

Ear

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Comments on Embryology of the Ear

External Ear

The auricle forms from the 4th week, forming 6 paired prominences of mesenchymal tissue of the first mandible and second hyoid brachial arch.

The following grow from the mandibular arch:

- **Tuberculum tragicum**
- **Tuberculum helcis**
- **Tuberculum helcis intermedium**

The following grow from the hyoid arch:

- **Tuberculum anthelcis**
- **Tuberculum antitragicum**
- **Tuberculum lobulare**

The auricle is formed by merging the above mentioned tuberculi until the 3rd month. The ectoderm of the first branchial sulcus forms the concha auricular. The auricle is primarily located caudally, but with the mandible evolving (until the 20th week) it migrates cranially to its common location. In 4-5 years old, the auricle has 80% of its size; in 9 years old, it has its common size.

The external auditory canal evolves from the ectoderm of the first branchial sulcus. The ectoderm forms a strip from the 4th week, which migrates medially to meet the endoderm from the first branchial fissure. This strip canalizes from the 28th week. The cartilaginous part

of the auditory canal corresponds with the primary auditory canal; the bone part corresponds with the epithelial strip. The fibrous layer stays between the auditory canal and the tympanic cavity, which is the base of the eardrum. Medially from the eardrum is located the endoderm of the first branchial exagination, from which the middle ear cavity is formed. The ossification of the bone part of the auditory canal starts in the 12th week.

Middle Ear and Eustachian Tube (ET)

The middle ear and ET evolve from the tubotympanal recessus formed by the 1st entodermal pharyngeal pocket, which appears in the 3rd embryonic week. This pocket contracts in the 2nd month into a bottle shape. The contracted part lengthens and forms the ET. Blind externally, it widens and is divided into 4 pouches: anterior, middle, posterior and superior, which form the tympanic cavity and pneumatisation of the temporal bone. The middle ear cavity is filled with the mucous mesenchymal tissue during its evolution. From the 3rd embryonic month this mesenchymal tissue dissolves and starts reabsorbing. The complete resorption lasts until the 1st year of age and sometimes even longer.

Musculus tensor tympani is formed from the mesoderm of the first branchial arch and is innervated by the mandible branch of n.V. Musculus stapedius evolves from the mesoderm of the 2nd arch and is innervated from n.VII.

Malleus and incus evolve from the 4th week from the Meckel's cartilage of the 1st arch (neck, head of malleus, short processus and body of incus) and from the Reichert's cartilage of the 2nd arch (manubrium mallei, long processus of incus and structures of stapes). The medial part of the stapedic plate and ligamentum anulare stapedis evolve from the otopocket. The ossification of the incus and malleus starts in the 15th week and in the 24th week it is complete. The stapes development starts in the 4th week as a stapedic circle around arteria stapedia. It ossifies between the 18th and 24th week.

Antrum Mastoideum

Antrum mastoideum evolves as an extension of the epitympanum between the 21st and 22nd week. The pyramid pneumatisation starts from the 28th week and the pneumatisation of mastoids starts from the 33rd week. The pneumatisation of the temporal bone is complete years after the birth and this process may not be ended even in adults.

The interior of ET is formed from persistent first pharyngeal evagination. The entodermal part of the evagination extends laterally and its distal part extends towards the middle ear. Musculus levator veli palatini and tensor veli are formed between the 10th and 12th week. ET grows and its lumen starts to evolve: in the 10th week it has 1mm, at birth it has 13mm. The most significant growth proceeds in the cartilaginous part. The skull base is flat in neonates so ET is practically horizontal in early childhood.

Nervus VII. and VIII.

N.VII. is a nerve of the 2nd branchial arch. It can be identified from the 3rd week together with the statoacoustic nerve as a cell aggregate – acousticofacial ganglion – ventrally from placoda oticum. They can be differentiated in the end of the 4th week. N.facialis is ventrally on the upper surface of the 2nd branchial arch. The motoric part of n.VII evolves separately

from neuroblasts in the upper part of rhombencephalon in the vicinity of pons Varoli (like n.VI) which can clarify damage of both nerves in Moebius's syndrome. Sensoric n.intermedius comes from ganglion geniculate from the 7th week. Abnormalities often afflict angle in the 2nd knee of the nerve during its course in the middle ear wall (60 grades compared to normal 120 grades), which moves the nerve between round and oval window.

Nervus VIII. Ganglion acusticofaciale n.VIII has a common base with ggl.geniculi n.VII, later separated. Ggl.n.VIII is placed to the medial part of the auditory pouch wall and divides itself into 2 parts: upper for ggl.vestibulare, lower for ggl.cochleare.

Inner Ear

The inner ear evolves between the 3rd and 4th week on the lateral part of head as an ectodermal thickening called acoustic placode. This structure gets deeper and forms a socket, its opening closes and forms a so-called otocyst coated with ectoderm and surrounded by mesenchyme. The surrounding mesenchyme starts to form a cartilaginous capsule of otocyst, which ossifies around the 17th week. During the 5th week the otocyst is differentiated by 3 plicae into dorsal vestibular part (utricle, semicircular ducts and ductus endolymphaticus) and ventral cochlear part (sacculus and ductus cochlearis). The membranous labyrinth is formed during the 6th month. The otocyst probably forms the cells of n.VIII. Ggl.n.VIII is divided into an upper (for utricle, lateral and superior semicircular canals) and lower part (for sacculus, posterior semicircular canal and cochlea).

Clinical Anatomy of the Ear

The ear is a peripheral auditory and balance analyzer. It consists of three major parts: the external ear, the middle ear and the inner ear. The external ear includes an auricle, the external auditory canal and the tympanum. The middle ear includes pneumatic system of the mastoid process, which is connected via aditus ad antrum with cavum tympani. The Eustachian tube connects the cavum tympani with the cavity of the middle ear. The inner ear is composed of bone and membranous labyrinth.

Auricle

The auricle is formed from elastic cartilage covered with skin. The perichondrium tightly adheres to the skin at the ventral side. The perichondrium and skin are dorsally connected with a thin layer of connective tissue. The earlobe is formed from fat tissue. The angle between the auricle and skull should not be more than 15 degrees. The following structures can be differentiated on the auricle: the helix, the anthelix and between them the fold (scapha).

The anthelix is ventrally divided into crura anthelicia; between them is located fossa triangularis. Concha auricular is divided by crus helices into cymba conchae and cavum

conchae. The entrance to the external auditory canal is surrounded by tragus and antitragus, they are divided by incisura intertragica. Incisura heliotragica is located between the helix and the tragus – a place of incision in the endaural approach to the middle ear. The form of the external ear allows sound concentration from outside into the auditory canal. The auricle is innervated from n.V, VII, IX and from the 2nd and 3rd cervical nerve (via n.auricularis magnus and n.occipitalis minor)

External Auditory Canal (Meatus Acusticus Externus)

The external auditory canal starts in the auricle, goes through the temporal bone and is ended by the eardrum. The outer third of the auditory canal is formed by the skin, which is connected with cartilage by a thin layer of fibrous tissue. The skin in this area contains hair follicles and glandulae ceruminosae et sebaceae. Gl.ceruminosae are modified apocrine glands producing secretion, which gets brown on air. This secretion together with sebaceous glands secretion forms the cerumen. It contains triglycerides and fat esters as well. Medial two thirds of the auditory canal are formed by a bone with skin of canal. The auditory canal is innervated by n.VII and IX.

Eardrum (Membrana Tympani)

The eardrum in adults is 9x10 mm in size.

- **Pars tensa** is formed by 4 layers. From the external auditory canal towards the middle ear: epithelium, radial and longitudinal fibrous layer and mucosa. The eardrum is connected with the auditory canal wall by the fibrous annulus fibrocartilagineus.
- **Pars flaccida** (Schrapnell's membrane) is located above the malleolar prominence between the stria malleolaris anterior and posterior. Pars flaccida is thinner than pars tensa; it does not have a longitudinal fibrous layer.

Cavum Tympani

- **Epitympanum** (atticus) is an area between the upper part of the eardrum and the tegmen tympani of the temporal bone.
- **Hypotympanum** is circumscribed by the lower edge of the eardrum and the base of the eardrum cavity
- **Mesotympanum** is the part of the tympanic cavity medially from the eardrum
- **Protympanum** lies ventrally from the eardrum and there is the ET orifice
- **Prussak's space** is circumscribed by pars flaccid, ligamentum mallei lateralis, the neck of the malleus and dorsally is opened to the epitympanum. It is the most frequent place of retraction pocket and cholesteatoma formation.

ET and the ventral part of the middle ear cavity are lined with columnar epithelium with cilia, which merge into ciliary epithelium of the nasal cavity and the nasopharynx. Towards the mastoid process we can see that epithelium gets low and flat and the number of cilia decreases. The dorsal part of the middle ear cavity is lined with one-layer cubic epithelium and in mastoideal pneumatisation we find flat epithelium without cilia.

Middle Ear Ossicles, Muscles and Nerves

Middle ear ossicles connect the eardrum and the oval window.

- **Malleus:** it is connected with the medial surface of the eardrum. Processus brevis and the inferior part of the manubrium (umbo membranae tympani) are tightly connected with the fibrous layer of the eardrum. The middle part of the manubrium is connected with the mucosal layer – stria mallearis. The neck and head are located in epitympanum. Manubrium mallei is connected with the incus.
- **Incus:** processus brevis goes dorsally, processus longus is connected with the stapes by processus lenticularis
- **Stapes:** the head of stapes is connected with the plate by two branches, anterior and posterior. The plate is held in the oval window by ligamentum annulare stapedis. The plate is about 1.5-3 mm in width.
- **M.tensor tympani** is located in the temporal bone in parallel with ET. After entering the middle ear its tendon goes towards the processus cochleariformis, is turned around it and is connected to the medial surface of the neck and the head of the malleus
- **M.stapedius** leaves eminentia pyramidalis on the dorsal wall of the middle ear cavity. A tendon is connected to the head and the posterior branch of the stapes.
- **N.VII (n.facialis)** goes out of pons and enters the upper frontal part of the internal auditory canal, where it is joined together with n.intermedius, which brings the olfactory fibers. It goes through the **Fallopí's canal**, comes out from the temporal bone in foramen stylomastoideum and goes along the dorsal part of m.digastricus. Its main stem enters glandula parotis, where it is divided into 6 branches for mimic muscles

In temporal bone are derived from n.VII:

- **N.petrosus maior** brings parasympathetic fibers to ggl.pterygopalatinum and innerves lacrimal glands and glands in mucosa of the palate and the nasal cavity. It goes towards canalis pterygoidei, where it is joined with sympathetic n.petrosus profundus and forms canalis pterygoidei
- **N.stapedius** innerves motorically m.stapedius
- **Chorda tympani** goes in the middle ear between the manubrium mallei and processus longus incudis. It goes out in fissure petrotympanica. It connects with n.lingualis and leads parasympathetic fibers into ggl.submandibulare (for submandibular

and sublingual salivary glands) and sensoric olfactory fibers (for anterior 2/3 of tongue) Recessus facialis is V-shaped area between chorda tympani and n.VII

- **Arnold's nerve (r.auricularis n.X)** is a branch of n.X, which contains also fibers from n.IX and goes above the vault of jugular bulb. The upper fibers are connected with n.facialis, lower ones lead sensitive information from the posterior surface of the external auditory canal. Arnold's reflex can be induced by irritation of the posterior wall of the external auditory canal.
- **N. tympanicus (Jacobson's nerve)** is a branch of n.IX. It forms the plexus in the middle ear. N.petrosus minor goes out of this plexus. It goes from the brain cavity through fissura petrotympanica and leads parasympathetic fibers into ggl.oticum. From this ganglion is parasympathetically innervated a specific salivary gland by n.auriculotemporalis.

Eustachian Tube (ET)

ET is a connection between the middle ear and the nasopharynx. ET has 3 parts – cartilaginous, junction and bony. The anteromedial part is cartilaginous and opens into the nasopharynx in the area of torus tubarius. The posterolateral part is bony and opens into the middle ear. The junction is located between the two previous parts and is called isthmus. Dilatation of the cartilaginous part is maintained in children only by m.tensor veli palatine with its medial fibers (m.dilator tubae), which is innervated from n.V (trigeminus). Children with malfunction of this muscle (e.g. with palatal fissures) have a dysfunction of ET. Adults have in addition m.levator veli palatine.

ET is rapidly lengthening in early childhood and in 7 years old children it has the same length as in adults. Neonates have a shorter ET by half in comparison with adults (18 mm : 35 mm). A short ET can be the cause of protective function disorder.

In young children, ET is horizontally placed with a maximum deviation of 10°. In adults, this deviation is 45°.

Physiological Functions of Middle Ear and Eustachian Tube

Middle ear functions

- The sound is transmitted with the help of the ossicles from the 52mm eardrum surface to the 3mm surface of the oval window. Sound energy is amplified this way to 17:1. In addition, manubrium mallei is 1.3 times larger than the long process of the incus, so another 1.3 times amplification of sound energy occurs. The total amplification is 22:1 which means 25 dB.
- There is a diffuse gradient between the atmospheric pressure and the mucosal circulation. Middle ear mucosa can continuously absorb gases, which permanently lower the middle ear pressure.

ET functions

- **Ventilation** function of ET is maintained by periodic opening of ET. It helps in pressure equalization between the middle ear and the nasopharynx (middle ear and atmospheric pressure)
- **Drainage** function of ET is maintained by the ciliary epithelium in ET and the anterior part of the middle ear (mucociliary clearance) and by the muscle functions (muscular clearance). Ciliary cells drain secretions into the nasopharynx, formed by calyciformis cells or formed during inflammations.
- **Protective** function of ET is maintained by the shape and the course of ET, muscle activity of the soft palate, immunologic and mucociliary function of the mucosa. It lowers the risk of infection penetration into the middle ear during inflammations of the respiratory airways.

ET function disorders

- Closed ET – opens less frequently than usually (in average once in 2 minutes in healthy people) according to the pressure gradient between the nasopharynx and the middle ear
- Open ET (patulous, semipatulous) – opens more frequently than usually or is permanently patent

Pneumatic System of Temporal Bone

The mastoid process is connected with the middle ear through aditus ad antrum. In adults, extensive pneumatisation occurs inside. Only a small central cavity is developed after the birth – antrum mastoideum, which is the base for other cavities. The development and the extent of the mastoid process pneumatisation depend on genetic factors and on the number and courses of middle ear inflammations. Children have a small mastoid process so pneumatisation is low. This small air volume can easily cause the development of low pressure in the middle ear in children. Pneumatization of most parts of temporal bone is complete between the 5th and 10th year.

Bone labyrinth

Bone labyrinth includes bone structures of pars petrosa ossis temporalis (vestibulum, cochlea and semicircular canals)

- **Bone cochlea:** it lies in the anterior part of the bone labyrinth and is formed by a spiral bone canal (ductus cochlearis), which whips around 5mm long bone axis (modiolus). Modiolus goes meatusocaudally from the anterior part of the upper wall of the inner auditory canal. Cochlea has 2.5 screws and is 31-33 mm long. Basal cochlear screw looks in the middle ear as a promontorium. The bone plate (lamina spiralis ossea)

goes from modiolus lengthwise cochlea, on it there are connected basillary and Reisner's membranes. Bone cochlea is divided by these into 3 spaces (scala tympani, scala media, and scala vestibuli). Between bone and membranous cochlea there is perilymph of similar composition as a cerebrospinal fluid (with high sodium and low kalium). Inside the membranous labyrinth there is endolymph, which has high kalium and low sodium (as a intracellular fluid)

Membranous cochlea

- It is located inside the bone cochlea

Basillary membrane: it is bound to lamina spiralis ossea and divides scala media and scala vestibuli. Externally located scala vestibuli connects with the oval window. Corti's organ is located on the basillary membrane in scala media

Reisner's membrane: it is bound to lamina spiralis ossea and divides scala media and scala tympani. Scala tympani is connected with the round window. Scala tympani and vestibuli are connected with helicotrema (scala communis) on the apex.

Membrana tectoria: it goes from ligamentum spirale, which is bound to lamina spiralis ossea.

The sound is transmitted from the stapedial plate via the oval window into the perilymph in scala vestibuli. Waves in the perilymph cause the irritation of frequency specific parts of the basillary membrane (lower frequencies on the base, higher on the apex)

- **Utriculus and sacculus:** these are vesicles containing neuroepithelial receptor cells in the macula sacculi et utriculi. These areas of sensoric epithelium produce gelatinous substance forming the otolith membrane. This substance contains crystals of calcium carbonate (otoconia). The macula sacculi is located in the ventral plane on the medial wall of the sacculus, macula utriculi is located on the anterolateral wall of the utriculi perpendicularly to the macula sacculi. Receptors in maculi contain ciliary cells, their cilia go into the otolith membrane with otoconia. Ciliary cells are surrounded by the auxiliary cells. The utriculus is ovoid and is sensitive to linear acceleration. The sacculus is smaller than the utriculus, is spherical and is connected with the cochlea via the ductus reuniens.
- **Semicircular canals:** there are three – upper, lateral and posterior. They have neuroepithelial cells in the ampullar endings connected with the utriculus. Upper and posterior canals are on the other ends connected into the common opening, which is located in the middle part of the utriculus. Receptor cells in ampullar endings are formed by ciliary cells. Their cilia are plunged in a gelatinous substance from polysaccharides and keratin. Semicircular canals are sensitive to angular acceleration.
- **Ductus and saccus endolymphaticus:** their main function is endolymph absorption and pressure equalization between the cerebrospinal fluid and the endolymphatic system.

Examination Methods of the Ear

They include sight (including otoscopy) and palpation, imaging methods and functional examinations. Hearing examinations are divided according to patient's compliance into subjective (patient responds during examination) or objective (patient's response is not needed; information is obtained by the doctor or a machine).

Examination of Eustachian Tube Function

- **Tympanometry**
- **Valsalv's maneuver:** expiration against the closed nasal entrance increases pressure in the nasopharynx and causes opening of ET and pressure increase in the middle ear
- **Politzeration:** pressure increase in the nasal cavity and the nasopharynx with the help of a ball inserted into one nostril with the concurrent closure of the other nostril. It leads to ET opening and pressure increase in the middle ear. At the same time, the patient utters syllables, which causes a velopharyngeal closure (in Czech: kuku, káva). The opening of ET and air passing into the middle ear cause murmur, which can be registered by the otophone (tube connecting the ear of the examined person and the doctor). The patient indicates a change in hearing
- **Catheterization:** with the help of metal catheters, ET opening can be sounded and with a ball we can increase pressure in the middle ear (similarly to politzeration). Examination is unilateral and anaesthesia is required (local or general).
- **Toynbee test:** swallowing in closed nasal entrance and mouth leads at first to positive pressure in the nose and the nasopharynx (1st phase), after that to pressure decrease (2nd phase). In the 1st phase the air can flow into the middle ear and cause overpressure. During the 2nd phase there is underpressure or overpressure in the middle ear persisted from 1st phase. In case of ET malfunction it is not opened and pressure in the middle ear stays the same. If eardrum is not damaged, test can be evaluated by tympanometry. If pressure in the middle ear decreases, the function of ET is probably normal. If pressure does not decrease it does not mean malfunction of ET, but another test must be performed.
- **Experimental examinations of ET function:** with X-ray contrast fluids we can examine the protective and drainage function of ET. A contrast fluid is applied into the nasopharynx and we observe its retrograde movement into ET. Protective function of ET is normal if contrast fluid cannot get into the bone part of ET during swallowing. Drainage function can be examined by contrast fluid application into the middle ear cavity and observation of its movement into the nasopharynx. We can use scintigraphy or microendoscopy as well. Sonotubometry is usable in research, however, it is not accessible for clinical usage.

Otoscopy

Examination of eardrum by sight: we use ear mirror (speculum), microscope or otoscope. During otoscopy it is essential to straighten the auditory canal, which has often a sigmoid shape. We do so by pulling the auricle dorsally and externally in adults or cranially and externally in children. The patient's position depends on their age and compliance.

Pneumotoscopy: we use otoscope connected with a ball, which can be used for pressure changes in the auditory canal. If we tightly occlude auditory canal we can observe mobility of eardrum with pressure changes.

Evaluation of otoscopic findings

We see Bezold's trias on a normal eardrum (prominentia mallearis, stria mallearis and light reflex). Prominentia mallearis is formed by processus brevis mallei, stria mallearis is formed by manubrium mallei, its end is tightly connected with eardrum and forms umbo membranae tympani. Light reflex comes up from light reflection, has a triangle shape with apex in the umbo and the base in the annulus. The eardrum can be divided by 2 imaginary axes (one goes through stria mallearis) into 4 quadrants: upper anterior, upper posterior, lower anterior, lower posterior.

- **Location of eardrum:** a normal eardrum should be in a neutral position with slightly protruding processus brevis mallei.
- **1. Retraction of eardrum** is usually a sign of underpressure or secretion in the middle ear or both – malleal prominence expressively protrudes and manubrium mallei looks like shortened and located more horizontally. If we evaluate the eardrum retraction in pars tensa we use a classification according to Sade. Pars flaccida retraction is classified according to Tose. Atrophic areas on the eardrum can form the so-called retraction pockets. They are classified according to Charachon.
- **2. Eardrum bulge** is noticeable mostly in its most supple parts – in the epitympanum and the upper posterior quadrant. Processus brevis mallei is indistinct. Eardrum bulge is caused by an increased pressure in the middle ear or by secretion or expansion.
- **Eardrum color** is grey. Yellowish or bluish color can be caused by secretion in the middle ear. Violet eardrum usually signals the presence of blood in the middle ear (hemotympanum).
- **Eardrum mobility** is evaluated by a pneumotoscopic examination or by tympanometry. Normal eardrum and ossicles move according to pressure changes in the external auditory canal. Mobility is influenced mostly by secretion, negative pressure in the middle ear or by fixation of the ossicles. Increased mobility is found in an atrophic eardrum or if the ossicle string is disrupted. Decreased mobility can be found in a thickened eardrum, myringosclerosis, otosclerosis and in children during the chronic secretory otitis.

Subjective Hearing Examination

Conversation with patient

Valuable information from the patient can be gained during history taking:

- Patient does not understand even loud speech with reading the lips
- Patient understands loud speech with reading the lips
- Patient understands loud speech without reading the lips
- Patient understands silent speech without reading the lips

We can also notice pronunciation defects (in higher frequencies defects the pronunciation of sibilants is slurred), change in speech melody (more serious hearing defects) or head turning (asymmetric affliction). Especially in children, hearing loss can cause uncertainty and fear because they do not understand what happens around them.

Classical hearing test

It is a basic hearing test. It is made by a loud or whispered voice (vox, vox sibilans – abbreviations V, Vs). The patient is turned with the examined ear to the doctor and with the face to the assistant who obstructs the other auditory canal in whispered voice or din ear (for the deafening, insertion of olive or Barany's device is necessary) in loud voice. According to the distance we make a description, e.g.:

- 6m Vs 3m – patient hears whispered speech on the right side from 6m and on the left side from 3m

The result is orientation and depends on the patient compliance, doctor and assistant experiences and the quality of the given room (silence and sufficient length). Differences between loud and whispered tests are now obsolete.

- The loud part is worse understood in case of lower frequencies affliction because loud speech has the majority of acoustic energy formed by vocals, which have significant formant structure with energy maximum between 100-1000 Hz.
- The whispered part is worse understood in case of higher frequencies affliction because whispered speech has the majority of acoustic energy formed by consonants, which have energy maximum between 2000-8000 Hz.

The sense of hearing test lies in the examination of the central hearing component. The more central defect is, the more the hearing is impaired and the less worsened the tone detection is. A typical example is aphasia when tone audiometry is normal but understanding is severely afflicted. On the other hand, understanding in a light and moderate conductive hearing loss is relatively good.

Tuning fork examinations

This examination was performed in the past by a set of tuning forks of different frequencies. At present, it is a special test with a restricted number of tuning forks (usually one). Their sense diminishes but even now they are a good guide before tone audiometry and are valuable in the understanding of diagnostics theory of conductive and perceptive hearing loss.

Rinne's test: this test informs us if hearing is better via air (the fork placed in front of the opening of the auditory canal produces sound which is transmitted through the auditory canal, eardrum, ossicles into the inner ear) or via bone (the fork placed on processus mastoideus vibrates with the whole skull and os petrosus where the membranous labyrinth is located and where the inner ear is stimulated). The energy needed for skull vibration and the sense of tone is about 40 dB bigger than in a healthy ear. Practical performance: vibrating fork is placed in front of the auricle and the patient tells us when he stops hearing it. Immediately after that the fork is placed on processus mastoideus and the patient tells us if he hears it or not. If not, hearing system is intact or perceptive hearing loss is present. If so, we reverse the course and if patient hears better through processus mastoideus, conductive hearing loss is present (but we cannot exclude a mixed hearing loss).

Weber's test: we place the fork on top of the head or on the forehead. The patient tells us in which ear he hears better.

- Conductive hearing loss – in the afflicted ear
- Perceptive hearing loss – in the healthy ear
- Mixed hearing loss – depends on both components

Results improvement can be reached by calibrated Weber's test when stimulus is led to the vibrator on the forehead from the audiometer. It allows examination of different frequencies and intensities.

Schwabach's test: it is not usually done, because it compares the hearing of the patient and the doctor

There are some specialized fork tests (Gelle, Cytovič, Frederici). However, they are suitable for only some diagnoses and are so complicated that we do not use them in children.

Tone audiometry

It is an examination of clear tones hearing. It is made by the audiologic nurse or the doctor with the help of audiometer, a machine with the generator of those tones. Sound is led in air (headphone, speakers) or in bone (bone vibrator placed on processus mastoideus – see Rinne's test). The examination is made in a silent room or better in an audiochamber (special, noise eliminating room). The patient responds to presented tones:

- Classic tone audiometry – pressing the button (from school children to adults)
- Classic tone audiometry in children – raising the hand (pre-school children)
- Tone audiometry with a game – building tower from cubes, etc., usually with speakers (headphones are felt negatively in children)
- Behavioral audiometry – non-specific reactions as a blink, activity interruption, turning round on noise (age of 6-24 months)

- Audiometry with a visual amplification – on the basis of conditional reaction, a child turns round on noise with a supposed award (e.g. a doll with blinking eyes), age of 6-24 months.

The result is a graph – tone audiogram:

- Axis X means frequencies of tones in Hz (Hertz) – 125, 250, 500, (750), 1000, (1500), 2000, (3000), 4000, (6000), 8000 – in brackets are optional frequencies, most important frequencies are 500, 1000 and 2000 Hz.
- Axis Y means intensities of the presented sound (in dB – decibel)
- Particular points determine the threshold of hearing – tone sense on certain frequency induced by as low as possible intensity. It is marked by:
 - Air conduction – circle and cross, frequencies are connected with a line
 - Bone conduction – brackets, frequencies are connected with a dash line

There are other tests used for a special audiologic test or for a hearing-aid devices application – the threshold for unpleasant listening, pain threshold, etc.)

Verbal audiometry

This is a group of examinations where the patient repeats words which are played in a variable intensity. It is the analogy of a classical hearing test made in a silent room or an audiochamber and stimuli intensities are calibrated. The source can be a microphone or a record on e.g. CD. Typical is a presentation of 10 words of the same intensity. From number of correct answers, a percentage of intelligibility is calculated. Basic described values are 50% intelligibility – threshold and the lowest value where the patient understands the most (usually 100%).

Presentation possibilities are the same as in tone audiometry:

- From speakers (so called free field)
- Into headphones
- Into bone vibrator (bilateral stimulation of the inner ear)

Meaning of verbal audiometry:

- Evaluation on hearing-aid devices effect
- Hearing examination of pre-school children (word repetition or showing them on pictures is for children simpler than signalization of tone detection)
- Examination of speech intelligibility – in case of normal hearing according to tone audiometry with the help of special verbal groups

Objective Audiometry

Tympanometry

Tympanometry is an objective examination method evaluating changes of pressure in the external auditory canal on the basis of sound reflection from the eardrum back to the tympanometer.

Tympanometer is a device which emits sound waves towards the eardrum, receives them and processes reflected waves and pressure changes in the external auditory canal. If pressures on both sides of the eardrum are the same, the maximum of sound energy goes into the inner ear (compliance (softness) of eardrum and ossicles is highest). The bigger the difference is between both sides of the eardrum, the more the compliance is decreased and the more the admittance (rigidity) is increased. The waves are reflected back to the tympanometer so that the tympanometer records several types of curves.

Tympanometric curves are usually evaluated by the classification according to Jerger: basic curves are A, B, C.

- **Curve A:** is physiological with peak in zero pressure. Peak means value of actual pressure in the middle ear (pressures on both sides of the eardrum are same). Curve As (peak is lower than 0.3ml on Y axis) is connected with increased rigidity of conductive system (e.g. otosclerosis). Curve Ad (has peak more than 1,2 ml above on Y axis) is connected with increased mobility of conductive system (eardrum atrophy, ossicle string disruption)
- **Curve C** means disorder of ET ventilation function. Peak on this curve is moved into the negative pressure values (C1 from -100 to -200 daPa, C2 from -200 daPa)
- **Curve B** has no peak. It means that eardrum reflects during different pressures the same amount of sound. It is caused by increased rigidity of eardrum-middle ear system, usually caused by secretion in the middle ear behind the intact eardrum (OMCHS – otitis media chronica secretorica)

Positive admittance can be found in amblyotia in acute otitis media; however, in case of this disease tympanometry is not performed because of pain.

The majority of tympanometers can detect actual volume where pressure change occurs (external auditory canal, possibly middle ear cavity) and reveal a small invisible eardrum perforation.

Modern tympanometers can examine even acoustic reflexes of the middle ear muscles (m.stapedius – innervation from n.VII and m.tensor tympani – n.V). These muscles act as protectors of auditory apparatus against loud noise. If function of outer, middle and inner ear is intact, stapedial reflex can be noticed in intensities over 80 dB, reflex of m.tensor tympani in intensities over 100 dB. Reflexes are bilateral during unilateral stimulation. If reflexes are noticed in hearing loss, it means that disorder is located behind reflex arches. If stapedial reflex is noticed on levels up to 60 dB above threshold we consider it a proof of recruitment. Decay reflex examines an increased fatigue of hearing. If values of stapedial reflex decrease more than by 50% in 10 seconds, it means an increased fatigue of hearing.

Otoacoustic emissions (OAE)

Sound produced by cochlea was described and measured by Kemp in 1978.

Otoacoustic emissions are generated as a nonlinear by-product of cochlear biomechanic activity on the level of external ciliary cells. They are produced only preneurally and do not show any ability to transmit sound. OAE examination is fast, non-invasive and objective.

There are 2 categories of OAE: spontaneous OAE (SOAE) or evoked OAE (EOAE). EOAE are important in clinical practice. They allow the assessment of the external ciliary cells function which generates the emission by mechanic touch as a response to sound presence. OAE are not noticed if hearing loss is greater than 30 dB.

- Use: hearing screening in neonates, stimulation examination, examination in ototoxic therapy, perceptive hearing loss

Examination of evoked potentials

Examinations: ERA (electrical response audiometry) or AEP (auditory evoked potentials) are based on average EEG record. We evaluate electric potentials with different latency:

- Short – electrocochleography, BAEP (or BERA) up to 15 msec
- Middle – MERA, SSEP, VEMP – 15-100 msec
- Long – LAEP (or CERA), P300, MMN – more than 100 msec

For a threshold determination, we evaluate the presence or latency of single waves. For auditory track description we describe latency, intervals and differences between sides.

- BAEP, **BERA** (brainstem AEP or ERA) evaluate potentials from brainstem. We describe 7 waves, the most constant is 5.wave, which originate from colliculus inferior, or waves 1 and 3.
- **Electrocochleography** evaluates potentials from cochlea – record is optimized maximally to the level of wave 1 of BAEP. The nearer to cochlea, the better the quality (best is transtympanic placement into round window)
- MERA (middle latency ERA) – classical examination of averaging is considerably afflicted by muscle artefacts (see VEMP), in general anaesthesia it is practically unnoticeable. It is not used.
- SSEP (steady state evoked potentials) – hearing threshold is determined according to the signal analysis (very fast and accurate examination, but we cannot describe auditory track).
- VEMP (vestibular evoked myogenic potentials) – is based on the disadvantage of MERA. Muscle artifact is generated on the basis of vestibule-spinal reflex during sacculus stimulation by great intensity (1000 dB nHL, 120 dB SPL). It is used in diagnostics of balance disorders.

- LAEP, **CERA** (long latency AEP, cortical ERA) evaluate potentials from auditory cortex (complex of waves P1N1P2). This examination is used especially in case of simulation, dissimulation, aggravation, because stimuli are the same as in tone audiometry and allow objective examination till the end of auditory track. Waves P300 and MMN are used in cognitive testing which allow examination of processing auditory signal by auditory track and cortex (significant in examination of intelligibility).

Examination of Vestibular Apparatus

History – essential is the description of circumstances, time, duration and character of problems development (balance disorders, nausea, vomitus, pressure in ear, tinnitus, hearing loss, collapse, headache).

Examination of vestibule-ocular reflexes

Examination of eye movements

Examination of spontaneous nystagmus:

- Barthels's glasses
- Frenzel's glasses
- Videooculography
- Electronystagmography

Examination of semi-spontaneous nystagmus:

- Position tests (slow position changes of body and head) – influence especially on utricle and saccule
- Positioning tests (fast position changes of body and head) – influence especially on semicircular canals
- Torsion test – diagnostics of vascularization defect of vertebral arteries
- Head shaking test

Examination of provoked nystagmus:

- Temperature tests (with warm and cold water or air)
- Rotating and pendulum provocation

Examination of vestibule-spinal reflexes:

Orientation neurologic examination (everything with exclusion of visual fixation):

- Hautant – with arms raised forward and watching the deviation

- Romberg - standing (in variable stances) and watching the traction or fall
- Target pursue – point at nose with index finger
- Examination of adiadochokinesis (synchronic turning of hands)
- Walking on straight line
- Unterberg-Fucuda – 1 minute march on place, pathologic is more than 45 degrees deviation

VEMP – stimulation of sacculus with a high intensity sound and registration of tonic responses of neck muscles

Posturography, stabilometry, craniocorpography – examinations of static or dynamic balance maintenance with the help of objective methods

Imaging Methods

Classical X-ray images are replaced by CT. However, Stenvers's projection is suitable for patients with cochlear implant because metal parts of implants can cause artifact during CT and make evaluation of CT difficult.

For a detailed structure of the temporal bone, HRCT (high resolution CT) is most suitable in the coronary and axial projection with scans 1-1,5 mm thin in bone algorithm.

For examination of the inner auditory canal, the brainstem and cerebellum area are suitable for magnetic resonance imaging – MRI. In comparison to CT, MRI can reach a better resolution of soft tissues and does not irradiate the patient. Disadvantages are: long examination time with the necessity of anaesthesia in small children and non-cooperative persons and a higher price.

Congenital Defects of Ear

Apostasis Auriculi

Congenital defect of anthelix is usually the cause of auricle displacement.

Therapy: plastic correction. The ideal age for surgery is before the school age (6 years)

Microtia et Atresia Meati Acustici Externi

Congenital defect of auricle development (microtia) or missing auricle (anotia) is often combined with the congenital defect of the external auditory canal (stenosis, atresia). The incidence of other congenital defects is increased (defects of the middle or inner ear). Auditory canal stenosis means that it is narrower than 4 mm.

Diagnostics: CT, objective hearing examination (exclusion of congenital defect of the middle and inner ear or the auditory track)

Therapy: it depends on examination results and on hearing affliction extent (unilateral or bilateral affliction): hearing-aid devices, cochlear implant, tympanoplasty, plastic of external auditory canal or auricle

Fistula Auris Congenital

During the development of the auricle, fistulae and cysts can develop in its surrounding. Most frequently we can find a preauricular fistula with an external opening placed between the tragus and the inner opening between cartilaginous and bone part of the external auditory canal. The most frequent complication is inflammation which causes secretion or sometimes swelling and erythema around the fistula.

Therapy: ATB in case of inflammation, incision in case of abscess. Extirpation of fistula in a still state with sparing n.VII

Genetic Defects of Hearing

Genetic hearing defects can be congenital or gained, conductive or perceptive (SNHL – sensorineural hearing loss) or mixed, stabile, fluctuating or progressing, unilateral or bilateral, symmetric or asymmetric, syndromic (over 400 syndromes) or non-syndromic. They are characterized by audiology, age, progression and type of heritability (80% are autosomally recessive, 18% are autosomally dominant, 2% are bound to chromosomes)

Diagnostics: CT, objective audiometry

Structural anomalies of inner ear

20% of children with SNHL have CT anomalies of inner ear:

- **Michel's type:** pyramid agenesis, outer and middle ear is usually normal. It can be confused with ossifying labyrinthitis. Heritability can be autosomally dominant or recessive. Changes develop during the 3rd week of development.
- **Mondini's type:** develops in 6th week of development. There are presented: 1 screw of cochlea, dilated ductus and saccus endolymphaticus, communication between scala tympani and vestibule. It is found in the following syndromes: Treacher-Collins, Pendred, Waardenburg, Wilderwanck, Branchio-oto-renal, and in CMV infection. Affliction is unilateral or bilateral.
- **Scheibe's type:** wrong differentiation of Corti's organ, we find malformation of tectorial membrane and Reisner's membrane collapse. It is the most frequent type of affliction, found in syndromes such as: Jervell, Lange Nielsen, Usher, Waardenburg
- **Alexander's type:** affliction of Corti's organ in the basal screw and dilated aquaeductus vestibule – typical bilateral affliction, cause perceptive hearing loss with fluctuating or progressing course, dizziness and balance disorders. It is often found in Pendred's syndrome.

- **Malformations of semicircular canals:** most frequent are malformations of lateral canal

Autosomally recessive diseases

- **Pendred's syndrome:** progressive SNHL, goiter as a result of iodine metabolism disorder
- **Jervell's and Lange-Nielsen's syndrome:** SNHL, lengthened QT interval on ECG
- **Non-syndromic hearing defects:** at least 20 loci were identified. Classification according to Königsmark and Gorlin:
 - Congenital severe hearing loss
 - Middle hearing loss
 - Impairment with early onset

Autosomally dominant diseases

- **Waardenburg's syndrome:** unilateral or bilateral SNHL (20% in type I, 50% in type II)
- Type II (mutation of MITF gene): pigment anomalies (white spots, heterochromia iridis, vitiligo).
- Type I (mutation of PAX3 gene): same as type I + dystopia canthorum
- **Stickler's syndrome:** small jaws with palatoschisis (Pierre-Robin sy), myopia, cataract, hypermobility and enlargement of joints, arthritis in early adults, mixed or SNHL (80%), gene mutation COL2A1 on chromosome 12
- **Branchio-oto-renal syndrome (Melnick-Fraser):** ear appendages, neck fistulae, renal anomalies, hearing loss of different type
- **Treacher-collins syndrome:** Microtia and atresia of the auditory canal, SNHL and CHL, hypoplasia of mandible, coloboma of lower eyelids, antimongoloid position of eyes
- **Non-syndromic hearing defects:** progressive hearing loss – variable age of onset, affliction of inner ear development, disorders on many different frequencies

Diseases bound to chromosome X

- **Norrie's syndrome:** SNHL, congenital or progressive blindness
- **Wilderwack's syndrome:** SNHL or mixed hearing loss + fusion of neck spondyles (Klippel-Feil sy), paresis of n.VI

- **Alport's syndrome:** progressive SNHL, affliction of kidneys
- **Oto-palato-digital syndrome:** ossicular anomalies, hypertelorism, palatoschisis, small nose, fingers anomalies
- **Turner's syndrome (X0):** SNHL or mixed hearing loss, gonadal dysgenesis, small stature, short neck. Multifactorial affliction.
- **Goldenhar's syndrome:** preauricular appendages, spine anomalies, epibulbar dermoid, coloboma of lower eyelid

Chromosomal diseases

Redundant or missing chromosomes can cause variable congenital defects.

- Down's syndrome: trisomia of chromosome 21
- Patau's syndrome: trisomia of chromosome 13
- Edward's syndrome: trisomia of chromosome 18

Trisomia of another autosome is lethal.

Congenital Defects of Auditory Nerve

Auditory neuropathy

Pathology in the area of synapse of inner ciliary cells and cells of the auditory nerve

Diagnostics: objective hearing examination, MRI of brain

Therapy: cochlear or stem implantation in case of bilateral affliction. If it is not successful – sign language.

Aplasia or hypoplasia of auditory nerve

Diagnostics: objective hearing examination, MRI of brain

Therapy: in case of bilateral affliction stem implant with dubious effect, sign language

Demyelination disease

See Neurology

Diseases of the External Ear

Cerumen

It is produced by gl.ceruminosae of the external auditory canal. Yellow-brown matter with fat particles can partially or totally obstruct the external auditory canal. It dries and oxidates (getting hard and black). It can be the cause of the inflammation of the external auditory canal or even hearing loss or tinnitus. Cerumen can be preferably removed by lukewarm water (cave thermal irritation of labyrinth). If we are suspicious of eardrum perforation, we use boric acid lotion. Hard cerumen can be partially dissolved by oil preparations.

Erysipel

Streptococcal infectious disease of skin. It causes painful, circumscribed erythema.

Therapy: preferably PNC, wide spectrum ATB

Eczema of the External Auditory Canal

Course: itching, increased sensitivity of auditory canal, discharge

Classification:

- Dry: increased production of skin epithelium
- Wet: blisters formation, discharge
- Mixed: combination of previous

Etiology:

- Contact: usually result of cosmetic preparations (soap, shampoo) usage
- Bacterial: in case of otitis externa, otitis media chronic mesotympanalis, otitis media chronic cum cholesteatomatae or otitis media acuta
- Atopic
- Seborrhoeic

Therapy:

- Contact: change of cosmetics
- Bacterial: therapy of primary disease
- Atopic and seborrhoeic: dermatological treatment

Otitis Externa

Otitis externa diffusa

- **Definition:** inflammation of the skin and the submucous tissues of the external auditory canal
- **Pathogenesis:** usually result of bathing in contaminated or chlorinated water. It is a disease of summer months
- **Course:** pain, erythema and stricture of the external auditory canal. In severe cases: auditory canal impassability caused by soft tissues swelling, conductive hearing loss, increased body temperature, erythema and swelling in the periauricular area (dif.dg.acute mastoiditis: tympanometry, X-ray, CT).
- **Diagnostics:** see Pathogenesis and Course. Pain increases by pressure on tragus or by auricle pulling.
- **Therapy:** local ATB in unguent (chloramphenicol) or drops (Garasone, Otosporin), analgetics, in severe cases ATB generally
- **Complications:** inflammation spread through Santorini's fissures into the skull base (otitis externa maligna) or into glandula parotis. Increased risk is present in patients with immunity defects or diabetes mellitus.

Otitis externa circumscripta

- **Definiton:** affliction of small skin glands (gl.ceruminosae) or hair follicles of the external auditory canal.
- **Course:** painful erythema and swelling in the cartilaginous part of the external auditory canal (folliculitis, furuncle). There is discharge after perforation or incision.
- **Diagnostics:** see Course. Pain increases by pressure on tragus or by auricle pulling.
- **Therapy:** incision, local ATB, analgetics, in severe cases ATB generally

Perichondritis Auriculae

Cartilage inflammation develops usually after ear injury, or e.g. after insect bite. Without therapy, the cartilage suffers from insufficient nutrition from perichondrium which leads to the cartilage destruction with a permanent deformation of the auricle

Therapy: ATB generally, punction, incision, drainage, remove destroyed cartilage.

Acute Inflammations of the Middle Ear

Otitis media acuta (OMA)

Definition: mucous inflammation of the tympanal cavity and the pneumatic system of the temporal bone, followed by suddenly developed symptoms of acute infection.

Pathogenesis:

- Infection spreads most frequently from nasopharynx through Eustachian tube due to a pressure gradient between the middle ear cavity and the nasopharynx (ET closed) or by snuffling (ET opened).
- Hematogenous infections during airways infection (flu, scarlet fever, variola)
- Infections of the middle ear in case of tympanic perforation or another communication between the middle ear and the outside environment

Etiology:

- *Streptococcus pneumoniae* (up to 60%)
- *Haemophilus influenzae* (up to 25%)
- *Moraxella catarrhalis*
- *Staphylococcus aureus*

After vaccination against *H.influenzae* and *S.pneumoniae* we suppose changes in the percentage of specific pathogens.

Symptoms and diagnostics:

Initial stage:

- Subjective problems: airways infection, pricking or pain in the ear, worsened hearing, nausea, restlessness

- Objective findings:

otoscopy: accentuated veins, red tympanum, which is in its normal position – malleolar prominence is distinct (processus brevis mallei)

audiometry: normal or conductive hearing loss

tympanometry: curve C. In neonates, toddlers and young children it is necessary to differentiate congestion of the tympanum caused by other condition, e.g. by scream.

Stage of advanced otitis: in this stage, bacterial superinfection appears on primary viral inflammation. Increased pressure of secretion leads to an increased pain because of peripheral sensitive nerves irritation. Risk of spreading inflammation to the bone with complications is also increased. If ET is opened, partial drainage of secretion to the nasopharynx is possible.

- Subjective problems: pain in the ear, hearing loss, nausea, vomiting (Arnold's reflex in *r.auricularis n.vagi* is irritated). Small children are increasingly restless especially in horizontal position because of increased head and neck vascularization.
- Objective findings:

Otoscopy: bulge of the tympanum with maximum in dorsal upper quadrant (processus brevis malei cannot be differentiated)

Audiometry: conductive hearing loss; in case of normal hearing Weber's test contralaterally lateralize into the sick ear.

Tympanometry: it is not made because of pain.

Stage of advanced otitis with perforation: increased pressure of secretion often causes tympanal perforation and drainage from the middle ear cavity into the external auditory canal. Infected secretion can cause inflammation of the external auditory canal.

- Subjective problems: hearing loss
- Objective findings:

Otoscopy: secretion in the auditory canal, maceration of the tympanum

Audiometry and Weber's test: same as previous stage

Tympanometry: is impossible due to tympanal perforation

Therapy:

- Initial stage: therapy of airways infection, strip with boric acid put on the tympanum, possibly ear drops. Analgetics, antipyretics, antihistaminics, fluids, vitamins, cold linen behind ear, increased position of the head. Examination after some time by general practitioner. If problems persist, examination by ENT specialist is needed.
- Stage of advanced otitis: paracentesis is fully indicated in the dorsal lower quadrant. It lowers risk of complication and relieves from pain. After paracentesis pain diminishes and body temperature decreases to normal. Secretion from the ear stops in a week. Intensive local treatment is necessary after paracentesis. We recommend repeated rinsing of the external auditory canal with lukewarm boric acid lotion in accordance with the intensity of the secretion from the middle ear, in first 3 days at least five times a day. Antibiotics are commonly not needed. During more serious course or other indicated cases amoxicillin is the antibiotic of choice. We can administer cephalosporin as well, in case of allergy macrolids. A follow-up examination is planned after 5 days after paracentesis. Patients are told that if problems are the same or are even worse, they must visit the specialist sooner. Hearing examination (in patients without compliance only tympanometry) is planned after inflammation healing – commonly 14 days after the first visit. The next check-up is planned according to the results of audiometry or tympanometry - once per 1-2 months.
- Stage of advanced otitis with perforation: same as stage 2, but without need of paracentesis

Otitis media acuta recidivans (OMR)

- **Definition:** advanced acute middle ear inflammation (paracentesis, spontaneous tympanic perforation) three times in 6 months or four times in a year.
- **Course:** possibility of otitis media chronica secretorica in coincidence (between relapse of acute inflammation of the middle ear tympanometry B, eventually conductive hearing loss), or without otitis media with secretion (tympanometry A and normal hearing)
- **Therapy:** insertion of ventilation tube premeatus, relapse of acute inflammation of the middle ear in most patients. In case we are not successful, imaging methods are needed (CT, X-ray) followed by surgical intervention (antromastoidectomy). Antibiotic premeatousion is ultimum refugium after immunological examination. OMR is one of the indications to vaccination against S.pneumonie.

Myringitis acuta

Definition: acute viral inflammation localized on tympanum itself

Pathogenesis: hematogenous infection during airways inflammation

Etiology: respiratory viruses

Course: pain of the ear during infection

Diagnostics: otoscopy – vesicle on the tympanum

Therapy: paracentesis, therapy of airways infection

Chronic Inflammations of the Middle Ear

Otitis media chronica mesotympanalis

- **Definition:** the presence of eardrum perforation lasting at least 3 months with a repeated secretion from the middle ear cavity and conductive hearing loss.
- **Pathogenesis:** malfunction of the Eustachian tube, repeated middle ear injuries or eardrum injury. The influence of the Eustachian tube malfunction type is unknown. In literature it is suggested that patients with an open Eustachian tube have chronic mesotympanic otitis more frequently than patients with a closed Eustachian tube.
- **Etiology:** in bacteriological examination we can find E.coli, Proteus, Pseudomonas and others.

Course: repeated discharge from the middle ear cavity into the external auditory canal, often after water penetration into the middle ear. In case of a long lasting inflammation there is a possibility of mucosal polyps and granulations development.

- **Diagnostics:**

Otoscopy: eardrum perforation of a variable range, commonly of a kidney shape

Tympanometry: classical curves cannot be examined, we can make only an auxiliary function – Eustachian tube examination, volume measurement (this helps differential diagnostics of retraction and perforation).

Hearing examination: commonly light or moderate conductive hearing loss

- **Therapy:** local therapy for premeatousion of middle ear discharge, similarly as during acute inflammation of the middle ear, local ATB drops, or possibly ATB generally. Surgical therapy – removing of middle ear polyps and granulations. If patient is without middle ear discharge for 3 months and the inflammation is cured, we call it a dry perforation of the eardrum (perforation membranae tympani or residua post otitidem) and as a definitive solution of this state myringoplasty is indicated. After closing the middle ear cavity by myringoplasty, hearing is improved and life comfort of the patient is increased because of premeatousion of repeated discharge from the middle ear.

Otitis media chronic secretorica

- **Definition:** secretion presence behind the solid eardrum without signs of acute infection. If it last at least 3 months, we assess it as a chronic inflammation. The presence of a secretion up to 3 months indicates subacute secretoric otitis. The secretion can be purulent, mucous, serous or combined.
- **Pathogenesis:** the malfunction of the Eustachian tube or acute middle ear inflammations lead to structural and functional changes of the middle ear and the eardrum. In the middle ear mucosa there are glandular structures formed, which are not present in a normal healthy mucosa. The malfunction of the ciliated epithelium leads to malfunction of the ciliated transportation to the Eustachian tube. The result is an accumulation of the middle ear glands secretion or the rest of secretion after acute otitis media. We can see atrophic spots and calcifications on the eardrum. Risk of retractions is increased in weakened parts of the eardrum.'
- **Etiology:** in middle ear secretion there was found DNA of H.influenzae
- **Course:** the beginning is asymptomatic, later light or moderate conductive hearing loss is present. Tinnitus and recurrent acute otitis media are possible. Eardrum retraction in atrophic spots can lead to adhesions or cholesteatoma.
- **Diagnostics:**
- Otoscopy: at the beginning there can be normal otoscopic findings (grey, shiny and contoured eardrum). However, a change of the eardrum color by secretion is typical. The eardrum can be yellowish or bluish, in some cases we can see a level of secretion. Reflex is fragmented, shortened or disappeared, eardrum is matt. The concavity of the eardrum leads to even a more horizontal location of the stria mallearis. Retraction pockets are an indication to a surgical treatment.

- Tympanometry: curve B or C2
- Hearing examination: light or moderate conductive hearing loss

Therapy:

- Conservative: stimulation of palatal muscles (chewing gum), antihistaminics, local corticosteroids. Politzeration can be made only in patients without an upper respiratory infect.
- Surgical
 1. Improving the Eustachian tube function: a sufficient Eustachian tube function can lead to an improved mucociliated transport from the middle ear cavity: surgical revision of the nasopharynx and the nasal cavity, improving of nose patency (by removing lymphatic tissue in the nasopharynx in childhood. Lymphatic tissue can cause a compression of the nasopharyngeal entrance of the Eustachian tube or can be the source of recurrent infections of the nasopharynx). Especially in adult patients, it is necessary to exclude a nasopharyngeal tumor.
 2. Improving hearing: by removing a secretion from the middle ear cavity and decreasing its production (myringotomy with secretion aspiration from middle ear or insertion of ventilation tube – risk of retraction pockets development is lower and aeration of the middle ear cavity is ensured).

According to studies which assess the effect of ventilation tube insertion - long term results of OMS therapy are not statistically different in patients with or without the ventilation tube. However, benefits of the ventilation tube are better hearing during disease and less extensive surgical interventions on the temporal bone.

Otitis media chronica cum cholesteatomatae

Definition: the presence of spinocellular epithelium in improper localization (in the middle ear cavity in case of cholesteatoma)

Classification: according to development mechanism we divide cholesteatomas into congenital or gained. According to the localization we divide them into:

1. Tensa cholesteatoma
2. Flaccida cholesteatoma
3. Sinus cholesteatoma
4. Cholesteatoma with solid eardrum

Tensa cholesteatoma comes from the upper frontal quadrant of the eardrum, sinus cholesteatoma from the upper dorsal quadrant, flaccida cholesteatoma from the Schrapnell membrane. Cholesteatoma with a solid eardrum can be congenital, posttraumatic or iatrogenous.

Pathogenesis:

- **Retraction theory:** long lasting ET malfunction and underpressure in the middle ear lead to structural changes of middle ear mucosa and eardrum – glands formation, atrophy, and calcification. The result is retraction pockets formation due to pulling atrophic areas medially.

Sadé differentiated retraction pocket into 4 groups. Retractions in pars flaccid can be classified according to Tose. Cholesteatoma is formed if epithelial cells from the external layer of the eardrum accumulate in those pockets. It happens often due to an increased proliferation during a chronic irritation. Retraction pockets forms a matrix which produces other epithelial cells and causes the growth of a whole formation. If fibrous membrane between the epitympanum and the mesotympanum is formed, it causes ventilation problems and underpressure in the atticum and the processus mastoideus, even if ET function is normal. This barrier isolating epitympanum is formed as a result of recurrent acute inflammations or repeated surgical interventions.

- **Proliferation theory:** In retraction pockets, a deep papillary growth of flagstone epithelium of the external eardrum layer was proved.
- **Implantation theory:** this applies especially during the iatrogenous or posttraumatic cholesteatoma behind the intact eardrum without retraction pockets. It supposes an implantation of flagstone epithelium in the tympanal cavity e.g. during paracentesis.
- **Metaplastic theory:** this describes rare cases behind an intact eardrum with adequate history. It supposes metaplasia of the middle ear epithelium to flagstone epithelium.
- **Congenital cholesteatoma:** it can be present in the middle ear e.g. intracranially. Congenital cholesteatoma can be termed hamartoma.

Course: a growing cholesteatoma causes the destruction of surrounding tissues including the bone with possible intratemporal and intracranial complications. Conductive hearing loss is frequent but it does not have to be present even in cases of a total destruction of the middle ear bones because cholesteatoma can conduct sound sufficiently well. If the inner ear is damaged by a cholesteatoma (bone destruction, toxic damage), vestibular symptoms or perception hearing loss can be present. During an infection a foul-smelling secretion can be present.

Diagnostics: otoscopy or microotoscopy, imaging methods (X-ray, CT), hearing examination

Therapy: a surgical intervention in the middle ear, small retraction cholesteatomas can be removed during microotoscopy. Cholesteatomas should be treated as soon as possible due to possible complications. We must count with a possibility of recurrence.

Otitis media chronic adhesive

Definition: the presence of adhesions in middle ear as result of changes during chronic inflammation

Pathogenesis:

- An adhesion of the retracted eardrum with middle ear structures, usually in the area of the long branch of the incus, the promontorium or the epitympanum. Blood supply disruption of the middle ear bones leads to their destruction.
- Fibrous strips formation which causes bones fixation in the middle ear as a result of chronic inflammation, presence of foreign body or postoperative.

Course: conductive hearing loss is present, often with signs of a decreased mobility of bone string. An increased mobility is present if the bone string is broken or the eardrum is atrophic. If the risk of retraction is increased, the risk of cholesteatoma development is also increased.

Diagnostics:

Otoscopy: retracted eardrum, retracted pockets or sometimes practically normal otoscopic picture

Audiometry: conductive hearing loss

Tympanometry: if bone fixation is present we find curves As or B, if breakage of bone string we find curve Ad.

Therapy: tympanotomy, tympanoplasty

Degenerative Disease of the Middle Ear

Otosclerosis

Definition: a degenerative affliction of the temporal bone in white race (10%). Women are afflicted twice as much as men. The frequency of the disease is lower in coincidence of morbilli vaccination.

Etiology: unknown, morbilli virus probably activates responsible genes (autosomally dominant heritability)

Pathogenesis: resorption of normal bone (osteolysis of osteocytes) and its replacement by sponge or sclerotic bone (unorganized bone with higher count of osteocytes and fibrous tissue). In 80-90%, the pathology is restricted on the ventral edge of the round window with the calcification of the ligamentum annulare. It can afflict the cochlea as well as the labyrinth. Pregnancy or hormonal changes can cause the disease progression.

Symptoms: usually around the age of 20:

- Slowly progressing, usually conductive hearing loss
- Paracusis Willisii: patient hears better in murmur
- Tinnitus or dizziness in more severe cases
- Syndrome “van der Hoeve” (otosclerosis + osteogenesis imperfecta)

Diagnostics:

- History: heritability, pregnancy, hormonal changes and medication

- Otoscopy: usually normal eardrum. IN 10% Schwartz's sign: black and blue color of the eardrum caused by an increased vascularization of the promontorium
- Examination of hearing: tone audiometry: conductive hearing loss occurs at the beginning in lower frequencies, later even in higher. Carhart's notch: a decrease in bone conduction in frequency of 2000 Hz. In case of cochlear affliction, mixed hearing loss in higher frequencies is present. During tympanometry, we can find curve A or As.

Therapy: hearing-aid devices, surgery (stapedotomy, stapedectomy)

Tympanosclerosis

Definition: formation of thick fibrous tissue in the middle ear usually with the content of calcium salts

Etiology: unknown, middle ear inflammations, postoperative states

Symptoms: conductive hearing loss if the middle ear bones are afflicted

Diagnostic: hearing examination, tympanometry (curve As or B)

Therapy: hearing-aid devices or tympanoplasty in case of conductive hearing loss

Myringosclerosis

Definition: formation of thick fibrous tissue in the eardrum usually with content of calcium salts

Etiology: unknown, middle ear inflammations, after surgery intervention, paracentesis or ventilation tube insertion

Symptoms: conductive hearing loss if affliction is extensive

Diagnostic: hearing examination, tympanometry (curve As or B)

Therapy: hearing-aid devices or myringoplasty in case of conductive hearing loss

Diseases of the Inner Ear

Acute hearing loss

Definition: acute hearing loss with minimum of 30 dB on 3 neighbour frequencies in 3 days.

Pathogenesis:

1. viral – serous labyrinthitis
2. impaired blood perfusion – increased risk in patients with hypercholesterolemia, hypertension, diabetes, age above 50 years, hypercoagulation factors
3. cochlear membranes damage – pyramid fractures, acoustic trauma, barotrauma, endolymphatic hydrops

4. immune affliction – autoimmune (with or without systemic autoimmune disease, rheumatoid arthritis, Cogan's syndrome, Wegener's granulomatosis, ulcerative colitis), or parainfection affliction of inner ear

Symptoms: hearing loss, speech understanding impairment, tinnitus, pressure in the ear, ear pain. In case of membranous labyrinth damage, signs of balance disturbance are present – dizziness, nausea, sight impairment (problems with focusing on moving objects, or in fast change of vision)

Diagnostics, therapy, complications and consequences according to specific chapters.

Autoimmune diseases of inner ear

Definition: (fluctuating progressive) acute sensorineural hearing loss, with 15 dB decrease in 1 frequency or 10 dB decrease in 2 or more frequencies in several days. It reacts to anti-inflammatory drugs, especially corticosteroids.

Pathogenesis:

- histamine induced vasodilatation on the base of I.type allergic reaction causes endolymphatic hydrops because of damaged ion and fluids exchange in endolymph (improving ear symptoms in allergies after antiallergic drugs administration)
- settling of circulating immunocomplexes in the inner ear arteries (e.g. in patients with systemic lupus erythematoses or Wegener's granulomatosis)
- production of antibodies against antigens of the inner ear (proved experimentally, but no test for clinical praxis exists)

Etiology: unknown (congenital, crossed antigenicity with wall of some bacteria)

Symptoms: usually bilateral hearing loss, tinnitus, pressure in the ear, rarely ear pain. In case of membranous labyrinth damage, signs of balance disturbance are present – dizziness, nausea.

Diagnostics: audiometry, exclusion of retrocochlear hearing loss (BAEP, MRI), in case of balance problems vestibulologic examination is suitable, inflammatory markers (sedimentation, CRP), antinuclear antibodies, rheumatoid factor, circulating immunocomplexes, parts of complement, other serologic examination according to rheumatologist or immunologist consultation. Hearing improvement (15 dB on 1 frequency or 10 dB on 2 or more frequencies) after corticosteroid administration is essential for diagnostics.

Therapy: corticosteroids per os, intravenously or local application into the middle ear (penetration to the inner ear via the round window). Not all patients respond to treatment. Not all of them who respond are without recurrence. The probability of success decreases with time from the onset of the symptoms. The efficiency borderline is about 30 days.

Complications and consequences: hearing loss, balance problems. Long term corticosteroid treatment has strong side effects which outweigh a possible profit from this therapy.

Labyrinthitis

Definition: an inflammatory disease of the membranous part of the inner ear and/or the labyrinth.

Pathogenesis:

- hematogenous dissemination
- passing from the middle ear through the natural connections of the middle ear and the perilymphatic space (foramen ovale and rotundum), or through the perilymphatic fistula
- from the central nervous system (subarachnoidal space) through the internal auditory canal and the aqueductus cochleae

Etiology:

- serous labyrinthitis – bacterial toxins, mediators of inflammation, viruses (CMV, HSV1, mumps, rubeola, influenza, parainfluenza, adenoviruses, coxsackie viruses, RS virus)
- syndrome Ramsay-Hunt (herpes zoster oticus) – reactivation of the latent viral infection (varicella-zoster), often many years after primary infection
- Suppurative labyrinthitis – the most frequent agents of acute and chronic inflammations of the middle ear (OMA), *Neisseria meningitidis* and others

Symptoms: dizziness, nausea, hearing loss, pressure in the ear, tinnitus, ear pain, fevers, meningeal signs, signs of respiratory infection, sight impairment

Diagnostics: otoscopy, hearing examination, meningeal signs, basic vestibulologic examination (nystagmus, Hautant's test, Romberg's test, test Unterberg-Fucuda), eye examination, lumbar puncture (serology, cultivation). In indicated cases CT, MRI or advanced vestibulologic examination.

Therapy: ATB in suppurative labyrinthitis, antivirals in indicated cases, corticosteroids, antiemetics, antivertiginous drugs, hydration. Surgical intervention in case of infection focus in the middle ear

Complication and consequences: hearing loss, ossification of the cochlea (makes cochlear implantation impossible), infection spread to meninges, disequilibrium (problem with balance even after considerable improvement of the dizziness)

Meniere's disease

Definition: hydrops (a pressure change between the endolymph and the perilymph) of semicircular canals in the inner ear without a known etiology (after the exclusion of all other causes in this chapter)

Classification:

Typical (presence of all symptoms based on the affliction of the cochlea and the vestibular part of the inner ear) and atypical

- hydrops cochlearis – only cochlea is afflicted (dizziness is not present)
- hydrops vestibularis – vestibular part of the inner ear is afflicted (hearing loss is not present)

Etiology: unknown, microvascular blockade is supposed, an endocrine influence is known (more frequent onset in menses or menopause)

Symptoms: fluctuating hearing loss, tinnitus, several minutes of rotatory vertigo, pressure in the ear. Symptoms are episodic.

Diagnostics: hearing examination, vestibular examination, electrocochleography and BERA. Glycerol or furosemide test (hydrops is confirmed if hearing improves after drug administration). MR of the brain to exclude pathological changes of the acoustic nerve or the brain. CT of pyramids if we have a suspicion on perilymphatic fistula.

Therapy: diet (salt, caffeine and alcohol restriction), diuretics, corticosteroids (per os, intravenously or into the middle ear), symptomatic therapy (antiemetics, sedatives). Destruction of the inner ear (surgically or chemically – aminoglycosides injection into the middle or the inner ear) in case of repeated recurrences with bad quality of life.

Ototoxicity

Acoustic trauma

See Hearing loss caused by noise

Pyramid fracture

Hearing loss caused by noise

Clinically, we differentiate 3 diagnostic units:

- acoustic trauma
- hearing loss caused by long lasting noise (especially professional) – the influence of recurrent and overpower noise in months and years
- socioacusis – the hearing of city dwellers is worse than the hearing of countrymen

Pathophysiology: temporary threshold shift (TTS) is caused by metabolic exhaustion of ciliary cells exposed to overpower noise. Lasting stress leads to microinjuries – stereocilia affliction of the inner and the outer ciliary cells, their apoptosis, rupture of Reisner membrane, destruction of ciliary cells which all results in a permanent hearing loss (PTS – permanent threshold shift). Affliction is biggest on frequencies of 3000-6000 Hz, often with greatest notch on 4000 Hz because of 2 reasons:

- protective function of reflexes of m.stapedius and m.tensor tympani in deep frequencies up to 1000 Hz
- interference of waves in basillary membrane on 4000 Hz according to Barány's theory

Etiology: the level of affliction and chance of improvement depends on noise dose (noise intensity x time of noise stress) Affliction is symmetric in case of long lasting stress. In clashes (short sound of high intensity). Affliction is usually unilateral depending on the direction of the incoming noise. Hearing loss can develop after 8 hours in 85 dB noise. Double energy of sound (about 3 dB increase) causes similar damage in half of time. Persistent noise is worse than intermittent. Other factors are: individual endurance connected with genetic influence and the global state of the organism (higher risk is in diabetics, patients with hyperlipidemia, with cardiovascular diseases, with tobacco abuse or in patients who have ototoxic drugs).

Diagnostics: subjective audiometry, tympanometry with stapelial reflexes. Retocochlear hearing loss must be excluded (BAEP, MRI) only in case of asymmetric hearing. Laboratory tests are useless.

Differential diagnostics: Simulation, dissimulation and aggravation must be excluded by cortical evoked potentials (LAEP, CERA) in case of professional diseases or money compensation.

Therapy: there is no proved effective therapy. We can offer corticosteroids, vasodilating drugs or hyperbaroxytherapy with corresponding information about side effects.

Premeatusion: it is the most important due to the minimal or no effect of any therapy. Maximum levels of noise which should be harmless (according to A filter):

- 16 h – 85 dBA
- 8 h – 90 dBA
- 6 h – 92 dBA
- 4 h -95 dBA
- 3 h – 97 dBA
- 2 h – 100 dBA
- 1,5 h – 102 dBA
- 1 h – 105 dBA
- 30 min – 110 dBA
- 15 min – 115 dBA

Presbycusis

Changes of hearing connected with old age. We have 4 types according to Schuknecht:

- sensoric – atrophy of the ciliary cells
- neural – atrophy of the acoustic nerve and the auditory pathway neurons (unlike others, it does not afflict hearing too much, but significantly impairs speech understanding)
- strial (metabolic) – atrophy of stria vascularis (maintains chemic and bioelectric balance in the inner ear)
- conductive (mechanic) – thick and rigid basillary membrane of the cochlea

Etiology: genetic predispositions, diabetes mellitus, arteriosclerosis, noise, ototoxic substances, stress

Diagnostics: hearing examination, other examinations only due to differential diagnostics

Therapy: there is no causal therapy. Premeatusion is important (elimination of etiological agents). Hearing-aid devices, cochlear implants, FN systems and others, reading the lips.

Tinnitus

Definiton: perception of sound whose source is not in the patient's surrounding, head or ears.

Objective tinnitus

We can hear or register by any other way, or logically connect with physical source of sound.

Etiology: the most frequent cause is vascular (turbulent flow in vertebral arteries and so called "jugulary venous noise"), less frequent is muscular (palatal myoclonus and degenerative neuromuscular diseases – see neurology)

Diagnostics: examination of the auditory canal and the middle ear, hearing examination, examination of the stapedial reflexes, Eustachian tube tests. If we suspect vascular etiology, angiography can be made.

Therapy: removal of the pathology of the auditory canal and middle ear, tenotomy of m.tensor tympani in case of palatal myoclonus, embolization in case of vascular etiology.

Subjective tinnitus

Source of sound does not exist – it is a phantom and the patient is aware of the nonexistence of the source (appropriately to their intellect, state and age). It usually is a simple sound (e.g. whistle).

Etiology: it follows the majority of diseases in this chapter. In any situation we do not sense any sound, compensatory sound perception develops – tinnitus. Tinnitus can develop as well in case of auditory nerve or auditory pathways damage due to signal desynchronization.

Diagnostics: ENT examination (objective tinnitus exclusion), hearing examination, comparison and tinnitus masking in sounds from audiometer

- Murmur is typical for disease of the auditory canal, the middle ear and the cochlea (in the apex).
- Whistle is typical for basocochlear diseases
- Fizzle is typical for diseases of the external auditory canal, the middle ear, the auditory nerve and the auditory pathways
- Tinnitus caused by the inner ear diseases can be better masked than from the auditory nerve or the auditory pathways

Objective diagnostics of subjective tinnitus is performed by functional imaging methods – PET or fMRI

Therapy: causal therapy according to etiological agent, „tinnitus retraining therapy“ (abreaction, autosuggestion, masking by bruit, etc.)

Differential diagnostics: hallucinations are differentiated according to aspect of patient on sound source and by character of the sound (as a running locomotive)

Tumors of the Ear in Children

They make only a very small percentage of children tumors of neck and head (1,5% of primary tumors). Embryonal development can explain some pathological states. Os petrosum ossifies enchondrally and is completely formed at birth. Squama, mastoids and os tympani ossifies endesmally and develop partially after birth. Their active growth centers are predisposing factors for a mesenchymal tumor development.

Congenital Malformations and Paraneoplastic Processes

Histiocytosis X

Ear symptoms: otorrhea and retroauricular swelling, rarely ear pain. In otoscopy there can be seen polyp, granulation tissue, eczema of the auricle and the auditory canal. Conductive hearing loss is present in case of the auditory canal and the middle ear infiltration by soft tissue. Perforation of the eardrum in secondary infection. Perceptive hearing impairment and vertigo are rare and are present in the labyrinth destruction and n.VII paresis.

Premeatusion of recurrence: control otoscopy once in a month, control CT once in a year

See also: Histiocytosis X in Tumors an expansive processes in children

Choristoma

Most frequent is salivary choristoma of the middle ear. It is related to abnormalities of n.VII or ossicles in patients with congenital anomalies of the external ear and the face. In children under 5 years, usually the left ear is afflicted (but both can be afflicted). It grows slowly and causes conductive hearing loss.

See also: Choristoma in Tumors an expansive processes in children

Hamartoma

The most frequent is a slowly growing extracanalicular osteoma of the mastoids and the squama. Symptoms slowly worsen conductive hearing loss.

See also: Hamartoma in Tumors an expansive processes in children

Teratoma

The most frequent is auro-nasopharyngeal hairy polyp.

See also: Teratoma in Tumors an expansive processes in children

Cystic lesions of petrous bone apex

They are probably the results of obstruction of petrous bone pneumatizations in children.

- congenital epidermoid (usually found in adolescents)
- cholesterol cyst (granulomatous lesion containing cholesterol crystals, bleeding is essential factor for development)

Symptoms: conductive hearing loss caused by OMS because of ET obstruction, headache because of bone destruction and meningeal irritation, diplopia from n.III and n.VI damage, hyperesthesia in face (n.V), dizziness if labyrinth veins are damaged

Diagnostics: CT, MRI

Differential diagnostics: cholesteatoma

Therapy: surgical, marsupialization of cholesterol cyst and extirpation of epidermoid cyst.

Children myofibromatosis

Temporal bone is usually one of many afflicted bones

See also: Children myofibromatosis in Tumors an expansive processes in children

Fibrous dysplasia

Temporal bone affliction: external auditory canal stenosis, retroauricular swelling, displacement of auricle, development defects of ossicles or inner ear

See also: Fibrous dysplasia in Tumors an expansive processes in children

Benign Tumors

Neurinoma n.VIII

Definition: slowly growing, usually unilateral, intracranial tumor from Schwann cells afflicting vestibular (95%) or cochlear (5%) part of nerve. 80% of tumors are located in the pontocerebellar angle.

Incidence: 0,7-1,0/100 000

Etiology: coincidence with neurofibromatosis type I (unilateral) and II (bilateral)

Symptoms:

- most frequent is asymmetric sensorineural hearing loss from cochlear nerve damage (slowly developing) or vascularization damage (acute onset)
- tinnitus, dizziness, balance disturbances, headache
- malfunction of n.VII
- can be asymptomatic (even in big tumors)

Diagnostics: MRI with gadolinium contrast, hearing examination (subjective and objective methods)

Therapy: according to age, size and symptoms:

- follow up
- Leksell gamma knife
- Microsurgery

Adenoma

It is present especially in adults, in children it is rare. Adenomas are nonvascular, gummy formations, which can fill up the whole middle ear and can spread to mastoids. They often tightly adhere to the mucosa, eardrum and ossicles. Bone destruction is a sign of malignancy.

Symptoms: progressive unilateral hearing loss with sense of fullness in the ear and with tinnitus

Therapy: complete resection. Recurrence is rare.

Paraganglioma

Synonym: chemodectom, glomus tympanicum (jugulare) tumor

Definition: red vascularized spherical tumor developing from neuro-ectoderm.

Pathogenesis: develops from paraganglions beside vena jugularis and near Jacobson's and Arnold's nerve. It occurs more frequently in women in the 5th decade, but some cases are known even in small children. There is a hereditary form as well.

Classification:

- glomus tympanicum tumor develops in middle ear from plexus tympanicus (Jacobson's nerve)
- glomus jugulare tumor develops in the area of jugular bulb

Symptoms:

- unilateral pulse tinnitus, sense of fullness in the ear
- conductive hearing loss, immobility of ossicles
- large tumor can cause bleeding, pain, perceptive hearing loss, dizziness and lesions of head nerves (n.VII)
- glomus tympanicum usually does not cause bone destruction
- glomus jugulare is often connected with demineralization or bone erosion
- sweating, hyperactivity, restlessness, tachycardia, palpitations, hypertension can be caused by catecholamine production (diagnostics – vanilla-almond acid in the urine)

Diagnostics:

- otoscopy: red and violet tissue behind the eardrum. Brown's sign – overpressure during pneumotoscopy whitens eardrum and stops pulsation if tumor is in contact with eardrum
- tympanometry: higher impedance and higher pulse of synchronic wave
- audiometry: conductive, rarely perceptive hearing loss
- imaging methods: CT, MRI, arteriography (embolization)
- laboratory tests: vanilla-almond acid in urine

Differential diagnostics: aberrant a.carotis interna, aneurysm, high bulb jugularis, persistent stapedial artery, hemorrhagic secretion

Therapy:

- surgical (laser), alpha and beta blockers if catecholamine is produced. Embolization is possible.

- Radiotherapy

Prognosis is bad.

See also: Paraganglion in Tumor in the area of external neck

Malignant Tumors

Rhabdomyosarcoma (RMS)

It is the most frequent malignant ear tumor and temporal bone in children. In the middle ear, embryonal RMS is the most frequent.

Symptoms: serohemorrhagic discharge from the ear, non-suppurative granulations, polyp or tumor in the external auditory canal, neurologic symptoms (n.VII). If the petrous bone is afflicted, ear pathology is often minimal. Headache, paresis of n.VI, trigeminal neuralgia and Horner's syndrome are still present. RMS is highly aggressive and causes local destruction. It has distant hematogenous metastases in 20% in the time of diagnostics (lungs, bones, bone marrow), regional lymphatic nodes are afflicted less.

Diagnostics: ENT, neurology, CT, MRI, lumbar puncture, biopsy, X-ray of chest and bones, liver and spleen examination, bone marrow puncture

Therapy: it depends on the localization and state, combined surgery, radiotherapy and chemotherapy

Prognosis: average survival is 7-12 months in case of ear RMS

See also: Rhabdomyosarcoma in Tumors and expansive processes in children

Other sarcomas: fibrosarcoma, malignant fibrous histiocytoma, liposarcoma, mesenchymal chondrosarcoma and extraskeletal Ewing's sarcoma are rare

Leukemia

Acute lymphatic or lymphoblastic leukemia afflicts the ear and the temporal bone in 20% of patients. Leukemic infiltrates can cause hemorrhagic ulcers in the middle ear and in the external auditory canal, thickening of the eardrum and the mucosa, damage of n.VII and n.VIII, hearing loss and dizziness.

Lymphoma

Primary is rare. Non-Hodgkin lymphoma can afflict the middle ear and the mastoids.

Other malignant tumors

Spinocellular carcinoma, melanoma, tumors of salivary glands (adenocarcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma) are extremely rare in children

Auricle Injury

Auricle injuries are divided into open and closed (blunt). Open injuries must be revised; disinfected and foreign bodies must be removed. Skin suture is made in local or general anaesthesia according to the extension of the injury and the patient's state. ATB is given generally.

Othematoma

In case of blunt injury (combat sports) hematoma is formed usually on the external side of the eardrum where the perichondrium is connected to the auricle less tightly than on the posterior side. Inflammation of the auricle cartilage can develop in case of open wound or infected hematoma (perichondritis auriculae). Without therapy the cartilage is destroyed and the auricle is deformed because of cartilage nutrition defect.

Therapy: generally ATB, puncture, incision, drainage, destroyed cartilage removal

Auditory Canal Injury

It happens most frequently during personal hygiene when manipulating with foreign body.

Symptoms: pain, bleeding. **Diagnostics:** otoscopy

Therapy: local ATB

Injury of Eardrum and Middle Ear

It happens most frequently during personal hygiene when manipulating with foreign body, in pyramid fractures and in case of barotrauma (sudden air pressure change in the auditory canal).

Symptoms: pain, bleeding, hearing loss, tinnitus

Diagnostics: otoscopy, CT

Therapy: ATB, myringoplasty, tympanoplasty

Premeatusion: do not insert foreign bodies into auditory canal

Inner Ear Injury

See Pyramid fracture and Acoustic trauma

Pyramid Fracture

It is usually present in polytrauma (car accidents, falls)

Diagnostics: ENT examination, otoscopy, CT of pyramids, hearing examination, neurological examination, examination of face and auditory nerve function. Liquorrhea can be proved by the presence of beta-2-transferrine

- longitudinal fractures of pyramids: during otoscopy we usually find eardrum perforation and liquorrhea from it

Therapy: ATB i.v. (with penetration to the liquor), let the liquor flows out of ear, do not obstruct the auditory canal (risk of infection). After the state of the patient improves, we can reconstruct the eardrum and the middle ear (tympanoplasty)

- transverse fractures of pyramids: the eardrum is usually intact. Inner ear affliction causes perceptive hearing loss or possibly vestibular symptoms. Affliction of the middle ear can be diagnosed during otoscopy – violet eardrum (hemotympanum) signs presence of blood with possible tinge of liquor in the middle ear. Liquor can flow out via ET into nasopharynx (paradox otoliquorrhea)

Therapy: ATB, after the state of the patient improves, we can reconstruct the middle ear (tympanoplasty)

- Oblique fractures of pyramids: combination of the previous two types

Otogenous Inflammatory Complications

Intratemporal Complications

They are the most frequent from all complications of the middle ear inflammations. If bone is destroyed, abscesses can develop (Besold's, Mourne's, Mouret's,...) and the inflammation can spread into the skull base and to the surrounding tissues and into the intracranium.

Acute mastoiditis

Definition: mucosa and bone inflammation of pneumatic system of temporal bone

Symptoms:

- erythema and swelling in retro(peri)auricular area
- displaced auricle
- descent of posterior upper wall of auditory canal
- conductive hearing loss
- signs of middle ear inflammation

Diagnostics: Decontoured eardrum or discharge out of the middle ear can be found during otoscopy. Pneumatic system of temporal bone is destroyed on CT or X-ray.

Therapy: antromastoidectomy, ATB

Chronic mastoiditis

Usually follows chronic or recurrent middle ear inflammations. It causes increased temperature, chronic ear pain and sensitivity above mastoid processus.

Therapy: antromastoidectomy, ATB

Paresis of n.VII

It is usually the result of toxic damage on the face nerve in often dehiscent Fallopian canal.

Diagnostics: see Differential diagnostics of n.VII paresis.

Therapy: antromastoidectomy, ATB, corticosteroids, nootropics, rehabilitation

Petrositis

It is an inflammatory affliction of pneumatized system of the petrous bone. It causes Gradenig's syndrome (n.VII paresis, trigeminal neuralgia, and discharge out of the middle ear)

Therapy: antromastoidectomy, ATB, corticosteroids, nootropics, rehabilitation

Labyrinthitis

It can be caused by toxic products of inflammation (fistula of the lateral semi-circular canal in the cholesteatoma, toxin transfer through the round window in acute inflammations) – serous or toxic labyrinthitis. Or the infectious agent can penetrate into the inner ear and cause diffuse inflammation – suppurative labyrinthitis. Serous and suppurative labyrinthitis can both be diffuse or circumscribed, acute or chronic. The end stage of chronic labyrinthitis is the sclerosis of labyrinth. We find sensorineural hearing loss, dizziness, balance loss, nystagmus, nausea, vomiting.

Diagnostics: hearing examination including tuning fork tests (Weber's test) and vestibular apparatus, imaging methods of temporal bone

Therapy: antromastoidectomy, ATB, corticosteroids

See also: labyrinthitis in Disease of the inner ear

Intracranial Complications

- meningitis – antromastoidectomy is indicated
- abscesses – epidural, subdural, brain, cerebellar – antromastoidectomy or neurosurgical intervention with abscess evacuation is indicated

- thrombophlebitis – sigmoidal sinus is usually afflicted. Thrombosis afflicts emissarium mastoideum as well and this causes increased palpable sensitivity during pressure on mastoid processus (Griesinger's sign)

Therapy: antromastoidectomy, hematological therapy (thrombolysis) if surgical intervention fails – thrombus evacuation

- hydrocephalus – obstructive in chronic thrombophlebitis

Differential Diagnostics of Ear Diseases

Differential Diagnostics of Hearing Problems

Classification:

- conductive – see diseases of outer and middle ear
- sensorineural (perceptive) – cochlear and retrocochlear – see diseases of the inner ear and the auditory nerve
- mixed – conductive + perceptive
- central – see neurology

Differential Diagnostics of Ear Pain

Definiton: ear pain is a subjective feeling determined by the patient and localized around the ear. In children non-speaking patients, we presume ear pain on the basis of local and global symptoms and clinical findings. The most frequent cause is an inflammatory affliction of the outer and the middle ear. Children suffer from ear pain usually in acute otitis media. They form about a half of the patients in ENT ambulances.

Classification:

- otogenous: often sharp, localized into the external auditory canal or into its very proximity
- non-otogenous: usually blunt, localized in the whole area of the ear and in addition the pain is present in another part of the head or the neck. This type of pain is mediated by neck nerves.

Otogenous pain

- Infection:

- Auricle inflammation (erysipelas, perichondritis)
- Auditory canal inflammation
- Obturating cerumen
- OMA
- Chronic otitis media
- Complication of middle ear inflammations
- Herpes zoster oticus
- Injury
 - Auricle injury
 - Foreign bodies
 - Barotrauma, acoustic injury
 - Burns and frostbites of auricle
 - Fractures of temporal bone
- Tumors
- Hydrops of the inner ear (M. Menière)

Non-otogenous pain

Pain transposed from swallowing or respiratory pathways and soft tissues of the neck:

- Odontogenous (tooth decay, stomatitis, teeth growth,...)
- Injury of facial skeleton (fractures of mandible joints, mandible luxation)
- Inflammations of oral cavity and pharynx
- Foreign body in pharynx
- Status post tonsillectomiam et adenotomiam
- Tumors of pharynx and larynx
- Neck lymphadenitis
- Parotitis
- Injury of neck spine
- Processus styloideus elongatus
- Rhinosinusitis
- Other (psychogenic, megrim, skull base tumor, neuralgia of n.occipitalis major, n.VII, ggl. Pterygopalatinum)

Peripheral vestibular syndrome

Peripheral vestibular apparatus affliction (semicircular canals, acculus, utricle) and vestibular part of auditory nerve

Etiology:

- labyrinthitis
- hydrops (M. Menière)
- neuronitis of n.VIII
- kinetosis
- benign position paroxysmal vertigo (BPPV)
- paroxysmal peripheral vertigo – mostly in patients with perceptive hearing loss, or changes in blood perfusion of vertebral arteries (degenerative disease of spine)

Symptoms: mostly acute onset of rotatory vertigo, balance loss connected with nausea and vomiting. In unilateral affliction there are present typical directional deviations. It lasts usually minutes (exceptionally days), fast relief from symptoms, good central compensation.

Diagnostics: vestibular examination, hearing examination, neurological examination, MRI of brain, CT of pyramids

Therapy: causal according to the etiology. Symptomatic (sedatives, antihistaminics, antiemetics, mineral balance)

Central vestibular syndrome

Affliction of the brain stem, the cerebellum and other brain structures participating in balance maintenance.

Etiology: CNS affliction – see Neurology

Symptoms: disequilibrium (balance uncertainty) rather than dizziness, slow onset, without nausea and vomiting. It lasts days to years. Balance deviations are nonspecific. Relief is very slow or does not occur at all. Central compensation is not possible.

Therapy: causal see Neurology, Neurosurgery. Rehabilitation is essential in residual affliction.

We must exclude the cause of n.VII paresis developed as a result of pathology in ENT area.

Classification:

- central
- peripheral

The most frequent causes of peripheral paresis:

- Bell's palsy – idiopathic, commonly result of viral infection
- Ear pathology – inflammations (OMA, mastoiditis, cholesteatoma), fractures of pyramids, tumors
- Parotis pathology – malignant tumors, injury, iatrogenous affliction (surgery)

Diagnostics:

- examination of n.petrosus major function – Schirmer's test (made by ophthalmologist)
- examination of n.stapedius function – tympanometry (stapedial reflex)
- examination of chorda tympani function – taste in first two thirds of tongue

Therapy: according to primary disease, ATB, corticosteroids, nootropics, and rehabilitation. Antromastoidectomy is indicated if an inflammatory affliction of the middle ear is present.

Basics of Ear Surgery

Surgical approaches into temporal bone

- retroauricular – skin incision about 5mm behind auricle
- endaural – skin incision in incisura heliotragica
- endomeatal – via meatus acusticus externus
- combined

Sanitation surgery

Antromastoidectomy

Pneumatization of the processus mastoideum removal with communication enlargement to the eardrum cavity (aditus ad antrum mastoideum) from retroauricular incision. The most frequent indication is mastoiditis and other inflammatory complications. In young children who do not have developed pneumatisation, it is called antrotomy.

Atticotomy

Atticotomy surgery (epitympanum) from endaural approach. The most frequent indication is lesser cholesteatoma.

Atticoantromastoidectomy

It is indicated in case of extensive cholesteatoma from a retroauricular or preaural approach. It contains the sanitation of the atticum and the pneumatisation of the processus mastoideus. The dorsal upper wall of the bone part of the external auditory canal has to be usually removed as well. If the auditory canal is reconstructed or the dorsal upper wall of the auditory canal was not removed, we call it a closed technique (it usually demands second-look operation up to 1 year). In other cases we call it an open technique. Classification according to Wullstein is now obsolete. For ossicles string reconstruction, artificial materials (metal, plastic) are used – piston, PORP, TORP.

Reconstruction Surgery – Tympanoplasty

Myringoplasty

Reconstruction in case of eardrum perforation. It is the simplest tympanoplasty. Small perforations can be resolved by removing margin of perforation and cover (silk, cigarette paper). For the reconstruction of the larger perforation, we use autologous materials. At present, „hard“ chondroperichondral graft from the tragus or the dorsal part of the auricle is preferred. The transplant stays in the position of the former eardrum (incisura tympanica).

Tympanotomy

The reconstruction of the ossicles can be performed after skin incision approx. 2 mm before eardrum and lifting off the eardrum. Autologous materials have a disadvantage in the form of insufficient nutrition and therefore they can be destructed. Artificial materials are better (they do not destruct).

'Comments' on nose embryology

Nose placodes are formed in 3rd week on the frontal prominence (olfaction area), which deepen and form olfaction sockets and olfaction vesicles.

- Frontal prominence is divided by 3 notches into 2 medial and 2 lateral prominences. Medial prominences deepen and turn caudally so they separate olfaction socket from primitive oral cavity – membrana bucconasalis.
- Cranial prominences (upper jaw) grow together with lateral prominences of the nose (line is base of sulcus nasolacrimalis) and with medial prominences (entrance to olfaction socket is formed and primitive nose entrance is circumscribed)
- After that both medial prominences grow together and they form the base of the philtrum. Lateral prominences form lateral parts of nose and medial parts of the face.

- Dorsum nasi develops from unpaired area triangularis located above medial nose prominences. This way formed flat middle part of the face rises with subsequent growth. Simultaneously grow together prominences of the first branchial arch for the upper and lower jaw and by this eyes are moved medially.
- Buconasal membrane rips in 4th week so that primitive choanas are formed. All olfaction sockets cause in advance prominences in form of sulcus vomeronasalis jacobsoni, which later closes itself into a canal inserted into the nose septum.