Inner Ear Disorders — Vertigo, Ménière's Disease, and More: Audiometry and Pathogenesis

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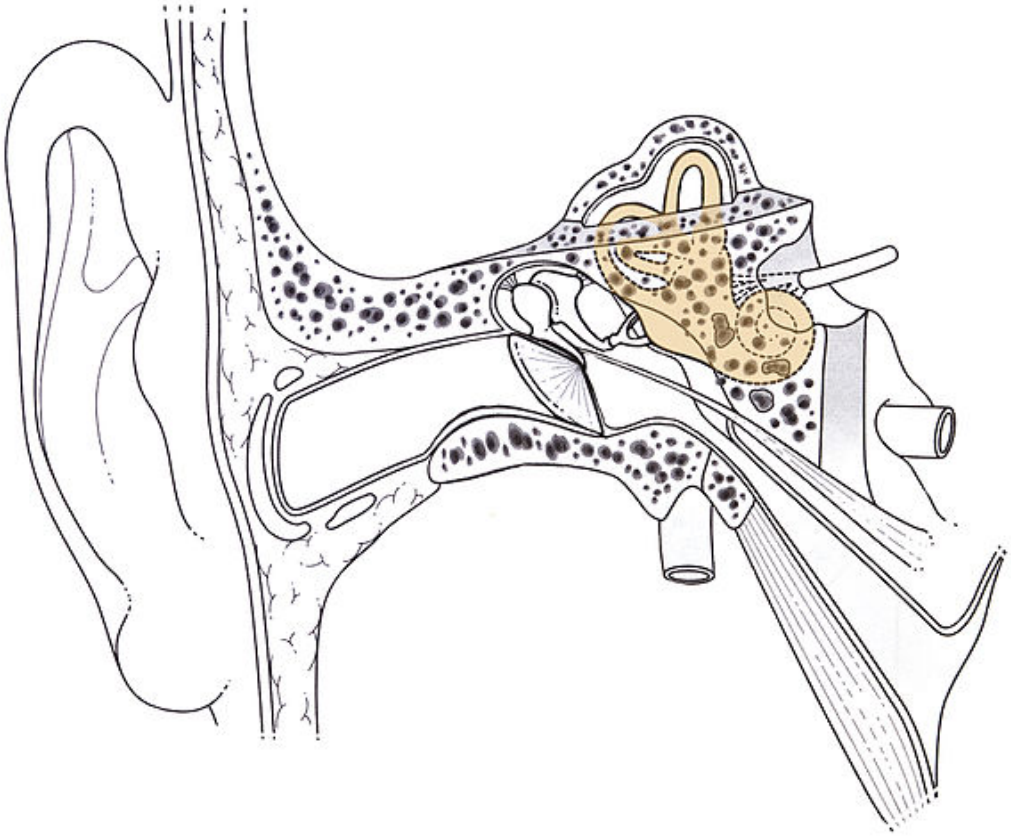
The sense of hearing connects us with our fellow human beings. Every dialogue, every conversation depends on it. Deaf people have the highest suicide risk. They are cut off from their social environment in a much more severe way than blind or paralysed people are. Unfortunately, age-related hearing loss 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 have various causes. This article will first differentiate between **conductive hearing loss** and **sensorineural hearing loss**.

The first stems from damage to the middle ear, which prevents the proper transmission of sound. With **conductive hearing loss**, the relevant stimulus, the sound, does not reach the hearing organ. With sensorineural hearing loss, however, the hearing organ is damaged, so sound does reach the inner ear but cannot be processed fully or at all, or sound might not be forwarded properly.

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

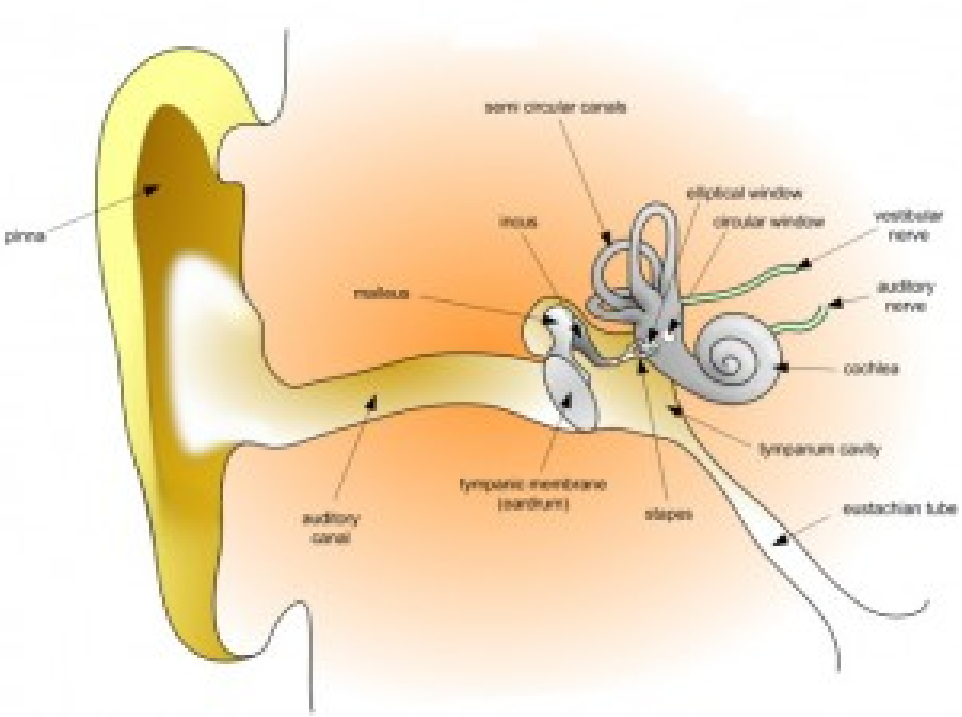


Image: Anatomy of the human ear. By Maksim, License: Public domain

Rinne and Weber: Middle Ear or Inner Ear?

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

The Weber test

Image: Tuning fork. By Wollschaf, License: CC BY-SA 3.0

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. In patients with unilateral hearing impairment, two possibilities exist:

- The sound is perceived only or more loudly by the affected ear: **conductive hearing loss/damage to the middle ear**.
- The sound is perceived only or more loudly by 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 as 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 as louder in the affected ear. If, however, the inner ear is causing the problem, it does not matter how the sound is being conducted. Hearing ability as such is impaired; therefore, the sound can be perceived only in the opposite, healthy ear.

The 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 or she is no longer able to hear the sound. At that moment, the still vibrating fork is placed in front of the patient's ear. Normally, the patient should hear the sound again, as **air conduction (AC) is better than bone conduction (BC)** and can transmit quieter sounds. This is referred to as **Rinne positive**. If the sound is **not heard by AC**, the diagnosis is **Rinne negative**. Both findings are possible with an impaired ear:

- **Rinne positive:** AC still works better than BC. 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:** AC does not work as well as BC. Because this is pathological, the damage has to be located in the middle ear. It is **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 (Hertz, or Hz) and intensity (decibels, or dB) of each tone can be regulated. Both ears are tested individually. This device can either measure AC using normal earphones or BC with a special bone vibrator that is placed on the mastoid.

Measurement of hearing thresholds

An audiometer produces a very low tone whose intensity is increased until the patient signals when a tone is heard. That **hearing threshold** then is recorded for each tone or frequency. To understand the following section, the reader 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 1,000 Hz because hearing is usually best at that frequency. Volume is increased in intervals of one dB, and the hearing threshold of the patient is recorded in decibels.

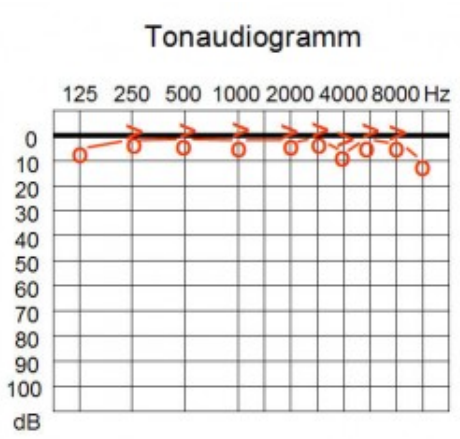
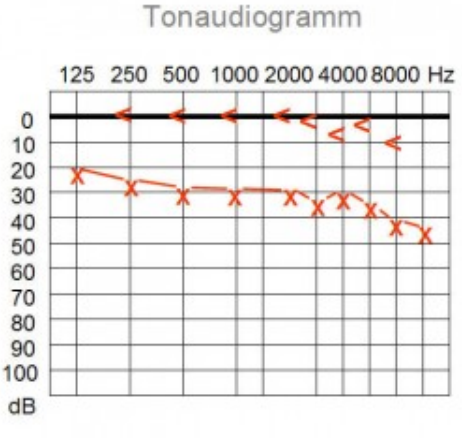
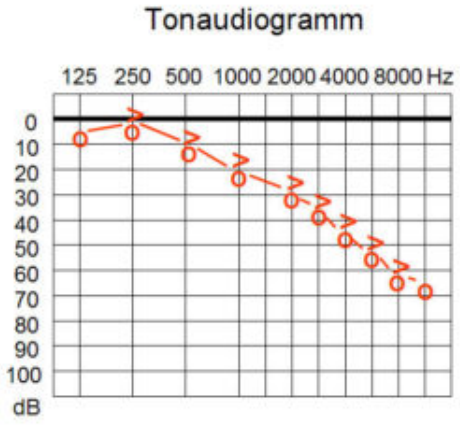
Evaluation

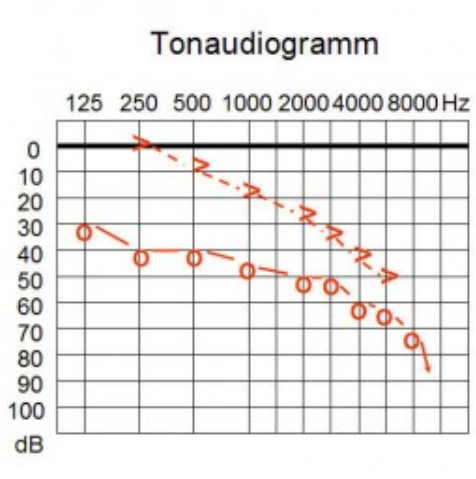
In many audiograms, the vertical axis is plotted from top to bottom, meaning that the further down on the audiogram, the louder the sound and the worse the sense of hearing. For an individual with normal hearing ability, the lines for AC and BC will be mostly parallel to the horizontal axis and close to the reference level. 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 BC is not affected.
- AC and BC are close together and both worse: **sensorineural hearing loss**; both ways of conduction are affected equally.
- AC and BC both worse, but AC more affected than BC: **the patient has a combination of conductive and sensorineural hearing loss**.

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

Audiograms

Normal hearing	 Image: Tone audiogram, normal hearing. By Welleschik, License: CC BY-SA 3.0
Conductive hearing loss	 Image: Tone audiogram, conductive hearing loss. By Welleschik, License: CC BY-SA 3.0
Sensorineural hearing loss	 Image: Tone audiogram, sensorineural hearing loss. By Welleschik, License: CC BY-SA 3.0

Combined hearing loss	 Image: Tone audiogram, combined hearing loss. By Welleschik, License: CC BY-SA 3.0

Pathogenesis of the Inner Ear

The ear is complex, and its malfunctions and disorders are diverse and numerous. This article focuses on the most common diseases tested during exams. Pathologies of the ear include hearing impairments as well as balance disorders.

Tinnitus: symptom without a disease?

Tinnitus is not a disease but a symptom. Tinnitus refers to sounds and tones that are perceived by the patient but not generated by an external source. Such sounds can be constant or transient and unilateral or bilateral. Tinnitus can affect a patient's quality of life to varying degrees, from hardly noticeable to suicidal thoughts.

Two types of tinnitus exist. **Objective tinnitus** can be measured by an examiner, for example with a microphone or stethoscope. The cause of objective tinnitus may be 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**, with rhythmic clicking sounds).

Generally, objective tinnitus is rather rare. Treatment involves removing causative factors, such as injection of botulinum toxin into the contracting muscles.

The exact genesis of **subjective tinnitus** has not been identified. The origin might be in the **cochlea**, **auditory nerves**, or **auditory pathway**. What provokes spontaneous activation of those structures is not totally understood.

Treatment of tinnitus

The objective of treating tinnitus is to help the patient live with the condition. In consultation with the patient, the healthcare professional should determine the likely source of the sound. The healthcare professional also should assure the patient that tinnitus is not associated with hearing loss or any other disease of the ear. As tinnitus can be associated with psychological problems, **psychotherapy** should be considered if and when a patient reports depression or any other psychological problem.

For treatment, the possibility is 'covering up' tinnitus with another external source of the sound, a so-called **tinnitus masker**. The produced frequencies (white noise) are similar

to the sounds of tinnitus. Because the new sounds have an external source, the brain actively suppresses those sounds, and they are no longer perceived and neither are the tinnitus sounds. Everyday tricks for managing tinnitus follow the same principle; for example, sleeping with open windows or applying **environmental noise**. Silence can be an issue for tinnitus patients because silence increases the perception of the tinnitus noise.

Vertigo: Malfunctions of the Vestibular System

An often forgotten but very 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 movement between oneself and the surroundings that is not actually there. V

Vertigo is not limited to a feeling of rotation (spinning); other forms include upward-lifting, swaying, rocking, or unsystematic movement. The following section will address two types of pathological vertigo, which both have their origins in the inner ear.

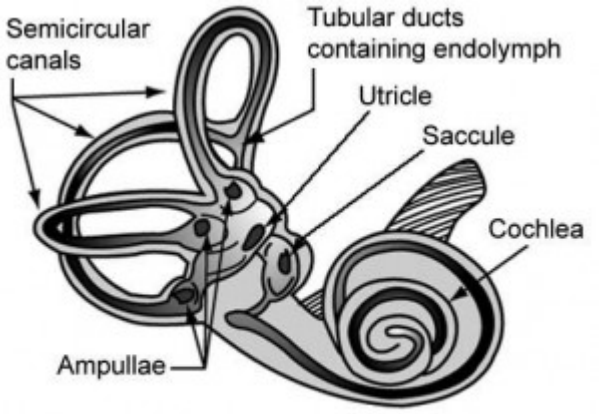


Figure 2: The Vestibular System - semicircular canals and otolith organs

Image: The vestibular system. By NASA, License: Public domain

Vestibular neuronitis

Vestibular neuronitis, or vestibular neuropathy, is usually a unilateral dysfunction of the vestibular system. The condition presents as an **acute and severe attack of vertigo**. **Patients are not able to stand up and have nausea**. The assumed cause is viral infection or reactivation which damages the sensory epithelium or the vestibular ganglia. One example of 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 30 to 60 years. After benign paroxysmal positional vertigo and Ménière’s disease, vestibular neuronitis is the third most common peripheral vestibular vertigo.

Symptoms and diagnosis of dysfunction in the vestibular system

Image: Frenzel goggles. By Polarlys, License: CC BY-SA 3.0

As described above, vestibular neuronitis is a sudden attack of severe vertigo associated with extreme nausea and vomiting. The condition often affects previously healthy persons. Patients tend to fall toward the affected side when standing or sitting. The sudden onset is an indicator of viral infection. During examination with Frenzel goggles, a patient experiences strong horizontal rotating nystagmus with fast phase oscillations toward the healthy ear.

The sense of hearing is not affected, pressure in the ear remains normal and there is no tinnitus found. **Only the vestibular organ** is affected. The dysfunction can be compensated by the central nervous system so that symptoms 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 balance training are important in order to speed up the compensation process. Patients with good general health are able to completely compensate for vestibular dysfunction.

Note: Because 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

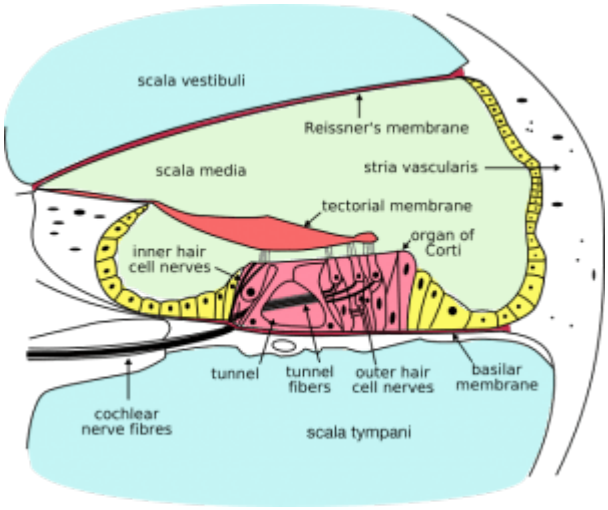


Image: Section through the cochlea (schematic). By Philipp N., License: CC BY-SA 3.0

This condition is a peripheral vestibular disorder and is usually unilateral. Patients suffer from vertigo and nausea for about one minute when changing position (e.g., getting out of bed) or attempting other movements (e.g., tilting the head backward to lookup).

This type of vertigo is triggered by inorganic particles (otoliths) that have migrated into the **endolymph** of one or more of the semicircular canals. Otoliths can dislodge from the otolithic membrane and migrate through the cupula into the lumen of a semicircular canal. Due to the anatomical position, mostly the posterior semicircular canal becomes affected.

Otoliths can become dislodged either because of trauma (e.g., a strong blow to the head) or spontaneously. Likelihood increases around the age of 60 years. Otoliths are sensitive to gravity; thus, they flow downward in the liquid when the person sits up. This causes strong movement within the endolymph, which would normally remain still. The nerve endings in the semicircular canal 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

Patient history is important. Typically, a patient has experienced vertigo in situations similar to those described above and symptoms have abated after one minute. In addition, certain **diagnostic maneuvers** can be performed.

Dix-Hallpike maneuver

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

Treatment

As the name says, benign paroxysmal positional vertigo is a benign type of vertigo, so symptoms disappear on their own after a few months. Should symptoms persist, possible interventions include the **repositioning maneuvers Epley or Semont**. The objective is to reposition otoliths from the semicircular canal back into the utricle. The patient's head is moved helically so that particles can be removed by the flow from the semicircular canal toward the side that is opposed to the cupula.

Afterward, the patient must sleep in a seated position for three days in order to avoid irritation of the posterior semicircular canal. The treatment is normally very efficient. In the rare case that a patient does not respond but needs a sense of balance for work, another treatment option is surgery to deactivate the posterior semicircular canal.

Ménière's Disease

Ménière's disease combines the pathogenesis of **hearing and balance**. The disease is caused by the accumulation of fluid in the endolymph, the so-called **endolymphatic hydrops**. One possible genesis of hydrops is insufficient reabsorption of endolymph in the **endolymphatic sac**. This strongly increases pressure in the endolymphatic system (the cochlea and vestibular organ).

The attacks of vertigo stem from a rupture of the Reissner membrane, which separates the endolymph from the perilymph, resulting 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. Hearing impairment affects mostly **low frequencies** and can, therefore, be easily differentiated on an audiogram from common hearing loss, which usually manifests in high frequencies.

Initially, the hearing will recover, leading to fluctuating hearing ability. After various seizures, however, permanent hearing loss occurs. Based on the symptom triad, unspecific nystagmus, and an audiogram, the disease can be easily diagnosed.

Treatment

Sedatives may alleviate a patient's acute complaints. Medication that stimulates blood circulation is supposed to improve reabsorption in the endolymphatic sac. Furthermore, betahistine (H1 receptor agonist) and diuretics may be prescribed. If symptoms persist despite those treatments, surgery can be considered.

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Notes