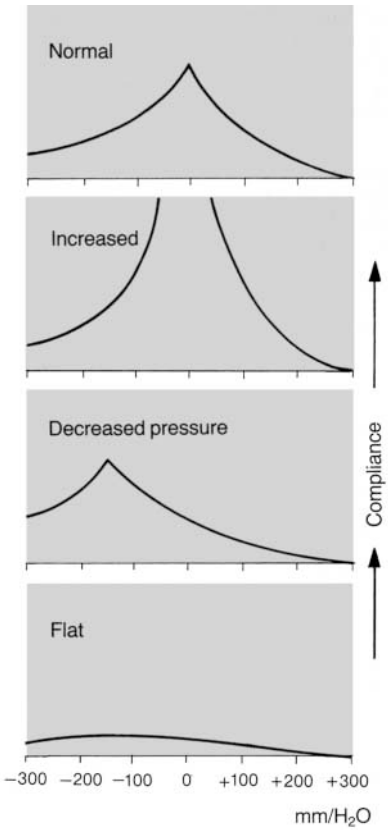

Fig. 5.**7** Tympanograms
From top to bottom: normal; increased compliance due to flaccidity or ossicular disruption; decreased pressure (TM retraction); flat tympanogram from dense effusion. (Source: Becker W, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994)

middle ear replacement with fluid, the compliance peak will flatten. A fluid-filled cavity with no air whatsoever will show a totally flat curve. The progression just described correlates very roughly with severity of the conductive loss. One other interesting variant occasionally shows up—the very tall peak. This is often seen with an overly compliant drum. It indicates either a disarticulation of the ossicles or a very thin, scarred area of "neomembrane," discussed earlier. Figure 5.**7** shows the four basic types of tympanometry curves (tympanograms) just discussed.

Treatment Summary

If a child has several months of abnormal ear findings despite appropriate antibiotics for acute otitis and sinonasal/adenoid infection, the family physician or pediatrician should consider ENT consultation. This brings us again to *ventilating tube insertion*. Most specialists feel that if a child endures continuously abnormal ears with hearing loss for 3 months or more, despite all conservative efforts, tubes are the answer. The recovery of hearing is usually immediate and noticeable. However, the tubes do not "cure" the problem. They are a temporary "umbrella under a leaky roof." The hope is that maturation of the eustachian tube and middle ear system will occur while they are in place.

There is evidence that *adenoidectomy* has a potentially permanent beneficial effect on both chronic effusion and repeated acute infection. The morbidity of this procedure is very low. Some would even prefer to avoid tube insertion and simply aspirate the ears when removing the adenoids. However, a myringotomy will heal very quickly, and fluid may reaccumulate if the removal is not immediately successful. Adenoidectomy is especially recommended for children who have recurrent middle ear problems after previous tube insertions. Incidentally, the tonsils have little to do with ear infections and should be spared if there is no other reason for removal.

Atelectasis and Retraction Pockets

We have not yet discussed some of the other dynamics that may affect middle ear aeration. The eustachian tube is not a conduit that is simply "open and normal," or "swollen shut." Middle ear pressure can change in a very positive way, as with the repeated Valsalva maneuver of frequent nose blowing. Extremely negative pressure can be generated by sniffing, a relentless habit in some patients. Such pressure variations, over time, may actually result in "stretching" of the drum, or flaccidity. Evidence shows that these activities may contribute to atelectasis, or deep retraction of the TM. Also, previous perforations that have healed, with loss of the fibrous layer, may be an underlying contributor. Simple chronic negative pressure is the only cause in many individuals.

Affected patients will usually have the same symptoms of plugged ears as those with TM retraction and middle ear fluid, and in fact, fluid may be present as well. Some patients with chronic atelectasis are accustomed to it and show no symptoms at all. The usual location is in the anterior or

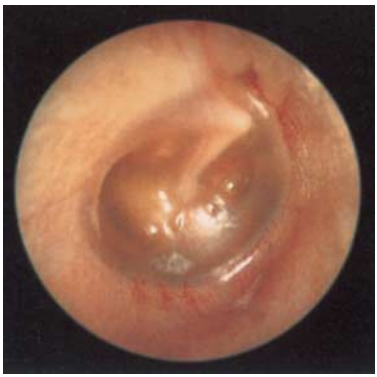

Fig. 5.**8** Posterior and anterior shallow retraction pockets, with serous effusion.
(Source: Hughes GB, Pensak MP. Clinical Otology. New York: Thieme; 1997)

inferior TM. Here, the drum appears to be very retracted and flattened out over the promontory. Sometimes, the inward retraction is gradual and diffuse, and other times, there is a sharp "step-off" between the non-retracted and atelectatic area, which may now be called a retraction pocket (Fig. 5.**8**).

A successful inflation by **Valsalva,** if possible, will produce a dramatic return of a retraction pocket to a normal position, and any hearing loss will usually improve. In fact, frequent performance of gentle inflation may result in good clinical improvement over the long run. Sniffing should be discouraged. Another treatment, aside from medical decongestive measures, is the insertion of a tube. In this case, the best option is a **T tube**, or any other type with a large inner flange. Grommet tubes, which have a smaller diameter and flanges, will extrude too readily. Insertion of the T tube is a surgically tricky process here; because the TM is often so thin and flattened out, it has the workablility of "wet tissue paper."

Adhesive Otitis Media

Retraction pockets that are posterior and superior are potentially dangerous. These are the ones located over the junction of the incus with the stapes at the entrance to the mastoid antrum, or more superiorly in the pars flaccida. The problem is that they tend to adhere to the ossicles and invaginate inward toward the mastoid antrum or attic (epitympanum) as time goes on. This leads to complications.

These pockets tend to occur in individuals with long-standing eustachian tube blockage causing chronic hypoaeration of the mastoid. Normal individuals develop a large volume of mastoid air cells as they mature and

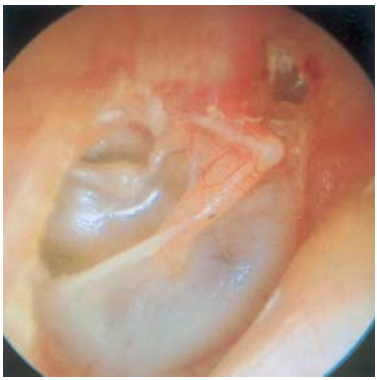

Fig. 5.**9** Inactive adhesive otitis media, with deep posterosuperior retraction pocket and incus erosion. (Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

these act as an "air cushion" for pressure changes. Sclerosis, or nondevelopment of the mastoid cells, predisposes to deep retractions here.

A patient with a quiescent retraction pocket posterosuperiorly may have the usual complaints related to negative pressure, or none at all. On examination, one will see an indentation of the drum over the incus–stapes joint, sometimes outlining these structures in sharp relief (Fig. 5.**9**). Sometimes, the pocket is hard to distinguish from a perforation, and in fact, a perforation may be present within the pocket. Diagnostically, negative pressure with the pneumatic otoscope may lift the drum off the ossicles if the pocket is not too deep, adherent, or perforated. If the drum is deeply adherent, then there is probably no mobility. The term for this occurrence is **adhesive otitis media**. (On Fig. 5.**9**, a deep retraction, adherent to the incus and stapes head, is shown. The end of the incus is slightly eroded. The stapedius tendon is the posterior horizontal structure.)

Inactive adhesive otitis may exist in a quiescent form for years with no damage and little, if any, hearing loss. Insertion of a long-lasting ventilating tube in the anterior drum may stabilize the situation. Unfortunately however, there is a tendency for infection and localized osteitis of the incus and stapes head to occur, with bone erosion and granulation tissue formation. This is now termed **active adhesive otitis**, and it often presents with weeping granulation tissue in the posterosuperior TM and distortion of landmarks here. With this, the patient complains of drainage that is sometimes bloody. There may be a red **aural polyp** (a large ball of granulation tissue) filling most of the posterior canal (Fig. 5.**10**). Once again, the microbes involved are the chronic ones, such as *Pseudomonas*.

Treatment with antibiotic–steroid eardrops may actually shrink away the polyp and improve the situation. It may also be removed, taking

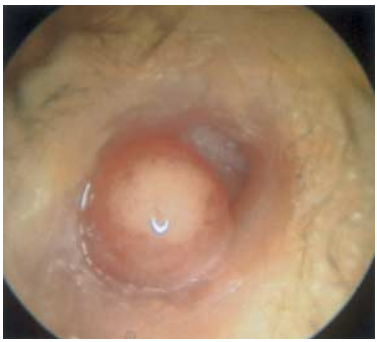

Fig. 5.**10** Active adhesive otitis media with aural polyp (granulation). (Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

care not to remove the stapes, which has happened because the polyp is often based here! No matter what treatment, there is a tendency for persistence and recurrences, with erosion of the lower incus. Similar occurrences and findings can be located in the pars flaccida over the neck of the malleus, and the same adhesion, osteitis, and granulation can occur here. Adhesive otitis in both areas tends to progress to cholesteatoma, and in fact, an aural polyp is likely to be hiding a cholesteatoma.

Cholesteatoma

Cholesteatoma, an otic epidermal cyst, is a complication of adhesive otitis that warrants aggressive surgical attention. Its formation occurs as follows. The posterosuperior retraction pocket of adhesive otitis insidiously invaginates deeply behind and around the ossicles, and "encystment" occurs. The epithelial lining of the invaginated cyst continues to produce keratin within its center, with expansion and bone erosion. Progressive enlargement into the mastoid antrum and/or epitympanum (attic) occurs, and more complications are possible. The actual histology of a cholesteatoma is similar to that of an epidermal inclusion ("sebaceous") cyst that can be seen under the skin anywhere else on the body. Unfortunately, the locale is not so benign.

The patient usually complains of chronic drainage and, possibly, a plugged ear. Early on, the hearing loss is mild. Sound can be conducted through the cholesteatoma to the stapes, even though the ossicles are eroding. Sometimes, erosion farther back in the mastoid antrum may involve the lateral semicircular canal and cause the vertigo of circumscribed labyrinthitis. Examination will most often show **posterosuperior drainage**, distortion, and possibly red **granulation**, as seen with active

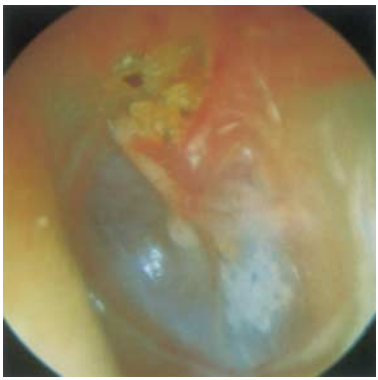

Fig. 5.**11** Epitympanic cholesteatoma, left ear.
(Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

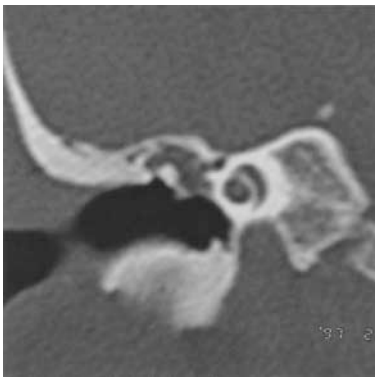

Fig. 5.**12** CT of epitympanic cholesteatoma. Note the density in the epitympanum around the head of the malleus and erosive blunting of the corner of the upper bony canal wall (scutum).
(Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

adhesive otitis. Characteristic wet flakes of **keratin** might be seen in the pocket (Fig. 5.**11**), but because they are not always seen, a scan may be necessary to confirm the diagnosis. On **coronal CT** scan of the temporal bones, the involved side will show a density in the epitympanum and mastoid antrum, with blunting of the scutum, the upper bony edge of the external canal (Fig. 5.**12**).

This type of cholesteatoma is the most common cause of **chronic mastoiditis** and has the potential for the following complications, which overlap those of acute mastoiditis:

1. Increased *conductive hearing loss* from erosion of the ossicles.
2. *Sensorineural hearing loss* from erosion into the labyrinth.
3. *Vertigo* from erosion into the labyrinth.
4. *Meningitis*, spread bacterially from the labyrinthine fluids or by direct erosion.
5. *Facial nerve paralysis*, by erosion and compression.
6. *Epidural* or *brain abscess*.
7. *Sigmoid sinus thrombophlebitis*.

In any patient who is severely ill with earache, fever, headache, stiff neck, or vertigo, look for a draining, distorted ear. Patients with a serious mastoid-related complication obviously needs careful workup with ENT consultation, hospitalization, and broad-spectrum IV antibiotics.

Hopefully, a patient's cholesteatoma will be picked up before these more serious complications occur. Corrective surgery (**mastoidectomy**) involves removal of the bone of the mastoid cortex and posterosuperior external auditory canal (EAC), with complete extirpation of the cholesteatoma. The eroded ossicles can sometimes be restored and repositioned and the drum grafted to restore adequate hearing (**tympanoplasty**). The surgery "exteriorizes" the region where the cholesteatoma existed, preventing its recurrence.

Several types of cholesteatoma exist. Those arising in the pars flaccida are called *primary acquired* or *attic retraction* or *epitympanic* cholesteatomas (Fig. 5.**12**). Those located in the pars tensa usually arise posterosuperiorly over the incus and stapes and are termed *secondary acquired* or *pars tensa* cholesteatomas. As these are similar enough in appearance, location, and clinical behavior, we will not dwell on their differences.

Congenital Cholesteatoma

A third type of cholesteatoma has nothing to do with chronic retraction, perforation, or adhesion. Its cause is uncertain, and it is much more rare than the first two types. This is the **congenital tympanic cholesteatoma** that may be discovered in a young child. It appears as a white, cystic structure seen beneath the TM, usually anteriorly. The drum external to it is grossly intact, and there is no chronic drainage, although middle ear fluid may be present due to coexisting eustachian tube obstruction. The histology is similar to the other types, however. Treatment is by surgical excision through a large opening in the drum, which usually heals without grafting procedures—tympanic epithelium in children is usually very regenerative. Congenital cholesteatomas can also occur as embryonic rests anywhere else in the temporal bone. These only show up if growth and erosion cause a complication.

Cholesteatomas from Ventilating Tubes

The fourth type of cholesteatoma is a very rare complication of ventilating tube insertion. This is seen as a white cystic structure, like the congenital one, adjacent to the tube insertion site. It may form while the tube is still in, or following extrusion. The cause is invagination and encystment of epithelium at the edge of the myringotomy site. This is more apt to happen with insertion of long-dwelling tubes. Treatment is by local excision through the drum, without mastoidectomy. These latter two types of cholesteatoma are not likely to cause the erosive complications of the first two unless huge, long-term growth is allowed to occur. Both may appear similar to white tympanosclerotic plaque.

Summary

The important thing for the clinician to recognize is that posterior or superior retraction pockets or perforations, especially with drainage and granulation tissue, are potentially dangerous. They may lead to, or actually hide, an active cholesteatoma, with all its potential complications. Diagnosis may be difficult due to the distortion that is usually seen, but the location should evoke suspicion. The primary physician might treat a draining abnormality in this area with eardrops as a first option, but a coronal CT scan and an ENT consultation is the safe route to take within a week of your first suspicion. Certainly, impending complications from a cholesteatoma and mastoiditis deserve immediate consultation. The congenital and tube-induced tympanic cholesteatomas are different; these appear white and cystic and are usually located under the anterior TM. ENT consultation is indicated for these, as well, but less urgently.

Unusual Eustachian Tube Disorders

Aerotitis

Two unusual middle ear pressure disorders should be discussed. The first of these is **aerotitis**, a form of barotrauma to the middle ear. The problem here is **rapid-onset negative middle ear pressure**. The typical scenario occurs with an airplane passenger who has an upper respiratory infection and swollen eustachian tube mucosa. On ascent, air pressure decreases outside the TM and middle ear pressure is relatively excessive. However, this readily decompresses down the eustachian tube during swallowing, and symptoms are not too severe. Unfortunately, the boggy tubal mucosa acts as a "one-way ball valve," and the real problem occurs when landing.

On descent, atmospheric pressure outside the drum increases rapidly and the swollen eustachian tube mucosa in the nasopharynx prevents aeration of the middle ear. The relatively large external pressure and low middle ear pressure collapse the drum inwards, and vessels in the middle ear may actually rupture and bleed, causing a hemotympanum. Thus, the individual has the sudden onset of a plugged ear that is often very painful. Middle ear findings of deep TM retraction, blood, and fluid can persist for days.

The primary practitioner can diagnose and treat this disorder with decongestants and the modified Valsalva maneuver. Refractory cases may require ENT consultation, with myringotomy as an option, although most cases spontaneously improve.

Prevention consists of a preflight dose of a 12-hour vasoconstricting nasal spray like oxymetazoline (Afrin), oral decongestants, and gum chewing while landing. The Valsalva maneuver to clear the ear can also be performed as needed, if possible. Incidentally, divers may experience similar barotrauma on deep descent. They are often well trained regarding the clearing maneuver, but it can be unsuccessful.

Patulous Eustachian Tube

A middle ear problem that is contrary to most of our discussion so far is the **patulous** (overly patent) eustachian tube. This is seen most often in older, debilitated individuals or individuals who have lost a lot of weight. It may also be precipitated acutely by a period of prolonged heavy exercise, with temporary dehydration and shunting of blood to the limbs and away from the head. In this disorder, **the eustachian tube is abnormally patent**; it remains open constantly. The symptoms are very annoying to those afflicted. Every breath, inhaled and exhaled, is felt and heard in the middle ear. Autophony is also present. One's own voice, and even those of others, sounds "hollow."

When suspected, the diagnosis can be made by visualizing the TM and asking the patient to occlude the opposite nostril and breathe in and out deeply. The examiner is often able to see the drum move in and out with respiration. Treatments in the past have focused on swelling the eustachian tube mucosa with chemical irritants or injecting Teflon at the orifice in the nasopharynx. Sadly, none of these have met with success. Temporary relief may be experienced when the patient lies down, due to venous engorgement of the eustachian tube. A **ventilating tube** may help, although the autophony is likely to continue due to ongoing tubal patency. Unfortunately, this is often a relentless disorder unless the weight loss or dehydration is reversed.

Otosclerosis

The term **otosclerosis** sounds like a hearing problem that develops in old age. In fact, it refers to a **conductive hearing loss** that usually develops in young adulthood. The loss is due to deposits of **abnormal bone around the footplate of the stapes**.* The stapes becomes progressively immobilized as time goes on, with advancement of the loss. These bony abnormalities can occur in other foci of the inner ear and cause cochlear hearing loss as well, but the stapes footplate is the usual location. Otosclerosis is the most frequent cause of conductive hearing loss in adults with a normal TM.

The cause of this disease is controversial. There is a well-documented familial tendency, and until recently it was thought of as a purely hereditary disorder. However, histologic examinations of the deposits of abnormal bone have shown inflammatory cells, and a new study has demonstrated measles virus RNA in stapes footplate specimens. It is possible that viral infection, autoimmunity, and heredity all play a part. Two-thirds of cases occur in women, and there is a much greater prevalence in whites than blacks. The hearing loss typically becomes apparent in the third to fifth decades of life. Otosclerosis is also associated with Paget's disease of bone and osteogenesis imperfecta.

The typical patient presents in her or his young adulthood with gradual hearing loss and, often, low or medium-pitched tinnitus. A **normal TM** is seen. **Tuning fork** testing will uncover the conductive loss, with the Weber test lateralizing to the bad side. A loss of 30 dB HL or more should make the Rinne test abnormal (bone conduction greater than air). A typical audiogram is shown in Figure 5.**13**. The disease is usually bilateral, but one side is often more affected than the other. Audiometry will further document the conductive loss, showing an "air–bone gap." Tympanometry usually shows a lowered compliance peak due to stiffening of the ossicular chain by stapes fixation.

Stapedectomy, surgical removal and grafting of the stapes with prosthetic or natural material, achieves a good result in a vast majority of patients, although there is a tiny risk of total hearing loss as a complication. A **hearing aid** or aids can be used if desired, but the loss tends to progress over the years. A number of studies show that supplemental **sodium fluoride** tablets, 2.2 mg daily, may retard progression of the disease, which can go on to develop foci in the cochlea. In fact, the widespread use of water fluoridation in recent years may be causing a decrease in incidence,

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* The bony deposits are actually "spongy bone" and purists use the term *otospongiosis*.

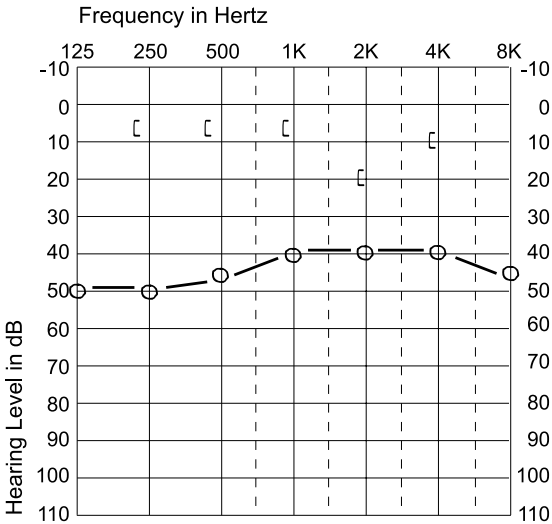

Fig. 5.**13** Audiogram of otosclerosis, right ear, with the air–bone gap of conductive hearing loss.

but this is pure speculation. Also, the study regarding measles virus notes that since the introduction of rubella vaccine, the incidence of otosclerosis is decreasing and the median age of discovery is rising.

Summary
Otosclerosis is the most common cause of unexplained conductive hearing loss in adults. The TM appears normal, and tuning forks will be the only diagnostic contributor on routine office examination. Treatment options are surgery or hearing aid(s). Daily sodium fluoride may retard progression of the disease. The ENT physician, when consulted, can explain these options to the patient.

Congenital Ossicular Abnormalities

A child may have a conductive hearing loss present from birth, with no middle ear effusion or retraction to explain it. "Juvenile" otosclerosis is possible, but this is extremely rare. The only other possibility is a congenital malformation of the middle ear ossicles. Some middle ear malformations readily stand out, with distorted TM landmarks or accompany-

ing abnormalities of the canal (as with **congenital atresia**, discussed in Chapter 4). Others, however, show a normal drum.

The most common deformity seen with patients that have a normal drum and landmarks is *ankylosis* of the malleus or incus heads in the epitympanum. Here, there is fusion of either ossicular head to surrounding bone in the attic, with otherwise normal anatomy. Other malformations may be more extensive and even involve absence of one or more ossicles. These are more apt to show distortions of landmarks when one examines the TM. An **audiogram**, **tympanogram**, and **CT scan** will help diagnose the hearing loss and image the deformities. Associated abnormalities in the location of the facial nerve may be present. **Surgical reconstruction** of these deformities to correct the hearing is often possible but caution is warranted to avoid injury to the facial nerve, which may be misplaced in some anomalies.

Middle Ear Tumors

The most common tumor of the middle ear, or hypotympanum beneath it, is **glomus jugulare**. This vascular neoplasm arises from neural crest cells in the adventitia of the jugular bulb, or from Jacobson's nerve on the promontory of the middle ear wall (in this case, called *glomus tympanicum*). It is often benign, but it may have malignant characteristics and slowly grow to tremendous size. In the literature, these tumors are staged according to their size and location.

A patient with a glomus tumor typically presents with **pulsatile tinnitus** and possibly, **autophony** from a conductive hearing loss, if the ossicles are involved. A **red or purplish mass** may be seen in the middle ear, usually inferiorly, behind an intact drum (Fig. 5.**14**). This mass may represent the entirety of the tumor or may just be the top of a huge one.

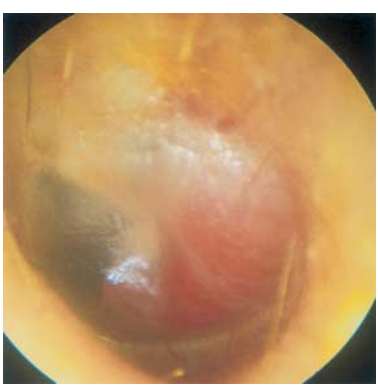

Fig. 5.**14** Glomus tumor of the lower left middle ear.
(Source: Sanna M, Russo A, DeDonato G. Color Atlas of Otoscopy. Stuttgart: Thieme; 1999)

Evaluation with a **CT scan with contrast** or **MRI** is essential. The stage I tumors confined to the middle ear may be removed with simple middle ear surgery, but the more advanced stages require skull-base surgery and/or irradiation. These tumors grow very slowly but, if untreated, may spread through the middle ear to the mastoid and even cause facial nerve paralysis. The ones arising from the jugular bulb may be hidden beneath the ear and yet massively involve the skull base and other cranial nerves, with a poor prognosis.

Briefly, other tumors rarely appear in the middle ear. These include benign connective tissue tumors, such as fibromas, and malignant ones like squamous cell carcinoma. Even metastatic tumors from other sites have been reported in the literature.

Spontaneous Perilymph Fistula

Only over the last several decades, with improvement in middle ear surgery techniques, has spontaneous perilymph fistula been discovered, documented, and surgically treated. A spontaneous fistula manifests itself mostly with inner ear symptoms and findings, but the cause actually lies somewhere between the middle and inner ear, and the surgical repair is done through the middle ear. Hence, it is discussed in this chapter. The lesion is a leak in the round window, or in the oval window adjacent to the stapes. The symptoms of hearing loss and dizziness can mimic Méniere's disease (see Chapters 7 and 8). In earlier years, its high rate of discovery around some teaching centers was a source of controversy.

This problem clinically presents with symptoms of **sudden hearing loss** and **vertigo**, usually **following forceful physical activity**. A violent cough, sneeze, or heavy exertion, producing a jump in pneumatic and venous pressure in the ear, brings on these symptoms. Divers, with their clearing maneuvers for pressure changes, are often prone. However, some cases are said to occur with a less noticeable incident, and thus high index of suspicion is needed to make the diagnosis.

On examination, a positive **fistula test** may point to it. Here, the pneumatic otoscope, with a good air seal, alternates moderate inward and outward pressure on the TM. When the test is positive, the patient will experience rocking vertigo accompanied by nystagmus. Swaying will occur if the test is done while the subject stands with the eyes closed. Tuning forks will show a variable magnitude **of sensorineural hearing loss**.

Early conservative treatment with bed rest and the head elevated is the first option, but many cases require **surgery**. Exploration of the middle ear reveals clear fluid oozing from the periphery of the footplate or from

the round window niche. Surgical patching of the leak is done with with connective tissue, such as fat obtained from the patient's ear lobe.

Summary
Recognition of spontaneous fistula requires a high index of suspicion. The rapid onset of hearing loss and vertigo may mimic a number of other inner ear disorders, such as Méniere's disease and sudden sensorineural loss. It is the history of some forceful precipitating physical activity, from a simple sneeze to a scuba diving clearing maneuver, along with a postive fistula test, that should alert the clinician. When suspected, the ENT physician should be consulted within a few days.

Overview of Mastoiditis

Having discussed a number of forms of mastoiditis, some clarification is in order at the end of this chapter. This diagnosis was ominous in the old days before antibiotics. It often led to primitive surgery, deafness, brain complications, and sometimes death. Times have changed. Now there are fewer acute cases and more refined treatments. As a review, some definitions and explanations are presented here.

1. *Acute mastoiditis* or *acute coalescent mastoiditis* refers to the purulent type that may occur following untreated severe acute otitis media. It is rare nowadays, but can be diagnosed by the physical findings and CT scanning.
2. *Chronic mastoiditis* refers to two types of ongoing infection in the mastoid antrum and/or air cells: a) One type is the erosive, expanding epidermal cyst—*cholesteatoma*. b) A second type occurs with chronic draining middle ear perforations. Here, purulent material and/or granulation are present in the middle ear and adjacent mastoid cavity. The term *chronic suppurative otitis media* overlaps this entity.
3. "Chronic mastoiditis" may also be diagnosed by a radiologist who, when reading routine X-rays or a CT scan, refers to an undeveloped mastoid cavity without a good "honeycomb" of air cells. This is actually more accurately termed a *sclerotic* mastoid. Inflammation is not necessarily present. However, the finding is abnormal and results from chronic eustachian tube disease with hypoaeration of the middle ear and mastoid.
4. "Mastoiditis" may be read by a radiologist when referring to any fluid density in the mastoid air cells. This is often a misleading overdiagnosis. To illustrate, if a CT scan is done on a patient with a sterile serous middle ear effusion, the fluid usually opacifies the mastoid air cells as well.

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6 Disorders of the Facial Nerve

The facial nerve, or cranial nerve VII, certainly deserves the attention of anyone who treats the ear. This structure is not involved with hearing or balance, but its disorders are often related to the ear due to its location. Most of it runs through a tunnel just deep to the tympanic cavity. The bony *fallopian canal*, 33 mm long, encloses it and separates it from the middle ear by only a millimeter or so over much of its course.

The facial nerve arises from the facial nucleus in the brainstem, then heads laterally through the internal auditory canal (IAC) along with the vestibuloauditory nerve (cranial nerve VIII). It then makes a sharp bend (the *genu*, meaning "knee") posteriorly, just deep to the neck of the malleus. This begins its horizontal portion, which courses back over the stapes in the oval window. Posterior to the oval window, it turns gently downward into its vertical portion and descends just behind to the middle ear cavity in its "mastoid segment." It exits the temporal bone between the tympanic ring (bony external canal) and the mastoid tip. From here, it angles forward, branching through the parotid gland to innervate the facial muscles (Fig. 6.**1**). This course makes it susceptible to damage by ear diseases and trauma, as well as by parotid gland problems. Thus, knowledge of it is essential to the otolaryngologist or any practitioner who deals with ears.

During its course through the middle ear, the cranial nerve gives off some specialized fibers that innervate other structures. Thus, there may be additional symptoms besides facial paralysis if the nerve malfunctions, as will be seen in the discussion of Bell's palsy. At the genu, branches to the lacrimal and parotid glands arise, promoting secretions there. The nerve to the stapedius, from its upper vertical portion, causes reflex contraction of this muscle to protect the ear from loud noise. Just below this, the chorda tympani arises, bringing taste sensation inward from the anterior tongue and sending secretory innervation outward to the lower salivary glands.

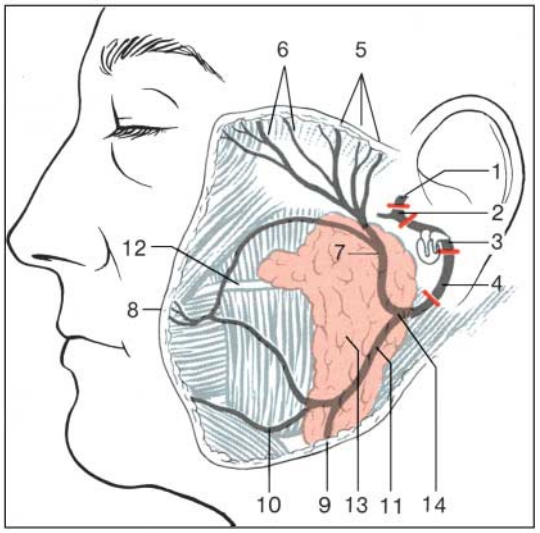

Fig. 6.**1** Anatomy of the facial nerve:
1. Entry outward into temporal bone from brainstem; 2. The genu (a much sharper turn and closer to the malleus than depicted); 3. Second turn to its vertical mastoid segment; 4. Exit from the temporal bone; 5. Temporal rami; 6. Orbital/brow rami; 7. Temporofacial trunk; 8. Buccal/oral rami; 9. Cervical rami; 10. Mandibular branch; 11. Cervicofacial trunk; 12. Parotid (Stenson's) duct; 13. Parotid gland; 14. Bifurcation within parotid gland.
(Source: Becker WW, Naumann HH, Pfaltz CR. Ear, Nose, and Throat Diseases. Stuttgart: Thieme; 1994.)

Bell's Palsy

Until recent years, Bell's palsy was thought to be idiopathic. ENT textbooks were dogmatic about ruling out all other causes of facial paralysis before diagnosing Bell's palsy. This is no longer true. The presentation of the disorder is so characteristic that it is not "taboo" to readily diagnose it, based on a careful history and examination. In addition, our recent sophistication in molecular biology suggests that a viral cause exists—most likely, herpes simplex is the culprit. Other factors or other viruses may be involved, however, and there is still controversy and ongoing investigation regarding this.

Clinically, the typical chain of events starts with a rapid onset of facial weakness, often following a period of stress or a viral illness. Several other symptoms may occur. A metallic taste and dryness of the mouth (chorda

Fig. 6.**2** Bell's palsy, with total left facial paralysis.
(Source: Hughes GB, Pensak MP. Clinical Otology. New York: Thieme; 1997.)

tympani involvement) are frequently noted at the onset. The patient might also note sensitivity to noise (stapedius paresis) or dryness of the eye (lacrimal branch involvement) on the affected side. These other symptoms, which relate to the aforementioned specialized nerve branches, are variably present. Pain, in or around the ear, is experienced about half the time, although it is not as severe as with *Ramsay-Hunt syndrome* (discussed next). The patient will often note drooping of the mouth, with drooling while trying to drink liquids, as the first symptom.

On examination, one will see unilateral facial weakness (Fig. 6.**2**). Notably, the eye will not close and the mouth droops. The weakness may be partial (paresis), or total and flaccid (paralysis).* Bell's phenomenon is usually present; the pupil deviates upward when an effort is made to close the eye. A careful ear examination should reveal no abnormality other than hyperacusis on the affected side, due to involvement of the nerve to the stapedius muscle. This can be demonstrated by placing a loud tuning fork next to each ear—the patient will wince on the affected side. A good examination should rule out other causes of facial paralysis, such as middle ear, mastoid, or parotid disease.

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* The *entire* half of the face is weak, as opposed to just the lower facial muscles, as with a cerebrovascular accident. This is due to unilaterality of the facial nerve nucleus, but crossing of corticomedullary fibers.

Once the diagnosis is made, one can discuss the prognosis with the patient. It is encouraging, but not perfect. When untreated, roughly 70% of Bell's palsies recover after 4 to 6 weeks, without residual facial deformity. Another 15% may last longer, leaving a slightly noticeable asymmetry. The worst 15%, however, have a prolonged course, with eventual recovery of flaccidity, but noticeable permanent asymmetry and spasm of the facial muscles. *Synkinesis* may be seen in these latter unfortunate patients. This is the tendency for more than one muscle group to contract at the same time. For example, the eye may squint while smiling.

Treatment can shorten the course of the disease and improve the prognosis. **Steroids** should be given over a period of at least a week, starting with a dose of about 60 mg of prednisone for adults. This should be continued for a longer period in refractory cases, with appropriate tapering. At least one recent study shows that the early use of **acyclovir**, 400 mg five times a day, in addition to steroids, improves the prognosis. This evidence supports the viral hypothesis. The flaccid eye may require patching or taping to prevent dryness and the risk of keratitis. Some recommend electrical muscle stimulation in prolonged cases. This may keep up muscle tone while the nerve recovers.

Numerous surgical facial nerve decompressions have also been done in the past for refractory and severe cases. The rationale was to allow the nerve "to expand," relieving pressure from the confining bony canal. Indeed, there were a number of reports of immediate, dramatic postoperative recovery. However, there was little agreement on surgical indications. Because evidence now suggests there is a viral cause and that steroids and acyclovir are of benefit, surgery is no longer a consideration in most cases.

The primary practitioner who is aware of the above symptoms and findings is certainly capable of diagnosing and treating this illness. Elective ENT follow-up is an option for refractory cases, and an ophthalmologist may even be needed to treat the drooping eye.

Ramsay-Hunt Syndrome

Ramsay-Hunt syndrome is caused by the **herpes** (or **varicella**) **zoster** virus. Like Bell's palsy, it presents with rapid onset of facial paralysis, but also with a **vesicular rash** (as seen with "shingles") erupting in the external ear or near it. There is almost always **severe otalgia**, and the virus affects other **adjacent cranial nerves** as well. Cranial nerve VIII is the most frequently affected, with sensorineural hearing loss and vertigo. Lesions of cranial nerves IX and X may cause pain in the pharynx, and involvement of cranial nerve V may give facial pain. The prognosis of Ramsay-Hunt syndrome is not as good as that for Bell's palsy. The clinical course is more

prolonged and the facial asymmetry is more apt to persevere. In addition, the hearing loss may be permanent. Vertigo and dysequilibrium from vestibular involvement can persist. In general, with unilateral vestibular loss, the opposite vestibule eventually compensates for most of the symptoms. However, this may take 6 months or more.

Treatment by the primary physician centers on **steroids** and **antivirals**, as with Bell's palsy. Steroids should be given over a long period, perhaps 2 weeks or more. Acyclovir is recommended as early as possible in the high-strength dosage—800 mg by mouth five times a day. Alternatively, Famvir, a newer antiviral in the same family, may be given. Total recovery, with no residual deficit, is less than 50%, with older age bringing a worse prognosis.

Infection and Facial Paralysis

As mentioned in Chapter 3, **acute otitis media** may rarely cause paresis, possibly by way of a bony dehiscence in the fallopian canal. If a face becomes rapidly weak on the side of an acute infection, **myringotomy and aspiration**, with a culture and appropriate **antibiotics**, are the accepted treatments. Most of these patients will recover. If facial weakness occurs 2 or 3 weeks after an acute otitis, be suspicious for **acute coalescent mastoiditis**. A thorough ear examination and work-up, with a CT of the mastoid, would be indicated. **Chronic mastoiditis**, either with granulation tissue or cholesteatoma, can also cause facial paralysis.

When facial paralysis is present in the face of positive middle ear or mastoid findings, as well as with trauma or evidence of tumor (discussed later), the primary physician should obtain early ENT consultation, as surgical intervention may be necessary.

Trauma and Facial Paralysis

Longitudinal and *transverse* **fractures of the temporal bone** may disrupt the facial nerve. Longitudinal fractures are more external, occurring in the plane of the external auditory canal (EAC). They are apt to disrupt the ossicles in the middle ear. They cause facial nerve problems about 20% of the time and are the much more prevalent type. Transverse fractures are rarer, traversing the petrous apex from anterior to posterior. These have a much higher incidence of facial paralysis, as well as vestibulocochlear damage. Immediate facial paralysis warrants surgical exploration and decompression. Delayed onset with incomplete paralysis will probably recover spontaneously.

Middle ear and mastoid surgery are the dreaded causes of facial injury in the ENT specialty. Mastoidectomy is the most common cause, usually

from an injury to the second turn, posterior to the stapes. This can be done in an effort to remove cholesteatoma or chronically infected tissue that closely involves the nerve. Re-approximation or grafting of the damaged ends can result in a recovery of function, although the result may take 6 months or more to show up; regeneration occurs at roughly 1 cm a month. Stapedectomy can also result in facial nerve injury. The fallopian canal is immediately superior to the oval window and can be dehiscent here, rendering it susceptible to damage. Distal to the ear, **parotid tumors**, or their surgical removal, can endanger the nerve. Adenoid cystic carcinoma is a major culprit in that it is malignantly invasive and tends to grow along perineurium, the lining of nerves.

Finally, traumatic **forceps deliveries** have caused neonatal facial paralysis, either by temporal bone fracture or by direct compression beneath the ear. These are also associated with hearing damage and are extremely rare. We might group penetrating injuries together with this; these may occur through the ear canal or beneath it at the stylomastoid foramen and posterior parotid gland.

Other Causes of Facial Paralysis

Compression by tumor can occur anywhere along the course of the facial nerve. Most proximally, an **acoustic neuroma** within the IAC can compromise it, usually late in the evolution of this slow-growing tumor. Early growth involves the vestibular and auditory nerves, causing hearing loss and dysequilibrium. Unfortunately, surgical removal of an acoustic neuroma can jeopardize a still-functioning facial nerve. In the middle ear, **glomus jugulare tumors** are the most frequent culprits, although squamous carcinoma, adenoid cystic carcinoma, and malignant metastatic tumors rarely may involve the nerve as well.

Sarcoidosis, Lyme disease, infectious mononucleosis, diabetes mellitus, and leukemia may all rarely involve facial palsy. Finally, there is a syndrome of recurrent facial paralysis with furrowed tongue and orofacial edema known as *Melkersson–Rosenthal syndrome*. This may be an autoimmune disease and is associated with angioedema. Here, surgical decompression of the nerve is indicated because the recurrences of the paralysis will eventually cause total denervation.

Summary
Sudden onset of facial paralysis, in the absence of any other diseases or precipitating factors, is probably Bell's palsy. A complete history and ENT examination should be done to rule out other causes. Facial weakness with severe pain may herald Ramsay-Hunt syndrome. The appearance of skin lesions, hearing loss, or vertigo will support this diagnosis. We now know that steroids and early antiviral medications may improve the outcome of both these diseases. When diagnosing a facial paralysis, one should beware of hidden middle ear, inner ear, or parotid lesions.

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7 Auditory Disorders of the Inner Ear

The inner ear, or labyrinth, lies deep to the middle ear cavity. Here, the complex sensory organ known as the *membranous labyrinth* is enclosed within the *bony labyrinth*.* The hearing and balance portions are in continuity with one another, and their tiny membranous channels share endolymph and perilymph. Many inner ear disorders affect both portions but, clinically, there is often a predominance of symptoms from one or the other. Hence, our discussion will be divided into two chapters. This one deals with problems that primarily affect the auditory division (i.e., the cochlea and auditory nerve). Chapter 8 deals with problems that mostly involve the vestibular division (i.e., the utricle, saccule, semicircular canals, and vestibular nerve).

At the outset, a couple of clinical points should be made about the inner ear as an isolated "compartment." Inner ear problems cause only a few specific symptoms. Sensorineural hearing loss and tinnitus may result from cochlear disease. Vertigo or poor equilibrium may result from vestibular disease. A sense of ear fullness may result from excessive endolymphatic pressure in either division.

There are no pain-sensing fibers in the labyrinth except for the ones carried *through* it from external structures via the facial nerve (cranial nerve VII). Thus, when a patient presents with ear pain as a predominant symptom, an inner ear problem is not the likely cause. Moreover, outer and middle ear problems, in general, do not cause inner ear problems. For example, middle ear fluid or acute infection should not cause sensorineural hearing loss or severe vertigo.

Having said this, some exceptions exist. Aggressive middle ear and mastoid problems (e.g. cholesteatoma or tumor) can directly invade the inner ear by bone erosion. Also, "inner ear" pain might be perceived via meningeal irritation or facial nerve compression—causes of this might include meningitis or abscess from mastoid disease, and large, deep tumors like acoustic neuroma. Finally, *chronic suppurative otitis media* may cause insidious cochlear hearing loss over the years, presumably by way of diffusion of toxins through the round window membrane.

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* The petrous portion of the temporal bone encasing the labyrinth is also known as the *otic capsule*.

Tinnitus

Webster defines tinnitus as a ringing or buzzing in the ear that does not result from an external stimulus. More loosely, it means any noise that is heard in the ear. Tinnitus is discussed here because it is experienced most often with cochlear hearing losses. However, as a symptom, it can accompany disturbances of the outer, middle, or inner ear, and even temporomandibular joint (TMJ), vascular, or metabolic disorders. Often it is just plain "idiopathic" and not associated with any objective disease. The mechanism producing it is much studied, but elusive. Extensive research has been devoted to finding a measurable electrical potential in the cochlea, auditory nerve, or brain to physically locate the source of tinnitus. Little understanding or localization has been gained so far.

However, it is well known that certain types of tinnitus are associated with certain conditions. Often, there is an underlying hearing loss when it is present. When patients complain of ear noise, you should ask them to describe, as accurately as possible, what they hear. The sound's character, duration, frequency of occurrence, and provoking activities all help the clinician figure out a cause and possible remedy.

Somatosounds

These *rarest* types of tinnitus, formerly termed **objective tinnitus,** are sounds in the ear that both the patient *and* the examiner can hear. The clinician can hear it by placing his ear close to the patient's, or by using a stethoscope on or adjacent to the patient's ear. The usual cause of somatosounds is myoclonus. Also, pulsatile tinnitus can occasionally be heard objectively.

Myoclonic Tinnitus

Palatal myoclonus is an extremely rare disorder, characterized by a series of rhythmic clicking sounds in a patient's ear, which may occur often and repeatedly. These are due to involuntary contractions of the muscles of the palate in the pharynx that transmit sound and pressure changes to the ear via the eustachian tube. Another form, **middle ear myoclonus**, refers to similar contractions of the tensor tympani or stapedius muscles. Both types of myoclonus might be heard objectively by the examiner. Sometimes, this disorder occurs in an individual on rare occasions and then goes away, similar to twitching of the eyelid. More often, it is a relentless condition that appears in middle or advanced age. A brainstem abnormality is the presumed explanation, and occasionally underlying disease, such as multiple sclerosis, has been demonstrated. Magnetic resonance imaging (MRI), with focus on the brainstem, should be con-

sidered. Unfortunately, no standard treatment protocol has been effective for idiopathic cases.

Pulsatile Tinnitus

Tinnitus that is vascular in cause can occasionally be heard as a somatosound. The presenting patient often complains of a rhythmic "swishing" or "beating" sound in one or both ears. The sound may not be audible to the patient all the time. Certain activities may precipitate it, such as lying down on the affected side or performing heavy exercise. Asking the patient if it coincides with his/her pulse helps to make the diagnosis.

On examination, pressure on the carotid bifurcation will often lessen the sound if the patient is experiencing it at the time. The stethoscope should be tried to see if it is loud enough to be heard objectively; if so, a serious workup is in order. Otoscopy may show up a middle ear abnormality. A **vascular tumor** (such as glomus jugulare) might be discovered behind the drum. Sometimes just plain middle ear fluid, or any other reason for conductive hearing loss and autophony, is the explanation; one tends to hear internal vascular sounds more loudly with these conditions.

A transmitted **carotid bruit**, as well as a **vascular loop** or **aneurysm**, may be the cause. This begs the question, "Should a CT or MRI with contrast be done to rule out all the dangerous vascular abnormalities?" Some advocate doing this in *all* patients with pulsatile tinnitus. However, I would advise clinical discretion here, basing one's judgment on the severity and persistence of the tinnitus and the physical findings. Relentless loud pulsation deserves such a workup.

On the other hand, the vast majority of patients with this symptom have intermittent symptoms and no demonstrable vascular abnormality. The cause is often a tortuous or sclerotic vessel near the ear, and many cases spontaneously improve with time. Discussion of possible causes with the patient, with a period of conservative observation, may avoid a costly study. **Aspirin**, one grain daily, may help when minor vascular compromise is suspected as the cause. An investigation of blood pressure, blood sugar, cholesterol, and cardiovascular status may reveal a contributing abnormality.

Subjective Tinnitus

Subjective tinnitus is inaudible to the examiner. It is the vastly predominant type and includes most cases of pulsatile tinnitus as well. The sounds heard take many forms, but a large number are high-pitched and occur in conjunction with high-frequency cochlear hearing loss.

The three main causes for high-pitched, nonpulsatile tinnitus are **presbycusis**, **noise-induced hearing loss**, and **ototoxicity**. The patient will usually describe a ringing or hissing sound, and interestingly, the pitch of the sound often corresponds to the frequencies that show the most hearing loss on audiometry. This type of tinnitus is usually continuous, but more noticeable when it is quiet. Unfortunately, presbycusis and chronic noise-induced loss are irreversible and little can be done to help the problem. Of course, patients with noise damage should be encouraged to avoid further exposure. Ototoxic agents should be avoided or discontinued. They will be discussed in more detail at the end of this subsection.

Tinnitus may also be heard as a low-pitched humming or buzzing. Ménière's disease, which affects the low and middle frequencies, usually causes a "roaring" or "seashell" noise. In other patients, insect-like sounds, "high-tension wires," pings, and all sorts of other noises may be experienced. One patient of mine some years ago repeatedly heard "Happy Birthday, Nathan," but this is more properly classified as a hallucination.

It must be emphasized that an **audiogram** is an essential part of a workup for tinnitus if the patient is looking for a good explanation. This will show if a hearing loss exists. As we stated, a deficit often shows up in the frequency range that roughly corresponds to the sounds experienced. The cause may then be identified, although an effective treatment might not exist. Recognizable types and patterns of cochlear loss are discussed under their headings throughout this chapter. Some patients experience tinnitus without any demonstrable loss, and obviously, this is more difficult to explain.

Treatment

When a recognizable hearing loss like presbycusis or noise damage is seen on the audiogram, the cause can be discussed with the patient. It is sometimes convenient to explain to the patient that "the damaged hair cells are making noise" when a cochlear loss exists, although this is not the whole truth. It is also helpful to reassure the patient that there is no ominous disease process present. Psychological factors may play a big role in one's perception and acceptance of tinnitus.

Regarding medical treatments, **antidepressant** and **antianxiety medications** may help. A broad range of other medications have been tried, including **vasodilators** (notably niacin), antihistamines, anticonvulsants, and calcium channel blockers, all with limited success. **Melatonin** has been mentioned as an effective therapy recently.

A number of nonmedical treatments exist. The use of pleasant background noise, such as soft music on a clock radio at bedtime, is good advice. The use of conventional hearing aids, if needed for hearing loss, often ame-

liorates the symptoms. Conditioning techniques to adapt to the noise have also been used. **Tinnitus retraining therapy** is a system of counseling in conjunction with the use of hearing aids and/or noise-generating devices. This has had success in reducing patients' awareness and annoyance by the tinnitus, although it can still be perceived. There are diagnostic and therapeutic centers in various cities, as well as the **American Tinnitus Association**, a research and support group with many regional chapters. This symptom is distressing to a number of individuals, and those afflicted should be treated with compassion.

Ototoxicity

Special mention should be made of medications that can damage the hearing, usually in the high frequencies first. The earliest sign of damage is high-pitched tinnitus. The most common offenders are the salicylates, aminoglycoside antibiotics, loop diuretics like Lasix, quinines, and some chemotherapy agents. Fortunately, the ototoxicity of salicylates is reversible when the drug is discontinued. Damage from the others is likely to be irreversible. One should ask the patient about all current medications and consult a drug reference book if there is any question. Caffeine, alcohol, and tobacco are also possible contributing agents that can be avoided.

Summary

The type of tinnitus that is heard often gives a clue to the cause. An examination should be performed, to rule out visible (or audible) abnormalities, and an audiogram. A diagnostic reason for the tinnitus may help put your patient at ease—many individuals are overly concerned that their ear noise portends a tumor or other serious medical problem. Perhaps a medication or other ingested substance is the culprit and can be eliminated. Most certainly, noise exposure should be discouraged. In some cases, antianxiety medications may help a distressed individual. Other medications, mentioned above, may be tried. The family practitioner might independently obtain an audiogram for a tinnitus patient in order to find a recognizable cause, although the experienced input of an ENT physician is usually useful. Tinnitus centers exist in some areas.

Presbycusis

If you are not a pediatrician, the predominant cause of hearing loss and tinnitus that you will encounter in your patients is presbycusis, the hearing loss of aging. This problem will significantly affect one out of three of us, if we get old enough. With presbycusis, there is a gradual loss in the high frequencies as time goes on, and this occurs at different rates in different people. Genetic factors, lifestyle, noise exposure, and medical conditions influence the progression. Many 80-year-olds still have normal hearing at all measurable frequencies; other 40-year-olds may start showing a drop, first at 8 KHz and then at 4 KHz. The loss is usually symmetrical bilaterally and progresses in a "sloping" fashion, with the higher frequencies affected more and earlier. Cochlear hair cell deterioration is the cause of the loss. Figure 7.**1** illustrates a typical audiometric progression.

The stubborn patient with **presbycusis** might be sent in by his spouse, denying that a problem exists and asserting that his wife mumbles. The more insightful patient typically complains that he has difficulty understanding speech, especially in background noise, even though the voiced vowels are heard. The statement "I hear them talking, but I don't know

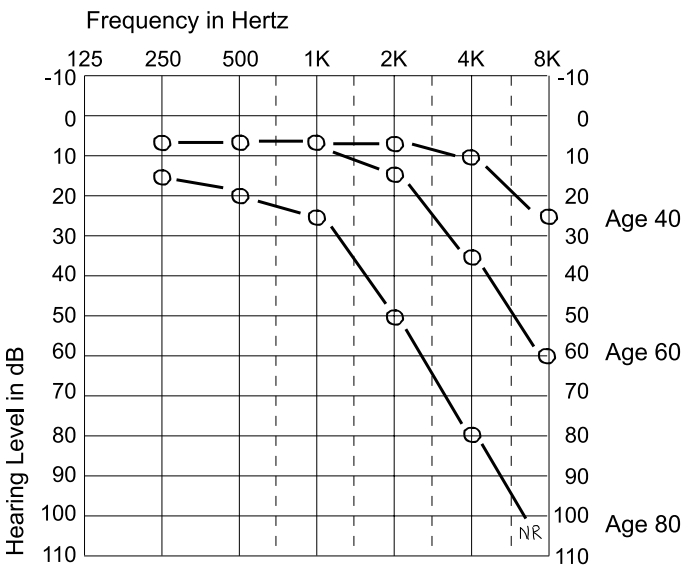

Fig. 7.**1** Audiogram showing examples of presbycusis progression, right ear.

what they're saying" reflects the fact that the high frequencies are impaired, with resulting decrease in ability to hear consonant sounds, as we discussed in Chapter 1.

On hearing evaluation, **speech discrimination testing** will usually show a drop, once 3 KHz and then 2 KHz are affected. High-pitched tinnitus is a frequent complaint. The interesting phenomenon of *recruitment* is often present, where there is an acute sensitivity to small increases in sound. This is exemplified by the individual who asks you to repeat something, and then says, "You don't have to scream!" when you speak just a little louder. Recruitment is seen in most cochlear hearing losses and can be demonstrated with the SISI test audiometrically. In this test, the subject with a cochlear loss is able to hear "tiny" 1-dB increases in presented tones that the subject with normal hearing cannot. A simple explanation for this has not been given.

At any rate, the progressive hearing loss is a nuisance, along with the ringing, loss of speech discrimination, and noise sensitivity. What can be done? Obviously, with milder losses, **listening strategies** will help. Avoiding situations of background noise, paying attention to people's lips, and positioning closer to the speaker are all good recommendations.

Hearing aids have improved greatly over recent years. The new digital aids can be tailored to an individual's pattern of loss, selectively amplifying appropriate frequencies. They even dampen sounds that are too loud. Nonetheless, individuals vary in their tolerance to hearing aid use. Some situations of background noise will make amplification uncomfortable, no matter how well the aid is customized to the wearer. Patients with precipitous drops in the high frequencies are apt to have more trouble with hearing aids than patients with "flatter" losses on the audiometric curve. (Of historical interest, the old-fashioned "ear horn" worked quite well, except for the social stigma.)

Summary

Nothing can be done to prevent presbycusis, but hearing aids can help tremendously. Listening strategies are valuable as well. The patient might benefit from the reassurance that these losses progress very slowly and that "total deafness" is not an inevitable outcome. The primary physician can diagnose and advise a patient with presbycusis (as well as one with noise-induced loss, to be discussed next). If hearing aid(s) are a consideration, the patient should have an ENT consultation and recommendation. Of course, empathic clinicians should avoid saying, "You're just getting old."

Noise-Induced Hearing Loss

"How much of the hearing loss of aging is due to noise exposure?" Actually, this is a difficult question that often arises in litigation, when an aging patient is seeking compensation for his hearing loss from years of industrial noise exposure. Quite certainly, the two types of loss coexist in many individuals and one aggravates the other. However, characteristic audiometric differences are noted in the pure forms. The early loss from noise damage shows up as a **dip at 4 KHz** most often, with recovery at 8 KHz. Sometimes the loss is greater at 3 KHz or 6 KHz but, in any case, a V-shaped **"audiometric notch"** is present, and this deepens as damage increases (Fig. 7.**2**).

A typical scenario of acute noise-induced loss might be as follows. An adolescent sits in the front row at a rock concert for 4 hours, with the speakers blaring at 105 dB HL. He comes home with his ears ringing, and this continues for several days. If the hearing is tested in the acute phase, a 40 dB HL drop at 4 KHz might show up on the audiogram. With luck, he completely recovers in a week, with a normal repeat audiogram, although some undetectable hair cell damage may now exist. This

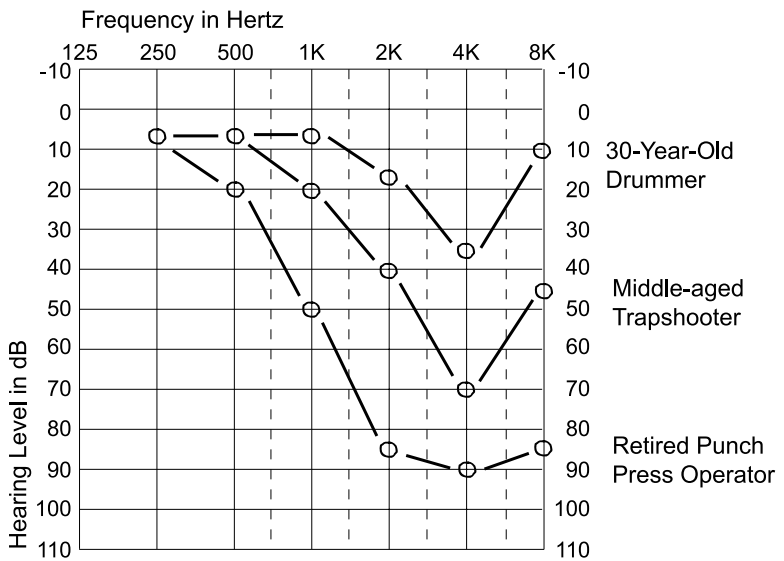

Fig. 7.**2** Audiogram showing examples of progression of noise-induced hearing loss, right ear.

occurrence is known as a **temporary threshold shift (TTS)**. The outcome can be worse. Even brief, but extremely loud, noises can cause unilateral or bilateral permanent loss. Recently, auto air bags deploying, with tremendous dB levels, have been responsible for a number of cases of permanent loss.

Chronic noise exposure, with its resultant damage, is dose related. However, individuals vary in susceptibility. Much research has been done on the levels of noise and duration of exposure needed to cause damage. Government regulators (i.e., OSHA) have developed exposure criteria requiring **ear protection** in the workplace. A few concepts are worth explaining. Hearing levels and noise levels are measured in decibels, which actually represent a logarithmic scale with respect to sound energy. An increase of 3 dB in noise level doubles the actual sound intensity. Also, the duration of exposure plays an equally important part. Hence, 96 dB HL of sound for 40 hours is considered twice as damaging as 93 dB HL for the same period. In other words, 5 years of work in a factory averaging 96 dB HL of noise causes the same damage as 10 years at 93 dB HL.

Research has shown that prolonged exposure to 90 dB HL or more may produce noise damage in some individuals, although the percentage of people affected is low. As the sound level and duration of exposure increase, so does the prevalence of damage. Many occupational settings have sustained noise at higher levels than 90 dB HL, and ear protection is mandated by law.

Preventing Noise Damage

The workplace is not, of course, the only source of potential noise-induced loss. Home power tools, chain saws, loud headphones, and firearms are other common menaces to hearing. When these devices are used, ear protection is the answer. (Obviously, the volume to one's headset should be kept down!) In-the-ear plugs and muff-type protectors are available at sports stores. They may be worn together to increase protection, although perspiration may be a problem. Noise levels might be decreased as much as 30 dB HL. For musicians who must hear without distortion, audiologists can fit specialized ear-protection molds that attenuate all the frequencies equally. It is important to advise the patient with chronic noise-induced loss that the damage is permanent and irreversible and emphasize the need to remedy the situation. The negative peak we see in early losses will deepen and widen over time if exposure continues, as shown in Figure 7.**2**

Nonorganic Hearing Loss

Occasionally patients will present with a complaint of hearing loss though, in fact, there is no loss. These individuals fall into two broad categories: those who consciously "fake" a loss (*malingerers*) and those who have psychological disturbances, usually of the *hysterical* type. The malingerers are often seeking compensation for some sort of injury, or have some other agenda whereby they will gain from their loss, such as a disability determination. The individuals with hysterical loss tend to be in the younger age group. These patients may be suffering from emotional stress and are not usually consciously shamming, although some secondary gain may be involved, as with other hysterical disorders. An office evaluation with simple clinical intuition and tuning fork testing may reveal inconsistencies with either type of patient, although the malingerers may be quite clever and convincing.

In either case, a good audiologist can perform special tests to prove and document the fictitious loss, whether unilateral or bilateral. The scope of this text precludes a detailed description of these procedures and how they work, but the **Stenger** and **Doefler–Stewart tests**, done with just a conventional audiometer, can catch the most skilled malingerers. **Auditory brainstem recording** (ABR) and **acoustic reflex measurements**, which do not depend on a patient's voluntary responses, can also be performed. ABR testing measures brainstem potentials in response to sound stimuli and is also a useful tool in the very young or mentally handicapped patient.

When malingering or hysteria is suspected, it is best for the primary physician to get early consultation from the ENT specialist, who can evaluate the patient before ordering the appropriate audiometric studies.

Congenital and Hereditary Disorders

Sensorineural hearing losses present at birth are congenital disorders. Some, but not all, of them, are **hereditary**, i.e., genetic, and the others are complications of pregnancy. Most are due to **cochlear defects**. There may be destruction of the vascular and membranous regions supporting the organ of Corti (Scheibe deformity) or loss of the neural elements, from the hair cells inward toward cranial nerve VIII (Mondini deformity).

It is important to discover congenital hearing loss as early as possible to promote rehabilitation and fit a hearing aid, if indicated. Speech awareness, the sooner the better, is critical for development of the auditory centers of the brain. An analogy with the eye exists: if a child with strabismus is not patched to train the nondominant eye, the visual cortex of the brain for that side does not develop. The auditory cortex is similar.

Congenital hearing losses are often suspected before 1 year of age. Unfortunately, many are not discovered until much later than this. Usually the mother another family member first notes a problem. It is then important for the clinician to heed the family's concerns and follow through with audiometric evaluation. An experienced audiologist can get a good idea about a potential loss with basic audiometric testing in very early infancy.

Excellent diagnostic information can be gained by **ABR**. This test measures auditory responses in the sedated infant, and characteristic curves can even pinpoint the problem to the middle ear, inner ear, or cranial nerve VIII. Also, **otoacoustic emissions** (OAE) tests are involuntary screening tests that identify spontaneous sound emissions from normally functioning cochlear hair cells. An increasingly large number of hospital neonatal units in the country screen all newborns using either ABR or OAE.

Predominant risk factors for congenital loss are:
1. Family history of hereditary loss.
2. Prematurity, with birth weight less than approximately 1.5 kg (3.5 lb).
3. Maternal rubella during pregnancy.
4. The presence of maxillofacial deformities.
5. Kernicterus, with bilirubin levels greater than 12 mg/dl.
6. Neonatal sepsis, especially with meningitis.
7. Forceps delivery, with temporal bone injury.

Hereditary, or familial, hearing loss may be present at birth, but also can develop during childhood or young adulthood. About 20% are reported as autosomal dominant, and 80% as recessive. The age of onset, severity, and audiometric patterns of loss vary greatly because many gene sites are possible. One pattern seen often is the "cookie-bite" audiogram, with an abrupt downward dip in the middle frequencies (Fig. 7.**3**).

Most of these losses do not involve syndromes with other organ systems, but a few do. Notable is *Waardenburg's syndrome*, an autosomal dominant disorder associated with widely displaced inner canthi of the eyes and a white forelock of hair. Another is *Usher's syndrome*, a recessive disorder with an eye problem, retinitis pigmentosa, as well. The severity of hearing impairment in these two is variable. In most familial disorders, the loss is bilateral. **Genetic testing** can now even localize and typify the gene involved.

There are syndromes of congenital hearing loss that do not involve the cochlea, but rather, the auditory nerve and more central pathways. **Auditory neuropathy** refers to an inability of the auditory nerve to send a good synchronous signal into the brainstem. These children have intact cochleas with normal OAEs, but abnormal ABRs. They can hear sounds of all frequencies, but fail to understand and develop speech. Kernicterus is a cause

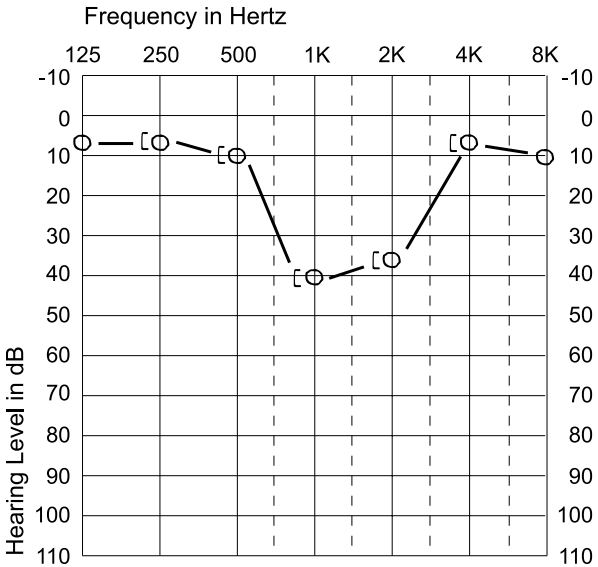

Fig. 7.**3** "Cookie-bite" pattern of hereditary hearing loss, right ear.

of some, but not all, cases. A number of afflicted individuals have other peripheral neuropathies, and there are probably genetic factors.

Some children have trouble with **central auditory processing**, a dysfunction somewhere between the brainstem and cortex. In general, they hear all frequencies normally and have good speech discrimination in quiet testing situations, but do poorly in situations of background noise or distractions. Their problems are milder than those of children with auditory neuropathy and are usually discovered as learning disabilities in school. They are then verified by specialized audiometric testing.

Treatment Summary
Great emphasis must be placed on the *early detection* of congenital hearing loss. Increasingly, hospital neonatal units are performing routine OAE or ABR screenings on newborns. Nonetheless, the medical practitioner should take seriously any concerned family member who suspects a loss and proceed with an audiology and ENT workup. Treatments for the disorders discussed above are tailored to the type of loss. Most mild to severe cochlear losses are best treated with early **hearing aid** fitting. Central auditory processing problems respond well to **FM listening devices** in the learning situation. These involve a microphone on the instructor and receiver earpieces on the student, eliminating background noise. Auditory neuropathy is seldom helped by aids, but instead by **visual cues** (cued speech, speech reading, or sign language) and possibly the **cochlear implant**, an electronic device surgically implanted in the inner ear by a very specialized otologic surgeon. The patient with profound hearing loss also has these last two options available. These treatments involve dedicated teams of audiologists, speech pathologists, and possibly specialized surgeons. They are mentioned only briefly in this text; the interested reader may look elswhere for further information.

Infectious Causes of Sensorineural Loss

When we speak of ear infections, we usually think of the middle ear and earaches. These are the good ones to have, because they seldom result in permanent damage. More ominously, the ones that involve the inner ear are likely to cause irreversible sensorineural loss. There are four general types of infectious diseases that may involve the labyrinth: systemic viral infection, meningitis, mastoiditis (with bacterial labyrinthitis), and syphilis.

Numerous viral infections, often occurring at a very young age, can wipe out the hearing. The damage is insidious; no ear pain is experienced. **Mumps** is probably the most notable. Often, the loss following mumps is unilateral, or worse in one ear. **Rubella** (German measles), rubeola (red measles), varicella (chickenpox), infectious mononucleosis, and the poliovirus have all been associated with unilateral or bilateral loss. Oddly, the lesions in the cochlea following these afflictions have a similar pattern of damage to that of the hereditary losses.

One viral illness behaves differently: **herpes zoster oticus**, or Ramsay-Hunt syndrome, discussed in the last chapter. This disease is clinically distinctive, being heralded by painful skin lesions showing up in the external

ear. It notably causes facial nerve paralysis, but it may also cause sensorineural loss by way of direct damage to the auditory nerve. The hearing loss that occurs here can spontaneously reverse, and treatment with high-dose **acyclovir** or **famcyclovir** may help.

Bacterial or viral **meningitis** has a high incidence of permanent damage to the labyrinth, especially in the very young. The meningeal infection spreads directly from the CSF into perilymph and causes severe labyrinthitis, with prolonged vertigo and permanent hearing loss. The damage may be unilateral or bilateral. Early treatment with intravenous steroids like **dexamethazone** may prevent this complication. In recent years, vaccines against *Haemophilus* and pneumococcus are decreasing the incidence of this, and other, ear complications.

Acute or chronic **mastoiditis** can penetrate the labyrinth. Cholesteatoma is the most common cause nowadays. The usual sequence of events begins with localized bone erosion of the lateral semicircular canal. This erosion may result **in circumscribed labyrinthitis**, symptomatic with positional vertigo and dysequilibrium. Fulminant bacterial spread throughout the labyrinth, known as **suppurative labyrinthitis**, may then ensue. This may also be a direct complication of acute coalescent mastoiditis. Here, the patient is severely ill with fever and vertigo. Profound permanent hearing loss and vestibular damage are inevitable, not to mention the threat of intracranial complications.

Syphilis (lues) is a rare infectious disease that can damage both the auditory and vestibular systems. The incidence is on the rise again, as is the incidence of other sexually transmitted diseases. It may be the congenital or adult type. The entire temporal bone, including the vascular supply to the labyrinth, becomes progressively infiltrated with perivascular lesions. The clinical picture is one of recurrent vertigo and fluctuating, but declining, hearing loss, mimicking Méniere's disease. An FTA–ABS serology test will confirm the diagnosis.

Summary
A hodgepodge of infectious conditions can invade the labyrinth and wipe out the cochlea and vestibule. The inner ear may resist penetration of middle ear problems, as we stated in the beginning of the chapter, but blood-borne infections can certainly penetrate it! Most of these are severe viral infections, and often they are seen in childhood. They tend to cause their rapid damage no matter what measures are taken. Thankfully vaccines have eradicated many of them in large areas of the world. Ramsey-Hunt syndrome deserves special mention, however, in that steroids and antiviral medications may help. Meningitis, mastoiditis with labyrinthitis, and syphilis are bacterial causes. Meningitis may also be viral and is more apt to cause hearing damage at a young age. Mastoiditis usually makes itself known with middle ear abnormalities, discussed in Chapter 5. Slowly progressive hearing loss with vertigo deserves a serology test to rule out syphilis, if your clinical suspicions are aroused.

Sudden Sensorineural Hearing Loss

This interesting phenomenon is not rare. A recent study estimates that there are 4000 new cases of sudden sensorineural hearing loss a year in this country. In my own practice, I have seen 10 to 20 cases a year in recent times. The term *idiopathic* is often used with this designation, although ongoing research supports various theories of causation. It is seen many times following viral respiratory infections, and occurs at any age, although the incidence increases greatly in the older population. No gender preponderance has been noted.

Many clinicians miss this disorder, mistaking it for a middle ear problem. Typically, a patient awakens with greatly decreased hearing in one ear. If it occurs while awake, there may be a "popping" noise, followed by roaring or humming tinnitus, with noticeable **sudden** loss of hearing. Dysequilibrium and vertigo are occasionally present at first, but they are usually transient, lasting an hour or less. The hearing, however, remains significantly decreased, along with persistent tinnitus. An important differential point in the history: autophony is not present because the loss is cochlear and not conductive.

On examination, the patient will have a normal TM, and tuning forks will show a substantial **sensorineural loss**, usually greater in the low frequencies. The Rinne should be normal at 512, and the Weber will lateralize to the uninvolved ear. If the fistula test (see Chapter 5) is positive, then there is a spontaneous perilymph fistula, eliminating this idiopathic diagnosis.

An **audiogram** should be done, and it will document the unilateral sensorineural loss. A downward-sloping (high-frequency) loss is seen occasionally, and it carries a worse prognosis than the usual low and mid-frequency loss. The clinical course is variable; 35% to 50% of patients have permanent loss, while the rest spontaneously recover. Certain factors influence the outcome. Old age, severe or high-frequency hearing loss pattern, persistent vertigo, diabetes, and hypertension all worsen the prognosis.

Theories of causation include viral, vascular, and autoimmune disease. Méniere's disease (differing from sudden sensorineural loss by its recurrent nature) and spontaneous perilymph fistula (often barotraumatic) must be eliminated. Most investigators conclude that no single cause exists, although most recent evidence points to a viral cause. It is proposed that there is viral disturbance of the microcirculation of the cochlea with vasospasm and blood sludging.

Numerous remedies have been used for this disorder, even including hyperbaric oxygen. However, **steroid therapy**, with or without additional remedies, has been shown in a number of studies to improve the outcome. Patients who present for treatment earlier (within 10 days from the onset of symptoms) have much better outcomes than those who present later on. Treatment recommendations from recent review articles include:

1. Oral steroid taper for at least 10 days.
2. Acyclovir 200–400 mg five times a day for 10 days.
3. Mild diuretics, such as hydrochlorthiazide.
4. Vasodilators, such as niacin 200 mg bid.

Summary
Without clinical acumen and hearing evaluation, sudden sensorineural hearing loss may be missed. Many times it is confused with ET and middle ear disorders. If we are aware of this entity, listen to the patient, examine the TM for mobility, and use our tuning forks, we can pick it up. An audiogram will then confirm it. Early diagnosis and treatment may help the outcome. The primary practitioner should recognize it early and treat it immediately with steroids and antivirals. An ENT physician can follow up.

Méniere's Disease

It may surprise you that we discuss this topic under "auditory" disorders, because the vertigo of Méniere's disease is the most notable symptom to the majority of clinicians. Actually, in the earlier stages, most of the patho-

logic process occurs only in the cochlea. This ailment is characterized by four classic symptoms: **hearing loss, tinnitus, ear fullness**, and **vertigo**. These symptoms are produced by **endolymphatic hydrops**, another name for the disorder, which is pathologically descriptive. To add to the confusion, another name, **Lermoyez's syndrome**, describes an earlier or milder form of it where only the cochlea has hydrops, vertigo is absent, and the hearing loss fluctuates.

In this disease, a localized disorder of circulation or electrolyte balance causes swelling of the endolymph-containing channels within the cochlea and vestibular apparatus. It has been proposed that the problem results from underlying metabolic, allergic, viral, or autoimmune disease. The onset of symptoms usually occurs in the thirties or forties, with no gender predominance. Eighty percent of the time, the disease is unilateral, and if the second ear becomes involved, this will occur within a few years of the first.

Typically, the first episode involves a sense of pressure in one ear accompanied by a low-pitched roaring or blowing sound. Slight hearing loss may be noted. This phase may last hours or days, and in some cases it may disappear, only to return again (Lermoyez's syndrome). In full-blown Méniere's disease, however, a violent attack of vertigo, usually with nausea and vomiting, follows the onset of the ear symptoms by a day or two. This may last several hours and then gradually subside. The vertigo usually abates by the next morning, although mild ear fullness and tinnitus may continue longer.

From here on, the clinical course is extremely variable. Recurrences may be frequent, infrequent but clustered, or sporadic. Over the years, the severity of the vertigo tends to decrease or become better tolerated, but the hearing often shows a downward trend. Early on, the hearing returns to normal after each bad spell, but eventually the drops linger, and severe to profound loss may be seen late in the disease. Some individuals never have severe vestibular symptoms, and others may have vertigo for more than a full day during a spell, with lingering dysequilibrium afterward.

On examination, there will be a normal drum, and tuning forks will show a unilateral **sensorineural loss**. The interesting phenomenon of **dyplacusis** will be seen: the same tone, say a 512 tuning fork, is heard as a different frequency in each ear. Presumably, this is due to the hydrops "stretching" the basilar membrane, changing cochlear tuning on the affected side. When vertigo is present, nystagmus may be seen, with the fast component beating toward the involved ear during an acute spell.

The audiologist is a valuable consultant here. **Audiometry** shows several characteristic findings, besides the expected sensorineural loss. The hearing loss often, but not always, predominates in the **low frequencies**

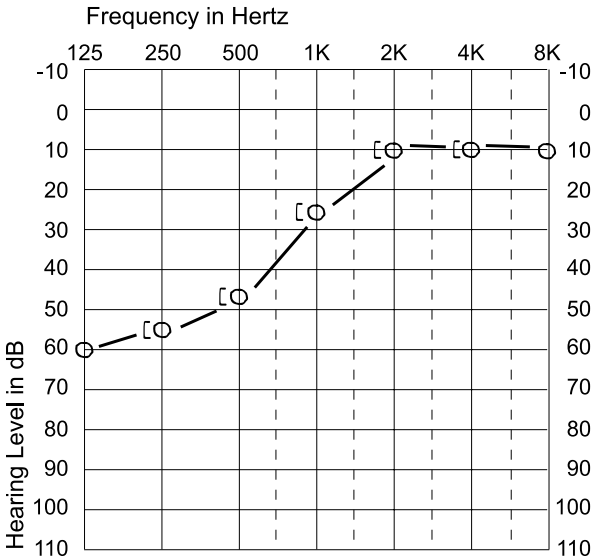

Fig. 7.**4** Hearing loss of early Méniere's disease, right ear.

(Fig. 7.**4**). Recruitment, mentioned in the presbycusis discussion, is demonstrated by a positive SISI test. The patient's labyrinthine function can be assessed by electronystagmography (**ENG**), which will be discussed in the next chapter.

A diagnostic workup regarding contributing factors is good practice. Possible metabolic and allergic problems, mentioned previously, should be investigated. **Thyroid function** and **glucose tolerance** tests are recommended by many. Hypothyroidism and diabetes have been associated with the disease. **Allergy testing** may pick up an abnormality. In addition, an **FTA–ABS** should be done to rule out syphilis, which can closely mimic the disorder. A serologic test for **autoimmune ear disease**, namely the 68kD antigen (see the next section), might also be done. A **lipid profile** may show hyperlipidemia, another possible contributor.

Various treatment modalities have been used for this disease. Some help the immediate symptoms; others alleviate it on a long-term basis. Some recommendations are:

1. A low-salt diet.
2. Daily diuretics (taking care to replace potassium if needed).
3. Meclizine, or other antihistamines, for the acute vertigo symptoms.

4. Tapering steroids for a flare-up, presumably treating an autoimmune or allergic cause.
5. Antianxiety medications, such as Xanax or Valium, to suppress the labyrinth and central vestibular centers.
6. Vasodilators such as niacin, used empirically to help blood flow, and thus fluid exchange, in the inner ear.
7. Bioflavonoid, presumably to improve labyrinthine circulation, in combination with vitamins.

Of the above treatments, the first three are most often recommended. In intractable cases with relentless vertigo, surgery may be performed. Recent research has shown that the vertigo may be reduced by **intratympanic injections** of **gentamycin**, a vestibulotoxic drug that diffuses through the round window membrane. The **endolymphatic shunt** operation can be done through a mastoid approach to decompress the endolymphatic sac. With either of these procedures, there is a risk for further hearing loss, but benefit for the disabling vertigo. If the hearing is totally gone and vertigo continues, **ablative procedures** that destroy the labyrinth or vestibular nerve may be performed.

Summary

Much research has been done on the cause and treatment of Méniere's disease. Clinical recognition of the entity rests on the *four* symptoms that should be present to make the diagnosis: *sensorineural hearing loss, tinnitus, aural fullness,* and *vertigo.* A key clinical point is that the ear symptoms during an attack are *unilateral*. The dizzy patient with vague *bilateral* ear symptoms probably does not have Méniere's disease! The fluctuating nature of the cochlear hearing loss, along with the violence of the vertigo, favors this diagnosis, as does the symptom of ear fullness. The hearing loss is fairly mild in the early stages of the disease, but increases with time. The periods of true vertigo usually last a few hours to a day. They tend to lessen in severity over the long run. Rarely, the second ear may become involved months or years later, which then favors the diagnosis of autoimmune inner ear disease, to be discussed next.

The astute and willing primary practitioner can diagnose this disorder, treat flare-ups, and pursue the blood tests we discussed for underlying causes. An ENT specialist should also be involved.

Autoimmune Inner Ear Disease

Autoimmunity is the cause of a number of cases of progressive bilateral sensorineural deafness. Since the mid-1980s, research has uncovered increasing evidence that this process may be responsible for **progressive sensorineural loss** in both ears, with or without vertigo, which is **steroid responsive**. Usually, one ear is involved first, then the other in a matter of weeks, months, or years. The symptoms in either ear may mimic Méniere's disease, with all four typical complaints.

A classic proven autoimmune disease involving the ear has been described for years, namely, **Cogan's syndrome**. This disorder consists of interstitial keratitis with vestibuloauditory dysfunction. Patients have severe eye involvement along with rapid deterioration of one or both inner ears over a period of just a few months. Some may develop polyarteritis nodosa, and studies of affected individuals show antibodies against the sclera and inner ear. Other systemic autoimmune diseases, such as Wegener's granulomatosis and lupus erythematosus, have also shown inner ear involvement.

However, a number of patients with no evidence of systemic disease have inner ear disease and show **positive serology for autoimmune disease** and respond to steroids. One study cites case reports with similar clinical histories. The typical individual had a sensorineural hearing loss in one ear that was fluctuating or progressive, sometimes with vertigo. Then, after a few months or years, the second ear developed a progressive loss, and by this time the loss in the first ear had usually become severe. The involvement of the second ear evoked clinical suspicion of autoimmunity, and thus serologic test batteries were done. The Western blot assay for inner ear antigen 68kD was often positive. Other more routine serologies, such as the antinuclear antibody (ANA), were occasionally positive as well. Serologic assay for inner ear antigen 68kD is available at several specialized commercial laboratories in the country.

When these study patients were given steroids, most recovered some hearing. Usually, 60 mg of **prednisone** was given daily for several weeks and then tapered to a low or moderate dose for a long period. In some patients, if the steroids were stopped, the hearing would drop off again. Other individuals were intolerant of prolonged steroids. Chemotherapy agents, notably cyclophosphamide (Cytoxan), were then used, which were often successful in maintaining the recovery of hearing.

Recently, **intratympanic injections of steroids** (dexamethasone or methylprednisolone) have been used for autoimmune hearing loss. This treatment is also being studied for sudden sensorineural loss, mentioned earlier. Further studies are underway regarding this relatively new procedure for both disorders.

Summary
The hearing loss and vertigo of autoimmune hearing loss may mimic Méniere's disease. A high index of clinical suspicion, usually prompted by *second ear involvement*, is needed to diagnose this disorder. Workup, after examination and audiometry, centers on serology. The antibody titer against *inner ear antigen 68kD* may be assessed in a few specialized serology laboratories. *Steroid responsiveness* also helps to make the diagnosis. Prolonged steroids or Cytoxan are reported as effective treatments. When the primary practitioner suspects and discovers this disorder, early ENT and even rheumatology consultation should be obtained.

Cerebellopontine Angle Tumors

The tumors discussed here are located in or near the internal auditory meatus, where cranial nerves VII and VIII exit the petrous temporal bone to enter the brainstem. **Acoustic neuroma** is the predominant one in this category, comprising about 85% of these tumors. Meningiomas, neurofibromas, and gliomas make up most of the rest. A rare congenital cholesteatoma may also appear as a space-occupying lesion anywhere in the temporal bone. The more accurate term for acoustic neuroma is **vestibular schwannoma**, because it usually arises from the sheath of the vestibular portion of cranial nerve VIII. This benign, slow-growing tumor is usually discovered in middle age, although it may have been evolving over years. It is most often unilateral, although it may be bilateral in individuals with neurofibromatosis.

Patients with acoustic neuroma have slowly progressive hearing loss and dizziness. **Dysequilibrium** (unsteadiness and poor balance) is experienced more than real vertigo. In the early stages, examination will uncover almost nothing except a **high-frequency hearing loss**, and this may only show up on an audiogram. A striking early finding, however, is abnormally **poor speech discrimination**, even when the hearing loss is fairly mild. Thus, the patient is apt to complain of hearing distortion, even though sounds seem loud enough. As the tumor grows, the hearing loss worsens, and pressure on the adjacent cranial nerve V may cause facial numbness, with characteristic **loss of the corneal blink reflex.** Compression of cranial nerve VII may cause facial paresis. All the while, there is poor balance, but not severe vertigo, even though the vestibular nerve is the original focus of growth! Very rarely, however, the tumor may hemorrhage and swell acutely, producing sudden hearing loss or vertigo, as with Méniere's disease or sudden sensorineural loss.

Special audiometric testing, including ABR, will strongly point to the diagnosis, but **MRI** is the gold standard for finding the smallest of lesions. We alluded to special audiologic tests earlier—**tone decay**, along with **acoustic reflex decay**, are important ones. They may be performed easily in an audiologist's booth and are specific for lesions of cranial nerve VIII. They are extremely cost effective compared to MRI, but not quite as reliable.

Traditionally, treatment has been **surgical removal** of the tumor. This can often preserve the hearing, but unfortunately there is some risk to a yet undamaged facial nerve. Improved operative techniques have decreased this risk, especially when the tumor is small. In recent years, radiation treatment via the **linear accelerator** or the **gamma knife**, has become available. For smaller tumors, these non-surgical treatments may be curative, or at least palliative over the long term. Finally, acoustic neuromas are so slow-growing that some older or chronically ill patients may opt for observation, with periodic scans to assess growth.

Summary

In growth and clinical presentation, acoustic neuroma is insidious. Typically, this tumor presents with a slowly-developing unilateral high-frequency loss with excessive distortion of sounds, and disequilibrium without vertigo. Audiometry, including the special test of tone decay, can lead to the diagnosis. MRI findings are characteristic. If the diagnosis is confirmed, referral to a subspecialist in neuro-otology should be the next step. Modern advances in radiotherapy have effected cures for smaller tumors.

Chapter Summary

In closing this presentation of the types of sensorineural hearing loss, let us emphasize that there is obvious overlap between some of the disorders we have discussed, especially with respect to symptoms. This applies to Ménière's disease, sudden sensorineural hearing loss, infectious sensorineural loss, autoimmune loss, spontaneous perilymph fistula (Chapter 5), and acoustic neuroma. Of interest, any of the categories of medical disease may be involved in causing a sensorineural hearing loss, including trauma, neoplasm, allergy, infection, heredity, autoimmunity, metabolic disease, and nutrition.

The important diagnostic point for us, as clinicians, is to determine whether or not the hearing loss is *sensorineural*. Patients with this type of loss should not be treated for a middle ear or eustachian tube problem.

Clinical knowledge and careful examination, with tuning fork evaluation, will avoid such misdiagnoses.

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8 Vestibular Disorders of the Inner Ear

We saved a discussion of vestibular disorders for last, and this is certainly not a popular way to conclude a textbook. Most clinicians know the sinking feeling when an individual comes into the office complaining of dizziness. The patient's history is often vague and hard to follow, yet despair and frustration are plainly expressed. Well, take heart—the truth will help to set you free. The more we know about dizziness and its possible causes, the easier it is to get things categorized and formulate a diagnosis. There is a finite number of well-described disorders of the vestibular system.

As caregivers, we must not forget that any type of dizziness can be very frightening to the patient; sympathy and understanding are in order while we try to obtain a history that can be hard to pin down. Localizing the cause of a patient's complaints to the vestibular system (the inner ear and its connections in the brainstem), or away from it, is a diagnostic priority. This chapter, along with a few sections in Chapter 7, describes the true vestibular disorders. A discussion of different types of dizziness precedes this.

Vertigo

The clinician should find out what the dizzy patient is experiencing, as accurately as possible. *A common clinical error is to label all dizziness as "vertigo."* Is there a sense of **motion**, either of one's own body or of one's surroundings? If so, true **vertigo** exists. The history in this case is usually clearer than with other forms of dizziness. The patient often vividly recollects when and where the first episode occurred, describing the spinning sensation and, usually, the accompanying **nausea**. If this is not the picture, one should ask the patient to describe the symptoms without using the word "dizzy." A good question is: "Does it feel like you or the room is moving or going around?" If an affirmation confirms that vertigo is indeed present, the problem almost certainly arises from the vestibular labyrinth or its connections in the brainstem.

Whether or not there is true vertigo, other questions regarding the patient's symptoms are in order. The circumstances of the onset of the first episode, its length, precipitating factors, and patterns of persistence

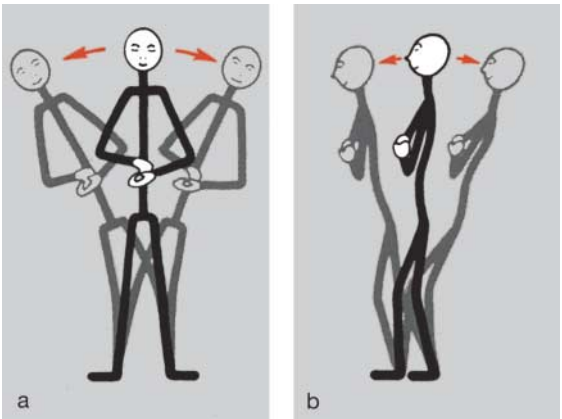

Fig. 8.**1a** and **b** The Romberg test. The patient is instructed to stand erect with the eyes closed, the feet slightly apart, and the arms folded. A peripheral labyrinthine lesion will cause swaying to the affected side (**a**); a central disorder is more likely to cause random imbalance in any direction (**b**).
(Source: Becker W, Naumann HH, Pfaltz CR. *Ear, Nose, and Throat Diseases.* Stuttgart: Thieme; 1994)

or recurrence all help point to a possible diagnosis. Are ear symptoms present?

Physical examination of the dizzy patient should include an assessment of balance and extraocular motion, as well as of the ears and hearing. One should look for nystagmus, especially during position change. The Hallpike maneuver is discussed later in this chapter. The Romberg test (Figs. 8.**1a** and **b**) should be done. With peripheral vestibular lesions, the patient tends to sway to the side of the diseased labyrinth, whereas in central balance disorders, the direction of sway is random. The sections on specific vestibular disorders discuss physical and test findings in greater detail. The most important diagnostic tool, however, is an accurate history taken by a caregiver who is aware of the disease entities to be discussed.

Disequilibrium

An important fact is that vertigo is the usual, but not the only, symptom pointing to the vestibular system. **Disequilibrium** (unsteadiness or loss of balance), without vertigo, can also be a sign of more chronic, insidious problems. In young and middle-aged patients with this symptom, certain specific diseases of the vestibular system should be ruled out. An acoustic

neuroma, chronic luetic labyrinthine disease, or even demyelinating disease in the brainstem (i.e., multiple sclerosis), may be present.

In very elderly people, dysequilibrium is more likely to indicate nonspecific multisystem deficiency, including poor circulation, poor vision, and generalized neurologic deterioration. An important clinical point should be made here. A common treatment error often occurs with this category of patients: many physicians prescribe meclizine or a similar drug. This antihistamine will not help a degenerating vestibule, but will cause drowsiness! Be advised that meclizine is intended for nausea and true vertigo, not imbalance.

Nonvestibular Dizziness

Balance and spatial orientation depend on three main systems: the vestibular apparatus, vision, and proprioception (position sense from the spine and pressure sensors of the extremities). Dizziness may result from problems involving any one, or more, of these systems. Furthermore, disease processes of other bodily systems, such as circulation, respiration, cerebration (including anxiety), and endocrine function, may cause dizziness by affecting the former ones! Numerous types of nonvertiginous dizziness may occur, and the more notable types will be discussed briefly. Again, emphasis should be placed on eliciting an accurate descriptive history from the patient.

A patient may be "lightheaded," feel vaguely disoriented, have visual blurring, become faint, lose consciousness (experience syncope), or have disequilibrium. Certain positions or activities might bring on the symptoms. Anxiety, with hyperventilation, may be present. Postural changes, moving the neck, or using the arms may be precipitating factors. The time line is also important. Some patients are vaguely dizzy around the clock for days, and others have symptoms during more specific time periods. These historical points will help to narrow down the differential diagnosis.

When a patient experiences **syncope**, end-organ inner ear disease can be ruled out; do not call the ear specialist first. Peripheral labyrinthine disease often puts the patient to bed with sickening discomfort, but it will never, by itself, cause loss of consciousness. Real syncope results from impaired oxygenation of the brain, by way of vascular compromise, blood pressure drop, hypoxemia, or cardiac arrhythmia. Faintness, or near-syncope, is closely related.

A workup of older syncopal patients should include Holter monitoring and blood pressure readings, both supine and erect. Postural hypotension may be occurring. Brainstem ischemia, cerebrovascular disease, or subcla-

vian steal syndrome might also be the cause. With younger patients, a good history may reveal that just anxiety, with resulting hyperventilation, caused the event. Look for sudden psychogenic vasovagal reactions as well, which may even be accompanied by seizure-like activity.

Blurring of vision does not relate to the vestibular system, and patients with this complaint deserve an eye workup. Other symptoms may be less specific. General disorientation or lightheadedness may be a sign of metabolic disease, for example, diabetes, nutritional disorders, or hypothyroidism. The more vague the symptoms and their time-line become, the more likely it is that stress, fatigue, or just plain psychiatric illness is the cause. However, a neurologic lesion may be the underlying cause. Certainly, when there is doubt about a source of dizziness, the urge to document things with a CT scan or MRI comes into play. The expense is great, but the reassurance value might make it worthwhile. Medicolegal considerations often oblige us to perform these expensive but reassuring studies.

We have mentioned some of the nonvestibular causes of dizziness. The purpose of this last chapter, however, is to describe disorders of the vestibule (utricle, saccule, and semicircular canals), vestibular nerve, and vestibular nuclei in the brainstem. A key point regarding these areas is that their diseases produce either **vertigo** or **disequilibrium**.

Nystagmus and Electronystagmography

Nystagmus, a rhythmic twitching of the eyes, is the only good physical clue that vertigo is taking place. It is not always obvious, so the patient or family members are not likely to report it. However, close physical examination during a spell of real vertigo should show it. Nystagmus is often horizontal, but it can also be rotary, or rarely, vertical. Certain positions may cause it, especially in benign paroxysmal positional vertigo, the most frequent cause of vertigo.

The physiologic explanation for nystagmus is complex. When something goes wrong in the vestibule, either from a disease process or an artificial stimulus, a message is sent to the oculomotor centers in the brainstem. In turn, these send an adaptive message to the eye muscles. A typical artificial stimulus would be spinning in place, as a child might do, sending the fluids of the semicircular canals into motion. Sudden cessation of the spinning will then result in classic vertigo with nystagmus. Similarly, a caloric stimulus, such as cold water in the ear, creates convection currents in the fluid of the lateral semicircular canal. In either case, the subject's neural pathways perceive a false sense of motion, and the vestibular-oculomotor connections cause the pupils to track sideways, with quick corrections in the opposite direction. This manifests itself as the "beating" nystagmus we

observe, with slow motion of the pupils in one direction and fast "beats" in the other.

All sorts of disorders can create *pathologic* nystagmus, but the above-mentioned stimuli, like spinning or caloric stimulation, should produce nystagmus as a *normal* response. Also, be aware that when one examines a normal patient's ocular motions, transient fatiguable nystagmus is often seen on lateral gaze to either side.

Electronystagmography (**ENG**) has been available for years. It involves measurement of nystagmus by placing electrodes adjacent to the eyes and recording ocular motion under varying circumstances. As mentioned above, abnormal nystagmus may occur in disease states, but normal nystagmus should be present with caloric stimulation to each ear, with symmetrical responses. Experience with ENG testing over the years has accumulated a vast body of knowledge, and as a result, a fairly standard test battery has been developed. Certain findings may point to more central brainstem pathology, and others to peripheral inner ear disease. Often, there are specific patterns for specific diseases. In the ENG test procedure, the electrodes sensing eye motion are placed on either sides of both orbits, as well as above and below, to record nystagmus in either axis. The impulses received from these ocular motions are measured and recorded, and patterns of disease states can emerge and be described.

The ENG assesses ocular motions under several circumstances. The first is with the patient sitting still and looking straight ahead. Abnormal **spontaneous nystagmus** may be seen in this neutral position. Next, the subject watches certain visual stimuli without moving the head. Here, "tracking" abnormalities relating to optic–vestibular–ocular connections are assessed, picking up possible problems in the central brain and brainstem, or even drug-induced disorders. The next set of tests involves moving the patient's head, or head and entire torso, into various positions to look for **positional nystagmus**. Here, certain disease states of the central pathways or peripheral inner ear may appear. The last test measures responses to **caloric stimulation**, using cold or warm air or water introduced into the ear canal. Body-temperature air or water elicits no convection currents in the semicircular canals, and thus no nystagmus. With cold or warm stimulation, normal ears will show symmetrical healthy nystagmus responses (although the subject has temporary vertigo). Absent or decreased responses in one or both ears, indicate pathology in the vestibular labyrinth(s).

When specialized and well-trained audiologists perform these tests, they can give us a great "read-out," even suggesting possible diagnoses. Figure 8.**2** shows an example of an audiologist's ENG report on a patient with significant findings.

Patient:	Jane Doe
Birthdate:	12/20/57
Date of Evaluation:	05/27/02
Physician:	Dr. Menner
referral reason:	Mrs. Doe has continued to experience feelings of dizziness since March 2000. The sensation is heightened by movement or chance in head position. ENG administered on 4/24/00 revealed a significant right unilateral weakness. Audiological test results today revealed a right moderate low-frequency loss.
Procedures:	Bithermal air calorics, saccade, tracking, optokinetic, gaze, and positional tests and Hallpike Maneuver.
Results:	—The caloric response of the right ear was 86% weaker than of the left ear. —The patient readily supressed the induced caloric nystagmus with visual fixation. —The results of the saccade, tracking, and optokinetic tests were within normals limits. —There was no nystagmus. —There was no nystagmus in the sitting, supine, head-right, head-left, right lateral, or left lateral positions. —The results of the Hallpike Maneuver were negative, i.e., no nystagmus was elicited following either the head-down-left maneuver.
Impression:	Abnormal ENG. The right unilateral weakness is consistent with right peripheral vestibular pathology involving the right labyrinth or vestibular nerve

Fig. 8.**2** Example of an ENG report.
(Source: Sheila Giovannini, Southern Tier Audiology, Elmira, N.Y.)

Posturography deserves brief mention. This is a newer form of balance assessment that evaluates spinal and peripheral mechanisms, as well as the vestibule. It is available at many centers and a database is building, but ENG is still the preferred test for vestibular assessment. Now, we will discuss clinical disease entities that cause vertigo.

Benign Paroxysmal Positional Vertigo

By far the largest number of patients with true vertigo suffer from **benign paroxysmal positional vertigo (BPPV)**. It was first described many years ago by Dix and Hallpike, but Dr. John Epley has re-investigated it more recently. He has developed a reliable curative procedure that can be done in the office. It has been found, with proven anatomic evidence, that the vertigo of these patients is due to otolithic debris (now termed canaliths) in the long limb of the posterior semicircular canal. These are displaced otoliths that are normally present over the stereocilia of the position-sensing hair cells in the utricle. Head trauma, or other less clear causes, put them in the wrong place. They create a physical "drag" on fluid motion in this semicircular canal, and the result is the classic symptom-complex seen in this disorder.

A typical patient complains of **vertigo lasting a few seconds** to a half-minute when the head **position changes** with respect to gravity. This usually occurs when lying down, arising from bed, looking upward, or bending forward. When supine, it is experienced when the individual rolls over to one specific side. A history of head trauma may exist, but not necessarily. The symptoms are often relentless, and over a period of time the patient learns to avoid the positions most likely to cause the vertigo, or else moves with great care in certain directions. There is **no associated hearing deficit**.

The **Hallpike maneuver**, a rapid shift from sitting to supine with the head hyperextended and turned 45° to the affected side, will produce vertigo and rotary nystagmus. Usually the nystagmus comes on after a lag period of a few seconds, and then abates after 30 seconds or less. This diagnostic maneuver can be done in the office. **ENG**, if using this maneuver during positional testing, will document the finding. The disease is usually unilateral, and the ear that is downward when the nystagmus occurs is the culprit.

Prior to the procedure developed by Epley, treatment consisted of reassurance and meclizine or the like, which did very little for this specific disorder. The clinical course remained chronic and recurrent. Now, there is the canalith repositioning procedure, a fairly simple manipulation designed to get the debris out of the posterior canal. This is often effective after the first try, and the success rate improves with repetition. Figure 8.**3** demonstrates the **Epley procedure**.

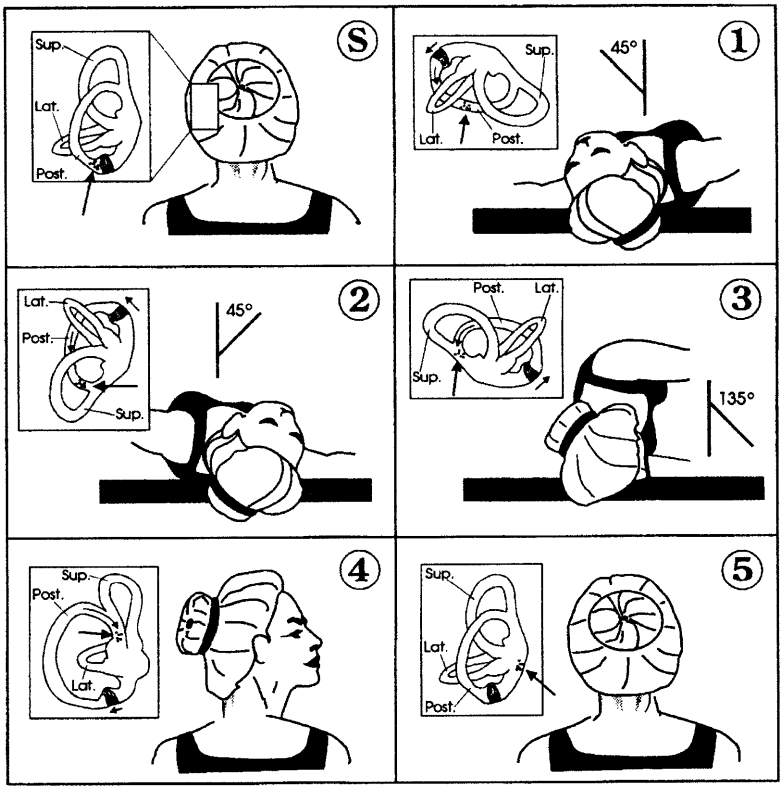

Fig. 8.**3** The Epley canalith repositioning procedure.
Positioning sequence for left posterior semicircular canal, as viewed by operator (behind patient). *Box*, Exposed view of labyrinth, showing migration of particles (*large arrow*). *S*, Start-patient seated (oscillator applied). *1*, Place head over end of table, 45 degrees to left. *2*, Keeping head tilted downward, rotate to 45 degrees right. *3*, Rotate head and body until facing downward 135 degrees from supine. *4*. Keepng head turned right, bring patient to sitting position. *5*, Turn head forward, chin down 20 degrees. Pause at each position until induced nystagmus approaches termination, or for T (latency + duration) seconds if no nystagmus. Keep repeating entire series (*1–5*) until no nystagmus in any position.
(Source: Epley JM, Particle Repositioning for Benign Paroxysmal Positional Vertigo. *The Otolaryngologic Clinics of North America*. 1996;29:2:327.)

Summary
Benign paroxysmal positional vertigo is the most common syndrome of vertigo. It is characteristically recognized by the symptom of true vertigo when lying down with the head turned to one side. Other position changes may also precipitate it. There are no other ear or hearing symptoms. A unilateral vestibular abnormality has been demonstrated, and Epley's well-described maneuver is often curative. The primary practitioner can diagnose it in the office by using the Hallpike maneuver, or by referral for ENG testing to document the nystagmus. The Epley maneuver can then be performed. If the clinician has any doubts, ENT consultation can be obtained.

Vestibular Neuronitis

Vestibular neuronitis is characterized by a sudden onset of severe vertigo without any auditory symptoms. Often following a viral infection, it is thought to be due to a unilateral postviral neuropathy of the vestibular division of cranial nerve VIII. There is some ambiguity in the literature regarding the terminology for this syndrome. It overlaps with *acute viral labyrinthitis*, *toxic labyrinthitis*, and *epidemic labyrinthitis*. These ailments generally behave the same way, but the cause may vary. Certainly, toxic labyrinthitis implies a nonviral cause: alcohol, drug ingestion, or occasionally, inhaled substances.

The clinical course of this disorder is characteristic, and similar to the other "labyrinthitis" labels. Often, there has been an upper respiratory infection or sinusitis 1 or 2 weeks before the attack. Frequently, the individual awakens from a sound sleep in the morning, or even in the middle of the night, with **sudden violent vertigo**. There is nausea and vomiting. The patient may state that she or he had to crawl to the bathroom to vomit due to the severity of the dizziness. There are **no specific ear symptoms**; tinnitus, hearing loss, or aural fullness is not experienced. Most of the time, the patient must spend a day or two in bed. Then, the symptoms start to subside, although **positional vertigo** may remain for a period of weeks, gradually abating with time. A month or more may elapse before things return to normal. Affected individuals may have rare recurrences during their lives. If there are multiple recurrences, other syndromes or diseases should be suspected. Recovery is usually complete.

During the acute stage, the patient will show **nystagmus**. In acute irritative lesions of the labyrinth such as this, the nystagmus usually beats (fast component) toward the diseased ear. ENG can document the spontaneous nystagmus at this time. Caloric tests will show diminished re-

sponses in one ear, demonstrating unilateral labyrinthine weakness. Audiometry shows **no hearing loss** in either ear.

Treatment of this disorder involves bed rest and the use of antivertigo medications such as **meclizine** (Antivert). **Promethazine** (Phenergan) or other antiemetics may also be used. The **antianxiety drugs** also have a place in treating all types of vertigo; they directly suppress the labyrinth while relieving emotional stress. As the term **toxic labyrinthitis** suggests, a potential toxic insult should be searched for. Alcohol, barbiturates, lead, arsenic, quinine, Dilantin overdose, or ototoxic antibiotics might be contributing agents. I have personally seen several patients who had developed this symptom pattern after inhaling the solvents of paint, stains, or adhesives in an unventilated area.

Summary

Vestibular neuronitis is probably a postviral inflammation of the vestibular nerve or its end organs in the labyrinth. This disorder is clinically similar to what used to be termed *viral labyrinthitis*. It presents with sudden vertigo, but no ear symptoms. Symptomatic treatment is indicated, and gradual resolution can be expected. Toxic causes for the symptom complex should be ruled out. The primary practitioner will often find the affected patient in the Emergency Room, and may consider hospitalization for a day or two. The ENT physician might be consulted for backup.

Ménière's Disease, Revisited

The four classic symptoms of true Ménière's disease are described in Chapter 7. Possibly, there is a limited form whose symptoms involve only the vestibular labyrinth. Some deny its existence. Nonetheless, there are occasional patients who have recurrent vertigo and aural fullness, lasting a few hours to a day, without any hearing loss or tinnitus. The vertigo is not positional. The time line and tendency for recurrences are similar to classic Ménière's disease, and these patients may respond to the usual treatments. A descriptive name for this disorder is **vestibular Ménière's syndrome**.

To add to the controversy, there may be a linkage between this syndrome and migraine headaches. Numerous reports show an increased prevalence of migraines in patients with both classic and vestibular Ménière's syndrome. It has been proposed that the cause involves spasm of the basilar artery, and there is probably a genetic predisposition, as with migraine headaches.

Cervical Vertigo

Patients with cervical vertigo experience their symptoms when the neck is turned in one or more directions. Three causes may come into play in this syndrome: *impaired vertebral artery circulation, cervical osteoarthritis*, and *whiplash injury*. When the vertigo is experienced on neck torsion, there may be mild nystagmus. Consistent ENG findings have not been precisely described in the literature, although head-turning may elicit measurable nystagmus. The pathophysiology for this type of vertigo is not absolutely certain, but it is likely due to impairment of circulation to the vestibular centers by way of vertebral artery compromise.

Affected patients often give a typical history. The vertigo occurs only on turning the head, usually in just one direction, regardless of overall body position. When whiplash injury (typically from vehicular accidents with severe neck flexion and extension) is the cause, the vertigo is worse when turning the head to the more painful side. Here, the symptoms slowly subside after a few months if there is no permanent cervical spine injury. Very prolonged cases of whiplash vertigo may mysteriously improve after successful litigation.

Vascular Disorders

Circulatory problems were discussed in the opening section as a cause of faintness and syncope. Some specific vascular disorders also cause real vertigo. **Vertebrobasilar insufficiency** (VBI) is one; it may cause recurrent acute vertigo, usually in response to postural changes or anything producing a decrease in blood flow to the brain. Ischemia to the vestibular nuclei is the cause. Most typically, the vertigo occurs when rising from a horizontal position. In severe cases, accompanying neurologic symptoms related to speech, vision, and sensation of the face or extremities may be present. ENG findings will document types of nystagmus that point to central (nonlabyrinthine) disease.

Wallenberg's syndrome is due to vascular occlusion of the posterior inferior cerebellar artery. In addition to vertigo from ischemia of the vestibular nuclei, there are other specific symptoms referable to associated lesions in the medulla. Ataxia, dysphagia, Horner's syndrome, with loss of pain and temperature sensation of the face and opposite side of the body, accompany the vertigo.

Subclavian steal syndrome, due to narrowing of the arteries to the upper limbs, causes shunting of blood in a retrograde fashion from the brainstem during vigorous use of the arms. This may result in vertigo, or even syncope.

In general, when vascular compromise is suspected, medications that impair platelet function may improve things. Short of full **anticoagulation**, daily aspirin may help, if not contraindicated. Some patients balk at this recommendation due to gastric symptoms, but it is unlikely that one grain a day would bother anyone, barring true aspirin allergy.

Circumscribed Labyrinthitis

Circumscribed labyrinthitis may be seen in patients with expanding middle ear and mastoid lesions. It is produced by *localized erosion of bone* into the labyrinth, without invasion of the endolymphatic portion. The usual cause is a **cholesteatoma** eating away at the lateral semicircular canal in the mastoid antrum, but any other expanding tumor, such as *glomus jugulare*, can be a cause.

The symptom is mild vertigo, which may be aggravated by pushing on the ear or getting water in it. Positional vertigo may also be present. Complaints of hearing loss or drainage, referable to the middle ear lesion, are often present. Ear examination will uncover a middle ear abnormality. If the primary physician encounters *true vertigo with evidence of middle ear disease*, the ENT physician should be called immediately.

A positive **fistula test** (see "Spontaneous Perilymph Fistula" in Chapter 5) is often present. There may be further erosion of the cholesteatoma or tumor into the *membranous* portion of the vestibule, with ensuing fulminant **suppurative labyrinthitis**. If this occurs, there will be extreme vertigo, with subsequent total loss of hearing and vestibular function.

Motion Sickness

Motion sickness is an abnormal response of an individual to the stimulus of prolonged motion. Usually it occurs in boats, automobiles, or airplanes. Those affected are prone to become dizzy, diaphoretic, and nauseated after an extended period of travel. Oddly, their dizziness is seldom actual vertigo, but more often a sense of imbalance and lightheadedness. These patients may show positional nystagmus on ENG testing and hip sway on posturography, even when symptoms are not present. Preventative treatment consists of oral **Dramamine** or **Phenergan**, or **scopalomine** skin patches.

Vestibular rehabilitation therapy (VRT) deserves mention as a therapeutic modality. Many medical centers have this capability within their rehabilitation departments. This treatment consists of a series of adaptive exercises for patients with *chronic vestibular dysfunction or damage*, including motion sickness. The exercises are not intended for patients

with acute vertigo. They are also not very helpful for those with central (brainstem) causes for their dizziness.

In addition to motion sickness, vestibular rehabilitation has proven effective for benign positional vertigo and any chronic loss of vestibular inner ear function, either unilateral or bilateral. The exercise protocols vary from program to program, but in general, they are designed to "challenge" the dizzy patient to perform tasks of balance with the eyes closed or with the head moving. Initially, the tasks will exacerbate a patient's symptoms, but the intent is for eventual adaptation and compensation. Clinical improvement is often seen in motivated patients.

In closing this discussion of vestibular disorders, be reminded that other diseases discussed in Chapters 5 and 7 may cause true vertigo or dysequilibrium. These include **spontaneous perilymph fistula**, classic **Méniere's disease, autoimmune inner ear disease, syphylis,** and **cerebellopontine angle tumors** (e.g. **acoustic neuroma**).

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Epilogue

Remember Pat, the patient mentioned in the preface with multiple ear symptoms? I hadn't yet completed the history, nor done an examination. (Somehow, the writing of this text interceded.) To continue where we left off, her symptoms and history initially seemed very compatible with temporomandibular joint (TMJ) syndrome. I doubted that ear fluid or infection would be seen.

Further careful questioning revealed that Pat was under more stress than usual in her life and that her dentist saw signs of wear from teeth grinding. She seemed tense and offered many complaints. She described her dizziness as lightheadedness rather than real vertigo. However, when quizzed about her right ear, she said that this recent fullness was worse than at other times, and that the hearing really seemed decreased and distorted. There was a roaring noise in it like that of a "seashell." Her own voice did not sound particularly loud in this ear, however.

On examination, both of Pat's tympanic membranes (TMs) looked slightly cloudy. However, the pneumatic otoscope showed nice mobility of the drums. Tuning fork testing showed the Weber test lateralizing to the left ear. The Rinne test was normal on both sides—air conduction better than bone. When asked to compare the loudness of the 512 fork on the right to the left, she said it was much quieter on the right and sounded like a different tone. Audioscope screening at 25 dB HL showed absent responses at 500 Hz and 1 KHz on the right. The rest of the examination was unremarkable, except for some subjective bilateral tenderness of the TMJs, with a little palpable crepitance on the right side. Obviously, there were TMJ findings, but there was also a right sensorineural hearing loss. This deserved a further workup.

A formal audiogram was done, and the right ear showed a 60 dB HL loss at 250 Hz, 45 dB HL at 500 Hz, and 35 dB HL at 1 KHz. The loss was sensorineural—no air–bone gap. All other frequencies in both ears were normal. Tympanometry showed normal middle ear pressure bilaterally. Thus, the cloudy drums were simply chronically scarred and there was no middle ear problem.

Pat had obvious signs and symptoms of a chronic TMJ disorder, but she also had acute cochlear hydrops (Lermoyez's syndrome) in the right ear! I wasn't sure whether the hydrops had occurred at other times in the past,

and whether or not Pat would develop full-blown Méniere's disease, with vertigo, in the future. Thankfully, however, a careful history and ear examination, backed by a good knowledge of ear disorders, kept me from missing the boat. After the audiogram, we discussed both diagnoses and started a treatment plan. Diuretics and tapering steroids would be prescribed for the hydrops, and advice was given about bruxism and the possible use of a night guard for the TMJ.

Pat's case is a good illustration of the challenges and possible pitfalls in diagnosing ear problems. Hopefully, reading this text has given you an increased knowledge of ear disease and its analysis. Your careful history, office examination, and preliminary hearing evaluation will likely pick up problems like Pat's.

Based on my experience, if diagnostic errors occur, it is usually in the following way. A patient's history, sometimes influenced by self-diagnosis, leads the clinician to an early conclusion. Then, the ear examination, falsely shaded by the initial impression, lacks completeness and accuracy.

Patients do not always fit into simple diagnostic slots, and neither did Pat. Some people offer the most incredible histories with multiple complaints. Other stoic individuals may give one-sentence, single-symptom histories. Experience shows that those patients with fewer and more specific complaints are more likely to have abnormal findings, but either type of patient may have a serious problem. In any case, knowledge and intellectual honesty can help us avoid a convenient, but inaccurate, diagnosis.

At this point, let me make one pitch for my specialty. If a clinician is unsure about an ear problem, consultation with an ENT person might be more useful and cost effective than continued treatment without a definite diagnosis, or a "shotgun" CT or MRI. I am grateful to all the physicians who have referred patients to me over the years; the challenges have made my life interesting and rewarding. It has been fun to put on the "thinking cap," treat these patients as friends, and then refer them back to their own physicians.

"Top Ten" Clinical Misjudgments in Ear Disorders

By way of review, it would be helpful to mention 10 of the most common oversights that I have seen some clinicians make over the years regarding specific ear problems. These errors have not occurred often, and usually the patient has been referred to a specialist in a timely fashion. Having read the text, you will be familiar with the disorders mentioned, but can always refer back to review an appropriate section, which will be cited in each example. The order in which these items are mentioned is not particularly significant in terms of frequency or importance.

1. Sudden sensorineural hearing loss is inappropriately diagnosed and treated as a middle ear disorder with decongestants, antihistamines, or even antibiotics. Accurate diagnosis and early treatment with systemic steroids and other measures can improve the outcome and prevent permanent loss. See pages 102–103.
2. Acute diffuse otitis externa (swimmer's ear) is treated with oral antibiotics that do not cover *Pseudomonas*, and no topical preparations are prescribed. This painful disorder is almost always caused by *Pseudomonas*. Appropriate antibiotic/steroid drops, on a wick if necessary, are the preferred treatment. See pages 38–39.
3. TMJ or myofacial pain syndrome is diagnosed and treated as an ear disorder. Quite often, a mildly scarred tympanic membrane is misinterpreted to indicate a middle ear problem, and the patient is treated for eustachian tube dysfunction or middle ear fluid. A thorough history and examination can prevent this error. See pages 26–28.
4. An elderly individual with generalized disequilibrium is treated with meclizine. This antihistamine is indicated for nausea and vertigo. It can actually make chronic disequilibrium worse by causing drowsiness. See page 113.
5. Chronic external ear infections with thick drainage are treated with antibacterial drops. Almost always, these are fungal infections. A culture for *all* organisms should be done, and then appropriate treatment, including topicals, can be started. See pages 41–43.
6. Hearing loss in a young child is ignored. The toddler seldom offers complaints, only the mother or another family member. The subtle findings of chronic middle ear fluid may be missed. Moreover, there may be a sensorineural hearing loss and no middle ear problem. A careful history, examination, tympanometry, and hearing evaluation will pick up most of these problems. See pages 65–67 and 97–99.
7. Benign positional vertigo is either missed, or on the other hand, overdiagnosed. This disorder is a frequent cause of recurrent vertigo, and the history and findings are characteristic. Electronystagmography (ENG) testing can clinch the diagnosis. The Epley maneuver, when used appropriately, is a reliable cure. See pages 117–118.
8. A foreign body is pushed further into the ear by a well-meaning caregiver. Determining the nature of the foreign body by history and examination should be followed by appropriate instrumentation. "Getting around behind" the foreign body, especially rigid ones like beads, is essential. ENT referral for the first try is usually the best option. See pages 46–47.

9. A cholesteatoma is missed. This invasive squamous ingrowth into the middle ear and mastoid can be interpreted as a routine middle ear infection. See pages 69–73.
10. Any type of ear complaint combined with any type of dizziness is diagnosed as "an inner ear infection." We should remember that the outer and middle ear "compartments" are separate from the inner ear, and that their problems seldom cause real vertigo. Familiarity with all the clinical chapters, especially 7 and 8, can give the reader a good knowledge of the causes of true labyrinthine vertigo.

Glossary

acoustic neuroma a benign tumor of cranial nerve VIII located in the internal auditory canal (IAC) or at the internal auditory meatus (cerebellopontine angle), also called *vestibular schwannoma*

acoustic reflex an involuntary bracing of the stapes by the stapedius muscle in response to a loud noise stimulus, which is measurable during tympanometry

acute coalescent mastoiditis a bacterial infection of the mastoid air cell system following a resistant or untreated acute otitis media

acute diffuse otitis externa an acute infection of the external auditory canal (EAC), usually by *Pseudomonas,* also known as *swimmer's ear*

acute otitis media an acute middle ear infection

adhesive otitis media a deep retraction of the eardrum that adheres to the ossicles or middle ear wall, either *active* (infected and draining, often with granulation) or *inactive* (clean and dry)

aerotitis a middle ear problem resulting from a rapid decease in middle ear pressure relative to the outer atmosphere, usually seen in flying

air–bone gap a difference in air and bone conduction on audiometry, indicating a *conductive hearing loss*

air conduction perception of sound through the outer ear inward; the way we normally hear

aminoglycosides antibiotics such as Tobramycin that are effective against *Pseudomonas* and other, usually resistant, Gram-negative bacteria

annulus a fibrous ring in the middle layer of the tympanic membrane located at its periphery

atelectasis deep retraction of the tympanic membrane, often with thinning due to loss or attenutation of the middle fibrous layer

atresia regarding the ear, a congenital malformation of the EAC with complete absence of a lumen

attic the cavity above the middle ear adjoining it, also called the *epitympanum*

audiogram a hearing test

Audioscope a device that screens hearing at a few selected frequencies

auditory brainstem recording a test measuring involuntary responses to sounds via electrodes on the skull, useful for the very young or mentally handicapped

auditory neuropathy hearing impairment due to pathology just deep to the cochlea, often of obscure cause
aural polyp a mass of granulation tissue in the ear, usually indicative of osteitis (inflammation of bone)
autophony the sense that one's own vocal sounds are loud and hollow, usually occurring with a conductive hearing loss
Bell's palsy a spontaneous unilateral paresis or paralysis of the face with no obvious cause
Bell's phenomenon elevation of the pupil when attempting the close the eye, often seen in Bell's palsy
bone conduction conduction of sound through the bone of the skull directly to the cochlea, usually done with a tuning fork or the bone transducer of an audiometer
branchial cleft cyst a congenital cyst of the neck; a *first branchial cleft cyst* is just below the external ear and closely associated with the facial nerve
bruxism clenching and/or grinding of the teeth
bullous myringitis a painful ear infection characterized by blebs on the tympanic membrane
caloric stimulation creation of convection currents in the lateral semicircular canal by introducing air or water into the ear that is not body-temperature, normally producing nystagmus
cauliflower ear a chronic deformity of the auricle usually resulting from a hematoma that has been neglected and become organized
central auditory processing disorder hearing dysfunction due to pathology somewhere in the central nervous system
cerebellopontine angle tumor a tumor located near the internal auditory meatus, often an acoustic neuroma
cervical vertigo vertigo secondary to cervical deformity or trauma characterized by dizziness on neck motion
cholesterol granuloma a long-standing solidified chronic middle ear effusion characterized by a bluish discoloration of the drum
cholesteatoma an epidermal inclusion cyst anywhere in the middle ear or temporal bone that tends to expand and cause bone destruction with complications
chorda tympani a branch of the facial nerve traversing the upper middle ear to bring taste from the ipsilateral anterior tongue
chronic mastoiditis chronic inflammation of the mastoid, usually with osteitis, and often associated with cholesteatoma
chronic otitis externa chronic inflammatory disease of the EAC, almost always fungal (*mycotic*)
chronic otitis media chronic inflammation of the middle ear associated with a long-standing perforation of the tympanic membrane

chronic stenosing otitis externa severe narrowing of the EAC secondary to chronic inflammation
circumscribed labyrinthitis an erosive localized inflammation of the labyrinth, usually at the lateral semicircular canal and caused by cholesteatoma, symptomatic with positional vertigo
cochlea the organ of hearing within the labyrinth of the inner ear whose outer bony surface appears similar to a shell
cochlear implant an electronic device surgically implanted in the inner ear for patients with profound hearing loss; the hearing is distorted but much improved
Cogan's syndrome autoimmune hearing loss associated with interstitial keratitis
conductive hearing loss hearing loss due to an obstruction or malfunction of the outer or middle ear
congenital cholesteatoma an epidermal inclusion cyst within the middle ear or elsewhere in the temporal bone that is not associated with an obvious defect in the tympanic membrane
Costen's syndrome temporomandibular joint (TMJ) disorder or syndrome
crepitance a crackling sound or tactile sensation
dyplacusis the hearing of a single tone as two different tones, one in each ear, often seen in Méniere's disease
disequilibrium difficulty with balance
eczematoid dermatitis a skin disorder characterized by scaling, itching, and weeping of the skin, sometimes allergic
endolymph potassium-rich fluid in the central membranous compartment of the inner ear
endolymphatic shunt inner ear surgery intended to reduce the *hydrops* of Méniere's disease in the membranous labyrinth
epidermal inclusion cyst a skin-lined cyst containing keratin and cholesterin crystals, found anywhere in the body, but also characteristic of cholesteatoma
epitympanum the cavity adjoining the middle ear from above, same as *attic*
Epley procedure a manipulation intended to cure *benign paroxysmal positional vertigo*
eustachian tube dysfunction any problem of the eustachian tube causing abnormal aeration of the middle ear cavity
exostosis in the EAC, a sessile rounded bony hump, often caused by chronic cold-water swimming
external auditory meatus the opening of the EAC
fallopian canal the bony canal enclosing the facial nerve just deep to the middle ear

fistula in the ear, a communication between the inner ear cavity and the middle ear or mastoid cavity

fistula test a diagnostic introduction of alternating positive and negative pressure to the ear with the pneumatic otoscope bulb in order to demonstrate vertigo if a fistula exists

glomus jugulare a benign but potentially aggressive vascular tumor in the hypotympanum, middle ear, or mastoid arising from the jugular bulb or Jacobson's nerve on the promontory

Gradenigo's syndrome involvement of the petrous apex with mastoiditis, producing persistent otorrhea, deep eye pain, and diplopia due to paresis of cranial nerve VI

Hallpike maneuver a diagnostic quick move from sitting to supine with the head turned to one side; vertigo and nystagmus are seen here in *benign paroxysmal positional vertigo*

hearing level an audiometric pure tone hearing measurement that adjusts sound pressure levels to human ear standards

hearing threshold level an audiometric hearing measurement of spoken words that adjusts sounds pressure levels to human ear standards

hemotympanum blood in the middle ear

hypotympanum the extension of the middle ear cavity below the level of the tympanic membrane

incus the second of the three middle ear ossicles, also known as the *anvil*

Jacobson's nerve and plexus the sensory branches of cranial nerve IX on the *promontory*

jugular bulb a curved bulge of the internal jugular vein beneath the hypotympanum

keloid a fibrous tumor arising from a skin defect that often occurs on the ear lobule at an ear piercing site

keratosis obturans a condition of accumulated squamous epithelium in the EAC that can erode bone

labyrinth the inner ear; a cavity containing the structures of the cochlea and vestibule

labyrinthitis any inflammation of the labyrinth; when used alone, often implying a self-limited viral infection

Lermoyez's syndrome endolymphatic hydrops limited to the cochlea without vestibular symptoms; its sensorineural hearing loss is usually fluctuating

light reflex a triangle of light on the anteroinferior drum that is present with healthy middle ears and is often, but not always, lost with pathologic middle ear conditions

malignant otitis externa a severe form of external otitis with osteomyelitis, usually occurring in diabetics or debilitated individuals, also called *necrotizing otitis externa* or *skull base osteomyelitis*
malingerer one who consciously fakes a physical disorder such as a hearing loss
malleus the first of the three middle ear ossicles, also known as the *hammer*
manubrium the handle of the malleus, the most prominent structure seen beneath the drum
mastoid the part of the temporal bone and its complex cavity adjacent to and adjoining the middle ear from posterior
mastoid antrum the largest part of the mastoid cavity, just posterior to the epitympanum
microtia a congenitally small, deformed external ear
Méniere's disease *endolymphatic hydrops*, a disease characterized by four symptoms: aural fullness, sensorineural hearing loss, tinnitus, and vertigo
modified Valsalva maneuver an attempt to inflate the middle ear cavity by occluding the nares, closing the mouth, and forcing air through the eustachian tube with positive pressure
mucous otitis media mucoid secretions in the middle ear
myoclonus involuntary rhythmic twitching of a muscle or muscles; for example *palatal* and *middle ear myoclonus*
mycotic otitis externa fungal otitis externa
myofascial pain syndrome chronic or recurrent pain in the muscles of mastication and of the face due to bruxism or muscle tension, closely related to TMJ disorder
myringitis inflammation of the tympanic membrane
night guard a prosthetic "bite block" worn at night to alleviate TMJ disorder when nocturnal bruxism is the cause
nystagmus rhythmic twitching of the eyes, usually due either to stimulation of the semicircular canal(s) or to a pathologic disorder of the vestibular system
organ of Corti a small compartment within the membranous cochlea where the hair cells and related structures are located; mechanical sound energy is transformed to neural impulses here
ossicles the sound-conducting bones of the middle ear: the *malleus* (hammer), *incus* (anvil), and *stapes* (stirrup)
ossicular disarticulation discontinuity in the chain of the three ossicles, occurring from congenital malformation, trauma, chronic infection, or erosion (e.g. by cholesteatoma)
otalgia pain in the ear

otic capsule the bony structure surrounding the cavity containing the soft tissue structures of the inner ear; the *petrous* portion of the temporal bone

otitis media any inflammatory involvement of the middle ear, expanded to include conditions of middle ear effusion

otoliths tiny calcium carbonate granules within the utricle and saccule whose weight shift stimulates the hair cells of these position-sensitive organs

otosclerosis a hereditary disorder of abnormal bone growth around the stapes footplate and occasionally in other locations; a frequent cause of progressive conductive hearing loss in adults

oval window the bony opening into which the stapes footplate inserts

papilloma a wart; when in the ear, it is often located at the external auditory meatus

pars flaccida the small, thin, two-layered portion of the tympanic membrane at its top

pars tensa the thicker three-layered portion of the tympanic membrane, comprising most of its surface

patulous eustachian tube an overly patent eustachian tube

petrous bone the pyramidal inner portion of the temporal bone encapsulating the inner ear structures

preauricular cyst a congenital cyst located deep to the skin just anterior to the upper auricle

presbycusis sloping high-frequency hearing loss, usually associated with aging, but genetically variable

promontory a rounded bony eminence over the cochlea that forms much of the medial wall of the middle ear

psoriasis a scaly skin condition that may affect the external ear

pulsatile tinnitus a thumping or "whooshing" noise in the ear that is synchronous with the pulse and vascular in cause

pure tone threshold the minimum sound intensity of a given tone that an individual can hear in an audiometric testing situation

quinolones new generation antibiotics that are effective against *Pseudomonas* and can be given orally, topically, or intravenously

Ramsay-Hunt syndrome *Herpes zoster oticus*, a painful viral infection located in the ear, mainly proliferating in the neural structures of the ear and in adjacent cranial nerves

recruitment the phenomenon of abnormal sensitivity to small increments in sound volume that is seen with *cochlear* losses and can be tested for audiometrically with special tests, like the SISI

referred pain pain felt in a given location that is transferred from another area due to shared innervation

retraction pocket a localized indentation in the tympanic membrane
Rinne test a tuning fork test for conductive hearing loss, it compares *air* to *bone* conduction
round window a window at the termination of the *scala tympani* located in a niche posteroinferior to the promontory
saccule a vestibular organ of the inner ear perceiving linear acceleration
scala tympani the upper perilymph-filled compartment of the cochlea
scala vestibuli the lower perilymph-filled compartment of the cochlea
scutum an anatomic spur corresponding to the superior-medial end of the bony EAC seen on coronal X-rays or scans of the middle ear; it is often significantly eroded by a *cholesteatoma*
sebaceous cyst a common term for *epidermal inclusion cyst*
seborrheic dermatitis a scaly skin condition that may affect the outer ear
semicircular canals the three canals of the vestibule; lateral (horizontal), superior, and posterior, that perceive rotational acceleration
sensorineural hearing loss hearing loss due to cochlear or auditory nerve (cranial nerve VIII) impairment
serous otitis media serous effusion in the middle ear
short increment sensitivity index a special audiometric test for recruitment, indicating a cochlear hearing loss
short process of the malleus a bony projection at the top of the manubrium of the malleus seen as landmark of the upper tympanic membrane
somatosounds sounds in or near one's ear that another can hear, formerly called *objective tinnitis*
speech discrimination the ability to understand speech, often tested audiometrically and expressed as a percentage score
speech reception threshold the minimum sound intensity of two-syllable words or phrases that an individual can hear and repeat in an audiometric testing situation
spontaneous perilymph fistula a rupture of the oval or round window membrane without externally inflicted trauma
stapedectomy surgical removal and replacement of the stapes, done for otosclerosis
stapedius muscle and tendon a bracing mechanism to tilt the stapes and stiffen the ossicular chain; contraction occurs reflexively to loud sounds
stapes the third of the three middle ear ossicles, also known as the *stirrup*
sudden sensorineural hearing loss a rapid-onset cochlear hearing loss without an obvious cause
suppurative labyrinthitis bacterial infection of the fluids of the labyrinth, often resulting in destruction of the inner ear organs

subclavian steal syndrome a vascular disorder with vertigo or syncope caused by shunting of blood away from the brainstem to the upper limbs
suppurative otitis media purulent drainage from the middle ear through a perforation
syncope fainting, or loss of consciousness
temporary threshold shift reversible temporary hearing loss from loud noise exposure
temporomandibular joint (TMJ) the articulation of the condyle of the mandible into its fossa in the temporal bone
tensor tympani the muscle of the middle ear that inserts into the malleus
tinnitus noise in the ear; *objective* tinnitus (extremely rare) can be heard by the examiner; *subjective* tinnitus is only heard by the individual experiencing it
tone decay a relatively inexpensive special audiometric test that localizes pathology to the auditory nerve
trigeminal neuralgia a pain syndrome originating from one or more branches of cranial nerve V, also known as *tic douloureux*
tuberculous otitis media a tubercular infection of the middle ear, characterized by multiple perforations of the tympanic membrane
tympanometer a device measuring the compliance of the tympanic membrane at varied external pressure settings, useful for uncovering middle ear effusions and other pathology; also known as an *impedance bridge*
tympanometry measurements done by a tympanometer
tympanoplasty surgical repair of the tympanic membrane and/or the middle ear ossicles
tympanosclerosis dense hyaline scar deposition within the tympanic membrane
umbo the small shiny circular depression of the tympanic membrane at the lower tip of the manubrium of the malleus
utricle a vestibular organ of the inner ear perceiving linear acceleration
ventilating tube a tube placed in the tympanic membrane as a presssure-equalizing vent, also known as a tympanostomy tube or myringotomy tube
vertebrobasilar insufficiency compromise of blood flow to the brainstem, possibly causing vertigo or disequilibrium
vertigo dizziness characterized by a whirling sensation with nausea, characteristic of vestibular dysfunction
vestibular Méniere's syndrome recurrent vertigo in a pattern similar to Méniere's disease, but without hearing loss; also termed *atypical Méniere's syndrome*

vestibular neuronitis a viral or postviral neuropathy of the vestibular nerve causing acute vertigo, without ear symptoms, which gradually resolves with time

vestibular rehabilitation therapy a protocol of adaptive exercises for patients with chronic vestibular dysfunction

vestibular schwannoma an anatomically accurate term for *acoustic neuroma*

Wallenberg's syndrome a symptom complex including disequilibrium that is due to occlusion of the posteroinferior cerebellar artery

Weber test a tuning fork test that lateralizes a hearing loss to one ear or the other, depending upon whether the loss is conductive or sensorineural

Acronyms and Abbreviations

ABR auditory brainstem recording
ANA antinuclear antibody
BAER brainstem auditory-evoked response (same as ABR)
BPPV benign paroxysmal positional vertigo
CBC complete blood count
CSF cerebrospinal fluid
CT computed tomography
dB decibel
EAC external auditory canal
ENG electronystagmography
HPV human papillomavirus
Hz hertz (cycles per second)
IAC internal auditory canal
KHz kilohertz (thousands of cycles per second)
MRI magnetic resonance imaging
MRA magnetic resonance imaging with angiography
OAE otoacoustic emission
OSHA Occupational Safety and Health Administration
SISI short increment sensitivity index
SRT speech reception threshold
TM tympanic membrane
TMJ temporomandibular joint
TTS temporary threshold shift
VBI vertebrobasilar insufficiency
VRT vestibular rehabilitation therapy

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