Introduction

Otolaryngologists are often requested to present lectures to medical students, other physicians and civic organizations. This Slide Lecture on Hearing Loss and Rehabilitation is the first in a series of topics designed to fill such requests. In addition, the Slide Lecture can be used as a self-study guide for medical students and new otolaryngology residents.

The selected slides represent many of the important clinical aspects of otology. It is hoped that they can be incorporated into the otolaryngologist's own collection and serve as an outline around which a lecture can be more easily prepared. The accompanying text is a version of the lecture presented by the author to medical students. Its emphasis and terminology must, of course, be modified according to the type of audience intended.
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Section I - Anatomy and Physiology of the Ear

A. External and Middle Ear

Slide 1 - Diagram of the external, middle, and inner ear

The ear is a compound organ contained within the temporal bone and responsible for the senses of hearing and balance. It can be divided into external, middle, and inner parts.

The external ear consists of the auricle and ear canal, which serve to collect the sound and actually amplify certain frequencies.

The middle ear is a small, air-containing space that includes the tympanic membrane, three ossicles - malleus, incus, and stapes - the eustachian tube, and a portion of the facial nerve. The middle ear system significantly enhance the transmission of sound to the inner ear by matching the impedance between air and water.

Slides 2 & 3 - Tympanic membrane

The tympanic membrane is a conical structure about one centimeter in diameter and one-tenth millimeter thick. It is divided into two parts, the larger pars tensa and the smaller pars flaccida located superiorly. The pars tensa is comprised of three layers: an outer squamous epithelium, continuous with the skin of the ear canal; a middle fibrous layer, consisting of both radial and circular connective tissue fibers; and an inner mucosal layer, which is part of the mucous membrane of the middle ear space.

The pars flaccida lacks the central fibrous layer and is not as strong or stiff as the remainder of the tympanic membrane. This anatomic fact is important in the pathogenesis of acquired cholesteatoma.

Slides 4 & 5 - Sound reflection at an air-water interface

Middle ear transformer mechanism

As sound travels from air to water, 99.9% is reflected because of the large difference in the mobility of the two media. This potential for loss of sound energy occurs in the process of hearing, since sound does travel from the air into the cochlear fluids. The middle ear system, however, enhances the transfer of sound into the cochlea, conserving most of the sound energy.

The most important factor in this transformer mechanism is the **hydraulic ratio**. This is the ratio of the area of the tympanic membrane, where the sound is collected, to the area of the oval window, where the sound is concentrated and enters the cochlea. This ratio is 17:1. Of lesser importance is the **lever effect**, which is the ratio of the length of the malleus handle to that of the long process of the incus - a value of 1.3. Together these mechanisms increase the transmission of sound into the cochlea 22-fold, minimizing the potential for energy loss. Of course, a hole in the tympanic membrane or damage to the ossicles will negate this transformer mechanism and result in a significant hearing deficit.
B. Inner Ear

Slide 6 - Diagram of the inner ear

The inner ear is a fluid-filled chamber containing the sensory structures for hearing and balance. The cochlea, a spiral-shaped tube approximately 3 cm in length, is the hearing portion of the inner ear. Three semicircular canals and two recesses, the utricle and saccule, contain the vestibular organs concerned with balance. The base or foot-plate of the stapes sits in an oval window or opening at one end of the cochlea and is in contact with the cochlear fluids.

Slide 7 - Schematic diagram of uncoiled cochlea

In this schematic diagram the cochlea has been uncoiled, and is represented as a long tube. Vibrations of the stapes produce fluid waves within this tube, stimulating the sensory elements (hair cells) arranged on the basilar membrane. At the other end of the cochlea is another opening, the round window, which is covered by a thin membrane that moves in and out in response to movement of the stapes.

Slide 8 - Histologic section of the inner ear

This slide shows a schematic cross section through the cochlea. The sensory cells of hearing are termed hair cells and are contained within the organ of Corti. They number approximately 16,000 and are arranged in four rows along the length of the basilar membrane. The hair cells transduce mechanical energy - vibrations of the basilar membrane caused by fluid movement - into electrical energy - nerve impulses within the VIII cranial nerve.

High-frequency sounds cause vibrations primarily at the base or beginning of the cochlea, and hair cells in that area are maximally stimulated. Responses to low-frequency sounds, on the other hand, are carried primarily by the nerve fibers innervating the hair cells at the apex of the coiled cochlea.

Section II - Evaluation of Hearing Loss

A. Physical Examination

1. Otoscopy

Slide 9 - Photograph of examination instruments

Examination of the ear requires a pneumatic otoscope, which provides illumination and magnification, and allows insufflation of air into the ear canal to evaluate the mobility of the tympanic membrane. As the ear is pulled upward and backward to straighten the cartilaginous portion of the ear canal, the largest possible speculum is inserted, facilitating visualization and any necessary cleaning of the ear.

The entire tympanic membrane should be carefully inspected, with particular attention given to its superior and posterior-superior aspects, where cholesteatomas are often first
manifest. Assessing mobility of the tympanic membrane is also critical, particularly when examining for fluid within the middle ear.

2. Tuning Fork Tests

**Slide 10 - Weber and Rinne tuning fork tests**

Tuning forks can be used to qualitatively assess hearing level and determine if the loss is primarily conductive or sensorineural. The two principal tuning fork tests used are the Weber and Rinne tests.

The Weber test is performed by placing the stem of a vibrating tuning fork on the middle of the skull such as the forehead or central incisor. If a unilateral conductive hearing loss is present, the sound will be louder in that ear. If a sensorineural hearing loss is present, the sound will be referred to the better hearing ear. A person with normal hearing will not localize the sound to either ear.

The Rinne test compares air- and bone-conducted sounds. A vibrating tuning fork is alternately placed on the mastoid and held beside the ear. The loudness or duration of the two sounds is then compared. In a normal hearing ear or in one with a sensorineural hearing loss, the air-conducted sound will be louder. In an ear with a conductive hearing loss, the bone-conducted sound dominates.

B. Audiological Tests

1. Basic Audiometry

**Slide 11 - Audiometer**

An audiologist assesses the hearing of a patient seated behind the glass window in a soundproof room.

An audiometer is used to measure hearing thresholds for pure tones and for speech. The various tones are presented both by air conduction, using headphones, and by bone conduction, using a bone vibrator resting on the skull. Sound presented by bone conduction vibrates the skull, and this vibration is transmitted directly into the cochlea, largely bypassing the middle ear system. The results, therefore, directly reflect the state of the inner ear - the sensory and neural parts of hearing. Sound presented through earphones reaches the inner ear through the normal pathway - the external ear canal, tympanic membrane, and ossicles. Any abnormality of the external or middle ear will therefore alter these results. It is thus possible to accurately differentiate a conductive hearing loss caused by various middle ear pathologic conditions from a sensorineural hearing loss caused by damage to the cochlea or VIII cranial nerve.

Although the testing is best performed in an alert patient who can signal his responses, valuable information can also be obtained from very young children using behavioural and conditioned responses.
Slide 12 - Audiogram showing a unilateral conductive loss

This is the audiogram of a patient with normal hearing in the left ear and a conductive hearing loss in the right ear. Across the top are the various tones tested, ranging from low to high frequencies. Down the side is the scale for intensity or loudness.

The left ear has normal hearing as indicated by the blue line across the top: both air- and bone-conducted sounds can be heard at a very soft level. The right ear has excellent hearing for bone-conducted sounds, meaning that the inner ear is functioning normally. For air-conducted sounds, which pass through the middle ear, the tones must be presented at a much louder level before they are heard. The difference is then a quantitative measure of the conductive hearing loss present.

2. Brain Stem Response Audiometry

Slide 13 - Auditory brain stem response (ABR) unit

An infant's hearing threshold is being tested with a portable ABR unit as part of a screening program for neonates at high risk for hearing loss.

Certain limitations are imposed on conventional audiometry because it requires active participation by the person being tested. Hearing thresholds in neonates or persons with multiple handicaps, for example, are particularly difficult to obtain.

In the 1970's ABR testing became available on a widespread clinical basis, and it has subsequently had a major impact on hearing evaluation. Briefly, several electrodes are positioned on the head, and clicks or tones are presented to an ear. Using computed averaging techniques, the minute VIII nerve and brain stem electrical discharges produced by the tones are extracted from all the background activity. In the normal ear, four or five distinct wave forms occurring in the first ten milliseconds after the stimulus can be recorded. No verbal responses are required from the patient; in fact, the individual can be asleep.

Two areas in which ABR testing has been particularly helpful are the early identification of hearing loss in young children and the diagnosis of tumors involving the VIII cranial nerve.

3. Impedance Audiometry

Slide 14 - Electroacoustic impedance meter

A portable electroacoustic impedance meter (tympanometer) is pictured.

Slide 15 - Schematic diagram of the impedance probe (probe tone generator, manometer, and microphone)

Another special diagnostic test that is completely objective is tympanometry. This test provides information on the compliance or mobility of the tympanic membrane, which is determined primarily by the state of the middle ear. For example, fluid in the middle ear
space will cause a stiff system or low compliance, whereas a dislocated ossicle will cause a hypermobile system or high compliance.

The procedure is performed by inserting a probe into the external ear canal, obtaining an airtight seal. The air pressure within the canal is then varied as a tone is presented. The acoustic energy from the tone will either be admitted to the middle ear or reflected back into the external canal, depending on the compliance of the tympanic membrane / middle ear system. This reflected energy is measured by a microphone within the probe.

The tympanic membrane will be most compliant when the air pressure is equal on both its sides. By varying the pressure in the external canal with the probe and measuring tympanic membrane compliance, an indirect measurement of middle ear pressure is readily obtained.

Section III - Pathology of the Ear

A. Diseases of the External Ear

1. Otitis Externa

**Slide 16 - Pseudomonas otitis externa**

Mid swelling and erythema of the external canal are present.

**Slide 17 - Pseudomonas otitis externa**

A more advanced stage of otitis externa is pictured, with cellulitis of the external canal and marked edema narrowing the lumen of the canal.

**Slide 18 - Otomycosis**

White mycelia and purulent discharge are present within the external canal.

Discussion

**Etiology.** Factors contributing to the development of otitis externa include moisture (humidity, swimming) and local trauma. The most common causative organisms are Gram-negative bacilli (especially *Pseudomonas aeruginosa*) and fungi (especially *Aspergillus*).

**Symptoms.** Pain and itching are the predominant symptoms. The patient may also have drainage, a feeling of fullness, and hearing loss.

**Signs.** Erythema of the skin of the ear canal (and tympanic membrane) and edema of the canal wall are usual. There may also be purulent discharge and, if a fungal infection is present, the presence of mycelia and spores.

**Treatment.** Careful and frequent cleaning of the ear canal, dry ear precautions, and local otic drops are the main-stays of therapy. Drops with antibiotics (ie, polymyxin and/or neosporin) or acidifying agents (ie, acetic acid or boric acid) are both effective. Analgesics
are frequently required. Systemic antibiotics are occasionally required for a periauricular cellulitis.

2. Malignant Otitis Externa

**Slide 19 - Malignant otitis externa**

Marked swelling of the external canal wall and granulation tissue are present.

**Discussion**

_Etiology._ Malignant otitis externa is a locally aggressive, necrotizing infection in a diabetic or immune-compromised patient. *P. aeruginosa* is the responsible organism, and it causes an osteitis and osteomyelitis of the skull base.

_Symptoms._ The usual symptoms of otitis externa, especially the pain, are magnified.

_Signs._ Granulation tissue is frequently present in the floor of the external ear canal at the junction of the bony and cartilaginous canals. Bony sequestra may form. Cranial nerve deficits (most commonly VII, but also IX, X, XI and XII) can occur, and when present portend a poor prognosis (mortality rates up to 60%).

_Treatment._ Regulation of the diabetes or other medical problems is necessary. Parenteral antibiotics, including an aminoglycoside such as tobramycin and a broad-spectrum, semi-synthetic penicillin such as ticarcillin, are required on a prolonged basis, often for four to eight weeks. Surgical debridement of devitalized tissue is also beneficial.

B. Diseases of the Middle Ear

1. Acute Otitis Media

**Slide 20 - Acute otitis media - stage of hyperemia (earliest stage of acute otitis media)**

There is inflammation, especially evident posteriorly, and slight bulging of the tympanic membrane.

**Slide 21 - Acute otitis media - early stage of exudation**

There is marked erythema and bulging of the tympanic membrane.

**Slide 22 - Acute otitis media - late stage of exudation**

The exudate in the middle ear is under considerable pressure. The tympanic membrane is severely inflamed and thickened. The skin of the membrane is beginning to desquamate and has a rough or corrugated appearance. Spontaneous perforation with release of the exudate is likely.
Discussion

Etiology. By age 4 years, seventy to eighty percent of children have had at least one episode of acute otitis media. This often follows an upper respiratory infection and most commonly is caused by Streptococcus pneumoniae or Hemophilus influenzae. In neonates approximately 20% of the infections are caused by Gram-negative enteric bacilli.

Symptoms. Fever, ear pain, and hearing loss are the predominant symptoms. Purulent drainage occurs if the tympanic membrane spontaneously rupture.

Signs. The tympanic membrane is red, thickened, bulging, and has poor mobility on pneumatic otoscopy. If a perforation is present, pulsating drainage from the middle ear space is often noted.

Treatment. Amoxicillin or an antibiotic with a similar spectrum are the drugs of choice. Analgesics are usually necessary. Myringotomy and aspiration of middle ear fluid for culture and sensitivity are often essential if the patient is a neonate or an immune-compromised host in whom unusual organisms may be present, or if any suppurative complications have occurred.

Complications. Complications include a persistent middle ear effusion, tympanic membrane perforation, ossicular damage from fibrosis or erosion, facial nerve paralysis, and central nervous system involvement (ie, meningitis).

2. Otitis Media With Effusion

Slide 23 - Otitis media with effusion - serous fluid

A yellow-tinged serous fluid is evident in the middle ear space. The tympanic membrane is retracted, causing the malleus to be very prominent.

Slide 24 - Otitis media with effusion - mucoid fluid

The tympanic membrane is opaque and has a gray-blue appearance suggestive of a more viscous or mucoid middle ear fluid accumulation. Again the tympanic membrane is retracted and the malleus is prominent.

Slide 25 - Ventilation tube insertion

The steps for insertion of a ventilation tube are illustrated. A small incision in the tympanic membrane is made inferiorly or anteroinferiorly, and the middle ear fluid is aspirated. A ventilation tube is then inserted within this opening. Most ventilation tubes will remain in place 6 to 12 months, then spontaneously extrude into the ear canal. In more chronic conditions, tubes that will remain in the tympanic membrane longer can be used.
Discussion

Etiology. The accumulation and persistence of fluid-serous or mucoid - in the middle ear space is termed otitis media with effusion or serous otitis media. This usually follows an acute otitis media (ie, a postinflammatory exudate). Poor eustachian tube ventilation of the middle ear is contributory. A metaplasia of the middle ear mucosa with an increase in the number of mucous cells and formation of mucous glands occurs. Although the effusion is not purulent, culturable bacteria (especially \textit{H. influenzae}) are present in up to 40% to 50% of cases. Otitis media with effusion is very common in children with a cleft palate (unrepaired or repaired).

Symptoms. Hearing loss is the dominant symptom. Behavioural disturbances may be the first evidence of such a hearing loss. Pain is unusual.

Signs. The tympanic membrane usually loses its translucency and becomes more opaque. An air fluid level or bubbles may be apparent, or the tympanic membrane may simply have an amber or gray-blue appearance. There is little or no mobility to the tympanic membrane with pneumatic otoscopy. A conductive hearing loss, primarily of low frequency, is present, and tympanometry shows stiff compliance.

Treatment. Decongestants and antihistamines in recent double-blind studies have not been proven efficacious, but may be considered when allergy is also present. An initial trial of antibiotics is reasonable since many effusions do contain viable bacteria. Myringotomy and ventilation tube insertion offer definitive treatment. If the middle ear effusion resulted from an acute otitis media at least ten to twelve weeks should elapse from the acute infection to permit spontaneous resolution. (Epidemiologic studies have shown that a middle ear effusion can be expected in 40% of children one month following an acute otitis media; most of these effusions will resolve over the subsequent four to six weeks.)

3. Barotrauma

Slide 26 - Barotrauma

The tympanic membrane is retracted causing the malleus handle to be very prominent; the tympanic membrane appears dark blue because of blood in the middle ear space.

Discussion

Etiology. Barotrauma usually occurs with rapid descent from an area of less atmospheric pressure to one of greater pressure; ie, descent in an aircraft or diving in water. As one ascends in the atmosphere, air pressure within the middle ear becomes greater than that in the external ear canal or nasopharynx; air will therefore passively move down the eustachian tube and into the nasopharynx. On descent, the middle ear pressure becomes negative relative to the ambient pressure. To equalize pressures, the eustachian tube must actively open to allow air in, a process that may be difficult for individuals with eustachian tube dysfunction or with an upper respiratory infection that produced edema around the
eustachian tube orifice. If the negative pressure within the middle ear persists, the tympanic membrane becomes retracted and a transudation of serum and blood occurs.

**Symptoms.** Pain, often severe, and a hearing loss (conductive) are the prominent symptoms. There is also frequently a sensation of fullness or pressure within the ear.

**Signs.** The tympanic membrane will be retracted and poorly mobile. The middle ear may contain serum and/or blood; if hemorrhage has occurred the tympanic membrane will have a dark blue color.

**Treatment.** Barotrauma can usually be prevented if avoidance of air travel during a significant upper respiratory infection is possible. The use of decongestants (systemic and nasal) at least one hour prior to descent, and frequent equalization of middle ear pressure by swallowing, Valsalva maneuvers, etc, are helpful. The effects of barotrauma are usually self-limited, although decongestants may speed recovery. Occasionally a myringotomy is necessary for relief of severe pain.

4. Tympanic Membrane Perforations

**Slide 27 - Traumatic perforation**

A linear tear of the tympanic membrane, which resulted from a slap to the ear by an open hand, is present.

**Slide 28 - Central perforation**

This large hole in the inferior aspect of the tympanic membrane resulted from otitis media. The whitish areas in the membrane represent tympanosclerosis, which is the deposition of calcium within the fibrous portion of the tympanic membrane in response to repeated inflammation.

**Slide 29 - Healed perforation**

The central perforation has spontaneously healed with a thin membrane. Usually there is poor or no regeneration of the middle fibrous layer, causing the healed area to appear atrophic and to have excessive mobility on pneumatic otoscopy.

**Discussion**

**Etiology.** Perforations of the tympanic membrane usually result from infection (suppurative otitis media) or trauma.

**Symptoms.** Hearing loss (conductive) is usually the predominant symptom. The loss will be greatest for large vs. small perforations (due to loss of the hydraulic ratio) and for perforations located posteriorly vs. anteriorly (due to loss of round window protection: sound now reaches the oval window and round window simultaneously, instead of the oval window preferentially, resulting in a cancellation of fluid movement within the cochlea).
Purulent drainage may also occur. The middle ear space is now vulnerable to external contamination and infection.

*Signs.* Trauma to the tympanic membrane usually creates either a linear tear or a small triangular flap of tympanic membrane. If secondary to infection, a round or oval perforation is typical. The tympanic membrane has no movement on pneumatic otoscopy. Depending on the state of the middle ear, the perforation may be dry and the middle ear mucosa seen through it normal, or there may be mucoid or purulent drainage from the middle ear with a hypertrophic, inflamed mucosa.

*Treatment.* Most traumatic perforations are small and heal spontaneously. Antibiotics, local or systemic, are unnecessary unless the ear was contaminated at the time of injury (ie, swimming accident) or is now actively draining. Occasionally the ragged edges of a traumatic membrane tear will be inverted into the middle ear and require eversion and approximation; this requires the use of a microscope.

In small perforations, healing is often promoted with the use of a paper patch. This acts as a flat surface for the ingrowth of epithelium from the edges of the perforation. Larger perforations require surgical closure.

a. Surgical Repair

Various grafting materials for repair of a tympanic membrane perforation have been used. These include skin, perichondrium, vein wall, fascia, and homograft tissue. Temporalis fascia and homograft tympanic membrane are most widely used today.

Many surgical techniques are employed, and the success rate for most is excellent (approximately 90%). The surgery may be performed either directly through the ear canal or by first making an incision behind the ear, reflecting the ear forward, then working through the ear canal. The grafting material is either placed directly on the surface of the tympanic membrane remnant or directly opposed to its underneath surface. In either case it is held in place by absorbable packing until healing has occurred.

The following series of slides show relevant steps from one technique of tympanic membrane grafting. The surgery is being performed on the right ear.

**Slide 30 - A postauricular incision is made.**

**Slide 31 - The temporalis fascia is obtained as the grafting material.**

**Slide 32 - The ear has been reflected forward.**

This view is through the ear canal. A large perforation incorporating most of the tympanic membrane is present.
Slide 33 - The fascia graft is positioned

The graft covers the perforation and is placed beneath the malleus handle for stabilization. The external canal will also be packed with an absorbable material to hold the graft in place until complete healing has occurred in four to six weeks.

Slide 34 - Tympanic membrane perforation (surgical photograph)

Slide 35 - Tympanic membrane perforation (surgical photograph)

Slide 35 - Fascia graft repair of tympanic membrane perforation (surgical photograph)

This photograph shows the temporalis fascia graft in place.

5. Cholesteatoma

Slide 36 - Cholesteatoma formation

This diagram shows the formation of a cholesteatoma from an attic retraction pocket. As the retraction in the pars flaccida deepens, it becomes more difficult for the desquamated epithelium to egress from the small opening of the pocket. The cholesteatoma, therefore, begins to grow, expanding into the middle ear.

Slide 37 - Small attic (pars flaccida) retraction

This is a photograph of a small attic (pars flaccida) retraction that is dry and without evidence of cholesteatoma. However, the potential for infection and cholesteatoma formation exists.

Slide 38 - Attic cholesteatoma

This is a photograph of an attic retraction pocket containing cholesteatoma debris

Slide 39 - Cholesteatoma (schematic diagram)

The full extent of a cholesteatoma is not readily appreciated on otoscopic examination of the ear. Usually the cholesteatoma will have enveloped the heads of the ossicles and extended back toward or into the mastoid air cells as seen in this schematic drawing.

Discussion

Etiology. A cholesteatoma is an accumulation of skin and epithelial debris in the middle ear, epitympanum, or mastoid. They can develop by an ingrowth of skin through a perforation at the margin of the tympanic membrane. More commonly, they result from a retraction pocket in the thin-walled pars flaccida. With poor eustachian tube function, this invagination of the pars flaccida deepens, and desquamated epithelium begins to accumulate.
The dangers of a cholesteatoma result from its destruction of bone. At the advancing edge of the mass, bone resorption occurs - ossicles are commonly destroyed and erosion into the inner ear, facial nerve, or cranial cavity can occur.

Symptoms. Purulent drainage is common. Cholesteatomas often become infected since the trapped keratin debris is a good culture media, especially when it becomes wet. Cultures reveal a mixed flora of aerobic and anaerobic Gram-positive and Gram-negative organisms.

A hearing loss (usually conductive) results from ossicular destruction. Occasionally the loss can be sensorineural from erosion by the cholesteatoma into the cochlea.

Dizziness can result from erosion of the cholesteatoma into the balance canals (the horizontal semicircular canal is the most vulnerable).

Facial paralysis can occur from pressure on the facial nerve after erosion of its bony canal by the cholesteatoma.

Pain is usually absent or minimal with a cholesteatoma.

Signs. The most common finding is a perforation or area of retraction in the attic or posterosuperior aspect of the tympanic membrane. Keratin debris and/or purulent drainage from this region are usually evident. Granulation tissue or a polyp may also be present.

Treatment. Cholesteatomas require surgery. The primary goal is to make the ear safe. Also to be achieved is an ear that remains dry and carefree and that has good hearing.

Many different operative approaches can be used successfully. For most cholesteatomas, surgery includes three main steps:

1) mastoidectomy - this involves widely opening the mastoid air cell system behind the ear; 2) removal of the cholesteatoma from the middle ear and mastoid; 3) reconstruction of the middle ear for the transmission of sound, ie, tympanoplasty and ossicular reconstruction.

   a. Ossicular Reconstruction

   The middle ear ossicles are most commonly damaged by chronic suppurative middle ear infection or by cholesteatoma. The incus, especially its long process which articulates with the stapes, is the most frequent ossicle involved.

   Ossicles can be dislocated by head trauma, and again, the most commonly injured is the incus.

   A damaged ossicular chain can be reconstructed with a variety of materials including bone (either the patient's own or homograft ossicles), cartilage, or manufactured alloplastic materials. Each material has advantages and disadvantages, but all can give good, but rarely perfect, functional results.
Slide 40 - Materials for ossicular reconstruction

Different materials for ossicular reconstruction are pictured, including cartilage, a homograft incus, and alloplastic materials (Plastipore).

Slide 41 - Ossicular reconstruction with Plastipore

This schematic diagram shows the use of a Plastipore strut between the tympanic membrane and stapes in the case of an absent malleus and incus, between the tympanic membrane and oval window in the case where all the ossicles are missing.

Slide 42 - Ossicular reconstruction with a fitted incus

6. Otosclerosis

Discussion

Etiology. Otosclerosis is a hereditary disease affecting the bone around the inner ear. Initially it causes a softening and resorption of bone. As the focus matures, new bone is formed and sclerosis occurs. Most commonly this process affects the stapes footplate, fixing it in place and preventing the transmission of sound into the inner ear.

The disease is inherited as an autosomal dominant trait with incomplete penetrance. Approximately one in 100 people have symptomatic otosclerosis; however, as many as 10% of the population have subclinical (histologic) otosclerosis. The incidence of otosclerosis in women is twice as that in men, and appears to be accelerated by pregnancy.

Otosclerosis generally becomes manifest by the fourth or fifth decade of life. In most cases both ears are involved, although not necessarily simultaneously or to the same extent.

Symptoms. A progressive hearing loss (conductive) in a patient without a prior history of ear disease is the typical presenting symptom. (Sensorineural hearing loss can also occur, but is less common.)

Tinnitus is present in some patients, and this symptom may improve with successful correction of the conductive hearing loss.

Signs. Usually the tympanic membrane appears normal. Rarely, the midportion of the tympanic membrane will have a reddish tinge due to an increase in vascularity of the mucosa over the promontory (Schwartzte sign).

Treatment. Surgical removal of the stapes and replacement with a prosthesis can be expected to improve hearing in approximately 90% of patients. Alternatively, a hearing aid can be satisfactorily used in many cases.

The following series of slides demonstrate one technique for stapedectomy.
Slide 43 - Canal wall incision

The middle ear is entered by elevating forward a flap of the canal wall skin and tympanic membrane.

Slide 44 - Middle ear exposure

Once the tympanic membrane has been elevated, the incus, stapes, and chorda tympani nerve can be seen. Removal of the stapes involves separation of the incudostapedial joint and severance of the stapedius tendon.

Slide 45 - Removal of the stapes

The stapes is fractured inferiorly, away from the facial nerve. The fixed stapes footplate remains, and must be removed by small hooks.

Slide 46 - Placement of the prosthesis

Removing of the stapes footplate opens into the inner ear. To prevent escape of perilymph and to form a new footplate, a soft tissue covering such as fascia or perichondrium is placed over the oval window. A wire prosthesis is then positioned between the incus and soft tissue to reestablish connection between the ossicular chain and the inner ear fluid.

C. Diseases of the Inner Ear

Slide 47 - Common causes for sensorineural hearing loss

1. Noise-Induced Hearing Loss

Slide 48 - An audiogram with a typical pattern of sensorineural hearing loss due to excessive noise exposure.

Loud noise can permanently damage the inner ear. Sensorineural hearing loss can occur from a single noise exposure (ie, gun blast) or it can be cumulative from years of intermittent occupational (ie, industrial plant) or recreational (ie, loud music, home power tools) exposure. Excessive noise causes a loss of hair cells (outer hair cells first), beginning in the basal region of the cochlea. The initial hearing loss is therefore limited to the high frequencies. Tinnitus may also occur, and can be the presenting symptom. With continued noise exposure, progressively lower frequencies are involved as the damage spreads toward the apex of the cochlea. Communication is affected as the speech frequencies are involved.

Noise damage is preventable if ear protectors (ie, ear muffs or ear plugs) are used.

2. Meniere's Disease

Slide 49 - Histologic section of the cochlea showing endolymphatic hydrops.

Note the bulging Reissner's membrane.


**Discussion**

*Etiology.* Meniere's disease is a disorder of the inner ear thought to be caused by altered fluid dynamics within the labyrinth. The disease is characterized by the triad of vertigo, sensorineural hearing loss, and tinnitus. Meniere's disease eventually involves both ears in 30% to 40% of cases. Syphilis of the inner ear can cause symptoms like those of Meniere's disease.

*Symptoms.* Episodic vertigo, typically lasting hours and often associated with nausea and vomiting, is generally the most severe and disabling symptom. In between attacks of vertigo, there may be little or no dizziness.

Hearing loss (sensorineural) is associated with the vertiginous episodes. Initially the hearing loss may fluctuate, returning to the normal range after the vertigo subsides. Eventually a permanent hearing loss of moderate to moderate/severe degree develops.

A roaring tinnitus is common, and often becomes louder with the vertigo.

Frequently, fullness or a pressure sensation within the ear is noted.

*Signs.* The otoscopic examination is normal. The sensorineural hearing loss is usually most severe in the low frequencies. Nystagmus may be present during an acute episode of vertigo.

*Treatment.* Since the underlying etiology of Meniere's disease is unknown, therapy continues to be empiric. Fortunately, most patients can be controlled on a medical regimen of salt restriction, a diuretic, and vestibular sedative. Surgery is directed primarily at controlling the vestibular symptoms. If hearing remains good, a section of the vestibular nerve offers the best control of vertigo. Endolymphatic drainage procedures (eg, endolymphatic sac surgery) continue to be widely used, but results are less predictive. If hearing is nonserviceable, destruction of the inner ear by a labyrinthectomy is effective in eliminating severe vertigo.

3. Acoustic Neuroma

**Slide 50 - Computed Tomography (CT) scan of an acoustic neuroma**

This is a CT scan following intravenous contrast showing a 2-cm tumor in the cerebellopontine angle of the posterior fossa. The tumor is centered over the internal auditory canal, and has the typical appearance of an acoustic neuroma.

**Discussion**

*Etiology.* Acoustic neuromas or schwannomas arise from the VIII cranial nerve, usually the vestibular portion. They account for about 10% of all intracranial tumors. They are benign, but can produce serious complications from compression of vital structures, especially the brain stem. Acoustic neuromas can occur bilaterally in patients with von Recklinghausen's disease.
Symptoms. Patients usually have unilateral hearing loss (sensorineural) and/or tinnitus. Vertigo is uncommon, but mild disequilibrium or unsteadiness is frequent.

Signs. Decreased corneal sensation or hypesthesia of the face occurs with larger tumors from involvement of the V cranial nerve. The VII cranial nerve is often severely compressed by the tumor, but facial weakness is rarely observed.

Diagnosis. Brain stem response audiometry is an excellent screening procedure for a possible acoustic neuroma in a patient with unilateral inner ear symptoms or findings. A CT scan with contrast (intravenous and/or air) or magnetic resonance imaging will give positive identification.

Treatment. Acoustic neuromas are not radiosensitive, but are amenable to surgical removal. Only in cases of very small tumors can hearing be preserved.

IV. Rehabilitation of Sensorineural Hearing Loss

Slide 51 - Hearing aid (schematic diagram)

The component parts of a hearing aid are illustrated in the schematic diagram.

Slides 52 & 53 - Examples of two types of hearing aids

Slide 54 - Nucleus 22-Channel Cochlear Implant System

The component part of an implant system are shown: internal receiver with attached electrodes, microphone, signal processor, and external transmitter.

Slide 55 & 56 - Schematic diagrams of an implanted cochlear prosthesis.

Discussion

Although medical or surgical therapy is rarely beneficial for sensorineural hearing loss (exceptions: penicillin and prednisone for inner ear syphilis; prednisone and/or cytoxan for autoimmune inner ear disease; surgical repair for a perilymph fistula), most patients, including infants, can benefit from a hearing aid. It is very important to fit babies with hearing aids as soon as a hearing loss is documented, so that the child will maximize his potential for speech and intellectual development. Hearing aids can be worn behind the ear or totally within the ear canal; they can also be part of an eyeglass frame. Since modern hearing aids can selectively amplify various frequencies, a person should undergo a complete hearing aid evaluation so that he can be individually fitted. Persons with sensorineural hearing loss often experience tinnitus, and the use of a hearing aid will frequently lessen this symptom as well.

In addition to the use of a hearing aid, other devices and educational techniques (ie, lip reading, signing, etc) are valuable for rehabilitating the person with a sensorineural hearing loss. For example, telephone amplifiers (portable and non-portable) can increase the loudness of the telephone signal. To further help with detection of the telephone ring a low frequency bell or a buzzer can be used for persons with a high frequency hearing loss; additionally, a
light system can convert the ring to a flashing light. Radio and television listening devices are also available. An example of the latter is an infrared system that converts audio signals to infrared waves which are transmitted to a headset worn by the patient. For the profoundly deaf, special telephone devices (TDDs) allow communication via a keyboard and printer.

The field of cochlear implantation shows great promise for the profoundly deaf. Most cochlear implant systems have five component parts: 1) a microphone to pick up the sound; 2) a processor for modifying the electrical signal; 3) a transmitter to relay (ie, by radiofrequency or electromagnetic induction) the signal to the implanted unit; 4) a receiver implanted in the temporal bone; and 5) an electrode(s) to deliver the electrical signal to the auditory nerve. Both single and multichannel devices are currently being evaluated. In some designs, the electrode system is placed within the scala tympani and in others is extra cochlear; usually positioned on the promontory near the round window.

With the present level of technology, consistent results in adult patients can be achieved: the cochlear implant recipient has a greater awareness of environmental sounds, and lip reading and voice production are improved. A small percentage of patients have limited speech discrimination without lip reading.