

All About the Temporal Bone: From Embryology to Congenital Pathology

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Disclosures

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Learning objects

1. Embryology & development of temporal bone
2. Normal anatomy of temporal bone
3. Radiologic findings and clinical features of major congenital abnormalities
 - 1) Inner ear abnormalities
 - 2) middle ear abnormalities
 - 3) External ear abnormalities
4. Take home notes

1. Embryology & development

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Embryology

1) Inner ear

- Membranous labyrinth
- Perilymphatic labyrinth
- Bony labyrinth

2) Middle ear

- Tympanic cavity
- Ossicles
- Eustachian tube

3) External ear

- Tympanic membrane
- EAC, Auricle

1) Inner ear development

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Embryology

- First one to develop
- From 2 – 3 weeks of gestational age
 - Otic placode → Otic pit → Otic vesicle
- Main three phases
 - ① Development (3rd – 8th weeks of gestational age)
 - ② Growth (8th – 16th weeks)
 - ③ Ossification (16th – 24th weeks)

1) Inner ear development

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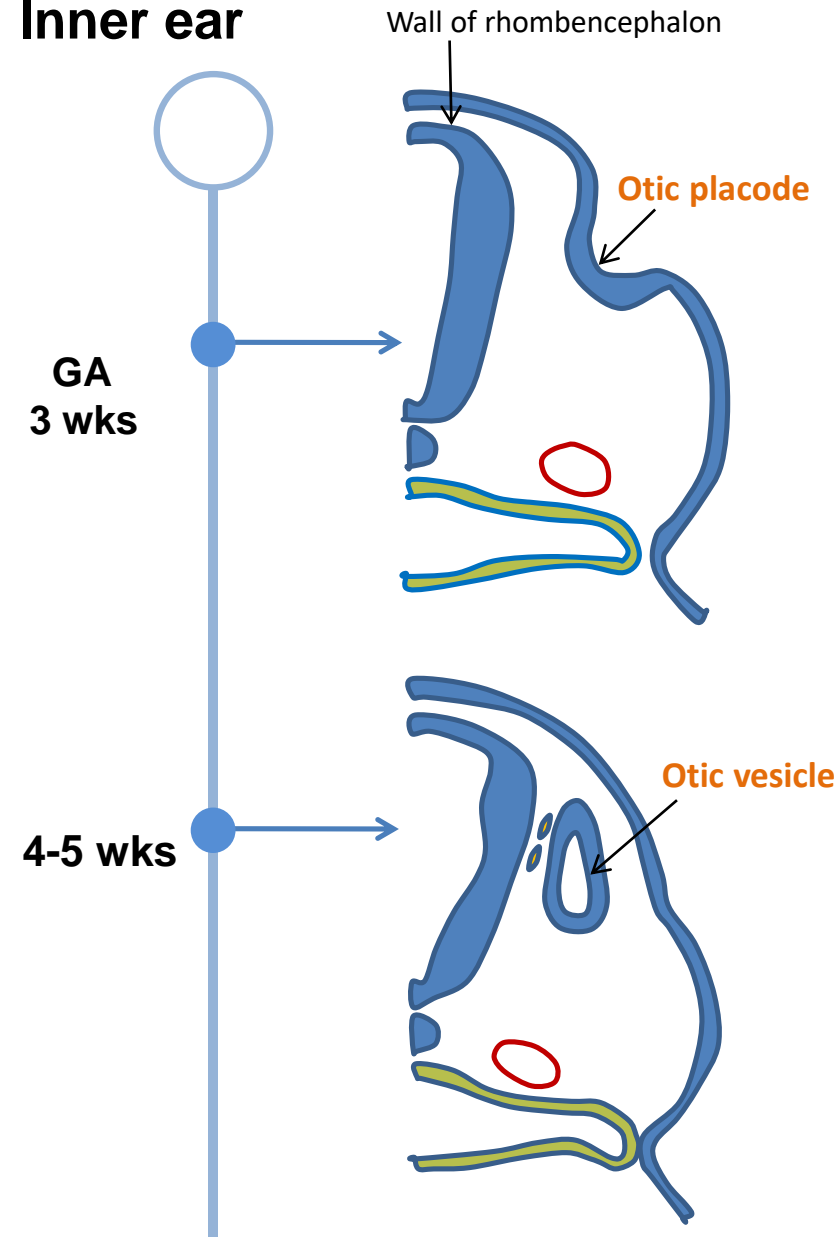
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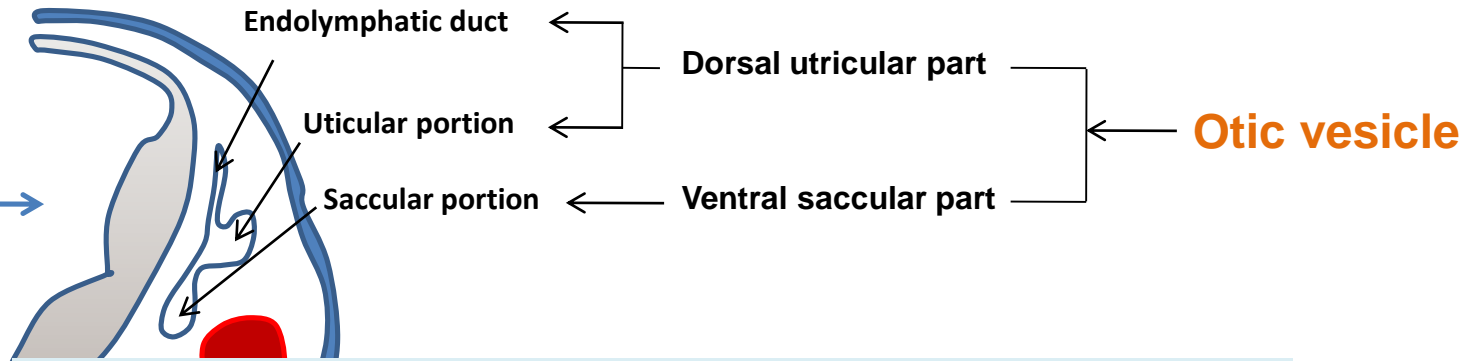
Embryology

Inner ear



- **Otic placode** from neuroectoderm
 - Invaginating from surface
 - Loss of connection from surface
 - Otic vesicle
- **Otic vesicle (otocyst)**
 - Constitutes primitive endolymphatic and membranous labyrinth

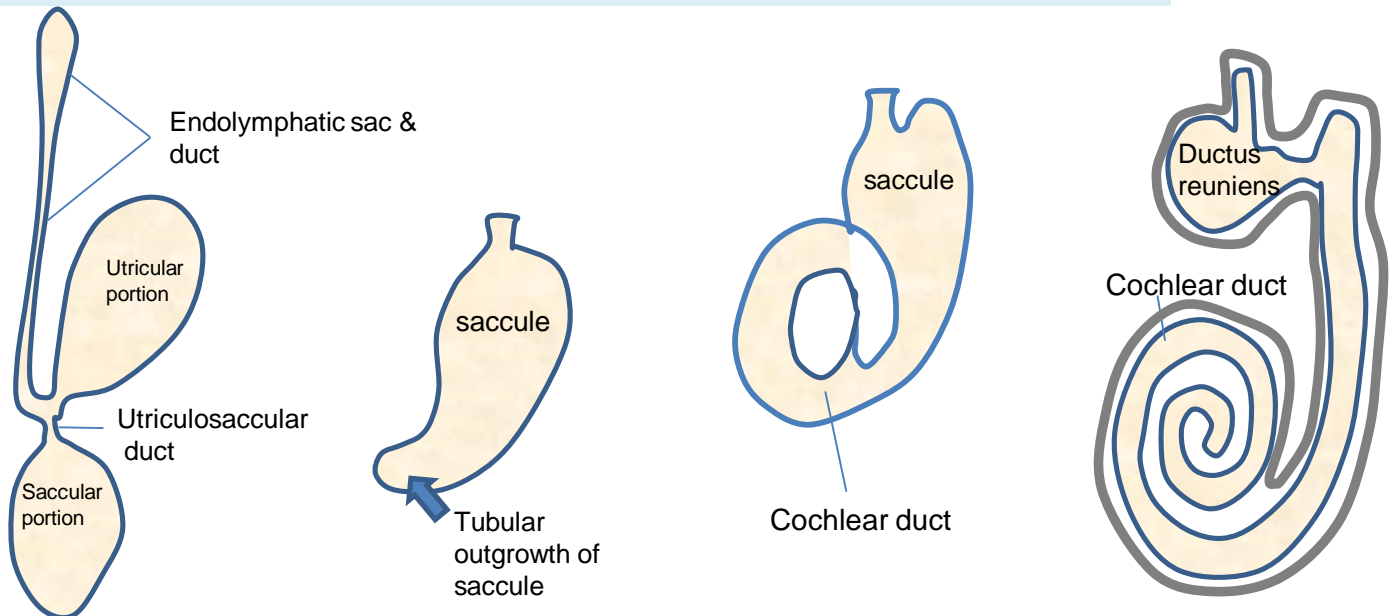
7 wks



- **Membranous labyrinth**

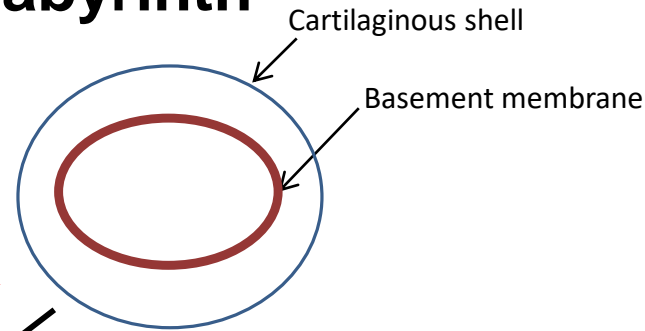
- Differentiate to cartilage encasing entire labyrinth
- Cartilaginous otic capsule ossifies
 - : But, endolymphatic duct & sac continues to change
 - : Mature anatomic configuration : Mid-term of fetal age (7 months)

7~8 wks



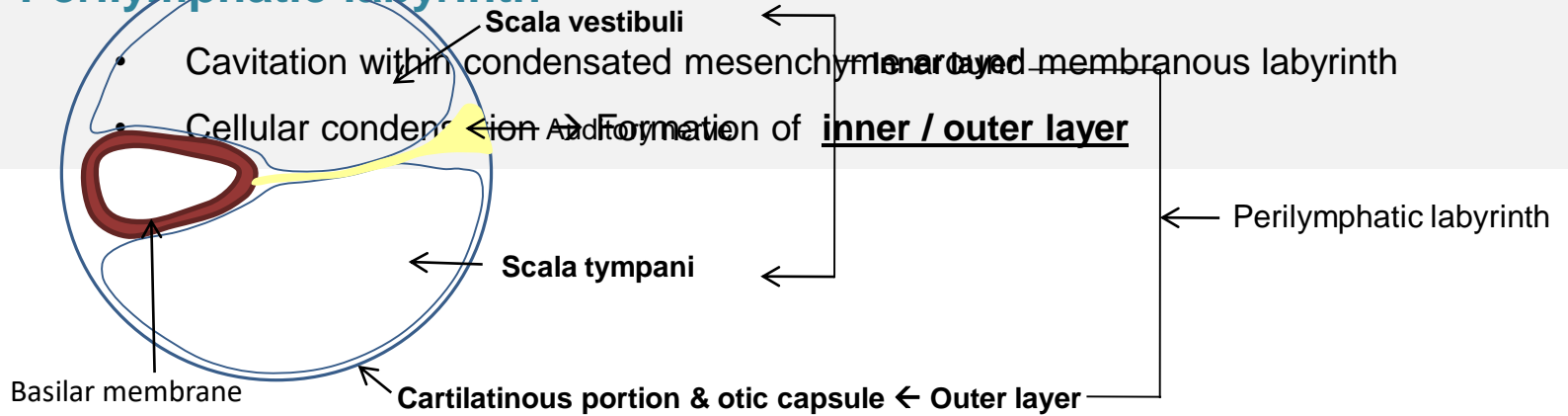
Perilymphatic labyrinth

6~8 wks



Cross section
of otic vesicle

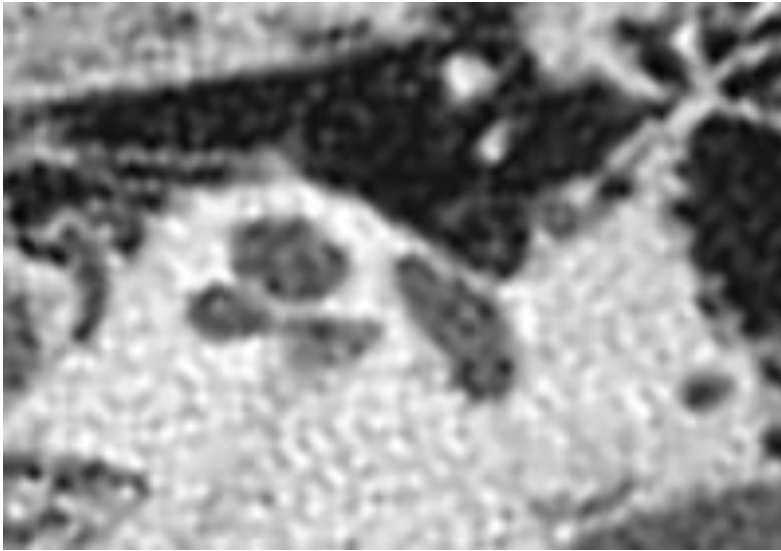
Perilymphatic labyrinth



16 wks

- Other 3 perilymphatic spaces into surrounding osseous otic capsule
- Cochlear aqueduct (perilymphatic duct)
 - : From scala tympani near round window to subarachnoid space
- Small fissula ante fenestram / Fossula postfenestram
 - : Focus of diseased bone in otosclerosis

- **Bony labyrinth**



- Ossification of otic capsule (16 – 23 weeks of fetal life)
- **Enchondral bone formation** → Inner ear structures cease to grow
- Limited osteogenic repair → Labyrinthine fistula when injured
- Membraneous bone
 - : Modiolus (communication with IAC) & interscalar septum
- Exception of three sites:
 - 1) Oval window
 - 2) areas of fissula ante fenestram,
 - 3) lateral most portion of lateral SCC

16~ 23
wks

2) Middle & external ear development

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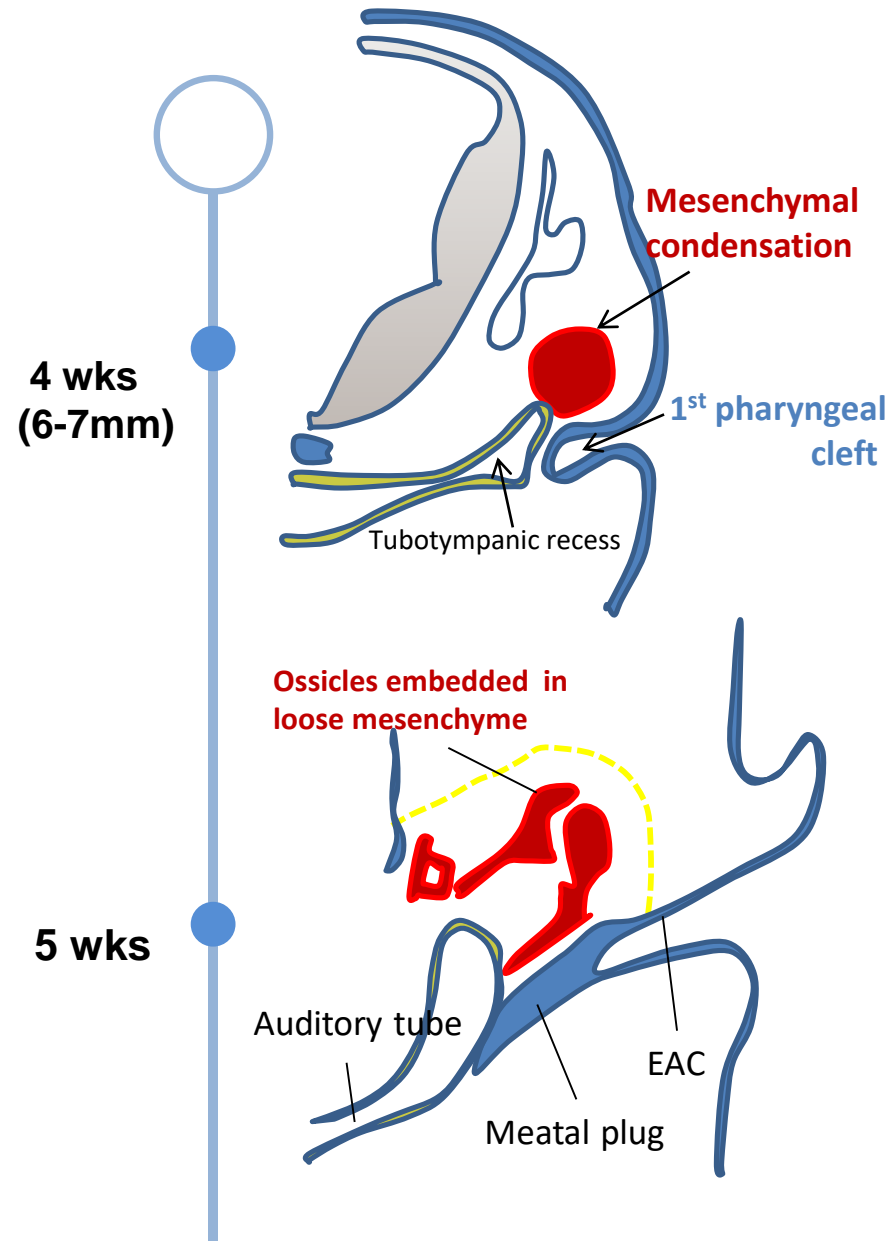
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Embryology

External ear development

- Begins at 3rd month of GA
- **External auditory meatus**
 - From **1st pharyngeal cleft**
 - Ectodermal cell proliferation
 - Meatal plug
 - Dissolved & form external auditory meatus
- **Tympanic membrane**
 - From 1st pharyngeal membrane btw cleft & pouch
- **Auricle**
 - From proliferation six mesenchyme in 1st & 2nd pharyngeal arches
 - Swelling & fusion to form auricle



- **Neonatal temporal bone**
 - Fully developed anatomic portions of hearing/vestibular system
Exception of osseous portion of EAC
 - Large squamous portion, diminutive tympanic portion
Formation & maturation of tympanic / mastoid portions
 - Pneumatization stimulated by middle ear aeration after birth
- **IAC**
 - Nearly adult vertical dimension at birth (> 4mm)
 - Length : increasing during childhood

2. Normal anatomy

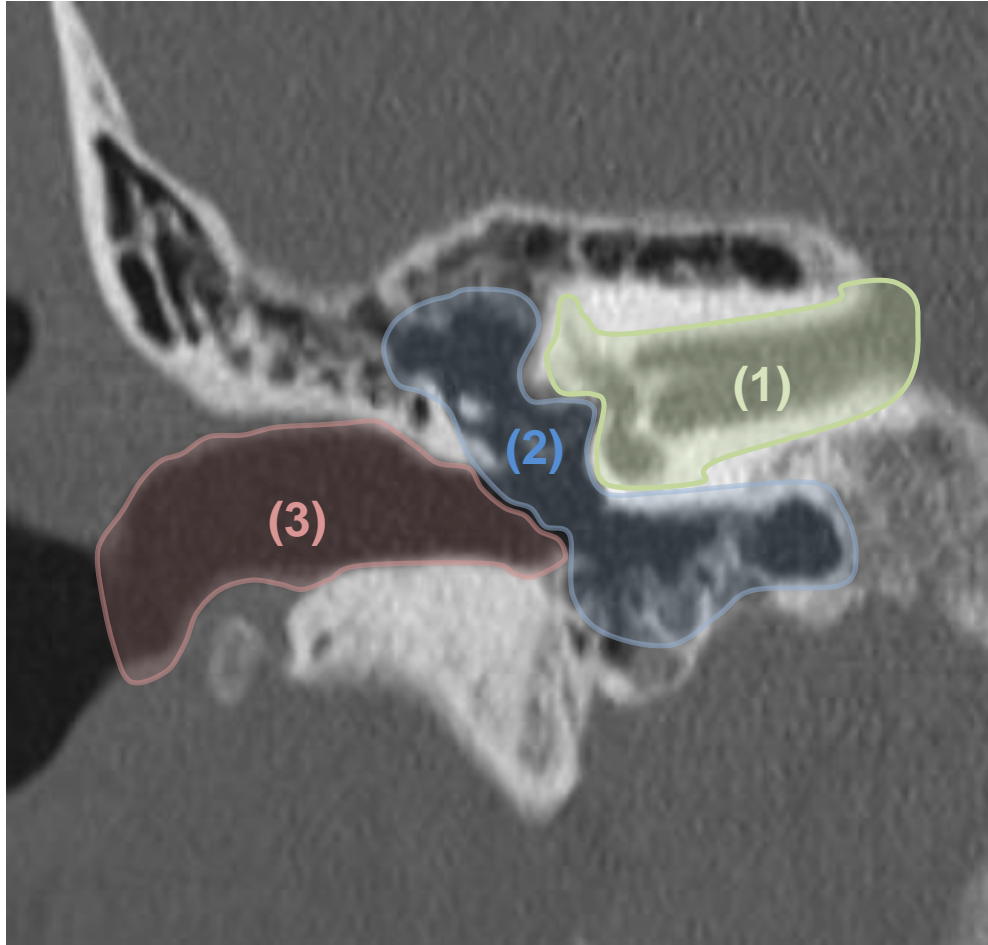
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Anatomy



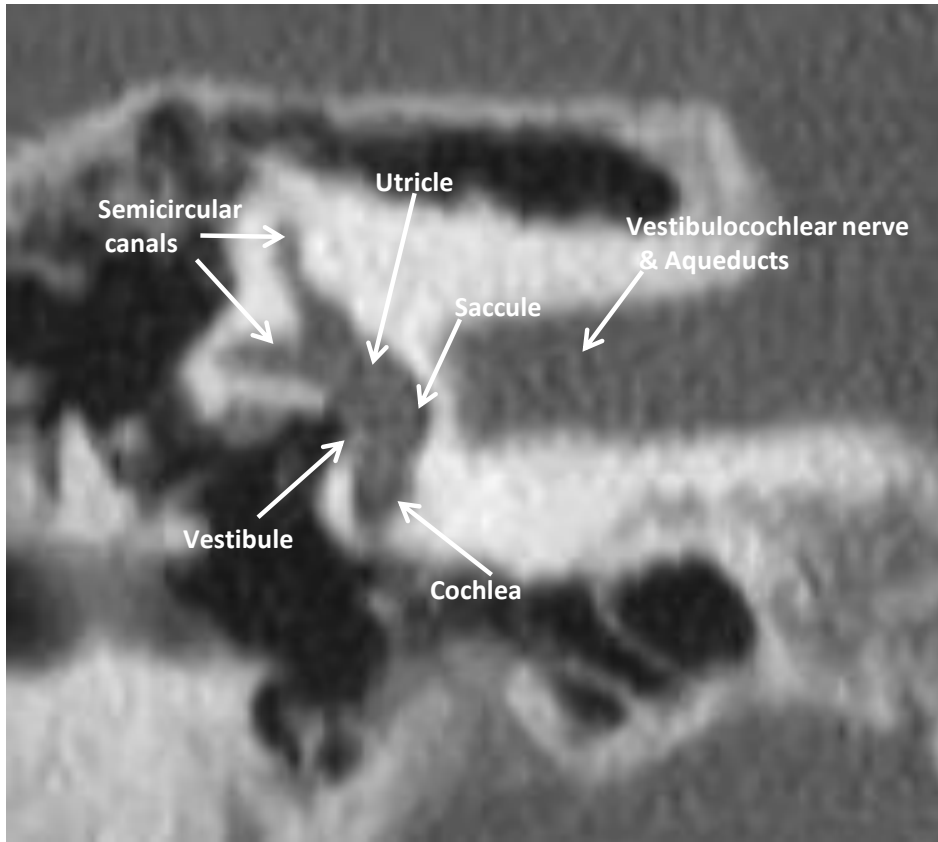
Normal temporal bone

1) Inner ear

2) Middle ear

3) External ear

Inner ear anatomy



1) Bony labyrinth

- Cochlea
- Vestibule
- Semicircular canals

2) Membranous labyrinth

- Cochlear duct
- Utricle & Saccule
- Endolymphatic duct

3) Perilymphatic spaces

- Oval & round window
- Cochlear aqueduct
- Vestibular aqueduct

1) Inner ear anatomy

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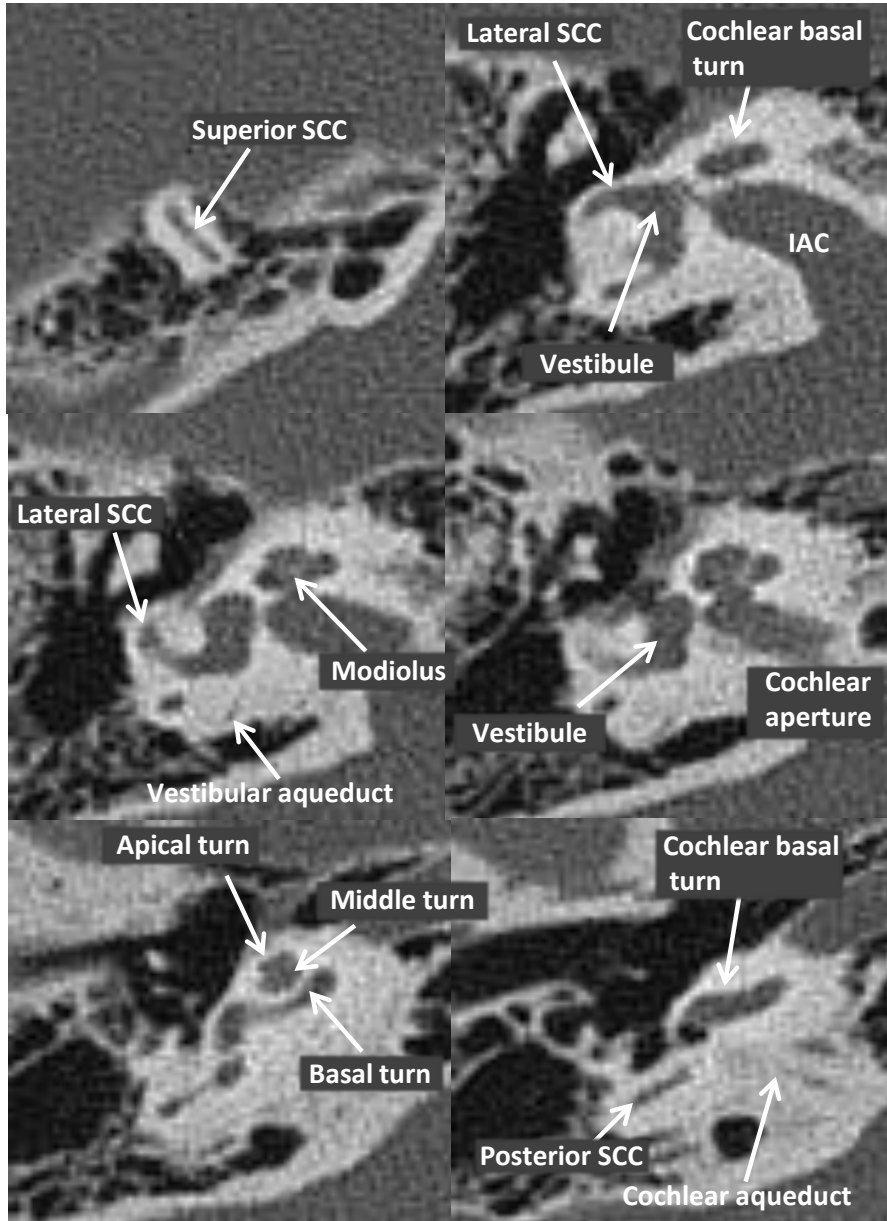
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Anatomy

Axial CT scans



• Bony labyrinth

- Cochlea, vestibule, semicircular canals

• Semicircular canals

- Three semicircular canals of bony labyrinth
- Vestibule is perilymphatic space of vestibule
- Continues posteriorly by SSC to IAC and apex
- Base of SSCs are angled to each other
- Intercanals angle anterior & posterior
- Bony cochlea : Around modiolus
(Conical central core)
- Basilar membrane : Divide canal into
scala vestibuli & tympani

1) Inner ear anatomy

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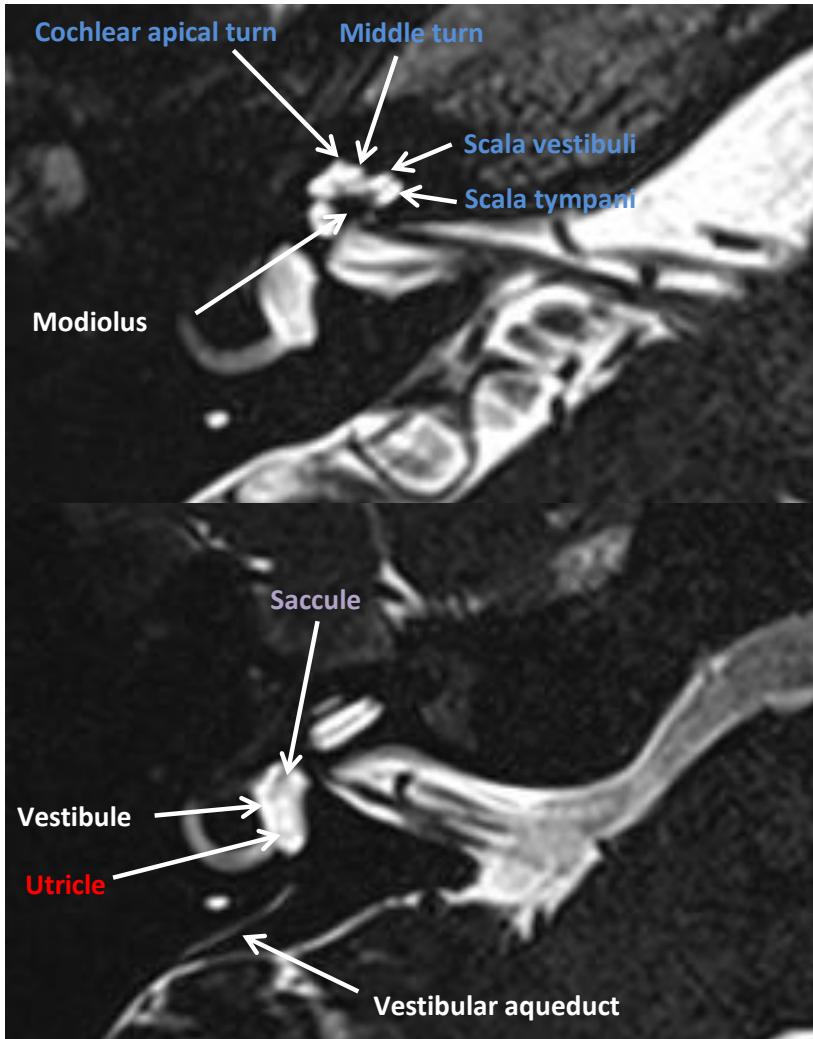
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Anatomy

Axial thin-section T2WI of MRI



• Membranous labyrinth

- **Continuous** openings of perilymphatic spaces
: Divides into Scala vestibuli & Scala tympani
- **Utricle**
• **Cochlear aqueduct**
: Opening of membranous semicircular canals
: Potential communication & equilibration
- **Saccul**
between perilymphatic & subarachnoid spaces
: Spherical recess near opening of scala vestibuli
- **Vestibular aqueduct**
: Maculae of utricle and saccule (Static balance)
: Ampullae of SCC (Angular acceleration)
- **Endolymphatic duct & sac**
: From vestibule, through vestibular aqueduct

1) Inner ear anatomy

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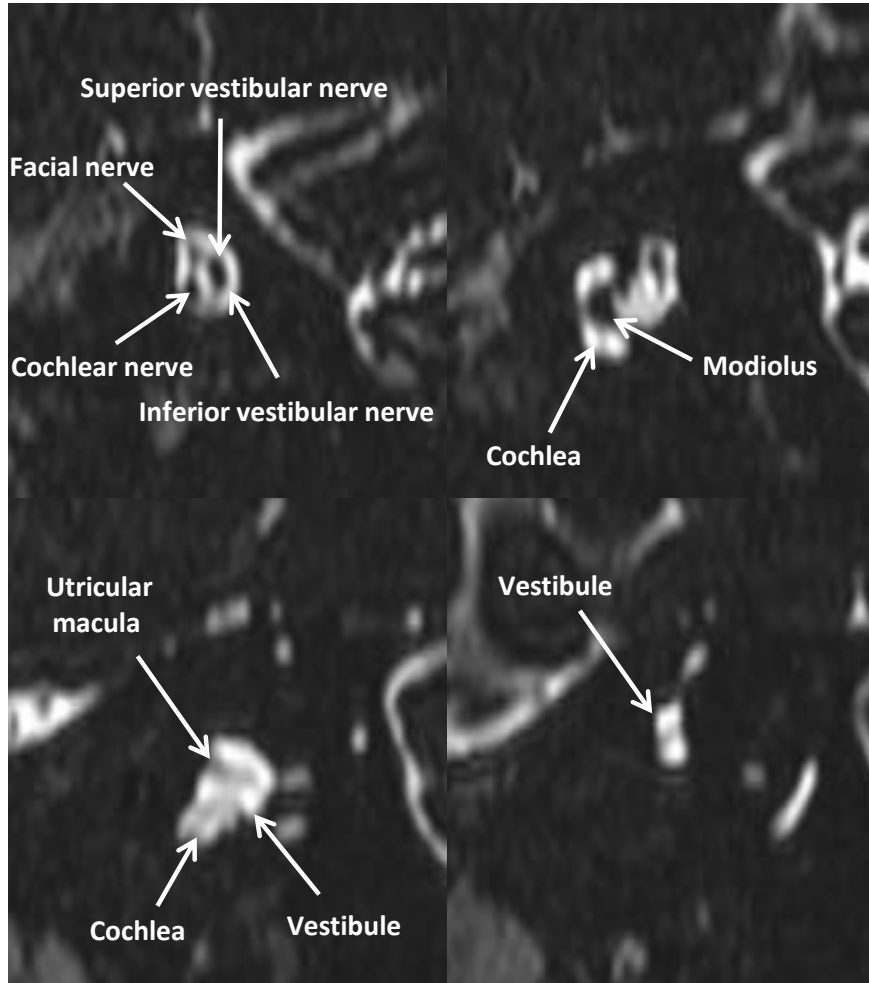
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Anatomy

3D heavily weighted T2WI with oblique sagittal reconstruction



- **Internal auditory canal (IAC)**

- Bony conduit transmitting **cranial nerve VII, VIII**
- From pontomedullary junction to inner ear
- Adult vertical dimension at birth
(Average : 4mm, < 2mm : hypoplasia)

2) Middle ear anatomy

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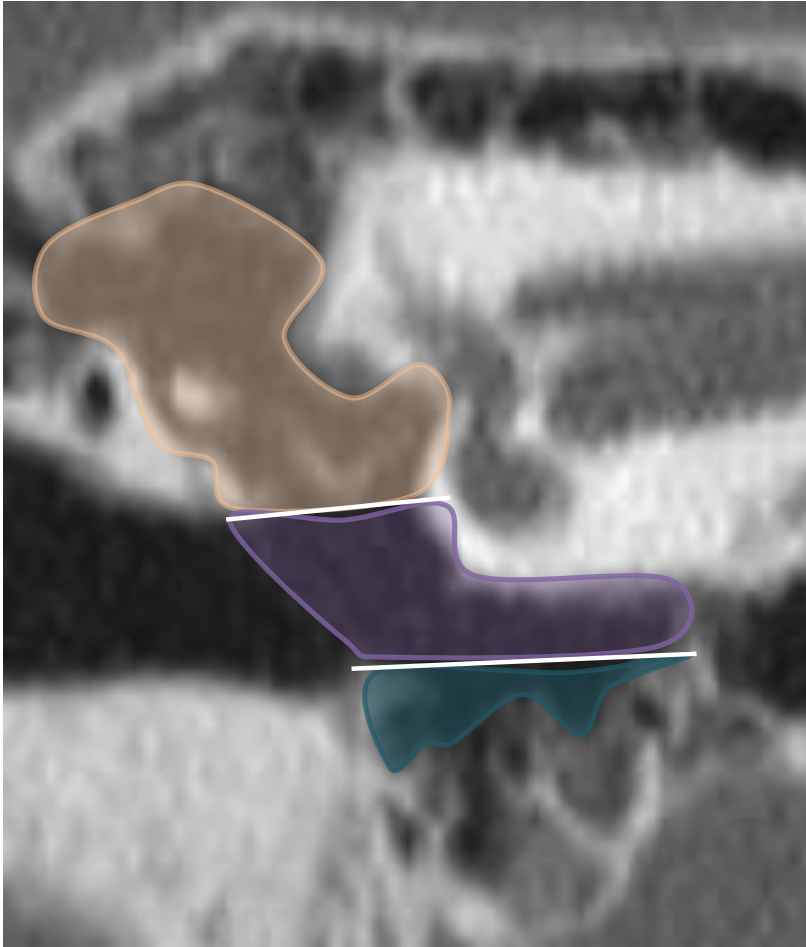
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Anatomy

Middle ear



- **Main function**
Ossicles transmitting vibration of TM to inner ear
- **Size**
Vertical & anteroposterior dimension : 15mm,
Transverse dimensions : 6mm
- **Space**
 - Epitympanum (above level of TM)**
 - Mesotympanum (tympanic cavity proper)**
 - Hypotympanum (below level of TM)**
- **6 walls (from petrous bone)**
 - : Anterior (carotid wall), posterior (mastoid wall),
roof (tegmen), floor (jugular wall),
medial (TM) and lateral wall (membranous wall)

2) Middle ear anatomy

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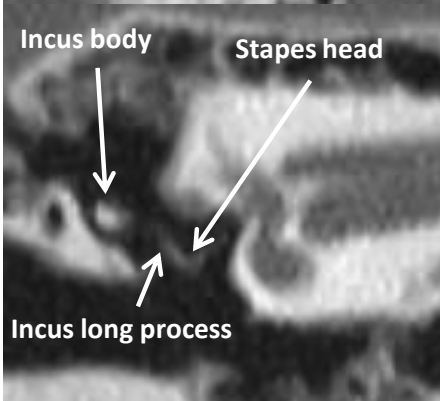
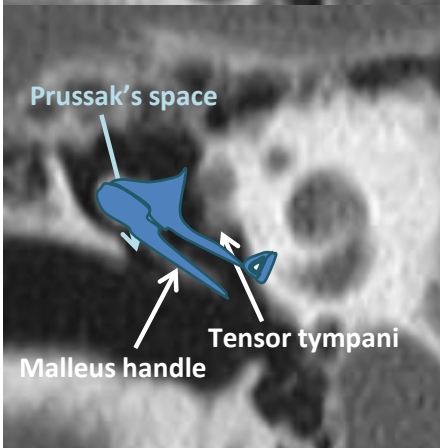
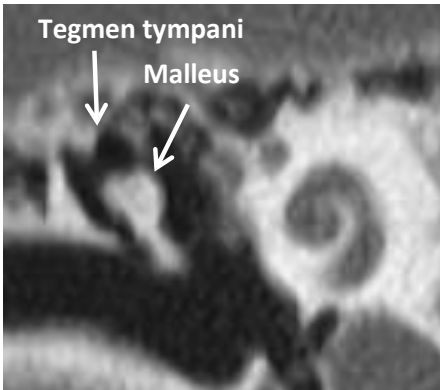
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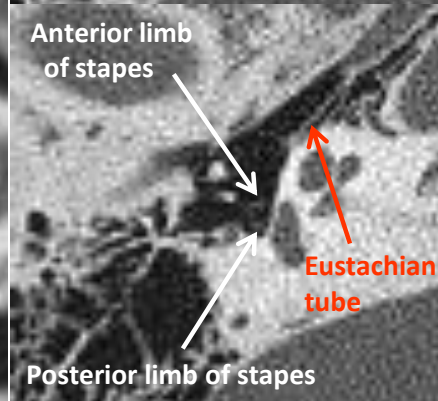
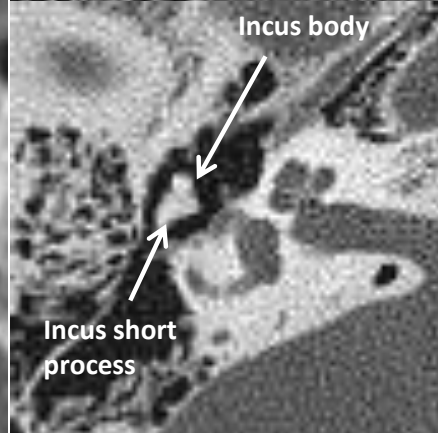
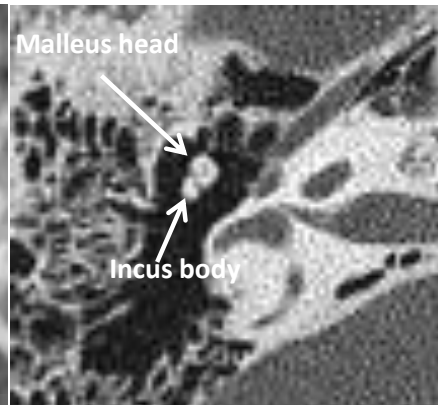
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Anatomy

Coronal



Axial



• Middle ear ossicles

- Osseous (1cm length)
: Transmitting & amplifying vibration of TM to inner ear

Cartilaginous portion (2.5cm length)

• **Malleus**

- To pharyngeal opening
- Head, neck
→ Tensor / levator veli palatini,
- Manubrium : Attached to TM
- Lateral process : Lateral process abuts TM
- Equilibrating pressures of middle ear and pharynx

• **Incus**

- Body, short process

• **Prussak's space**

- Long process : Descends nearly vertically behind
- Small recess medial to pars flaccida of TM
- Lenticular process : Articulate with head of stapes
- Common location for cholesteatoma

• **Stapes**

- Head : Articulation with incus
- Neck, anterior & posterior crura
- Base : To oval window with edge of cartilage

3) External ear anatomy

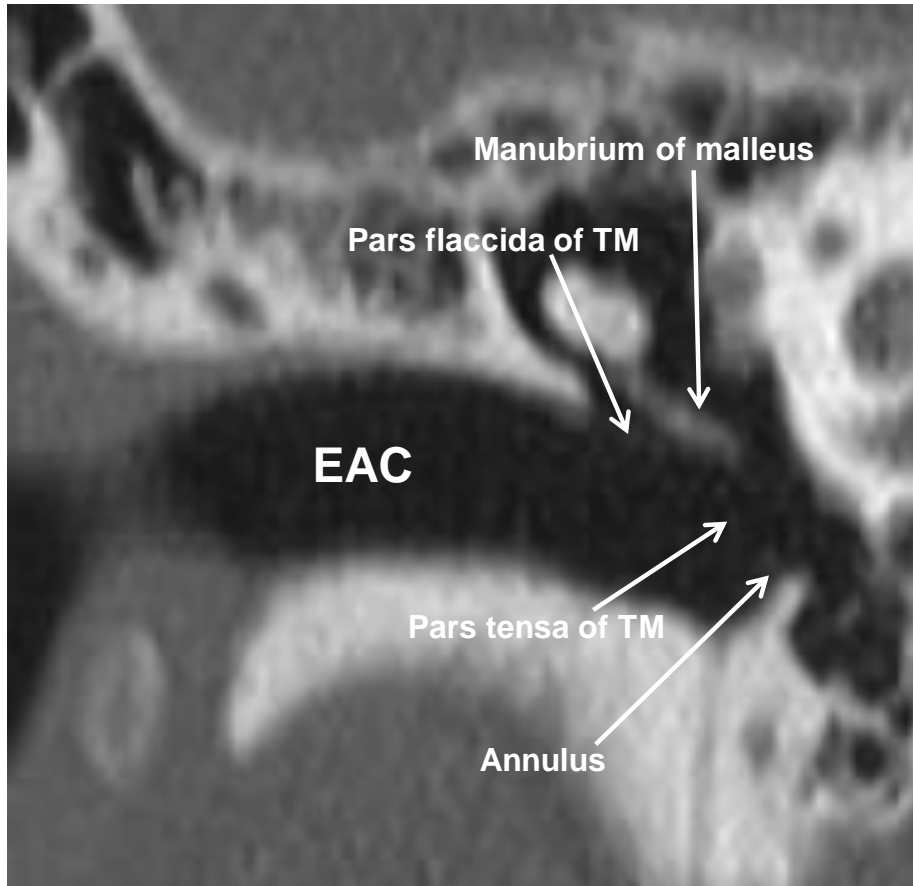
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Anatomy



- **External auditory canal (EAC)**
 - Fibrocartilaginous (lateral), bony portion (medial)
- **Tympanic membrane**
 - Thin membrane forming medial boundary of EAC
 - Manubrium of malleus attached to TM
 - **Pars flaccida**
 - : Superior-anterior portion of tympanic membrane
 - : Triangular thin lax zone by notch of Rivinus
 - : Under the scutum
 - **Pars tensa**
 - : Remaining inferior portions of tympanic membrane

3. Major congenital abnormalities

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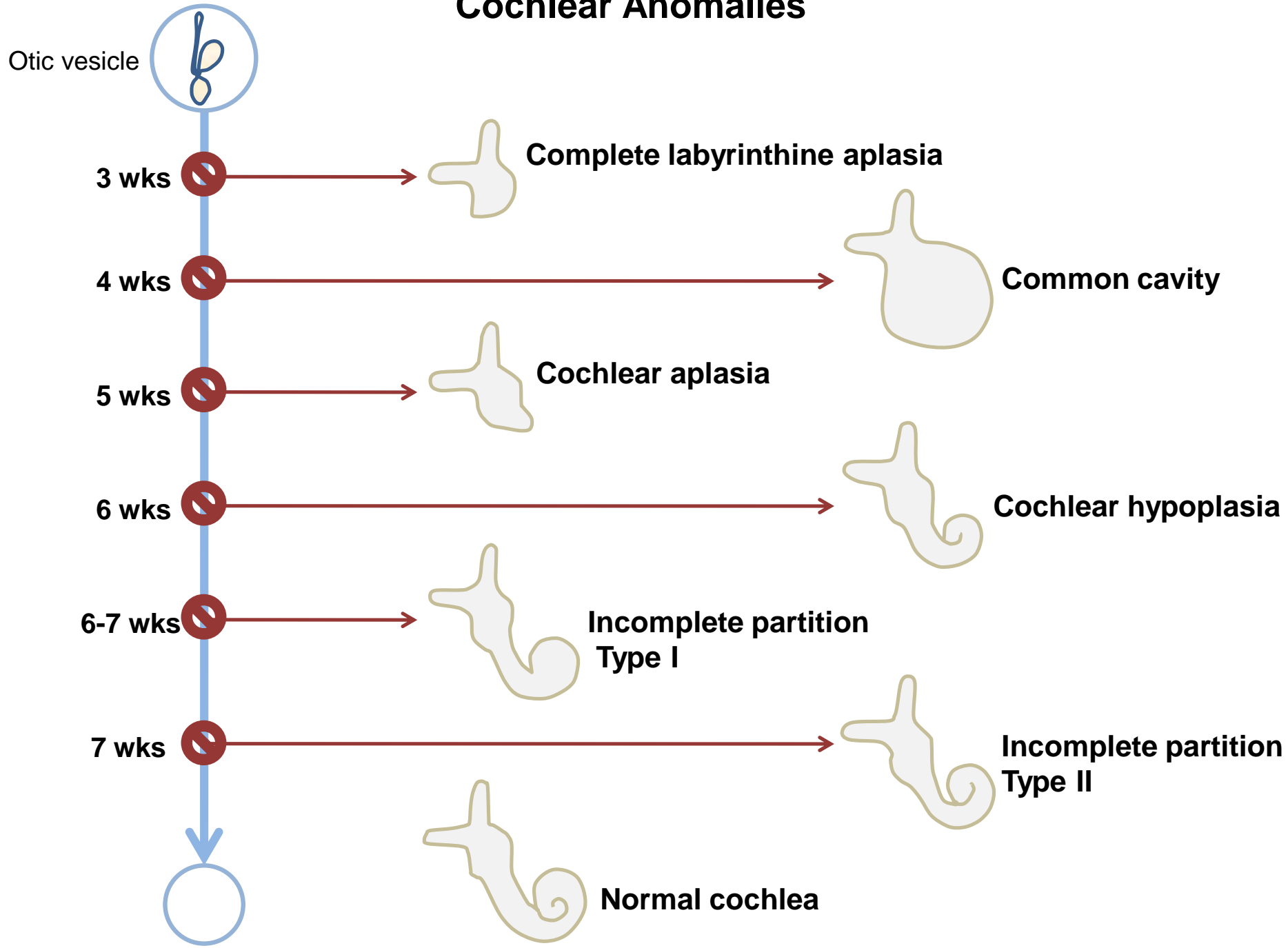
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Major anomaly

- Inner ear abnormalities : 30%
- External & middle ear abnormalities : 60%
Due to developmental arrest during embryogenesis
: From otic placode & vesicle
- Complex type : 10%
 - Bilateral in 65%
 - Membranous labyrinthine abnormality in 80%
: Scheibe dysplasia, Siebenmann-Bing
 - **Combined bony labyrinthine abnormality in 20%**
→ Radiologically detectable on CT / MRI

Cochlear Anomalies



1) Inner ear abnormalities (Cochlear)

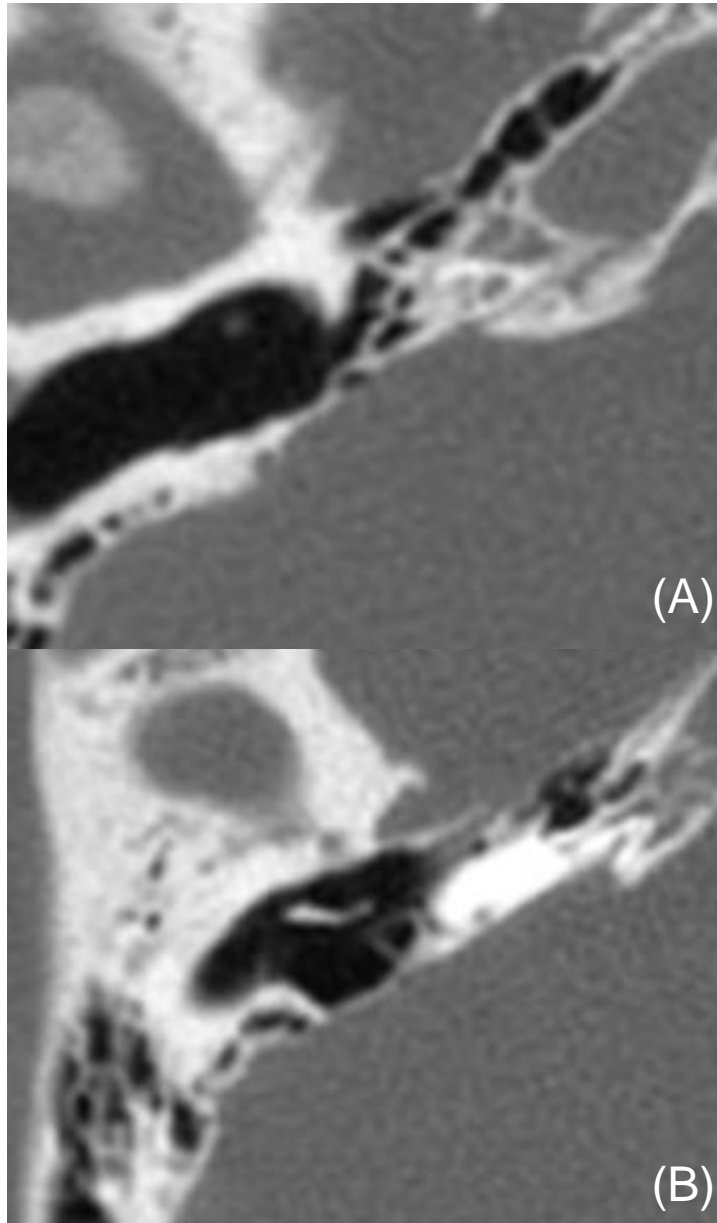
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Major anomaly



- **Complete labyrinthine aplasia (Michel's aplasia)**

Michel's aplasia

Axial CT scans (A, B) showed near complete absence of the differentiated inner ear structures including cochlea and vestibule. Note hypoplastic petrous bone.

: Single / several cystic cavity in place of inner ear

1) Inner ear abnormalities (Cochlear)

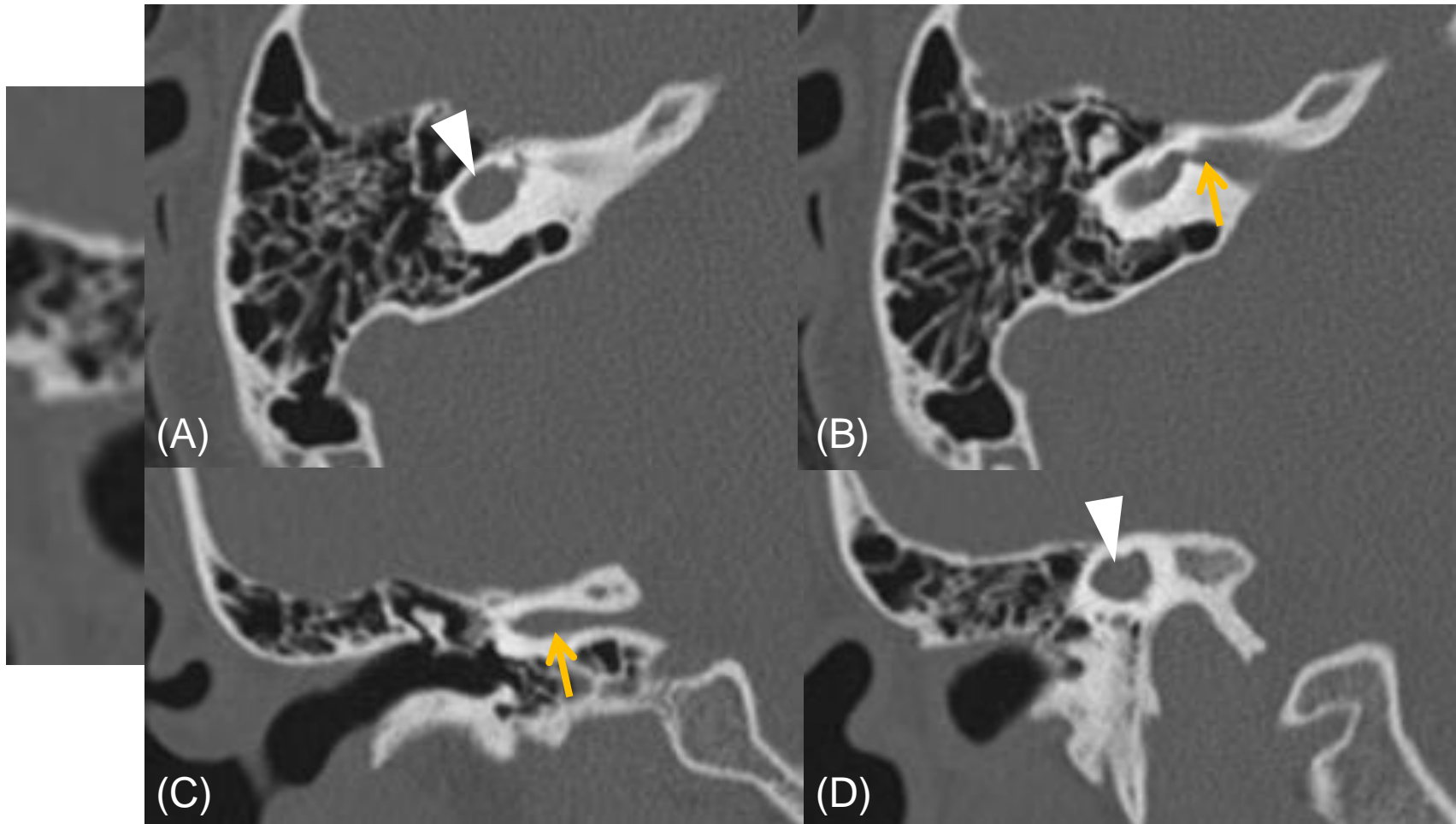
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Major anomaly



15/M, Common cavity type of cochlear-vestibular malformation.

Axial (A, B) and coronal (C, D) CT scans presented no distinct cochlea, vestibule, and semicircular canals and It formed large common cavity (White arrowheads). The single cavity structure showed connection with IAC (Yellow arrows). Note that there was not any cochlear structure anterior to the IAC, where the cochlea is normally situated.

1) Inner ear abnormalities (Cochlear)

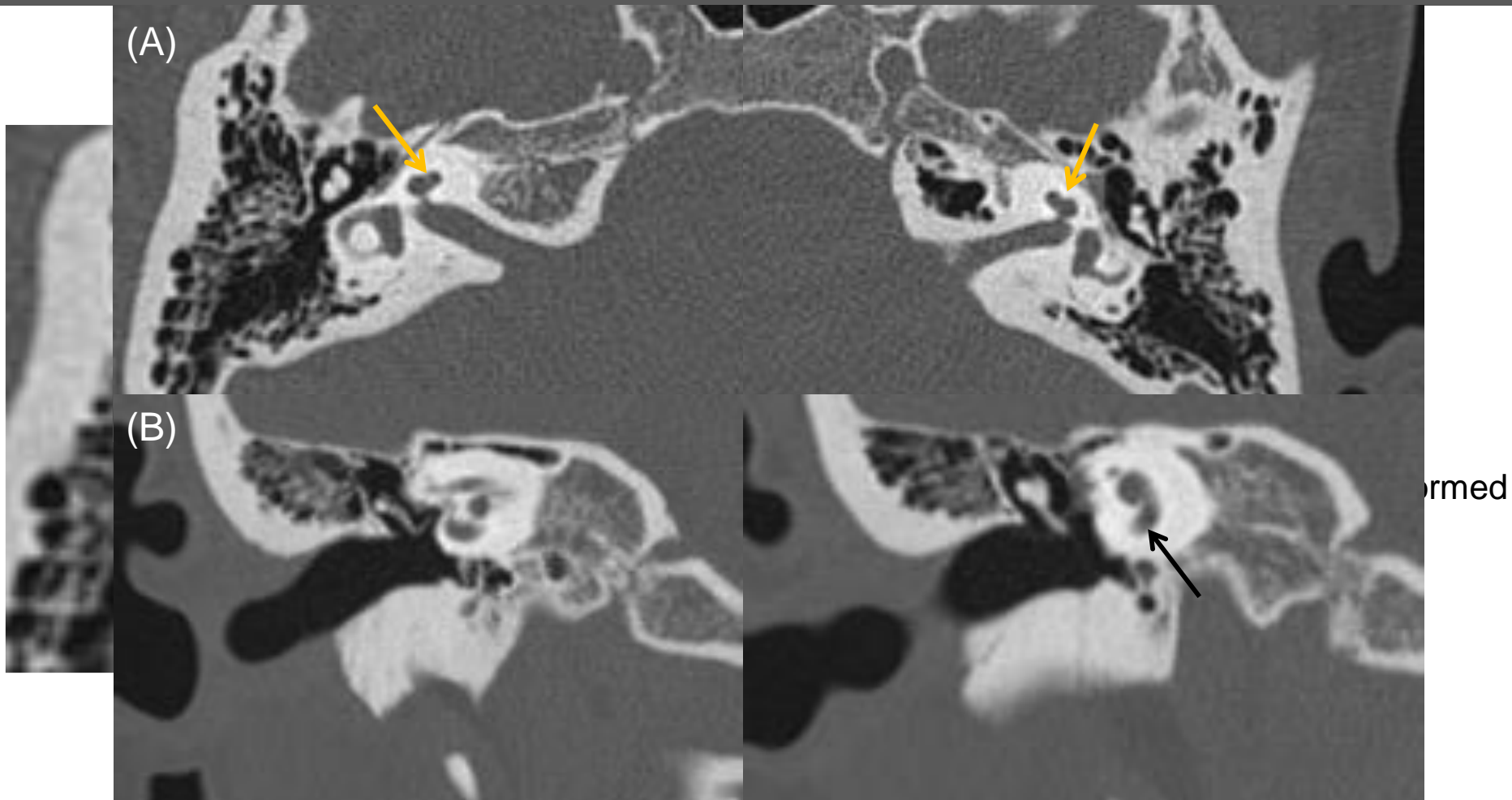
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Major anomaly



16/F, Cochlear hypoplasia.

On axial (A) and coronal (B) CT scans, aplastic apical turns of cochlea could be recognized in both sides (Yellow arrows in A). There were also reduced maximal dimension of basal turns (Black arrow in B).

1) Inner ear abnormalities (Cochlear)

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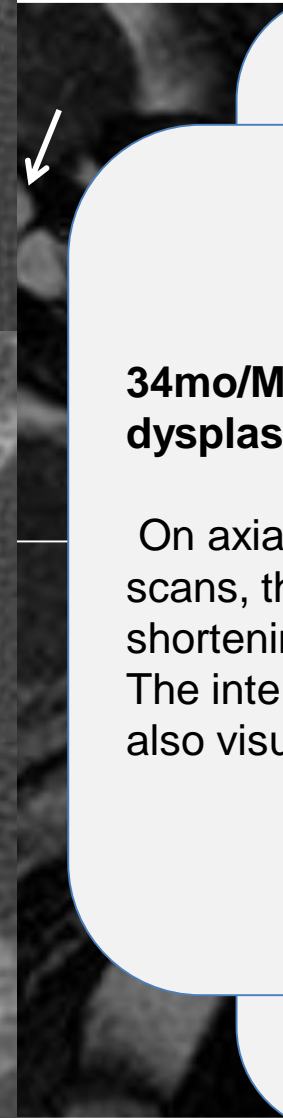
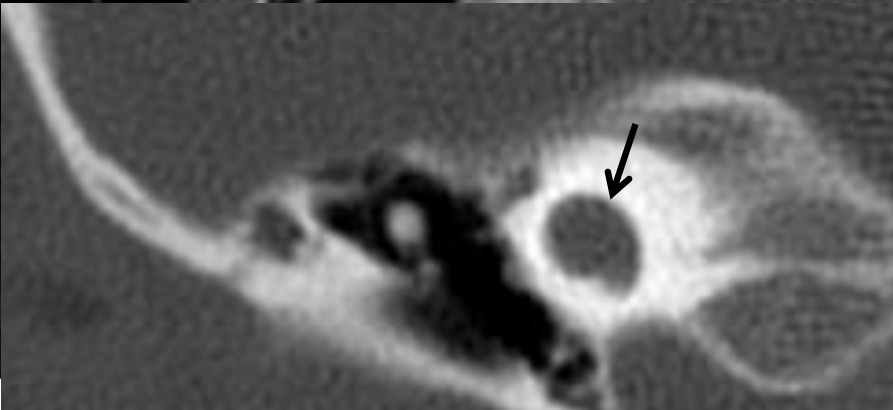
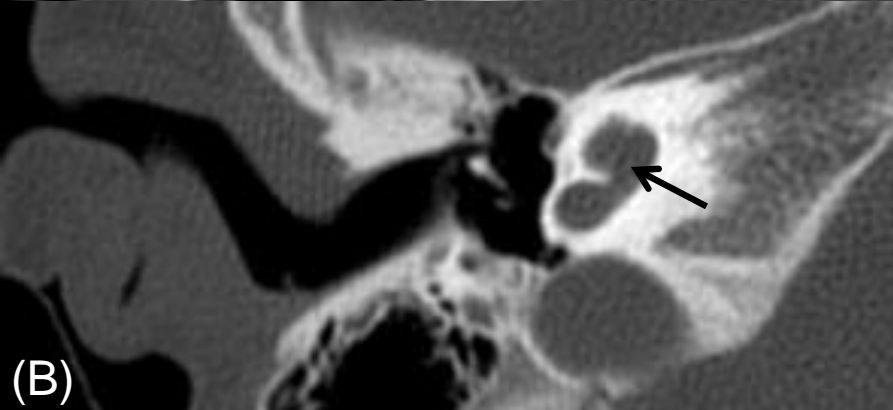
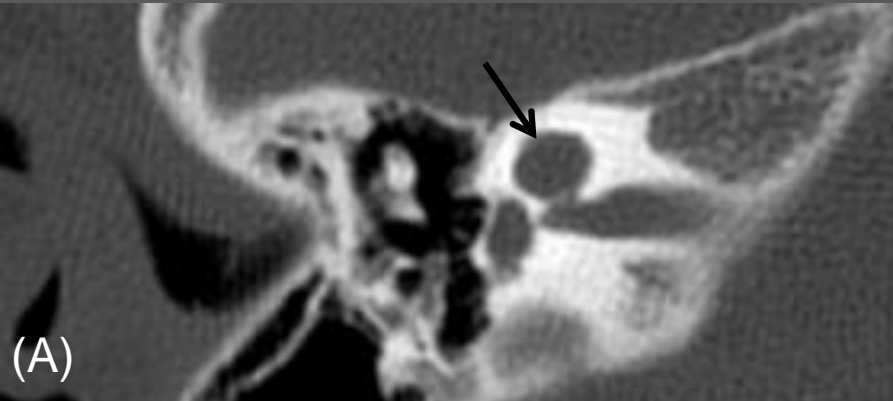
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Major anomaly

Ty



34mo/M, Bilateral Mondini dysplasia and large vestibule

On axial (A, B) and coronal (C) CT scans, the right cochlea showed shortening along its modiolar axis. The interscalar septum was not also visualized.

1) Inner ear abnormalities (Vestibular)

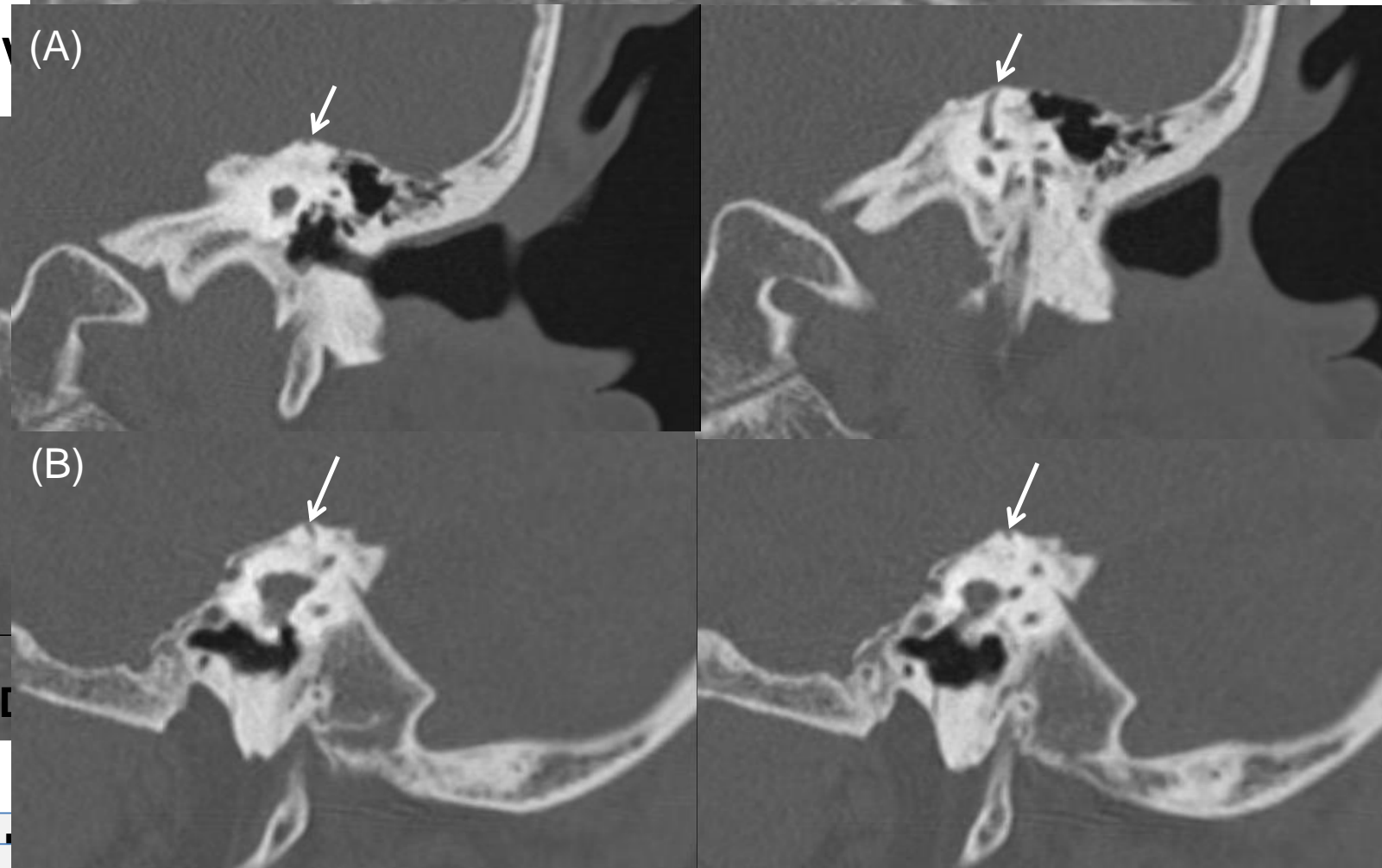
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Major anomaly



19/F, SCC dehiscence

On coronal(A) and sagittal (B) CT scans, there was bony dehiscence of the left superior semicircular canal (Arrows). She suffered from sound-induced dizziness.

1) Inner ear abnormalities (Aqueduct)

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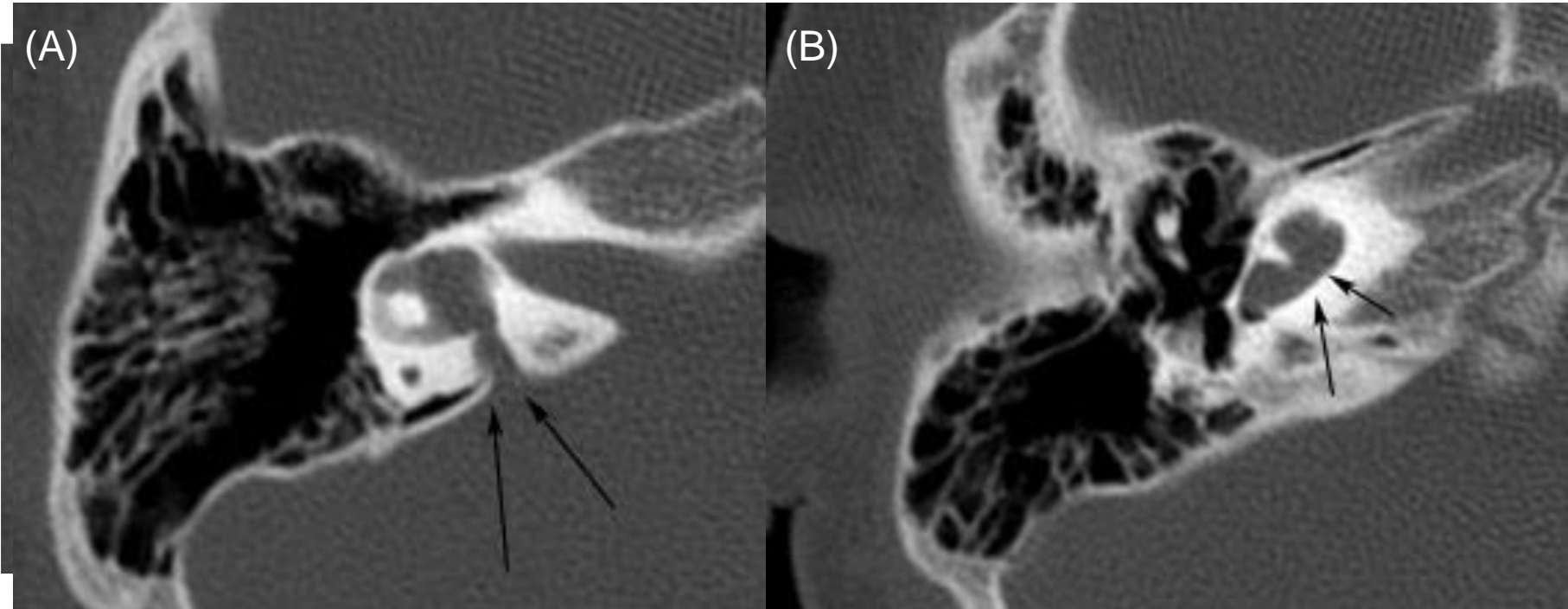
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Major anomaly

Aqueductal anomaly



3/F, Mondini dysplasia with enlarged vestibular aqueduct.

On axial CT scans (A and B), the vestibular aqueduct (Arrows in A) was enlarged and widely opened, larger than normal posterior SCC. In addition, insufficient cochlear turns (Arrows in B) were also noted, suggesting Mondini dysplasia.

1) Inner ear abnormalities (IAC)

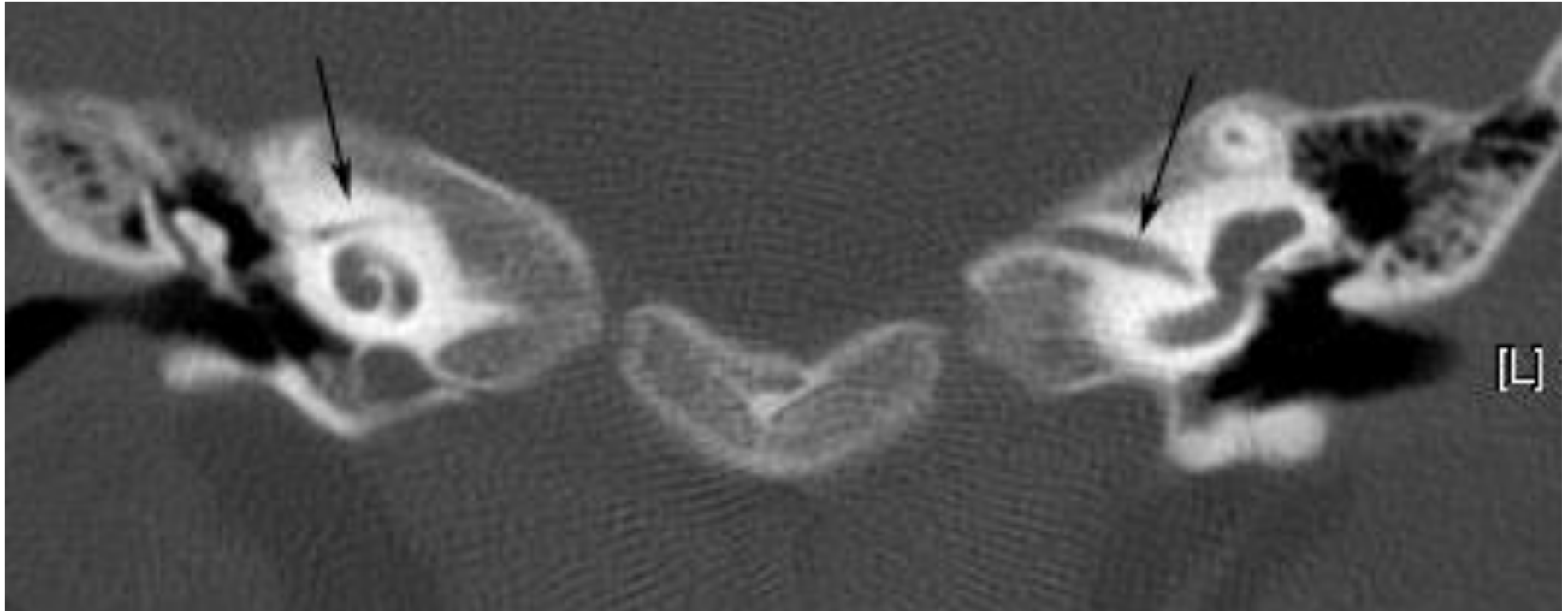
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Major anomaly



12mo/F, Bilateral small IAC

Coronal CT scan showed narrowed bilateral internal auditory canals (Arrows), measuring maximal diameter of 1.3 mm in right side and 1.6 mm in left side (Normal range : 2 – 8 mm). She had sensorineural hearing loss on both sides.

2) Middle ear abnormalities

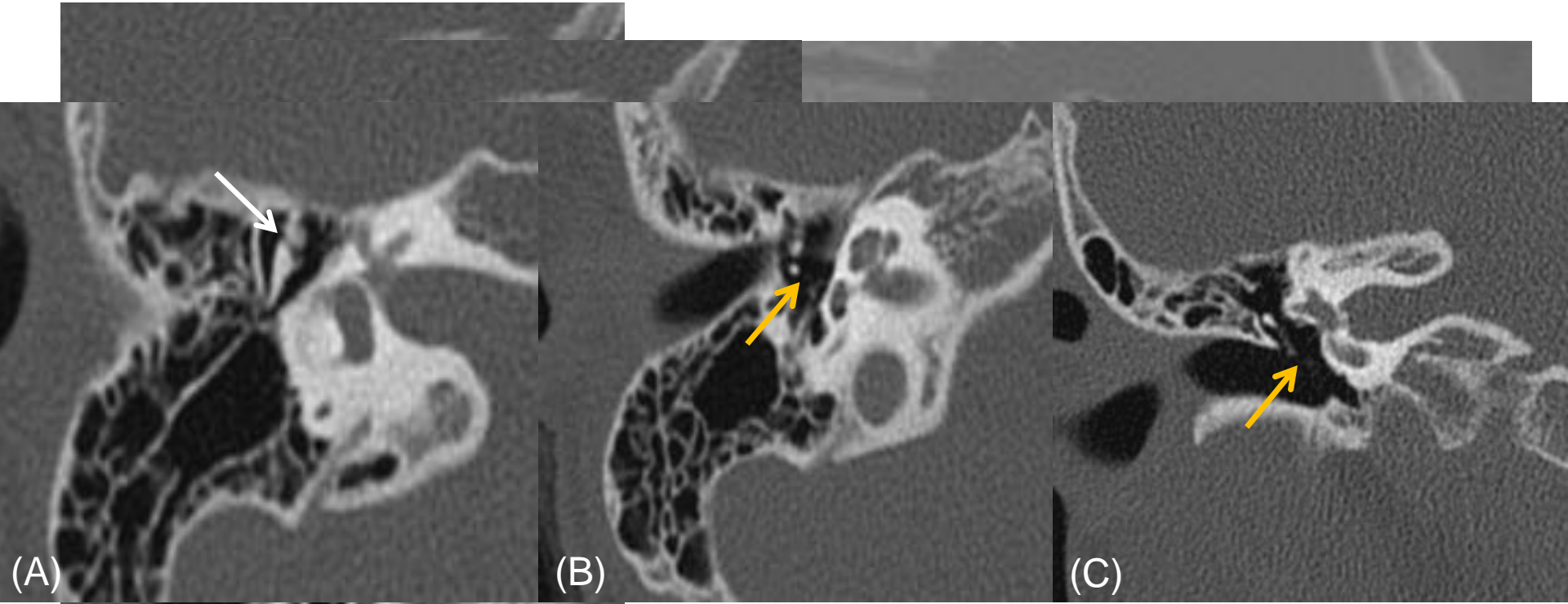
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Major anomaly



13/F, Isolated middle ear anomalies

On axial CT scan (A), superiorly displaced incus was noted on the right side. There was also incudostapedial subluxation (Yellow arrows) on axial (B) and coronal (C) CT scans. The surgical report described that there was not only incudostapedial subluxation, but also absence of lenticular process of incus.

3) External ear abnormalities

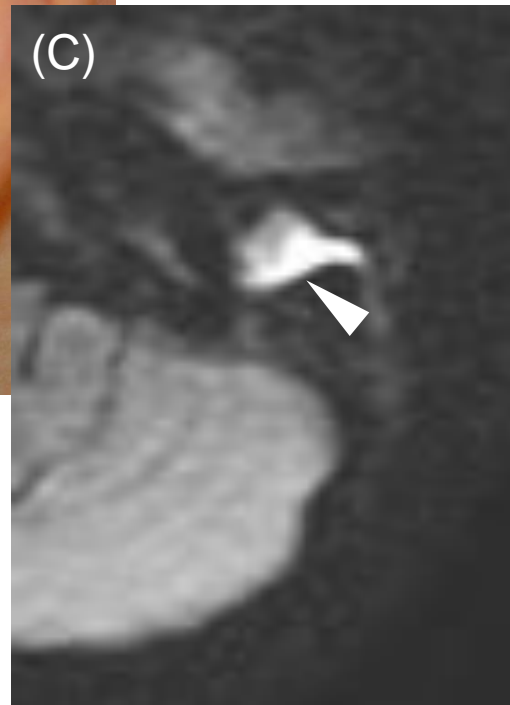
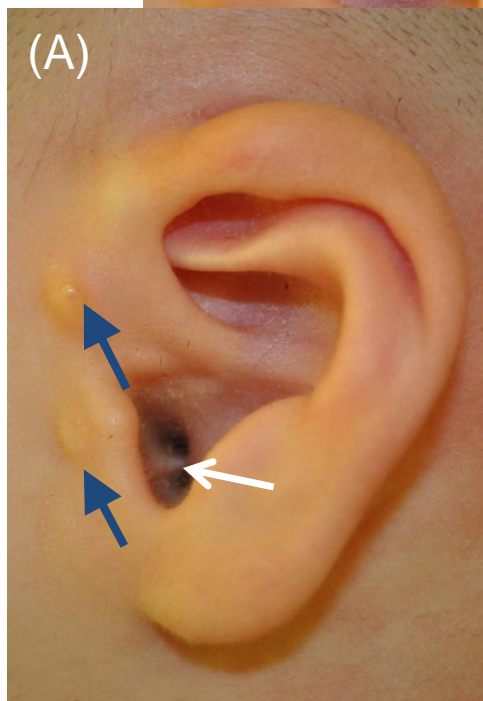
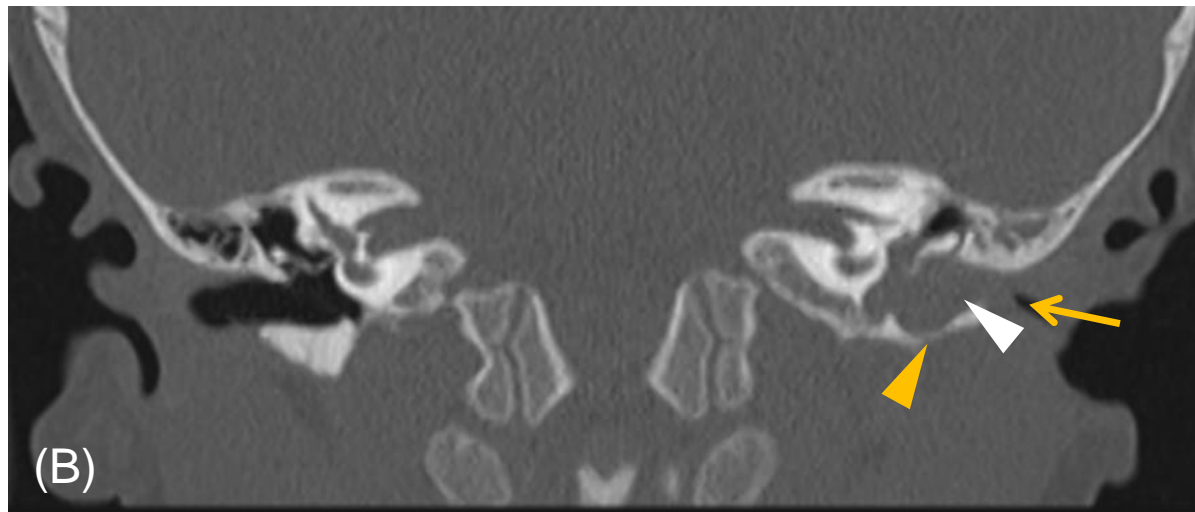
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Major anomaly



2/F, Left bifid orifice and stenosis of EAC with cholesteatoma

She had bifid orifice (White arrow in A) in EAC and skin tags (Blue arrows in A) in external ear. Coronal CT scan (B) showed atresia of left external auditory canal (Yellow arrow) with inner soft tissue density (White arrowhead), accompanying adjacent bony erosion of EAC (Yellow arrowhead). On diffusion-weighted image of MRI (C), the lesion showed diffusion restriction (White arrowhead), suggesting cholesteatoma in EAC and middle ear cavity.

- Temporal bone is one of the most elaborate and complicated structure in human body
- Development of the temporal bone
 - Inner ear –Neuroectoderm to cochleovestibular apparatus
 - Middle & external ear – 1st & 2nd branchial arches and pouch
 - Vestibular aqueduct – Last embryogenic structure, when enlarged, high suspicion for other inner ear abnormalities
- Knowledge of embryology of temporal bone assists understanding various congenital anomalies

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