CONGENITAL DEFECTS OF EAR

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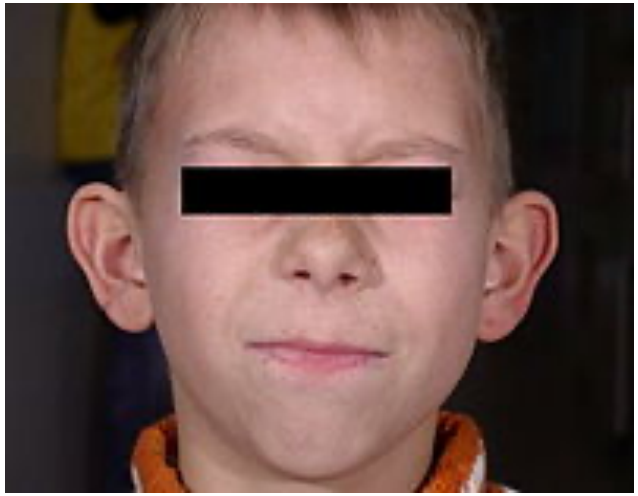
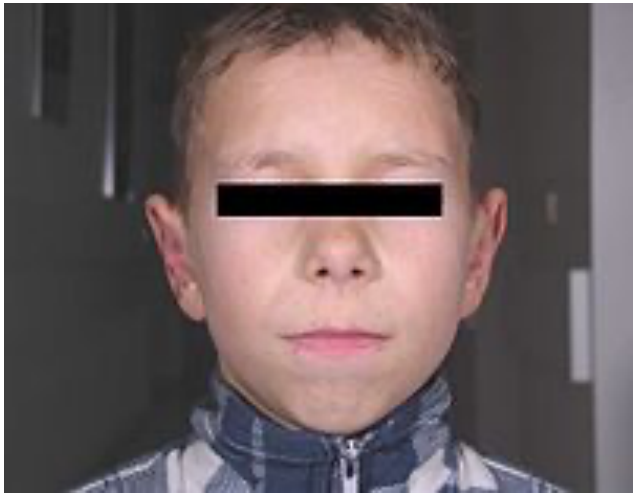
Seminar for medical students 2019/20

APOSTASIS AURICULI

- Deformity of shape, cosmetic defect
- **Therapy:** plastic correction – otoplasty:
- Children from the age of 6 years

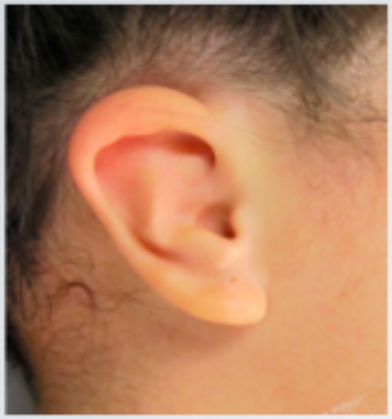




MICROTIA AND ATRESIA OF EAC

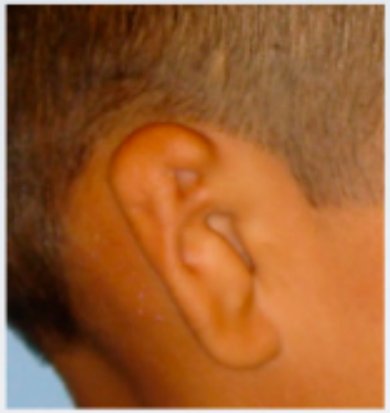
- Congenital defect of auricle development **(microtia)** or missing auricle **(anotia)** is often combined with the congenital defect of the EAC **(stenosis, atresia)**
- Auditory canal stenosis means that it is narrower than 4 mm
- **Dg:** CT, objective hearing examination (exclusion of congenital defect of the middle and inner ear or the auditory track)
- Conductive hearing loss

MICROTIA (I-III), ANOTIA (IV)



Grade 1

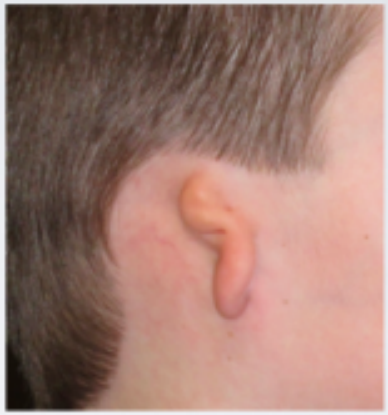
Smaller than normal, but the ear has mostly normal anatomy



Grade 2

Part of the ear looks normal, usually the lower half

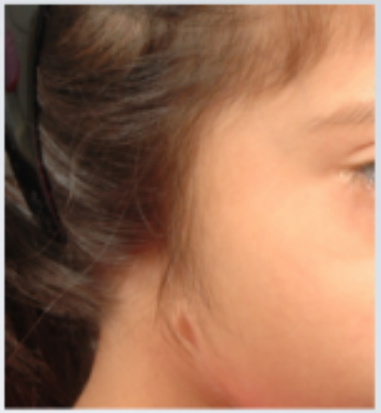
The canal may be normal, small or completely closed



Grade 3

Just a small remnant of "peanut-shaped" skin and cartilage

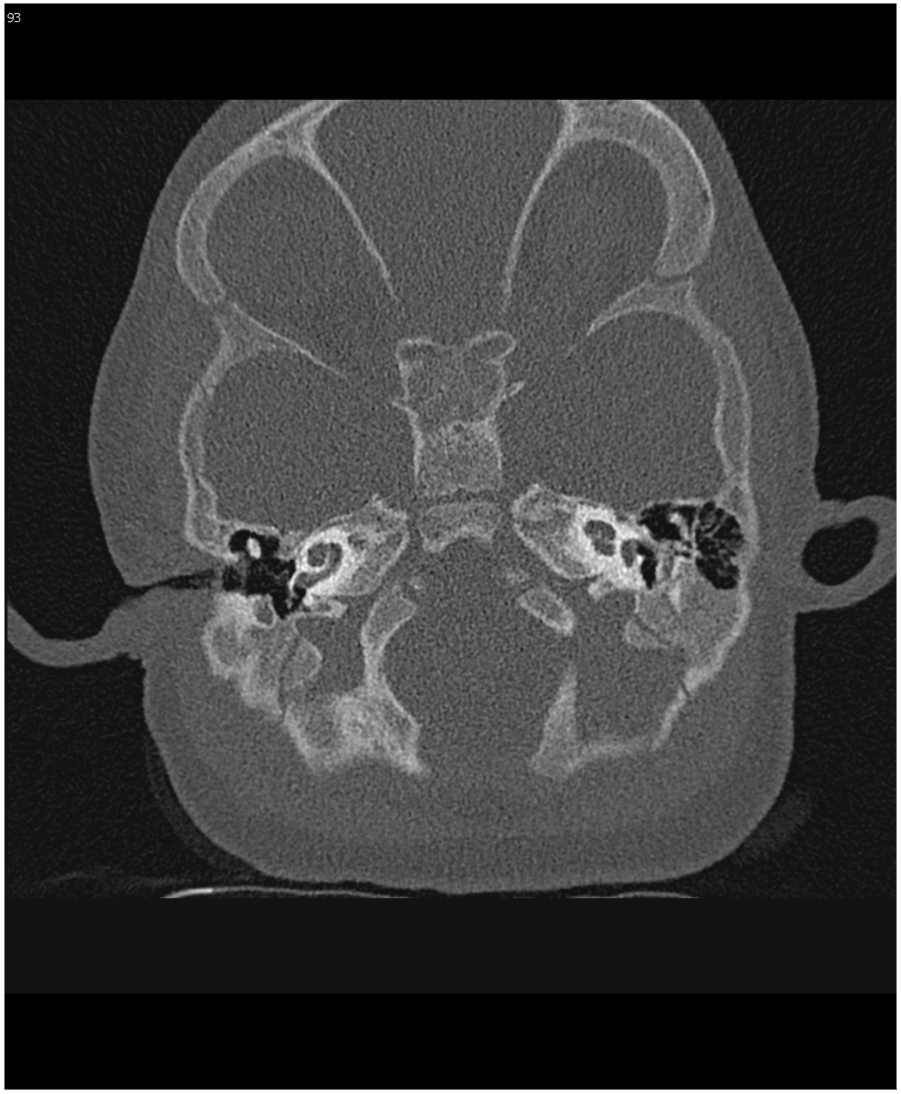
There is no canal, which is called aural atresia



Grade 4

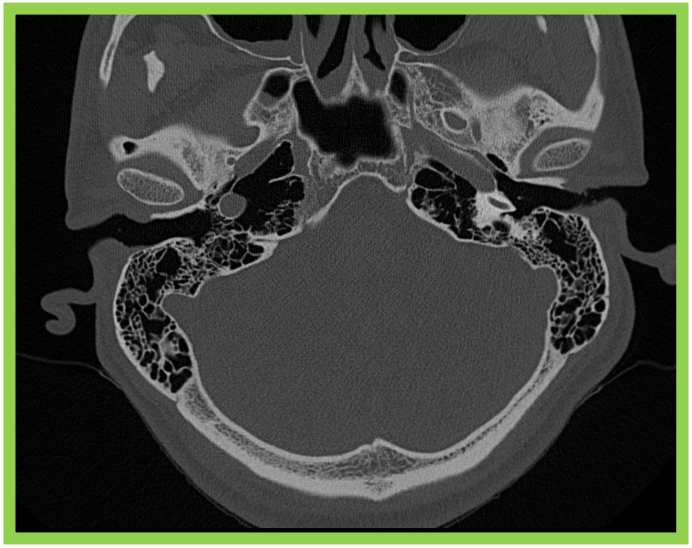
Complete absence of both the external ear and the ear canal, also called "anotia"

ATRESIA OF EAC



HRCT of the Temporal Bone

Right side – stenosis of EAC
Left side – atresia of EAC



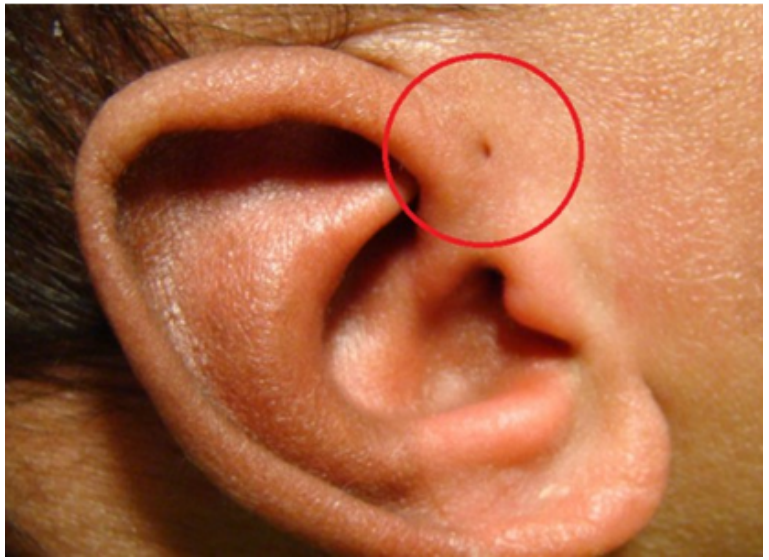
THERAPY

- It depends on examination results and on hearing affliction extent (unilateral or bilateral affliction)
- Aim: provide communication
- Hearingaid devices (BAHA)
- Surgery: tympanoplasty, plastic of external auditory canal or auricle

FISTULA AURIS CONGENITAL

- External opening is placed near the tragus and the inner opening between cartilaginous and bone part of the EAC
- Complication – inflammation (secretion, swelling or erythema)
- Th: Exstirpation,

  ATB (inflammation)



STRUCTURAL ANOMALIES OF MIDDLE EAR

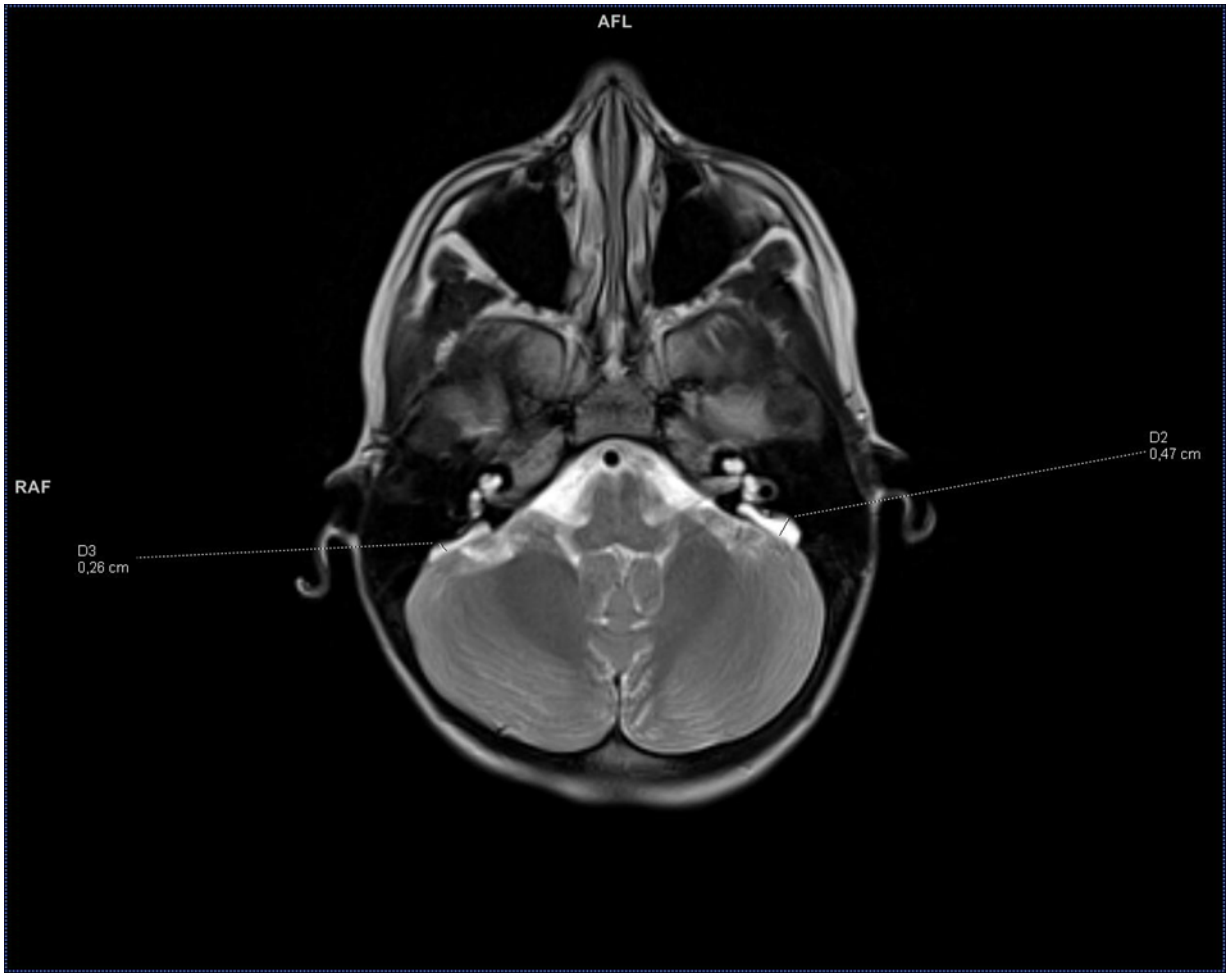
- Usually connected with atresia of EAC and anomalies of auricle
- Isolated / part of syndroms (more common, e.g. Treacher Collins)
- Usually unilateral
- Hearing-impairment
- Dg: CT, objective hearing examination
- Th: hearing-aid devices, tympanoplasty

STRUCTURAL ANOMALIES OF INNER EAR

- 20% of children with SNHL (sensorineural hearing loss) have CT anomalies of inner ear

- Cochlear anomalies
- Enlarged vestibular aqueduct (EVA)
- Semicircular canal dysplasia

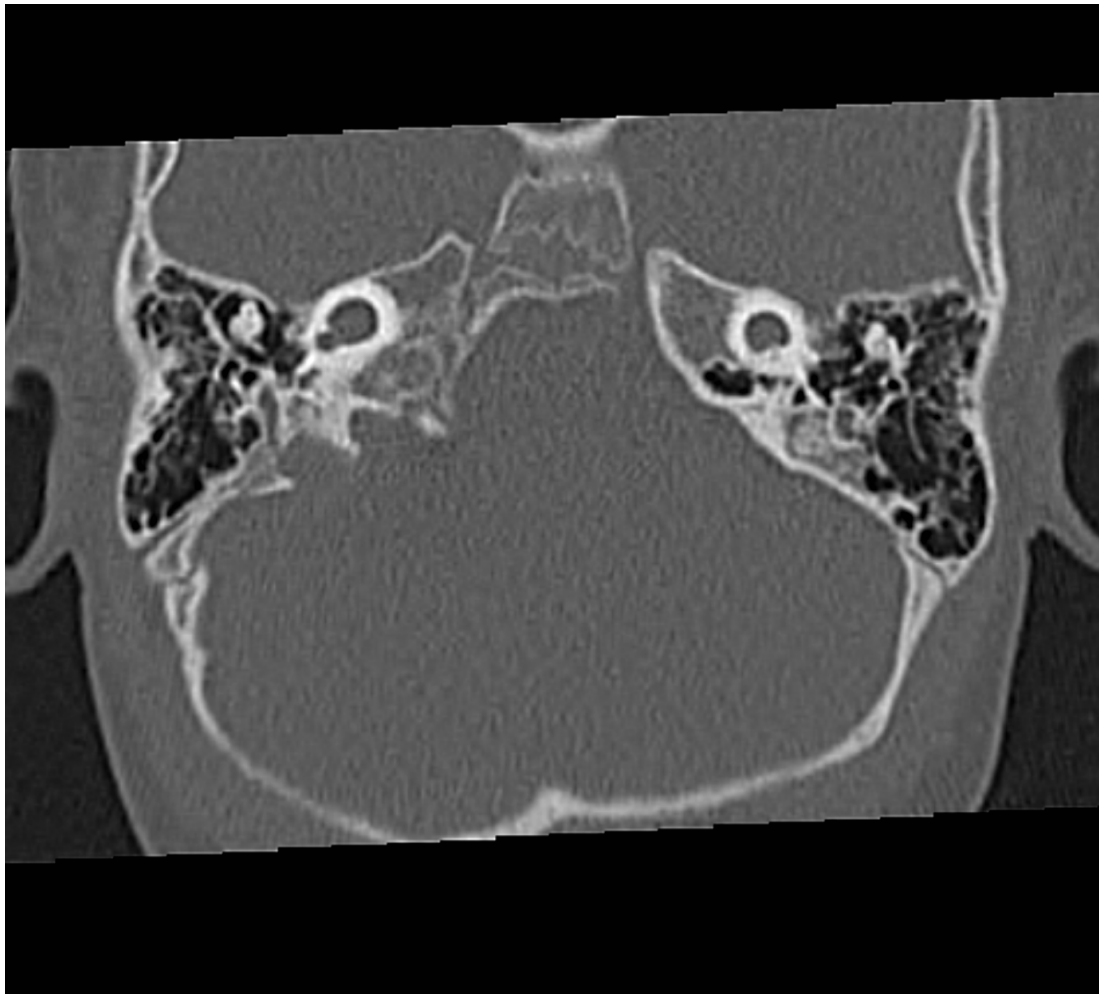
ENLARGED VESTIBULAR AQUEDUCT (EVA)

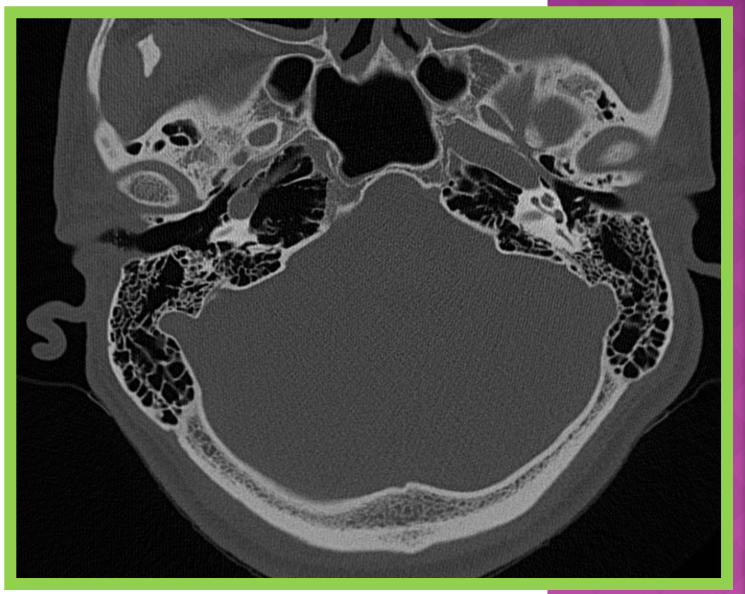


COCHLEAR ANOMALIES

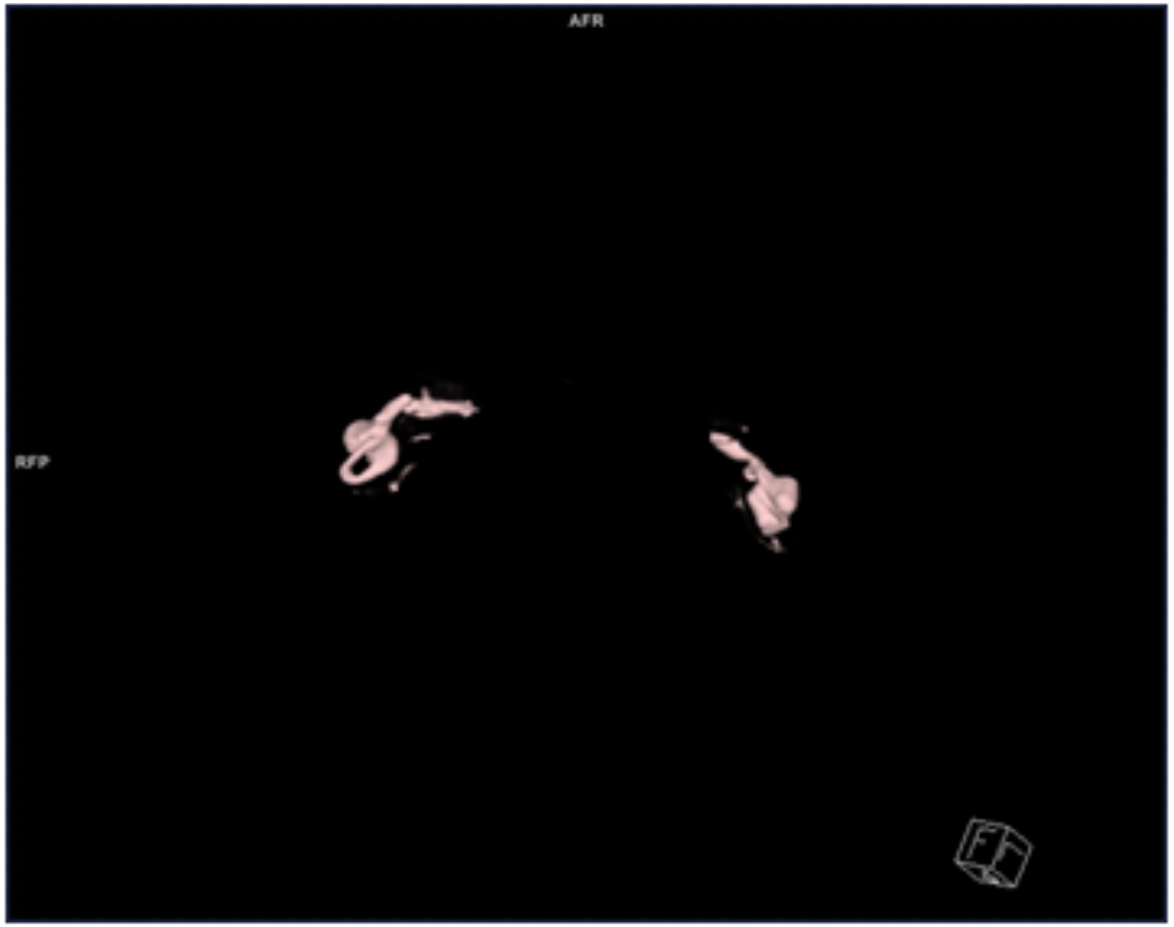
- **Michel deformity** or complete labyrinthine aplasia (cochlea + vestibulum)
- **Cochlear aplasia**
- **Common cavity malformation** to the cochlea and vestibule
- **Cochlear hypoplasia**
- **Cochlear incomplete partition type I** (including cystic cochleovestibular anomaly)
- **Cochlear incomplete partition type II (Mondini dysplasia)**

COCHLEAR APLASIA

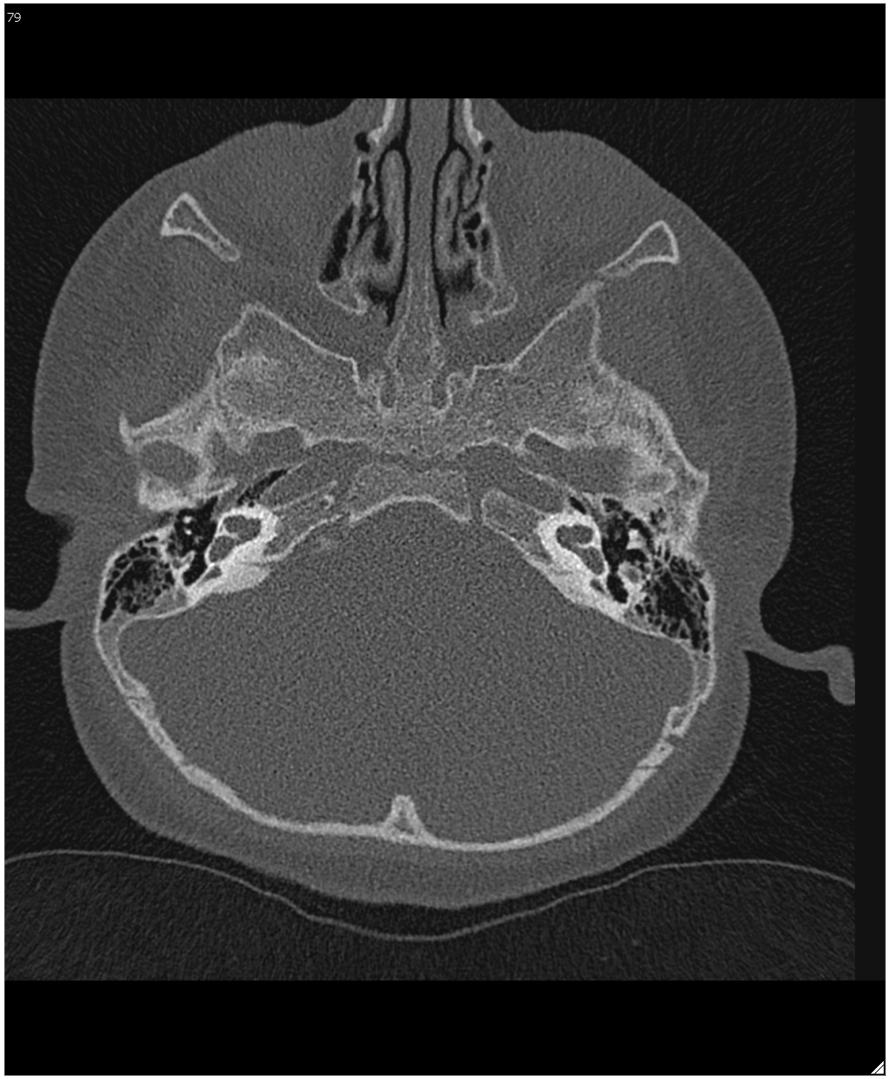




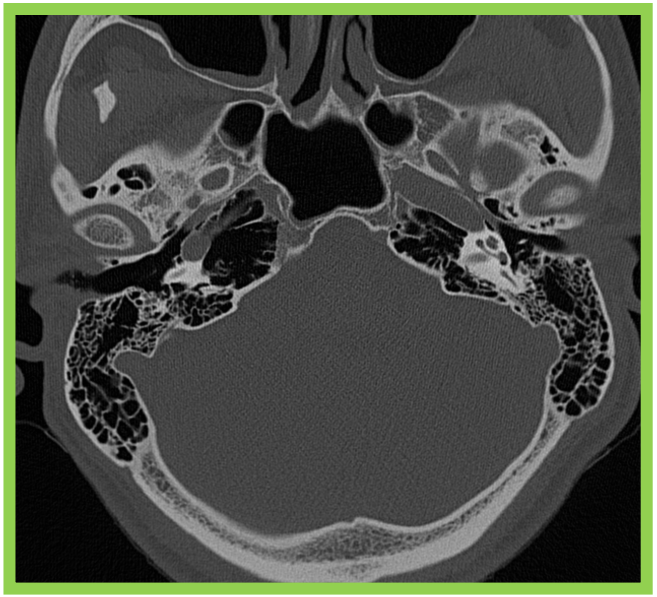
COCHLEAR APLASIA



MONDINI´S DYSPLASIA

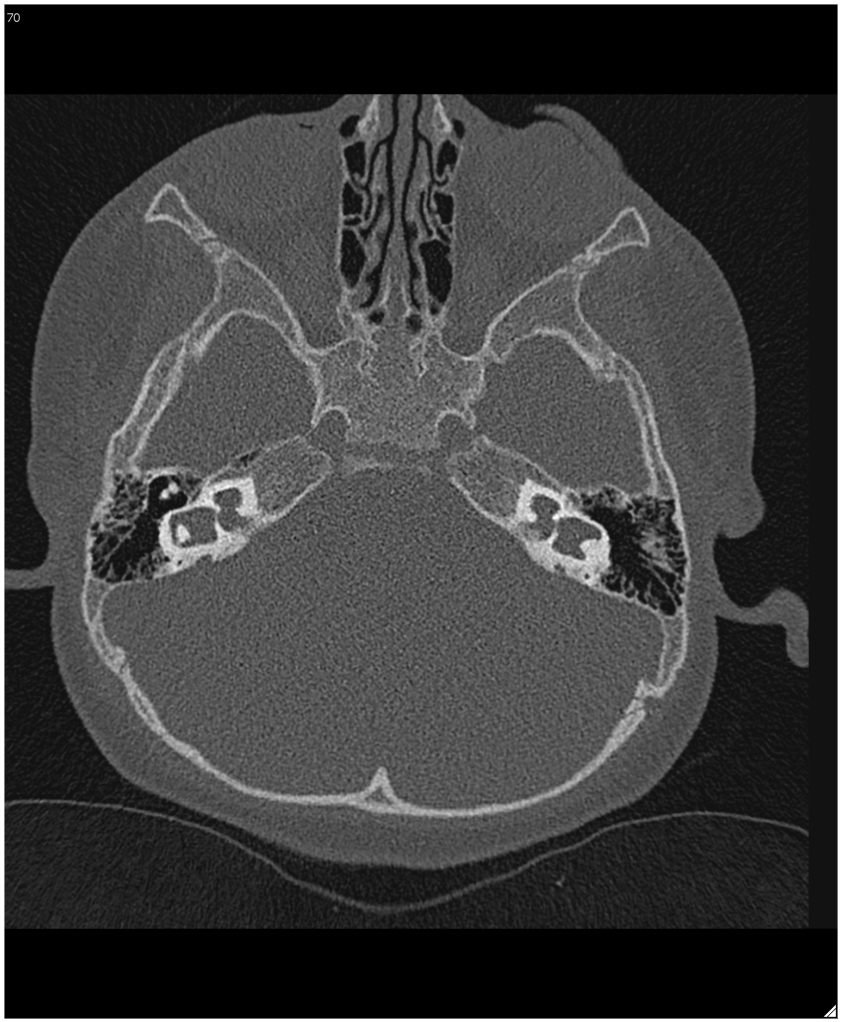


1,5 screw of cochlea,
dilated aqueductus

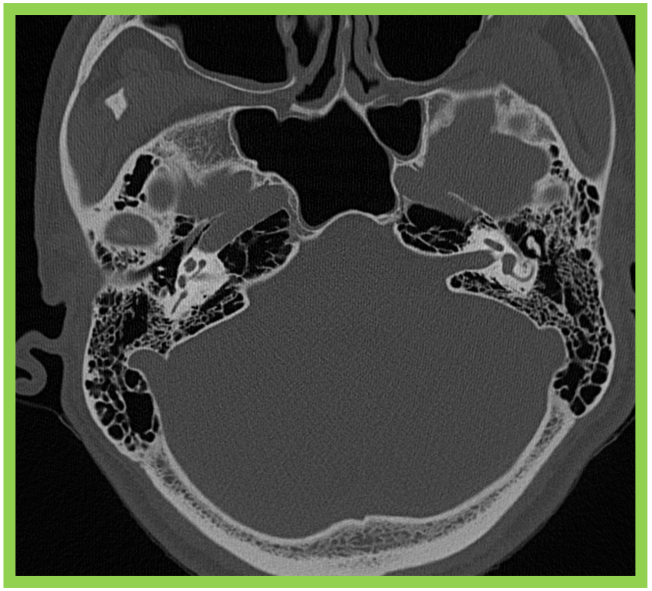


Normal cochlea 2,5-2,75 screw

MONDINI´S DYSPLASIA



Dilated vestibulum



Normal vestibulum

DEFECTS OF HEARING

- **60% congenital**
- Damage of auditory organ during development (1. trimester, prenatal period)
- **Genetic hearing defects**

  Non-syndromic 70%

  80% autosomally recessive **(Connexin 26 - protein)**

  19% autosomally dominant

  1-2% X chromozom

  Syndromic 30%
- **Infection:** Rubeola, CMV, toxoplasmosis…
- **40% gained**
- Perinatal period – premature, asphyxia…
- Postnatal period – meningitis, injury…

SYNDROMS CONNECTED WITH ANOMALIES OF THE EAR

Anomaly of external + middle

- **Treacher-collins syndrome**

Anomaly of inner ear

- **Pendred´s syndrome:** EVA + Mondini dysplasia, affiction of thyroid gland, 2. most common
- **Usher´s syndrome:** partial or total hearing loss and vision loss that worsens over time
- **Waardenburg´s syndrome**: unilateral or bilateral SNHL, pigmentation changes
- **Alport´s syndrome:** progressive SNHL, affliction of kidneys

CONGENITAL DEFECTS OF AUDITORY NERVE

- Auditory neuropathy
- Aplasia or hypoplasia of auditory nerve
- Demyelinization disease

DIAGNOSTIC OF HEARING IMPAIRMENT (NEWBORNS)

- **Newborn hearing screening**
- **Otoacoustic emissions (OAE)**

- Otoscopy, tympanometry
- Examination of evoked potentials (BERA - Brainstem Evoked Response Audiometry)
- CT / MRI, genetic examination