CT and MR Imaging In Patients Undergoing Evaluation for Cochlear Implantation

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Learning objectives

After viewing this exhibit, the viewer should be able to

- Identify the normal anatomy of the inner ear, and cochlea on both CT and MR Imaging
- Assess and classify congenital and acquired abnormalities and pathologies of the inner ear, cochlea and vestibulocochlear nerves
- Discuss the suitability or otherwise of patients being evaluated for cochlear implantation and make recommendations to the treating physician

Background

- Sensorineural hearing loss has an estimated prevalence of 2 to 3 in 1000 neonates and deafness is associated with major developmental and socioeconomic implications
- Cochlear implantation has become a recognised treatment option for many of these patients William FHouse developed the first cochlear implant system in 1960s. FDA approval for implants was granted in 1985 for adults and 1990 for children
- Cochlear implantation is indicated in severe to profound hearing loss which is not adequately treated with standard hearing aids
- A cochleostomy is done followed by insertion of an electrode array, usually via the round window into the scala tympani and basal turn of cochlea
- Various types of electrodes are available and the choice depends not only on the surgeons preference, but also on the anatomy of the cochlea

Role of Pre Implant Imaging

1. To Rule out Conditions which may contraindicate implantation
   - Cochlear Aplasia
   - Cochlear Nerve Absence
   - Severe Cochlear Ossification

2. To Image Conditions which may modify the surgical approach
   - Congenital Anomalies of Cochlea
   - Choose The Better ear for implantation
   - Cochlear Fibrosis
3. Image Conditions Associated with Increased incidence of Complications

- Dilated Vestibular Aqueduct
- Absent Modiolus -CSF/Perilymph Communication

Findings and procedure details

Imaging Modalities and Normal Anatomy

Computed Tomography

- High Resolution CT using sub milimetre collimation and small field of view
- Multiplanar reconstructions using Minimum intensity projections and Volume Rendered Images
- CT provides superb bony detail. The axial and coronal CT anatomy are displayed in figure 2, 3 and 4

Magnetic Resonance Imaging -

- Minimum field strength of 1.5 Tesla. A heavily T2W sub millimetre sequence is always acquired (CISS/FIESTA/DRIVE) with a screening T2W sequence of the brain for visualisation of membranous cochlea and cisternal segments of the facial and vestibulocochlear nerves.
- Oblique reformats are acquired perpendicular to internal auditory canal (IAC) for visualisation of cochlear nerve in IAC
- VR, MIP and MPR images of cochlea for morphology of cochlea (Figs 5 and 6)

Imaging Check List

Cochlea -Morphology, Turns, Interscalar Septum, Modiolus, Signal

Cochlear Nerve

Internal Auditory Canal

Vestibular Aqueduct

Semicircular Canals

Round and Oval Window

Facial nerve course
Embryology of the Inner Ear - (Figure 7)

Malformations of the Inner Ear

Fig. 8: Image showing timings and line diagram appearance of major cochlear anomalies


(From References 7&8)

Jackler (6) in 1987 proposed an embryologic basis for cochlear malformations. This classification was modified by Sennaragolu and Saatci in 2002 (7) and this classification is the most widely accepted at present and is the basis for further discussion in this presentation

Labyrinthine Aplasia - (figure 9)

Failure of development of otic vesicles at 3rd week thus all inner ear structures are absent (8) or

May be associated with skull base anomalies, hypoplastic petrous apex, aberrant ICA etc

Absence of cochlear promontory differentiates it from Labyrinthitis ossificans. This malformation is a complete contraindication for cochlear implantation

Cochlear Aplasia (figure 10,12)
Complete absence of cochlea. \((7,8,9)\) Vestibule and semicircular canals variably formed. Differentiate vestibule from cochlea by position of structure anterior or posterior to the IAC. IAC is anterior to cochlea whereas vestibule is posterior to it. Differentiate from cochlear ossification by presence of promontory

Cochlear implant is not indicated in this malformation

**Common Cavity** (figure 11,12))

In common cavity malformation, all the inner ear structures fuse into an irregular common cavity. The IAC is either enlarged or small and generally communicates with the common cavity and may give rise to a gusher during cochleostomy. Cochlear implant may be beneficial in such patients. A full band electrode is usually indicated in these patients and they have a variable response to implantation

**Cochlear Hypoplasia** (figure 13,14)

A hypoplastic cochlea is smaller than normal and has a reduced number of turns. It may be represented only by a small cystic cavity, however the cochlea is seen anterior to the IAC

**Incomplete Partition (type I)** (figure 15,16)

Also known as cystic cochleovestibular malformation. In IP (type I) Cochlea and vestibule lack internal architecture. The cochlea is cystic without a modiolus. The vestibule is also dilated with the dilated cochlea and vestibule giving a figure of eight appearance. The IAC is usually wide with communication between IAC and Cochlea Semicircular canals may be dysplastic. Vestibule is not dilated. High risk of gusher and post operative meningitis during surgery. Usually a full band electrode is used for such patients. Has a worse response to CI than IP II.

**Incomplete Partition (typell)/Mondini Malformation** (figure 17)

Mondini malformation has 3 major components (i) normal basal turn with cystic apex due to incomplete partitioning of apical and second turns (ii) minimally dilated vestibule (iii) enlarged vestibular aqueduct. Basal turn is usually preserved. These patients show good results after cochlear implantation

**X Linked Deafness** (figure 18)
Sometimes known as Incomplete Partition type III, X linked deafness is a genetic disorder associated with a mutation in the POU3F4 gene located on the X chromosome.

Male patients present with progressive mixed hearing loss. Female carriers have normal or mildly impaired hearing.

High risk of gusher during surgery \(^{(11)}\)

Perilymph communicates with the subarachnoid space with increased intra cochlear pressure and progressive hearing loss

**Cochlear nerve Deficiency** \(^{(12)}\) (figure 19.20)

Thickness of the normal cochlear nerve should be equal to the thickness of the adjacent facial nerve.

The division of the vestibulocochlear nerve into its divisions occurs at a variable length and is complete only in the distal portion of the IAC \(^{(12)}\)

Normal Internal auditory canal should be more than 3mm in diameter. Vestibulocochlear nerve development is related to development of IAC, and cochlear nerve deficiency is more commonly associated with small or hypoplastic internal auditory canals. Acquired cochlear nerve degeneration may be seen in patients with normal sized canals. Patients in whom cochlear nerves are not clearly identified on MRI should undergo further testing before undergoing cochlear implantation.

**Enlarged Vestibular Aqueducts** (figure 21)

Enlarged vestibular aqueducts are diagnosed either when they are more than 1.5 mm in size, or when larger than the adjacent semicircular canals.

Patients present with fluctuating sensorineural hearing loss.

Associated with incomplete partition (Type II) and Pendreds syndrome.

At increased risk of CSF gushers during cochlear implantation, but do well audiometrically \(^{(9)}\)

**Lateral Semicircular Canal Dysplasia** (figure 22)
Abnormalities of the lateral semicircular canals are the most commonly seen inner ear malformations, possibly because it is the last to develop. It may or may not be associated with decreased hearing.

**Absent Lateral Semicircular canal** (figure 23)

Absent lateral semicircular canals may be syndromic and are associated with CHARGE syndrome - Coloboma, Heart Defects, Choanal atresia, Mental retardation, Genitourinary and ear anomalies. Cochlear anomalies and absent cochlear nerves with stenotic IAC are often seen as in our patient.

**Obliterative Labyrinthitis** (figures 24 to 27)

Meningitis in childhood may cause labyrinthitis with inflammation of the membranous labyrinth. Generally begins in the basal turn of the cochlea\(^9\).

It causes replacement of the fluid in the labyrinth with fibrous and later osseous material which damages the nerve cells and organ of corti.

CT may not reflect the fibrous component of the oblitative labyrinthitis although it is very good at picking up calcium.

MRI is the gold standard for evaluation of fibrous obliteration of the membranous labyrinth.

MR Imaging is more sensitive than CT in detecting early fibrotic changes in patients suspected to have oblitative labyrinthitis. MR picks up loss of hyperintense signal whereas CT picks up later stages with calcification.

MR imaging can precisely show the diameter of the cochlea and length of the affected segment and thus provide a road map for the surgeon as to how far to drill and where to perform the cochleostomy.

**Miscellaneous Conditions**

A number of other conditions may be encountered while evaluating patients for sensorineural hearing loss including **Otosclerosis** (figure 28) which leads to demineralisation of cochlea and stapes fixation, **Osteogenesis Imperfecta** (figure 29) which has similar imaging findings, and in our case the patient had an associated
perilymph fistula. **Neurofibromatosis** (figure 30)- which may present with hearing loss. These conditions are illustrated in this review, but the list is by no means exhaustive.

**Images for this section:**

![Diagram showing Internal and external portions of a cochlear implant.](image)

**Fig. 1:** Diagram showing Internal and external portions of a cochlear implant. The microphone worn on the ear picks up sounds which are digitised by the speech processor. The transmitter sends these signals through the scalp to the receiver and the impulses are sent into the electrode array which directly stimulates the auditory nerve.
**Fig. 2:** Axial CT images showing Normal Axial anatomy of the Inner Ear. Sections through the superior aspect of the IAC reveal the Facial nerve exiting the canal to form the genu. The 'signet ring"appearance of the Lateral Semicircular canal is also seen. Subsequent sections reveal the tympanic and descending segments of the facial nerve canal, the apical, basal and second turns of cochlea and the vestibule.
Fig. 3: Coronal CT Sections Showing Normal Anatomy The 'snake eyes' of the facial nerve (FNv) formed by the labyrinthine and tympanic segments is noted. The turns of the cochlea are well seen on the coronal images, as are the oval window and the descending facial nerve canal. Scrolling through coronal images is an easy way of confirming completeness of cochlear turns.

Fig. 4: Volume Rendered CT Images of cochlea revealing normal cochlear turns, vestibule and semicircular canals.
Fig. 5: Axial and Coronal Oblique T2W images The Images reveal superb detail with delineation of the normal cochlear turns, vestibule and semicircular canals. The interstellar septum is seen as a hypo intense line in the bright fluid signal in the cochlea
Fig. 6: Coronal Oblique Sections through IAC and Axial MIP T2W Images The coronal oblique sections reveal four hypo intense structures in the internal auditory canal. The cochlear nerve is aneroinferior, the facial nerve is anterosuperior and the superior and inferior vestibulocochlear nerves are seen along the posterior aspect of IAC. The modiolus is well seen on the axial MIP images.

Embryology (5)

- Ectoderm forms Otic Placode (3rd Week)
- Otic Placodes invaginate to form Otocysts
- Otocysts + Surrounding mesenchyme = Otic Capsule
- Endolymphatic Sacs formed followed by Cochlea and Vestibule (5th Week)
- Membranous Cochlea 1 to 1.5 turns by end of 6 Weeks
- 2.5 turns by end of 7th week
- Superior SCC, then Posterior, then Lateral - Adult appearance by 8th week

Fig. 7

<table>
<thead>
<tr>
<th>Cochlear Deformities</th>
<th>Gestational Age at Arrest (wk)</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Michel deformity</td>
<td>Third</td>
<td>Very rare</td>
</tr>
<tr>
<td>Common cavity</td>
<td>Between fourth and fifth</td>
<td>25%</td>
</tr>
<tr>
<td>Cochlear aplasia</td>
<td>Fifth</td>
<td>Uncommon</td>
</tr>
<tr>
<td>Cochlear hypoplasia</td>
<td>Sixth</td>
<td>15%</td>
</tr>
<tr>
<td>IP I (pseudo-Mondini)</td>
<td>Between sixth and seventh</td>
<td>Uncommon</td>
</tr>
<tr>
<td>IP II (classic Mondini)</td>
<td>Seventh</td>
<td>55%</td>
</tr>
</tbody>
</table>
**Fig. 8:** Image showing timings and line diagram appearance of major cochlear anomalies

**Fig. 9:** Michel Aplasia Axial CT and Coronal CT and MRI reveal non visualisation of ANY internal ear structure. The petrous bones appear hypoplastic abnormal in morphology

**Fig. 10:** Cochlear Aplasia CT images reveal absent cochlea with a red circle at the expected site. Vestibule is small and dysplastic
**Fig. 11:** The MIP MRI images reveal an irregular common cavity without any obvious internal architecture (red arrow) An irregular Semicircular canal is seen (yellow arrow) with a dilated IAC (green arrow)

**Fig. 12:** Common Cavity and Cochlear Aplasia Assymetric fluid attenuating structures in the inner ear of a patient.(Red arrows) No well defined cochlea is seen although a portion of the cystic structure just extends anterior to the IAC (green arrows) On the left side the structure appears continuous with the IAC. The VR images reveal a common cavity with a dysplastic semicircular canal (yellow arrow)
Fig. 13: Hypoplastic Cochlea CT and MRI reveal a small IAC with a cystic structure anterior to the IAC suggestive of a hypoplastic Cochlea (red arrows)

Fig. 14: Cochlear Hypoplasia MRI of another patient showing a small Left IAC (green arrow) with a hypoplastic cochlea (red arrow) and dysplastic cochlear on the left side on the right side(yellow arrow)
Fig. 15: Incomplete Partition Type I (IP type I) Axial MR shows dilated cochlea (red arrow) and vestibule (yellow arrow) giving a figure of eight appearance. The funds of the IAC appears continuous with the cochlea. MIP images confirm the findings and show lack of internal cochlear architecture along with irregularity of the Semicircular canals and dilated IAC.
Fig. 16: Incomplete Partition (Type I) Axial CT images reveal a dilated Internal Auditory Canal, (blue arrow) with dilated cystic appearing cochlea (red arrows) and vestibule (yellow arrows) suggestive of IP (I). Volume Rendered MRI images of another patient reveal dilated cochlea (red arrow) and vestibule (yellow arrow) with dysplastic semicircular canals (blue arrows).
**Fig. 17:** Mondini Malformation (Incomplete partition type II) Axial & Coronal CT images reveal a cystic cochlear apex (red arrows) with no differentiation of the apical and second turns. A grossly enlarged vestibular aqueduct is also seen (yellow arrows) Coronal Oblique MIP MRI images in another patient reveal a cochlea with 1.5 turns.

![Fig. 17](image)

**Fig. 18:** X linked Deafness (Incomplete Partition type III) Axial and coronal T2 W images reveal symmetrically dilated internal auditory canals (red arrows) and communication between the fundus of IAC and the cochlea. (yellow arrows)

![Fig. 18](image)

**Fig. 19:** Normal Cochlear Nerve MIP image in the left shows a normal sized IAC with the cochlear nerve seen extending upto the cochlea. High resolution T2 W oblique sagittal image reveals the cochlear nerve (green arrow) and the adjacent facial nerve (yellow arrow) to be of equal calibre.

![Fig. 19](image)
**Fig. 20:** Cochlear Nerve Deficiency Two patients with markedly reduced size of internal auditory canals (red arrows). In the patient on the left, only a single nerve (yellow arrows) is seen extending from the cistern into the IAC. In the patient on the right, although two nerves were seen, the cochlear nerve could not be clearly identified. Both patients fared poorly on further testing which suggested deficiency of the cochlear nerves.

**Fig. 21:** Enlarged Vestibular Aqueducts Axial T2W MRI (right) and HRCT (left) reveal dilated endolymphatic ducts (red arrows) and sacs (yellow arrows) The adjacent semicircular canals are marked by green arrows
Fig. 22: Lateral Semicircular Canal Vestibular Dysplasia Enlarged vestibule with dilated semicircular canal (red arrow) Lateral semicircular canal vestibular dysplasia
Fig. 23: Coronal MIP MRI Absent lateral semicircular canal with small cochlea and small IAC in a patient with CHARGE syndrome
Fig. 24: Obliterative Labyrinthitis Coronal (top) and axial CT images reveal a normal cochlea on the left side (green arrows) and almost completely ossified left cochlea (red arrows). The site of cochlea is marked by an oval in the bottom left image.

Fig. 25: MRI in Obliterative Labyrinthitis Coronal and Axial MRI (top) and MIP images (bottom) in a young child with history of meningitis. Images reveal areas of fibrosis/ossification suggestive of obliterative labyrinthitis, as loss of signal. Almost complete ossification of the left cochlea is seen in this patient. The right ear shows preserved cochlear signal except for loss of signal in the posterior semicircular canal (green arrow).
Fig. 26: Obliterative Labyrinthitis MRI findings in a patient with hearing loss reveal irregular areas of signal reduction and areas of loss of signal in the semicircular canals (yellow arrows) and cochlea (red arrows) bilaterally. The right cochlea and the left semicircular canals are most involved.

Fig. 27: CT Vs MRI in Obliterative Labyrinthitis. Coronal CT reveals normal attenuation in the basal turn of the cochlea. No calcification is seen (green arrow). The coronal MIP image reveals a short segment in the basal turn with loss of normal hypo intense signal (red arrows). The same finding is seen on the T2W base images (red arrows).
**Fig. 28:** Otosclerosis Axial CT images reveal pericochlear lucencies around Bilateral cochleae suggestive of Otosclerosis (red arrows)

**Fig. 29:** Osteogenesis Imperfecta with Perilymph fistula MIP MRI images reveal fluid around the basal turn of the cochlea and pericochlear region in a known case of osteogenesis imperfecta
Fig. 30: Neurofibromatosis Contrast Enhanced MRI reveals enhancing masses in the CP angle cisterns extending into the IAC with Enlarged Internal auditory canals suggestive of bilateral acoustic neuromas - Neurofibromatosis 2
Conclusion

- CT and MRI imaging are essential for pre-operative imaging of the cochlea and inner ear in order to provide a satisfactory outcome in patients being evaluated for cochlear implantation.

- Though both are complementary modalities, CT has advantages in visualising the bony structures whereas MRI has the upper hand in visualisation of the neural structures including cochlear nerves and early cochlear fibrosis. Both modalities together provide a detailed view of the morphology of the inner ear and the operative field to the surgeon.

- The demonstration of congenital or acquired anomalies of the inner ear may lead to rejection of a potential cochlear implant candidate, may alert the surgeon to possible complications such as gushers, may lead to a change in size or type of electrode or may lead to modification of surgical approach or site of cochleostomy.

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