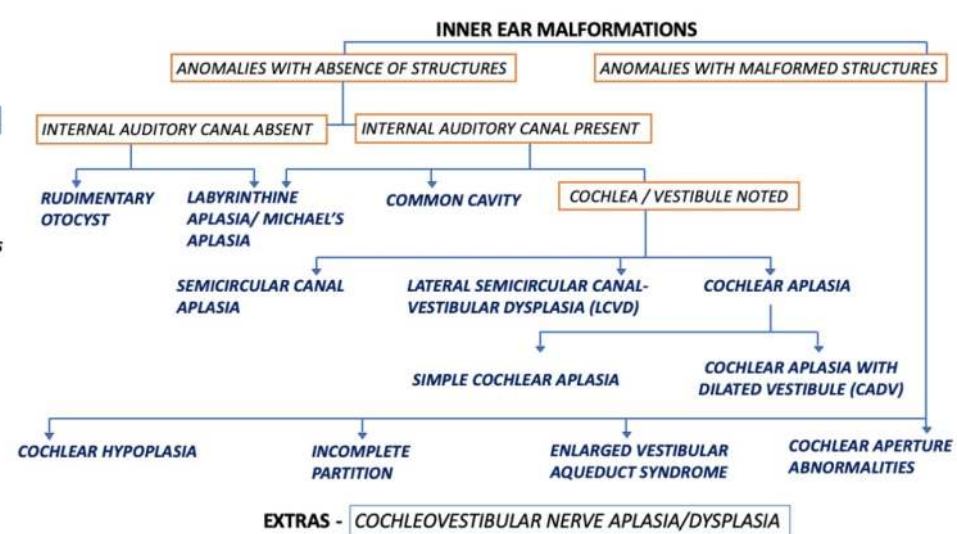
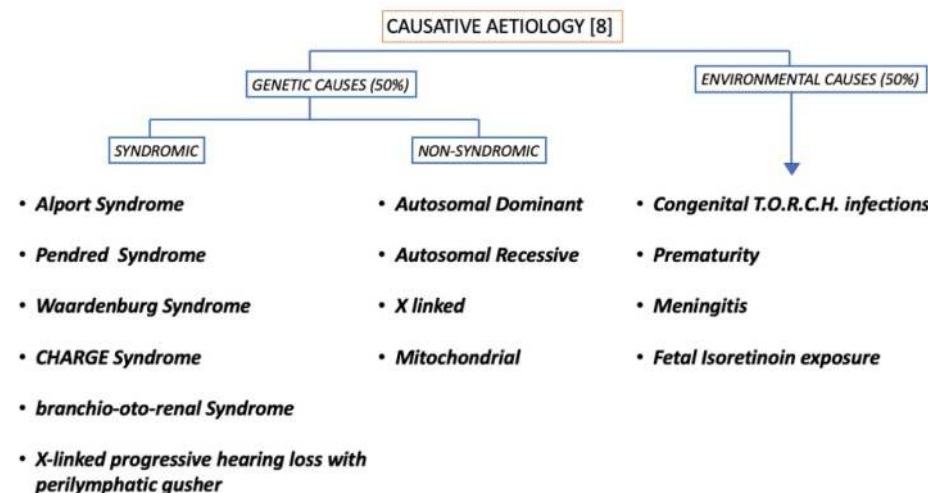


INNER EAR MALFORMATIONS: MAKING WAY THROUGH THE LABYRINTH

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LEARNING OBJECTIVES

- To synthesize an *easy-to-remember Imaging Approach* to Inner Ear malformations (IEM)
- To establish confident and reproducible *parameters for evaluation* of IEM on CT/MR
- To *differentiate between difficult imaging differentials* like Cochlear Hypoplasia Type IV vs Incomplete Partition Type II, Cochlear Hypoplasia Type II vs Incomplete Partition Type I, Common Cavity vs Cochlear Aplasia with Dilated Vestibule.
- To assess *imaging predictors of poor post-surgical outcome*
- To enumerate and showcase common syndromic associations of IEM.



PLANES/VIEWS FOR EVALUATION

STENVER PLANE

Orientation: Axis of modiolus/perpendicular to Superior SCC axis.

Use: Integrity of bony plate over Superior Semicircular Canal
Cross-sectional Evaluation of the Cochlea and Facial Nerve

COCHLEAR VIEWS

MID-MODIOLAR VIEW

Orientation: Axial Plane at the level of modiolus

Use: Visualization of the two parts bony modiolus (medial and lateral) and cochlear aperture.
Characterization of IP abnormalities

ROUND WINDOW NICHE VIEW

Orientation: Just inferior to the Mid-Modiolar View

Use: Visualization of two parts of inter-scalar septum (medial and lateral)
Characterization of IP abnormalities

POSCH'S PLANE

Orientation: Along the Petrous Ridge

Use: Evaluation of the vestibular aqueduct

PARAMETERS OF EVALUATION I

COCHLEAR HEIGHT

Coronal Section

Normal: 5.3 mm with SD - 0.5 mm

Objective marker of cochlear hypoplasia

COCHLEAR APERTURE

Axial Section: Mid-modiolar view

Normal: 2.1 mm with SD - 0.4 mm

Predictor of cochlear nerve aplasia
Prognostication of future implant surgery

INTERNAL AUDITORY CANAL

Axial Section with longest view

Stenotic when < 2 mm.

Predictor of cochleo-vestibular ne aplasia

LENGTH and WIDTH OF BASAL TURN

Axial Section

Length: 8.6 mm with SD - 0.4 mm
Width: 2.1 mm with SD - 0.1 mm

Objective marker of cochlear hypoplasia

HEIGHT and WIDTH OF APICAL TURNS

Axial Section: Mid-modiolar view

Height: 3.2mm with SD - 0.4 mm
Width: 4 mm with SD - 0.4 mm

Objective marker of cochlear hypoplasia

VESTIBULAR AQUEDUCT/ ENDOLYMPHATIC DUCT

Posch's Plane

Dilated when >0.8 mm in this plane / wider than posterior semicircular cana

Identification of Enla Aqueduct Syndrome
Identification of "Trag ear" and post-op complications (CSG G)

PARAMETERS OF EVALUATION II

WIDTH OF POSTEROMEDIAL LATERAL SCC

Axial Section

Enlarged when > 1.8 mm.

Identification of semicircular canal malformation.
Identification of "fragile ear" and post-op complications (CSF Gusher)

ANGLE BETWEEN TYMPANIC AND LABYRINTHINE SEGMENTS OF FACIAL NERVE

Axial Section

Abnormal when obtuse angled

Identification of "fragile ear" and post-op complications (CSF Gusher)

BONY ISLAND WITHIN LATERAL SCC

Axial Section

Dilated vestibule causes lateral semicircular canal assimilation
Enlarged vestibule - area of bony island < 6 mm²

Identification of Lateral Semicircular Canal - Vestibular Dysplasia

ENLARGED VESTIBULE

Axial Section

Length: 5.8 mm with SD - 0.6 mm
Width: 3.4 mm with SD - 0.2 mm

Identification of isolated enlargement of the vestibule without assimilation of lateral semicircular canal (Eg: Thalidomide)

MALFORMATIONS

ANOMALIES WITH ABSENCE OF STRUCTURES

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    graph TD
        AWAS[ANOMALIES WITH ABSENCE OF STRUCTURES] --> IACA[INTERNAL AUDITORY CANAL ABSENT]
        AWAS --> IACP[INTERNAL AUDITORY CANAL PRESENT]
        IACA --> RO[RUDIMENTARY OTOCYST]
        IACA --> LA[LABYRINTHINE APLASIA/ MICHAEL'S APLASIA]
        IACP --> CC[COMMON CAVITY]
        IACP --> CV[COCHLEA / VESTIBULE NOTED]
        CV --> SC[SEMICIRCULAR CANAL]
        CV --> LSCV[LATERAL SEMICIRCULAR CANAL-VESTIBULAR DYSPLASIA (LCVD)]
        CV --> CA[COCHLEAR APLASIA]
    
```

DIFFERENTIAL

- RO:** Ovoid structure in inner ear Semicircular canals +/-
- LA:** No inner ear structure visualized
- CC:** An ovoid structure replaces inner ear region.
Internal Auditory Canal meets the ovoid structure in CENTRE.
Semicircular canals +/-
Facial Nerve abnormal course.
- CV:** An ovoid structure replaces inner ear region.
Ovoid structure POSTEROLATERAL to the Internal Auditory Canal
Semicircular canals +/-
Facial Nerve abnormal course.
COCHLEAR APLASIA WITH DILATED VESTIBULE

ALL CANALS - CHARGE SYNDROME
Superior Semicircular Canal - Alagille, Waardenburg Syndrome
Posterior Semicircular Canal - Thalidomide toxicity

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        CV --> CA[COCHLEAR APLASIA]
    
```

DIFFERENTIAL

- SC:** One/None semicircular canal identified, except lateral semicircular canal.
- LSCV:** Lateral Semicircular Canal not identified. Other canals normal.
- CA:** Cochlea not identified. Facial Nerve anteriorly displaced.

COCHLEOVESTIBULAR NERVE APLASIA/DYSPLASIA

TYPE I

- UNDIVIDED COCHLEOVESTIBULAR NERVE "UCN"
- ABERRANT COCHLEAR BRANCH "ACB"
- HYPOPLASTIC COCHLEAR BRANCH "HCB"

TYPE II

- Normal vestibule
- Simple Cochlear Aplasia
- Dilated vestibule
- Cochlear Aplasia with Dilated Vestibule (CADV)
- Common Cavity

MALFORMATIONS

ANOMALIES WITH MALFORMED STRUCTURES

STRUCTURE	TYPE I	TYPE II / MONDINI	TYPE III / X LINKED DEAFNESS
A. Cochlea	Normal in size, but CYSTIC	Normal in size	Normal in size
B. Modiolus	NOT VISUALIZED	NOT VISUALIZED (AT APEX)	NOT VISUALIZED
C. Inter-scalar Septum	NOT VISUALIZED	NOT VISUALIZED (AT APEX)	NOT VISUALIZED
D. Vestibule	DILATED	Normal	Normal
E. Vestibular Aqueduct	Normal	DILATED	Normal
F. Semicircular Canals	Normal	Normal	Normal
G. Internal Auditory Canal	Normal	Normal	Normal
H. Stapes Footplate	DEFECTIVE (encloses a cystic structure - characteristic)	Normal	Normal
I. Facial Nerve	Normal	ANTERIORLY DISPLACED	SUPERIORLY DISPLACED
J. Cochleo-Vestibular Nerve	Normal/Absent	Normal/Absent	Normal/Absent

MALFORMATIONS

ANOMALIES WITH MALFORMED STRUCTURES

COCHLEAR APERTURE ABNORMALITIES

- Cochlea normal in size, morphology but **APERTURE < 1.4mm**
- Associated IAC narrowing +/-
- Vestibule and aqueduct, Semicircular Canals, Internal Auditory Canal, Facial and Cochleo-Vestibular Nerves NORMAL

ENLARGED VESTIBULAR AQUEDUCT SYNDROME

- Vestibular Aqueduct Enlarged
- Cochlea, Vestibule, Semicircular Canals, Internal Auditory Canal, Facial and Cochleo-Vestibular Nerves NORMAL

DIFFERENTIAL

Type A: Labyrinth abnormal.

Type B: Labyrinth normal.

MALFORMATIONS

ANOMALIES WITH MALFORMED STRUCTURES

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MALFORMATIONS

ANOMALIES WITH MALFORMED STRUCTURES

COCHLEAR HYPOPLASIA

Abnormal cochlea morphology and **DECREASED** size

STRUCTURE	TYPE I	TYPE II
Cochlea	BUDS FROM I.A.C.	SMALL AND CYSTIC
Modiolus	ABSENT	ABSENT
Inter-scalar Septum	ABSENT	ABSENT
Vestibule	Normal	Normal
Vestibular Aqueduct	Normal	DILATED
Semicircular Canals	Normal	Normal
Internal Auditory Canal	Normal	Normal
Stapes Footplate	Normal	DEFECTIVE
Facial Nerve	NORMAL/ABSENT	NORMAL/ABSENT
Cochleo-Vestibular Nerve	NORMAL/ABSENT	NORMAL/ABSENT

MALFORMATIONS

ANOMALIES WITH MALFORMED STRUCTURES

COCHLEAR HYPOPLASIA

Abnormal cochlea morphology and **DECREASED** size

STRUCTURE	TYPE III	TYPE IV
Cochlea	SMALL IN SIZE (Decreased number of turns)	SMALL IN SIZE, WITH ANTEROMEDIAL DISPLACED (Hypoplastic Apical Turns)
Modiolus	POSITIVE	POSITIVE
Inter-scalar Septum	POSITIVE	POSITIVE
Vestibule	NORMAL/HYPOPLASTIC	Normal
Vestibular Aqueduct	Normal	Normal
Semicircular Canals	NORMAL/HYPOPLASTIC	Normal
Internal Auditory Canal	Normal	Normal
Stapes Footplate	FIXED	Normal
Facial Nerve	Normal	ANTERIORLY DISPLACED
Cochleo-Vestibular Nerve	NORMAL/ABSENT	NORMAL/ABSENT

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Internal Auditory Canal	Normal	Normal
Stapes Footplate	FIXED	Normal
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I. Facial Nerve	Normal	ANTERIORLY DISPLACED
J. Cochleo-Vestibular Nerve	NORMAL/ABSENT	NORMAL/ABSENT

	RIGHT	LEFT
Pinna		
Tympanic Membrane		
EAC - Course		
Caliber		
Morphology		
Canalization		
Middle ear cavity size		
Epi tympanum		
Mesotympanum		
Hypotympanum		
Ear ossicles - Density		
Integrity		
Outline		
Foot Plate of Stapes		

	RIGHT	LEFT
Foot Plate of Stapes		
Joints- Incudo-malleal		
Incudo-stapedial		
Facial Recess		
Sinus Tympani		
Mastoid Air cells-Grade of Pneumatization		
Type of Pneumatization		
Aditus Patani?		
Korner's Septum seen?		
Tegmen Tympani		
Tegmen Mastoideum		
Smoothen Plate		
Round and Oval Window		

	RIGHT	LEFT
Cochlea - Dimensions		
Location in relation to IAC		
No. of Turns		
Basal turn width		
Modiolus		
Interscalar Septum		
Aperture width		
Vestibule		
Lateral Semicircular Canal - Normal/Dilated		
Area of Bony Island within		
Ampulla of semi-circular canal		
Posterior Semicircular Canal		
Superior Semicircular Canal		