Chapter 2. Ear

I. Introduction

A. Anatomy of the ear

1. The auricle, or pinna, is the projecting cupped appendage that, along with the external canal, constitutes the external ear. It is composed of skin, subcutaneous tissue, fat, rudimentary muscles, and cartilage. The cup shape assists in the capture of sound waves. Although normal variations occur, certain components are common to the auricle:

   a. Helix - the rolled edge of the periphery.
   b. Antihelix - shaped like a "Y"; within confines of the helix.
   c. Concha - bowl-shaped depression in center.
   d. Meatus - entrance to the external auditory canal.
   e. Tragus - also called "goat's beard" because hair often projects from the cartilaginous prominence located anterior to the meatus.
   f. Lobule - fleshy inferior portion of auricle.

2. External auditory canal. The external auditory canal is a tubular conduit connecting the sound-filled environment to the tympanic membrane. It is lined by skin. In the outer one-third, the skin is cushioned with fat, cartilage, connective tissue, and muscle. In the medial two-thirds, the skin is very thin and adherent to the bony canal. The lateral canal often has hair and normally contains cerumen glands. The canal augments hearing by functioning as an air conduit for sound. The canal is often convoluted, requiring manipulation for cleansing or for viewing the tympanic membrane.

3. The tympanic membrane is the receptive diaphragm for auditory impulses and is the "window" to the middle ear. The pars tensa is a cone with its apex at the umbo (see C) and is composed of three layers: outer (squamous), middle (two fibrous layers, one with concentric and one with radial orientation), and inner (mucous membrane). The pars flaccida (see D) lacks the middle (fibrous) layer. The tympanic membrane collects sound waves in a complex fashion. Its large size, compared to the size of the stapes foot plate, allows for a pressure gradient of 17-fold. Certain landmarks should be noted:

   a. Annulus - rolled edge at periphery.
   b. Manubrium - handle-shaped process of the malleus in apex of tympanic cone.
   c. Umbo - end of manubrium in center of the tympanic membrane.
   d. Pars flaccida - Shrapnell's membrane.
   e. Incus - vertical bone in superoposterior quadrant seen through tympanic membrane.

4. Middle ear. The contents of the middle ear space are the malleus, incus, stapes, middle ear muscles (stapedius and tensor tympani), chorda tympani nerve, various smaller blood vessels, nerves, and mucosa. The middle ear or tympanic cleft is a mucosal-lined cavity containing the ossicles and their related tendons and muscles. It extends anterior and posterior, inferior and superior to the tympanic membrane. The superior portion of the tympanic cavity located above the pars tensa portion of the tympanic membrane is known as the epitympanum,
or attic, and opens into the mastoid antrum through the additus. The additus extends posterior between the epitympanum and the mastoid antrum.

The middle ear cavity is entered in its anterior portion by the eustachian tube, which provides ventilation and pressure equalization of the middle ear and mastoid. In addition, it prevents reflux of nasopharyngeal contents into the middle ear cleft. At birth, the mastoid consists only of the antrum, a superior cavity that opens into the additus.

Beginning at birth and continuing for several years, the mastoid cells form as outgrowths of the antrum that extend to all portions of the temporal bone and to the zygoma, in extensively pneumatized bones. There is great variability in individual mastoid dimensions. These cells are lined with mucosa and are subject to the same disease processes as the middle ear mucosa.

The primary function of the middle ear is to conduct sound from the external environment to the fluids of the inner ear. The function of the mastoid air cells is unknown, but they are thought to participate in the pressure-regulating mechanism.

5. Inner ear

a. Anatomy. The inner ear consists of a bony labyrinth filled with perilymph surrounding a membranous labyrinth filled with endolymph. The labyrinth is composed of three semicircular canals, a vestibule, and a cochlea. The three semicircular canals are at right angles to each other. Each canal contains a crista, or sense organ. When fluid moves in these canals, the sense organs are stimulated and send electrical impulses through the vestibular nerve. The cochlea makes two and one-half turns and contains the inner and outer hair cells within the organ of Corti, which connect the fibers of the cochlear division of the eighth cranial nerve. The vestibule contains the oval window, in which the stapes is situated and also the utricle, a sensory receptor responsive to positional change, and the saccule, whose function is unknown.

b. Physiology

(1) Hearing. The auricle acts as a sound-localizing device, although it probably is not as effective in humans as it is in animals with larger, more mobile appendages. The external ear acts as a conduit to transmit sound waves to the tympanic membrane. Vibration of the tympanic membrane initiates ossicular motion, which transmits sound via the oval window to the perilymphatic fluid, which in turn displaces endolymph fluid and the basilar membrane, activating auditory receptors (auditory hair cells). Hair-cell motion initiates activity in the auditory nerve (eighth cranial nerve) that, via complex neural pathways, arrives in the auditory cortex and is perceived as sound.

(2) Equilibrium. The three semicircular canals each contain a crista, or sense organ. These sense organs are responsive to fluid movement in the endolymph caused by rotational acceleration. Each crista has a resting potential that is either increased or decreased, depending on the direction of fluid motion. Each canal is located in a plane that is approximately 90 degrees in relation to the others and is maximally stimulated by rotation in
the plane in which the canal lies. This location enables the body to sense the direction of rotation. The sense organ of the macula responds to linear acceleration.

Through reflex connections to the extraocular muscles, the eyes attempt to provide visual fixation when the head rotates. Reflex connections to muscles produce a "righting" reflex and other postural adjustments. The visual system and peripheral sensors also contribute independently to the balance mechanism. Aberrations in these areas can similarly be responsible for imbalance or disequilibrium.

6. The facial nerve is encased in the temporal bone for a distance of 37-45 mm. This is the longest bony enclosure of a nerve in the human body, and makes the facial nerve subject to injury from swelling or trauma to the temporal bone. The nerve enters the internal auditory meatus superior to the cochlear nerve and travels through the internal auditory canal, passing next through the labyrinthine portion of the temporal bone for a short distance to reach the geniculate ganglion. Here the nerve turns sharply posterior (first genu) and passes superior to the oval window (tympanic segment). It then turns inferiorly (second genu), traveling vertically through the mastoid (mastoid segment), and exits through the stylomastoid foramen. The nerve has three primary branches in the temporal bone:

   a. The greater superficial petrosal nerve branches at the level of the geniculate ganglion and controls lacrimation.

   b. The stapedial branch exists in the mastoid segment and controls the stapedius muscle.

   c. The chorda tympani nerve, which branches just above the stylomastoid foramen, carries taste to the anterior two-thirds of the tongue.

7. The carotid artery is located in the petrous apex. Rarely is it involved in diseases of the ear, but it can be involved in severe temporal bone injuries involving the petrous apex or in extensive resection of the temporal bone.

8. The jugular bulb, located on the floor of the middle ear space, usually lies inferior to the tympanic membrane. Occasionally, the jugular bulb is positioned more superiorly and may be noted on examination of the tympanic membrane as a bluish discoloration below the umbo.

9. The sigmoid sinus is the S-shaped portion of the lateral sinus that is responsible for the majority of the venous drainage from the head to the jugular vein. It passes through the posterior wall of the mastoid and may be injured in a surgical procedure or involved in extensive infection of the mastoid cells, producing sigmoid sinus thrombosis.

10. The dura of the middle cranial fossa is closely related both to the mastoid cells and to the middle ear cleft, being separated by a thin layer of bone, the tegmen tympani. The tegmen tympani is penetrated by small veins and arteries that can transmit an infection from the middle ear or mastoid to the meninges and brain.
B. General signs and symptoms of ear disease

1. Otalgia. Ear pain can be caused by pathology directly related to the ear, the periauricular area, or distant sites (referred otalgia). With otalgia, it is imperative that, before treatment is initiated, a definitive diagnosis be established. Below are areas to be considered, other than the ear and periauricular area, that can cause otalgia. The ear receives innervation from cranial nerves V, VII, IX, X, XI, and from cervical nerves C2 and C3. Any of the other areas innervated by these nerves can refer pain to the ear.

a. Oral cavity

(1) Dental infection.
(2) Glossitis and stomatitis (particularly herpes).
(3) Neoplasia.

b. Pharynx (naso-, oro-, and hypopharynx)

(1) Malignancy, especially in the pyriform sinus.
(2) Pharyngitis.
(3) Retropharyngeal or peritonsillar abscess.
(4) Tonsillitis.
(5) Posttonsillectomy, adenoidectomy.

c. Esophagus

(1) Foreign body.
(2) Tumor.
(3) Esophagitis.

d. Larynx

(1) Tumor.
(2) Mucosal ulceration.
(3) Cricoarytenoid arthritis.
(4) Laryngitis.
(5) Epiglottitis.

e. Neuralgia

(1) Trigeminal.
(2) Geniculate.
(3) Glossopharyngeal.
(4) Sphenopalatine.
2. **Ototorhea.** Drainage from the ear is a common complaint. Depending on the associated history and physical examination, this symptom may indicate serious disease. It is important to document the nature of otorrhea, related symptoms, and events such as trauma that may have been precursors. The following types of otorrhea warrant consideration:

   a. **Cerumen** is the most common cause of otorrhea. The color varies from brown to pale yellow. The consistency varies from liquid to solid. Water in the ear or otic drops can increase the discharge.

   b. **Blood.** Although the primary cause is trauma (slap, instrumentation), acute perforations, external otitis, and tumors can also cause bleeding. Except in severe trauma and with clotting disorders, bleeding is rarely severe.

   c. **Serum** is seen occasionally with the rupture of a bleb from bullous myringitis; however, serum usually indicates a dermatitis affecting the external canal.

   d. **Pus** in acute otitis media is usually viscous, yellow or white. In chronic otitis media, the color changes to yellow gray or greenish and is thinner. "Pus" from external otitis is usually cheesy in nature.

   e. **Cerebrospinal fluid (CSF)** is usually clear and may be profuse. A sample should be taken for laboratory analysis (sugar, sodium, protein, and cells). A history of trauma, surgery, or tumor is often present. A prior history of meningitis may be obtained.

3. **Hearing loss.** As opposed to otalgia, hearing loss always indicates a disease process somewhere in the acoustic pathway (external ear to cerebral cortex). Hearing loss is usually described as conductive, sensorineural, or mixed (a combination of sensorineural and conductive). A hearing loss is often difficult for the patient to describe and, indeed, is frequently brought to the physician's attention by a family member or friend. The patient often denies a hearing loss, saying that people are speaking unclearly.

   a. **Conductive hearing loss** results in the interference of transmission of sound energy from the outside environment to the receptor organ (cochlear hair cells). Interference can occur anywhere from the auricle to the organ of Corti. Conductive hearing loss is frequently a temporary or correctable condition.

   b. **Sensorineural hearing loss** results from defects both in the transmission of sound energy into electrical impulses and in the transfer of these impulses to the auditory cortex. This hearing loss results from a variety of causes, including trauma, viral diseases, ear infection, and the aging process (presbycusis) (see IX.A.8.).
c. Mixed hearing loss indicates an additive effect of a conductive and sensorineural hearing loss. In a mixed hearing loss, it is important to differentiate whether the loss is primarily conductive (ie, correctable) or sensorineural.

4. Vertigo may best be described as a sensation of motion of either the patients or the environment. Severe vertigo with nystagmus and vomiting always indicates a disease process involving the peripheral vestibular apparatus. Vertigo may be the only symptom of ear disease, or it may be combined with other symptoms, such as hearing loss, otalgia, or otorrhea. Severe vertigo, particularly when acute, must be regarded as a significant symptom of ear pathology, and a thorough evaluation performed. Vertigo should be distinguished from dizziness.

5. Tinnitus is an altered sound perception not associated with an external stimulus. It can be correlated with systemic considerations; eg, sensorineural hearing loss, medications, temperature elevation, headache syndromes, vertigo, etc. It can also occur as an independent entity. Subjective and objective tinnitus have been defined:

a. Signs and symptoms

(1) Subjective tinnitus is perceptible only to the affected individual.

(2) Objective tinnitus can be identified by others.

b. Diagnostic tests

(1) Audiologic - many patients have an associated high frequency sensorineural hearing loss.

(2) Auditory brainstem response (ABR) - appropriate for all unilateral tinnitus.

(3) MRI (enhanced) - indicated for abnormal ABR or other suggestive audiometric data for acoustic neuroma.

(4) Hematology screen - appropriate in all instances - can define multiple medical precursors; eg, hyperlipidemia, diabetes, Paget's disease.

c. Treatment - The object is symptom control; however, cure is rarely effected.

(1) Identify and control medical correlates, temporomandibular joint (TMJ), hypertension, etc.

(2) Reduce environmental noise exposure - provide ear defenders as indicated.

(3) Avoid exposure to aminoglycosides and other medications known to cause tinnitus.

(4) Resolve if possible all remediable otologic pathology; eg, chronic suppuration, effusion, negative middle ear pressure.
(5) Tinnitus maskers - popular but do not meet most patient's expectations. These devices provide external noise and, via residual inhibition, can decrease awareness for some patients.

(6) Electrical stimulation - investigatory; the short-lived response is not infrequently associated with tissue damage.

(7) Biofeedback or relaxation techniques - of occasional benefit, best performed by competent professional.

(8) Medications - generally directed toward controlling the affect - none with clear superiority.

(9) Cognitive therapy - an affect-control modality.

II. Physical examination

A. External evaluation

a. **Position.** The top of the auricle should not fall caudal to a line drawn from the occiput to the lateral canthus of the eye. Such "low-set" ears can signal other congenital anomalies. The angle that the auricle makes with the side of the head varies. The condition of "lop-ear" from excessive protrusion is correctable.

b. **Consistency.** The ears of neonates are almost alarmingly soft and can maintain an iatrogenic crease for an extended period. Adult ears take on the normal "springy" consistency associated with cartilage.

c. **Size and shape.** A great deal of variation occurs. The auricles should be symmetric. Congenital anomalies that can occur are multiple and range from insignificant to complete absence of pinnae.

2. **The mastoid, postauricular sulcus, squamosal, and zygomatic areas** should be checked routinely for pathology of skin or underlying bone. A postauricular scar may be present - an indication of prior otologic surgery that the patient may have forgotten.

3. **Seventh cranial nerve function.** The nerve can be injured anywhere in its tortuous course through the temporal bone. All branches should be checked by raising eyebrows, lid closure, whistle or pucker, smile, scowl, and neck muscle tightening. Testing taste, lacrimation, and stapedial muscle function can pinpoint the site of injury.

B. **Otoscopy.** Several methods are available to illuminate the external canal and tympanic membrane. With use of any method, however, two principles should be kept in mind: (1) the bony external canal is tender when manipulated, and must be instrumented gently. (2) Patients should be advised in advance of any instrumentation to prevent a startle and sudden head movement.
The pinna should be positioned to open the meatus, to straighten the canal, and to provide a direct visual path to the tympanic membrane. In the child or adult, the pinna should be pulled posteriorly and superiorly. In the neonate, the lobule may require a tug directly caudal. One method of examination is the head mirror with reflected light. This method allows two-handed manipulation and is well suited for cerumen removal, wick placement, and other manipulation. In common practice, the conventional electric otoscope is an all-purpose instrument. It should have several features:

- Small handle ("C" batteries).
- Intense light (halogen is preferred).
- Air seal head (for pneumatic otoscopy).
- Open head (for instrumentation).
- Rubber tubing and bulb (for pneumatic otoscopy).

Other methods such as loupes, headlight, and the suspension microscope are available to the otologist. Micro-otoscopic telescopes are also available.

After removal of cerumen and after good visualization of the canal and tympanic membrane are achieved, the anatomy is evaluated and a drawing made. Osteomas and exostoses of the canal wall should be included in the drawing. The following manipulations can then be performed.

C. **Pneumatic otoscopy** is performed routinely on all patients. A handheld bulb is used for changing pressure in the external canal. A tight seal is essential. The tympanic membrane should be observed to move actively and crisply in both directions. The examiner can be misled and assume normal mobility (eg, a retracted tympanic membrane will move out with negative pressure, then passively return, without the need of positive pressure). Immobility or sluggish movement can be detected. The patient can perform his or her own pneumatic otoscopy. A Valsalva maneuver with a pinched nose and closed mouth can autoinflate the middle ear with tympanic membrane motion. This is a reasonable measure of eustachian tube function. An alternative method is to swallow with an occluded nose and mouth.

D. **Palpation.** Occasionally, the canal or tympanic membrane requires manipulation. A small suction tip (No. 20) can be used to check for mobility, for the presence of a perforation, or for other pathology. Although the drum is sensitive, gentle palpation is tolerated by most patients.

E. **Examination of the child.** The child often sets the tone of the examination. The examiner must be prepared to kneel on the floor, to sit in the chair while the child stands, or to place the child on a lap. Games are useful such as watching the painless "Tinkerbell" otoscope light on the child's hand, blowing out the light, and looking for bunnies in the ears. Children like to squeeze the bulb for the "wind" in their ears.

Should care and cleverness prove unsuccessful, the child should be "papoosed" by the parent. The parent sits on the chair, the child is on the lap. The child's legs are placed between the parent's clamped legs. The head of the child is snuggled sideways on the upper chest, and the parent hugs the patient. An assistant then controls the head.
F. Tuning fork testing helps to define normal from abnormal hearing, conductive loss versus a sensorineural loss, and the frequency range of the loss. These tests provide a gross estimate, but are not a substitute for an audiogram. Tuning fork tests are usually unsuccessful under the age of 5 years.

1. Rinne. Positive if air conduction is greater than bone conduction (a rare label for the normal to be "positive"). The tuning fork, preferably 512 Hertz (Hz), is placed on the mastoid tip firmly (almost to the point of discomfort) and then placed near the meatus so that the two prongs of the fork are aligned with the direction of the ear canal. (Rotate a tuning fork near your ear and note the change of loudness.) The patient states which position sounds louder.

   Air > Bone (Rinne is positive). Normal or sensorineural loss of that ear.

   Bone > Air (Rinne is negative). Conductive loss of that ear.

2. Weber. Tests for symmetry of hearing. The tuning fork is placed on the forehead or on the central incisor. The patient reports subjectively in which ear the tone is loudest. The test is fraught with error, because the patients may "not hear anything" or, in the case of a conductive loss, are afraid to report that the "bad" ear hears "better".

   a. Symmetric can be interpreted as normal hearing or symmetric hearing loss (either conductive or sensorineural).

   b. Lateralization toward the poorer-hearing ear usually indicates a conductive component (try this with a finger in one of your ears). Lateralization toward the better-hearing ear may indicate a sensorineural component in the opposite ear.

G. Clinical speech testing. Clinical speech testing may be performed by whispering simple words into an ear. The opposite ear should be "masked" by producing a noise in it. The otologist commonly uses a Barany Box, which produces 100-110 decibels (dB) of sound at ear level. Other noisemakers may also be used, such as a partially occluded suction tubing (producing a hissing noise). It is helpful to practice this technique on normal patients, always keeping the same distance from the ear and varying the intensity of the noise. The hand should be held in such a way as to prevent the patient from lip reading. This whispered voice test is much more accurate than having the patient listen to a watch tick. Bisyllabic words of equal stress (spondaic words) should be used when testing. (Examples are baseball, airplane, cowboy, railroad, eardrum, ice cream, hotdog.)

   Results should be expressed as normal hearing or as mild, moderate, or severe hearing loss. A person skilled in this method can estimate the hearing loss (in decibels) with surprising accuracy.

H. Audiometry should be considered as an extension of the physical examination. The audiometric tests cannot diagnose a disease process but should be used in conjunction with the history, physical examination, and other testing to arrive at a diagnosis (Table 2-1).
Table 2-1. Audiologic evaluation of cochlear and retrocochlear disorders

<table>
<thead>
<tr>
<th>Test</th>
<th>Cochlear lesions</th>
<th>Retrocochlear lesions (eight nerve)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pure tone audiometry</td>
<td>Sensorineural hearing loss</td>
<td>Sensorineural hearing loss</td>
</tr>
<tr>
<td>Speech discrimination</td>
<td>Good</td>
<td>Poor</td>
</tr>
<tr>
<td>Recruitment</td>
<td>Normal</td>
<td>No</td>
</tr>
<tr>
<td>Stapedial reflex</td>
<td>Impaired</td>
<td>Yes</td>
</tr>
<tr>
<td>Tone decay</td>
<td>Ménière's syndrome</td>
<td>Acoustic schwannoma</td>
</tr>
</tbody>
</table>

1. Pure tone testing presents a single-frequency tone to the patient through a headphone. The intensity of this tone is then varied until the tester determines the lowest intensity (in dB) that is audible. This testing is repeated in each ear at various frequencies, usually 250, 500, 1000, 2000, 4000, 6000, and 8000 cycles per second (cps). The test is then repeated using a bone conduction vibrator placed over the mastoid bone (usually in this test only 250-4000 cps are tested). In sensorineural hearing losses and in normals, bone conduction and air conduction (headphones) are equal. With conductive hearing losses, bone conduction scores are better than air conduction scores. Air conduction scores can never be better than bone conduction scores. The results are expressed as decibels of hearing loss with a range of 0-100 dB (the smaller the number, the better the hearing). The normal adult range is approximately 0-20 dB, and the children's range is 0-15 dB.

2. Speech reception threshold (CRT) is found by giving the patient a list of spondaic words (see G) at a frequency equal to a 1000-cps signal. The intensity of the stimulus (words) is then varied until a level is reached at which the patient can repeat half of the test items. This level is known as the speech reception threshold and is expressed in decibels. The test estimates the patient's handicap in connected conversation. The SRT for each ear should approximate (± 10 dB) the average for the pure tones at 500, 1000, and 2000 cps in each ear. (The range from 500-2000 cps is often called the speech frequency.)

3. Speech discrimination tests are used to test the clarity of articulated speech. A list of monosyllabic words that represent the phonetic balance of spoken English is used. The words are given at a comfortable intensity above the speech reception threshold (usually about 40 dB). The results are reported as a percentage of the words of the list that are repeated correctly. Normal discrimination is 90% or above, with most normal-hearing individuals scoring 96-100%. For the non-English-speaking, separate word lists must be used that approximate the phonetic balance of their native tongue. Unfortunately, such lists are not available for all languages.

4. Audiometric testing to identify the site of a lesion is frequently performed to determine if a hearing loss is caused by a cochlear or retrocochlear lesion (eighth nerve to auditory cortex). Some of these tests are listed below; however, their usefulness has greatly diminished now that auditory evoked response testing is available.
a. SISI (short-increment sensitivity index)
b. Tone decay
c. Bekesy
d. Recruitment testing.

III. Radiographic evaluation of the temporal bone is extremely difficult because of the small size of the structures being evaluated and the numerous overlying shadows. The routine use of x rays on an emergency basis (except in ideal circumstances) should be avoided.

A. Plain films usually consist of three to four view of the ear. The size and aeration of the mastoid can be determined, as can (sometimes) breakdown of the cell partitions caused by acute mastoiditis. In addition, large erosives lesions can be identified.

B. Computed tomography (CT), usually with iodinated intravenous contrast, gives the best bony definition of the temporal bone. Assessment of congenital abnormalities as well as bone destruction by tumor is best done by scanning. Soft tissue definition is acceptable.

C. Magnetic resonance imaging (MRI) with gadolinium gives excellent soft tissue definition of structures in and around the temporal bone. Inflammatory lesions and tumors are well defined.

D. Angiography of temporal bone lesions - especially glomus tumors - may help in defining their extent and blood supply. Newer noninvasive MRI-angio techniques may replace this modality.

IV. Special testing

A. Calories. Nystagmus can be elicited by instilling water of a temperature different from the body temperature into the external ear canal. This condition is produced by a change in temperature in the lateral semicircular canal, causing endolymphatic flow in the canal and stimulation of the sense organ (crista). Nystagmus is described according to the direction of the fast component. Cold water produces a quick component away from the ear tested; conversely, warm water produces a quick component toward the ear testedes (COWS: cold - opposite; warm - same). By comparing the length and intensity of nystagmus in each ear, a gross measurement of vestibular function can be made. Ice water (2 mL) can be used; however, it may stimulate severe vertigo with vegetative symptoms of nausea, vomiting, and diaphoresis. Water caloric are contraindicated in tympanic membrane perforations, temporal bone fracture, and CSF leaks.

B. Electronystagmogram (ENG) is based on the principle that the eye is a dipole, with a positive charge at the cornea and a negative charge at the retina. By placing recording electrodes around the eye, movements, including nystagmus, will cause a deflection in current that can be recorded. Caloric stimulation (30 and 44°C), positional testing, optokinetic testing, pendulum tracking, and spontaneous nystagmus are all recorded as part of the ENG report.

Currently, the ENG is the preferred method of vestibular testing because it gives a permanent record. The intensity (slow-phase velocity) of the nystagmus can be measured
accurately. The electroneystagmogram can give a better overall functional picture of the entire balance system.

C. **Brainstem auditory-evoked response (BSER), or auditory brainstem response (ABR),** is a development that uses a computer to average random cortical electrical activity (EEG). A series of stimuli (clicks) is presented to the ear to be tested, and scalp electrodes monitor the response. By analyzing the wave thus produced, a "map" of the auditory pathways can be produced. Because no patient response is necessary, this method is invaluable in testing neonates, young children, and others in whom an accurate response to conventional audiometry is questionable. Site of lesion testing, including the diagnosis of acoustic neuromas, is another important application of the ABR. Because patient movement can affect the outcome of the ABR, sedation may be required in some patients. The waveforms produced (I-V) are currently thought to represent synaptic connections in the auditory and other nuclei in the brainstem.

D. **Tympanometry, acoustic impedance, acoustic reflex.** Tympanometry is accomplished by using a probe that seals the ear canal. Varying pressure is then introduced into the canal, and the compliance of the eardrum is recorded on a graph. The normal ear shows a smooth, bell-shaped "peak" of compliance; fluid or other mass effects in the middle ear produces a flattened curve. Clues to various other ear pathology (eustachian tube dysfunction, ossicular discontinuity) can be gained by analysis of this graph.

Using the same equipment, high intensity (≥ 85 dB) sound can be introduced into the ear to produce a notch on the graph caused by contraction of the stapedius muscle. Since this muscle is innervated by a branch of the seventh cranial nerve, this test can be used for topographic testing. Decay of this reflex is often seen with acoustic neuromas. Because this reflex occurs at about 80 dB above threshold, a rough estimate of hearing sensitivity can be obtained in some patients. About 5% of the population have absent stapedial reflexes with otherwise normal ears.

E. **Posturography and rotation testing** have recently been added to the vestibular test armamentarium. They help to further evaluate vestibular reflex responses. Their limitations, however, are the same as with any reflex test. They do not actually measure vestibular output, and thus many factors can interfere with their validity.

V. Diseases of the auricle

A. **Preauricular appendages (accessory auricles).** Preauricular appendages are small, skin-covered tags that appear in the preauricular area on a line drawn from the tragus to the corner of the mouth. They may contain small pieces of cartilage.

1. **Signs and symptoms.** Except for their appearance, preauricular appendages are usually asymptomatic, but the examiner should be cautioned to look for other anomalies.

2. **Management.** Removal is not indicated unless the appendages are cosmetically desirable.
B. **Preauricular pits.** Preauricular pits commonly occur at the root of the helix, although they may occur in other locations. They can descend down to the lower border of the tragus and can contain glandular structures.

1. **Signs and symptoms.** Purulent drainage with swelling and pain can occur when the pits become infected.

2. **Management.** When these tracts become repeatedly infected, surgical excision is necessary.

C. **Auricular atresia (aplasia).** Auricular atresia may be unilateral or bilateral, and can range from mild malformations to complete absence of the auricle. It is frequently associated with anomalies of the external canal, middle ear, and temporal bone. Hearing loss is frequent. Atresia can be inherited, associated with embryopathies (rubella, thalidomide), or chromosomal abnormalities. For this reason, careful evaluation for other anomalies is mandatory, particularly in regard to derivatives of the first and second branchial arch.

1. **Signs and symptoms** are purely cosmetic unless a hearing loss is present.

2. **Management.** Restoration of hearing must take precedence, but surgical management must be carefully coordinated between the otologic and reconstructive surgeon.

D. **Trauma to the auricle**

1. **Lacerations** can range from simple lacerations to complete avulsion and are often associated with multiple trauma.

   a. **Management**

   (1) Careful cleansing of the wound with removal of foreign debris is necessary. In extensive injuries, general anesthesia may be required. Local blocks may also be used in the cooperative patient. Except in simple lacerations, local infiltration should be avoided because it distorts the anatomy and may disrupt the blood supply to the cartilage. The cartilage should be sutured only if necessary to reform the contour of the ear. If cartilaginous suturing is necessary, fine (5-0 or 6-0), noncolored suture should be used to prevent its showing through the skin. If possible, the perichondrium should be closed using fine, absorbable suture. Knows should be buried and the skin approximated with interrupted 6-0 monofilament nylon. A sterile pressure dressing (mastoid type) should then be applied.

   (2) Avulsion of the auricle must be repaired in the operating room with a team well versed in reconstructive techniques. The auricle can be preserved in sterile, iced saline until reconstruction can be accomplished.

   (3) With lacerations to the auricle, prophylactic antibiotics are not indicated except in dirty wounds (eg, human bite). Penicillin is the drug of choice in these instances.

2. **Hematoma or seroma** usually occurs with trauma to the auricle that produces hemorrhage under the perichondrium or skin.
a. **Signs and symptoms.** The hematoma or seroma is often blue, round, and smooth. Pain may be present. Because it disrupts the blood supply to the cartilage, prompt treatment is required to prevent aseptic necrosis and deformity (cauliflower ear).

b. **Management.** If hematomas or seromas are seen early in their course, before clot formation has occurred, aspiration with an 18-gauge needle and application of a pressure dressing may be sufficient. Careful follow-up is necessary to assess reaccumulation of the fluid. If the fluid reaccumulates or if aspiration is unsuccessful, incision and drainage, with the placement of drains, is indicated and should be performed in the operating room. Particularly resistant cases may require the placement of through-and-through mattress sutures over a bolus of cotton to ensure a good result. Fluid removal from the area should be Gram stained, cultured, and appropriate antibiotics begun. If there is evidence of infection (eg, purulent aspirate or cellulitis), intravenous antibiotic coverage should be started immediately to cover *Staphylococcus* and *Streptococcus* until culture results are obtained.

3. **Burns.** As with burns to other areas of the body, severe deformity is usually due to secondary infection and cartilaginous necrosis.

a. **Signs and symptoms.** The ear may be reddened, have vesicles, or be shiny white, depending on the degree of the burn. First- and second-degree burns are usually painful, whereas third-degree burns are not. A large number of patients will develop suppuration and chondritis, regardless of the depth of the burn.

b. **Management.** The burned area should be cleansed, an antibiotic ointment applied, and a light sterile dressing placed over the ear. Pressure should be avoided to prevent further embarrassment of the blood supply. Antibiotics should be used only when there is evidence of infection. Any debridement should be delayed to allow the devitalized areas to demarcate.

4. **Frostbite** occurs frequently to the auricle due to its protrusion and relatively poor blood supply.

a. **Signs and symptoms.** The auricle becomes white, with a slightly shiny appearance. There is loss of sensation to the affected area. Bullae may be present.

b. **Management.** Gradual rewarming is advisable using tepid compresses. Thereafter, the ear is treated like a burn. Antibiotic cream is applied to breaks in the skin, and a light sterile dressing is placed over the ear. Pressure is to be avoided. No debridement should be performed until viability is determined.

E. **Auricular chondritis (auriculitis).** Auricular chondritis may be caused by a spreading external otitis, trauma, or insect bite, but is often idiopathic.

1. **Signs and symptoms.** The entire ear is red and tense, including the lobule. Tenderness is usually present, but may not be severe. The periauricular soft tissues may be involved. *Streptococcus pyogenes* is often the causative organism.
2. Management consists of IV semisynthetic penicillin in high dosages and treatment of any underlying causes such as external otitis. Bacitracin or other antibiotic ointment should be used on skin breaks.

3. Complications. Lack of prompt and effective treatment results in loss of cartilage and in auricular deformity.

F. Polychondritis (relapsing). Relapsing polychondritis involves the ear as well as other cartilages (septal, costal). In contrast to auriculitis, the lobule is spared. Systemic steroids in high doses is the treatment of choice.

G. Tumors. Both basal cell and squamous cell carcinomas often involve the pinna since it is exposed to sunlight. Biopsy is necessary to determine the proper treatment. Squamous cell tumors may involve local nodes (mastoid, high cervical, parotid). Nodal disease must be resected in block or irradiated.

VI. Diseases of the external canal

A. Cerumen impaction. Cerumen is a normal finding in ear canals. It acts as a protection from maceration and lubricates the skin. Normally, cerumen migrates laterally and is discharged from the canals. In certain patients, however, this mechanism is less efficient. This problem can be aggravated by the use of cotton-tipped applicators, which tend to pack the wax into the canal.

1. Signs and symptoms consist of hearing loss, pressure sensation, or otalgia.

2. Diagnosis is made by the appearance of the wax, which varies from almost white to dark black-brown.

3. Management consists of removal with instrumentation (eg, curette, suction) or by irrigation. Removal can often be facilitated by the use of softening drops (glycerol peroxide, liquid diocyl sodium (Colacel)). Good illumination and exposure are necessary for this procedure. Irrigation should be avoided in patients who give a history of infection, bleeding, or perforated tympanic membrane. In some patients with recurrent impactions, self-administration of softening drops and gentle irrigation with a bulb syringe can avoid numerous trips to the doctor.

B. Trauma. Most trauma to the external canal is caused by instrumentation of the ear canal, either by the patient or by the physician. Cotton-tipped applicators are a common offender.

1. Signs and symptoms. The appearance of a laceration or hematoma in the skin of the canal makes the diagnosis.

2. Treatment consists of antibiotic drops and water precautions in simple lacerations and hematomas. More complex lacerations, particularly circumferential lacerations, should be treated by packing the external canal with a Merocel wick and using antibiotic ear drops to prevent canal stenosis.
C. **Foreign bodies** are extremely common in younger children, but may be seen in any age group. The foreign body may consist of anything that is small enough to enter the canal.

1. **Signs and symptoms.** The history, particularly in young children, is often not helpful in establishing the diagnosis. Symptoms consist of hearing loss, pain, or drainage.

2. **Management.** In an adult or cooperative patient, gentle removal with a foreign body curette, suction, or forceps (alligator-type) is often possible. In less cooperative patients and in those patients in whom the foreign body is wedged into the canal, operative removal under anesthesia with magnification is indicated.

Vegetable foreign bodies (eg, dried beans) swell after insertion and often require operative intervention. Extreme caution and gentleness must be exercised in foreign body removal. Imprudent attempts at removal have resulted in severe lacerations of the canal, tympanic membrane perforations, ossicular disruptions, and facial nerve injury. Proper equipment and expertise are essential.

D. **Furuncle.** Single or multiple furuncles are common in the external portion of the ear canal.

1. **Signs and symptoms.** Furuncles appear as localized swellings that may be fluctuant. Tenderness to palpation or insertion of an ear speculum is often marked.

2. **Management** consists of drainage of fluctuant areas, heat, and the use of a topical antibiotic (eg, Bacitracin). Systemic antibiotics are necessary only with cellulitis or systemic symptoms (eg, fever) and should consist of antistaphylococcal drugs. Narcotics may be necessary for 24-48 hours for pain control.

E. **External otitis (diffuse)** also known as "swimmers' ear." This condition is very common, particularly during the summer months. Water maceration or trauma (or both) are often etiologic.

1. **Signs and symptoms** consist of itching, pain (often severe), a plugged sensation in the ear, and a discharge, which is often cheesy.

2. **Diagnosis.** Physical examination elicits pain on auricular movement or tragal pressure. The canal is diffusely swollen and tender and may be completely closed. Desquamated debris is usually present in the canal. The tympanic membrane may be obscured by debris or swelling. Cultures usually grow *Pseudomonas, Proteus* or, less frequently, *Staphylococcus* and *Streptococcus*.

3. **Management** consists of gentle cleaning of the canal and topical antibiotic drops containing a steroid such as polymyxin B-neomycin-hydrocortisone (Cortisporin Otic Suspension) to reduce swelling. With marked swelling of the canal, a Merocel wick should be inserted to allow the drops to be delivered to the entire length of the canal. A wick of sufficient length should be inserted so that the patient may remove it in 48 hours. Drops are placed on the wick 4 times/day, and thereafter in the canal for a total of 7-10 days. Instructions to observe strict water precautions are of importance (ie, no swimming, and
inserting a vaseline-coated plug in the canal before showering or washing hair). Cotton-tipped applicators or other manipulation by the patient should be avoided. Pain often lasts for 3-4 days after beginning treatment, and should be controlled with sufficient pain medication. Systemic antibiotics are necessary only for cellulitis extending outside the canal, in diabetics or immunosuppressed patients. Systemic antibiotics alone are never sufficient treatment for external otitis. If symptoms persist after 1 week of medical treatment, reexamination is essential. If adequate visualization of the tympanic membrane is not possible initially, a follow-up examination should be performed. It may be necessary to remove debris from the canal on several occasions during the course of treatment.

F. Necrotizing external otitis (sometimes confusingly referred to as malignant external otitis). This condition is an external otitis that has spread outside the confines of the external canal to involve bone, mastoid cells, and periaural soft tissue. It is usually diagnosed in diabetics or those immunosuppressed (including a few reported cases in newborns). Early reports cited a 50-80% mortality, but this rate has been improved with adequate and prompt therapy. Marked reduction in the incidence of necrotizing otitis externa can be achieved with prompt initiation of oral antipseudomonal drug therapy (ciprofloxacin) in those patients at risk for the disease with symptoms of early otitis externa.

1. Signs and symptoms. Pain is usually more severe than with simple external otitis and is often described as "deep" or "boring". The disease process usually begins with an external otitis, but tends to progress on standard medication.

2. Diagnosis

   a. Physical examination reveals granulation tissue at the junction of the bony and cartilaginous canal. There may be exposed bone in the canal. Facial nerve paralysis may be present.

   b. Computerized axial tomography (CAT scan) can aid in the diagnosis by showing destruction of bone.

   c. Cultures should be obtained. Pseudomonas or Proteus are most often the causative organisms.

3. Management consists of high doses of tobramycin and ticarcillin intravenously (often for weeks), topical aminoglycoside drops, and judicious debridement of devitalized bone or areas of accumulated pus. Oral antipseudomonal drugs such as ciprofloxacin may replace IV antibiotics in early cases, and may also allow for early cessation of IV medication. In the diabetic, careful control of the diabetes aids in recovery. The immunosuppressed patient needs aggressive medical intervention.

G. Exostoses. Exostoses are seen as smooth subcutaneous swellings of the bony external canal and are usually asymptomatic unless they entrap water, causing external otitis. In rare cases, exostoses completely close the ear canal, causing a conductive hearing loss, wherein surgical removal is indicated.
H. Dermatitis. As a skin-lined tube, the external canal is subject to dermatitis. Since it is a closed pouch and thereby more prone to maceration by moisture, dermatitis may affect the canals alone. Seborrhea, atopic dermatitis, and psoriasis are common and may predispose to recurrent suppuration. Management involves specific measures for the underlying etiology, as well as topical treatment of any associated external otitis.

I. Tumors. Although rarer than those of the auricle, basal cell and squamous cell carcinomas can involve the external canal. Neoplasia should be suspected when otitis externa is refractory to therapy. Persistent granular or necrotic tissue should be biopsied. An adequate biopsy will establish the diagnosis. Radical surgery is usually required.

VII. Diseases of the tympanic membrane

A. Bullous myringitis. Bullous myringitis may be of viral etiology, although some reported cases have been caused by Mycoplasma infections. Haemophilus influenzae infection can present with tympanic membrane bullae in a child.

1. Signs and symptoms. Pain and a full feeling in the ear are common. The blebs can rupture spontaneously, causing a small amount of serous or serosanguineous drainage. Sensorineural hearing loss has been found in up to one-third of affected patients.

2. Diagnosis is made by the appearance of one or more "blebs" that are thin walled and involve only the squamous layer of the tympanic membrane. There may be an associated effusion in the middle ear. Unless there is a secondary infection, the pain subsides in 24 to 48 hours. The fullness may persist for several weeks.

3. Management consists of symptomatic treatment. An audiogram should be performed. If a new sensorineural component is identified, viral titers should be obtained (ie, EBV and CMV) and adjunctive steroid therapy considered. Narcotic agents may be required for pain control. Puncture of the blebs with a fine needle or myringotomy knife may provide pain relief, but is not usually recommended. Since only the squamous layers of the tympanic membrane is involved, careful puncture of the blebs will not produce a perforation. Antibiotics (eg, ampicillin) are of value when a concomitant otitis media is present.

B. Granular myringitis is an unusual disease of unknown etiology.

1. Signs and symptoms. Symptoms consist of itching, mild pain, and otorrhea. The otorrhea is usually sparse. Symptoms have often been present for many months before the diagnosis is made. The tympanic membrane is covered with granulation tissue that is often obscured by the discharge.

2. Management consists of long-term antibiotic steroid drops (4-6 weeks) and weekly cleaning. Broad-spectrum systemic antibiotics, culture-directed, are sometimes beneficial. In resistant cases, operative curetting of the granulation tissue and coverage with split-thickness grafts become a consideration.
3. **Complications.** Untreated cases can give symptoms for years and ultimately may heal by squamous overgrowth, producing a markedly thickened tympanic membrane and mild conductive hearing loss.

C. **Perforations (traumatic)** result from either direct trauma (eg, cotton-tipped swabs) or pressure transmitted to the closed canal (slap, explosion).

1. **Signs and symptoms** consist of pain, bleeding, a hollow feeling in the ear, and hearing loss. The appearance varies but usually consists of an irregularly shaped perforation with hemorrhage at the edges. Ossicles and other middle ear structures may be visible through the perforation.

2. **Diagnostic tests.** The initial evaluation must include an audiogram to rule out an associated ossicular discontinuity or sensorineural hearing loss. Associated vertigo warrants immediate attention by a specialist. Temporal bone x rays may be necessary to exclude a temporal bone fracture.

3. **Management.** The uncomplicated traumatic perforation usually heals spontaneously. The rate of healing depends on the size of the perforation. Perforations can heal in a few days or may take weeks to months. Perforations that have not healed after 6 months of observation can be repaired. Antibiotic drops are indicated only if there is contamination of the perforation by water or debris. Systemic antibiotics are not indicated. Pain medication may be necessary for the first few days following a perforation. The patient must observe water precautions (vaseline-impregnated cotton plug) until the perforation has healed and should be followed at regular intervals until healing is complete. An audiogram should be obtained at the beginning and end of treatment. With vertigo and a hearing loss - either sensorineural or conductive - an ossicular disruption or perilymphatic fistula is suspect. Emergent surgery may be necessary in this setting.

D. **Slag burns** are a unique types of traumatic perforation caused by hot metal (slag) burning through the tympanic membrane. These burns are usually seen in welders.

**Management.** Early operative intervention is indicated to remove the slag from the middle ear and to close the perforation. These perforations rarely heal spontaneously.

E. **Tympanosclerosis.** Tympanosclerosis is a pathologic condition of the tympanic membrane (and occasionally of the middle ear) consisting of chalky white, plaquelike patches occurring at any site within the membrane. The patches consist of hyaline degeneration of the membrane with calcium deposition and usually result from repeated bouts of inflammation. When localized to the eardrum, tympanosclerosis represents a benign condition.

1. **Signs and symptoms.** Hearing loss is not evident unless extensive involvement of the entire tympanic membrane is present. It is differentiated from cholesteatoma by its chalky white, plaquelike appearance as compared to the pearly white, cheesy appearance of cholesteatoma.

2. **Management.** Treatment is not indicated.
VIII. Diseases of the middle ear and mastoid

A. Acute otitis media (suppurative) is an acute infection involving the middle ear (and mastoid) that is seen in all age groups. It is particularly prevalent in children during the winter months. Acute otitis media often follows or coincides with a viral upper respiratory infection (URI). Children with AIDS (acquired immunodeficiency syndrome) often have recurrent acute otitis media (AOM) as their initial presenting symptom.

1. Signs and symptoms consist of an acute onset with variable ear pain, pressure sensation, or hearing loss. Drainage may be present depending on the stage of infection, and the process may be unilateral or bilateral. The stages consist of:

   a. Hyperemia, a reddened, thickened tympanic membrane.

   b. Exudation with serous fluid in the middle ear space.

   c. Suppuration in which the fluid becomes purulent and the tympanic membrane may perforate.

   d. Resolution in the uncomplicated case in which the tympanic membrane heals, the fluid becomes thin and serous, finally resolving. Physical findings depend on the stage of disease in which the ear is inspected. Resolution can occur at any stage depending on the virulence of the organism, the host resistance, and antibiotic usage.

2. Bacteriology. The most prevalent organisms cultured in acute otitis media are *Streptococcus pneumonia*, nontypable *Haemophilus influenzae* and, to some extent, *Moraxella catarrhalis*. *H. influenzae* type B, group A streptococcus, *Staphylococcus aureus*, gram-negative enteric bacilli and anaerobic bacteria are far less prevalent. In infants and neonates, group B streptococcus and *Escherichia coli* assume more import.

3. Management

   a. Antibiotics. Amoxicillin (30-40 mg/kg/day) is the drug of choice in children under 12 years of age. It has better absorption and fewer side effects than ampicillin, and can be given 3 times/day instead of 4. Treatment should be continued for 10 days at least. In adults, amoxicillin is also the drug of choice. Penicillin-allergic patients should receive trimethoprim-sulfamethoxazole (TMP-SMZ). Up to a 10% failure rate is anticipated for the above medications due to resistant organisms. Frequent examinations are essential, and alternative drug treatment must be considered. These include amoxicillin-clavulanate and oral cefalosporins.

   b. Myringotomy is indicated to establish a bacteriologic diagnosis in patients not responding to conventional medication, in those immunosuppressed, in the neonate, and if complications ensure. Pain control should be obtained via medication, narcotics if necessary.

   c. Topical antibiotics. Drops may be indicated if there is a perforation or if drainage has produced a secondary external otitis. Decongestant preparations that include antihistamines do not shorten the course of the disease. Vasoconstriction (pseudoephedrine) may alleviate
associated symptoms of pressure or the nasal congestion of an upper respiratory infection (URI).

B. Mastoiditis (acute coalescent). Mastoiditis is an unusual entity since the advent of antibiotics. Untreated, about 1-5% of cases of acute otitis media progress to mastoiditis. Treated, the incidence is much lower. The pathogenesis involves a blockage of the additus with granulation tissue or swollen mucosa so that free drainage of purulent material cannot occur. This complication leads to pressure in the mastoid cells with breakdown of cell partitions.

1. Signs and symptoms include continued pain, low-grade fever, malaise, and hearing loss. Drainage is inconsistently present, but when present has been noted to change from thick, mucopurulent secretions to thinner, foul-smelling secretions. Physical examination reveals a thickened, sometimes bulging tympanic membrane. There is thickening of the mastoid cortex with a somewhat doughy feel, a sagging of the posterosuperior canal wall, and later a protrusion of the ear.

2. Laboratory data include an elevated white blood count (WBC) with a left shift. X rays show breakdown of the normal cell partitions of the mastoid, best demonstrated on CT. (It should be noted that in acute otitis media with effusion, x rays will show clouding of the mastoid. This clouding should not be confused with acute mastoiditis, because it represents fluid in the mastoid cells, not cell breakdown.)

3. Management. Unless an abscess is present, the management includes a myringotomy to decompress the middle ear and provide for culture and sensitivity. Pending culture-directed specificity, an acceptable initial choice of antibiotic for intravenous usage is ceftriaxone, which covers most pathogens. If the process does not resolve, a complete (cortical, simple) mastoidectomy should be performed.

C. Complications of acute otitis media. With the exception of mastoiditis, complications can occur at any stage of acute otitis media. Whether or not complications occur depends on the virulence of the organism, the resistance of the host, anatomic abnormalities, and the institution of appropriate antibacterial therapy. The complications can be divided into:


2. Intracranial complications. Meningitis, epidural abscess, subdural abscess, brain abscess, and sigmoid sinus thrombosis. It should be noted that otitis media is still the most common cause of meningitis (excluding meningococcus) and is the most common cause of brain abscess.

3. Treatment consists of managing the complication and directing attention to the otitis media. A myringotomy is indicated to establish bacteriologic specificity. Facial nerve paralysis usually resolves spontaneously after adequate treatment of the otitis media. All of these complications, of course, constitute medical emergencies.
D. Acute necrotizing otitis media. This unusual form of otitis media occurs most frequently in children with severe systemic disease (eg, measles).

1. Signs and symptoms. In a few hours, a large perforation develops and may be associated with destruction of the ossicles. The perforation is frequently kidney shaped. The organism involved is most often beta-hemolytic streptococcus, although *S. pneumoniae* has been cultured.

2. Treatment consists of high doses of a semisynthetic penicillin. Secondary operative repair of the perforation must await the appropriate age in children (> 10 years).

E. Eustachian tube dysfunction. Eustachian tube dysfunction has a wide clinical spectrum from very mild to chronic otitis media, as described below.

1. Signs and symptoms. The mildest symptoms consist of a blocked or hollow feeling, pressure, mild otalgia, and occasional crackling or popping noises in the ear. These often accompany a URI or allergy.

2. Diagnosis. Otologic examination is normal except that the drum moves sluggishly or not at all during a Valsalva maneuver. Tympanometry may reveal a flattened curve or negative pressure. Audiologic examination is normal.

3. Management. These mild symptoms are self-limiting in most patients. Antihistamines and decongestants may help in lessening the symptoms but not their duration. If the symptoms are allergy-related, the underlying cause should be treated. Repeated Valsalva maneuvers may alleviate the symptoms.

F. Hyperpatent (patulous) eustachian tube is usually seen in patients who have undergone a rapid weight loss or suffer from disorders of muscle wasting. Estrogen has also been associated with the syndrome.

1. Signs and symptoms may be much the same as with eustachian tube dysfunction - a "hollow" or "stopped-up" sensation and pressure. Patients often state that they can hear their own breathing. Short periods of recumbency relieve the symptoms temporarily.

2. Diagnosis is made by observing the tympanic membrane while the patient occludes one nostril and breathes with the mouth closed. The drum will move with respiration.

3. Management consists of treating (or removing) the underlying cause (such as birth control medications). With persistent symptoms, tympanotomy tube insertion may give relief. Teflon injections near the eustachian tube orifice are now used infrequently. Rarely, however, is this disorder a persistent problem.

G. Otitis media with effusion (OME). OME describes a nonpurulent effusion in the middle ear space. The fluid varies from thin to mucoid. Mucoid fluid usually signifies a more chronic process. The fluid is secondary to obstruction of the eustachian tube. Defining the cause of the obstruction is indicated prior to initiating therapy whenever possible. The varied etiologies include inflammation (bacterial, viral, allergic), congenital malformation, polyps or
tumors of the nasopharynx, hypertrophied adenoids, cleft palate, radiation, endocrine, or iatrogenic. Serous fluid is commonly seen in the resolution stage of acute otitis media. OME is the most common cause of hearing loss in children. It is less frequent in the adult and, when seen, the nasopharynx should be carefully evaluated for malignancy.

1. **Signs and symptoms.** The tympanic membrane may appear normal. Usually it is slightly retracted even in early onset. Fluid, with or without bubbles, may be seen and may be amber in color. With thicker fluid, the amber color is not prevalent, and the drum is dull. Prominence of the vessels is not infrequent, but in contrast to acute otitis media, the margins are distinct.

2. **Diagnostic tests.** Pneumatic otoscopy reveals diminished or no movement of the tympanic membrane. Tuning fork tests and audiometry usually reveal a conductive hearing loss. Conductive loss on audiometry should not exceed 30-40 dB with serous otitis media.

3. **Management**

   a. In older children autoinflation and politzerization of the eustachian tube are possible, sometimes effecting resolution. If the fluid results from a resolving otitis media, the process is usually self-limiting, 90% in 3 months. Decongestants and antihistamines have not proved to be effective management for OME. Recognizing that a small percentage of serous effusions contain bacteria or may be associated with nasopharyngeal or eustachian tube infection, often a 6- to 8-week therapeutic trial of an antibiotic (amoxicillin or TMP-SMZ) is warranted. Pressure-equalizing (PE) tubes (ventilation tubes) are indicated in refractory cases to alleviate hearing loss and arrest development of permanent tympanic membrane and ossicle malfunction. Adenoidectomy is probably beneficial in selected instances of recurrent OME and acute otitis media.

   b. For adults an underlying cause should be determined if possible. CT scans of the nasopharynx and sinuses are indicated to rule out infection or tumor. With a suggestive history, an allergy evaluation is indicated. Myringotomy with or without tube insertion may be indicated in selected instances.

H. **Barotrauma (aerotitis).** Barotrauma results from a change in atmospheric pressure with an occluded eustachian tube. This usually occurs during scuba diving or during descent when flying.

1. **Signs and symptoms** include pressure, pain (often severe), and hearing loss. Often, there is a concurrent URI or other cause of eustachian tube congestion. Examination reveals a dull drum with fluid behind it. The fluid may be bloody. Hemorrhagic areas in the tympanic membrane are frequent. Tuning fork tests and audiometric evaluation usually define a conductive hearing loss.

2. **Management.** The fluid can take several weeks to clear. Simple decongestants may alleviate the pressure sensation. Mild analgesia may be necessary in the acute phase. Instruction in pressure-equalizing techniques (Valsalva; gum chewing; prophylactic vasoconstrictors; both oral and nasal) is warranted to prevent further episodes. Pressure change with severe nasal congestion should be avoided.
I. Chronic otitis media. As opposed to acute otitis media, chronic otitis media is a surgical disease in most instances. While the name suggests a lengthy process, it would be more accurate to say that irreversible changes in the middle ear or mastoid have taken place. Depending on the process, the disease can be either dangerous (implying an extending process) or benign. A chronic perforation that is dry and noninflamed is an example of a benign process, as is chronic adhesive otitis media, in which fibrous tissue has replaced a chronic inflammatory process. Cholesteatoma, on the other hand, is an example of a potentially dangerous process. In this disease, squamous epithelium enters the middle ear and mastoid and expands, causing bony erosion. This entrapped skin may become infected, which hastens the erosive process and can spread to surrounding structures (labyrinth, meninges, sigmoid sinus, brain facial nerve).

1. Signs and symptoms may be noticeably absent in chronic ear disease. Pain is unusual, except when there is active acute infection. Discharge may be present and intermittent. Hearing loss is inconsistent. The presenting complaint may be a complication (eg, vertigo or facial nerve paralysis) without any noted prior history of ear disease. Physical examination may reveal a perforation or cholesteatoma. A cholesteatoma in this setting is most often identified in the pars flaccida (attic) and may be obscured by a crust. There may be an associated discharge, which must be removed for the correct diagnosis to be made.

2. Diagnostic tests

   a. Audiometry can identify a mild to marked hearing loss, usually conductive or mixed, depending on the extent of destruction.

   b. CT scanning of the temporal bone can identify bony destruction in the attic and mastoid area.

3. Management consists initially of treatment of any associated acute infection with both systemic antibiotics (culture appropriate) and topical antibiotic drops. Subsequent surgery is often required. Evidence of a complication such as vertigo, facial nerve paralysis, or brain abscess requires immediate intervention by an otolaryngologist.

J. Tuberculous otitis media is more frequent in children, but may occur in any age group. It can occur with or without evidence of pulmonary or other site involvement.

1. Signs and symptoms consist of drainage from the ear and occasional lymphadenopathy. Pain is not common, but hearing loss is frequent. Early in the disease, the tympanic membrane appears grayish-yellow. Tuberculous otitis media usually progresses to multiple perforations.

2. Diagnosis is made by acid-fast smear and culture.

3. Management is first medical, with antituberculous drugs; and second, surgery with repair of the tympanic membrane and ossicular chain after the infection has been completely eradicated. HIV infection should be excluded.
K. Syphilis can mimic tuberculosis in the clinical appearance of the ear.

1. **Signs and symptoms.** In addition to the signs and symptoms in J.1., a fluctuating sensorineural hearing loss and vertigo may be present in both the tertiary and congenital forms (see IX.A.3.c.). An osteitis of the ossicles is present.

2. **Diagnostic tests.** Diagnosis is made either by darkfield examination of the exudate or by serologic tests.

3. **Management** consists of treatment of the syphilis with penicillin or steroids or both.

L. Conductive hearing loss. In addition to the processes mentioned in VI-VIII, certain disease entities cause conductive hearing losses. The remainder of the examination is usually normal.

1. **Otosclerosis** involves fixation of the stapes by the otosclerotic process and is most often bilateral.

   a. **Signs and symptoms.** This entity presents as a progressive hearing loss, conductive in nature. A family history of hearing loss is often present. Females are more frequently affected than males (2.5:1.0). The remainder of the examination is normal.

   b. **Treatment** consists of surgical replacement of the fixed stapes or the use of a hearing aid.

2. **Other ossicular fixations** can occur as congenital aberrations from inflammation or trauma. The diagnosis is made at the time of surgical exploration.

M. Tumors

1. **Malignant.** As with the auricle, squamous cell carcinoma and basal cell carcinoma can involve the middle ear, either primarily or by extension from surrounding structures. Malignant parotid tumors can likewise involve the temporal bone. Metastatic disease to the temporal bone has been reported.

   a. **Signs and symptoms** include pain and drainage (from secondary infection) that is sometimes bloody. Since these tumors are often associated with chronically draining ears, pain in these patients should raise the suspicion of malignancy and lead to biopsy.

   b. **Other malignant tumors, lymphomas, and rhabdomyosarcomas** infrequently involve the ear, and then usually in a younger age group.
2. Benign tumors

a. Polyps can protrude through a perforation and imply a chronically infected area.

(1) Symptoms are usually those of a draining ear.

(2) Treatment consists of topical antibiotic and steroid drops. Polyps should not be pulled out, because they can be attached to middle ear structures. However, judicious surgical removal may aid in the diagnosis and also can enhance drainage and, thereby, treatment. A polyp frequently results from an underlying chronic otitis media and should be evaluated as noted above (see I.).

b. Glomus tympanicum and jugulare (chemodectomas). These tumors usually arise from paraganglionic cells in the tympanic plexus of the middle ear. Glomus jugular tumors are chemodectomas arising from the jugular bulb.

(1) Signs and symptoms are that of a stuffy feeling, hearing loss, and typically a pulsating tinnitus. On physical examination, a bluish mass is visible behind and intact tympanic membrane.

(2) Diagnostic tests. Plain radiographs may reveal bony destruction. Angiography, contrast study CT scans, or MRI scans can demonstrate the lesion.

(3) Management primarily consists of surgical extirpation. Biopsy is usually contraindicated since the tumors are very vascular. Radiation therapy or embolization should be considered in large tumors.

c. Congenital cholesteatoma is a misnomer, in that these "tumors" are formed from embryonic cell rests of ectoderm and may involve any area of the temporal bone.

(1) Signs and symptoms depend on the size of the lesion and the area involved. Usually, a white mass is identified behind an intact tympanic membrane in children. In adults, congenital petrous apex cholesteatomas can present with hearing loss or facial paralysis.

(2) Management is surgical.

IX. Diseases of the inner ear. Diseases of the inner ear may involve the cochlea, producing sensorineural hearing loss, or the vestibule, producing vertigo, or both. Some disease processes described below are assigned arbitrarily to one system or another, even though both systems may be involved.

A. Diseases of the cochlea

1. Sudden hearing loss (idiopathic sudden deafness). The syndrome known as sudden hearing loss (SHL) can be defined as the sudden onset of a sensorineural hearing loss without preexisting ear pathology or another accountable etiology. To establish a diagnosis of SHL, known etiologies for acute sensorineural deafness must be excluded. The more common etiologies are listed below.
a. **Infection.** Mumps, herpes zoster, CMV, meningitis, encephalitis, syphilis, otitis media.

b. **Trauma.** Head injury (with or without fracture), noise trauma, barotrauma.

c. **Vascular.** Embolism, coagulopathy, cerebrovascular accident.

d. **Otologic.** Ménière's disease, acoustic neuroma, perilymph fistula, cholesteatoma.

e. **Other.** Multiple sclerosis, malignant tumor (metastatic or primary), drug toxicity, Cogan's syndrome (hearing loss associated with nonsyphilitic interstitial keratitis), diabetes, autoimmune disorders. Idiopathic SHL is most often unilateral, but may be bilateral in a small number of cases. Controversy exists as to its etiology, but viruses and vascular syndromes are most often postulated.

(1) **Signs and symptoms.** The patient typically gives a history of an abrupt onset of hearing loss over a period of minutes to hours. Depending on the severity and rapidity of onset, associated symptoms may be vague. Tinnitus often accompanies the loss. Pain is virtually never present. Vertigo, usually brief in duration, occurs in a significant number of patients. The otologic examination is negative except for the hearing loss.

(2) **Diagnostic tests.** Audiometric testing typically shows a sensory loss, cochlear in origin, that ranges from mild to severe. Calorics and ENG may reveal a canal paresis but also be normal. ABR confirms the cochlear nature of the hearing loss. Radiographs are helpful in excluding other causes of hearing loss (cholesteatoma, acoustic neuroma), but are normal in SHL. Other laboratory data may be normal. The sedimentation rate can be elevated.

(2) **Management.** Recovery typically occurs in 50-60% of patients. Young patients with mild hearing loss, particularly in those who have less loss in the higher frequencies (upward sloping loss), have a better prognosis. Recent evidence indicates that if the patient is seen within 24 hours of the onset of SHL, high-dose steroids (60 mg prednisone/day), tapered over a 2-week period, can improve the recovery rate.

2. **Noise-induced hearing loss.** Exposure to extreme noise can diminish the ear's capacity to detect pure tones. This chance can be temporary or permanent.

a. **Temporary threshold shifts**

(1) **Short-term masking** lasts a fraction of a second. It is thought to be due to the refractory period of nerve fibers after discharge. The tone most affected is the tone of the "noise", up to a level of input of 70 dB. Above 90 dB, a tone half an octave above the noise is the tone that is most affected. Recovery is swift, being exponential in time, and is independent of the length of the inducing noise.

(2) **Ordinary temporary threshold shifts** last usually more than 2 minutes, but less that 16 hours. The threshold shift increases linearly with intensity above a level of 70-75 dB. Below 70-75 dB no threshold shift is seen, even with indefinite exposure. High frequencies produce a greater response than low frequencies, with the greatest sensitivity at 3000 Hz.
Maximum shift is seen at a frequency half an octave above the present noise. Therefore, the greatest effects to broad-band noise will be seen at frequencies between 4000 and 6000 Hz.

If exposure is intermittent, the effect is the same as that seen for the mean exposure level over that time period. As the quiet intervals between noise exposure lengthen, partial recovery occurs and may lead to an underestimation of the noise effects that are actually seen. The above reasoning does not hold well for impulse noise either. These effects seem to be poorly predicted at present due to difficulty in quantifying the impulse parameters.

3) **Prolonged temporary threshold shifts.** If a threshold shift of greater than 40 dB is produced, recovery is neither complete in 16 hours nor exponential. Instead, recovery is linear and may require days to weeks before full recovery occurs.

4) **Permanent threshold shift.** When noise exposure is loud and long enough, complete recovery does not occur, leaving a permanent threshold shift in pure tone perception. Whether this is due to a buildup of minor traumas or to a few major events is still debatable. In one of the few studies attempting to deal with this complex problem, Passchies and Vermeer have shown that exposure for 8 hours/day for 5 days/week for 10 years, at levels of 80-90 dB, results in a permanent shift in hearing thresholds, the effect being greatest at 4000 Hz and proportional to the level of exposure. This finding correlates with the 4000-Hz notch typically associated with noise-induced hearing loss. Recruitment and paracusia, signs of cochlear injury, often accompany the losses and worsen the disability. More annoying to many is the tinnitus that also occurs.

b. **Pathology.** Noise trauma is associated with outer hair cell injury (see I.A.5.). This injury can range from simple disruption of the stereocilia to total hair cell loss or disruption of the organ of Corti. The exact mechanism of injury is still unsettled, with the line between temporary and permanent shifts far from clear. With exposure to a sudden loud noise, the response seems to be mechanical; however, with long-term exposure, two theories have been proposed.

1) **Mechanical theory.** Long-term exposure is accompanied by episodes of occasional loud noise leading to intermittent loss of hair cells. The loss gradually accumulates to a significant level.

2) **Chemical theory.** Long-term exposure is accompanied by gradual buildup of toxic metabolites or depletion of required chemicals necessary for maintenance of cell function and viability. Ischemia may be a contributing factor. Hyperlipidemia both in children and adults increases the cumulative negative effect of noise exposure. The triad of hyperlipidemia, hypertension, and noise exposure has an additive deleterious effect, which is probably vascular in origin.

Regardless of which theory is used, the focal point is the hair cell, with the outer hair cells being affected before the inner hair cells.

c. **Management.** Treatment necessitates reduction in noise exposure below the level that causes permanent threshold shifts. This reduction may be difficult to attain. Commercial earplugs may be of value if a noisy environment cannot be avoided. Specially designed
acoustic plugs for musicians, which give better balance to the sound reduction, are available. Patients must be aware of the negative impact of hyperlipidemia and hypertension with continued noise exposure. Treatment of these associated conditions should be instituted. As for treatment of an already existing hearing loss, little except hearing aid amplification and lip reading is of benefit.

3. Inflammation

a. Bacterial. Sensorineural hearing loss due to bacterial infections can be the result of either direct bacterial invasion or of diffusion of bacterial toxins into the inner ear. Bacteria can invade via the bloodstream, CSF, cochlear aqueduct (channel connecting CSF to perilymphatic space), internal auditory canal, or directly through the middle ear. This invasion can produce a serous or suppurrative labyrinthitis.

(1) Serous labyrinthitis is caused by a diffusion of bacterial toxins into the inner ear, producing an inflammatory response.

(a) Signs and symptoms are those of a sensorineural hearing loss and vertigo. A marked nystagmus and sensorineural loss are found on physical examination. Serous labyrinthitis is usually associated with an acute otitis media or chronic otitis media with acute infection.

(b) Management should include IV antibiotics such as ampicillin in high doses, myringotomy to promote drainage and to obtain culture, surgical removal of chronically infected tissue, or all three, when appropriate. A partial to full recovery of the inner ear function is possible if treatment is prompt.

(2) Suppurative labyrinthitis

(a) Signs and symptoms. The symptoms and physical findings are the same as with serous labyrinthitis, except that the patient is more toxic and the hearing loss is profound. WBC is elevated with a left shift.

(b) Treatment is the same as with serous labyrinthitis and is directed to prevent the spread of infection to adjacent structures (meninges, brain). An antibiotic with less bacterial resistance than ampicillin, such as ceftizomine, should be considered in this setting. Recovery of hearing does not occur.

(3) Chronic otitis media. Mild to moderate sensorineural hearing loss can occur with low-grade infection due to diffusion of bacterial toxins through the round, or oval, window into the cochlea. The vestibule is not involved.

(a) Signs and symptoms are a slowly progressive sensorineural hearing loss (with or without a superimposed conductive loss due to destruction of the ossicles).

(b) Treatment is as for chronic otitis media, to produce a safe, dry ear (see VIII.I.).
**b. Viral.** Mumps, rubella, rubeola, varicella, influenza, herpes zoster, cytomegalovirus, adenovirus, and others have been shown to produce a sensorineural hearing loss through invasion of the inner ear. Effective management is through vaccination programs; however, steroids may have efficacy in the acute situation. Several of the viruses are important enough to deserve special mention.

1. **Mumps** is the most common cause of unilateral sensorineural hearing loss in children. Rarely does it cause bilateral loss.

2. **Rubella** produces a symmetric, bilateral loss, greatest in the higher frequencies.

3. **Herpes zoster (Ramsay Hunt syndrome)** produces burning aural pain, vesicular eruptions in the external canal and concha, and sensorineural hearing loss. Vertigo and facial paralysis are often associated with the infection. Early treatment of patients with acyclovir and prednisone has markedly improved the prognosis for return of facial nerve function.

**c. Granulomatous disease.** **Syphilis** can cause sensorineural hearing loss in either the congenital or acquired form. In either case, there is a mononuclear cell infiltration of the inner ear, leading to an obliterative endarteritis. A progressive endolymphatic hydrops is also seen.

1. **Signs and symptoms** include a fluctuating hearing loss and episodic vertigo. Tinnitus is also present. The symptoms, therefore, mimic Ménière's disease. In the congenital form, pathologies such as Hutchinson's teeth and saber shins may be present. A conductive component of the hearing loss may be present if there is an associated osteitis.

2. **Diagnosis.** Examination of the ear may be normal. The diagnosis is confirmed by a positive FTA-ABS serology. (A negative VDRL or RPR card test cannot exclude the diagnosis.)

3. **Management** is long-term therapy with penicillin and systemic steroids. The length of treatment is debatable. Treatment can stop the progression of the hearing loss and, in some cases, afford partial recovery. Vertigo may continue despite therapy and should be treated with meclizine, Dramamine, or similar medications.

4. **Trauma**

   **a. Temporal bone fractures** can be classified as longitudinal, transverse, or mixed, depending on their angle to the axis of the temporal bone.

   1. **Longitudinal** fractures produce a conductive hearing loss and are discussed in Chapter 1.

   2. **Transverse** fractures occur in about 15% of temporal bone fractures and are usually caused by a blow to the occiput. The fracture line extends from the foramen magnum across the petrous apex and through the internal auditory canal. The eustachian tube or jugular foramen is often involved.
(a) **Signs and symptoms.** A direct fracture of the inner ear is common, producing a profound sensorineural hearing loss and vertigo. The tympanic membrane is often intact. Facial nerve paralysis occurs in about 50% of these injuries.

(b) **Management** consists of decompression and, if necessary, reapproximation of the facial nerve. The hearing loss is irreversible. Vertigo is treated by bed rest, meclizine, Dramamine, or diazepam.

(3) **Mixed** fractures are the most frequent types of injury and can produce any symptoms noted previously as well as other neurologic abnormalities.

**Management** depends on the extent of injuries.

(4) **Labyrinthine concussion.** Closed head injuries can cause sensorineural hearing loss by sudden deceleration and rapid alteration of the hydrodynamics of the inner ear. In rare cases, the seventh cranial nerve may be sheared. Vertigo may be present along with the hearing loss.

**Management** is directed at control of vertigo through antivertiginous drugs (meclizine, Dramamine, or diazepam).

5. **Nonorganic hearing loss.** A decrease in hearing not associated with organic changes may be deliberate (malingering) or not. The ability to verify this type of hearing loss is directly correlated with the audiologist's suspicions and ingenuity in testing the subject. Several types of testing are available.

a. **Stenger.** A tone is presented to the better ear at a level of 10 dB above threshold, so that the subject responds 100% of the time. A tone is then simultaneously presented to the poorer ear, starting at 0 dB and gradually increasing. The level at which the patient ceases to respond or notes a change to the better hearing ear should be 10-20 dB above the measured threshold of the poorer ear.

b. **Delayed feedback.** The patient reads a paragraph. His voice is played back with a short delay (0.15-0.18 seconds) at a decibel level below the measured threshold. He will become confused or slow down his reading if the actual threshold is below the level of the playback.

c. **Swinging voice test.** A story is read to the patient with key words entering only one ear at a time. The patient is then tested for comprehension.

d. **Auditory brainstem response (ABR).** This test is an objective recording of auditory-evoked brainstem responses. The accuracy of this test in determining hearing thresholds is well established and it should be considered definitive in diagnosing a nonorganic loss.

e. **Management** in malingering consists only of establishing normal thresholds with ABR. When a nonorganic loss lacks a conscious component, psychiatric evaluation is necessary to determine and to treat the underlying cause.
6. Endocrine

a. Diabetes cause diffuse vascular changes throughout the body, and the inner ear is no exception.

(1) Signs and symptoms. A bilateral progressive sensorineural hearing loss greater in the higher frequencies often accompanies diabetes. Sudden hearing loss has also been reported. Vertigo may be present. No correlation exists between the duration or severity of the diabetes and the severity of the hearing loss. It is not known if better control of the blood sugar levels decreases progression of the hearing loss.

(2) Management consists of amplification when necessary.

b. Thyroid

Myxedema. Hypothyroidism causes an increase in acid mucopolysaccharide accumulation. Hyperthyroidism has not been associated with hearing loss.

(1) Signs and symptoms. If acid mucopolysaccharide deposition occurs in the eustachian tube or middle ear, a serous otitis with a conductive hearing loss ensues. The evolution of the sensorineural component remains unclear.

(2) Management consists of thyroid hormone replacement, which in some instances partially reverses the hearing loss.

7. Congenital

a. Hereditary. Genetic causes of sensorineural hearing loss can be grouped in many ways to emphasize associated organ systems involved, mode of inheritance, or pathologic changes.

(1) Mode of inheritance. Defects are either dominant, recessive, X-linked, or involved quantitative changes in chromosomal material. The type of hearing loss seems to show some relationship to the mode of inheritance.

(a) Dominant syndromes usually show flat hearing losses and may be progressive throughout one's life. Their penetrance is variable, leading to a large range of disabilities. Examples are Waardenburg's disease, Schäfer's syndrome, Huntington's chorea, and von Recklinghausen's disease.

(b) Recessive syndromes tend to show loss of hearing greater in high frequencies with some retention of low-frequency hearing. They tend not to be associated with other congenital lesions and not to be progressive throughout life. Examples are Hurler's syndrome, Morquio's syndrome, Tay-Sachs disease, Wilson's disease, and Usher's syndrome.

(c) X-linked syndromes are less common. There seems to be retention of hearing at all frequencies (Hunter's and orofacial digital I syndrome).
(d) Quantitative changes in chromosomal material are evident in the trisomy syndromes. These syndromes tend to have severe hearing loss associated with multiple organ defects, such as trisomy 13 and trisomy 18.

(2) Associated organ system. See Genetic and Metabolic Deafness by Konigsmark and Gorlin.

(3) Pathologic changes. Temporal bone studies show that the pathologic correlates in congenital hearing loss can range from submicroscopic lesions not visible by present methods of study to total agenesis. Several abnormal patterns of developments seem to occur more frequently.

(a) Scheibe's deformity. There is a normal bony labyrinth, a normal utricle and semicircular canals, and a limited development of the stria vascularis with few hair cells and support cells in the organ of corti. Reissner's membrane is collapsed on the stria and organ of Corti.

(b) Mondini deformity. Vestibular structures may be underdeveloped. Only the basal coil of the cochlea is developed with the mid- and apical turns incomplete, making the cochlea only about one and one-half turns. The saccule and endolymphatic sac are dilated.

(c) Michel's deformity. There is a total lack of development of the inner ear.

b. Acquired. There are many causes of congenital hearing loss that are not genetic in origin. Below is a list of the most common.

(1) Rubella (German measles). When contracted by the mother during the first trimester of pregnancy, rubella causes a group of defects. Temporal bone studies show anything from complete agenesis to the more common Mondini deformity. There is degeneration of the membranous portion of the cochlea and saccule. The hearing loss is usually asymmetric between ears, with a flat audiogram. Other associated anomalies include mental retardation, microcephaly, cardiac defects (usually patent ductus arteriosus), dental defects, and congenital cataracts.

(2) Erythroblastosis fetalis. Lesions in this condition are thought to be due to a high bilirubin level and can be prevented by keeping the level low with exchange transfusions and ultraviolet lighting. The loss often occurs in higher frequencies and may relate to changes in the cochlea. Mental retardation and cerebral palsy are also associated with this condition.

(3) Thyroid disease (cretinism). A mixed sensorineural and conductive loss is present in cretin children. The middle ear can be quite deformed, without significant change in the inner ear. Mental and physical growth are also retarded.

(4) Birth injury. Birth injury includes prematurity, hypoxia, prolonged labor, toxemia of pregnancy, and anesthesia. The hearing loss, when present, is usually bilateral, symmetric, and mainly in the higher frequencies.
(5) **Drugs.** The effects of most ototoxic drugs given to the pregnant mother are not known; however, two drugs - thalidomide and quinine - have had unfortunate clinical trials. Thalidomide can produce any type of inner ear lesion, and quinine usually produces a bilateral deafness.

(6) **Meningitis** can produce a mild to profound hearing loss due to spread of infection to the labyrinth, producing a labyrinthitis.

(7) **Management** should be directed at early diagnosis and appropriate amplification and education. Evoked response audiometry can establish a diagnosis early in life and enhance the chances for a child attaining full potential. Cochlear implantation appears to hold promise for children 2 years of age and older with profound sensorineural hearing loss.

8. **Presbycusis** is a loss of the ability to perceive or discriminate sounds due to the aging process. It is the most common cause of hearing loss in humans. The audiometric pattern is dependent on the type or combination of types of presbycusis present.

Discrimination may or may not be significantly impaired. The diagnosis is based on purely clinical grounds, mainly by the exclusion of all other possible causes of hearing loss. Positive proof of the diagnosis is found only at autopsy with temporal bone sections. Changes in the middle ear are known to occur with aging. The tympanic membrane thickens and the ossicular joints can undergo arthritic changes. There is no evidence, however, that these alterations have a significant effect on hearing. There are at least four histologic variants in the inner ear and its neuronal connections that alone or in combination seem to explain the audiometric findings of presbycusis.

a. **Sensory.** Characterized by atrophy of the organ of Corti in the basal end of the cochlea with loss of hair cells and support cells. There is also neural degeneration, which is thought to be secondary to hair cell loss. The audiogram shows a loss of hearing in the high frequencies - 4000-8000 Hz.

b. ** Neural.** Due to loss of neurons in the cochlear nerve, especially in the basal turn. Minimum loss in the pure tones is noted, but marked loss of discrimination is present.

c. **Strial atrophy** caused by atrophy of the stria vascularis. While this atrophy is greatest at the apical end, the loss is equal for all frequencies because the endolymph is distributed throughout the cochlea.

d. **Cochlear conductive.** Atrophy of the spiral ligament alters the shape of the cochlear duct, leading to loss of hearing. This degeneration is greatest at the basal end and progresses to the apical end. The audiogram shows a straight line greatest at the higher frequencies.

e. **Management** consists of careful evaluation and appropriate amplification.

9. **Ototoxicity.** Toxic effects on the inner ear have been observed with a wide variety of therapeutic and chemical agents. Aminoglycoside antibiotics, chloramphenicol, erythromycin, ethacrynic acid, furosemide, salicylates, and quinone are but a few drugs in
common usage that produce ototoxicity. In addition, chemotherapeutic agents used in the
treatment of cancer (eg, nitrogen mustard and cisplatinum) can be toxic to the inner ear. The
toxicity usually includes both the cochlea and vestibule, but in some drugs one effect may
precede the other. The effects may be temporary (salicylates, ethacrynic acid) or permanent
(aminoglycosides) and, in most patients, are directly related to the blood levels of the agent.
In some instances, two agents given together may produce more severe ototoxicity than blood
levels of either agent might suggest (eg, aminoglycoside and furosemide).

a. **Signs and symptoms** are usually that of tinnitus, vertigo, or unsteadiness and
hearing loss. Since most of these agents are used in the extremely ill, the symptoms can be
overlooked.

b. **Diagnosis.** Examination reveals normal canals and drums. Nystagmus may be
present. Tuning fork testing is consistent with a sensorineural hearing loss. The diagnosis can
be confirmed by serial audiograms and vestibular testing that shows progression of the loss.

c. **Management.** Prevention is the only management. Serum levels of ototoxic drugs
should be carefully monitored, when appropriate, and the patient questioned frequently about
ototoxic symptoms. Ototoxic drugs, particularly aminoglycosides, should not be used if
another less toxic agent will produce similar therapeutic results.

10. **Autoimmune hearing loss.** Progressive sensorineural hearing loss that occurs over
a period of months may have an autoimmune etiology.

a. **Signs and symptoms.** The pattern of loss is that of bilateral progressive
sensorineural hearing loss. The loss is commonly asymmetric and may fluctuate. The trend,
however, is a fairly rapid loss unassociated with any obvious etiology.

b. **Associated symptoms** may include those found in many autoimmune disorders,
including myalgias, arthralgias, dry eyes, and dry mouth, to name a few.

c. **Diagnostic tests.** An autoimmune hearing loss is considered when faced with a
progressive sensorineural hearing loss. An elevated sedimentation rate, positive antinuclear
antibodies, positive rheumatoid factor, and positive lupus erythematosus (LE) prep all give
supportive evidence for the entity, but are not specific or sensitive. More sophisticated
laboratory tests are being developed to bridge the diagnostic gap.

d. **Treatment** for autoimmune hearing loss involves the use of high-dose steroids. The
exact dosage and duration of treatment are still being evaluated. Treatment for 1-3 months is
not uncommon. Chemotherapeutic agents are also being evaluated to prevent deafness in
patients who cannot tolerate long-term steroid therapy or who fail to respond to steroids.

11. **For profound hearing losses,** traditional management has always centered on
hearing aids and rehabilitation utilizing the other senses. Cochlear implants have now added
to this rehabilitation armamentarium. Cochlear implants can give patients environmental
awareness and, at times, an understanding of sounds. They are presently approved for children
over 2 years of age and for adults. Future developments may allow for their use in less
severely handicapping losses and for younger children.
B. Diseases of the vestibular system (vertigo)

1. Signs and symptoms. Dizziness is an extremely frequent complaint. In evaluating this symptom, it is first necessary to identify those patients in whom dizziness is a symptom of a vestibular disorder. Careful questioning is often necessary. Vertigo is defined as a sensation of motion when no motion is present and almost always indicates disease of the vestibular system. The motion may be described as the patient moving or his or her environment moving. When motion is not noted by the patient, both vestibular and nonvestibular causes of dizziness may be accountable. Some of the more common nonvestibular causes of dizziness are listed below:

   a. Eye disorders (refractive, muscle imbalance).
   b. Hypotension (postural and nonpostural).
   c. Anemia.
   d. Syncope.
   e. Hypoglycemia.
   f. Paroxysmal atrial tachycardia.
   g. Heart block.
   h. Drug-induced causes (barbiturates, tranquilizers).
   i. Psychogenic.
   j. Hyperventilation.

2. Peripheral versus central vertigo. The history can be very helpful in distinguishing peripheral vertigo (diseases of the end organ) from central vertigo (eg, eighth cranial nerve, brainstem vestibular nuclei, medial longitudinal fasciculus, cerebellum, vestibulospinal tract). Although not completely foolproof, the characteristics outlined below are helpful:

<table>
<thead>
<tr>
<th>Severity</th>
<th>Peripheral</th>
<th>Central</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Most often very severe</td>
<td>Less severe, usually no nausea or vomiting</td>
</tr>
<tr>
<td></td>
<td>with associated nausea and vomiting</td>
<td></td>
</tr>
<tr>
<td>Hearing loss</td>
<td>Often present</td>
<td>Seldom present (except with acoustic neuromas)</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>Often present</td>
<td>Seldom present (except with acoustic neuromas)</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>Horizontal (if present)</td>
<td>Horizontal or vertical (if present).</td>
</tr>
</tbody>
</table>

3. History

   a. Distinguish true vertigo (the illusion or sensation of whirling or falling) from syncope, disequilibrium, or lightheadedness. Does the patient have a sensation of motion or of objects moving about him or her? Associated illness or incident (eg, URI, otitis media, during airplane descent or exertion).
b. Characteristics of attacks:

(1) Episodic or continuous.
(2) Onset and duration (minutes, hours, days).
(3) Severity (nausea, vomiting, ataxia).
(4) Vertigo with nausea and vomiting is most likely end organ disease in the absence of acute central nervous system (CNS) problems.
(5) Aura or prodroma (eg, tinnitus, fullness, hearing loss).
(6) Course (subsiding, unchanged, worsening).

c. Associated symptoms

(1) Hearing loss or fluctuation.
(2) Tinnitus.
(3) Otalgia or otorrhea.
(4) Facial palsy.

d. Presence or absence of symptoms of central nervous system disease, including loss of consciousness, sensory deficits, convulsions, confusion, memory loss, dysphagia, paralysis, or history of head trauma.

e. Drugs. A number of agents, such as anticonvulsants, alcohol, salicylates, sedatives, tranquilizers, and certain antibiotics, can induce vestibular toxicity.

4. Physical examination

a. Complete head and neck examination, including the nose, paranasal sinuses, pharynx, oral structures, and larynx. The neck, orbits, mastoids, and temporal squamae are similarly auscultated for bruits. The postauricular area, auricle, external auditory canal, and tympanic membrane are carefully inspected. Cerumen is removed to adequately examine the external canal and tympanic membrane. Discharging ears should be appropriately cultured.

Pneumatic otoscopy aids in identifying perforations, granulomas, and middle ear pathology. If there is evidence of a perforation, previous surgery, head trauma, or barotrauma, a fistula test is performed. This test is best performed by insufflation with a rubber bulb fitted with an olive tip; however, a pneumatic otoscope (Siegle) will suffice, provided a good seal in the canal is achieved. When positive pressure is applied, the eyes are carefully examined for any nystagmic movement. If a fistula is present, the eyes will beat to the opposite side, and the patient will report intense vertigo similar to his or her complaint.

The use of +20-diopter Frenzel lenses enhances the examiner’s perception of the nystagmus. Weber and Rinne tuning fork tests are performed with a 512-cps tuning fork. A clinical estimate of the patient's speech reception threshold is obtained.
b. Neurologic examination

(1) Cranial nerves

(a) First cranial nerve. Olfactory nerve. Smell - test patient with scents of coffee, tobacco, cloves, and mint. Standardized tests of olfaction are also available.

(b) Second cranial nerve. Optic nerve.

Assess degree of spontaneous nystagmus.
Vision.
Pupillary responses.
Light reflex, consensual reflex, accommodation.
Fundoscopic examination.

(c) Third, fourth, and sixth cranial nerves. Oculomotor, trochlear, and abducent nerves. Eye motion and conjugate motion.

(d) Fifth cranial nerve. Trigeminal nerve.

Open mouth against resistance.
Corneal reflex.
Facial sensation.

(e) Seventh cranial nerve. Facial nerve.

Facial motion.
Taste.
Hitselburger's sign. Sensation of posterosuperior EACs (external auditory canals).

(f) Eighth cranial nerve. Vestibulocochlear nerve. Tuning forks; masked speech testing.

(g) Ninth cranial nerve. Glossopharyngeal nerve. Palatal and pharyngeal reflex; position of soft palate.

(h) Tenth cranial nerve. Vagus nerve.

Vocal cord examination.
Motion of soft palate.

(i) Eleventh cranial nerve. Accessory nerve. Forced motion of head; shoulder shrug.

(j) Twelfth cranial nerve. Hypoglossal nerve. Motion and strength of tongue.
(2) Cerebellum function tests

(a) Finger-to-nose test.

(b) Adiadochokinesia (ability to perform rapid alternating movements).

(c) Romberg test.

(d) Tandem studies.

(e) Deep tendon reflexes.

c. Nystagmus refers to eye movements that are sustained, involuntary, and rhythmic with a speed that is different in the two directions of motion (fast and slow components). Traditionally, nystagmus is described by the direction of the fast component. As described by this method, nystagmus can be horizontal (right to left beating), vertical (upward or downward beating), or rotary (clockwise or counterclockwise beating). Spontaneous nystagmus indicates a vestibular system disorder (either central or peripheral), with the exception of congenital-familial nystagmus and the nystagmus observed with blindness.

(1) Examination for spontaneous nystagmus. The patient is seated or supine with the head immobile. The patient's eyes must not move except to follow the examiner's finger. The finger should not be closer than 18 in from the patient and is moved from left to right, superiorly to inferiorly, and vice versa in sweeps no greater than 30 degrees from the neutral gaze. End-point nystagmus at the extremes of lateral gaze is not uncommon and not considered pathologic. Again, the use of a Frenzel lens enhances the examiner's ability to detect nystagmus by magnification and by interfering with the patient's ability to "fixate" and suppress the nystagmus (a known characteristic of peripheral lesion-induced nystagmus).

(2) Examination for positional nystagmus. Patients often complain that their vertigo is related to head position or changes in position. Positional vertigo can occur in disorders of the labyrinth or of the central nervous system. The Nylen-Bárány test is performed by moving the patient from the sitting position to a lying position with the head extended 45 degrees backward. The test is performed with the head extended, turned to the right, then to the left. Symptoms of nystagmus and vertigo are noted. A Frenzel lens should be used. The latency, duration, direction, and fatigability of the nystagmus are recorded.

d. Diagnostic tests. In addition to the clinical tests mentioned above, patients with vestibular system disorders may require one or more of the following tests to establish the diagnosis.

(1) Audiometry, including site of lesion testing.
(2) Electronystagmography.
(3) Auditory brainstem response (ABR).
(4) CT scan, MRI scan.
(5) Spinal tap, especially if tumor or subarachnoid bleeding is suspected as a cause for vertigo.
(6) Electroencephalography (EEG).
(7) More sophisticated vestibular testing may be indicated in patients with persistent symptoms of dysequilibrium or in whom a diagnosis has not been made. These include harmonic acceleration testing, head autorotational testing, dynamic posturography, and tracking testing.

5. Specific disorders of the vestibular system. The disorders characterized below are not meant to be a complete treatment of vestibular pathology, but only to represent some of the more common or classic disorders. Other disorders of the inner ear described previously can also present with vertigo as the primary symptom. For a more thorough discussion of vestibular disorders, the reader is referred to the bibliography at the end of this section.

a. Ménière’s disease (endolymphatic hydrops) is the classic and probably most common disorder of the peripheral vestibular system.

(1) Signs and symptoms. The classic triad of Ménière's disease is vertigo, tinnitus, and sensorineural hearing loss. The vertiginous episodes are usually sudden in onset and severe with nausea and vomiting. They last minutes to hours with lingering symptoms of unsteadiness for days. These episodes are usually preceded by a fullness in the ear or increasing tinnitus. Hearing loss usually fluctuates, becoming worse near the time of the attacks and then initially returning to or near to normal. The loss is of the sensorineural variety, greater for low frequencies early, later changing to a flat configuration. The hearing loss tends to be progressive and can lead to profound hearing loss, but rarely to total deafness. Discrimination decreases and recovers with the pure tones. Recruitment is also present. Nystagmus is evident, usually away from the involved ear during the attack. Caloric response may be normal early but decreases as the disease progresses. The frequency of attacks is variable. Ten to thirty percent of people with Ménière's are affected bilaterally.

Three isolated variants exist:

(a) Cochlear Ménière's syndrome has two components - fluctuating sensorineural hearing loss and tinnitus.

(b) Vestibular Ménière's syndrome has a sense of aural pressure or fullness and vertigo.

(c) Tumarkin's crisis (drop attacks) has a sudden violent episode of vertigo, during which the patient falls to the ground. The attack is characteristically brief, and the patient is always alert.

(2) Pathologic studies show endolymphatic hydrops with few changes in the sensory and neural structures. It is postulated that attacks are due to rupture of the membranous labyrinth, with mixing of endolymph and perilymph.

(3) Management. No completely effective medical treatment exists that will halt the attacks and prevent progressive hearing loss. Since the attacks vary widely as to severity and frequency, the management should be individualized. A patient with mild, infrequent episodes can reasonably be managed with reassurance. Conversely, those with frequent severe attacks must be considered for a more aggressive medical or surgical approach. Management of acute
attacks consists of treatment with antivertiginous medication (Dramamine, meclizine, diazepam, scopolamine) and correction of any fluid and electrolyte imbalance that might occur from vomiting. Bed rest, sedation, and hydration are the mainstays of severe attacks, and hospitalization is sometimes necessary. Long-term medical management to reduce the number of attacks or to halt the progression of the hearing loss (or both) is more controversial. However, smoking must be stopped and caffeine consumption decreased. Salt intake should be decreased as well. Although several surgical procedures are promising in this respect, evaluation by a competent otologic surgeon is necessary to choose the procedure that will best benefit the patient.

b. Vestibular neuronitis

(1) Signs and symptoms. Vestibular neuronitis is a disease characterized by severe vertigo, sudden in onset, with no associated hearing loss. The attack is usually protracted and may last from a few days to several weeks. Relapses in the first 6 months are not unusual. A viral cause is suspected.

Nystagmus is usually evident and is horizontal. Other neuropathies are not present. The otologic examination is normal.

(2) Diagnostic tests. The audiogram is normal. Calorics and ENG show a depressed vestibular response in the affected ear.

(3) Management of the acute attack is similar to Ménière's disease (antivertiginous drugs, hydration). The disease is usually self-limiting, but can become chronic and disabling. In this rare instance, vestibular nerve section can be beneficial.

c. Benign paroxysmal vertigo (positionai). Patients with numerous disease processes may be subject to postural vertigo (cerebellar-pontine angle tumor, multiple sclerosis, vascular insufficiency). Some peripheral diseases such as Ménière's disease may be exacerbated by changes in position. Benign positional vertigo may be brought on by head trauma, ear infection, or may occur spontaneously.

(1) Signs and symptoms consist of vertigo with the head in a certain position. Other head positions do not precipitate the attack. No hearing loss or other neurologic symptoms are noted. The nystagmus created is usually rotary and is produced by placing the head in a hanging position with the affected ear down. There is a latent period of 2-20 seconds prior to the onset of nystagmus, and the nystagmus is of short duration (usually less than 1 minute). Repeated positioning of the patient produces decreasing nystagmus and symptoms (fatigue).

(2) Management. The disease is usually self-limiting, lasting 6 months or less. Avoidance of the precipitating head position usually suffices. Vestibular rehabilitation exercises can be helpful. Disease lasting longer than 6 months, particularly in the younger patient, should be reevaluated for central causes. Operative section of the nerve to the posterior semicircular canal (singular nerve) has been advocated for persistent cases, but this therapy is controversial.
d. **Motion sickness.** Motion sickness is produced by a mismatch of vestibular, visual, and somatosensory inputs, such as induced in a ship or automobile. The vertigo is variable, depending on the intensity of the mismatched stimuli and the patient's response. It is diminished by providing improved visual input of the surroundings and often responds to symptomatic medical treatment such as with transdermal scopolamine, Dramamine, or meclizine. Prophylactic usage of medication often eliminates or greatly diminishes the dizziness in patients prone to motion intolerance.

e. **Perilymph fistula.** Even mild trauma such as sneezing, coughing, or vigorous nose blowing can produce a leak of perilymph in the areas of the oval or round windows. More commonly, a history of implosive trauma, such as diving or plane flight, or vigorous exertion is elicited.

   (1) **Signs and symptoms** are episodic vertigo, especially with motion or straining, aural fullness, tinnitus, and mild or fluctuating hearing loss.

   (2) **Diagnosis** is history-dependent, but confirmed by a "fistula test" in which the patient strains while holding the nose, inducing increased middle ear pressure. This reproduces the symptoms and can be documented subjectively or objectively by simultaneous ENG. Often, caloric weakness is also found.

   (3) **Treatment** may require exploratory tympanotomy to find and seal the fistula; however, most perilymphatic fistulas heal spontaneously. Disabling vertigo, progressive hearing loss, or persistent symptoms necessitate surgical intervention.

6. **Central vestibular disorders**

a. **Acoustic neuroma.** Better named vestibular schwannoma, the acoustic neuroma is a tumor of Schwann cell origin, usually occurring on the vestibular nerve. It comprises 8% of all brain tumors and about 80% of all cerebellar-pontine angle (CPA) tumors. Although the tumor is benign, it can be very destructive due to its location. The tumor occurs with about equal frequency on both the superior and inferior divisions of the vestibular nerve. It can occur anywhere along the length of the nerve, and thus symptoms can be those of a lesion in the cochlea, in the internal auditory canal, or in the cerebellopontine angle.

   (1) **Signs and symptoms** are usually unilateral sensorineural hearing loss, unilateral tinnitus, or unexplained vertigo. Progression of symptoms is usually slow, but hemorrhage into the tumor may produce a sudden exacerbation. Fifty percent of patients notice hearing loss as a first symptom, with it being sudden in onset 5-10% of the time. Tinnitus can be the only symptom, or it can be associated with the hearing loss. Vertigo rarely is the initial complaint but is frequently present at the time of diagnosis. The description of the vertigo is usually a vague sensation of motion. In the past, trigeminal symptoms of facial pain or numbness were seen in up to three-fourths of people with neuromas, but now with earlier diagnosis, this figure is decreasing. Facial weakness is uncommon, with less than 10% of patients showing this symptom.
(2) Diagnostic tests

(a) ENG (electronystagmogram). Approximately 70-80% of patients show spontaneous nystagmus. Calorics are decreased in 50% of patients with small tumors and 90% of those with large tumors.

(b) Audiology. Approximately 96% of patients have a hearing loss. It is a high tone loss in 64%, a flat loss in 20%, a low tone loss in 8%, and a trough-shaped loss in 8%. There is a marked decrease in discrimination greater than expected for the given hearing loss. Testing to differentiate cochlear from retrocochlear lesions is only suggestive, since 20% of acoustic neuromas affect the cochlea. Evidence of retrocochlear alterations is noted by absent stapedial reflexes and presence of tone decay (adaptation). Cochlear signs of involvement showing recruitment may also be positive.

(c) Auditory brainstem response (ABR) is one of the most accurate noninvasive studies. A delay in wave V (the presumed inferior colliculus wave) on one side of greater than 0.4 msec over the opposite side is highly suggestive of a lesion impinging on the eighth cranial nerve.

(d) X rays can show widening and shortening of the internal auditory canal, seen in 55% of acoustic neuromas on plain x rays.

(e) CT scan. With improved techniques, CT scans are one of the more accurate noninvasive tests available.

(f) MRI scan with gadolinium is the most accurate test for detecting acoustic neuromas. Three- to four-mm tumors can consistently be imaged.

(g) Cerebrospinal fluid. Lumbar puncture may show an increase in CSF pressure and usually shows a CSF protein greater than 50 dL.

(h) Posterior fossa myelogram. Although this test can detect 95% of all acoustic tumors with virtually no false-negatives and only 5% false-positives, MRI with gadolinium enhancement is the imaging study of choice.

(3) Management. Surgery is the major form of therapy, with the approach dependent on the size and location of the tumor. Proton beam and "gamma knife" radiation have been recently added to the treatment regimen. Long-term results with these modalities, as well as a lack of known incidence of complications, make their treatment role unclear at present.

b. Multiple sclerosis is a demyelinating disease that primarily affects white matter in the CNS in young patients (less than 40 years of age).

(1) Signs and symptoms. Charcot's triad of nystagmus, scanning speech, and intention tremor make the diagnosis easy when all are present; however, this is not the case early in the disease. Vertigo is present in about 30% of patients with multiple sclerosis. Nystagmus is present in 40-70% and can be of any type, but dissociated nystagmus is pathognomonic of the disease. Hearing loss, when present, is usually unilateral, high frequency, and sudden with
recovery after a variable time. The diagnosis must often wait for a more typical syndrome to appear, although ENG and evoked response audiometry may help to localize the lesions.

(2) Management. There is no uniformly beneficial treatment. Currently, several experimental regimens are being investigated. The patient should be under the care of a neurologist.

c. Vascular disorders. Dysequilibrium can be induced by vascular insufficiency and is usually seen in elderly patients.

(1) Signs and symptoms depend on the anatomic area affected by the diminished blood flow.

(a) Vertebrobasilar insufficiency symptoms include vertigo, hemiparesis, visual disturbances, dysarthria, facial numbness, ataxia, and headache. These result from transient episodes of ischemia. Decrease in hearing is rare because the posterior inferior cerebellar artery is involved; however, if the anterior inferior cerebellar artery is occluded, decreased perfusion of the inner ear will also occur.

(b) Wallenberg syndrome (lateral medullary syndrome) produces symptoms due to infarction of the lateral portion of the medulla. These include vertigo, nausea, vomiting, nystagmus, ataxia, diminished sense of pain and temperature on the ipsilateral face and contralateral body, dysphagia with ipsilateral palatal and vocal cord paralysis and Horner's syndrome.

(c) Occlusion of the internal auditory artery or anterior vestibular artery can produce sudden vertigo with nausea and vomiting, often without deafness.

(d) Subclavian steal syndrome symptoms are vertigo, occipital headache, diplopia, blurring of vision, and pain in the upper extremity. A difference of 20 mmHg in the systolic blood pressure between the two arms with a diminished radial pulse is diagnostic.

(e) Cervical vertigo is due to cervical spondylosis and intervertebral disc degeneration. Symptoms are produced due to lack of proprioception of the neck musculature, as well as vascular compression or spasm of the vertebrobasilar system. Symptoms include vertigo, syncope, headache, visual disturbances, tinnitus and, occasionally, hearing loss.

(f) Migraine due to constriction and subsequent dilatation of the vertebrobasilar vessels produces vertigo, dysarthria, ataxia, visual disturbances, and intense unilateral pulsatile headache.

(g) Cardiovascular abnormalities can cause dizziness or lightheadedness. Presyncopal symptoms also include nausea, pallor, diaphoresis, and blurred vision. True vertigo is not common. Some causes are cardiac arrhythmias, cardiac sinus arrest, aortic stenosis, and cardiac sinus syncope. Vascular hemodynamic disorders include such entities as orthostatic hypotension, vasovagal episodes, hypoglycemia, and atherosclerosis.
(2) **Diagnosis** is most often based on a thorough history and physical examination, often with neurological consultation. Angiography may be needed.

(3) It is critical to identify patients with **vascular etiology of dizziness** before the serious sequelae of brainstem stroke ensues.

(4) **Treatment** is both medical and surgical, requiring appropriate neurological and vascular surgical consultation.

d. **Epilepsy.** Temporal lobe epilepsy can produce variable degrees of dysequilibrium.

(1) **Signs and symptoms** can be true vertigo, mimicking Ménière's disease, or mild dysequilibrium. Often "absences" with repetitive movements such as chewing, facial grimacing, or lip smacking are seen, as well as auditory hallucinations.

(2) **Treatment** is directed to the seizures disorder, using such medications as phenytoin or carbamazepine.

e. **Hyperventilation.** Lightheadedness is often associated with episodes of hyperventilation due to anxiety or emotional upset.

(1) **Signs and symptoms** are lightheadedness, circumoral or digital paresthesias, diaphoresis, trembling, acute anxiety, and palpitations.

(2) **Diagnosis** is made by having the patient recreate the attack by hyperventilation for 3 minutes or more.

(3) **Treatment** is by reassurance or a method of rebreathing (into a bag), which prevents hypocapnia and alkalosis due to inhalation of expired carbon dioxide.

7. **Facial nerve paralysis.** Although not technically an otologic disorder, the course of the facial nerve through the temporal bone and its frequent association with diseases and trauma to the ear make it appropriate to discuss. Disorders of the facial nerve outside the temporal bone are discussed in Chapter 6. Attention should also be directed to the description of the anatomic course of the nerve described in this chapter.

a. **Topognostic testing.** Location of the point of damage to the nerve by various techniques has historic value, but little prognostic value. Most lesions of the facial nerve in Bell's palsy are at the geniculate ganglion, where the facial canal is the narrowest. Loss of function of the stapedius reflex, salivary flow, lacrimation, and taste probably have more to do with differential response of various size nerve fibers carrying these responses, rather than location of the lesion.

b. **Electroneurography (ENG)** is presently the best clinical test for monitoring facial nerve function. It involves stimulation of the facial nerve near the stylomastoid foramen with recording responses on the face. Comparison of the electrical response with the other side gives an estimate of the degree of degeneration of the facial nerve. Degeneration of greater than 95% leads to poor expected recovery. Less than 90% degeneration usually bodes well
for a good recovery. Test results are a reflection of injury 2-3 days prior to testing, because it takes that long for the neural elements to degenerate.

c. Electromyography. This procedure is used to test the musculoelectrical activity. An electrode must be placed within the substance of the muscle and its potential recorded. If total interruption of the nerve has occurred, no motor unit activity will be seen approximately 2-3 weeks after the injury. If degeneration occurs but the lesion is not complete, fibrillation potentials will be recorded.

d. Etiology. It is important to assess not only the site of injury to the facial nerve, but also the etiology so that appropriate therapy can be instituted. Only after all possible causes have been ruled out can a diagnosis of idiopathic facial paralysis can be considered (Bell's palsy).

e. Trauma

(1) Trauma at birth. The extratemporal portion of the facial nerve may be injured during delivery, especially when the forceps are inappropriately applied. Unless a specific laceration occurs, facial paralysis is transient. Facial paralysis may also occur as a congenital neurologic deficit, but is most frequently associated with multiple neuropathies.

(2) Intratemporal. Injury to the facial nerve within the temporal bone most often is a result of surgical trauma. This occurs most frequently in the vertical segment of the facial nerve during mastoid surgery, although dehiscence of the bony covering of the facial nerve in the horizontal segment may allow for injury to occur during middle ear procedures, such as stapedectomy. Surgical removal of an acoustic neuroma may also result in transient or permanent facial paralysis.

Temporal bone fractures may result in loss of facial nerve function. Facial nerve paralysis may occur with both transverse and longitudinal fractures. Although up to 90% of all temporal bone fractures are longitudinal, approximately 15% of these fractures have concomitant facial nerve injury. Up to one-half of transverse temporal bone fractures may occur with a facial nerve paralysis that may be immediate, due to transection or bony impingement on the nerve. These fractures often require immediate surgical intervention for repair. Delayed paralysis is most often due to posttraumatic edema. The question of immediate or delayed surgery must be based on the individual patient, as well as the evidence of progressive degeneration.

(3) Extratemporal. Injury to the facial nerve distal to the stylomastoid foramen most often occurs during parotid surgery. Prevention entails identification of the nerve at the outset of the procedure. Extratemporal injury may also occur following trauma to the face. Attempts at reanastomosis or nerve grafting should be done as soon as the appropriate diagnosis is made in order to allow the maximum return of function. (See Chap 6, III.A.).

f. Infection

(1) Acute infection (bacterial). Otitis media, whether acute or chronic, is the most common cause of facial nerve paralysis from bacterial origin, especially in the young child.
Signs and symptoms consist of an acute otitis media, often with a bulging drum and a relatively rapid onset of facial nerve paralysis. Chronic disease often implicates a cholesteatoma with either an infectious neuritis or compression from an enlarging cholesteatoma.

Management. Myringotomy to decompress the middle ear and mastoid and appropriate antibiotics usually result in recovery of the nerve in acute otitis media. Cholesteatomas with facial paralysis require emergent surgery.

Acute necrotizing (malignant) otitis externa is an infectious process originating within the external auditory canal that spreads to the contiguous cartilage and bone. The most common organism is *Pseudomonas aeruginosa*. The facial nerve may be involved either within the soft tissue of the parotid at the stylomastoid foramen or within the fallopian canal (canal of the facial nerve) (see VI.F.).

Management consists of the topical and IV antibiotics with judicious debridement.

(2) Viral. Herpes zoster oticus (Ramsay Hunt syndrome) is a viral disorder involving the facial nerve and associated with hearing loss, vertigo, and herpetic rash around the pinna. Although it is known that this is a viral disorder, assessment of surgical intervention and appropriate therapy is often similar to that of Bell's palsy.

Management. Early treatment with acyclovir and prednisone offers the best chance for recovery of facial function. Surgical decompression of the facial nerve is of limited value.

Guillain-Barré syndrome may represent a central nervous system viral disorder, often accompanied by facial paralysis. The therapy in these situations is supportive.

(3) Chronic infection most often entails a chronic otitis media with an associated cholesteatoma. Facial nerve paralysis is not usually sudden unless an acute process intervenes. Therapy should be directed at removal of the infectious process with careful surgical identification of the facial nerve. The presence of facial paralysis with a purulent otorrhea should be viewed as a significant problem and as an indication for immediate hospitalization.

(4) Idiopathic (Bell's palsy). The term Bell's palsy should be used only for those situations when the etiology of the facial paralysis cannot otherwise be ascertained. It is perhaps the most common of all the groupings of facial paralysis. Current theories as to the etiology of this disorder have not been proved or confirmed; however, Bell's palsy may represent an isolated viral neuritis or a single manifestation of a polyneuritis. Vascular ischemia may also play a role, causing the facial nerve to swell within its bony canal.

Diagnosis. The diagnosis is made by exclusion of all other etiologies of facial paralysis. Enhanced MRI scanning may show increased activity around the geniculate ganglion region.

Management remains controversial; however, if corticosteroids are used, they should be limited to those in whom the facial paralysis is diagnosed early (within 1-7 days). Facial nerve decompression is probably not beneficial.
(5) Miscellaneous causes

**Neoplastic.** Any neoplasm of the temporal bone can involve the facial nerve secondarily; however, a neuroma of the facial nerve must be a prime consideration. Other possible lesions are glomus tumors, squamous cell carcinomas primarily arising from the external auditory canal, congenital cholesteatoma, meningioma, and acoustic neuroma. Eosinophilic granuloma must be considered in children with multiple punched-out lesions of the squamous portion of the temporal bone. Biopsy of the lesion is diagnostic.

**Metabolic.** Facial paralysis is occasionally associated with sarcoidosis (Heerfordt's syndrome), Wegener's granulomatosis, periarteritis nodosa, and diabetes. **Therapy** is directed at the underlying disorder. Facial nerve decompression is rarely indicated in such a situation.