

Temporal Bone Anomalies

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A. **Developmental defects of EXTERNAL and MIDDLE ear** (both are derived from first and second branchial arches and clefts):

- 1) deformity of auricle
 - 2) partial or complete atresia of external auditory canal
 - 3) ossicular abnormalities
- **conduction defects** - may be surgically correctable.
 - may be accompanied by anterior **malposition of facial nerve** (CN7 location must be identified preoperatively!).

Atresia of external auditory canal (coronal CT) - external canal is not visible because of bony atresia (*white arrow*); mastoid segment of CN7 canal (*arrowheads*) is anteriorly displaced into same plane as vestibule* - very susceptible to injury as atresia plate is surgically removed.

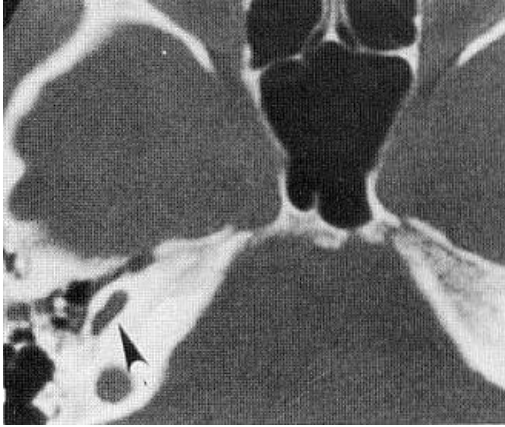
*normally, canal is at level of posterior semicircular canal.



B. **Developmental defects of INNER ear** (derived from auditory vesicle) - vary from abnormalities involving solely semicircular canals to complete aplasia of otic capsule.

- **sensorineural deafness** - not surgically correctable (may be helped by cochlear implants).
- **MONDINI defect** - partial or total absence of cochlear turns.

Mondini's defect (axial CT) - right cochlear turns are absent except for portion of basal turn (*arrowhead*); left cochlea was normal:



Source of picture: John H. Juhl "Paul and Juhl's Essentials of Radiologic Imaging", 7th ed. (1998); Lippincott Williams & Wilkins; ISBN-10: 0-397-58421-0 >>

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