Congenital and Acquired Hearing Loss: a Review with Emphasis on MRI Findings

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LEARNING OBJECTIVES

• To become familiar with the normal anatomy of the inner ear & of the 8th cranial nerve.
• To understand the embryology of the inner ear as it relates to congenital hearing loss.
• To review examples of various common causes of congenital and acquired hearing loss with emphasis on their MRI findings.

NORMAL ANATOMY

MRI: CISS axial images showing the normal inner structures and nerves. Vestibule (V), Cochlea (C), Cranial Nerve VIII (8), Lateral Semicircular Canal (LSC), and Modiolus (M).

NORMAL ANATOMY

MRI: Axial CISS image shows (clockwise): anterior inferior cerebellar artery (AICA), cochlear nerve (CN), scala vestibularis (SV), modiolus (M), basilar membrane (BM), scala tympani (ST), vestibular nerve (VN), and vestibular ganglion (VG).
NORMAL ANATOMY

MRI: Sagittal reformation from CISS axial images shows: facial Nerve (FN), Cochlear Nerve (CN), Vestibular Nerve Inferior Division (INF), and Vestibular Nerve Superior Division (SUP).

EMBRYOLOGY OF THE INNER EAR

• Three primary phases of development include:
  – Development- weeks 4-8 of life
  – Growth- weeks 8-16 of life
  – Ossification- weeks 16-24 of life

DEVELOPMENT

The Otic placode develops during weeks 4-8 of life, beginning as a plaque of neural ectoderm lying between 1st branchial groove & hindbrain.

DEVELOPMENT

Otic placode invaginates to form the otic pit.
DEVELOPMENT

• Enlargement & invagination of the otic pit leads to separation from overlying tissues to form the otocyst.
• The otocyst divides into a dorsal pouch (becomes utricle & semicircular canals) & a ventral pouch (becomes cochlear duct & saccule).
• The Endolymphatic duct arises separately.
• At end of week 8, membranous labyrinth & cochlea are completely formed
• Cartilagenous condensation & ossification begins at this time as well.

GROWTH- INNER EAR

• Occurs between weeks 8-16 of life.
• The otic capsule recruits vascular channels via the fissula ante fenestrum. This structure is located in the anterior aspect of the oval window (remember that later in life it plays an important role in the development of otospongiosis of the fenestral type.)

OSSIFICATION

• Cartilagenous condensation continues.
• Ossification occurs via 14 separate ossification centers (no growth plates).
• Complete by 24 weeks; fetus is then capable of hearing.
• Ossification complete as endochondral bone.

IMAGING FEATURES OF SPECIFIC RELATIVELY COMMON CAUSES OF CONGENITAL AND ACQUIRED HEARING LOSS
COCHLEAR APLASIA

- Due an arrest of development in the 5th week.
- Unusual lesion, representing 3% of cochlear malformations.
- Characterized by absent cochlea with remnants of vestibule & semi-circular canals.

MRI in cochlear aplasia: CISS axial images demonstrate an absent cochlea on the right (circles). Compare with left cochlea.

ABSENT COCHLEAR NERVE

- May be isolated or in combination with lack of cochlear formation
- Due to lack of cochlear stimulus to formation of intracochlear apparatus
- CT may show absence of bony canal at center of basal turn of cochlea
- MRI may show absent nerve
- Critical to establish presence of nerve before cochlear implantation

Absent Cochlear Nerve. CT: The cochlear nerve canal is absent (arrow) and the cochlea is small. MRI: Axial CISS image demonstrates absence of the cochlear nerve on the right, compare with normal left cochlear nerve (arrow).
COCHLEAR DYSPLASIA

• Most common inner ear malformation (55%):
  – Incomplete partition of cochlea.
• Classic example is Mondini deformity:
  – Insult during 7th week of life.
  – Cochlea develops only 1.5 turns, lacking spiral lamina & interscalar septum.
  – Apical & middle turns are confluent, basal turn is preserved.
  – Associated anomalies of vestibule, semicircular canals & endolympathic in 20% of patients.
  – Development of organ of Corti & auditory nerves is variable.

COCHLEAR DYSPLASIA WITH ABSENT SEMICIRCULAR CANALS

CT: The cochleas (circles) are dysplastic and lack internal structures. The vestibules are large. MRI: Axial CISS images demonstrate enlarged and featureless cochleas (circles) and vestibules. The nerves (arrows) are present.

LARGE VESTIBULAR AQUEDUCT SYNDROME

• Most common imaging found abnormality in pediatric congenital hearing loss.
• Insult during 4-8 weeks.
• Vestibular aqueduct is large when >1.5 mm at midpoint.
• Size of dilated endolympathic sac has no correlation with size of duct.
• Often presents as decremental hearing loss.
• Cochlear dysplasia always present.
Mild Enlargement of Vestibular Aqueduct and Cochlear Dysplasia

CT: The opening for vestibular aqueduct is enlarged (arrow). The cochlea is dysplastic with fusion of its middle and apical turns. MRI: Axial CISS image shows enlarged endolymphatic sac with high T2 signal (arrow).

LARGE ENDOLYMPHATIC SAC

MRI: T2 axial image shows enlarged aqueducts (white arrows) and a very large right sac (black arrow). The cochleas are dysplastic.

MARKEDLY ENLARGED ENDOLYMPHATIC SACS

MRI: Axial CISS image demonstrates markedly enlarged endolymphatic sacs bilaterally (arrows).

LARGE VESTIBULAR AQUEUDCTS

CT: Spontaneous pneumo-labyrinth in a patient with vestibular aqueduct syndrome shows air in endolymphatic sac (black arrow) and sac (white arrow). Air is also seen in the vestibule while the cochlea is dysplastic.
**COMMON CAVITY DEFORMITY**

- Deformity results from insult during 4-5 weeks of life between formation of otocyst & differentiation into primordia of inner ear structures.
- Entity represents 25% of cochlear malformations.
- Size of cyst usually < than 1 cm.

**LABYRINTHITIS**

- Inflammatory disease of membranous labyrinth associated with hearing loss & vertigo.
- Classified by agent: meningogenic (bacterial,viral) or etiology (serous, suppurative).
- Begins with inflammation, proceeds to fibrous stage, and may culminate in ossifying labyrinthitis over months to years.

**FIBROSING LABYRINTHITIS**

- MRI: Axial CISS image shows decreased T2 signal in modiolus of cochlea (circle) with small amount of fluid in the scalae tympani and vestibularis. CT: Hazy increased density is seen in the cochlea (arrow) corresponding to early ossification of the inner structures of the cochlea.
FIBROSING LABYRINTHITIS

MRI: Axial CISS image shows subtle decreased T2 signal in scalas due to thickening of the modiolus and basilar membrane.
CT: Hazy increased density is seen in the modiolus region (arrow) secondary to early ossification.

LABYRINTHITIS OSSIFICANS

MRI: Axial CISS image (center) shows decreased T2 signal in the cochleas (circles). The right one is partially seen while in the left only minimal residual high T2 normal signal is present. CT: Increased bony density is seen in the cochleas (arrows). Note that the extent of calcifications does not correspond with the degree of the abnormality as seen on MRI due to the fact that MRI shows not only ossification but also the effects of fibrosis.

LABYRINTHITIS OSSIFICANS

CT: Axial images demonstrate markedly increased bony density in the cochleas which are nearly obliterated by ossification. The vestibules and semicircular canals are also affected.

VIRAL LABYRINTHITIS

• It is probably the most common cause of infectious labyrinthitis.
• Radiologic are findings becoming more apparent as imaging technology improves.
• Sensorineural hearing loss is most often accompanied by vertigo.
• Look for enhancement in the eighth cranial nerve and/or inner ear structures.
Presumed viral labyrinthitis. MRI: T1 post-contrast images demonstrate abnormal enhancement in the vestibules and cochleas bilaterally (circles).

COCHLEAR OTOSPONGIOSIS

- Also known as retrofenestral otosclerosis.
- Replacement of normal endochondral bone with Haversian bone, most likely secondary to an inciting inflammatory event.
- Female predominance with majority of cases bilateral.
- Appears in second to third decade of life.
- Presents as focal or diffuse demineralization of the otic capsule with abnormal enhancement.

Cochlear otospongiosis. CT: Coronal and axial images demonstrate diffuse demineralization of the otic capsule (circles).

MRI: T1 axial and coronal post-contrast images demonstrate abnormal signal in the otic capsule (circles).
CRANIAL NERVE VIII SCHWANNOMA

- Intralabyrinthine schwannomas are much more rare than those in the internal auditory canal.
- May originate from the cochlear nerve, the utricle, cochlea, and the vestibule.
- Presents as erosive or expansile mass with intense contrast enhancement.

CONCLUSIONS

- Knowledge of inner ear anatomy and embryology are fundamental to a basic understanding of the causes of congenital and acquired hearing loss.
- As many of the causes of hearing loss present with similar clinical findings, familiarity with imaging findings is critical in making the correct diagnosis.

REFERENCES

- www.med.unc.edu/inner-ear