Hearing Disorders in Children and Adults
Importance of hearing

• Communication is based on speech and hearing
• Continuous contact with the environment
• Consequences of severe deafness in childhood ⇒
  delayed development of speech, reading difficulties, intellectual and emotional disorders, behavioral disturbances, articulation problems
• Severe deafness in adults⇒
  Isolation, depression, secondary speech disorders
Statistical data

- WHO (2001): 50 million hard of hearing patients all over the world, 30% severely deaf, 20% children
- 10% of the population is involved
- Incidence of hearing disorders in infants in Hungary is 1%
  Distribution of hearing disorders according to localization:
  - sensorineural: 60%
  - conductive: 30%
  - mixed: 10%
Normal pure tone threshold audiogram
What do you have to think of if the patient complains about sudden deafness?

- wax (impacted cerumen)
- otitis externa, furuncule of the external meatus
- acute purulent otitis media (children)
- tubal occlusion
- labyrinthitis (complication of chronic otitis)
- acute sensorineural hearing loss (microcirculation disorder)
- acoustic neuroma
- Meniere`s disease
- perforation of the tympanic membrane
- labyrinth commotion
- pyramidal fracture
- acute acoustic trauma
What do you have to think of if the patient complains about progressive hearing loss?

• Presbyacusis
• Noise induced hearing loss
• Otosclerosis
• Hereditary deafness
• Acoustic neurinome
• Chronic purulent otitis media
• Tympanosclerosis, adhesive process
What is an emergency at a hearing impaired patient?

- Suspicion of acute sensorineural hearing loss
- Trauma (skull base fracture, perforation of tympanic membrane, acoustic trauma)
- Pain+discharge+vertigo+facial palsy
- Pain+discharge+vertigo
- Pain+discharge+fever
- Pain+discharge
- Pain
Diagnosis of hearing loss

- Case history
- ENT physical examination
- Audiological examination
- Radiological assessment
- Otoneurology
- Neurology
- Internal diseases
- Laboratory examinations
Characteristical hearing losses

- High frequency hearing loss: noise trauma, presbyacusis
- Low frequency hearing loss: M. Meniere
- Speech discrimination loss: lesion of the inner ear or n.VIII.
- Fluctuating hearing loss: M. Meniere
- Unilateral hearing loss: M. Meniere, acoustic neurinome
- Bilateral hearing loss: presbyacusis, noise induced hearing loss, ototoxic lesions
What do we have to ask from the patient?

• Since when does the patient have hearing loss?
• Is it unilateral or bilateral?
• Is it permanent or intermittent?
• How severe is the hearing loss?
• Is there a discharge, tinnitus, vertigo, pain?
• Did the patient have ear surgery or any otological event previously?
Conductive hearing loss

- **Etiology:** diseases of external and middle ear
- Usually surgically treated
- Speech is soft and monotonous
- Good speech comprehension with appropriate amplification
- No significant articulation disorder
Conductive hearing loss
Otitis externa ekzematosa  Otitis externa diffusa
Otitis externa mycotica

Otitis externa diffusa
Conductive hearing loss 1.

- Wax
- Foreign body
- Severe otitis externa
- Atresia
- Ossicular abnormalities
- Osteoma, exostosis
- Tumors of the external and middle ear
Foreign body in the external meatus (bead)
Multiple exostoses in the ear canal
Conductive hearing loss 2.

- Traumatic perforation of the tympanic membrane
- Tubal catarrh, serosus otitis media
- Acute purulent otitis media
- Chronic purulent otitis media
- Otosclerosis
- Tympanosclerosis, adhesive process
- Disruption of the ossicular chain
- Longitudinal pyramidal fracture
Serous otitis media
Acute purulent otitis media
Chronic mesotympanic otitis media
Tympano sclerosis
Sensorineural hearing loss

- **Etiology:** diseases of inner ear and upper auditory pathways
- **Usually treated with conservative methods**
- **Speech is much too loud**
- **Speech comprehension problems**
- **Sometimes articulation problems**
Sensorineural hearing loss
Sensorineural hearing loss 1.

- Inner ear malformations
- Hereditary hearing loss
- Prenatal acquired (rubella, toxic damages, fetal hypoxia, irradiation)
- Perinatal (fetal asphyxia, erythroblastosis foetalis)
Sensorineural hearing loss 2.

Postnatally acquired hearing losses
- Trauma (labyrinth commotion, pyramidal transverse fractures)
- Noise-induced
- Presbyacusis
- Toxic lesions (aminoglycosids!!!, dihydrostreptomycin, cytostatics)
- Microcirculation disorders (hypertonia, arteriosclerosis, diabetes, lipid metabolism disorders, nicotin)
- Infections (meningitis, mumps, herpes zoster, Lyme-disease, influenza)
- M. Meniere
- Tumors (acoustic neuroma), metastasis, congenital cholesteatoma
- Sclerosis multiplex
Herpes zoster oticus
Noise-induced hearing loss
Presbyacusis
Speech audiogram
Sensorineural hearing loss 3.

**Therapy**

- **Importance of prevention** (noise, ototoxic antibiotics)
- **Acute sensorineural hearing loss**: vasodilator infusions, vitamins, corticosteroids, treatment of internal diseases
- **Chronic hearing loss**: hearing aid, in case of symmetrical hearing loss bilaterally
- **In selected cases of profound bilateral hearing loss of cochlear origin**: cochlear implantation
Mixed hearing loss
Acute sensorineural hearing loss

- It is a medical emergency!
- Symptoms: feeling of pressure in the ear, tinnitus, severe hearing loss
- Disorder of microcirculation of the inner ear (occasionally autoimmune disease)
- Generally unilateral sensorineural hearing loss
- Differential diagnosis: acoustic neuroma, acute tubal catarrh, impacted cerumen, viral infections
- Immediate vasodilatation or low-molecular weight infusions, corticosteroids
Hearing loss in childhood

When to suspect it?

- If a newborn consistently doesn`t react to noise
- If the tone of crying is unusual
- Delayed development of speech
- Slurred articulation
- Acoustic inattention
- Unusual behaviour (introversion or aggressivity)
Risk factors of hearing loss in childhood

- Intrauterin, peri- és postnatal infections
- Neonatal hyperbilirubinaemia
- Craniofacial disorders
- Hearing loss in family history
- Symptoms of well known syndromes combined with hearing loss
- NICU treatment lasting more than 48 hours
- Syndromes with progressive hearing losses
- Neurodegenerative illnesses
- Parental suspicions
Diagnosis and treatment of hearing impaired children

• In case of suspicious symptoms and risk factors assessment of hearing

• **Immediate therapy according to cause and degree of hearing loss**

• **Aim:** prevention of auditory deprivation, acquiring speech, hearing training, attending normal schools
Prevention of hearing loss

- Protection of hearing against noise, screenings
- Avoiding ototoxic antibiotics (aminoglycosids)
- In case of acute hearing loss immediate hospitalisation
- Early hearing screening
- Close observation of children with risk factors
- Rehabilitation of patients with hearing aids
- Education of people about hearing
Cochlear implant

Electrical device, which in case of bilateral hearing loss/deafness of cochlear origin delivers sound and speech information directly to the auditory nerve by means of small electrical currents.
Indication of CI

Bilateral sensorineural deafness of cochlear origin with little or no benefit from a hearing aid.
Cochlear implant

1. Sound waves are picked up by the microphone
2. Signal sent to speech processor
3. Speech processor codes the signal
4. Coded signal sent to transmitter
5. Signal sent through intact skin to receiver
6. Receiver decodes the signal
7. Electrodes stimulate the auditory nerve
8. Nerve impulses sent to the brain
Classification of patients

- **Prelinguals**
  *patients getting deaf before having acquired speech* (<3 years old)

- **Perilinguals**
  *patients getting deaf during the period of speech development* (between 3-5 years of age)

- **Postlinguals**
  *patients getting deaf after having acquired speech* (>5 years old)
Who are candidates for CI?

• Prelingual children maximum under 6 (“the younger the better”)
• Postlingual patients of any age
• Rarely older prelingual patients
• Reimplantation cases
Cochlear implantation
Selection process before CI

- Audiological evaluation
- Electrophysiological measures
- Promontory test/electroaudiometry
- Psychological evaluation
- Complete medical checkup
- CT scan (in some cases MRI)
Contraindications

- Poor anesthetic risk
- Severe mental retardation
- Severe psychiatric disorders
- Organic brain syndromes
- AIDS
- Active systemic disease
- Active middle ear disease
- Etc.
Surgery

- Operations are performed in general anesthesia combined with local infiltration
- Perioperative antibiotic prophylaxis
- Soft surgery
- Antiemetics administered intravenously before the end of surgery
- No drainage
- In case of uneventful recovery, patient is discharged from the hospital after 2-3 days
- First fitting after 4 weeks, sometimes sooner
Results after cochlear implantation (PTA)

- Excellent speech comprehension without lip-reading
- Better speech production
- Use of telephone
- Good speech comprehension even in difficult circumstances (noise, etc.)
Bilateral cochlear implantation

- Better speech comprehension in noise
- Localization of sounds
- Speech comprehension equals that of normal listeners