

European Course on
Pediatric Neuroradiology in
Latin America (ECPNR-LATAM)
3rd Cycle - Module 1

HEARING LOSS

**Eloisa Santiago
Gebrim**

Idealização:



Organização:

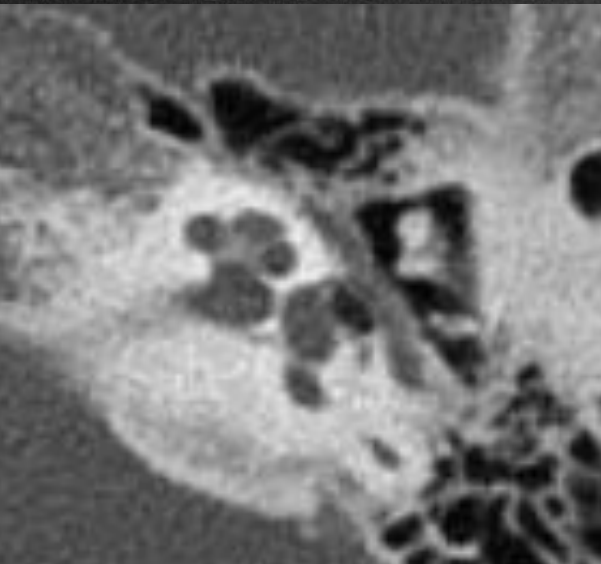


HEARING LOSS

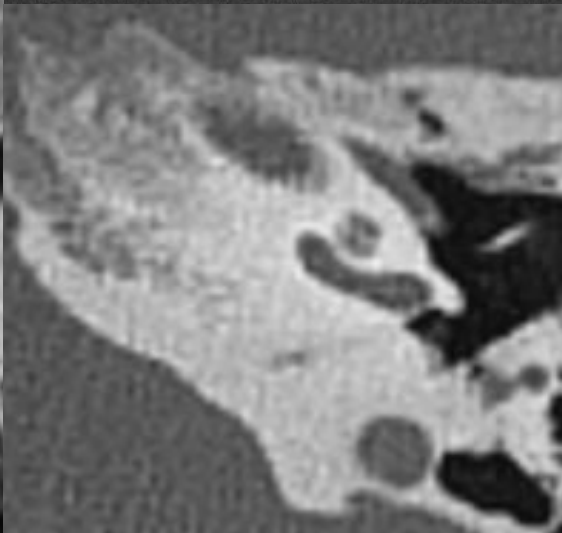
- Prevalence: 1,33 per 1000 live births
3,5 per 1000 adolescents
- Causes: environmental, genetic or mixed
- Hearing loss: sensorineural, conductive or mixed
- Syndromic 20%

Sensorineural hearing loss

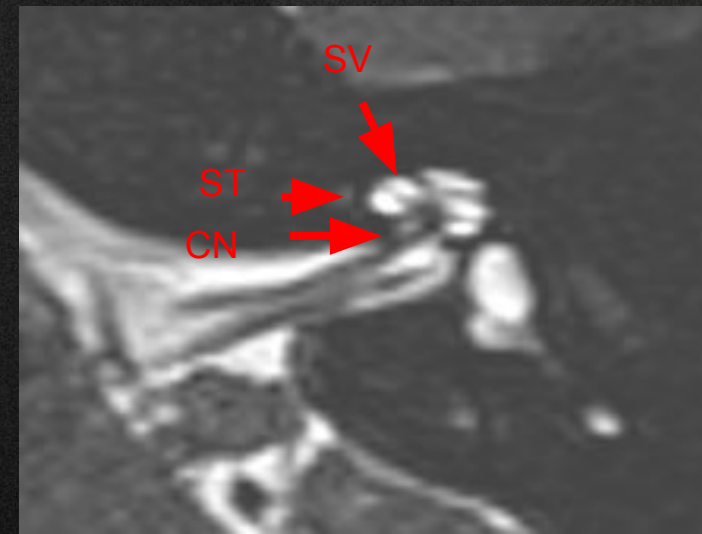
- Inner ear malformations
- 80% radiologically occult abnormality affecting the membranous labyrinth.



Mid modiolar view



Round window niche



ST
CN
SV

Inner ear malformations

Classification
proposed by
Sennaroglu

- **Complete Labyrinthine Aplasia**
- **Rudimentary Otocyst**
- **Cochlear Aplasia**
- **Common Cavity**
- **Cochlear hypoplasia - 4 types**
- **Incomplete partition type 1**
- **Incomplete partition Type 2**
- **Incomplete partition Type 2**
- **Enlarged vestibular aqueduct (EVA)**
- **Coclear aperture CN canal**
- **CN abnormalities**

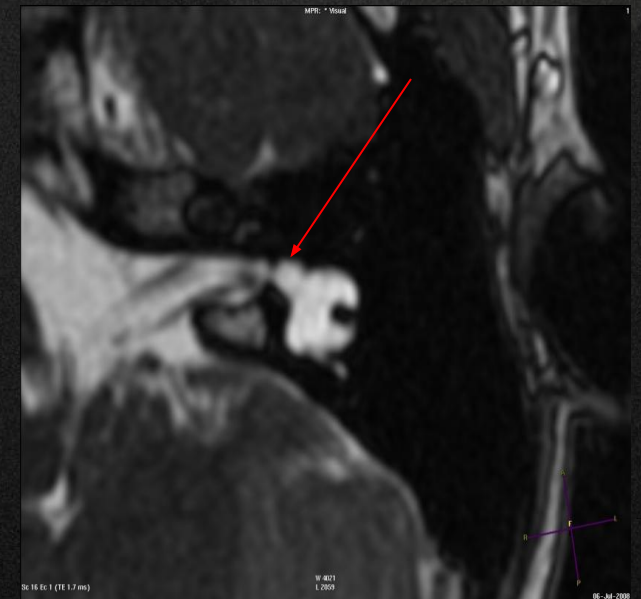
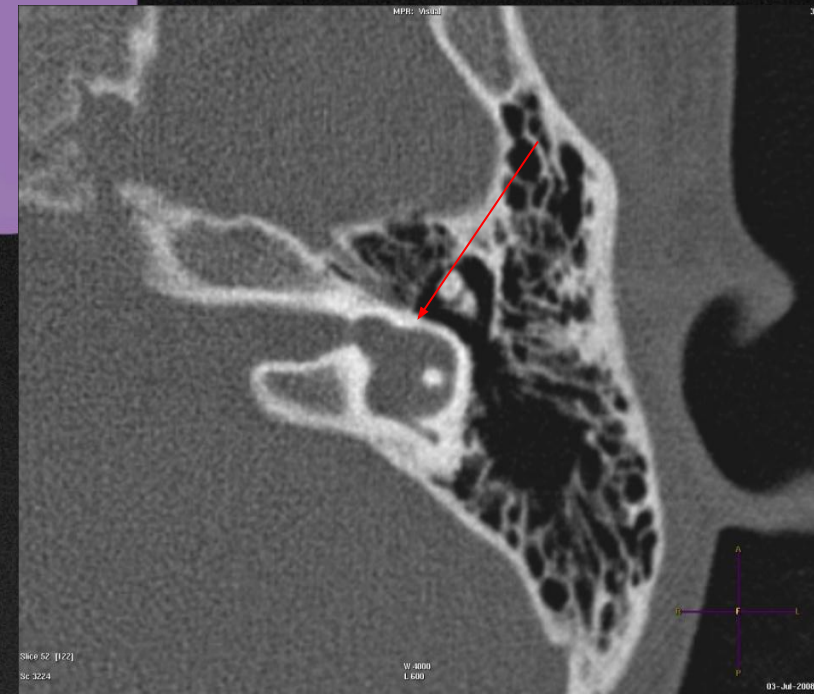
Complete Labyrinthine Aplasia

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- MICHEL DEFORMITY
- Complete absence of inner ear structures
- Internal auditory canal markedly reduced or absent
- Cochlear nerve absent
- Development of middle ear ossicles are usually normal.

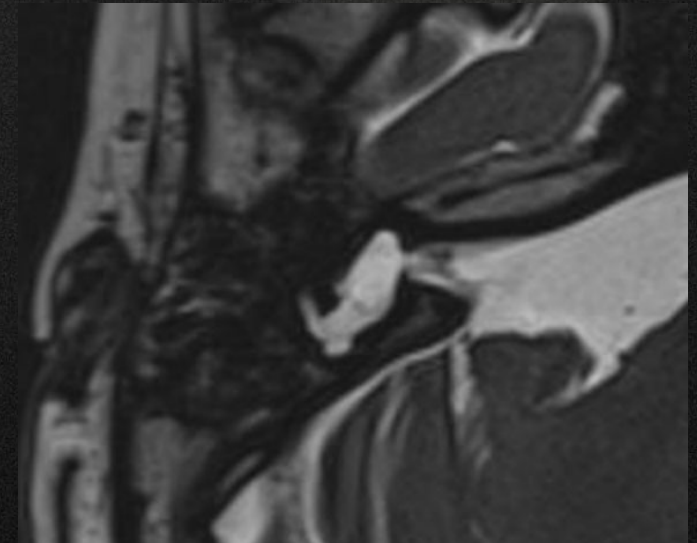
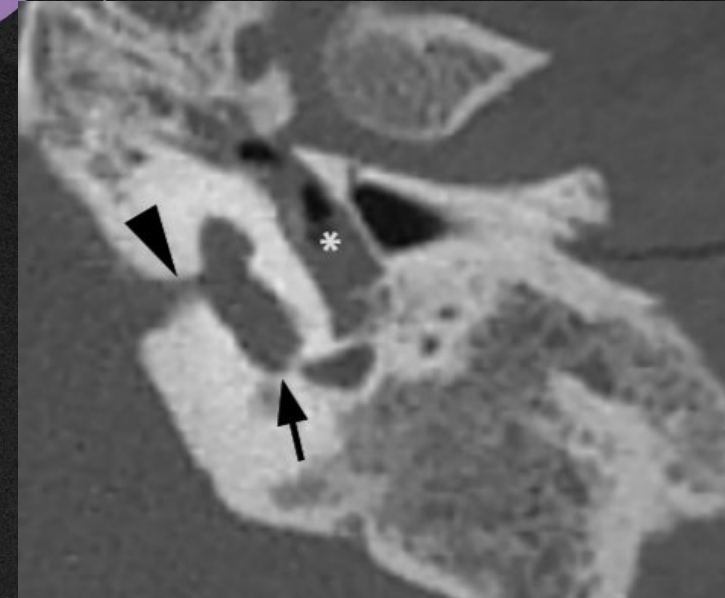
Cochlear Aplasia

- Absence of the cochlea.
- Vestibule and SCCs are in their normal anatomic location - at the posterolateral part of the fundus of IAC
- Vestibule may be normal or dilated



COMMON CAVITY

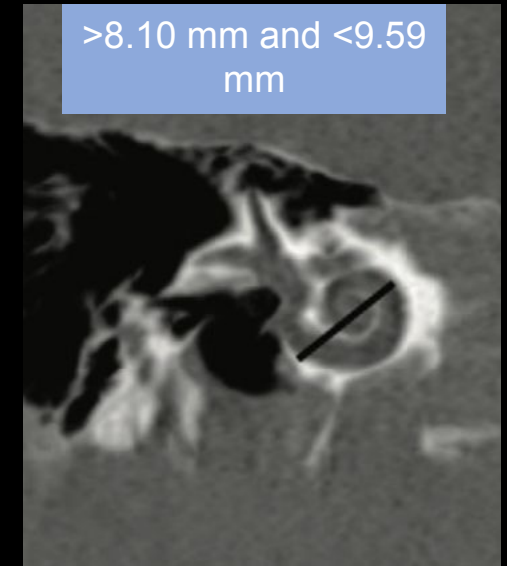
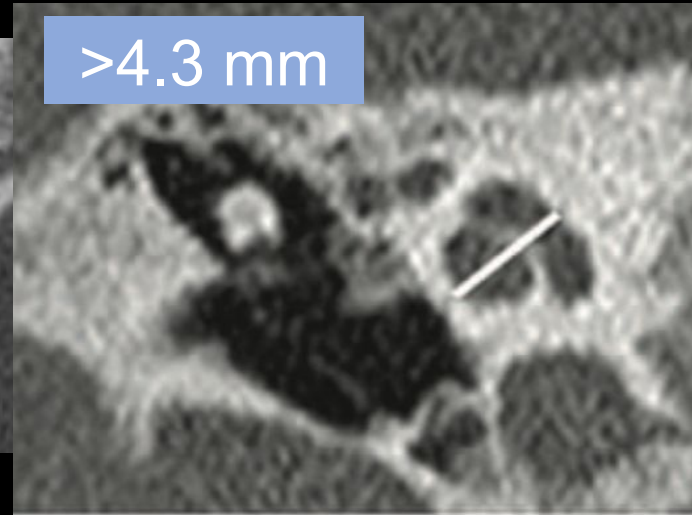
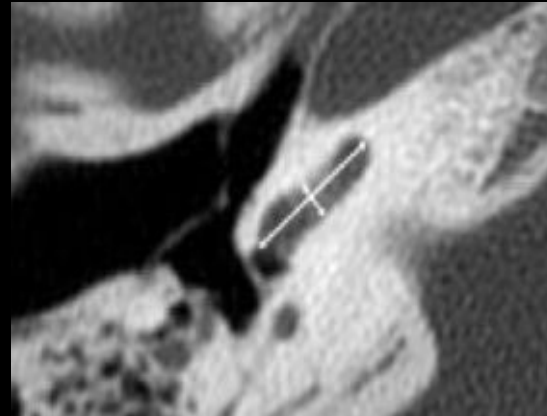
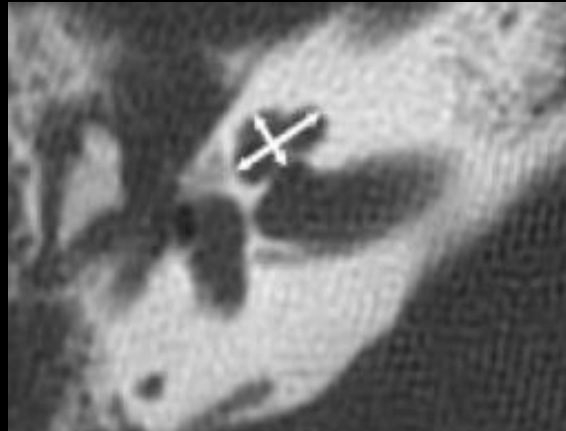
- A single, ovoid or round cavity, representing cochlea and vestibule
- Absence of differentiation between the cochlea and vestibule.
- The internal auditory canal (IAC) is positioned in the midportion of the cavity.
- It is important to differentiate a common cavity from cochlear aplasia with a dilated vestibule.
- The semicircular canals may appear normal, although they are usually malformed.



Cochlear Hypoplasia

- 15% - 23,5% of inner ear malformations
- Recognition of cochlear hypoplasia is essential during pre-implantation evaluation.
- Electrode array length should be adjusted according to cochlear size.

Measurements of the cochlea



Establishment of normative cochlear and vestibular measurements to aid in the diagnosis of inner ear malformations

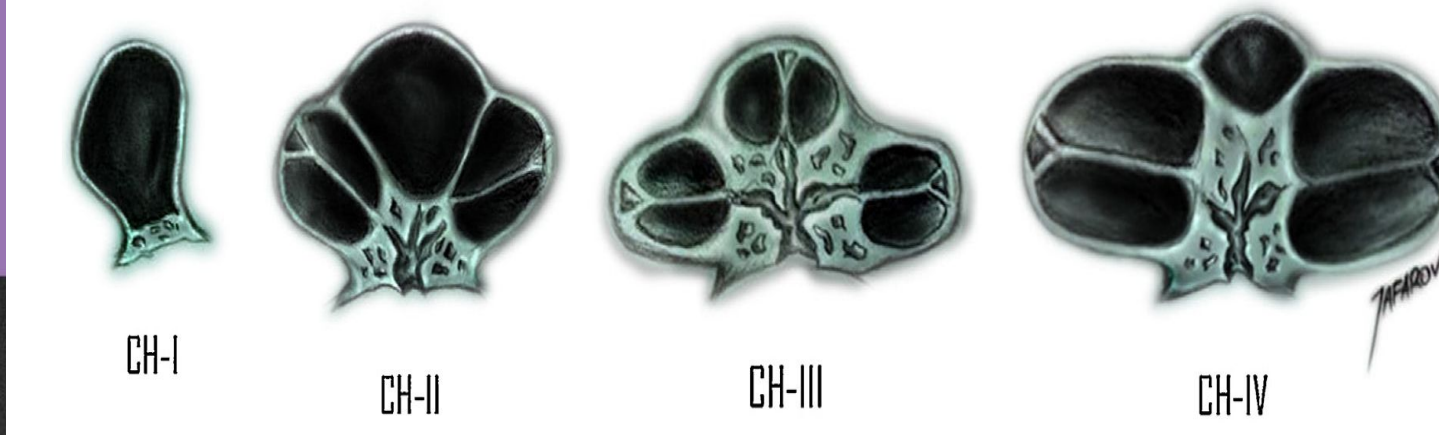
DERK PURCELL, BS, JACOB JOHNSON, MD, NANCY FISCHBEIN, MD, and ANIL K. LALWANI, MD, San Francisco, California

Do Measurements of Inner Ear Structures Help in the Diagnosis of Inner Ear Malformations? A Review of Literature

*Felice D'Arco, *Giacomo Talenti, *Rahul Lakshmanan, †Kate Stephenson, ‡Ata Siddiqui, and *Olivia Carney

Otolaryngol Head Neck Surg
. 2003 Jan;128(1):78-87.

Cochlear Hypoplasia



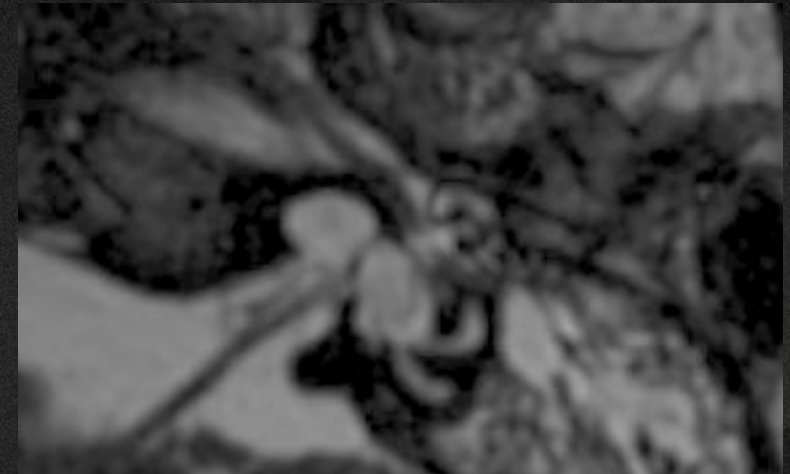
- CH-I Bud-like cochlea - small round or ovoid cochlea in shape, arising from the IAC
- CH-II - Absence of the modiolus and interscalar septum
- CH-III - The cochlea has fewer turns (less than 2 turns) with a short modiolus.
- CH-IV - The cochlea has a normal basal turn, but middle and apical turns are severely hypoplastic and located anterior and medially rather than in their normal central position.

INCOMPLETE COCHLEAR PARTITION-TYPE I

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CYSTIC COCHLEOVESTIBULAR MALFORMATION

- The modiolus is entirely absent; the cochlea has a cystic appearance; and the vestibule is often dilated.
- Basal turn of cochlea is not identified.
- Spontaneous CSF fistula and recurrent meningitis due to abnormal communication between the fundus of the internal auditory canal and the cochlea, in addition to a defect in the stapes footplate



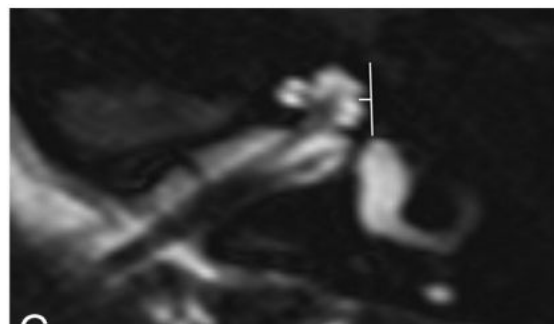
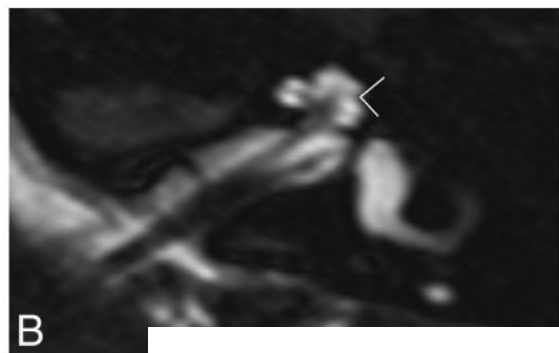
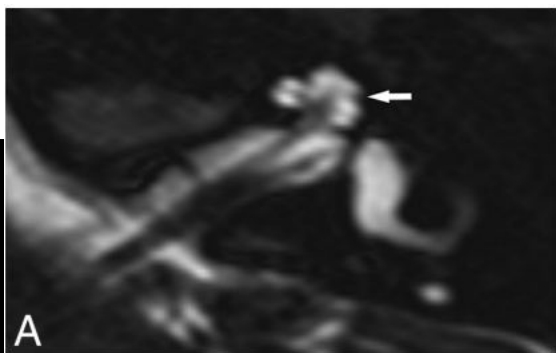
INCOMPLETE COCHLEAR PARTITION-TYPE II

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- Confluence of middle and apical turns of the cochlea to a deficiency of the apical part of the modiolus and the interscalar septa
- Normal basal turn of the cochlea
- Loss of the normal contour of the cochlear lateral wall
- Classic Mondini: type II partition, enlarged vestibule, and enlarged vestibular aqueduct.

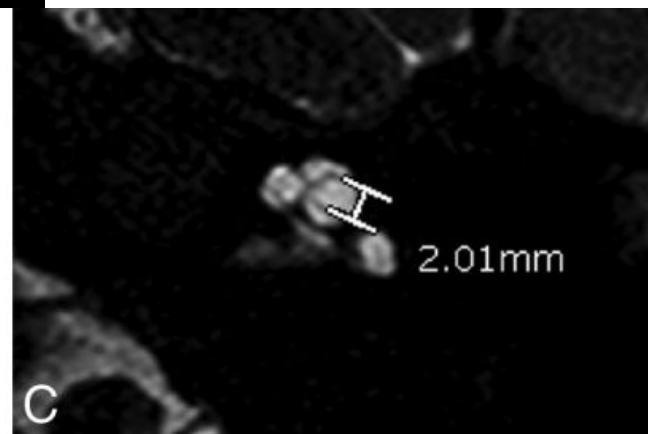
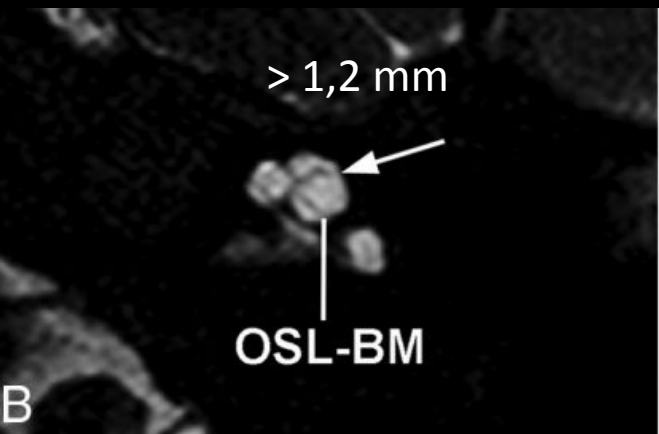
Evaluation of the Normal Cochlear Second Interscalar Ridge Angle and Depth on 3D T2-Weighted Images: A Tool for the Diagnosis of Scala Communis and Incomplete Partition Type II

T.N. Booth, C. Wick, R. Clarke, J.W. Kutz, M. Medina, D. Gorsage, Y. Xi, and B. Isaacson



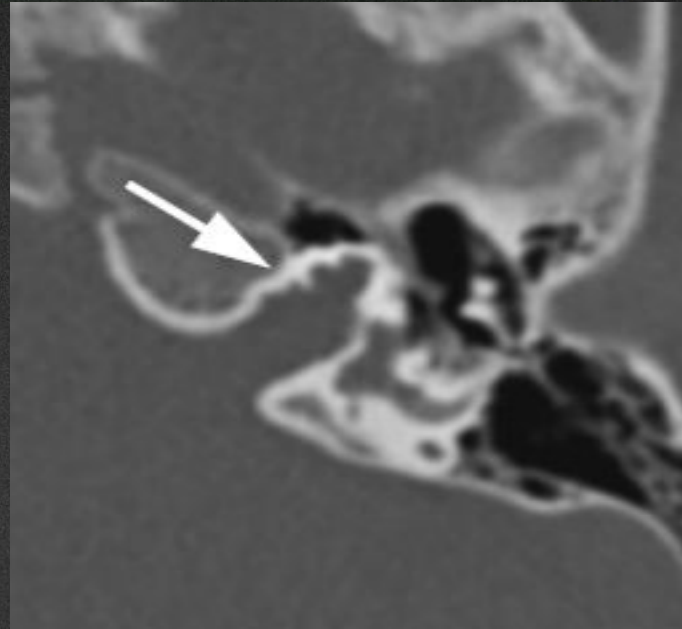
Measurement for Detection of Incomplete Partition Type II Anomalies on MR Imaging

K.L. Reinshagen, H.D. Curtin, A.M. Quesnel, and A.F. Juliano



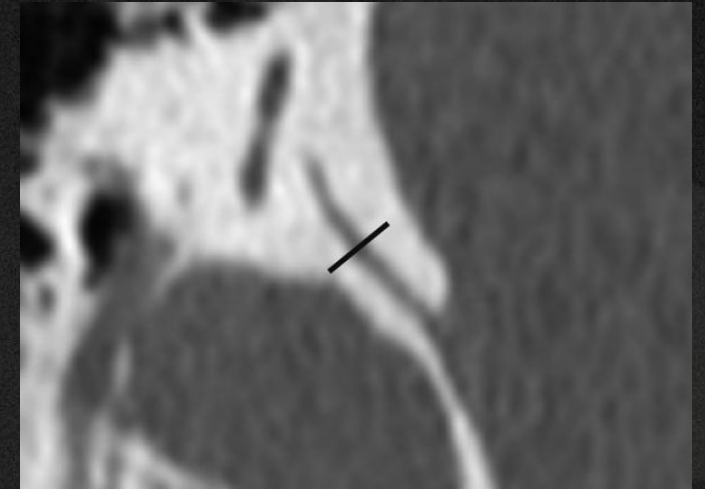
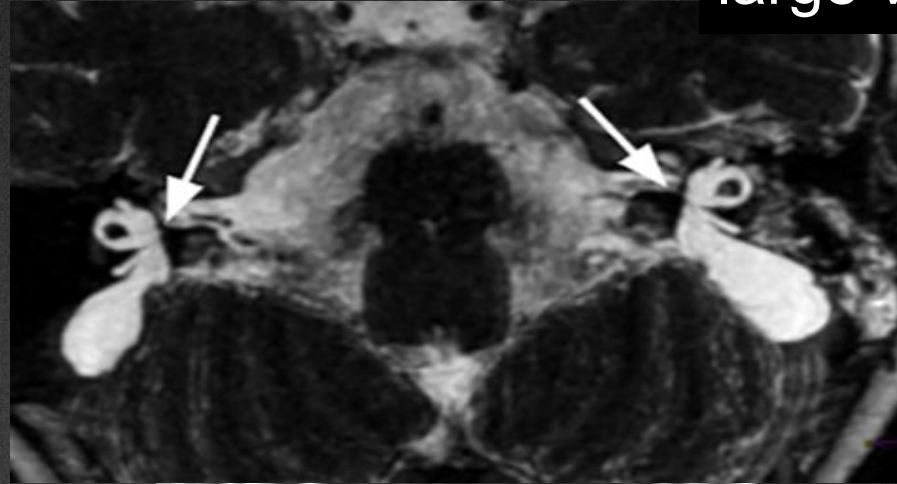
Incomplete Partition Type III

- X-linked deafness.
- Modiolus absent with a wide communication between the cochlea and the internal auditory canal (IAC).
- The otic capsule around the cochlea is thin.
- Enlargement of the IAC.
- Dilatation of the labyrinthine segment of the facial nerve



Enlarged Vestibular Aqueduct

Most common abnormal imaging findings in children with sensorineural hearing loss is a large vestibular aqueduct.



Measured in the oblique plane 45°
Normal between 0,3 and 0,9 mm

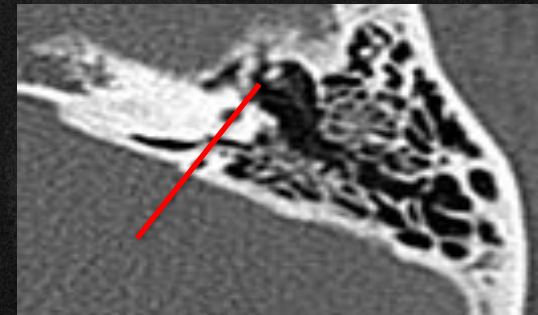
VALVASSORI CRITERION

Normal vestibular aqueduct up to 1.4 mm on the midpoint btw labyrinth and operculum.

CINCINNATI CRITERION

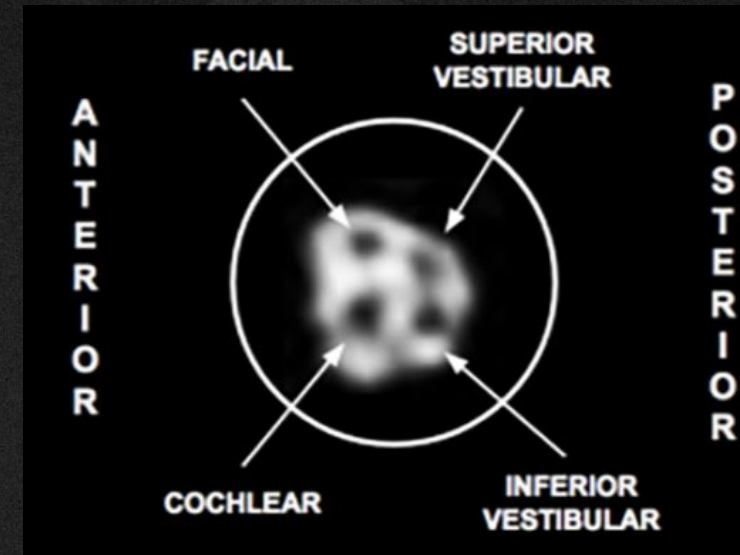
1.9 mm at the operculum
0.9 mm at its midpoint.”

Compared with the ascending portion of the posterior semicircular canal



Cochlear nerve deficiency

- Definition: absence or reduction in the caliber of the cochlear nerve.
- IAC may be also hypoplastic (less of 2,5 mm) or normal
- MRI is the method of choice to determine cochlear nerve integrity
- Normal Cochlear Nerve (CN): is **similar in size** to the ipsilateral facial nerve or slightly larger.
- Located in the anterior inferior part of the IAC
- CT: stenosis of cochlear nerve aperture (if less than 1,5 mm)



BRANCHIO-OTO-RENAL SYNDROME

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- EYA1 gene and SIX homolog family (AD)
- Hearing loss, auricular malformations, branchial arch closure defects and renal anomalies.
- Cochlear hypoplasia (type 4), deviation of the labyrinthine facial nerve canal medial to the cochlea
- Funnel-shaped IAC with a large porus acousticus
- Others findings: eustachian tube dilation, stenosis or atresia of the external auditory canal, middle ear and ossicular chain abnormalities

CHARGE SYNDROME

- **C**oloboma
- **H**ear defects
- **A**tresia of the choanae
- **R**etarded growth and development
- **G**enital hypoplasia
- **E**ar anomalies
- **CDH7** gene present in 2/3 of cases
- Low-set and misshaped pinnae
- Small tympanic cavity
- Ossicular chain: ossicular fixation, hypoplasia of the incus and stapes
- Absence of the round and oval windows
- **Inner ear:** SCC aplasia with associated vestibular dysplasia
- Cochlear nerve deficiency
- Abnormalities of cochlear partitioning

Waardenburg Syndrome

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- Lateral displacement of the inner canthi of the eye
- Heterochromia
- SNHL
- White forelock
- Imaging findings: **aplasia or hypoplasia of the posterior SCC**
- Vestibular aqueduct enlargement, widening of the upper vestibule, IAC hypoplasia, decreased modiolus size,



Pendred Syndrome

- Most common cause of syndromic congenital SNHL
- Thyroid endocrine abnormality and goiter
- Imaging findings:
 - Modiolar deficiency and vestibular enlargement (100%), absence of the interscalar septum between the
 - upper and middle cochlear turns (75%), and enlargement of
 - the vestibular aqueduct (80%).
 - Endolymphatic sac enlargement

Conductive hearing loss

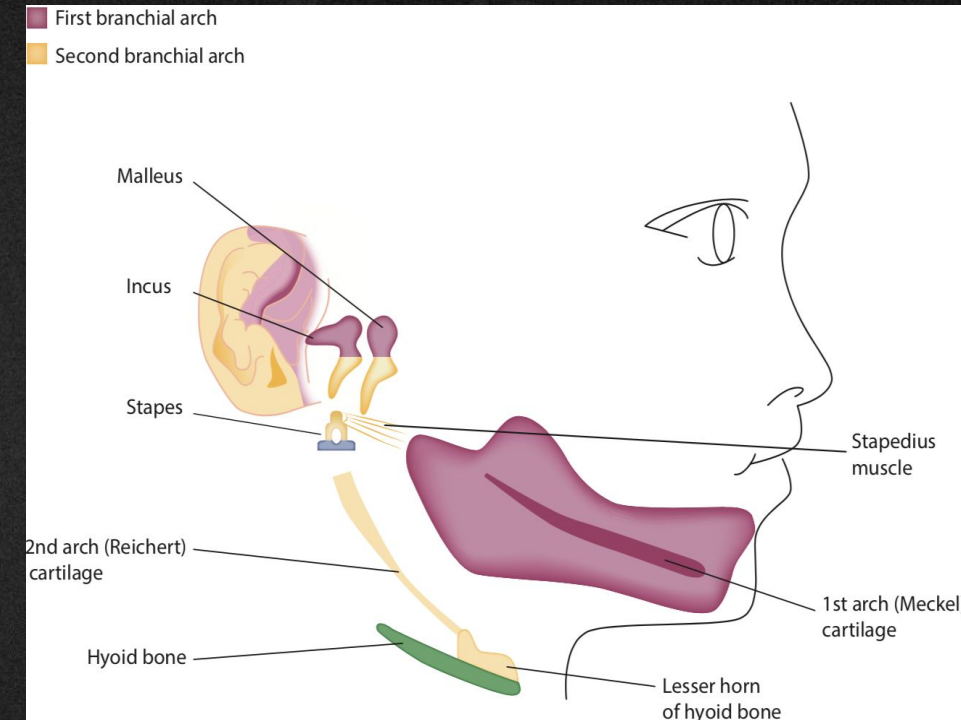
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- Congenital external and middle ear malformation
- Syndromes
- Chronic inflammatory disorders

External and middle ear malformation

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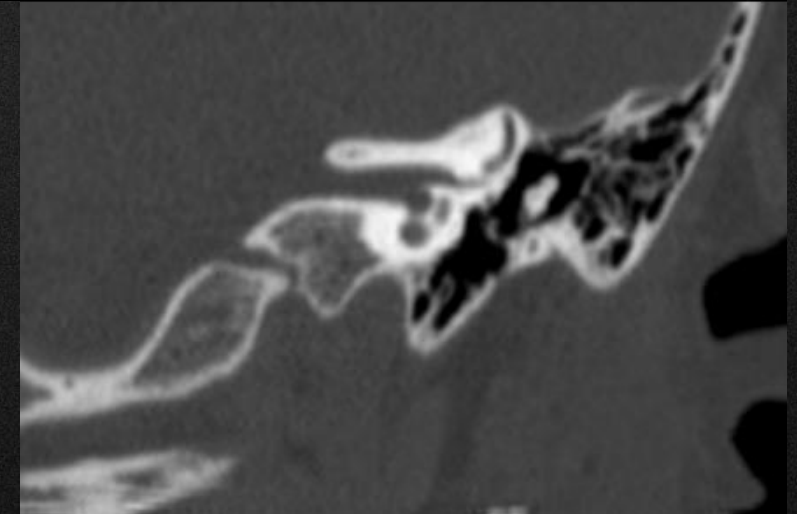
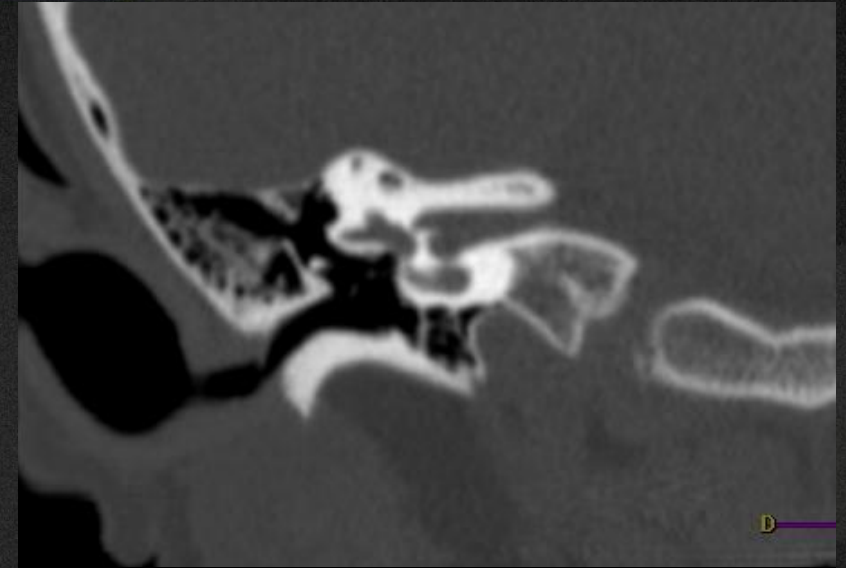
- The structures of the middle and external ear are derivatives of the first and second pharyngeal arches
- External ear anomalies are associated with malformations involving middle ear.
- Anotia, microtia, a misshapen pinna



External Auditory Canal Atresia

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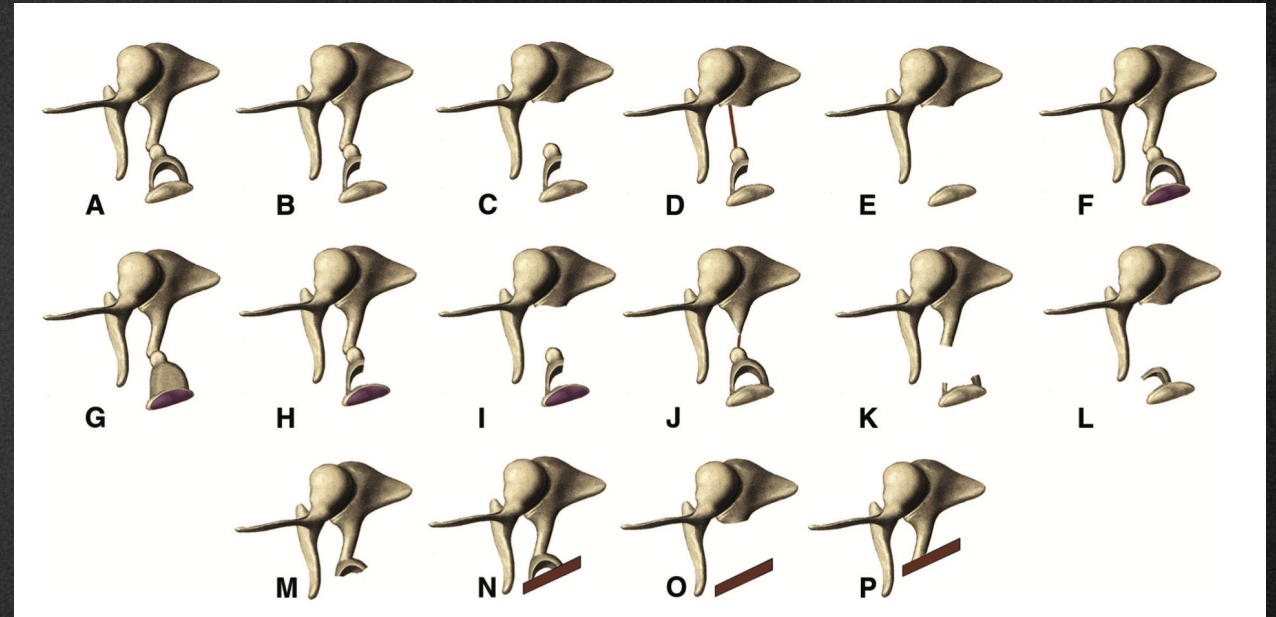
- Complete or incomplete atresia of the EAC
- May be bony or membranous
- Ossicular abnormalities:
 - malleus and incus may be fused together
 - malleus with a rudimentary handle.
- Stapes may be normal/dysmorphic or absent
- Tympanic segment of the facial nerve canal may be inferiorly displaced at the level of the round window and dehiscent
- Mastoid segment is usually anterolaterally displaced.



Ossicular Malformations

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- Rare cause of CHL
- Stape footplate fixation CT is normal
- Fixation of malleus
- Ankylosis malleus and incus
- Incudostapedial discontinuity



Neurographics 2016
September/October; 6(5):332–349

Hemifacial Microsomia and Oculoauriculovertebral spectrum

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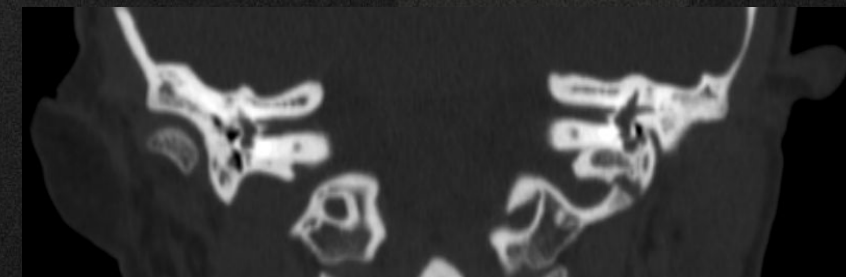
- Goldenhar syndrome
- Hypoplasia of the maxilla and mandible
- Temporomandibular joint deformities
- Isolated tags/pits/appendages
- Unilateral or bilateral stenosis or atresia of the external auditory canal, a small middle ear cavity, ossicular malformation, and atresia or stenosis of the oval window.
- Inner ear anomalies also sometimes occur in patients with GS.



Treacher Collins Syndrome

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- Downward slanting of the palpebral fissures, eyelid coloboma, micrognathia, microtia and other ear deformities, facial bone hypoplasia, involving mandible, zygomatic complex and maxilla. Conductive hearing loss and palate cleft are often present



Cornelia de Lange Syndrome

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External auditory canal stenosis

Cochlear hypoplasia

Obliteration of the tympanic cavity

Malformation of the malleus and incus

Incudial cleft

Facial nerve dehiscence

Vestibular dysplasia with medial protrusions

Hearing Loss

- Hearing loss has a wide range of causes.
- Imaging plays a crucial role in the detection and classification of anomalies, allowing proper surgical planning, especially prior to cochlear implantation.

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eloisa.gebrim@hc.fm.usp.br



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