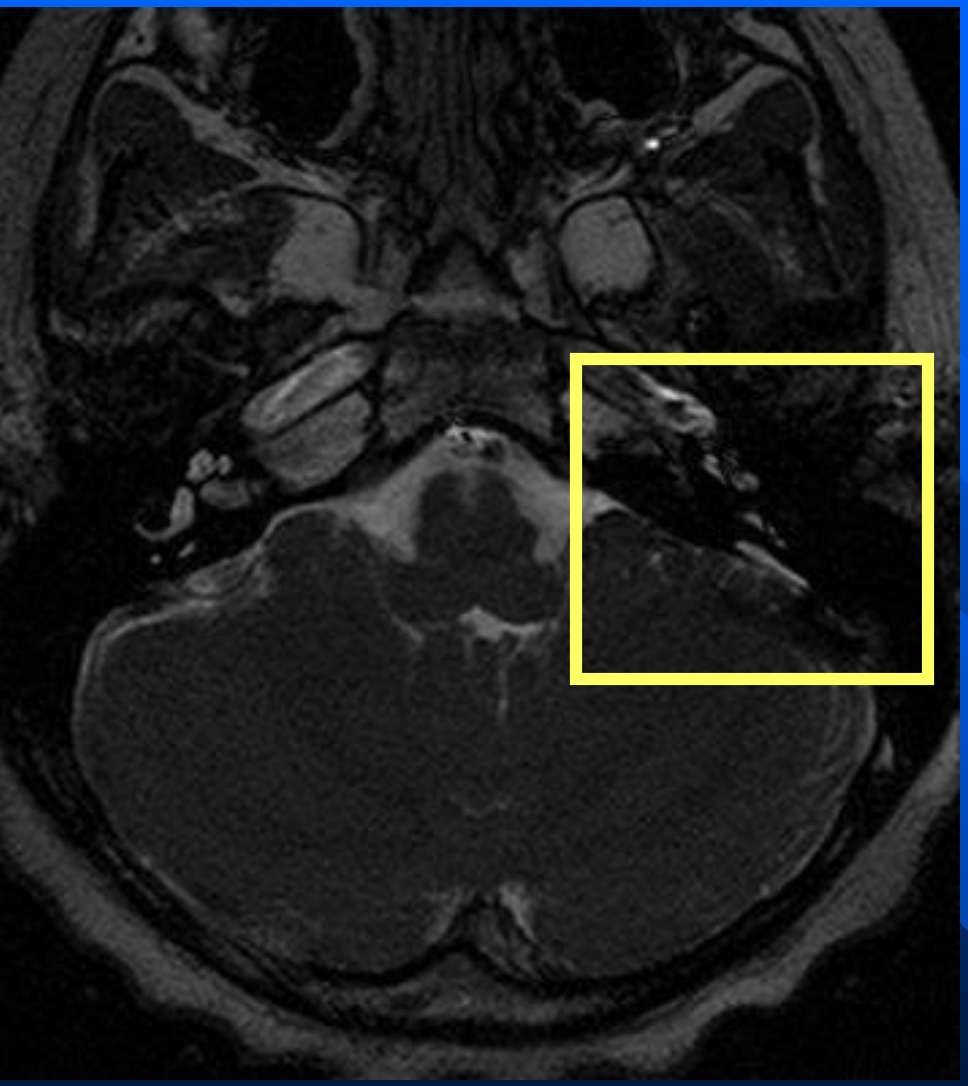
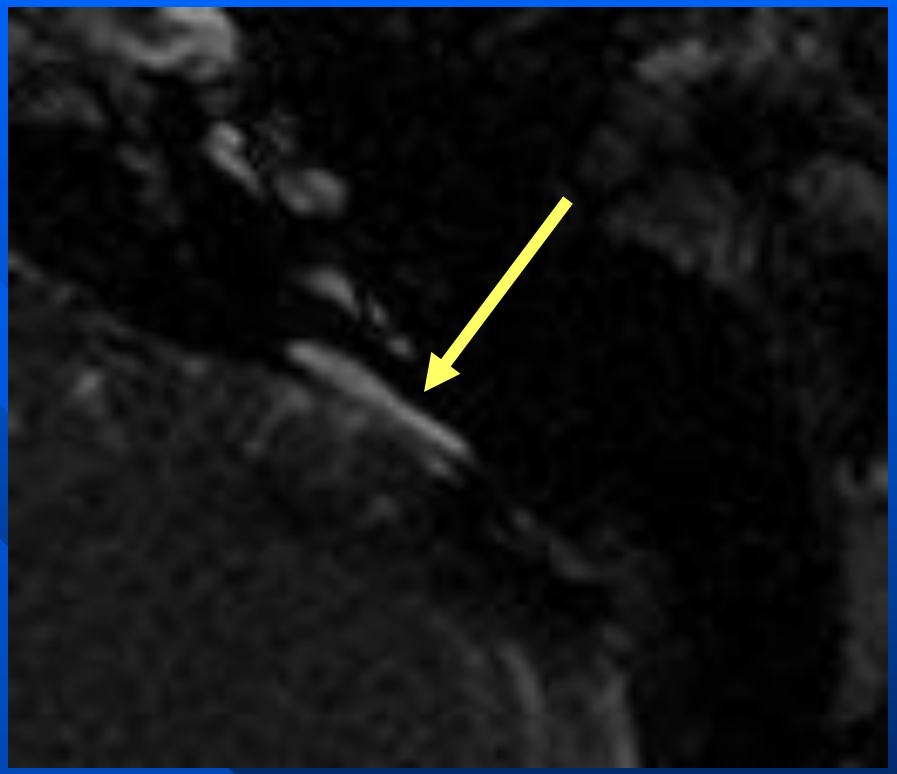
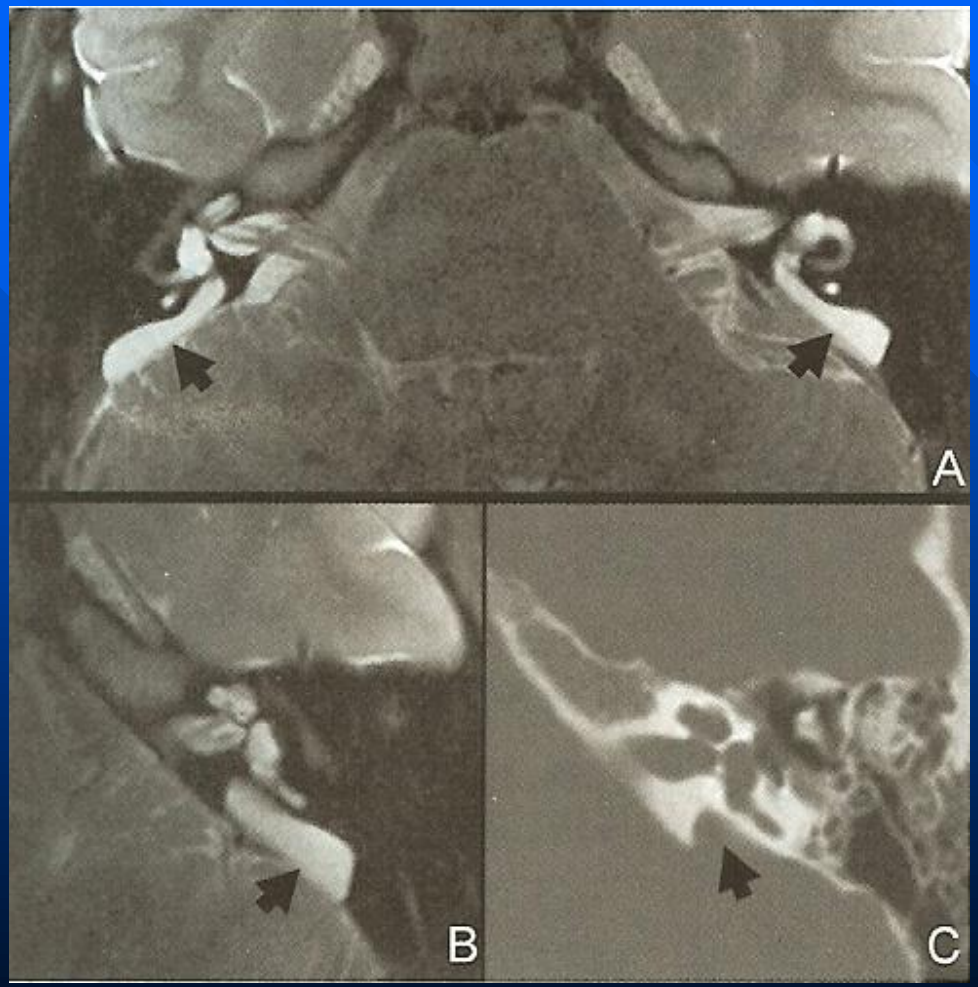
Large Endolymphatic Sac Anomaly

- *Large vestibular aqueduct syndrome*
- *Arrested development of inner ear with large sac and mild cochlear dysplasia **(8-9 week arrest) autosomal recessive***
- *Acquired SNHL (90%); 2nd to minor head trauma, the cochlea is **fragile***
- *Best imaging clue is large sac **mid duct > 1.5 mm or exceeds the posterior semi-circular canal***
- *90% are bilateral*
- *75% have associated cochlear dysplasia.*
- *Cochlear implant when SNHL is profound.*

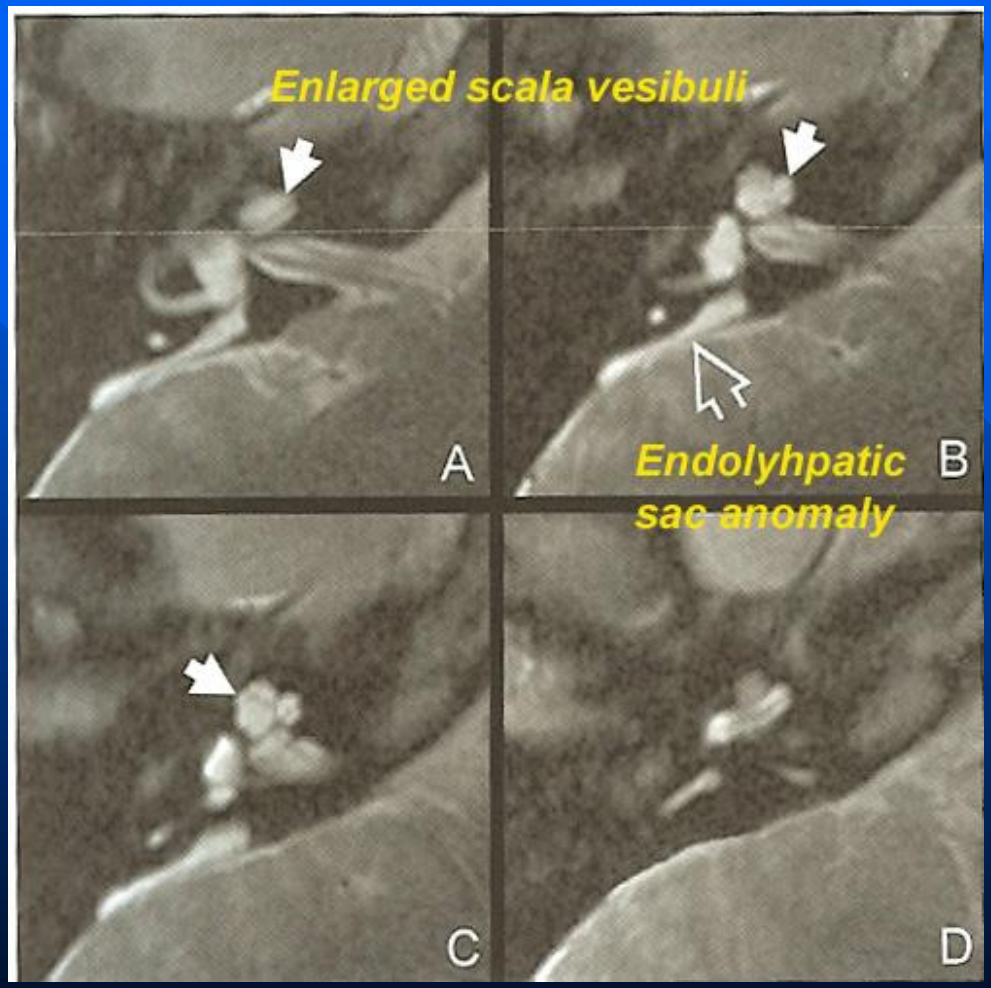


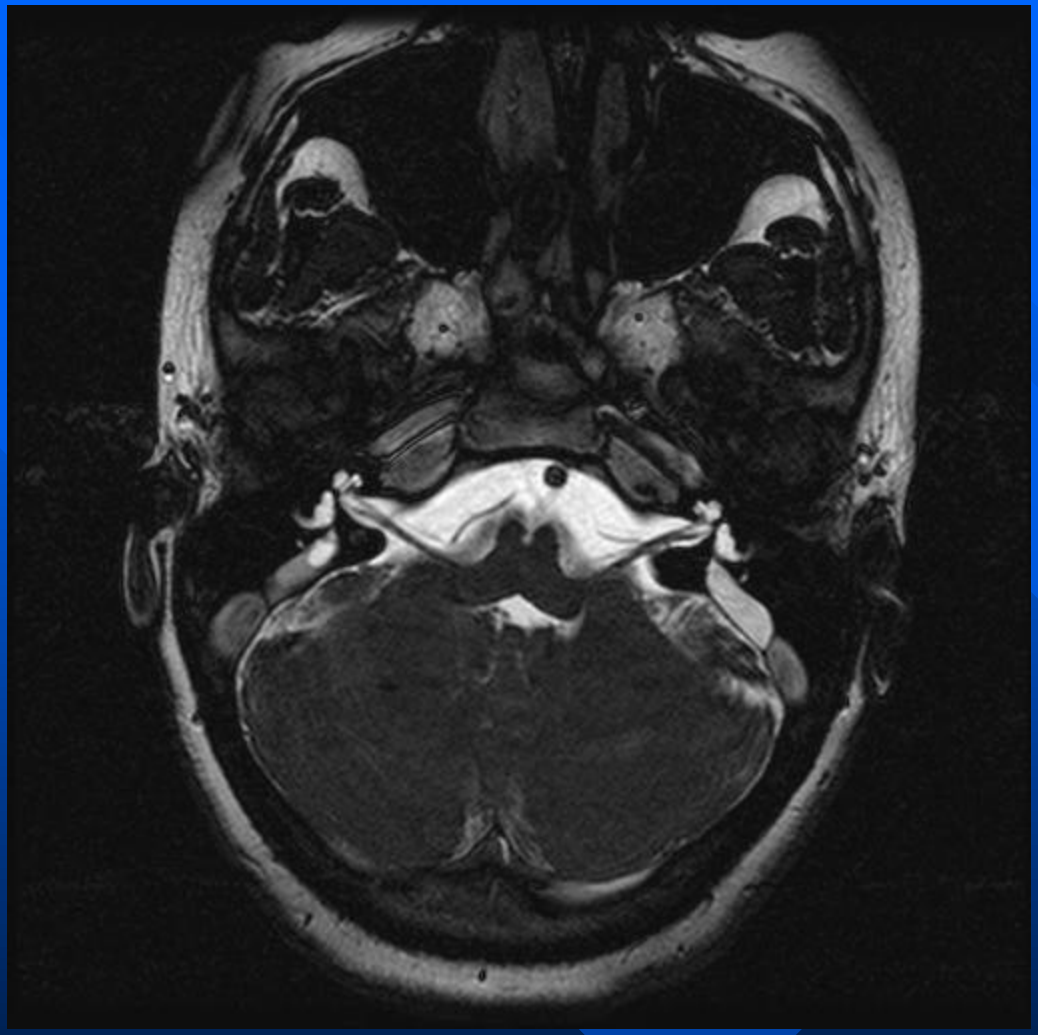


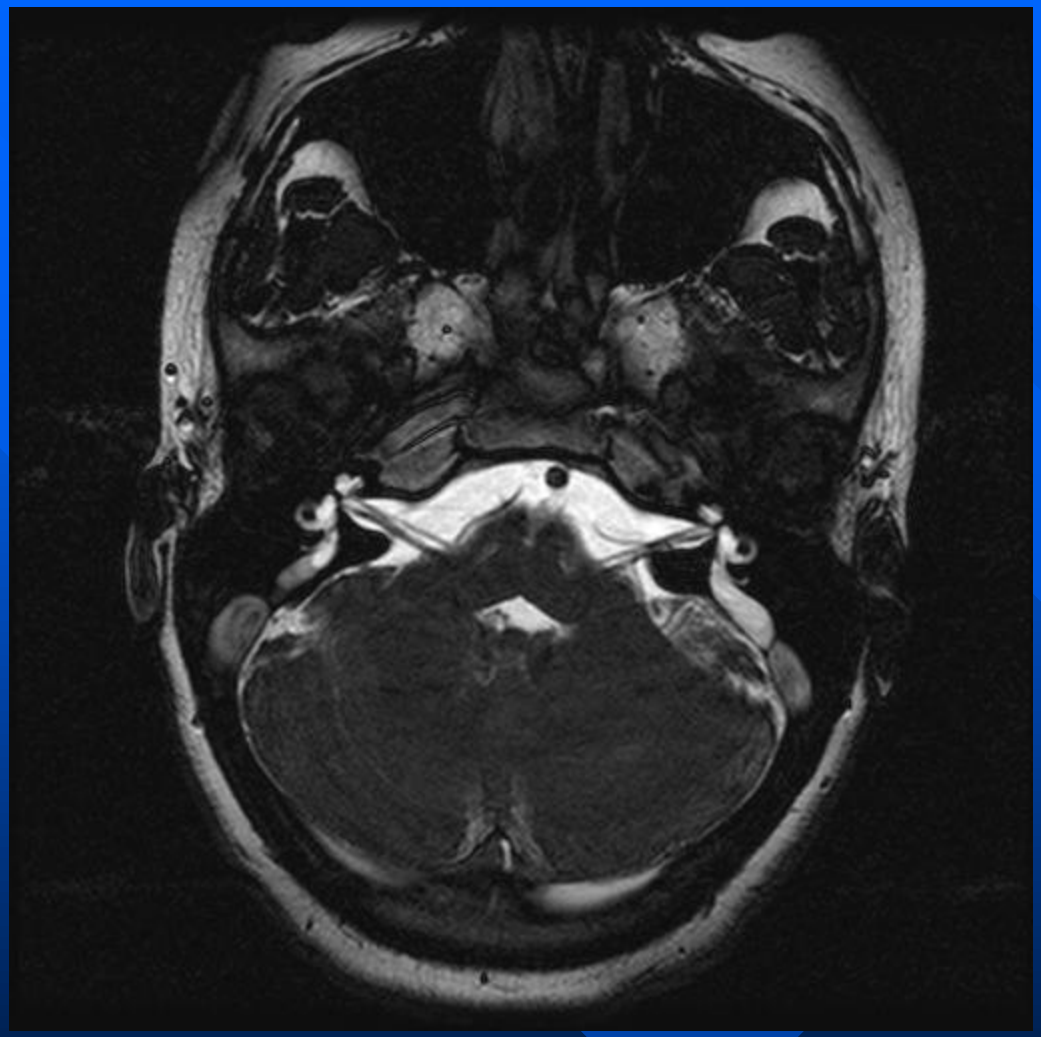
Endolymphatic sac anomaly

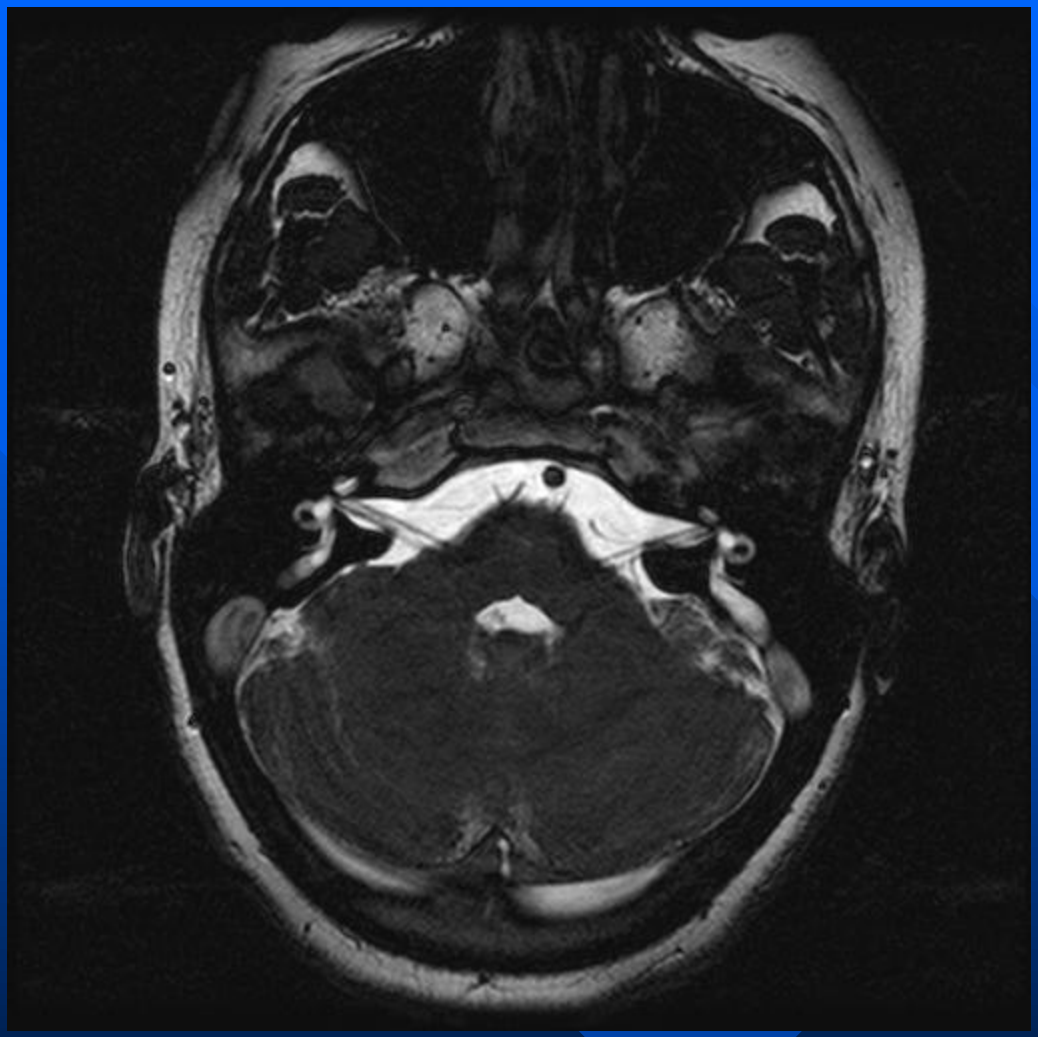


Mild Cochlear Dysplasia









Large Endolymphatic Duct & Sac

- Clinical Features

– Unilateral or bilateral (90%) SNHL
– Hearing may be present at birth
– Progressive or sudden hearing loss
  » often secondary to mild trauma
– Treatment options limited to cochlear implantation

Large Endolymphatic Duct & Sac

- Imaging Features

– CT: Large *bony* vestibular aqueduct

– MR: Large endolymphatic duct & sac

– MR: Cochlear anomalies common

» Absence of modiolus, partition defect

– MR is *preferred* imaging tool

"Cochlear Anomalies Associated with Large Vestibular Aqueduct Syndrome"

- *Davidson HC, Harnsberger et al*
  - » AJNR 1999 Sep;20(8):1435-4
  - – 63 ears with LEDSS; 60 controls
  - – *76%* had cochlear anomalies
    - » Modiolus deficient or absent, scalar chamber asymmetry, dysmorphism,
  - – 40% had vestibular anomalies
    - » Enlarged lateral SCC

Pendred Syndrome

- SNHL, usually congenital
- Bilateral Dilated Vestibular Aqueduct +/- Cochlear hypoplasia
- Euthyroid Goiter in late childhood/early adulthood