The sense of hearing connects us with our fellow human beings. Every dialogue, every conversation depends on it. Deaf people have the highest suicide risk. They are cut off from their social environment in a much more severe way than blind or paralysed people are. Unfortunately, age-related hearing loss and balance disorders are not rare at all and they have an enormous effect on the quality of life—even without a complete loss of hearing. The diseases and the respective diagnostic tests which are going to be mentioned here will give you an overview of the wide field of diseases of the inner ear, their diagnosis and treatment possibilities.

Causes for Hearing Impairments

Hearing impairments may have various reasons. In order to address this huge field, we will first differentiate between conductive hearing loss and sensorineural hearing.
**Loss.** The first stems from a damage to the middle ear, which prevents the proper transmission of sound.

The relevant stimulus, the sound, does not or not completely reach the hearing organ—while the latter is in itself functional. In case of a sensorineural hearing loss, however, it is this very hearing organ that is damaged. The sound does reach the inner ear but it cannot be processed fully or at all, or it might not be forwarded properly.

Due to the extensive and rather different issues that these two disorders encompass, this article concentrates on sensorineural hearing loss and the inner ear as an organ.

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**Rinne and Weber – Middle Ear or Inner Ear?**

For the detection of a pathogenesis of the inner ear, there are two classic methods of audiometry available. Both hearing examinations use with a tuning fork.

**Weber Test**
The vibrating tuning fork is placed on the midline of the patient’s head. Healthy individuals hear the bone-conducted sound equally on both sides. With patients with a unilateral hearing impairment, there are two possibilities:

- The sound is perceived only/louder on the affected side: **Conductive hearing loss/damage to the middle ear**
- The sound is perceived only/louder with the unaffected ear: **sensorineural hearing loss**

**Explanation:** If the sound transmission is blocked, the stimulus that is conducted from the head midline through the bone is perceived louder. Loss of sound through the blocked middle ear is prevented and the impact on the patient’s cochlea is enhanced. Therefore, the sound is perceived louder in the affected ear. If it is however the inner ear that is causing the problem, it does not matter how the sound is being conducted. The hearing ability as such is impaired, and therefore, the sound can only be perceived in the opposite, healthy ear.

**Rinne Test**

The vibrating tuning fork is placed on the patient’s mastoid bone behind the ear. The patient is asked to say when he is no longer able to hear the sound. At this moment, the still vibrating fork is placed in front of the patient’s ear. Normally, the patient should now hear the sound again, as **air conduction is better than bone conduction** and can transmit quieter sounds. This is referred to as **Rinne positive**. If the sound is **not heard by air conduction**, the diagnosis is **Rinne negative**. Both findings are possible with an impaired ear:

- **Rinne positive:** Air conduction and the middle ear still work better than bone conduction. Damage has to be located in the inner ear (**sensorineural hearing loss**), which means that both ways of conduction are affected in the same way.
- **Rinne negative:** Air conduction does not work as good as bone conduction.
Since this is pathological, the damage has to be located in the middle ear. It is a conductive hearing loss.

Audiometry

A more precise diagnostic tool for hearing impairments is pure tone audiometry. An audiometer is an electronic device that produces acoustic stimuli, i.e. pure tones at intervals of one octave. The frequency (Hz) and intensity (decibels) of each tone can be regulated. Both ears are tested individually. This device can either measure air conduction using normal earphones, or bone conduction with a special bone vibrator that is placed on the mastoid.

Measurement of Hearing Thresholds

The audiometer basically produces a very low tone whose intensity is increased until the patient signals when a tone is heard. This hearing threshold is then recorded for each tone or frequency. For the following section, one should be familiar with the physical parameters of hearing (sound pressure level, frequency).

Implementation

The lowest sound is defined by the hearing threshold of a healthy teenager. It is used as a reference level on the audiogram. Measuring starts at a frequency of 1000 Hz since hearing is usually best at this frequency. The volume is increased in steps of 1 dB, and also the hearing threshold of the patient is recorded in decibels.

Evaluation

First, an important pointer: In many audiograms, the vertical axis is plotted from top to bottom, meaning the further down on the audiogram, the louder the sound and the worse the sense of hearing. For an individual with normal hearing ability, the lines for air conduction (AC) and bone conduction (BC) will be mostly parallel to the horizontal axis and close to the reference level. A hearing loss will manifest on the graph as one of the following types of deviations:

- AC is markedly worse than BC: conductive hearing loss; the hearing via bone conduction is not affected.
- AC and BC are close together and both worse: sensorineural hearing loss; both ways of conduction affected equally.
- AC and BC both worse, but AC more affected than BC: a combination of conductive and sensorineural hearing loss.

Typically, the curve drops most in the sector of high frequencies since high-pitched tones are more difficult to perceive than low-pitched tones (typical of presbyacusis). Curves that deviate from this pattern might be an indicator of an hereditary hearing disorder or Ménière’s disease.

Audiograms
Pathogenesis of the Inner Ear

As complex as the ear is, as diverse and numerous are its malfunctions and disorders. The focus will be on the most common diseases which are often tested in exams. Pathologies of the ear include hearing impairments as well as balance disorders.

Tinnitus: Symptom Without a Disease?

Tinnitus is mentioned here even though it is not a disease but a symptom. It refers to sounds and tones that are perceived by the patient but which are not generated by an external source. The sound can be constant or transient, unilateral or bilateral, and it can affect the patient’s living quality in varying degrees, from hardly noticeable to suicidal thoughts.

One can differentiate between two types of tinnitus. The objective tinnitus can be measured by the examiner, for example with a microphone or stethoscope. The cause of this type of tinnitus may be the blood flow from adjacent blood vessels (vascular tinnitus - always pulse synchronous) or contractions of the muscles of the middle ear or the palate (muscular tinnitus - rhythmical clicking sounds).

Only in very few cases can produce the sounds by themselves, their origin is assumed to be located in the external hair cells. Generally, the objective tinnitus is rather rare. It is treated by removing the causing factors, for example by injection of botulinum toxine into the contracting muscles.

The question of the exact genesis of the subjective tinnitus has not been resolved so far. It might have its origin in the cochlea, the auditory nerves, or the auditory pathway. What provokes the spontaneous activation of those structures, is as of yet not totally understood.

Treatment of Tinnitus

The objective in treating a tinnitus is not to get rid of it but rather to help the patient to live with it. In a consultation with the patient, it should be determined where the source of the sound is probably located, and he/she should be assured that the tinnitus does not carry any danger of leading to hearing loss or any other disease of the ear. As a tinnitus can be associated with psychological problems, psychotherapy should be taken into
consideration if and when the patient reports depression or any other psychological problem.

Another possibility is “covering up” the tinnitus with another external source of sound, the so-called tinnitus masker. The produced frequencies (white noise) are similar to the ones of the tinnitus. Because they have an external source, the brain actively suppresses them and they are no longer perceived. Everyday tricks for managing tinnitus follow the same principle; for example, sleeping with open windows or environmental noise. Silence can be an issue for tinnitus patients as it increases the perception of the tinnitus noise.

Vertigo: Malfunctions of the Vestibular System

An often forgotten but just as important function of the inner ear is balance. Disturbances of the sense of balance can lead to vertigo. Vertigo (from Latin vertigo = dizziness) describes the sensation of a movement between oneself and the surroundings which is actually not there. Importantly, vertigo is not limited to a feeling of rotation (spinning); other forms include upward lifting, swaying, rocking, or unsystematic movement. In the following, two different types of pathological vertigo will be addressed which both have their origin in the inner ear.

Vertibular Neuronitis

Vestibular neuronitis or vestibular neuropathy is a usually unilateral dysfunction of the vestibular system. It presents as an acute and severe attack of vertigo. Patients are not able to stand up and have nausea. The assumed cause is a viral infection or a reactivation thereof which damages the sensory epithelium or the vestibular ganglia. One example for a virus that can remain latent in vestibular ganglia is herpes simplex. Another possibility is a vascular genesis of the disease. However, the exact pathomechanism remains unknown.

Epidemiology of Vestibular Neuronitis

The age of onset is between the age of 30 and 60 years. After the benign paroxysmal positional vertigo and Ménière’s disease, vestibular neuronitis is the third most common
As described above, vestibular neuronitis is a sudden attack of severe vertigo associated with extreme nausea and vomiting which affects previously healthy persons. Patients tend to fall towards the affected side when standing or sitting. The sudden onset is an indicator for a virus infection. During examination with Frenzel goggles, a strong horizontal rotating nystagmus with fast phase oscillations toward the healthy ear can be observed.

The sense of hearing is not affected; pressure in the ear or tinnitus are not found either. Only the vestibular organ is affected. The dysfunction can be compensated by the central nervous system so that symptoms will often gradually improve after a few days. This can be supported by training the sense of balance.

**Treatment**

Treatment first concentrates on alleviating the patient’s suffering with sedatives and anti-emetics. When compensation starts, ambulation and balancing training are important in order to speed up the compensation process. Patients with a good general health are able to completely compensate the vestibular dysfunction.

**Note:** Since the disorder is assumed to be a post-infectious autoimmune reaction, the administration of glucocorticoids has been proven helpful in containing the immune system.

**Benign Paroxysmal Positional Vertigo**
This condition is a peripheral vestibular disorder which is usually unilateral. Patients suffer from vertigo and nausea for about 1 minute when changing position (e.g., getting out of bed) or other movements (for example, tilting your head backwards when looking up).

This type of vertigo is triggered by inorganic particles (so-called otoliths) that have migrated into the endolymp of one or more of the semicircular canals. The otoliths have dislodged from the otolithic membrane and migrate through the cupula into the lumen of a semicircular canal. Due to the anatomical position, it is mostly the posterior semicircular canal that becomes affected.

Otoliths can become dislodged either because of a trauma (e.g., a strong blow to the head) or spontaneously. The likelihood for this to happen increases around the age of 60.

Now, if there are particles in the endolymp of the semicircular canals, the following happens when sitting up in bed: Otoliths are sensitive to gravity; thus, they flow downwards in the liquid when sitting up. This causes strong movement within the endolymp, which would normally remain still, and thus the nerve endings in the semicircular canals receive the message that there is a rotating movement even though there is none—leading to the sensation of vertigo.

Symptoms and Diagnosis of Benign Paroxysmal Positional Vertigo

Anamnesis starts with the patient's history. Typically, the patient has experienced vertigo in the situations described above which has abated after one minute at the latest. In addition to history taking, certain diagnostic maneuvers can be performed.

Dix-Hallpike Maneuver

This test is performed with the patient sitting upright. The patient's head is rotated 45 degrees toward the affected side. Then a quick change of position follows in which the patient lies on the unaffected side. Patient stays in this position for about 20 seconds and his/her eyes are observed. Patients with benign paroxysmal positional vertigo will show a rotational nystagmus. Sometimes, the testing has to be interrupted if the patient's vertigo symptoms become more than bearable. After lying on his side, the patient goes back to the upright position and the head back to the starting position.
Treatment

It is a benign type of vertigo, which means that symptoms will disappear on their own after a few months. Should symptoms be persistent, there are possible interventions which include the **repositioning maneuvers Epley or Semont**. The objective of these procedures is to reposition the otoliths from the semicircular canal back into the utricle. For this purpose, the patient’s head is moved helically so that particles can be removed by the flow from the semicircular canals towards the side that is opposed to the cupula.

Afterwards, the patient will have to sleep in a sitting position for three days in order to avoid irritation of the posterior semicircular canal. This treatment is normally very efficient. In the rare case that a patient should not respond to it but needs the sense of balance for his/her work (e.g., as a roofer), another treatment option is a surgery which deactivates the posterior semicircular canal.

Ménière’s Disease

This disease combines a pathogenesis of **hearing and balance**. It is caused by an accumulation of fluid in the endolymph, the so-called **endolymphatic hydrops**. One possible genesis of this hydrops may be insufficient reabsorption of endolymph in the **endolymphatic sac**. This strongly increases the pressure in the endolymphatic system (i.e., cochlea and vestibular organ).

The attacks of vertigo stem from a rupture of the Reissner membrane, which separates the endolymph from the perilymph, and thus results in a mixture of endolymph and perilymph with a changed ion concentration.

Epidemiology of Ménière’s Disease

Women are slightly more affected by Ménière’s disease than men. The peak incidence is between the ages of 40 and 60 years.

Symptoms and Diagnosis of Ménière’s Disease

Ménière’s disease is characterized by a **symptom triad of vertigo, tinnitus and hearing impairment**. Vertigo can last from a few minutes to several hours. The hearing impairment mostly affects **low frequencies** and can therefore be easily differentiated on an audiogram from common hearing loss, which usually manifests in high frequency areas.

Initially, the hearing will recover, which leads to a fluctuating hearing ability. After various seizures, however, a permanent hearing loss remains. Based on this symptom triad, and unspecific nystagms and an audiogram this disease can be readily diagnosed.

Treatment

In order to alleviate the patient’s acute complaints, sedatives can be given. Medication that stimulates the blood circulation is supposed to improve the reabsorption in the endolymphatic sac. In a next step, betahistine (H1-receptor agonist) and diuretics are prescribed. If symptoms persist despite these treatments, surgery can be considered.
Popular Exam Questions on Diseases of the Inner Ear

Solutions can be found below the references.

1. A patient suffers from hearing loss in his right ear. The doctor presumes a damage to the middle ear. Which of the following findings would support this assumption?
   A. Rinne test is negative; Weber test shows lateralization to the left side.
   B. Rinne test is positive; Weber test shows lateralization to the right side.
   C. A conductive hearing loss cannot be diagnosed by Weber or Rinne tests.
   D. Rinne test is negative; Weber test shows lateralization to the right side.
   E. Rinne test is positive; Weber test shows lateralization to the left side.

2. Which statement concerning audiometry is incorrect?
   A. Hearing threshold close to the reference line indicates a good sense of hearing.
   B. Hearing impairment in the area of low frequencies is typical of Ménière's disease.
   C. If conductive and sensorineural hearing loss are combined, air conduction and bone conduction will both be affected.
   D. When assessing the auditory threshold, the volume is increased in steps of 1 decibel.
   E. In case of conductive hearing loss, only the curve of bone conduction drops.

3. Which association between symptom and disease is correct?
   A. Tinnitus – vestibular neuronitis
   B. Vertigo for several hours – benign paroxysmal positional vertigo
   C. Rotational vertigo – tinnitus
   D. Tinnitus – Ménière’s disease
   E. Vertigo after standing up – vestibular neuronitis

References


Correct answers: 1D, 2C, 3D

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Notes