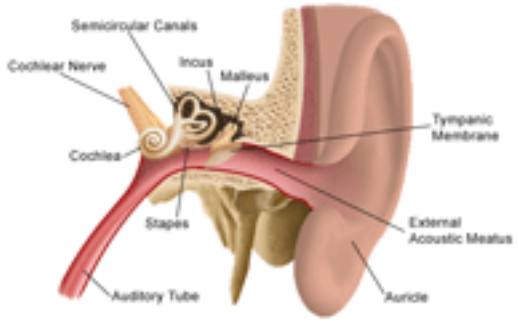
Ear Conditions

Anatomy



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The ear is the organ that is responsible for hearing and maintaining balance.

The different parts of the ear include:

- The external or outer ear, which is subdivided into the pinna or auricle, and external auditory canal or tube.
- The tympanic membrane (eardrum) is a thin film of tissue which separates the external ear from the middle ear.
- The middle ear (tympanic cavity) is an airspace that contains the ossicles (hearing bones) which are responsible for transmitting the sound waves into the inner ear. There are three ossicles named the malleus, incus and stapes. The eustachian tube is a canal that links the middle ear with the area found at the back of the nose (called the nasopharynx). The eustachian tube helps to equalize the pressure in the middle ear by allowing air to flow in or out of the middle ear. Equalized pressure is needed for the proper transfer of sound waves.
- The inner ear, consists of the cochlea and the labyrinth. The cochlea is the organ that receives and uses the sound waves from the middle ear to stimulate the nerve that is responsible for hearing. The labyrinth consists of the vestibule and semicircular canals which are responsible for informing the brain of the body's movement in different directions (up, down, left, right, and rotation).

Ear Conditions

Outer ear conditions

- **Otitis externa** is an infective condition of the outer ear (pinna/auricle and external auditory canal/tube. It is characterized by swelling and redness of the external ear, a discharge form the ear and moderate to severe pain. Sometimes there are underlying risk factors for developing this condition, for instance injury to the ear (especially the use of ear buds and other objects to try and remove wax, or to scratch an itchy ear), eczema or other skin conditions, and diabetes mellitus. In severe cases (especially in the case of abscess formation), the infection can cause destruction of the cartilage, with eventual and deformity of the ear.

- **Perichondritis** is chronic inflammation of the perichondrium (layer of tissue tightly adhering to the cartilage of the ear). It causes fluid accumulation between the cartilage of the pinna/auricle and the perichondrium. If this condition is not treated early by draining the collection, the blood supply to the cartilage will be impaired and the cartilage will break down causing deformity of the ear.

- **Blood accumulation (hematoma)** between the cartilage and perichondrium can occur as a result of an injury to the ear. The blood collection needs to be drained as soon as possible, otherwise the blood supply to the cartilage will be impaired and the cartilage will break down, causing deformity of the ear.

- **Growths in the ear canal** are mostly harmless, but if the ENT specialist finds that the growth has suspicious features, a malignancy has to be excluded with a biopsy and histological analysis. Although cancers of the outer ear most commonly arise from the skin (squamous or basal cell carcinoma), a tumor can arise from any of the other surrounding tissues. Growths can cause obstruction the ear canal resulting in build-up of wax and dead skin cells, as well as impair hearing. Bony growths (osteomas or exostosis) are mostly harmless and only need attention if complications arise.

Conditions of the Eardrum (Tympanic Membrane)

- An **acute infection of the eardrum** is called **myringitis**. It can be extremely painful. The eardrum becomes red, thickened, and may have blisters on it. The blister contains fluid or blood, and once it had ruptured, the pain becomes much less intense. **Chronic myringitis** is rare and poorly understood in its isolated form. It is not clear whether it is caused by an infection or by chronic inflammation of another source. Mostly it is associated with outer or middle ear infections.

- A **perforation of the eardrum/torn eardrum** can be caused by different types of trauma. Traumatic causes include a sudden pressure change in the external ear canal and direct trauma. Sudden pressure changes can be caused by (for example) a hit to the ear, a bomb explosion or sudden cabin pressure changes during a flight. Direct trauma can be caused by a foreign object in the ear or a fracture of the temporal bone (bony encasement of the ear structures. Pressure changes inside the middle ear can also cause an eardrum to burst. The most common cause is an acute middle ear infection where pus accumulates in the middle ear space. A perforated eardrum can result in hearing loss and/or a chronically discharging ear due to the exposure to organisms from the outside.

Conditions of the Middle Ear

- An acute infection of the middle ear is called an **acute otitis media**. The cause is either due to viruses or bacteria that enter the middle ear via the ventilation tube from the area behind the nose, or due to poor ventilation of the middle ear, causing fluid to accumulate and become infected. The middle ear becomes inflamed and often filled with pus. Acute middle ear infection can be very painful, and may cause fever and listlessness.

- When the middle ear becomes chronically inflamed the condition is called **chronic otitis media**. There are two types of chronic otitis media that differ significantly.

    - In the case of **chronic otitis media with effusion** (glue ear), poor ventilation of the middle ear causes a thick fluid to accumulate in the middle ear. The poor ventilation is caused by an inability of the ventilation tube (eustachian tube) to allow air to flow into the middle ear and drain the fluid that accumulates. This is a common problem in children for several reasons related to the early development of the ventilation tube. A glue ear may be unproblematic and will only need to be followed up by an ENT. However, if it

causes pain, discomfort, conductive hearing loss (inhibition of sound to be transmitted from the eardrum to the inner ear), or recurrent acute infections, it should be treated. Sometimes medical treatment is all that is necessary, but other times it may be necessary to make a small hole in the eardrum (myringotomy) to allow the fluid to drain and the middle ear to ventilate. A small tube (grommet) is usually inserted to keep the hole open.

  - **Chronic suppurative otitis media** (chronic infection with constantly draining ear) is characterized by a chronically infected, discharging ear through a perforated/torn eardrum. It may be that the eardrum ruptured during an acute infection of the middle ear or due to previous trauma. The middle ear is now open to organisms from the outer ear, which causes chronic infection and pus formation. Another cause of a chronically discharging ear is a cholesteatoma. This condition is almost always characterized by some degree of conductive hearing loss (inhibition of sound to be transmitted from the eardrum to the inner ear).

- **Eustachian tube dysfunction** (abnormal function of the ventilatory tube that communicates with the middle ear) is usually associated with children, where it causes chronic otitis media, but may also be present throughout life. It causes difficulty with equalizing (especially with changes in altitude), intermittent pain and mild hearing loss.

- A **cholesteatoma** is a benign skin cyst that grows inside the middle ear behind the eardrum, which can be intact or perforated. A cholesteatoma typically occurs in children and adults who have underlying ventilation problems with their middle ears. Sometimes it is something that the patient is born with (congenital cholesteatoma). Cholesteatomas are often associated with a chronically infected and discharging ear. Although it grows slowly, the content of the cholesteatoma has a tendency to destroy bone as it enlarges, which causes a significant risk of damage to the middle ear structures. It may even erode and destroy the thin bone covering the inner ear structures, the brain, the large blood vessels that lies in close relationship to it and the nerve that is responsible for facial movement (facial nerve). These consequences can be debilitating and even life-threatening. This is why this condition must always receive the urgent attention of an ENT. Symptoms may include a discharging ear, pain, hearing loss, dizziness, paralysis of the facial muscles, and sometimes infection that spreads to the brain.

- **Masses in the middle ear** are usually benign (rarely cancerous). It may arise from any of the tissues within and around the middle ear. The most common tumor is a glomus tumor which is very vascular (lots of blood supply) and develops in relation to the nerves running through the middle ear. However, although they are rare, there is a vast amount of different types of masses that may occur in the middle ear.

Symptoms may include hearing loss, fullness of the ear and tinnitus (noise originating from the ear). In more advanced cases nerve deafness, balance problems and impaired movement of the face may be present. The brain and several other nerves may be affected in very advanced cases.

Conditions affecting the ossicular bones/hearing bones

Ossicular abnormalities include all the conditions that affect the hearing bones in some way. These conditions include absence, breakdown, dislocation and/or fixation of the hearing bones. The hearing bones are essential for good hearing and therefore abnormalities of these structures always result in some degree of conductive hearing loss (inhibition of sound to be transmitted from the eardrum to the inner ear).

- **Complete absence of one or more of the hearing bones** is rare, but most commonly caused by a congenital disorder (condition that has been present since birth). It can be the only abnormality that the patient has, or it can be part of a syndrome (part of a combination of other abnormalities). Unless associated ear problems are evident at birth, it is only picked up during later childhood.

- **Erosion of one or more of the hearing bones** is often found in patients with chronic middle ear infections or the presence of a cholesteatoma. Chronic infection causes an inflammatory reaction that can erode bone over a long period of time. A cholesteatoma can also cause bone erosion by chronic inflammation, but also by production of additional elements that disrupts bone and by destroying the bone in its pathway as it grows. Any mass within the middle ear can destroy bone in its pathway as it grows. Some rare conditions of bone deformation can also cause the ossicles (made of bone) to be abnormal.

- **Dislocation of the hearing bones** can be caused by severe ear trauma, which can tear the joints between the bones. Chronic infection can destruct the part of the hearing bones that connect at a joint, causing separation between the bones.

- **Fixation of the hearing bones** can occur when chronic inflammation causes fibrotic tissue to form around the bones. Fibrotic tissue causes tissues to adhere to each other tightly, keeping them from moving freely. Another condition that causes fixation of the stapes (smallest of the hearing bones), is otosclerosis. In the case of otosclerosis, abnormal bone forms around the area where the smallest hearing bone (stapes) communicates with the fluid of the inner ear organ (cochlea), which prevents the transmission of sound waves.

Conditions affecting the inner ear (hearing structures)

Conditions affecting the inner ear hearing organ (cochlea), causes neural hearing loss, because it causes damage to the nerve endings situated in the cochlea. Abnormalities of the cochlea inhibits the ability of the nerve endings to transmit sound to the auditory nerve (hearing nerve). There is a myriad of causes for nerve deafness, which includes:

- Age related hearing loss.
- Noise exposure related hearing loss.
- Hearing loss in the family.
- Damage to the hearing structure (cochlea) from acute or chronic infections.
- Very advanced cholesteatoma or other masses that affect the cochlea.
- Severe head trauma.
- Previous ear surgery.
- Severe meningitis (infection of the lining of the brain), especially if caused by TB.
- Use of medication that can cause hearing loss.
- Hearing loss that is due to birth abnormalities.
- Systemic diseases, like HIV or diabetes mellitus.
- Auto-immune diseases (damage caused to the body by its own immune system).
- Meniere's disease is a condition that causes hearing loss, vertigo (spinning sensation) and tinnitus (noise in the ears) by causing abnormal pressure fluctuations in the inner ear.

Conditions affecting the inner ear (balance structures)

Damage to the labyrinth causes a sense of rotation (vertigo) and balance problems. The labyrinth in the inner ear consists of several structures (see anatomy) that play a big role in our ability to sense motion (up, down, left, right, and rotation). There are several conditions that can affect this structure, including:

- Primary bacterial infection of the labyrinth (labyrinthitis).
- Secondary bacterial infection of the labyrinth due to spread from a nearby area, like the brain.
- Benign paroxysmal positional vertigo (BPPV), which is caused by abnormal stimulation of the nerves in the circular canals, likely due to loose crystals.
- Meniere's disease is a condition that causes hearing loss, vertigo (spinning sensation) and tinnitus (noise in the ears) by causing abnormal pressure fluctuations in the inner ear.
- Previous ear surgery.

- Severe head trauma.
- Leak around the area of communication between the middle and inner ear (perilymphatic fistula).
- Functional vertigo (increased awareness of balance or perceived imbalance).

Conditions affecting the hearing and balance nerves (audiovestibular nerves)

There are several conditions that can cause **damage to the nerves** responsible for transmitting the sound and movement impulses from the inner ear to the brain, for example:

- Vestibular neuritis is understood to be a viral infection of the vestibular nerve (balance nerve), causing swelling of the nerve in its canal (not associated with hearing loss).
- Viral infection of both the hearing and balance nerves caused by the Herpes Zoster virus and also called Ramsey Hunt syndrome (associated with hearing loss and possibly sores of the ear, face or mouth on the affected side).
- Tumors (masses) growing anywhere along the course of the nerves.
- Migraine.
- Stroke.
- Brain damage.
- Multiple sclerosis or any other conditions that causes deterioration of nerve function.

Hearing loss

The normal lower limit of the hearing range is 0-20 decibels (dB), where 0 dB is the threshold for the perception of sound at a given frequency for people with normal hearing. Deafness is a partial or complete loss of hearing, also known as hearing impairment. Hearing loss is measured in decibels hearing loss (dB HL). It can be graded as follows:

- 20-40 dB HL: mild, cannot hear whispers.
- 41-70 dB HL: moderate, cannot hear conversational speech.
- 71-95 dB HL: severe, cannot hear shouting.
- >95 dB HL: profound, cannot hear sounds that would be painful to others.

There are two **types of deafness**:

- *Conductive hearing loss* occurs when there is a problem in the transmission of sound waves through the external and middle ears.
- *Sensorineural hearing loss* refers to problems occurring in the cochlea, cochlear nerve or brain stem.

Causes for **sensorineural hearing loss** include (many have been mentioned previously):

- Age related hearing loss.
- Noise exposure related hearing loss.
- Hearing loss in the family.
- Damage to the hearing structure (cochlea) from acute or chronic infections.
- Very advanced cholesteatoma or other masses that affect the cochlea.
- Severe head trauma.
- Previous ear surgery.
- Severe meningitis (infection of the lining of the brain), especially if caused by TB.
- Use of medication that can cause hearing loss.
- Hearing loss that is due to birth abnormalities.
- Systemic diseases, like HIV or diabetes mellitus.
- Auto-immune diseases (damage caused to the body by its own immune system).
- Meniere's disease is a condition that causes hearing loss, vertigo (spinning sensation) and tinnitus (noise in the ears) by causing abnormal pressure fluctuations in the inner ear.

Although symptoms improve between episodes, it causes progressive hearing loss and the vertiginous symptoms can become debilitating.

Causes of **conductive hearing loss** include (many have been mentioned previously):

- Obstruction of the external ear canal (hard impacted wax/cerumen, foreign bodies, swelling of the ear canal secondary to infection, growths in the ear canal).
- Abnormalities of the eardrum, for example a perforation/hole or scarring.
- Middle ear conditions which include ear infections, abnormal ventilation of the middle ear, cholesteatoma and abnormalities of the ossicles (hearing bones).
- Birth abnormalities.

There are several **investigations** that can be used to establish the extent and maybe find the cause of the hearing loss:

- Audiogram.
- Oto-acoustic emissions.
- Tympanogram.
- Auditory brainstem testing.
- A CT/MRI scans.
- Blood tests.

An *audiogram* measures your ability to perceive sound at different frequencies presented at different degrees of loudness (dB).

In small children, it is not possible to do such an assessment. Therefore, the response of the inner ear structures is measured by recording the *oto-acoustic emissions*. Oto-acoustic emissions (OAEs) are low-level sound emitted by the cochlea either spontaneously or evoked by an auditory stimulus. It is not a very specific test, but serves as a good screening tool in children.

A **t***ympanogram* tests the mobility of the eardrum, which is often requested when poor ventilation of the middle ear is suspected or when there is fluid in the middle ear.

If there is still no clarity on whether there is hearing loss, the *brainstem responses to auditory stimuli* can be measured with electrodes placed on the head.

Sometimes a *CT or MRI scan* is requested to evaluate all the structures of the hearing system.

In certain circumstances *blood tests* can give some valuable insight if an auto-immune condition, certain infections, metabolic disorders, or inheritable congenital problems are suspected.

Treating hearing loss is aimed at the underlying cause. If the hearing is still not adequate after treatment or surgery, hearing amplification can be considered. There are several types of instruments that can be used to amplify hearing, including hearing aids that are worn inside or behind the ear, and implantable hearing aids (bone-conduction hearing aid and cochlear implant).

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Tinnitus

Tinnitus is the perception of a sound in the ears which is not generated by the environment. It is important to differentiate between **pulsatile tinnitus** (throbbing sound/rhythmic pulsation) and **non-pulsatile tinnitus** (constant sounds), as the causes and management are very different.

Pulsatile tinnitus (throbbing sound/rhythmic pulsation) usually originates within the blood vessels inside the head or neck region. The noise of the turbulent blood flow can be heard in the ears. This results from either increased blood flow or a narrowing of the opening of the blood vessel, both of which result in turbulent blood flow that can be heard in the ears. Some of the possible causes include:

- Pregnancy.
- Hyperthyroidism.
- Hypertension.
- Anemia.
- Atherosclerotic disease of the carotid arteries, or the smaller vessels that lie in close association with the hearing structures.
- Conductive hearing loss (inability of the ear to carry the soundwave from the environment to the inner ear structures) can cause body sounds to be perceived more intensely.
- Vascular abnormalities in the brain (aneurysms or other lesions).
- Vascular tumors in close relation to or involving the ear structures (eg. Glomus tumors).
- Benign intracranial hypertension, where there is increased pressure of the cerebrospinal fluid that bathes the brain.

Non-pulsatile tinnitus may be characterized by a ringing, buzzing, whistling, humming, clicking or hissing sound. Although bothersome, tinnitus usually isn't a sign of something serious. Treating an identified underlying cause sometimes helps. Other treatments reduce or mask the noise, making tinnitus less noticeable. Non-pulsatile tinnitus may be caused by:

- Hearing loss.
- Noise exposure.
- Certain medications.
- Temporomandibular joint disorders (joint between the lower jaw and the skull).
- Meniere's disease (condition of increased pressure in the inner ear structures).
- Masses/tumors involving the acoustic nerve (hearing nerve).
- Often the cause is unknown.

Treatment of tinnitus involves treating any underlying causes. In the case of non-pulsatile tinnitus, there may not be a treatable cause. Adjusting to tinnitus can be assisted by psychotherapy and tinnitus-retraining therapy.