Radiologic Evaluation of Cochlear Incomplete Partitions: An Overview

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Presentation Overview

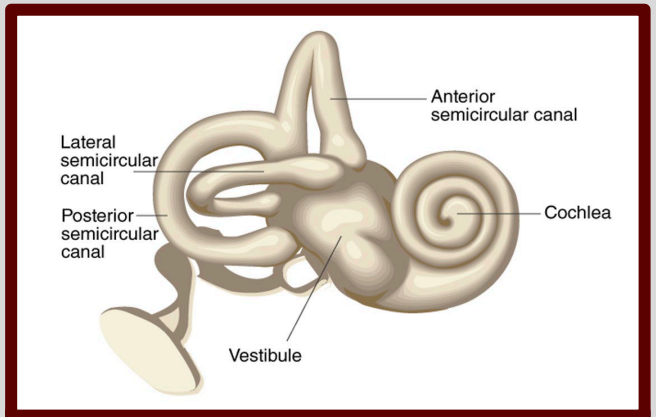
- What are cochlear incomplete partitions?
- Cochlear Anatomy
- Key Evaluation Criteria
- IP-I
- IP-I vs Common Cavity
- IP-II
- IP-III
- Surgical considerations
- Teaching points

Cochlear Incomplete Partitions

- The cochlear incomplete partitions are a group of congenital bony cochlear malformations
- Group includes cochlear incomplete partition type I (IP-I), type II (IP-II), and type III (IP-III)
- Each member of this group is characterized by distinct etiology and anatomic features which result in varying degrees of sensorineural hearing loss
- Radiologists have a vital role in diagnosing these conditions through accurate image interpretation which influences clinical decision making and ensures optimal management
- Thin bone slice CT and T2-weighted steady state free precession MRI (FIESTA, CISS) are optimal for imaging assessment

Cochlear Anatomy

- Snail-shaped structure of the inner ear within the petrous part of the temporal bone
- Composed of a hollow osseous spiral containing fluid filled chambers, hair cells, and specialized membranes which convert mechanical sound vibration into electrical nerve impulses
- Important terms:
  - Modiolus: Bony pillar around which the cochlea spirals
  - Lamina Cribrosa: Base of modiolus
  - Interscalar septum: Separates and defines each turn of the cochlea (apical, middle, and basal turn)
  - Vestibule: Located at the base of the cochlea, connects cochlea and vestibular apparatus
  - Cochlear aperture: Opening where cochlear nerve exits the cochlea



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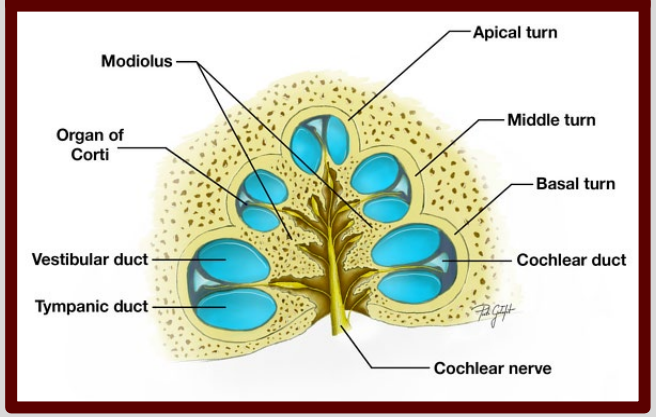

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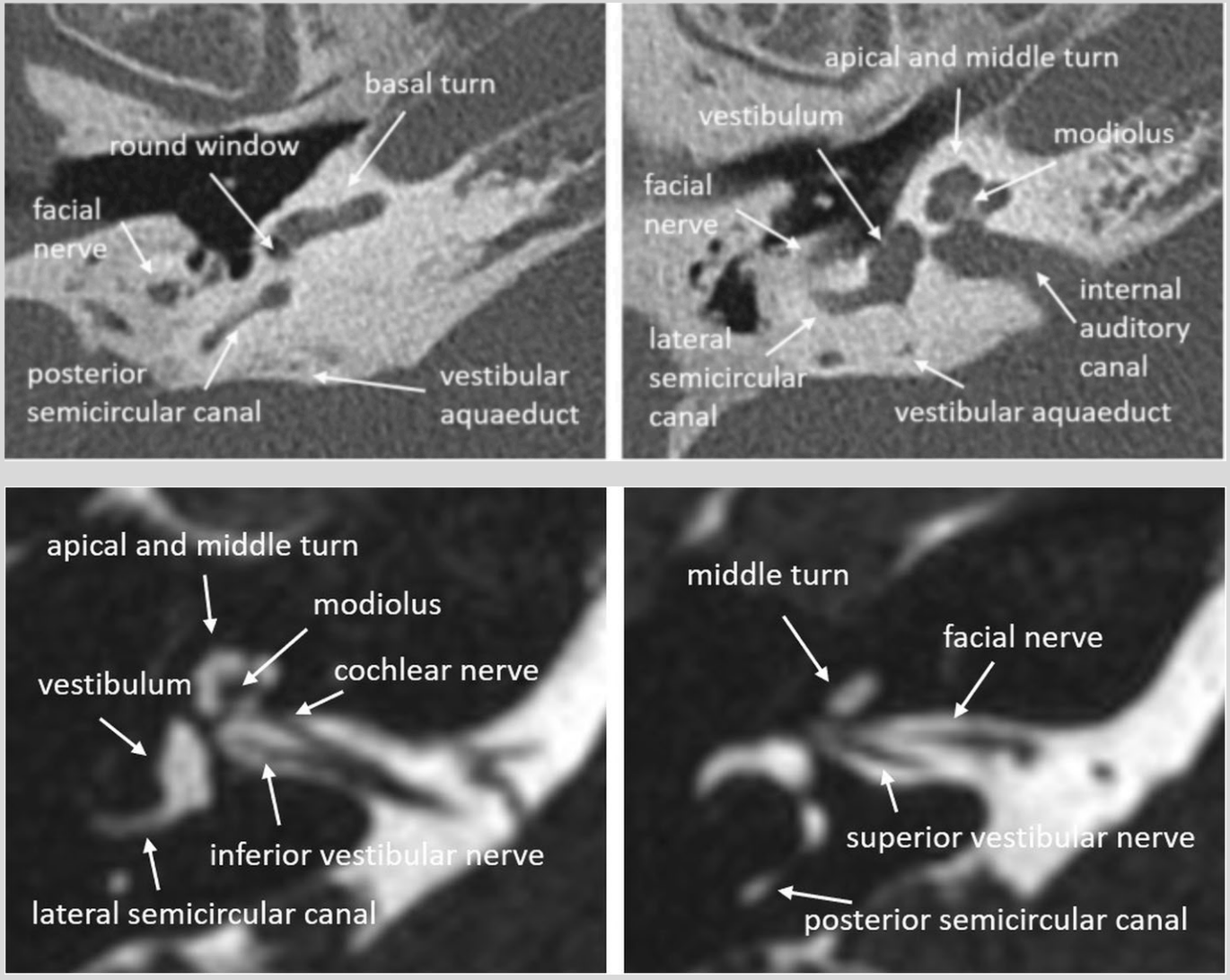
Cochlear Anatomy

- Cochlear size
  - Must be of normal size to classify as cochlear incomplete partition
    - If small, consider cochlear hypoplasia instead
  - Outer dimensions typically measure approximately 10mm×8mm×4mm
    - Corresponds to 33×27×13 voxel widths on 0.3mm CT
  - Good rule of thumb: Height should measure >4.3mm in the coronal plane, regardless of age
- Cochlear turns
  - 2½ to 2¾ turns which spiral around the modiolus
  - Divided into the basal, middle, and apical turns by the interscalar septum
    - Interscalar septum has 3 ridges: R1, R2, and R3
      - R1 is posteromedial between the lower basal and lower middle turns
      - R2 is lateral between the upper basal and upper middle turns
      - R3 is medial between the lower middle and apical turns

Cochlear Anatomy on Imaging

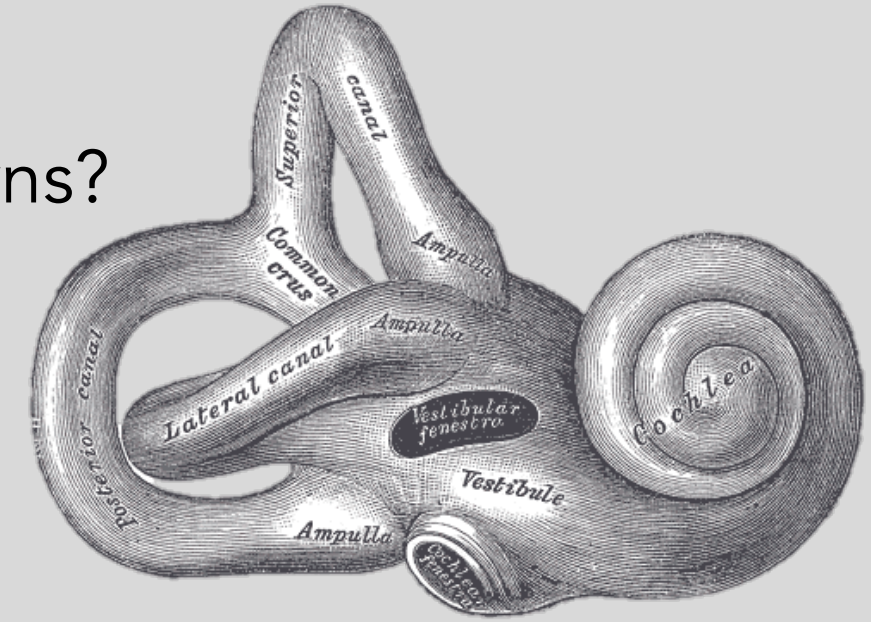


HRCT axial images of normal cochlear anatomy in a 74-year-old man

MRI 3D-CISS axial images of normal cochlear anatomy in a 40-year-old woman

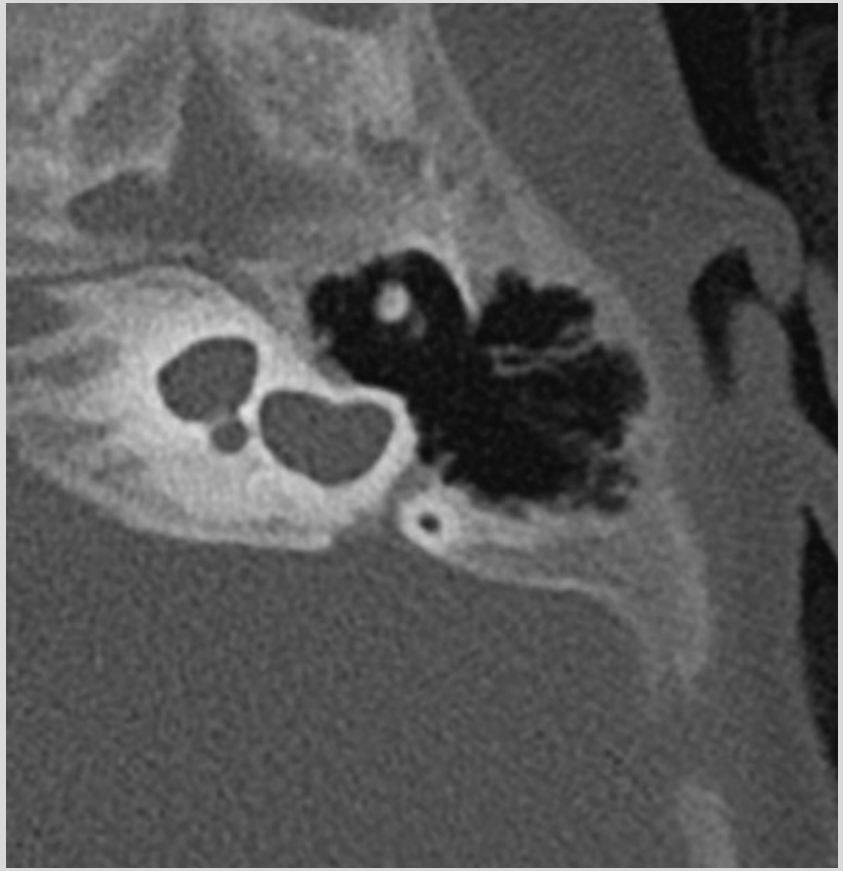
Evaluating Cochlear Malformations

- Is the cochlea normal in size?
- Is the cochlea separate from the vestibule?
- Is the modiolus present?
- Is the interscalar septum present?
- Number and morphology of cochlear turns?
- Is the cochlear aperture patent?
- Are the facial and cochleovestibular nerves clearly visualized?



IP-I (Cochleovestibular Malformation)

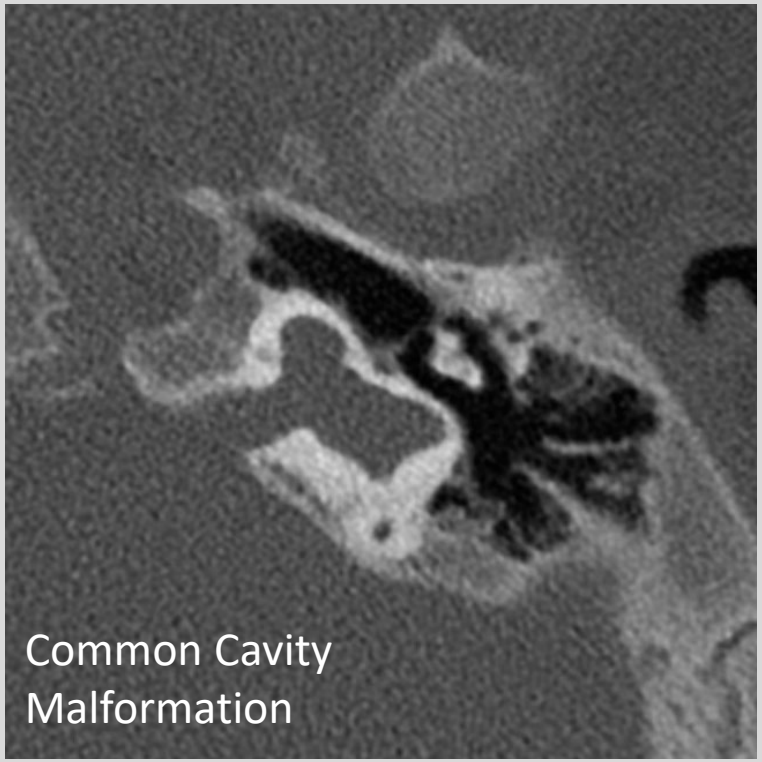
- Caused by failure of the modiolus and interscalar septum to form
- Results in a single, cystic appearing cochlear cavity
- Often presents with a dilatated but separate vestibule, leading to a “figure 8” appearance on imaging
- IP-I is associated with profound hearing loss and cochlear implantation is typically the primary treatment option
- No known genetic associations

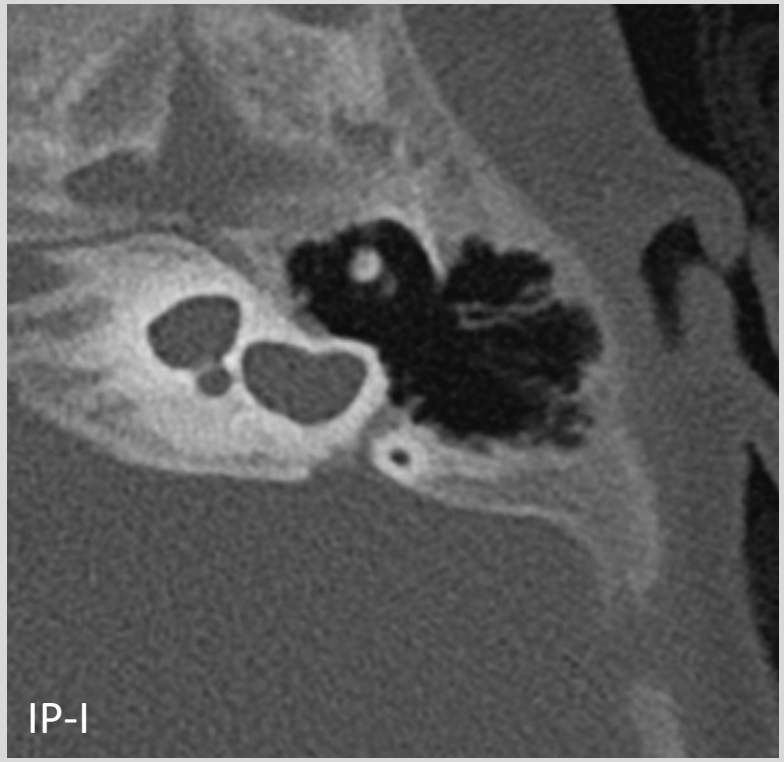


IP-I. Separate cochlea and vestibule in a 6-week-old female with sensorineural hearing loss

IP-I vs Common Cavity Malformation

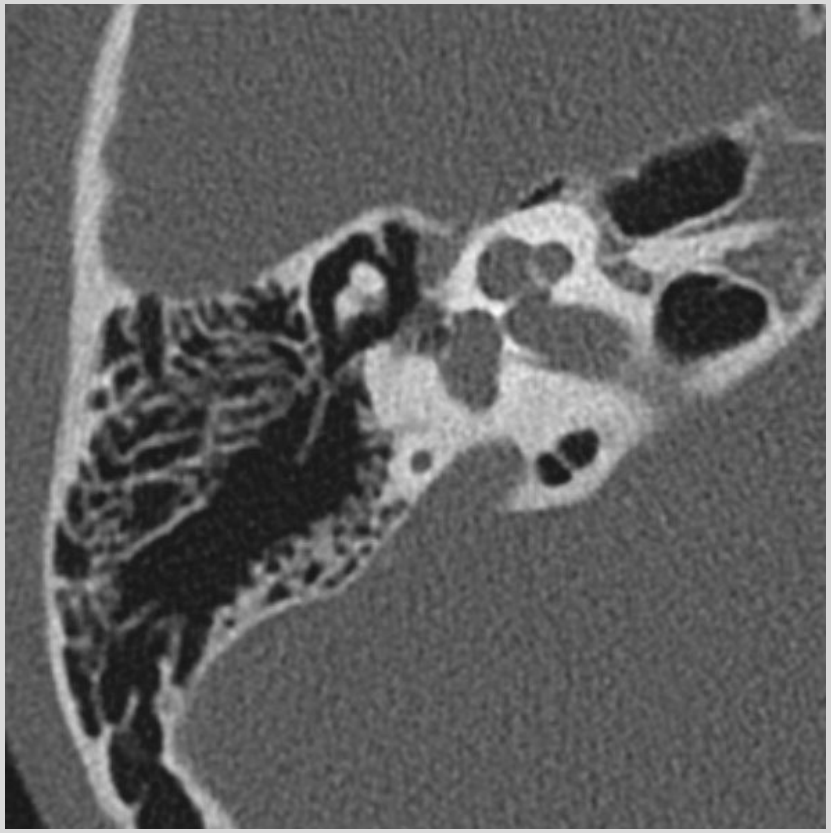
- IP-I should be differentiated from common cavity malformation
- In common cavity malformation, the cochlea and vestibule are fused





Cochlear Incomplete Partition Type II IP-II

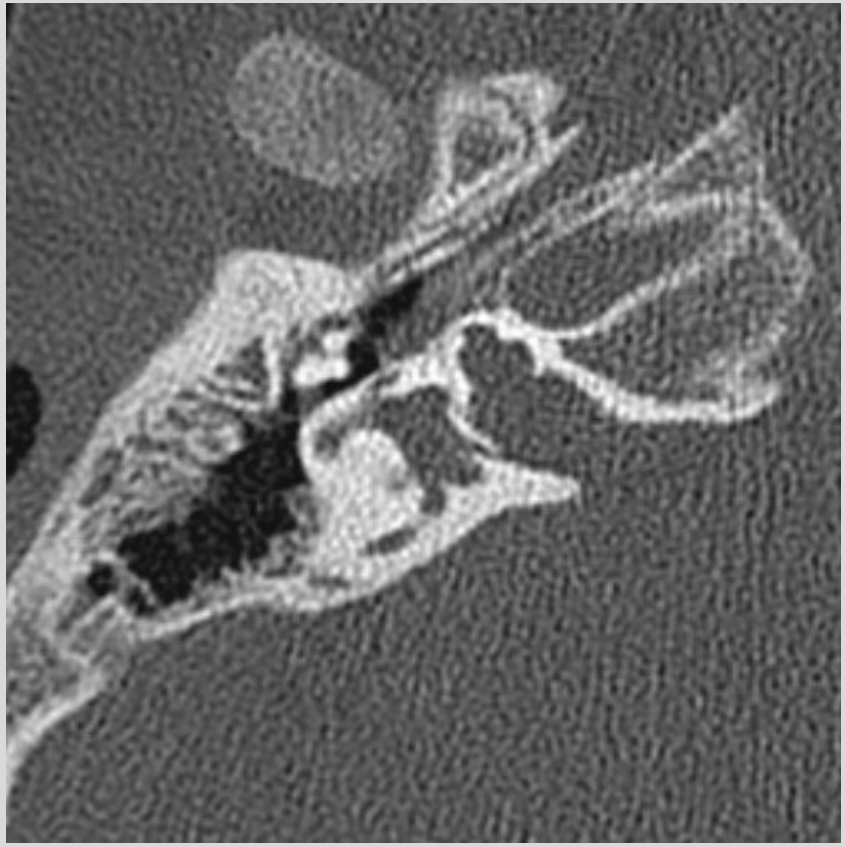
- Defective formation of the apical modiolus and interscalar septum, resulting in fusion of the apical and middle cochlear turns with a normal appearing basal turn
- This malformation gives the cochlea a characteristic “baseball cap” appearance
- Typically associated with an enlarged vestibular aqueduct and mild vestibular dilation
- Hearing loss is variable but is often progressive, eventually requiring use of hearing aids or cochlear implantation
- Differential considerations include cochlear hypoplasia and brachio-oto-renal syndrome (BOR)
    - BOR is characterized by an unwound appearance to the cochlea, ossicular anomalies, and enlarged Eustachian tubes, features atypical for IP-II
- Associated with mutations in SLC26A



IP-II. Absent apical interscalar septum and modiolus in a 9 y/o female with sensorineural hearing loss

Cochlear Incomplete Partition Type III IP-III

- Characterized by the complete absence of the modiolus with preserved interscalar septa, resulting in the pathognomonic “cork-screw” appearance of the cochlea
- Also features a thin otic capsule and bulbous internal auditory canal with a constellation of other findings possible
- Associated with mixed conductive and sensorineural hearing loss
- Mild to moderate disease may be managed with hearing aids while severe hearing loss makes patients candidates for cochlear implant
- X-linked, associated with mutations in POU3F4 and COL4A6



IP-III. Absent modiolus with preserved interscalar septa showing classic corkscrew appearance in a 3 y/o male with mixed hearing loss

Surgical Considerations for Cochlear Implants

- Radiologists must identify findings which may affect surgical planning and management
  - IP-I
    - Absence of the modiolus makes straight electrodes preferable
    - Increased risk of meningitis, spontaneous CSF leaks, and CSF gushers due to defects in the lamina cribrosa
  - IP-II
    - Electrode insertion challenging due to abnormal coiling, care is needed to avoid misplacement or trauma
    - CSF leakage due to the incomplete partition, requires close monitoring during and after surgery to manage potential complications
  - IP-III
    - Close monitoring for cerebrospinal fluid (CSF) leak or gusher

Teaching Points

- IPs are a subgroup of congenital bony malformations involving the cochlea
- IP-I results from failure to form the modiolus and interscalar septum, resulting in a single and cystic appearing cochlear cavity that gives a “figure 8” appearance on imaging
- IP-I should be differentiated from common cavity malformation
- IP-II occurs when the interscalar septa to fail to form, resulting in a “baseball cap” appearance on imaging
- IP-III is an X-linked genetic disorder resulting in complete absence of the modiolus with preserved interscalar septa, resulting in the pathognomonic “cork-screw” appearance of the cochlea
- Severity of hearing loss and treatment options vary among IP-I, IP-II, and IP-III
- Recognizing the distinct characteristics, genetic associations, and surgical challenges associated with each type of cochlear incomplete partition is essential for optimizing patient outcomes
- Accurate radiologic identification of cochlear incomplete partitions is crucial for clinical decision making and management

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