Dissertation on

"PRE-OPERATIVE HIGH RESOLUTION CT&MR IMAGING FINDINGS IN CHILDREN WITH BILATERAL PROFOUND SENSORINEURAL HEARING LOSS"

Dissertation submitted in partial fulfillment

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MADRAS MEDICAL COLLEGE, CHENNAI.

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CERTIFICATE

This is to certify that this dissertation entailed **"PRE-OPERATIVE HIGH RESOLUTION CT&MR IMAGING FINDINGS IN CHILDREN WITH BILATERAL PROFOUND SENSORINEURAL HEARING LOSS"** submitted by **Dr.R.KALAIMANI,** appearing for M.S. ENT., Branch IV Degree examination in April 2018 is a bonafide record of work done by her under my direct guidance and supervision in partial fulfillment of regulations of the Tamil Nadu Dr. M.G.R. Medical University, Chennai, Tamil Nadu, India.

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ABSTRACT

"PRE-OPERATIVE HIGH RESOLUTION CT&MR IMAGING FINDINGS IN CHILDREN WITH BILATERAL PROFOUND SENSORINEURAL HEARING LOSS"

BACKGROUND: In 2012, world health organization (WHO) estimated 360 million people in the world suffer from disabling hearing loss. About 50% of the patients with sensorineural deafness are congenitally deaf and 30% of these congenitally deaf patients have syndromic deafness. High resolution computed tomography (CT) and magnetic resonance imaging (MRI) of the temporal bone are the imaging modalities of choice to evaluate the ear structures and nerves for cause of hearing loss and are now routinely being performed.

AIMS: To evaluate the role of various imaging modalities and are findings in pre- operative evaluation of cochlear implant in children with profound sensorineural hearing loss.

MATERIALS AND METHODS: In our study a total of 70 children with profound sensorineural hearing loss were included. All the children underwent both the modalities of radiological investigations (HRCT& MRI).

RESULTS: Majority of our children 58 (82.9%)showed normal HRCT &MRI of the temporal bone .A total of 17.1% (12 children) demonstrated various bony malformations of the cochlea-vestibular system, internal auditory canal and cochlear nerve aplasia **.** Majority of children had multiple abnormalities. 3

children (4.2%) of children had bilateral cochlear nerve aplasia. One child(1.4%)had complete labyrinthine aplasia, which are the absolute contraindication for cochlear implant surgery.4 children(5.7%) had Unilateral cochlear nerve aplasia with bilateral abnormal cochlea and vestibule, 2 children(2.8%) had bilateral Incomplete partition type 1,One child had Incomplete partition type 1 one side with common cavity other side, One child had Bilateral dilated vestibular aqueduct. Both modalities provided critical information on abnormalities of the otic capsule, pneumatisation of the mastoid, middle ear abnormalities, cochlear ducts patency and vascular abnormalities-thus helping to assess the suitability of the ear for implantation, determine the side to be implanted and to find any associated abnormality which could adversely influence the surgery.

CONCLUSION: HRCT Temporal bone and MRI of the inner ear are complementary to each other and provide exquisite vital anatomical details and information. Hence they are now considered as baseline investigations and are mandatory prior to cochlear implant surgery. It also helps in choosing the candidacy for surgery, side selection, and deciding the surgical technique in cochlear implantation.

KEYWORDS: Sensorineural hearing loss, HRCT, MRI

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1.INTRODUCTION

Hearing loss is the most common sensory deficit in human today. In 2012, world health organization (WHO) estimated 360 million people in the world suffer from disabling hearing loss. This constitutes 5.3% of world population. In India the prevalence and incidence of the hearing impairment is substantially high and it is preventable. In India 63million people (6.3%) suffer from significant hearing loss, which ranges from 4% in urban to 11% in rural and slum areas. 4 in every thousand children suffer from severe to profound hearing loss. About 50% of the patients with sensorineural deafness are congenitally deaf and 30% of these congenitally deaf patients have syndromic deafness. In India hearing impairment is second most common cause of disability.

Diagnostic imaging plays an important role in the evaluation and management of hearing loss. High resolution computed tomography (CT) and magnetic resonance imaging (MRI) of the temporal bone are the imaging modalities of choice to evaluate the ear structures and nerves for cause of hearing loss and are now routinely being performed. Also in general, to evaluate the bony structures of ear including the external auditory canal, middle ear ossicles, mastoid, petrous apex and bony labyrinth HRCT is a better modality, whereas for the membranous

labyrinth (luminal obstruction), identification of cochlear nerve MRI should be the imaging modality of choice. However, many a times both modalities are used together to complement each other. MRI with its higher soft tissue resolution and tissue characterization capabilities would be more frequently used in cases of sensorineural hearing loss where the pathology is generally in the cochlea or retrocochlear region(In the 8^{th} nerve or brain).

In cases of bilateral SNHL, the radiological imaging may be abnormal in 6.8% to 12.8% and in up to 30% of children.

It is vital to obtain a good quality high resolution CT and MRI of temporal bone in patients with SNHL with accurate technical parameters. Most centers are now equipped with multi slice spiral CT scanners, which is essential as reconstructions can be obtained in any plane with isotropic voxels in high end CT scanners without loss of resolution. MRI should be obtained on a high tesla strength magnet, 1T (tesla) or higher. In our institute, 64 slice spiral CT and 1.5T MRI machine are used.

2.**AIMS AND OBJECTIVES**

1. To pre-operatively evaluate the cochlea-vestibular anomalies, middle ear abnormalities, cochlear duct patency, presence of cochlear nerve, course of facial nerve and other abnormalities
2. To know the type of pneumatization of the mastoid
3. Finally to make surgical decisions regarding candidacy for surgery, side selection and surgical technique in cochlear implantation.

3.REVIEW OF THE LITERATURE

Robert K.Jackler et al (1987) explained the incidence of congenital inner ear malformations. 80 to 90 % of the inner ear anomalies are limited to the membranous labyrinth .10 to 20 % of inner ear anomalies are associated with radiographically detectable malformations, in which cochlear malformations are most common. But many cases demonstrate abnormalities in more than one portion of the inner ear.

Thomas L Eby et al (1996) studied the development of the facial recess and its implications for CI surgery. Facial recess was measured in 123 temporal bones ranging from 8 weeks in utero to 7 years after birth. They were found no growth after birth and concluded that the facial recess appears to be adult size at birth.

Woolley et al (1999) studied the pre-operative CT scan of temporal bone and it is uses in candidacy selection for cochlear implant surgery. In their study 6% of the children had high riding jugular bulb, 1.6% of children had anteriorly placed sigmoid sinus, 22% of the children had inner ear anomalies, and 8% of the children had cochlear ossificans.

D E Bamiou et al (1999) studied the HRCT temporal bone findings in children with bilateral sensorineural hearing loss. In their study, they reported 28.4 % of children have abnormal CT findings. Similar study done by **Woolford et al** who reported 29.5 % abnormal CT scans, **Shusterman et al** 12.85% and **Zalzal et al** reported an incidence of 6.8% abnormal CT findings in children with bilateral SNHL.

Harker et al (1999) assessed the perilymphatic gusher during cochlear implantation surgery in children with large vestibular aqueduct. They did 5 CI surgeries for a patient with LVAS. All children had uneventful surgical procedures without perilymphatic gushers. They concluded perilymphatic gushers may not be common with the LVAS syndrome.

L Sennaroglu et al (2002) did study to investigate the isolated absence of the cochlear nerve in the presence of normal CT in pre-operative evaluation of cochlear implant candidates. They reported in their study that isolated absence of the cochlear nerve was not detected in patients with congenital hearing loss who had normal temporal CT findings. CT scan was done for 27 children, 14 children had normal CT & MRI findings.13 children had abnormal inner ear finding on CT scan. Of these only 4 children had absent cochlear nerve.

B R Gastman et al (2002) did a study, to evaluate the risk of carotid injury in cochlear Implant Surgery. The distances between the basal turn of the cochlea and the internal carotid artery varies from 0.25 mm up to 7 mm. If the distance was small in between these two, there is increased chance of injury to the carotid artery during drilling of the basal turn of the cochlea and improper placement of the electrode.

MH.Khalessi et al (2003) correlating the CI surgical findings in patients with inner ear malformations. From 1997 to 2002 they were performed six CI surgery in children with inner ear anomalies, in which CSF gusher occurred in 4 patients (3 children with enlarged vestibular aqueducts, 1 child with common cavity), Facial nerve paralysis occurred in 2 patients (one with common cavity, another with LVA).In this study 27 percent of the children with inner ear malformations had facial nerve anomalies.

A Abdullah et al (2003) reviewed the preoperative High Resolution CT and MR Imaging in 46 Cochlear Implant patients. In their study majority of patients 73.9% (34 patients) showed normal findings in HRCT temporal bone,10.9% (5 patients) had labyrinthitis ossificans, 4.3% (2 patients) had mondini's deformity,4.3 % (2 patients) had middle ear effusion, one patient each had single cochlear cavity, hypoplasia of the internal auditory canal, high jugular

bulb. They concluded that above findings contributed to surgical decisions regarding candidacy selection, side selection, and surgical technique in cochlear implant surgery.

James et al (2004) have assessed the safety of cochlear Implantation surgery in children aged 12 months or younger is reviewed with radiological assessment of mastoid bone anatomy and surgical outcome. Under one year of age, approximately one third of the mastoid is pneumatised and one-third marrow filled at the level of the round window niche. At the age of 2 years pneumatization increases to approximately 60%. But pneumatization was always adequate for safe identification of surgical landmark during surgery**.** No complications occur in his study. They concluded CI surgery is safe even under the one year of age.

MiklosToth et al (2005) studied the development and surgical anatomy of the round window niche. The uneven growth of different walls of the round window niche can alter the shape of the entrance, which results in eight different types of niches. In their study normal shape of round window 34.8%, most common variants are open fundus 17%, followed by anterior septum 11.8%, and extremely narrow 11.2%. The rare variant is bony membrane.

A Chaturvedi et al (2006) evaluated the role of HRCT & MRI imaging modalities in pre and post-operative evaluation of CI candidates. HRCT has a main role in detection of inner ear anomalies and considered superior to MRI for detection of a high riding jugular bulb, tracing the course of the facial nerve. Potential advantage of MRI is in detection of labyrinthine ossification, large vestibular aqueduct and identification of cochlear nerve. They concluded that HRCT is recommended in all children for cochlear implant work up. MRI is advised in children with post meningitic deafness and doubtful HRCT findings.

Sennaroglu et al (2006) reported the surgical aspects cochlear implantation in inner ear malformation. CI is surgically feasible in children with inner ear anomalies like common cavity, incomplete partition types, and LVA, but surgeon should be ready to make the modifications in the surgical approach.

UdiCinamon (2009) studied the growth rate and size of the mastoid air cell systems and bone. He concluded three phases of mastoid pneumatisation from birth till reaching adult size.

Santhosh S Gupta et al (2009) reviewed the CT & MRI of congenital temporal bone anomalies in cochlear implantation surgeries. He said non visualisation of cochlear nerve in MRI, may be due to that the nerve is so thin, that it is beyond the resolution of a 1.5 T MRI or the cochlear nerve fibres traverse along the vestibular nerve. When the cochlear nerve is not well seen, intracochlear electrical stimulation to determine the auditory nerve action potential and auditory brainstem response may be a valuable test before performing CI surgery.

Santosh Gupta et al (2010) studied the imaging in sensorineural hearing loss patients. To evaluate bony labyrinth, CT is a better modality, whereas for the membranous labyrinth and suspicion of retrocochelear pathology MRI should be the best imaging technique. They concluded both CT and MRI are complimentary to each other and can be used together.

David R Friedmann et al (2011) evaluated radiological study on development of jugular bulb. They reported in their radiological study, children younger than 2 years of age did not demonstrate bulbous enlargement of the jugular bulb. They said, the jugular bulb is a dynamic

structure that is not present at birth, develops after 2 years and it is stabilized in adulthood.

Tarik EI Hadi et al (2012) studied the etiopathogenesis of spontaneous tegmen defect and semicircular canal dehiscence. Due to same embryological origin and common anatomical location both are exposed to the same etiopathogenic factors leading to defect in the both tegmen and semicircular canal. In their study 56.5% of the patients had SCCD combined to spontaneous tegmen defect. So we should always look for SCCD in case of STD is present in the HRCT temporal bone.

Gomes ND et al (2013) highlighted the importance of radiological imaging to select the candidacy for cochlear implantation surgery. They considered as absolute contraindications are cochlear nerve aplasia, labyrinthine or cochlear aplasia. Relative contraindications are labyrinthitis ossificans and complicating factors in CI surgeries are hypoplasia of the mastoid, aberrant course of facial nerve, otomastoiditis, otosclerosis, dehiscent jugular bulb, enlarged endolymphatic sac.

Ambrose Lee et al (2014) done a study, aimed to establish the frequency of postoperative bony dehiscence overlying the mastoid portion of the facial nerve after round window membranous cochleostomy using HRCT.

They found a radiological dehiscence rate of 40% but none of the patients in their series sustained a facial palsy. They concluded unroofing the bone overlying the nerve does not pose undue danger during cochlear implant.

Santosh S Gupta et al (2015) studied HRCT &MRI temporal bone in patients prior to cochlear implant surgery to assess the status of the inner ear structures. Their study showed on imaging, about 20% of patients with congenital sensorineural hearing loss had congenital malformations of the inner ear.

A S Jallu et al (2015) studied the radiological imaging evaluation of paediatric sensorineural Hearing loss. In their study 25% of the children have radiologically detectable inner ear malformations. The most common are large vestibular aqueduct, cochlear anomalies, IAC stenosis with cochlear nerve hypoplasia.

Sanjay vaid et al (2015) studied the role of HRCT and MRI of the temporal bone in predicting and grading the degree of difficulty of cochlear implant surgery. Image based10 – pointing scoring chart was used, which alerts ad prepare the surgeon to encounter the difficulties during surgery.

Kimberley S Noji et al (2015) explained the cochlear nerve better visualised in the direct parasagittal T2-weihgted 3D TSE (with DRIVE) images of the IAC when compared to reconstructed axial image. The appearance of cochlear nerve in the reconstructed image is highly variable and it might be underestimate the presence of cochlear nerve.

Sennaroglu et al (2016) highlighted the importance of pre-operative radiological findings in children with narrow facial recess undergoing CI surgery. They explained HRCT temporal bone provides adequate information about facial recess. If it is narrow CI surgery difficult to perform or facial nerve injury will occur. In this situation surgeon preoperatively can plan the alternative approach to reach the round window niche, which are the removal of part of the EAC and subsequent reconstruction with bone or cartilage graft, temporary anterior mobilization of the EAC, suprameatal approach, transcanal approach, Combined transcanal/transmastoid approach, Subtotal petrosectomy with middle ear obliteration, retrofacial and middle fossa approach.

KeremOzturk et al (2016) studied the anatomy of facial recess during temporal bone cadaveric dissection (totally 24 temporal bone). They reported

total round window exposure can be achieved through the facial recess in 79.2%.The RW was partially visible in the remaining temporal bones 20.8%. The unexposed part of the RW lay posteromedial to the FN in these five bones.

4. IMAGING TECHNIQUES

HRCT and MRI are currently the most widely used techniques for imaging the temporal bone. CT and MRI studies are complementary. Each technique has advantages and disadvantages, and often more than one examination is necessary for a complete temporal bone evaluation.

Imaging of children with congenital sensorineural hearing loss is frequently performed in an attempt to determine an underlying pathology. Both high resolution computed tomography scan (HRCT) of the temporal bone and magnetic resonance imaging scan (MRI) of the inner ear have been used in this set of children with certain advantages and disadvantages of each. The HRCT scan reveals many types of bony inner ear malformations and MRI scan provides better visualization of the membranous labyrinth and the status of vestibulocochlear nerves.

Computed Tomography:

The temporal bone has a high inherent radiation attenuation contrast, having both the dense bone in the body and air-filled spaces. High-resolution CT images are usually acquired with thin sections (0.5 to 1 mm) and special bone algorithms for high detail.

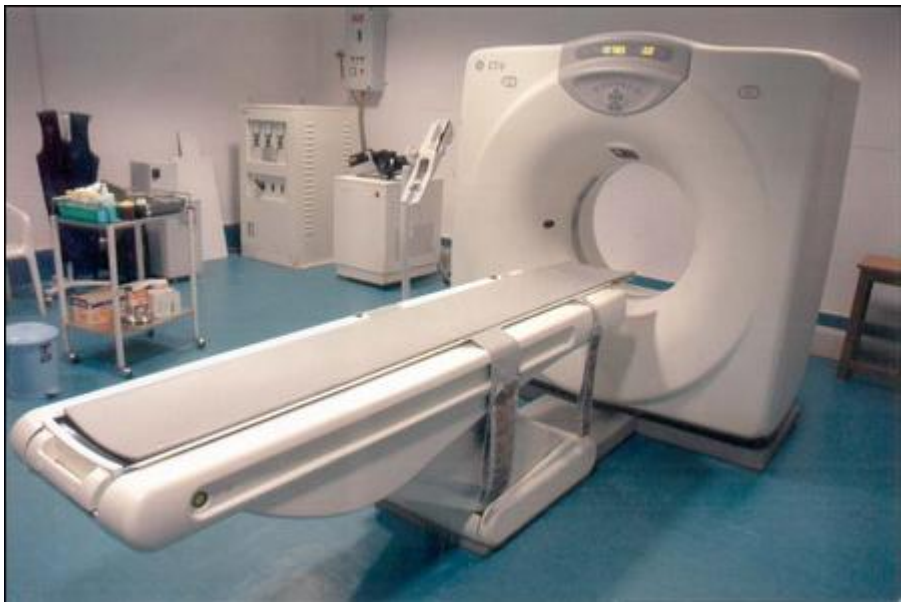


Figure 1: Photo of a CT Scanner

High-resolution CT of the temporal bone, followed by image reconstruction in both the axial and coronal planes, is required to evaluate the inner ear and its malformations. Axial scanning is performed in planes parallel to the infraorbitomeatal line, 30 Degree to anthropological base line.

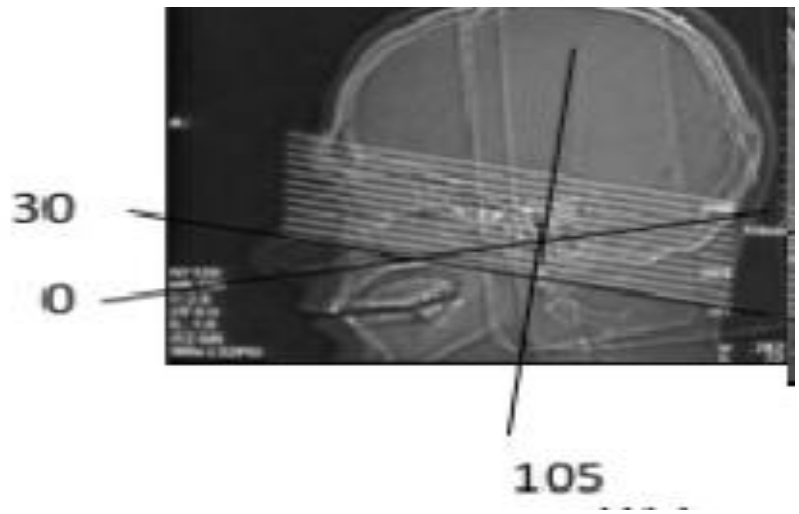


Figure 2: planes of CT Scanning (Axial &Coronal planes)

On a multidetector CT scanner, the raw axial image data set can be reconstructed with a section thickness of as little as 0.3 mm to obtain high-quality coronal reformatted images. A 512 × 512 matrix is used, and all the images are reviewed with a high-resolution bone algorithm and a small field of view (9 cm) for separate documentation of the right and left ears. Axial images are obtained from the top of the petrous apex to the inferior tip of the mastoid bone. Coronal reformatted images are obtained from the anterior margin of the petrous apex to the posterior margin of the mastoid. Coronal planes patient head extended in prone or supine with 105 Degree.

Magnetic Resonance Imaging:

The use of a 1.5- or 3-T MR imaging system is preferred for inner ear examinations, and sedation is used in most children. A thin-section gradient-echo sequence that is heavily T2 weighted is best suited for evaluation of the fluid-filled spaces of the membranous labyrinth and the eighth cranial nerve. A section thickness of as little as 0.4–0.7 mm is preferred for optimal delineation and to allow the generation of high-quality multiplanar reformatted images. A small field of view is used, and a volumetric acquisition is performed in the axial plane with sagittal and coronal reformatting. Oblique sagittal reformatted images are obtained in

planes perpendicular to the course of the seventh and eighth nerves in the internal auditory canal (IAC) and cerebellopontine angle. Routine axial T2-weighted imaging of the brain should be performed in all patients to exclude central nervous system causes of sensorineural hearing loss.

HRCT &MR Imaging should be used in the preoperative evaluation of candidates undergoing cochlear implantation. Imaging findings are crucial in the indication or contraindication for such surgical procedure.

Cochlear nerve aplasia, labyrinthine and cochlear aplasia's are still considered as absolute contraindications. Cochlear dysplasia, labyrinthitis ossificans are the relative contraindications. Complicating agents in the temporal bone assessment, namely, hypoplasia of the mastoid process, aberrant facial nerve, otomastoiditis, otosclerosis, dehiscent jugular bulb, enlarged endolymphatic duct and sac.

5.APPLIED ANATOMY AND RADIOLOGICAL ANATOMY OF EAR:

EXTERNAL EAR:

External ear consists of auricle, external auditory canal and tympanic membrane

External auditory meatus:

It is 2.5 cm in length, from the bottom of concha to tympanic membrane. It has two parts (a) cartilaginous and (b) bony parts. In young infants bony canal is not developed.

Tympanic membrane (Ear drum):

It separates the external auditory meatus from the middle ear. It is divided in to two parts, pars tensa, pars flaccida. Periphery of the pars tensa membrane is thickened to form a fibrocartilaginous part which is called tympanic annulus.

In HRCT Temporal bone facial recess is measured from distance between posterior tympanic annulus to vertical segment of facial nerve.

MIDDLE EAR:

This is an irregular shaped air-filled cavity within the petrous portion of the temporal bone. Anatomically and radiologically middle ear is divided into three parts (i) Epitympanum (ii) Mesotympanum (iii) Hypotympanum.

Epitympanum lies between tegmen tympani which form the roof and floor is defined by line between scutum and tympanic segment of facial nerve.

Mesotympanum is situated between the scutum and tympanic segment of facial nerve above and line between tympanic annulus &base of cochlear promontory below.

Hypotympanum is the shallow trough in floor of middle ear cavity.

TEGMEN:

Tegmen tympani and antri are the thin plate of bone, which separates the epitympanum and mastoid antrum from middle cranial fossa.

Medial part of the tegmen develops from the cartilaginous otic capsule. So their common anatomic location and embryological origin of the tegmen and superior semicircular canal are exposed to the same potential etiopathogenic factors .So development of spontaneous tegmen defect and superior semicircular canal defect occurs together.

Always look for SCC defect in case of spontaneous tegmen defect is present by HRCT temporal bone with multiplanar reconstructions.

JUGULAR BULB:

It is lies in the floor of the middle ear. Thin plate of bone separates the hypotympanum from the jugular bulb.

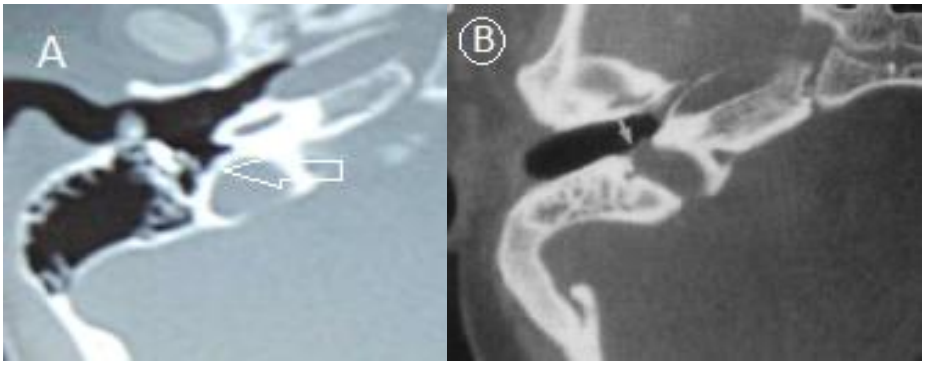


Figure 3(A, B).Normal jugular bulb (white arrow in A)
High-riding jugular bulb in the hypotympanum, Note the
Absence of bone over the jugular bulb. (white arrow in B)

Jugular bulb is a dynamic structure that forms after two years and stabilizes in adulthood. Radiologically children < 2 years did not demonstrate the typical bulbous enlargement of jugular bulb.

An erect posture causes an ascending negative pulse wave originating from the heart and traversing upwards to strike the jugular sinus at the jugular foramen of cranial base causing expansion of jugular bulb. This occurs once the infant assumes an erect posture. Relatively longer left branchio cephalic vein the energy of this venous pulsation generated from

the heart might disappear on left side, so larger jugular bulb and high riding jugular bulb more common on right side compared to left side.

Position of jugular bulb is important in cochlear implant surgery. HRCT temporal bone give information about jugular bulb.High-riding jugular bulb has been defined as a jugular bulb that extends above the inferior bony annulus or the superior margin of the jugular bulb extend above the floor of the ipsilateral internal auditory canal or above the level of the basal turn of the cochlea. It is reported to occur in approximately 6% of the general population. A high riding jugular bulb may prevent access to the round window niche in facial recess approach.

SIGMOID SINUS:

The position of the sigmoid sinus is highly variable, an anteriorly displaced sigmoid sinus is towards the external auditory canal form the less room for cortical mastoidectomy, may restrict the approach to the facial recess in cochlear implant surgery.

The sigmoid sinus forms a shallow indentation on the posterior aspect of the mastoid in the axial cuts of HRCT. If sinus courses lies more anteriorly and produces a deep groove in the mastoid, it is best seen in the axial sections of HRCT temporal bone.

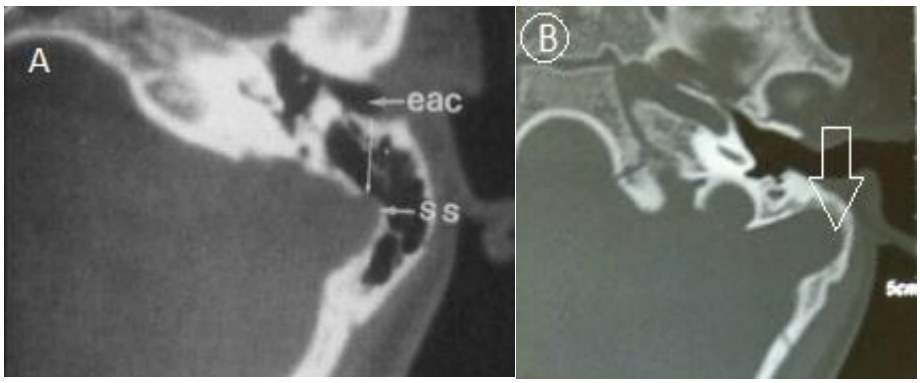


Figure 4; The position of the sigmoid sinus was calculated by measuring from the posterior bony external auditory canal to the most anterior extension of the sigmoid sinus (white arrow in A); Anteriorly displaced sigmoid sinus(white arrow in B)

Radiologically the position of the sigmoid sinus was calculated by measuring the distance from the posterior bony external auditory canal to the most anterior extension of the sigmoid sinus. This measurement was taken at the level where one could identify the basal turn of the cochlea, sigmoid sinus, and bony EAC in the same cut. Distance between the sinus and bony external auditory canal < 10 mm , considered as an anteriorly displaced sigmoid sinus .

PROMONTORY:

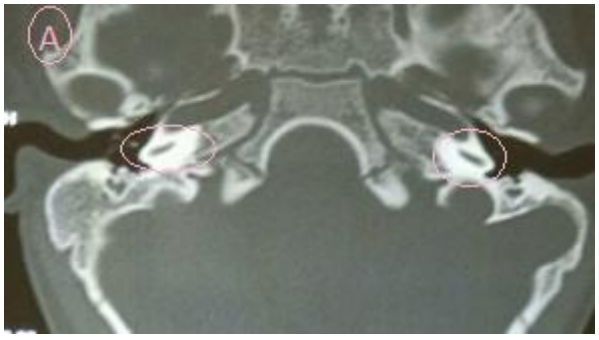


Figure 5: HRCT Axial image shows promontry

with basal turn of cochlea(Pink arrow)

It is a smooth rounded bony projections covering the basal turn of cochlea. Least covered portion of the cochlea lies behind the apex of the promontory. Lower half of the basal turn of the cochlea can be approached from that facial recess during cochlear implantation.

OVAL WINDOW:

It lies above and behind the promontory, closed by footplate of stapes and annular ligament. It opens in to vestibule of inner ear.

ROUND WINDOW:

It is a 2–3mm long and about 1.5mm wide channel connecting the middle and inner ears. It is present behind and below the oval window which is covered by secondary tympanic membrane.

Embryology:

At 8^{th} to 15th week of gestation the whole round window is formed by the portion of cartilage of the otic capsule. Around 16^{th} week of fetal life, ossification centres of the otic capsule start to establish the bony round window. Major changes are completed before birth, since the round window has the same form, size and feature at birth as in adults. Anterior, superior and posterior walls are first to appear while the inferior wall is completely absent at this time. In the middle of the inferior wall a stick-shaped bony elevation, the fustis, indicates

the former cartilage. A process of the otic capsule, called the cartilage bar, this forms the inferior wall of the round window niche. The anterior and superior walls of the niche formed by intramembranous ossification, whereas the posterior and inferior walls predominantly formed by enchondral ossification.

Uneven growth of different walls of the round window niche can alter the shape of the entrance, which results in eight different types of niches namely; extremely narrow, descending tegmen, anterior septum, bony membrane, open fundus, exostosis, jugular dome and trabeculae.

Extremely narrow :

When all the walls are thick, the niche becomes very narrow. Its surrounded by compact bony surface without a fustis .

Descending tegmen :

The tegmen of the niche stretches backward, results in lengthening of niche. It may stretch to varying degrees which results in a V-shaped transformation of the niche's form.

Anterior septum:

This is most frequent variation of the round window niche. Entrance of the niche is formed by lamellar growth of the postis anterior which covers the majority of the opening of the round window. The shape of the postis anterior shows a wide variability from a continuous plate to the trabecular form.

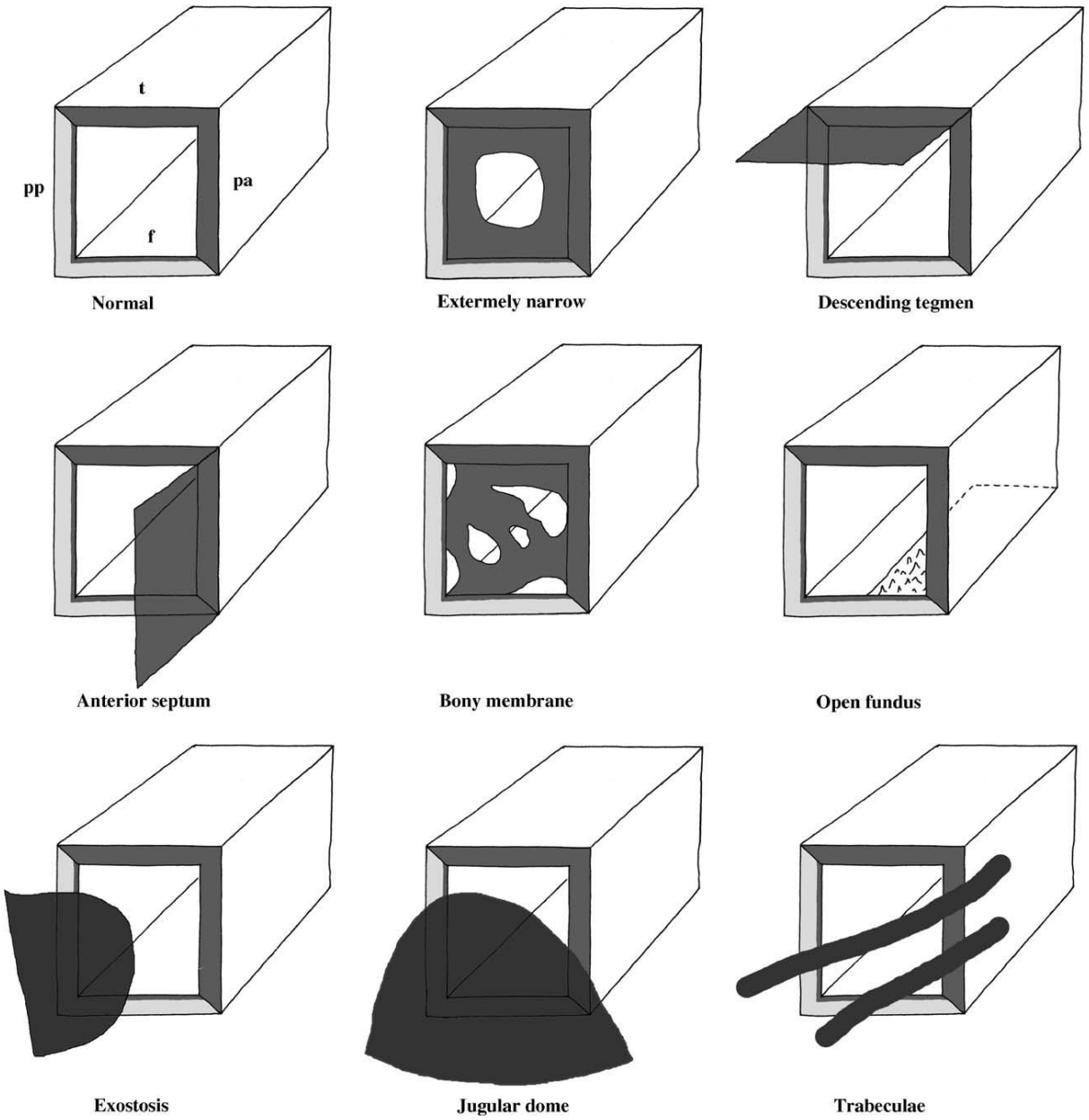


Figure 6; Various types of round window.

Bony membrane:

It is rarest variation .The pseudo membrane is partially or completely ossified in the round window's entrance and forms a thin bony plate

Open fundus:

Lack of ossification lateral to the fustis results in the absence of bony tissue between the fustis and the anterior wall which occurs when the inferior wall remains incomplete .

Exostosis :

Structures outside the niche and surrounding the round window can modify the form of exostosis near the entrance of the niche.

Jugular dome:

Less frequent but clinically very important variation, the jugular dome can partially or even fully hide the round window niche.

Trabecules:

The niche is protected by trabecules of varying sizes.

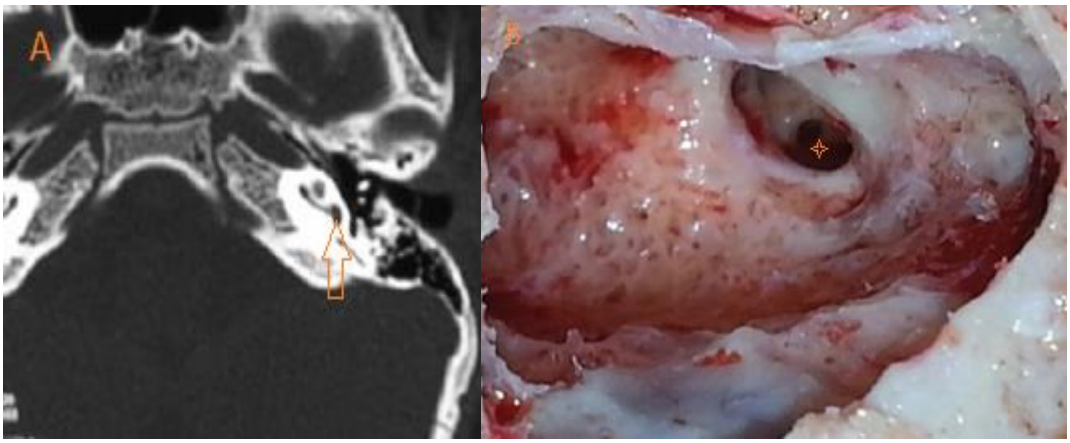


Figure 7(A&B); A-HRCT Axial image shows round window niche with bony overhang (Orange arrow): B – After posterior tympanotomy round window (Orange star)

During cochlear implantation the electrode array inserted either through the round window or cochleostomy into scala tympani. Round window

insertion, reduces insertion trauma and could potentially preserve residual hearing. Hence more surgeons are using the round window approach for Cochlear implantation.

FACIAL RECESS:

It is a surgically defined space rather than an anatomic space, is the route used in posterior tympanotomy to perform cochlear implant surgery. Facial recess bounded superiorly by the fossa incudis, medially by the descending segment of facial nerve, laterally by the chorda tympani nerve, inferiorly chorda-facial angle. Extended facial recess is defined as space between facial nerve and posterior tympanic annulus, in which chorda tympani nerve is sacrificed.

Development:

Between the developing facial canal and the tympanic ring is the cartilage bar of the second branchial arch, known as Reichert's cartilage. This cartilage attaches to the otic capsule and descends, lies between the facial nerve at the level of the stapedius muscle and the tympanic annulus. This position places it directly in the area that will create the facial recess.

Facial recess is completely developed at birth and appears to be adult sized at term. In infants, the facial recess reaches **3.25** mm at the oval window and **2.62**mm at the round window. The extended facial recess reaches **3.79** mm at the oval window and **3.04** mm at the round window.

The facial recess was measured from the epineurium of the facial nerve to the chorda tympani nerve. The extended facial recess measurement was taken from the epineurium of the facial nerve to the sulcus of the posterior annulus.

HRCT temporal bone demonstrate the facial recess,even with thin slice temporal bone CT visualisation of chorda tympani nerve is not always possible, so it is measured in HRCT as the distance between the facial nerve and the posterior tympanic annulus. It is measured in the axial plane at the level where the round window and the basal turn of the cochlea are seen. If the distance is >3mms labelled as normal and wide facial recess .Distance less than 3 mms, it is labelled as narrow facial recess.

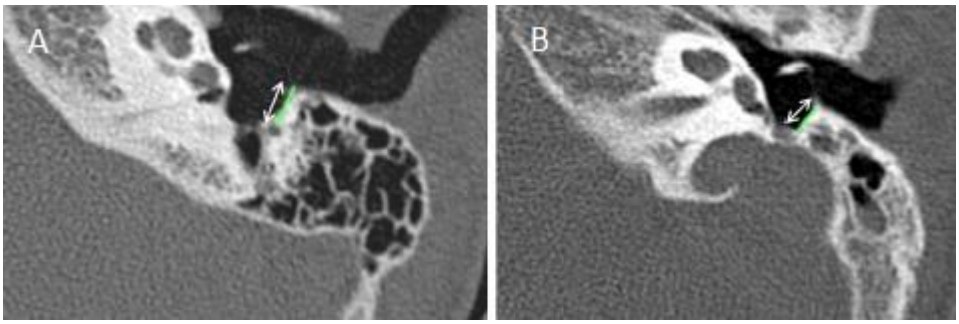


Figure 8(A&B): HRCT Axial image A-Wide facial recess B-Narrow facial recess

Facial recess typically provides adequate exposure to the round window niche. Cochlear implantation surgery is difficult to perform in children with a narrow facial recess.

MASTOID PNEUMATISATION:

The antrum is the first mastoid cell which can be recognized at 21–22 weeks of gestation and fully organized at 34 weeks . At birth, usually no additional air cells and only mesenchymal tissue may fill the middle ear cleft. This mesenchymal tissue is quickly cleared, by absorption and redistribution, during growth and expansion of the mastoid air cells.

Three phase of mastoid pneumatization from birth to until reaching adult size. The infantile stage of the mastoid air cells is rapid growth upto one year followed by a linear growth till the age of six years, then slow incremental growth to adult size at puberty.

If the child has well pneumatized mastoid cavity and the approach to the facial recess is easily carried out during CI surgery. The HRCT temporal bone image provides the ability to assess mastoid pneumatization and size of cavity before implantation, which is particularly important in younger children or children with a history of chronic ear disease.

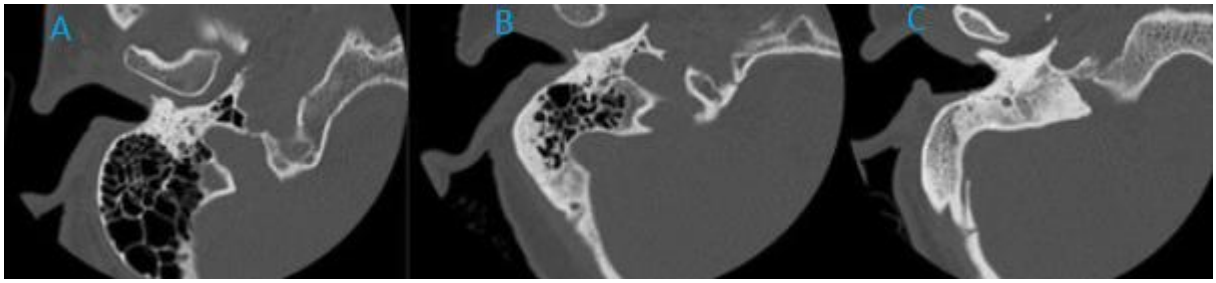


Figure9: Axial images of HRCT scan showing mastoid that is (**a**) well aerated, (**b**) moderately aerated, and (**c**) poorly aerated

Mastoid pneumatisation are graded into three based on the degree of pneumatization in relation to the sigmoid sinus. Grade 1 well pneumatised mastoid cavity with pneumatization that extended posterior to the sigmoid sinus. Grade 2 pneumatization was a moderately pneumatized mastoid cavity with pneumatisation that extended up to but not beyond the sigmoid sinus. Grade **3** was a mastoid cavity that was sclerotic with very poor pneumatization.

FACIAL NERVE:

It is the nerve of the second branchial arch. It develops from the collection of neural crest cells called as the fascioacoustic primordium around the third week of gestation and is in close proximity to the otic placode. In the new born facial nerve anatomy approximates that of an adult, except for its location in the mastoid, which is more superficial.

The facial nerve arises from the motor nucleus which resides in the lower pons below the fourth ventricle. The facial nerve course divided into intracranial, intratemporal and extratemporal portions.

The intracranial portion of the facial nerve is approximately 24 mm, which runs from the brain stem to the porus of the internal auditory meatus. Then it is runs in the anterosuperior segment of the internal auditory meatus (lAM) from porus to fundus in a distance of 5- 12 mm.

The intratemporal portion of the facial nerve runs from the entrance of the fallopian canal at the fundus of the lAM to the stylomastoid foramen. The length of this portion is 28-30 mm and is divided into labyrinthine, tympanic and mastoid segments.

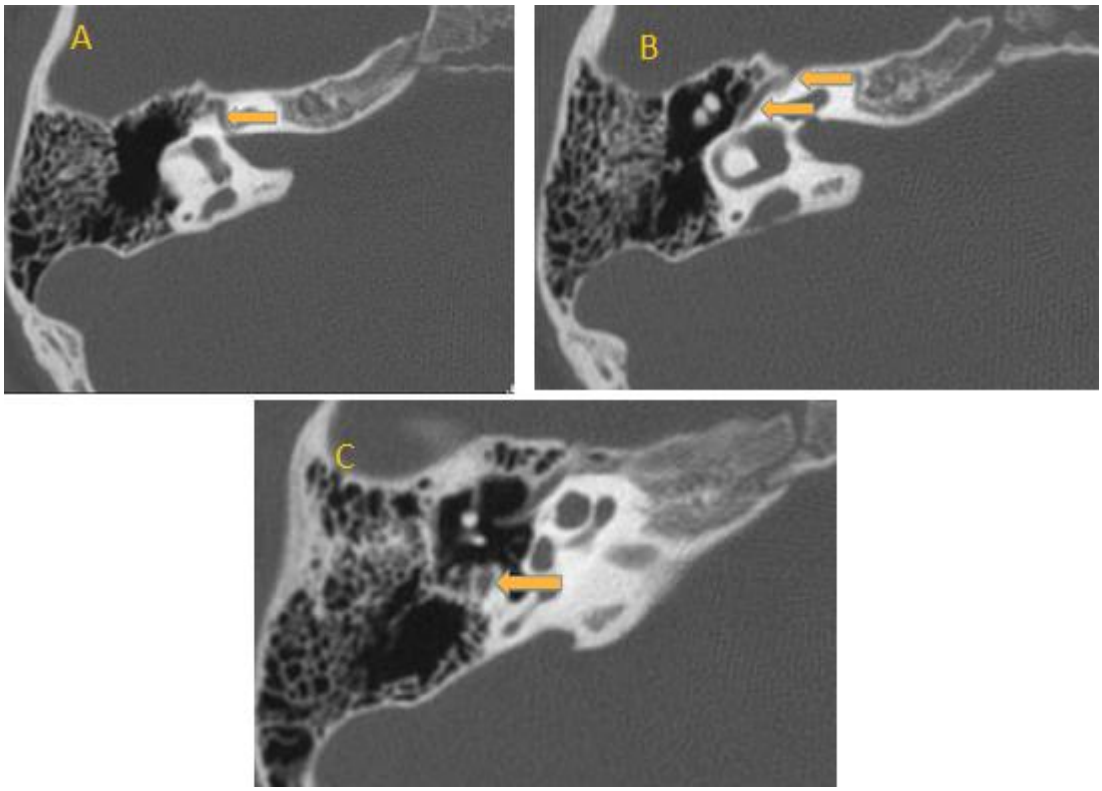


Figure 10: HRCT Axial image showing varies segment of facial nerve (Yellow arrow) A-Labyrinthine segment&1st genu; B-Tympanic segment; C-Mastoid segment

The labyrinthine segment is the shortest (3-5 mm) and thinnest part of the facial nerve within the Fallopian canal. At the distal end, the geniculate ganglion forms part of a sharp turn, the first

genu of the facial nerve.

The tympanic segment of the facial nerve (8- 1 1 mm) is from the geniculate ganglion to second genu which is runs above and medial to the cochleariform process and lies beneath the lateral semicircular canal and
above the oval window.

The mastoid segment (13–15 mm)extends from the second genu to the stylomastoid foramen. The nerve lies in vertical position, runs downward in the posterior wall of the tympanic cavity and the anterior wall of the mastoid to exit at the base of the skull the stylomastoid foramen.

The chorda tympani nerve has a variable take off and can come off anywhere from the mastoid segment of the facial nerve, but usually 6 mm above the stylomastoid foramen. It runs superiorly, along the facial nerve, but angles anteriorly where it enters the mesotympanum through the iter chordae posterius.

Developmental facial canal dehiscence is most common at the tympanic segment compare to mastoid segment, making the nerve vulnerable to injury during middle ear surgery. The most common site of injury during cochlear implantation is the mastoid segment of facial nerve.

Landmarks for identifying facial nerve (1) the second genu hugs the inferior aspect of the lateral semicircular canal and this is extremely constant (2) The pyramidal eminence, The nerve is lateral and posterior to the pyramidal process

(3) The nerve located surgically by the interval between the short process of the incus laterally and the lower border of the lateral semicircular canal medially.
(4) Digastric ridge, which lies medial aspect of the mastoid tip, which is most important land mark for mastoid segment of facial nerve.

During cochlear implantation (CI), the surgeon operating very close to the mastoid portion of the facial nerve (MPFN) while creating the posterior tympanotomy (PT). CI is a safe procedure, the estimated rate of facial nerve paralysis of 0.3%. We are routinely using the round window membrane (RWM) cochleostomy approach for electrode insertion because it enables the surgeon to see the scala tympani directly and is a well-established method of electrode insertion.

Round window membrane cochleostomy difficult to perform unless the surgeon is able to see enough of the RWM, which is lies more posterior than in a conventional cochleostomy. For better surgical access to the round window niche requires a ‘perfect’ PT with an intact tympanic annulus and posterior canal wall, with maximum thinning of the bone overlying the chorda tympani and the MPFN.

It is imperative to evaluate the position of the facial nerve on HRCT temporal bone. Abnormalities of the facial nerve as it courses through the temporal bone with or without congenital malformation of inner ear are best evaluated with

HRCT, which allows assessment of the size and morphology of the facial canal. Aberrant course of the facial nerve can be quite unusual and at increased risk of injury during implantation surgery.

The dehiscent facial nerve canal most commonly along the tympanic segment, this place of facial nerve may be greater risk during surgery .If the nerve may protrude through the defect and may be mistaken for a soft tissue mass on HRCT.HRCT is the best modality for evaluating for dehiscence.

The facial nerve may have an abnormal course through the temporal bone. Because the second branchial arch, which gives rise to the facial nerve, and the first pharyngeal pouch, which gives rise to the EAC, both are developing simultaneously in utero, an abnormal course of the facial nerve is commonly seen with congenital aural atresia and microtia children.

The mastoid segment of the facial nerve may be anterolaterally displaced, with a slightly more horizontal course as it descends.

For safe and successful implantation careful preoperative mapping of the course of the facial nerve canal is necessary. Careful review of the position of the facial nerve is also warranted in children without inner ear malformations, as there may be dehiscence of the tympanic portion that may be encountered during the approach to the cochleostomy site.

INNER EAR:

It is lies in the petrous portion of temporal bone. It consists of two parts (1)osseous periotic labyrinth (2) membranous otic labyrinth. Osseous labyrinth has 3 main parts ; vestibule ,three semicircular canal and cochlea .

Embryology:

Inner ear development begins early in embryogenesis. Approximately third week of gestation, an either side of the rhombencephalon otic placode arise from the surface ectoderm. This otic placodes subsequently invaginate and form the otic pit, which then gets detached from the surface to form the otic and auditory vesicles. At around the fifth week, diverticulum buds from the otic pit form the endolymphatic sacs, followed by the cochlea and vestibule. The membranous cochlea achieves 1 to 1.5 turns at the end of 6 weeks, and 2.5 turns are formed at the end of the 7th week. The semicircular canals start to develop from the utricle segments of the otocysts at 7–8 gestational weeks. The superior canals form first, followed by the posterior and then the lateral canals.

The inner ear structures have an adult configuration by the end of 8 weeks. By the end of the eighth week, the membranous labyrinth has developed its characteristic convoluted shape. Ossification of the otic capsule develops gradually around the membranous labyrinth and it is completed by birth. Late second and early third trimesters maturation of

the sensory epithelium occurs long after formation of the membranous labyrinth. At 26 to 28 weeks of gestation, hair cell and auditory neural development are completed. Normal human fetus may be able to hear 2.5 to 3 months before birth.

CONGENITAL INNER EAR MALFORMATIONS:

Malformations limited to the membranous labyrinth are at cellular level which account for more than 80% of cases of congenital deafness. Since bony labyrinth is normal, there is no radiological abnormality.

Malformations of the membranous and bony labyrinth, account approximately 20% of congenitally deaf children, demonstrate radiologically anomalous inner ears.

Classification:

(1) Malformations Limited to the Membranous Labyrinth

- Complete membranous labyrinthine dysplasia(Siebenmann-bing)
- Limited membranous labyrinthine dysplasia
    - Cochleosaccular dysplasia (Scheibe)
    - Cochlear basal turn dysplasia(Alexander)

(2) Malformations of the Osseous and Membranous Labyrinth

- Complete labyrinthine aplasia (Michel)
- Cochlear anomalies
  - Cochlear aplasia
  - Cochlear hypoplasia
  - Incomplete partition
  - Common cavity
- Labyrinthine anomalies
  - Semicircular canal dysplasia
  - Semicircular canal aplasia
- Aqueductal anomalies
  - Enlargement of the vestibular aqueduct
  - Enlargement of the cochlear aqueduct
- Internal auditory canal anomalies
  - Narrow internal auditory canal
  - Wide internal auditory canal
- Eighth nerve anomalies
  - Hypoplasia
  - Aplasia

COMPLETE MEMBRANOUS LABYRINTHINE DYSPLASIA:

It is very rare, described first by Siebenmann and Bing. It has been reported in literatures usually in association with cardioauditory (Jervell and Lange-Nielsen) and Usher syndromes.

Cochleosaccular Dysplasia (Scheibe's Dysplasia)

In this type the Dysplasia is limited to the cochlea and the saccule, Here the SCCs and utricle appear normal. There is absence or partial development of organ of Corti, cochlear duct may be collapsed or there may be an endolymphatic hydrops with Reissner's membrane adherent to the limbus. Cochlear changes may be severe in the base turn and gradually lessen in intensity toward the apex, or they may be severe throughout. The saccule usually is collapsed and has degenerated sensory epithelium. Auditory neuronal survival is variable.

Cochlear Basal Turn Dysplasia

Here the dysplasia is usually limited to the basal turn of the cochlea. It is related to familial high-frequency SNHL. The affected persons are usually asymptomatic or have normal hearing.

MALFORMATIONS OF THE MEMBRANOUS AND OSSEOUS LABYRINTH:

Inner ear anomalies that deform the otic capsule are recognized and differentiated during life through radiographic imaging. Approximately 20% of congenitally deaf children demonstrate radiographically anomalous inner ears.

Malformations appears to arise from a premature arrest in the development of one or more components of the inner ear . It occurs during embryogenesis, particularly between the fourth to eighth weeks of gestation. As a general rule, the earlier the developmental arrest, the more severe the deformity and the worse the hearing.

Whereas some malformations of inner ear involve only one portion of the inner ear or more than one component results in combination of anomalies. Between the fourth and fifth weeks of development, the sphericotocyst develops the three buds that ultimately form the cochlea, SCCs, and VA. An inner ear malformation may be limited to one of these anlages, may involve a combination of two, or may even affect all three.

The frequent coexistence of anomalies involving all three cochlea, SCCs, and VA has several possible explanations: (1) the anomaly is genetically predetermined (2) an insult to the embryo occurred before the fifth week (3)

each of the buds was susceptible to some teratogenic influence at a later stage of development.

Majority of malformations of inner ear are bilateral and symmetrical. In children who have radiological detectable anomaly on only one side, the opposite “normal” inner ear has a hearing loss in approximately 50% of children.

Complete labyrinthine aplasia (Michel's aplasia)

First described by Michel, it is a very rare and most severe deformity of the membranous and osseous labyrinth. Before the formation of an otic vesicle there occurs a developmental arrest resulting in a complete absence of inner ear structures. Michel’s aplasia has been reported in association with anencephaly and thalidomide exposure. It has been associated with external ear abnormalities. The incidence of Michel’s aplasia is overestimated in the radiographic imaging because it is confused with labyrinthine ossification. In the latter condition, which usually is acquired during life, a sizable and dense otic capsule is evident radiographically. In michel’s aplasia, the otic capsule is entirely absent , Such ears are, of course, uniformly deaf.

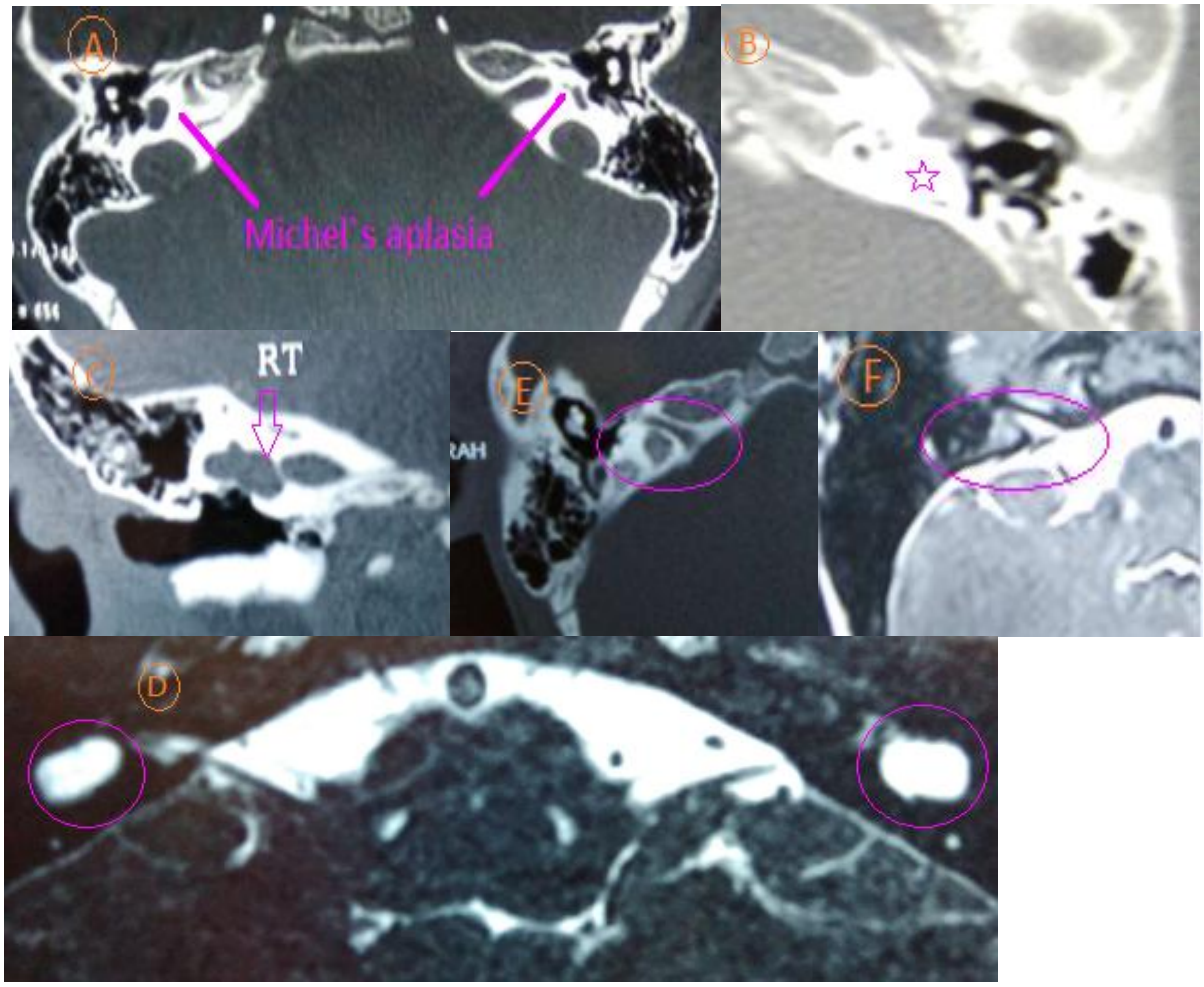
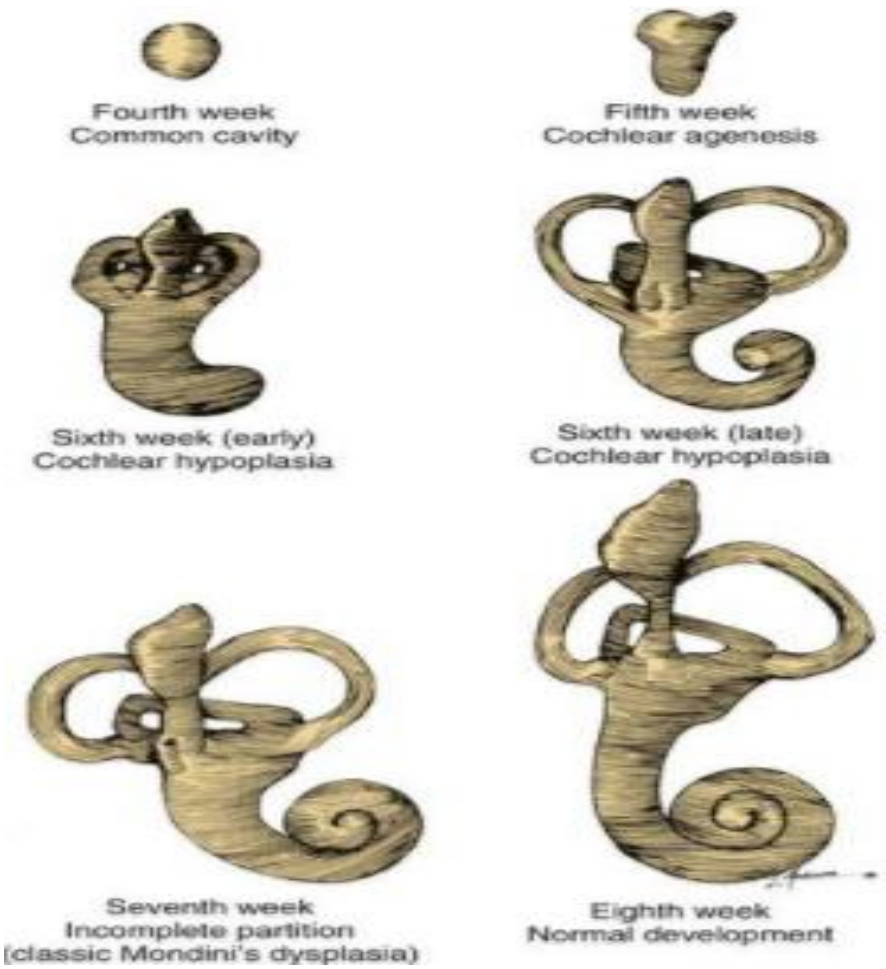


Figure 11 (A-F): Michel deformity. HRCT-Axial image shows bilateral Michel`s aplasia Right side replaced by cystic structure (Fig A&B): HRCT coronal image a cystic structure is seen replacing the inner ear (Fig C): HRCT axial images (E) shows a narrow Right internal auditory canal with a Michel deformity. Axial 3D-FIESTA images (D) show absence of the entire vestibulo-cochlear structures bilaterally: Axial 3D-FIESTA images (F) the internal auditory canals are small on right sides

COCHLEAR ANOMALIES:



Cochlear Aplasia:

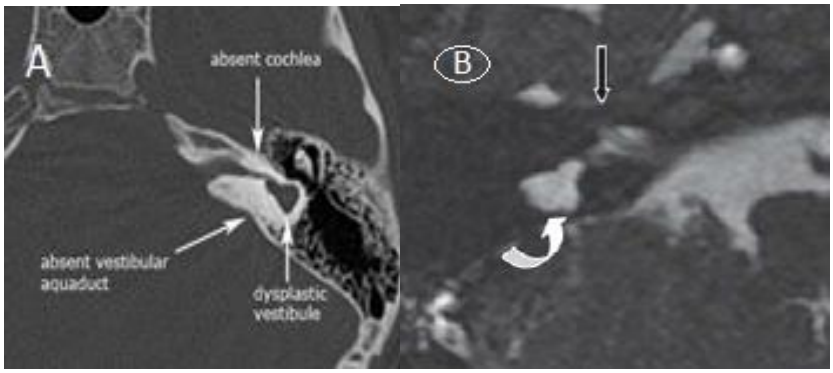


Figure12 HRCT axial image & axial 3D-FIESTA images shows
Absent cochlea, dysplastic vestibule

In this type cochlea is completely absent, it occur as a result of arrest in the development of the cochlear bud at the fifth week of gestatation.

Radiographically, only a vestibule and SCCs are either normal, dilated or hypoplastic. To differentiate cochlear aplasia from labyrinthine ossification, it is necessary to assess the amount of otic capsule bone anterior to the internal auditory canal (IAC). In cochlear aplasia, the otic capsule is absent, whereas in labyrinthine ossification, it is dense and of normal dimensions. Ears with cochlear aplasia are devoid of auditory function.

Cochlear Hypoplasia

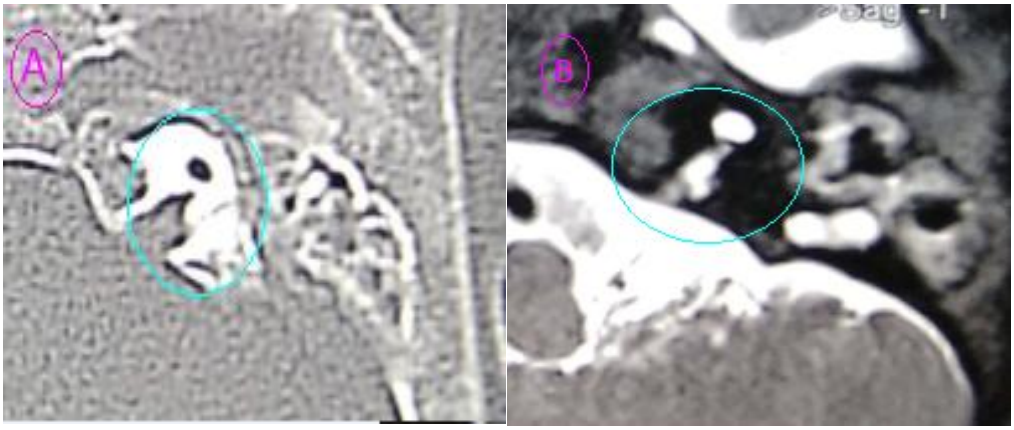


Figure 13 HRCT axial image (A) & axial 3D-FIESTA images (B) shows cochlear hypoplasia(Blue round)

An arrest during the sixth week of gestation results in a hypoplastic cochlea consisting of a single turn or less. This account approximately 15% of all cochlear anomalies. Radiographically, a small bud of variable length (usually 1 to 3 mm) protrudes from the vestibule . The vestibule frequently is enlarged, with semicircular malformations in approximately one half of the cases. Histologically small cochlea lacking a modiolus or other internal architecture. Hearing is variable and may be remarkably good in view of the minute size of

the cochlea. The variability of hearing is accounted for by the degree of membranous labyrinthine development within the truncated cochlear lumen.

According to radiologic literature, as well as the author's own radiological (Sennaroglu et al) data, three different types of cochlear hypoplasia can be identified.

Type I (bud-like cochlea): The cochlea is like a small bud arising from the IAC Internal architecture is severely deformed. no modiolus or interscalar septa can be identified.

Type II (cystic hypoplastic cochlea): The cochlea is smaller in its dimensions with no modiolus and interscalar septa, but its external architecture is normal There is a wide connection with the IAC. The vestibular aqueduct is enlarged and the vestibule is minimally dilated .This is the type of hypoplasia where a gusher and unintentional entry of the electrode into IAC are possible.

Type III (cochlea with less than two turns): The cochlea has a shorter modiolus and the overall length of the interscalar septa is less, resulting in a smaller number of turns (less than two turns). The internal and external architecture (modiolus, interscalar septa) is similar to that of a normal cochlea, but the dimensions are less and hence the number of turns is less .The vestibule and the SCCs are hypoplastic.

INCOMPLETE PARTITION TYPES:

An arrest at the seventh week of gestation results in incomplete partition types. Sennaroglu and Saatci have subtyped the incomplete partition deformity into three variants

Type I: (Cystic Cochleovestibular malformation:

It lacks the entire modiolus and interscalar septa and demonstrates a cystic appearance; it has been associated with dilated vestibule. Cochlear implant indicated if cochlear nerve is present.

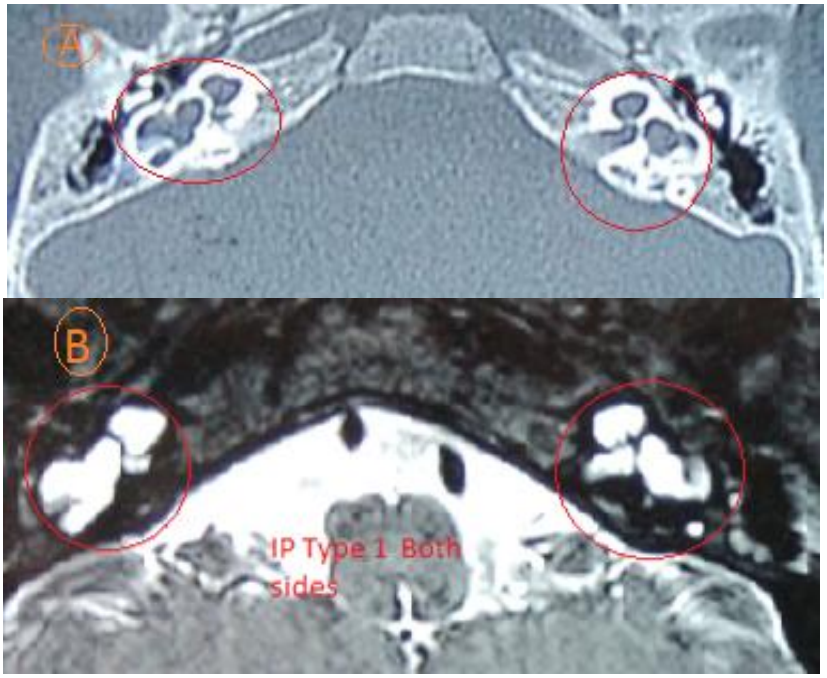


Figure 14: HRCT axial image (A) & Axial 3D-FIESTA images (B) shows IP-I Both sides(Red arrow)

Type II (Mondini type):

In this type cochlea consists of 1.5 turns, and the interscalar septum ,osseous spiral lamina are absent. The basal cochlear turn appears normal, but the middle and apical turns coalesce to form a cystic apex. The modiolus is present only at the level of the basal turn. This is the most common type of cochlear malformation, accounting for more than 50% of all deformities.

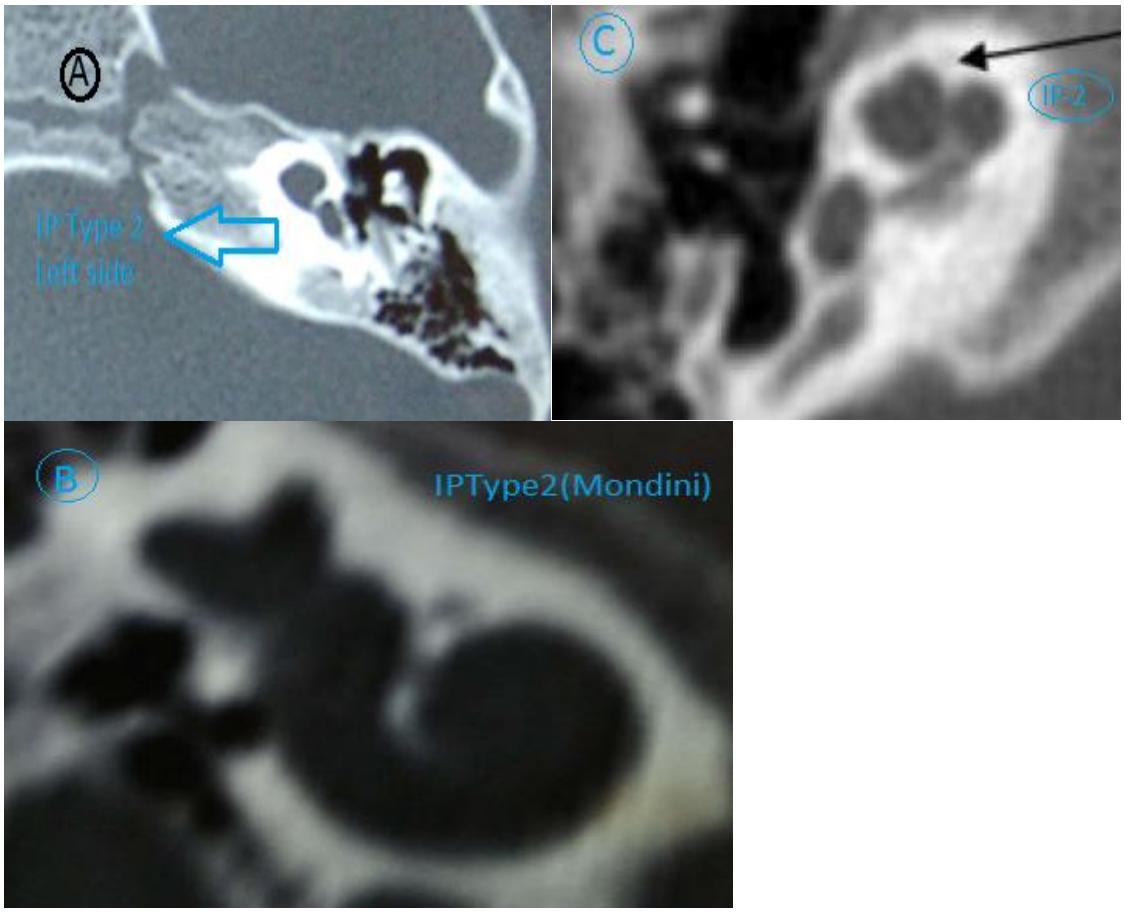


Figure 15 (A-C): Incomplete partition type II. Axial HRCT images (A, C) show fusion of the middle and apical turns of the cochlea (arrow in A&C)

Radiographically, the cochlea is smaller than normal and partially or completely lacks an interscalar septum. Care must be taken in counting the number of

cochlear turns radiographically because this may be difficult to determine even using high-resolution CT. The radiographic diagnosis depends more on cochlear size and the absence of a scalar septum than on the number of cochlear turns perceived.

Type III variant(X-linked deafness):

In this type modiolus is deficient and partial interscalar septation at the cochlea's periphery. It accounts 2% of cochlear malformations. Organ of corti development is variable, auditory function also is variable, ranging from normal to profound SNHL.

COMMON CAVITY

An arrest at the fourth week of gestation (otocyst stage) results in common cavity ear in which the cochlea and vestibule are confluent, forming an ovoid cystic space without internal architecture. Alternatively, this deformity may result from aberrant development at a later stage. Radiographically an empty ovoid structure typically longer in its horizontal dimension, the size of the cyst may vary, it averages 7 mm vertically and 10 mm horizontally

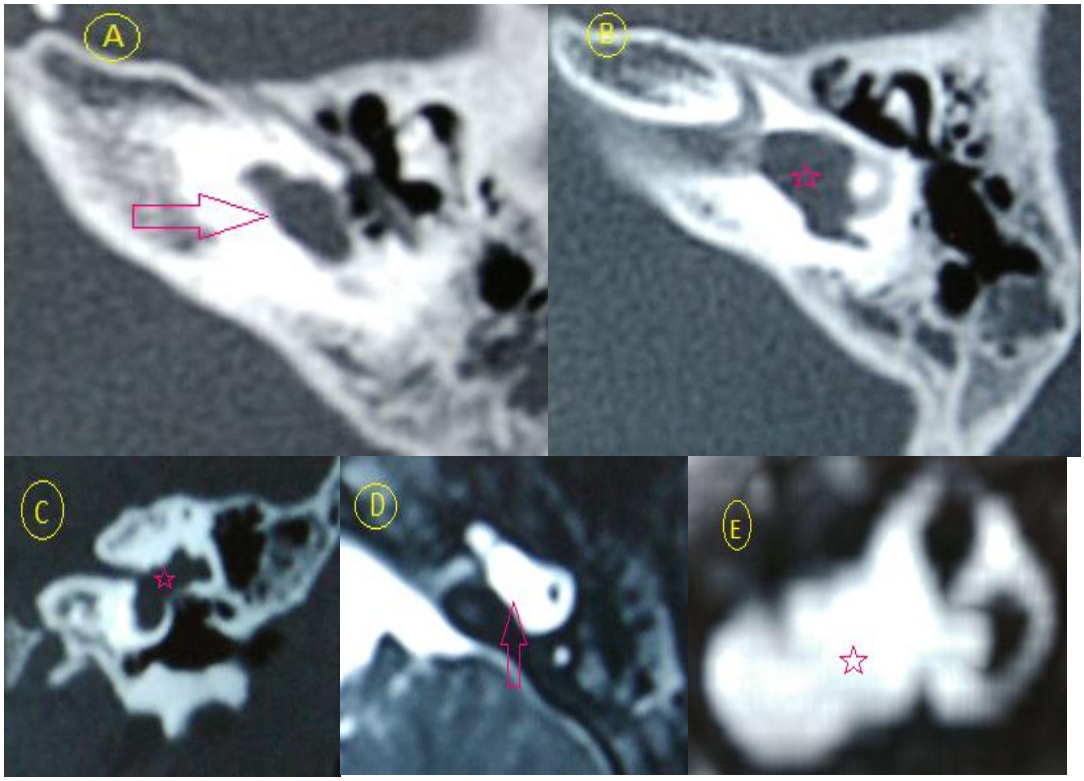
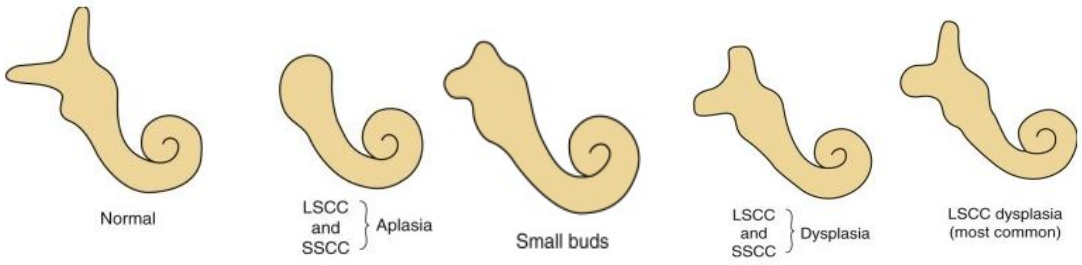


Figure 16 HRCT axial (A&B) images, coronal (C) image shows common cavity: Fig D Axial 3D-FIESTA images shows common cavity with IAM; Fig e3D-MIP reconstruction of the images showing common cavity

Radiographically it is quite easy to misdiagnose a dysplastic lateral SCC as a common cavity deformity. The key to differentiating between the two is that a common cavity cochlea lies predominantly anterior to the IAC on axial-plane CT, and a dysplastic vestibular system lies posterior to it.

Labyrinthine Anomalies:



Semicircular Canal Dysplasia

During the sixth week of embryological development, the budding SCC normally forms a semicircular evagination from the vestibular anlage. The central portion of the pocket-shaped protrusion adheres, leaving a peripheral semicircular tube. When this central adhesion fails to occur, SCC dysplasia results . Dysplasia of the lateral SCC is a common type of labyrinthine anomalies than the posterior or superior SCC, apparently because it forms earlier in embryogenesis. Approximately 40% malformed cochlea will have an accompanying lateral SCC dysplasia, Occasionally, It exists as the sole inner ear malformation. The typical radiographic appearance of SCC dysplasia is that of a short, broad cystic space confluent with the vestibule.

Semicircular Canal Aplasia

SCC aplasia arises from a failure in the development of the vestibular anlage before the sixth week of gestation. SCC aplasia is only one-fourth as common as SCC dysplasia.

MALFORMATIONS OF THE VESTIBULAR AND COCHLEAR AQUEDUCTS:

Enlargement of the Vestibular Aqueduct

The VA developed from a diverticulum formed in the wall of the otocyst during the fifth week of gestation. It begins as a short, broad pouch but

gradually elongates and form characteristic J shape of adult. A premature arrest in development results a VA that is abnormally short and broad.

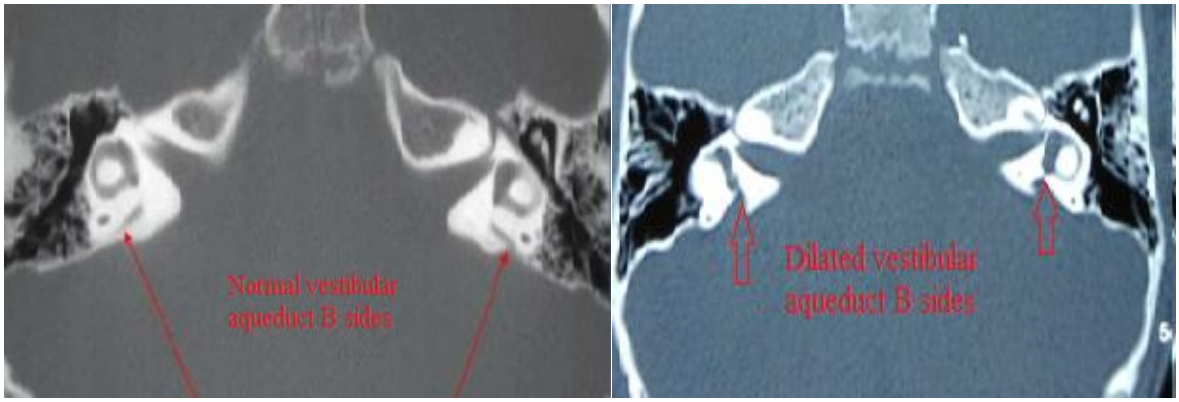


Figure 17 HRCT Axial image shows normal &dilated vestibular aqueduct

Enlargement of the VA is the most common radiographically detectable inner ear malformation. The advent of high-resolution CT in the axial plane has made assessment of the VA much easier. The diameter of a normal VA, when measured halfway between the common crus and its external aperture, is between 0.4 and 1 mm. Enlargement of the VA is diagnosed when its diameter exceeds 2 mm, although enlarged VAs may exceed 6 mm in width. Whereas the VA is well visualized on axial CT, the dilated endolymphatic sac is better seen with T2-weighted magnetic resonance imaging (MRI). VA enlargement most often accompanies malformation of the cochlea or SCC. It also may be the sole radiographically detectable abnormality of the inner ear in a child with hearing loss.

Large VA stems from an abnormal communication between the subarachnoid space and the fluid chambers of the inner ear. Serial MRI images show variability in both the size and signal characteristics of the enlarged sac. Presence of cerebrospinal fluid (CSF) under pressure, with consequent "gusher," within the inner ear has been observed in ears with large VAs during cochlear implantation. This anomaly typically consists of a defect of the cochlear modiolus at the distal end of the IAC. The large VA syndrome typically is bilateral. Affected children usually are born with normal or mildly impaired hearing that gradually deteriorates through childhood into adolescence and early adulthood. Cochlear implantation has been quite successful in children with large VA.

Enlargement of the Cochlear Aqueduct

Enlargement of the CA are misinterpretations of the wide internal funnel that opens into the posterior fossa. In healthy people the radiographic diameter of this aperture averages 3 to 4 mm but ranges from radiographically invisible to more than 10 mm. For enlargement of the CA to be diagnosed radiographically, the intraosseous portion coursing toward the vestibule must be enlarged beyond 1 mm, the practical resolution limit of contemporary CT scanners. If criteria analogous to those for enlargement of the VA are used, then an enlarged CA must have a diameter exceeding 2 mm throughout its course

between the inner ear and the posterior fossa.CA less than 1 mm in diameter is undetectable radiographically.

DEVELOPMENTAL ANOMALIES OF THE INTERNAL AUDITORY CANAL :

Wide Internal Auditory Canal

A congenitally large canal may be an incidental finding in healthy individuals. When a large IAC (larger than 10 mm in diameter) accompanies a malformation of the inner ear.

Narrow Internal Auditory Canal

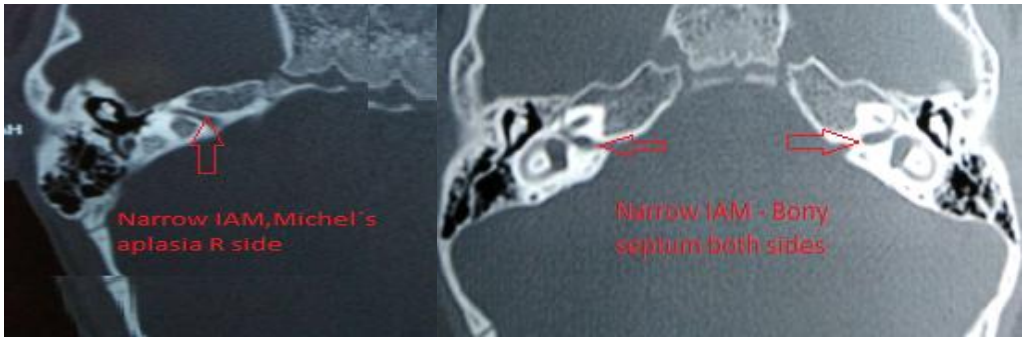


Figure 18 HRCT Axial image shows narrow IAM

A narrow IAC may indicate a failure of eighth cranial nerve development. When a child has normal facial function and an IAC less than 3 mm in diameter, it is likely that the bony canal transmits only the facial nerve . A narrow IAC may accompany inner ear malformations or may be the sole radiographically detectable anomaly in a deaf child.

A narrow IAC has been considered a relative contraindication to cochlear implantation, because it suggests that the eighth nerve may be insufficiently developed to conduct an auditory signal. After cochlear implantation, some patients with narrow IACs have experienced facial pain and twitching without useful auditory sensation.

ANOMALIES OF THE EIGHTH NERVE

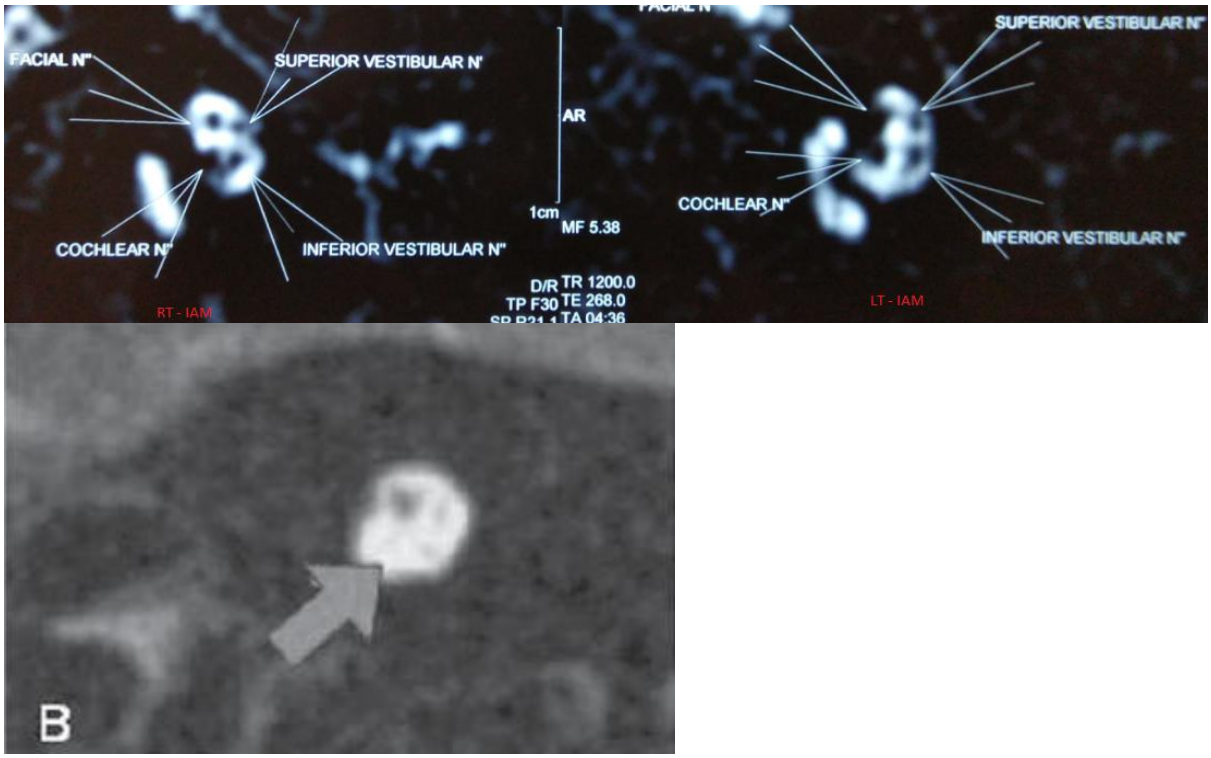


Figure 19 Sagittal MRI T2-weighted images. A: Anatomical distribution of the nerves inside the internal auditory canal. B: Absence of the cochlear nerve (arrow).

Hypoplasia and aplasia of the eighth nerve are often, but not always associated with congenital narrowness or even absence of the IAC. Similarly, although eighth nerve maldevelopment frequently accompanies malformation of the inner ear, the presence of a normal cochlea and semicircular canals does not guarantee normal development of the audiovestibular nerve. High-resolution, thin-section

MRI with T2-weighted sequences currently is the best means of assessing the fine anatomy of the eighth nerve in the IAC. MRI is warranted before cochlear implantation when the bony IAC is narrow on the CT scan, with severe types of inner ear malformation, and in syndromes known to be associated with maldevelopment of the eighth nerve (e.g., the CHARGE association, Mobius's syndrome). Unilateral cochlear nerve aplasia, sometimes familial, is increasingly recognized as an important cause of congenital unilateral profound SNHL. The internal auditory canal frequently is normal. Increasingly refined MR techniques may reveal hypoplasia of the auditory nerve as more common than previously recognized.

ANATOMY OF INNER EAR

The inner ear is present in the petrous apex of the temporal bone, and is situated in a bony structure called the osseous or bony labyrinth. The labyrinth consists of three continuous sections: the vestibule, the cochlea, and the semicircular canals.

Cochlear Anatomy:

At the basal end of the cochlea is the round window membrane, which communicates with the middle ear space. The cochlea is snail-shaped and has a wide diameter at the base, which further narrows for two and three-fourth turns till it reaches its apex.

The core of the cochlea is the modiolus, which is highly porous bone, and allows passage of auditory nerve fibers as they travel from the internal auditory meatus to the hair cell synapse. Further extending from the modiolus into the osseous labyrinthine space is the osseous spiral lamina, which coils around the centre of the cochlea. This provides partial division of the upper and lower cochlear chambers into the scala vestibuli and scala tympani, which at the apex of the cochlea, communicate with each other at the helicotrema. The basilar membrane is the lower border of the membranous labyrinth encasing the scala media. The scala tympani is closed by secondary tympanic membrane. It is also connected with the subarachnoid space through the *aqueduct of cochlea*

The widths of the spiral lamina and basilar membrane are inversely related along the length of the cochlea. The spiral lamina wider at the base and narrowing toward the apex, while the basilar membrane narrower at the base and wider at the apex. This is one of factors for the frequency specificity of basilar membrane motion.

HRCT temporal bone axial image demonstrate the cochlea anterior to internal auditory meatus.

Cochlear aqueduct:

The cochlear aqueduct which is at the basal turn of the cochlea , is a bony channel that allows communication between the perilymphatic fluid and cerebrospinal fluid of the subarachnoid space.

Labyrinthitis Ossificans

Children with labyrinthitis ossificans presented with SNHL and a history of meningitis, sickle cell disease, autoimmune diseases, and otitis media and cholesteatoma. Labyrinthitis ossificans (LO) are demonstrated by radiological imaging. Not only the late stage but also the early fibrous stage of LO that can make electrode insertion at cochlear implantation difficult or even impossible. LO can occur as early as 2 weeks following meningitis .

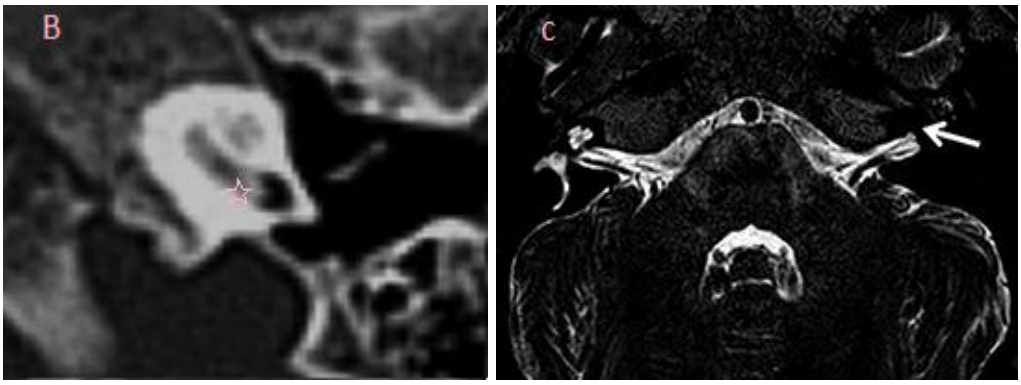


Figure 20:HRCT & MRI Axial image shows labyrinthine ossificans

HRCT is insensitive to the fibrous stage, while detected on MRI by a loss of normal fluid signal within the membranous. The most common site of Labyrinthine ossificans is proximal scala tympani within the inferior basal turn.

High-resolution MRI is imperative to delineate the two main scalar chambers and degree of involvement, which is helpful in determining surgical approach. At the time of MR imaging in children for meningitis, axial 2–3 mm post-contrast images

through the temporal bones may be helpful to detect the acute inflammatory phase and predict the development of SNHL, which can be suggested by presence of labyrinthine enhancement . This may help to rapidly direct these

Children to the otolaryngologist for implant evaluation.

Once osseous obstruction is present, the success of cochlear implantation decreases.

VESTIBULE:

The initial point of communication between the middle and inner ears is at the oval window of the vestibule .The vestibule is situated between the internal auditory meatus anteromedially and the middle ear cavity laterally .The cochlea is anterior to the vestibule. It is connected to the vestibule by the narrow ductus reuniens. Posterior and lateral to the vestibule are the mastoid air cells. Medially is the posterior cranial fossa, into which the endolymphatic duct and sac extend beneath the dura.

SEMICIRCULAR CANALS:

There are three semicircular canals, which are sensitive to angular accelerations. Each canal is at approximately right angles to the other two. Each canal is maximally sensitive to rotations that lie along the plane of that canal, and because of this arrangement the three canals can specify the direction and amplitude of any arbitrary head rotation. Each of the canals acts as an integrating accelerometer. The necessary stimulus for the canal is an angular acceleration, but the information that is encoded by the firing of the afferent nerve fiber , and is more closely related to angular velocity. The canals are organized into functional pairs. Any rotation in that plane is excitatory to one of the canals of the pair and inhibitory to the other. In the horizontal system the two horizontal canals form a functional pair. In the vertical system, the anterior canal on one side is parallel and coplanar with the posterior canal on the opposite side.

Internal auditory meatus:

It is a bony canal within the petrous portion of the temporal bone which transmits the nerves and vessels from within the posterior cranial fossa to the auditory and vestibular apparatus.

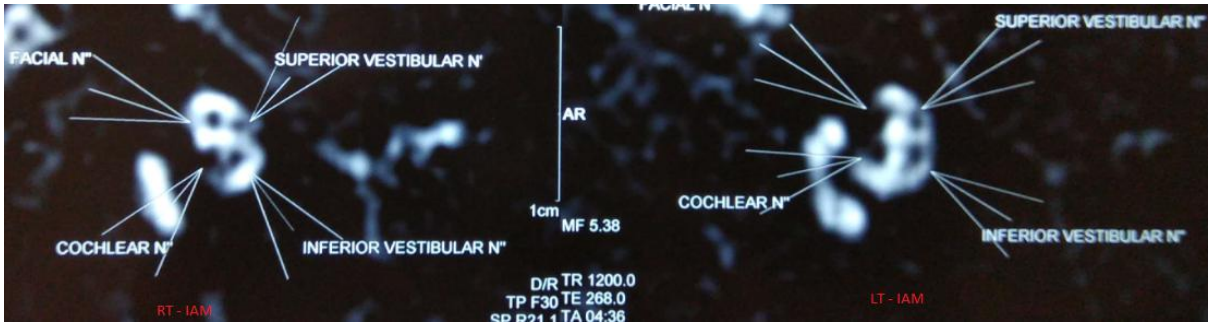


Figure 21; MRI Sagittal View - Internal Auditory Meatus with four nerves

The medial opening of the IAM is porus acousticus which is located within the cranial cavity, this margin of the opening are smooth and rounded, and the distance of the canal is short approximately 10 mm running laterally to the bone. Lateral end of the canal narrows which is called as fundus, where the canal splits into three distinct openings, one of which is the facial canal.

The contents of the IAM include, facial nerve, vestibulocochlear nerve, vestibular ganglion, labyrinthine artery .There are five nerves that run through the IAM separately which are motor root of facial nerve, nervus intermedius, cochlear nerve, superior and inferior vestibular nerve.

Their position is most constant in the lateral portion of the meatus which is anatomically divided by the crista falciformis. This horizontal ridge divides the canal into superior and inferior portions. The superior portion further divided vertically by ridge of bone called as Bill's bar. The facial nerve is running in the anterosuperior segment of meatus, superior vestibular nerve lies in the posterosuperior. . cochlear nerve and inferior vestibular nerve lies in the inferior portions, in which the cochlear nerve is situated anteriorly.

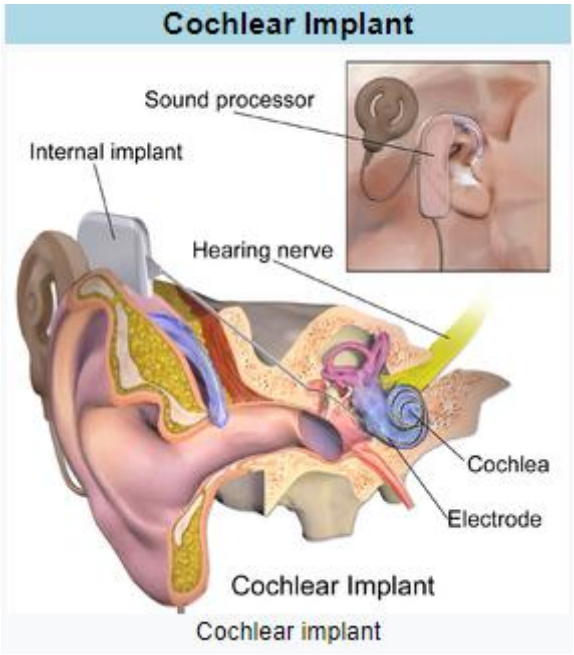
CLASSIFICATION HEARING LOSS:

Hearing loss is classified as conductive, sensorineural and mixed hearing loss. The degree of hearing loss classified by world health organization in pure tone audiometry, 10 to 25dB- normal hearing, 26 to 40dB – mild hearing loss, 41 to 55dB moderate hearing loss,56 to 70dB severe hearing loss, 71 to 90dB profound hearing loss, 91dB and above total hearing loss.

Sensorineural hearing loss is due to dysfunction of the inner ear or auditory nerve. It may be congenital or acquired. 50% of the all cases of congenital hearing loss due to environmental factors, such as congenital hyper bilirubenemia, ototoxic medication exposure, neonatal hypoxia, viral infection and meningitis. Other 50% of cases attributed to genetic causes. Of this genetic cases 30% syndromic (Branchio-oto-renal, stickler, waardenberg, treacher Collins, penred syndrome) 70% non syndromic (75 to 80% autosomal dominant, 20% autosomal recessive, 2 to 5% X-linked, < 1% mitochondrial inheritance).

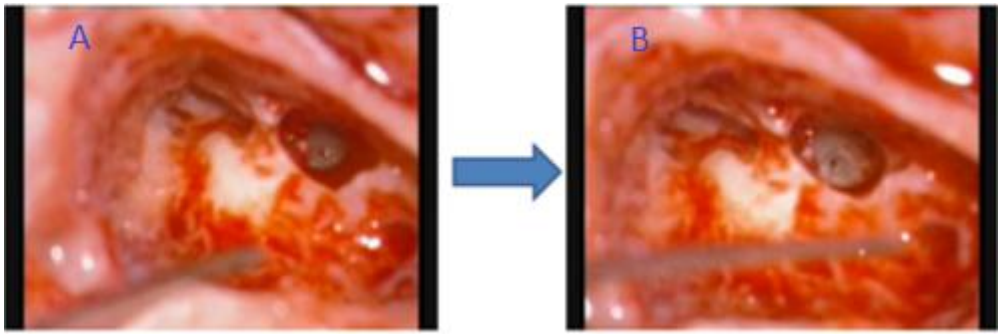
COCHLEAR IMPLANT SURGERY:

The majority of CI operations with or without malformations can be done via the classical transmastoid-facial recess approach. Sometimes, the presence of complex malformations conformed by radiological imaging makes this approach impossible and the surgeon must be ready to modify the surgical approach.



Cochlear implant

CI surgery done under the general anaesthesia, Lazy double flap incision made in the post auricular region then palva flap is elevated. Mastoidectomy is performed, the cavity should not be saucerized. The facial recess is identified and widely opened. Visualisation of anatomy of round window, if clearly seen remove the anterior lip of the round window to insert the electrodes otherwise make the cochleostomy inferior to the inferior attachment of round window membrane.



A- RW membrane not visible. B: Conventional bony cochleostomy performed with drilling for insertion of the electrode array.

The surgical drill is used to create a well to exactly fit the device. After fixation of stimulator the electrode array should be inserted as atraumatically as possible in to the scala tympani and force should never be used. Closure should be accomplished in layers.

SCORING CHART:

Sanjay vaid et al (2015) studied the role of HRCT & MRI of the temporal bone in predicting and grading the degree of difficulty of cochlear implant surgery. The authors assigned 10-point scoring chart of HRCT and magnetic resonance imaging MRI imaging findings in patients being assessed preoperatively for cochlear implantation. This chart helps in objectively assessing the degree of difficulty of the surgical procedure and alerts the surgeons to any potential intraoperative complications.

10-point scoring chart based on HRCT and MRI imaging findings are

(1) Grades of mastoid pneumatisation; Authors assign a score of ‘0’ for a well pneumatized mastoid, and a score of ‘1’ for a non or hypo pneumatized mastoid.

(2)Facial recess; if wide facial recess author assign it a score of ‘0’. If the facial recess measures less than 3 mms, it is labelled as narrow with a score of ‘2’.

(3) Mastoid segment of facial nerve canal; A normally positioned descending facial nerve is assigned a score of ‘0’. If the descending segment of the facial nerve canal overhangs the round window niche, a score of ‘2’ is assigned.

(4). Position of the jugular bulb; If it`s normal position the score of‘0’. If the jugular bulb extends above these anatomical landmarks, it is a high riding jugular bulb and assigned a score of ‘1’

(5) Posterior wall of external auditory canal/sigmoid sinus lines; one line is drawn along the posterior wall of the external auditory canal and the second line is drawn tangential to the sigmoid sinus. These lines are drawn on the axial HRCT image where the tympanic annulus and the handle of malleus are visible in the same plane. Basal turn of cochlea between these two lines, and assigned a score of ‘0’ and if basal turn of cochlea outside these two lines assigned a score of ‘1’

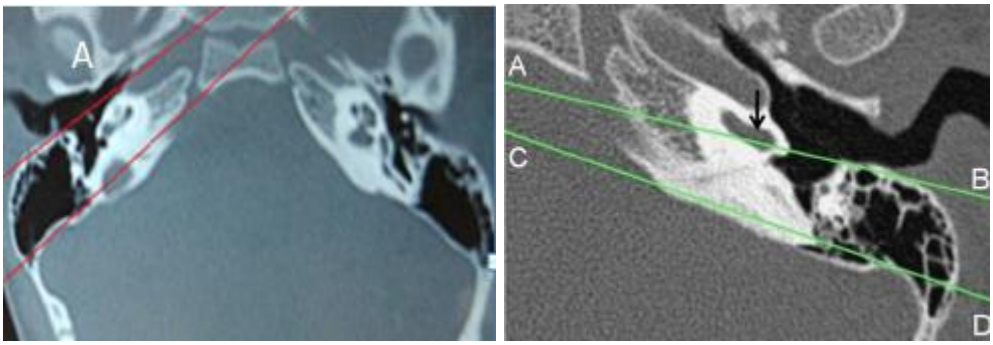


Figure 22: HRCT Axial image A- The posterior canal wall/sigmoid sinus lines: B-The posterior canal wall/long axis of the basal turn of cochlea

(6) Posterior wall of external auditory canal/long axis of the basal turn line; one line is drawn along the posterior wall of the external auditory canal, and the second line is drawn along the center of the long axis of the basal turn of cochlea. These lines are drawn on the axial HRCT image depicting the entire basal turn of the cochlea. Normally, these two lines run parallel to each other these cases are assigned a score of '0'. If the lines intersect, such cases are assigned a score of '1'.

(7)Relative position of the basal turn of cochlea to the malleoincudal joint in axial plane; Ice cream cone is usually never identifiable in the same axial HRCT image that depicts the basal cochlear turn. These cases are normal and assigned a score of '0'. If the cochlea is rotated away from its normal, the superior 'ice-cream cone 'is identified in the same axial section as that of the basal cochlear turn and these cases are assigned a score of '1'

(8) Lines along the anterior margin of the IAC (rotated cochlea); normally line is drawn along the internal auditory canals are parallel to each other and do not intersect. These cases are assigned a score of '0'. If the internal auditory canals

are mal-angulated anteriorly, these lines will intersect one another and are assigned a score of ‘1’.

(9) Associated congenital anomalies of the temporal bone; a score of ‘0’ was assigned to normal imaging without inner ear anomalies. Patients with isolated LVAS, Mondini deformity and bulbous IAC were assigned a score of ‘1’. Patients with IP-Type I, IP-Type III and common cavity were assigned a score of ‘4’.

(10) Associated acquired abnormalities of the temporal bone (labyrinthitis ossificans and Otosclerosis) The authors assign a score of ‘0’ when cochlear ossification and otosclerosis are not present. Labyrinthitis ossificans (Balkany Grade 1) are assigned a score of ‘2’. Labyrinthitis ossificans (Balkany Grade 2)/otosclerosis are assigned a score of ‘4’.Labyrinthitis ossificans (Balkany Grade 3) are assigned a score of ‘6’.

After grading the pre-operative imaging based on the 10-point scoring chart

The author concluded that children who have PDS between 0 and 3(Grade 1) have uneventful and uncomplicated surgery with the lowest intraoperative times. Children with PDS between 4 and 7 alert the surgeon to moderate surgical difficulty and longer intraoperative times. PDS of 8 and above indicate prolonged and difficult surgery. We were followed this scoring system also in our study.

6. MATERIAL AND METHODS

STUDY DESIGN:

Prospective study

STUDY SETTING:

Upgraded Institute of otorhinolaryngology, Rajiv Gandhi Government General Hospital, Chennai-600 003

STUDY PERIOD:

October 2015 to September 2017

STUDY SUBJECTS:

Children with bilateral profound sensorineural hearing loss are admitted for pre-operative evaluation of cochlear implant surgery during the study period at Upgraded institute of otorhinolaryngology, Rajiv Gandhi Government General Hospital, Chennai, who satisfy the inclusion criteria.

SAMPLE SIZE:

30-40 children

INCLUSION CRITERIA:

- Children aged between one to six years
- Bilateral profound sensorineural hearing loss with >90dB in better ear
- No appreciable benefit with digital hearing aid
- Motivated parents for surgery
- No medical contraindications

EXCLUSION CRITERIA:

- Children aged more than six years
- Benefit with digital hearing aid
- Parents not willing for cochlear implant surgery

DATA COLLECTION:

Data will be collected from the patients using semi-structured questionnaire.

Clinical examination and other relevant investigations will be done. Data will be collected from radiological department.

DATA ANALYSIS:

Data will be entered into Microsoft excel sheet and data analysis will be done by using appropriate statistical packages.

Quantitative and qualitative data will be expressed in Mean and proportions. Appropriate tests of significance will be used.

BENEFIT TO COMMUNITY:

Selection of candidacy for cochlear implant surgery, which will be helpful for preventing hearing handicap and to have better quality of life.

ETHICAL CONSIDERATIONS:

Approval from Institutional Ethics Committee.

Informed written consent will be taken from the parents/ guardian of the study participation before the study process.

The information collected will only be used for the study purpose and strict confidently will be maintained throughout the study.

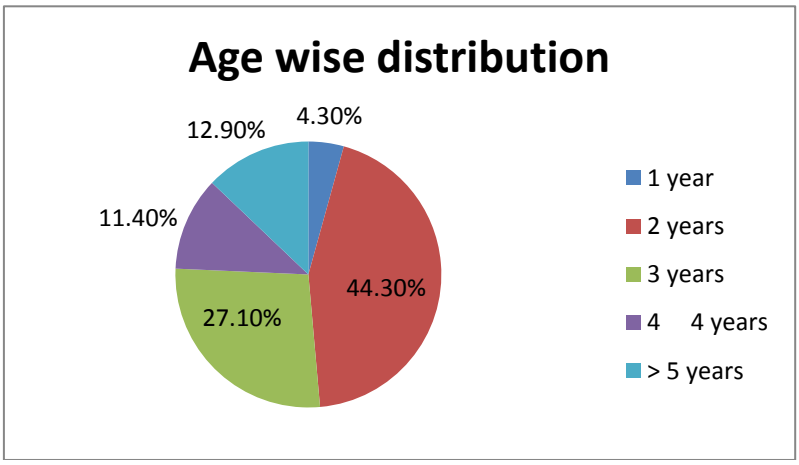
7.RESULTS AND ANALYSIS

Totally seventy children with profound sensory neural hearing loss had participated in the study. Characteristics and imaging features of the study participants are describes as per the following:

1. AGE WISE DISTRIBUTION:

Table-1: Percentage distribution of study participants by age group:

Age group	Frequency	Percent
1 year	03	4.3
2 years	31	44.3
3 years	19	27.1
4 years	08	11.4
≥ 5 years	09	12.9
Total	70	100.0



Among the study patients, Majority (44.3%) were in 2 years age group, followed by 3 years age group (27.1%) 5 years (12.9%) and4 years(11.4%) . Majority of the children participated in the study were 2-3 years of age (71.4%).

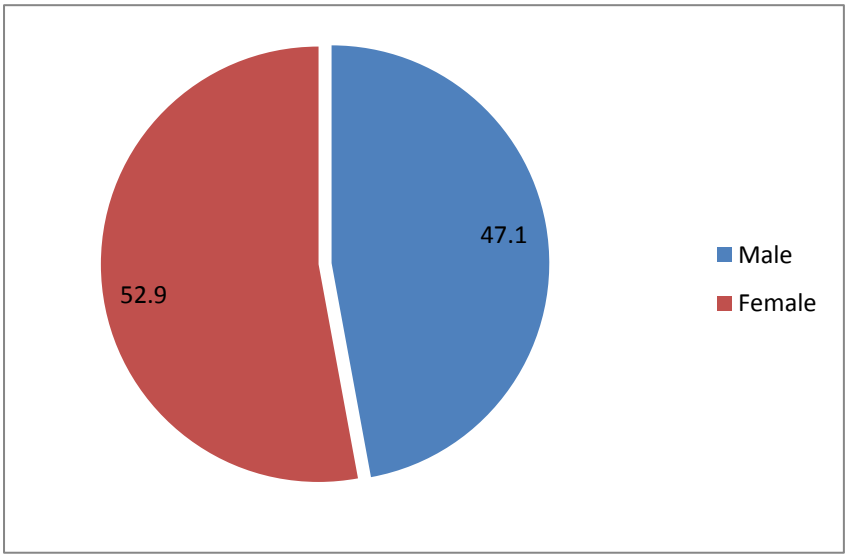
2. SEX DISTRIBUTION:

Table-2: Percentage distribution of study participants by sex:

Sex	Frequency	Percent
Male	33	47.1
Female	37	52.9
Total	70	100

Regarding sex distribution of participants, the females are slightly higher than males.

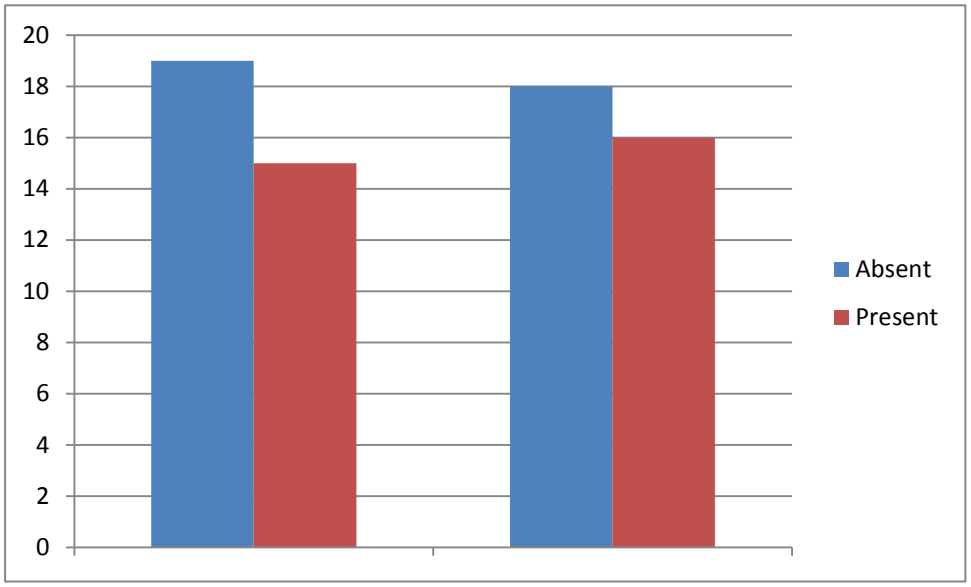
Percentage distribution of study participants by sex:



3.AGE WISE MASTOID PNEUMATIZATION:

***Table-3: Percentage distribution of study participants by* mastoid pneumatization in <2years of age:**

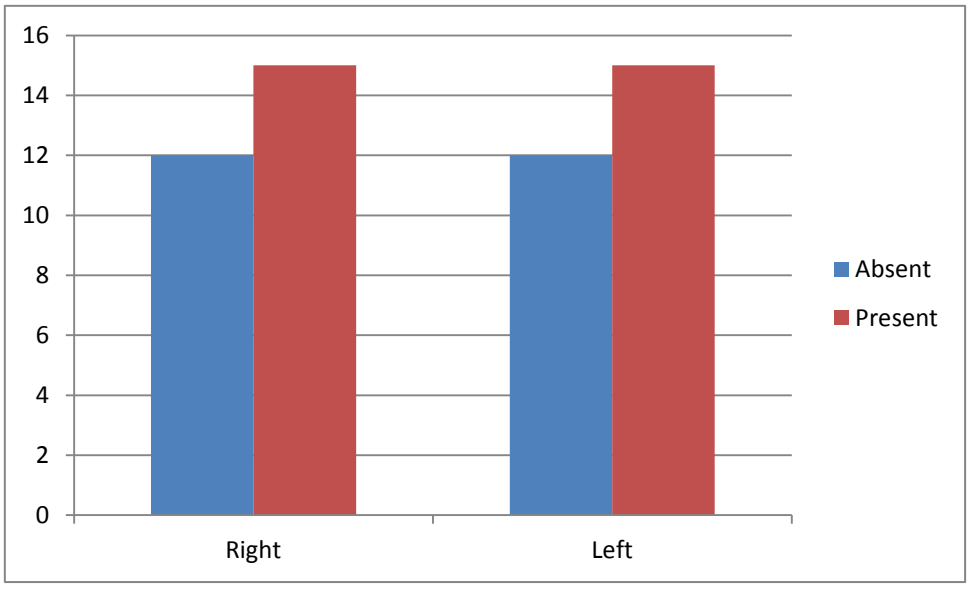
MASTOID PNEUMATIZATION	Right		Left	
	Frequency	Percent	Frequency	Percent
Absent	19	55.9	18	53.0
Present	15	44.1	16	47.0
Total	34	100.0	34	100.0



Among study subjects, majority of the children were below 2 years of age group. The CT scan findings of those children, well pneumatized mastoid (considered as present) in Right side 44.1%,Left side 47%.Hypo&non pneumatized mastoid(considered as absent in CT scan) in Right side 55.9% Left side 53%.This is due to increased marrow content of the mastoid.

***Table-4: Percentage distribution of study participants by* mastoid pneumatization in 2-4years of age:**

MASTOID PNEUMATIZATION	Right		Left	
	Frequency	Percent	Frequency	Percent
Absent	12	44.4	12	44.4
Present	15	55.6	15	55.6
Total	27	100.0	27	100.0

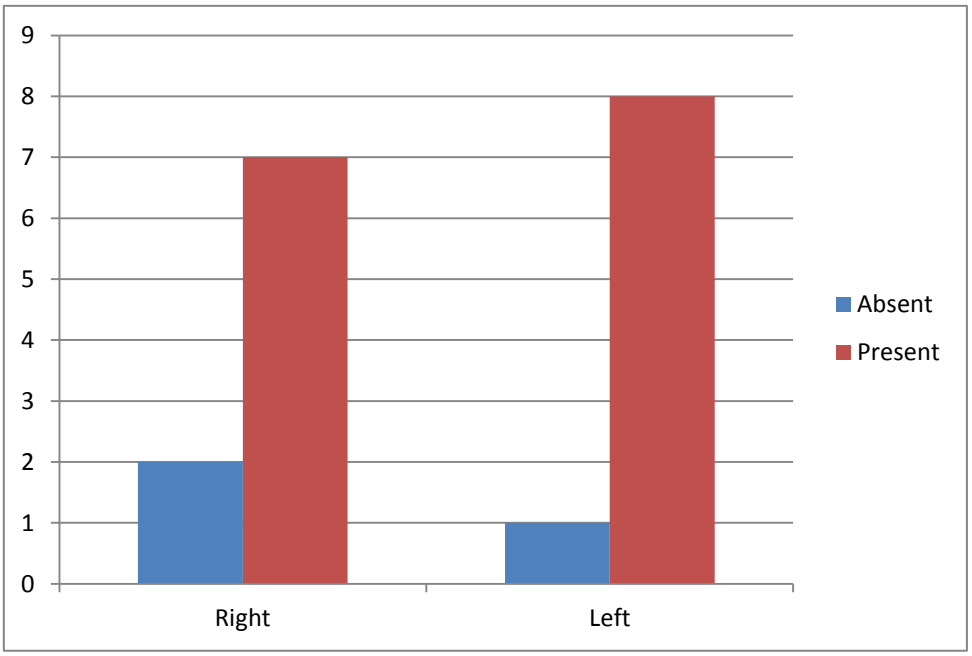


Out of 70 children, 27 subjects comes under the age group of 2 to 4 years.

Mastoid pneumatization is present 55.6% of the children on both sides.

***Table-5: Percentage distribution of study participants by* mastoid pneumatization in 4-6 years of age:**

MASTOID PNEUMATIZATION	Right		Left	
	Frequency	Percent	Frequency	Percent
Absent	2	22.2	1	11.1
Present	7	78.8	8	88.9
Total	9	100.0	9	100.0

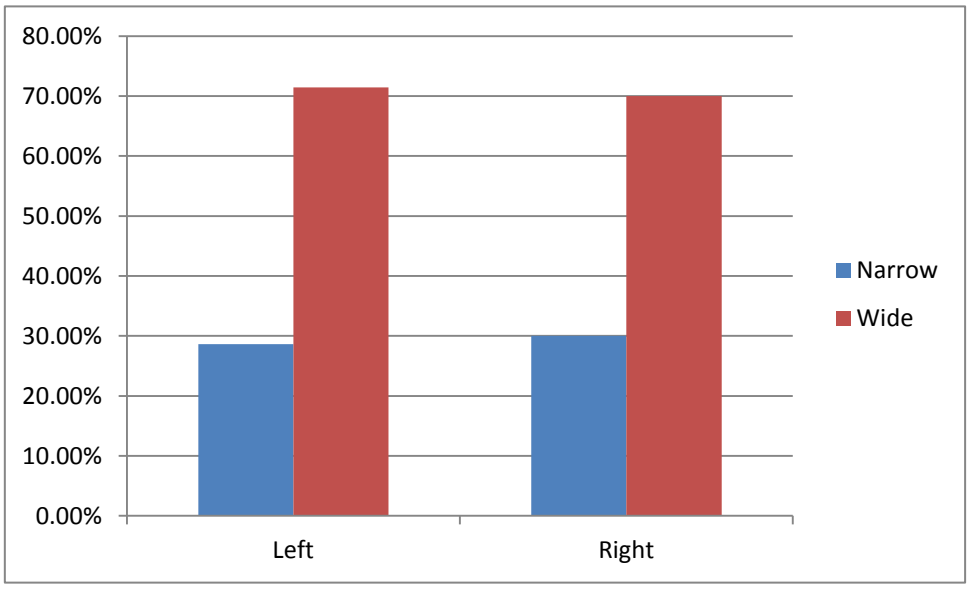


Among study subjects, only 9 children were 4-6 years of age group. Mastoid pneumatization is present in Right side 78.8%, Left side 88.9%.This is shows the increase in pneumatization of the mastoid bone and reduction in marrow content that occurs at age.

4. FACIAL RECESS:

Table-6: Percentage distribution of study participants by facial recess:

FACIAL RECESS	Right		Left	
	Frequency	Percent	Frequency	Percent
Narrow	21	30.0	20	28.6
Wide	49	70.0	50	71.4
Total	70	100.0	70	100.0



Among the study subjects, 70% on Right side,71.4% on Left side had wide (>3mm) facial recess.30% on Right side,28.6% on Left side had narrow (<3mm)facial recess.

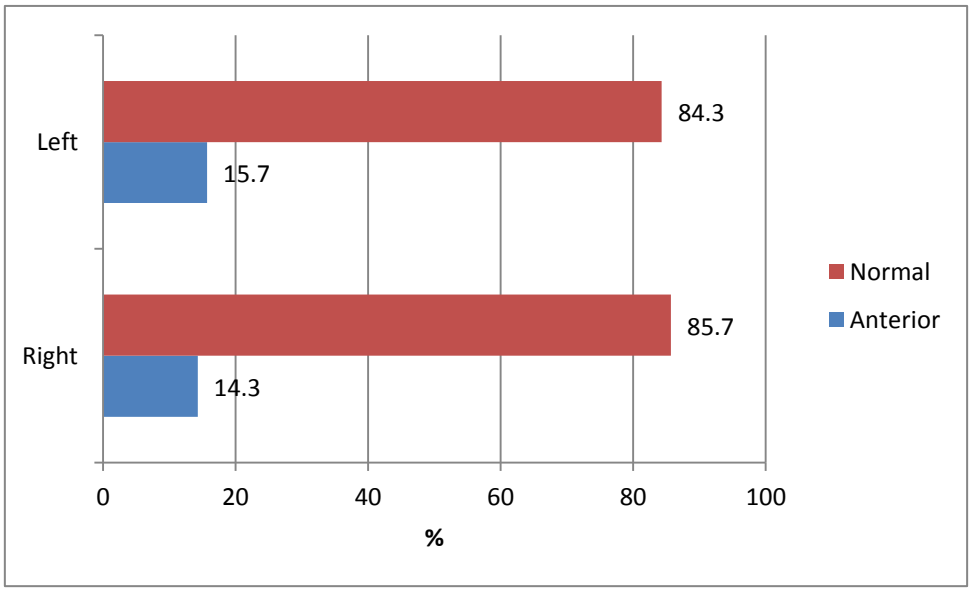
5.SIGMOID SINUS:

Table-7: Percentage distribution of study participants by sigmoid sinus:

SIGMOID SINUS	Right		Left	
	Frequency	Percent	Frequency	Percent
Anterior	10	14.3	11	15.7
Normal	60	85.7	59	84.3
Total	70	100.0	70	100.0

70 children CT scan were reviewed. Of this anteriorly displaced sigmoid sinus on right side 14.3%, Left side15.7%.

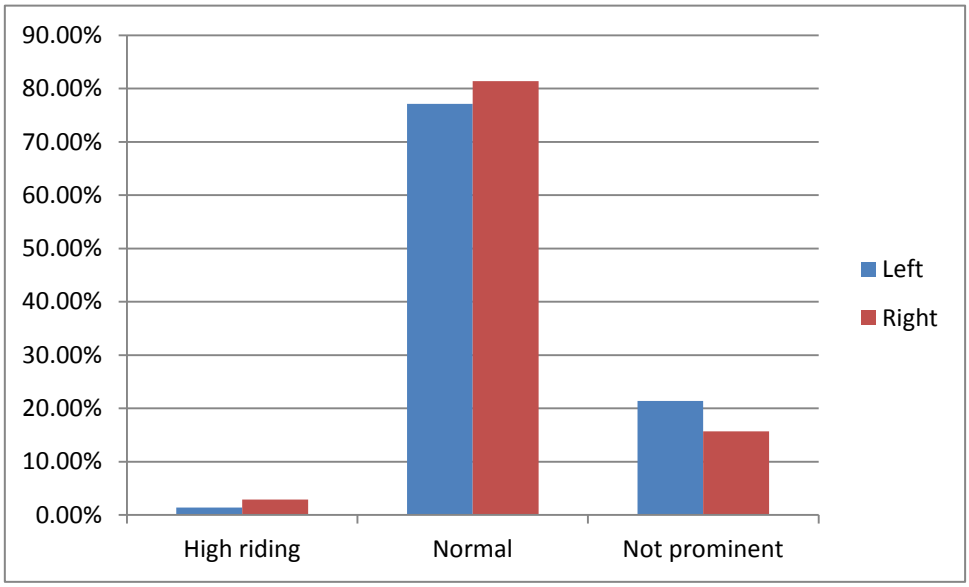
Percentage distribution of study participants by sigmoid sinus:



6. JUGULAR BULB:

Table-8: Percentage distribution of study participants by jugular bulb:

JUGULAR BULB	Right		Left	
	Frequency	Percent	Frequency	Percent
High riding	2	2.9	01	1.4
Normal	57	81.4	54	77.1
Not prominent	11	15.7	15	21.4
Total	70	100.0	70	100.0



Among 70 children participated in the study only 2.9% on right side, 1.4% on left side had high riding jugular bulb. Not prominent in HRCT 15.7% on Right side, 21.4% on Left side, which is most commonly found less than 2 years of age group due to typical bulbous enlargement not developed.

7. ROUND WINDOW NICHE:

Percentage distribution of patients by round window niche:

Right **Left**

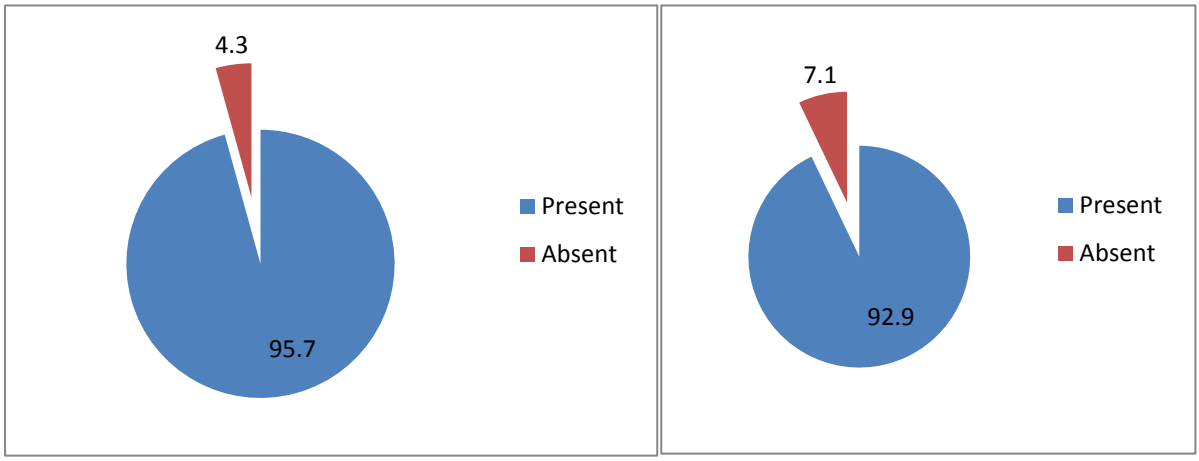


Table-9: Percentage distribution of study patients by round window niche:

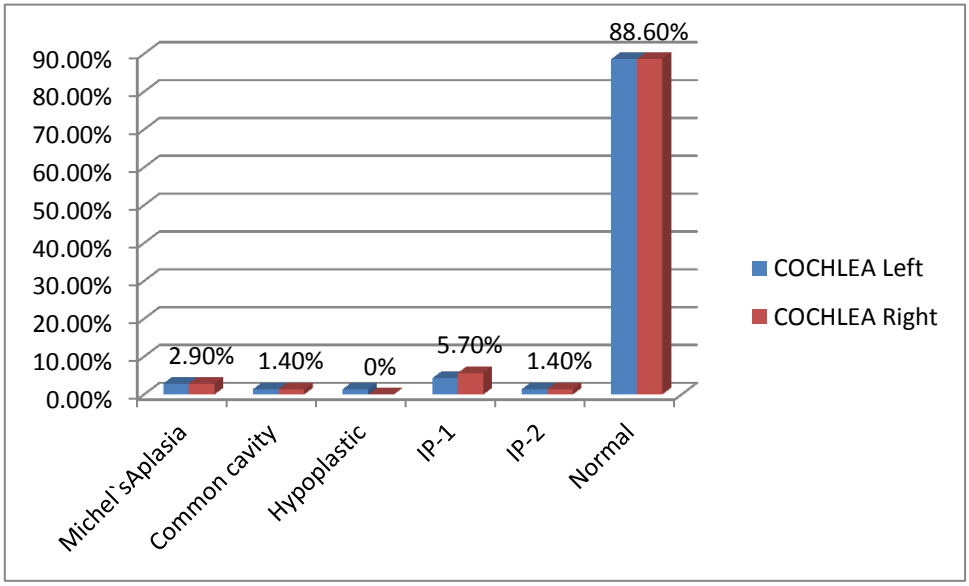
ROUND WINDOW NICHE	Right		Left	
	Frequency	Percent	Frequency	Percent
Absent	3	4.3	5	7.1
Present	67	95.7	65	92.9
Total	70	100.0	70	100.0

Among study subjects 4.3% right side, 7.1% left side the round window niche found to be absent. It is associated with inner ear malformations.

8. COCHLEA:

Table-10: Percentage distribution of study subjects by cochlea:

COCHLEA	Right		Left	
	Frequency	Percent	Frequency	Percent
Michel's Aplasia	2	2.9	2	2.9
Common cavity	1	1.4	1	1.4
Hypoplastic	0	0	1	1.4
IP-1	4	5.7	3	4.3
IP-2	1	1.4	1	1.4
Normal	62	88.6	62	88.6
Total	70	100.0	70	100.0

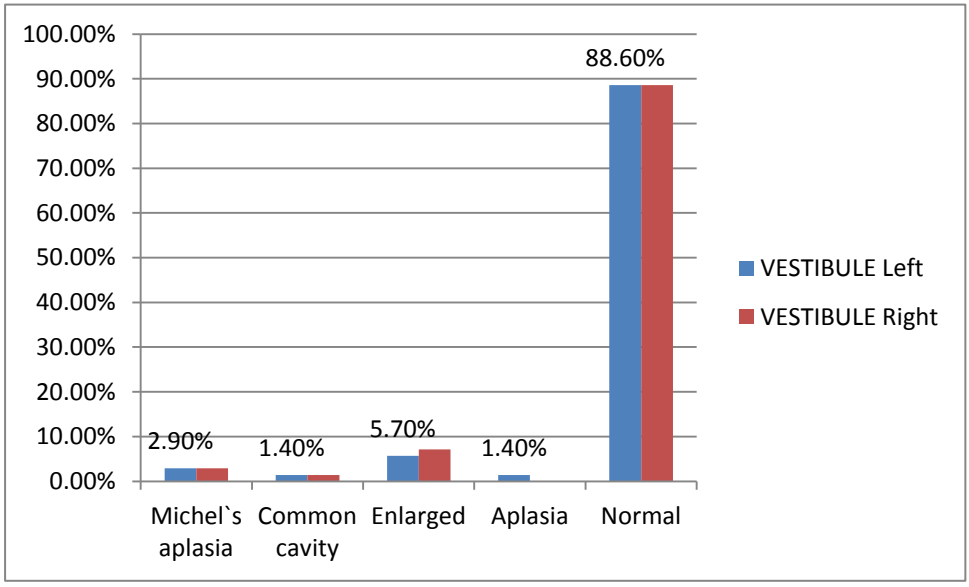


Evaluated 140 inner ears in 70 children with profound sensorineural hearing loss. All patients were studied with HRCT temporal bone & MRI inner ear. Out of 70 subjects, 4 inner ears showed Michel`s aplasia, 12 inner ears had cochlear malformations, associated with multiple abnormalities.

9.VESTIBULE:

Table-11: Percentage distribution of study participants by vestibule:

VESTIBULE	Right		Left	
	Frequency	Percent	Frequency	Percent
Michel`s aplasia	2	2.9	2	2.9
Common cavity	1	1.4	1	1.4
Enlarged	5	7.1	4	5.7
Aplasia	0	0	1	1.4
Normal	62	88.6	62	88.6
Total	70	100.0	70	100.0

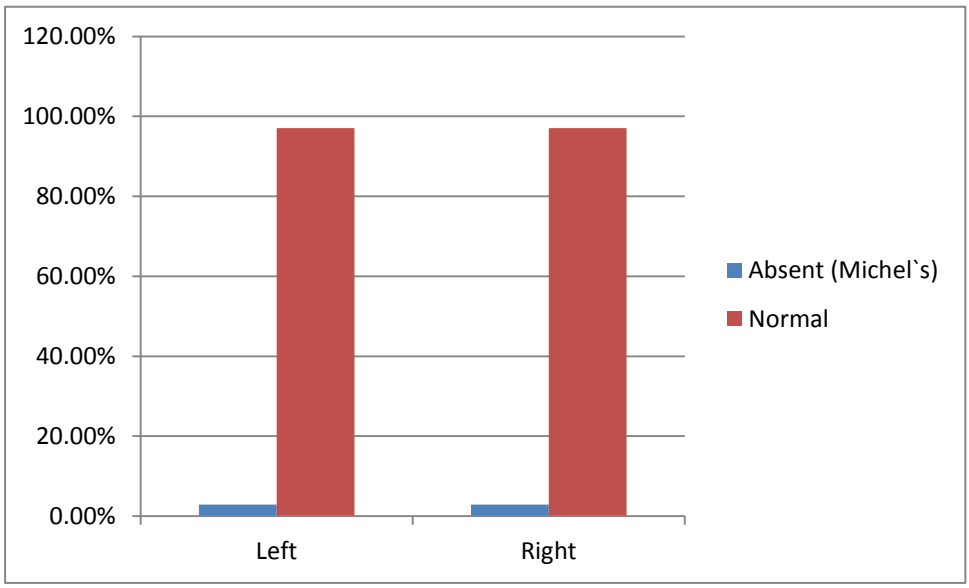


Among study subjects, 12 inner ears had vestibular anomalies along with other abnormalities,

10. COCHLEAR AQUEDUCT:

Table-12: Percentage distribution of study participant's by cochlear aqueduct:

COCHLEAR AQUEDUCT	Right		Left	
	Frequency	Percent	Frequency	Percent
Absent (Michel`s)	2	2.9	2	2.9
Normal	68	97.1	68	97.1
Total	70	100.0	70	100.0



Cochlear aqueduct is absent in 4 inner ears in 3 children with congenital deafness. One patient had bilateral complete labyrinthine aplasia; another two children had unilateral labyrinthine aplasia with absent cochlear aqueduct.

11. VESTIBULAR AQUEDUCT:

Percentage distribution of study participants by vestibular aqueduct:

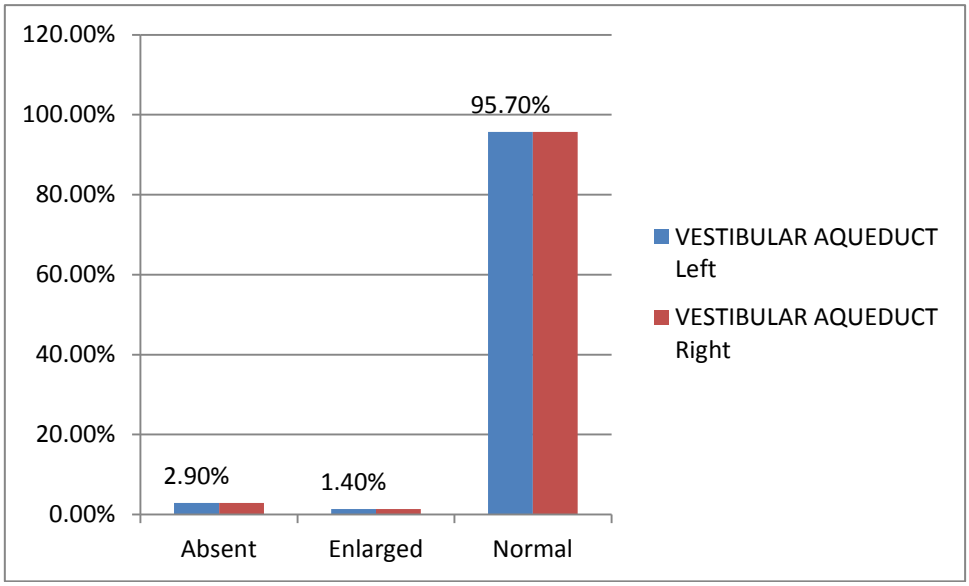


Table-13: Percentage distribution of study participant's by vestibular aqueduct:

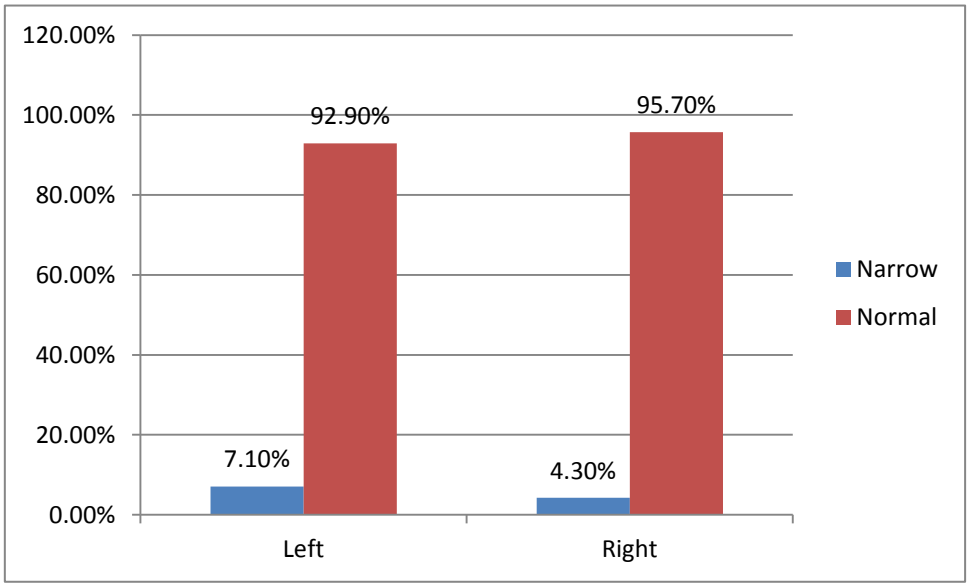
VESTIBULAR AQUEDUCT	Right		Left	
	Frequency	Percent	Frequency	Percent
Absent	2	2.9	2	2.9
Enlarged	1	1.4	1	1.4
Normal	67	95.7	67	95.7
Total	70	100.0	70	100.0

Absence vestibular aqueduct associated with Michel`s aplasia. One children (1.4%) had bilateral enlarged vestibular aqueduct.

12. INTERNAL AUDITORY MEATUS:

Table-14: Percentage distribution of study participant's by internal auditory meatus:

IAM	Right		Left	
	Frequency	Percent	Frequency	Percent
Narrow	3	4.3	5	7.1
Normal	67	95.7	65	92.9
Total	70	100.0	70	100.0

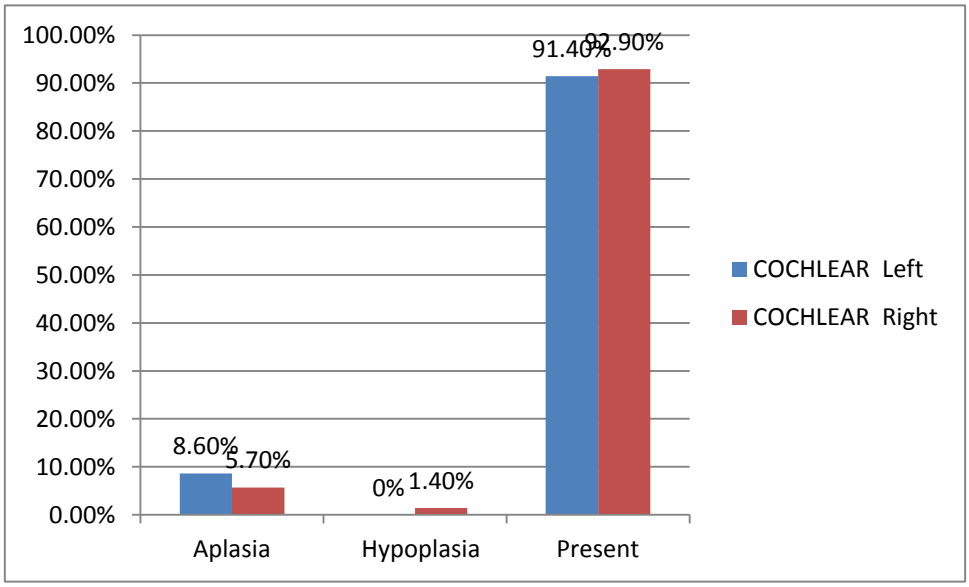


Out of 140 inner ears, narrow IAM were identified in 8 ears out of 6 ears has been associated with cochlear nerve aplasia.

13. COCHLEAR NERVE:

Table-15: Percentage distribution of study subjects by cochlear nerve:

COCHLEAR NERVE	Right		Left	
	Frequency	Percent	Frequency	Percent
Aplasia	4	5.7	6	8.6
Hypoplasia	1	1.4	0	0
Present	65	92.9	64	91.4
Total	70	100.0	70	100.0

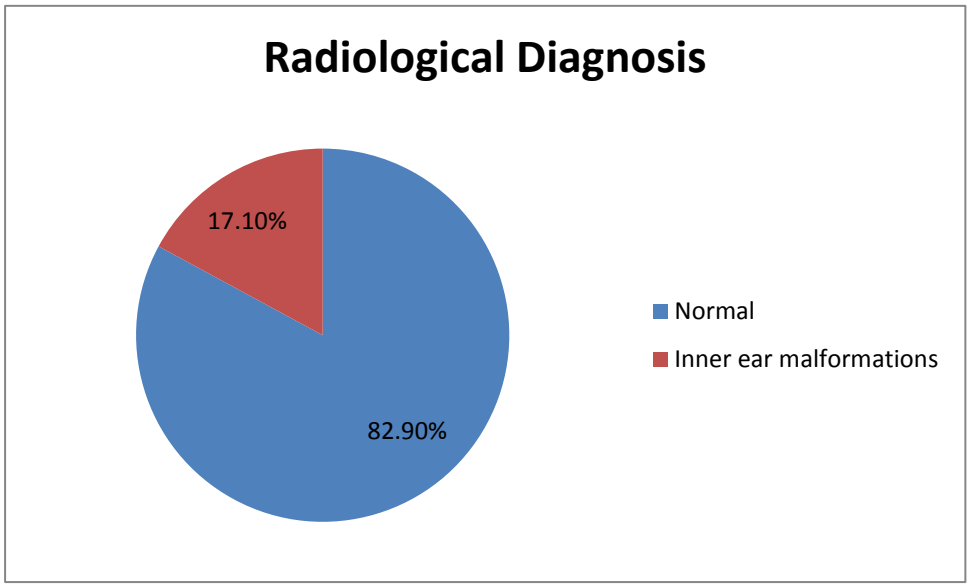


Among the study participants, 5.7% Right side, 8.6% side had cochlear nerve aplasia (3 children with bilateral cochlear nerve aplasia, 4 children with unilateral cochlear nerve aplasia). 1.4% Right side (one child) had hypoplastic cochlear nerve, cochlear implantation done in same side.

14. INNER EAR MALFORMATIONS:

Table-16: Percentage distribution of study subjects by inner ear malformations:

RADIOLOGICAL DIAGNOSIS	Frequency	Percentage
Normal	58	82.9
Inner ear Malformations	12	17.1
Total	70	100.0



Out of 70 children 82.9% (58 patients) demonstrated normal inner ear findings in HRCT & MRI temporal bone. **17.1% (12 children) demonstrated various bony malformations of the cochlea vestibular system, internal auditory canal and cochlear nerve aplasia.**

Table-17: Percentage distribution of study subjects by inner ear malformations:

Radiological diagnosis	Frequency	Percentage
Normal	58	82.9
Bilateral cochlear nerve aplasia	1	1.4
Bilateral cochlear nerve aplasia with narrow IAM	2	2.8
Unilateral cochlear nerve aplasia with abnormal cochlea and vestibule both sides.	4	5.7
Bilateral Michel`s aplasia	1	1.4
Bilateral Incomplete partition type 1	2	2.8
Incomplete partition type 1 one side, common cavity other side	1	1.4
Bilateral dilated vestibular aqueduct	1	1.4

Unilateral cochlear nerve aplasia associated with bilateral cochlea vestibular malformations (4 children) were the most common radiological abnormality in our study. The next common anomalies are bilateral cochlear nerve aplasia with narrow IAM.

Table-18: Percentage distribution of overall evaluation of inner ear malformations:

TYPE	Frequency	Percentage
Michel`s aplasia	4/49	8.2
Cochlea	12/49	24.5
Vestibule	12/49	24.5
Vestibular aqueduct	2/49	4.1
Internal auditory canal	8/49	16.3
Cochlear nerves	11/49	22.4

Total of 49 malformations were detected in 24 abnormal inner ears in a total of 12 patients. 4 of 49(8.2%) was Michel's aplasia, 12of 49 (24.5%) inner ear malformations showed malformations of cochlea and in 12 of 49 (24.5%) inner ear malformations vestibule was abnormal. In 2 of 49 (4.1%) inner ear malformations vestibular aqueduct was abnormal, In 8 of 49 (16.3%) inner ear malformations internal auditory canal was found to be malformed. In 11 of 49 (22.4%) cochlear nerves anomalies were present.

15. COCHLEAR IMPLATATION:

Table-19: Percentage distribution of study subjects by side selected for cochlear implantation:

Surgery	Number	Percentages
Right Cochlear Implantation	60	85.7
Left cochlear implantation	2	2.9
planned for CI on R side	2	2.9
planned for CI on L side	1	1.4
Not done	4	5.7
Not done due to willingness issue	1	1.4

Table-20: Percentage distribution of study subjects by Electrode insertion:

Electrode insertion	Frequency	Percentage
RW Insertion	55	88.7
PR Cochleostomy	07	11.3
Total	62	100

Of the 70 children recruited for the study, only 62 children had undergone the prescribed surgical procedure most commonly RW insertion (55 children-88.7%).

Table-21: Percentage distribution of study subjects by potential difficulty score to predict the degree of difficulty of cochlear surgery:

Score	Frequency	Percentage
Grade 1(0-3 score)	47	72.3
Grade 2(4-7 score)	16	24.6
Grade 3 (>8 score)	2	3.1

There were 47 children (72.3 %) classified as Grade 1, 16 children (24.6%) classified as Grade 2 and 2 children (3.1 %) classified as Grade 3. Out of the 47 children in Grade 1, it was observed that 39 children had uneventful surgery and 8 children had prolonged surgery .Out of these 8 children, 5 children had descending segment of the facial nerve canal overhangs the round window niche,2 children had increased marrow content of mastoid and one child had congested middle ear mucosa. Out of the 16 children assigned as Grade 2,10 children had minor surgical difficulties as predicted by specific imaging findings on the scoring chart, and 4 had uneventful surgery, 2 children are awaited for implant.2 children assigned as Grade 3, one child had prolonged and difficult surgery due to inner ear abnormalities. Another child is awaited for implant.

8.DISCUSSION

A Prospective study was done, a total of 70 children (140 ears) with the age group of less than 6 years with bilateral congenital severe to profound sensorineural hearing loss were radiologically evaluated with HRCT of temporal bone and MRI of inner ear, at Upgraded Institute of Otorhinolaryngology, Rajiv Gandhi Government General hospital, Chennai. A total of 70 children were evaluated for the study satisfying the inclusion criteria. For all children studied the radiological findings of mastoid pneumatization, facial recess, sigmoid sinus, jugular bulb cochlea-vestibular anomalies, vestibular aqueduct, presence of cochlear nerve, course of facial nerve and other abnormalities. Finally we made surgical decisions to select candidacy for surgery, side selection and surgical technique in cochlear implantation.

In our study, among the study participants, Majority (44.3%) were in 2 years age group, followed by 3 years age group (27.1%) 5 years (12.9%) and4 years (11.4%). Majority of the children participated in the study were 2-3 years of age (71.4%).

In our study the mastoid pneumatization was evaluated by dividing the children in to three groups. The first group less than 2 years in which

well pneumatized mastoid 45.6%, Hypo and non pneumatization mastoid 54.4%. Second group between 2-4 years of age in which pneumatization was good in 55.6%, poor pneumatization in 44.4%.The third group >4 years of age well pneumatization 83.4%, Hypo & non pneumatization 16.6% of children. In our study mastoid pneumatization increased with age. During surgery we were encountered increased bleeding in poor pneumatized mastoid due to increased marrow content compare to well pneumatized mastoid.

It is correlates with the **James et al (2004)** have assessed the safety of cochlear Implantation surgery in children aged 12 months or younger is reviewed with radiological assessment of mastoid bone anatomy and surgical outcome. At the age of 2 years pneumatization increases to approximately 60%. But pneumatization was always adequate for safe identification of surgical landmark**.** No major complications occur due to poor pneumatisation. , so they concluded CI surgery is safe even under the one year of age.

In our study facial recess is considered as wide where the distance between the mastoid segment of facial nerve and posterior tympanic annulus is >3mm, narrow facial recess if <3mm.Among the study subjects, 70% on Right side, 71.4% on Left side had wide (>3mm) facial recess. 30% (21 children) on Right side, 28.6% (20 children) on Left side

had narrow (<3mm) facial recess**. *There is no difference between right and left side.***

In our institution all cochlear implantation performed through a transmastoid facial recess approach. Electrode insertion into the scala tympani is done either via the round window or a cochleostomy made anteroinferior to the round window. Out of 60 cochlear implantation ,we did 58 CI surgery on the Right side when identical finding in both ear and handedness of the surgeon to facilitate device manipulation. 2 CI surgery done left side because one child had poor pneumatization with narrow facia recess on Right side, well pneumatised mastoid with wide facial recess on Left side, Another child had narrow facial recess both sides but poor pneumatisation, anteriorly lying sigmoid sinus and high riding jugular bulb on Right side, these factors are favourable on Left side so we selected left side for implantation.

CI done with narrow facial recess of 19 children, during surgery we were encountered difficult to reach round window but facial nerve sheath exposed to assess the round window only in 7 children, in which 2 children had temporary facial palsy.

Sanjay vaid et al in 2015reported 30.9% narrow facial recess in their study between the age group of 9 months to 13 years of age. Lastly the author concluded narrow facial recess with other unfavourable factors, the surgeon was experienced the difficulties during surgery and prolonged operating time.

Olivier Deguine et al in 1995reported there wass no significant difference in results between children implanted on their dominant side and children implanted on their non-dominant side. The authors suggest implanting the better ear, provided there is no significant hearing in that ear. When both ears are identical, the side of implantation should be the side of handedness laterality to facilitate device manipulation (a practical reason).

In our study the sigmoid sinus was anteriorly displaced in CT scan on right side 14.3%, Left side15.7%.If sigmoid sinus was displaced anteriorly that there was no room available between EAC and the sigmoid sinus. CI surgery may not be feasible in this situation. Fortunately we did not encounter this type of severe case. We were experienced mild difficulty in doing posterior tympanotomy due to forward lying sigmoid sinus.

This is similar to the finding of **Atilla et al** who reported 12.4% anteriorly displaced sigmoid sinus**. Rebekah Clarke et al in 2017 & Audie L. Woolleyet al 1997** reported an incidence of 1.6% abnormal position of sigmoid sinus in CT scan.

Among study subjects the high riding jugular bulb only 2.9%(2 children) on right side, 1.4% (one child) on left side. Unfortunately we did not select the candidacy for cochlear implant surgery in the high riding jugular bulb side. One child had HJB along with other unfavorable peripheral factors on right side, we did surgery for that on left side. Another child with bilateral HJB is awaiting implant. **Rebekah Clarke et al in 2017**reported incidence of high riding jugular bulb3.5% to34%

In our study out of 70 children, 58 children (82.9%) were normal and **12 children (17.1%) were found to be congenital abnormal inner ear malformations.** All the 12 children had bilaterally abnormal inner ear (24 inner ears) with a majority of children demonstrating multiple abnormalities.

On evaluation of anomalies, out of 24 abnormal inner ears, in 4(16.7%) inner ear was found to be complete labyrinthine aplasia (Michel`s aplasia).

On evaluation of cochlear anomalies out of 24 abnormal inner ears 12(50%) cochlea found to be abnormal. Abnormalities of cochlea includes the cochlear hypoplasia, incomplete partition type-I (IP-I), incomplete partition type-II (IP-II), and common cavity deformity.

In 1(4.2%) inner ear, the cochlea was hypoplastic (type 1 –bud like cochlea).

In 7 (29.2%) inner ears, cochlea had no turn or only a bony mass without any turn was visualized so it was classified as incomplete partition type-I (IP-I).

In 2 (8.3%) inner ears, cochlea was of incomplete partition type-II (IP2-Mondini deformity) in this type the cochlea consists of 1.5 turns in which the middle and apical turns coalesce to form a cystic apex, accompanied by a dilated vestibule and enlarged vestibular aqueduct.

In 2 (8.3%) of cases cochlea was classified under the common cavity as there was cystic cavity representing the cochlea and vestibule, without showing any differentiation into cochlea and vestibule.

Vestibular anomalies, out of 24 abnormal inner ears in 12 (50%) inner ears vestibule was found abnormal. In 11 (45.8%) inner ears vestibule was dilated and in the rest 61(4.2%) it was aplastic.

Vestibular aqueduct anomalies, in 2 out of 24 (8.3%) of abnormal inner ears the vestibular aqueduct was found to be abnormal. Vestibular aqueduct was found to be dilated in both ears with normal vestibule.

On evaluation of internal auditory canal (IAC) anomalies, in 8 out of 24 (33.3%) of abnormal inner ears the internal auditory canal was found to be abnormal. In 6 (25%) inner ears IAC was found to be narrow in lumen. In 2

(8.3%) inner ear, complete bony septum was present in the proximal part of IAM.

On evaluation of the cochlear nerve , in all cases where IAC was malformed, vestibulocochlear nerves were also malformed expect for 2 children where IAC was found to be normal but cochlear nerves were aplastic in 2 inner ears, hypoplastic in 1 inner ear. Out of 24malformed inner ears, 11 (45.8%) inner ears had nerve anomalies. In10 (41.7%) of cases cochlear nerves were absent.in 1 inner ear(4.1) thin in diameter but well visualized (hypoplastic).

An overall evaluation of the malformations, a total of 49 malformations were detected in 24 abnormal inner ears in a total of 12children.4 of 49(8.2%) malformed inner ear showed complete labyrinthine aplasia (michel`s aplasia). In 12 of 49 (24.5%) showed malformations of cochlea. In 12 of 49 (24.5%) anomalies inner ear vestibule was abnormal. In 2 of 49 (4.1%) vestibular aqueduct was abnormal. In 8 of 49 (16.3%) inner ear malformations internal auditory canal was found to be malformed. In 11of 49 (22.4%) cochlear nerves anomalies were present.

Sangeet Kumar Agarwal et al in 2014 done a clinical study of radiological assessment of the Indian children with congenital sensorineural hearing loss in Sri Ganga Ram Hospital, New Delhi, A total of 280 children (1-14 years) with

congenital deafness (158 males and 122 females), between January 2002 to June 2013 were included in the study and they were assessed radiologically by HRCT scan of temporal bone and MRI with 3D scan of inner ear. **The congenital inner malformations were found 14.3% (4o children)** .Out of 560 inner ears we found 78 anomalous inner ears. Out of these 78 inner ears 57 (73%) had cochlear anomaly, 68 (87.1%) had anomalous vestibule, 44 (56.4%) had abnormal vestibular aqueduct, 24 (30.7%) had anomalous IAC, and 23 (29.4%) had abnormal cochleovestibular nerves.

D E Bamiou et al (2000) reported 28.4% congenital inner ear malformations of which dilated vestibular aqueduct (60%) most common anomalies, this is similar to the **woolford et al** who reported 29.5%.**Shusterman et al** 12.9%, **Zalzal et al** 6.8% of children found to be inner ear malformation.

In our study out of 70 children, 60 (85.6%) children received an implant after radiological imaging. In 58 out of 60(82.9%) children received cochlear implant in Right ear. In 2 out of (2.9%) 60 children received an implant in left side. 4 children (5.7%) were rejected for cochlear implant on the basis of imaging. Remaining 3 children (4.3%) are awaiting implants and for a child CI was not done due to willingness issue.

In 58 out of 60 (82.9%) children received cochlear implant in Right ear. In our institution when the peripheral and central factors are identical in HRCT scan of temporal bone and MRI with 3D scan of inner ear on both sides, routinely Right ear was selected for implantation to easy handling of surgeon.

In 2 out of (2.9%) 60 children received an implant in left side after imaging, because of unfavourable peripheral factors like one child had narrow facial recess on both sides and absent pneumatisation, anteriorly displaced sigmoid sinus, high riding jugular bulb on right sides, but these factors are favourable on left side. Another child had narrow facial recess& overhang of facial nerve on RW niche on right side, facial recess was wide on left side.

Olivier Deguine et al in 1995reported there wass no significant difference in results between children implanted on their dominant side and children implanted on their non-dominant side. The authors suggest implanting the better ear, provided there is no significant hearing in that ear. When both ears are identical, the side of implantation should be the side of handedness laterality to facilitate device manipulation (a practical reason).

In 4 out of 70children (5.7%) were rejected for cochlear implant on the basis of MRI findings. There are all 4 children had bilateral cochlear nerve aplasia with or without IAM abnormalities.

In 3out of 70 children (4.3%) are awaiting implants. This all 3 children have multiple inner ear malformations. For 2 children we are planned to do implantation on Right side (one child withIP-1 R Side, Michel`s aplasia L Side, cochlear nerve aplasia L Side&IP-1 R side, common cavity L side, cochlear nerve aplasia L side), we are selected Left side for a child with common cavity R side,IP-1 L side.

In our institution 4 children with inner ear malformations underwent cochlear implantation. Malformations are includes two incomplete partition type -1, one IP-1 with hypoplastic cochlear nerve, one dilated vestibular aqueduct.

Among the 62 children implanted, Round Window approach was used in 88.7% (55children) and 11.3% (7 children) through cochleostomy approach.

Naresh Panda& Mohan kameswaran et al in 2017 studied the evaluation of round window accessibility for electrode insertion. The round window approach has been advocated as a method to reduce insertion trauma and preserve residual hearing. However accessing the round window membrane could be difficult due to the bony overhang of the round window niche. Additionally the variable size and orientation of the round window may make electrode insertion difficult and challenging.

After achieving adequate posterior tympanotomy, the round window visibility was graded as Grade I >50% of the round window membrane is visible. Grade II 25% to 50% of the round window membrane is visible. Grade III <25% only a glimpse of the round window membrane can be seen and Grade IV Round window membrane is not visible.

They concluded Grade IV type of round window membrane either a round window insertion or a standard cochleostomy antero-inferior to the round window niche was performed. In Institute 1, 3 out of 9 (33%) required a standard cochleostomy whereas in Institute 2, 13 out of 38(34%) patients of grade IV needed a cochleostomy.

9.CONCLUSION

HRCT Temporal bone and MRI of the inner ear are complementary to each other and provide exquisite vital anatomical details and information. Hence they are now considered as baseline investigations and are mandatory prior to cochlear implant surgery. It also helps in choosing the candidacy for surgery, side selection, and deciding the surgical technique in cochlear implantation.

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ABBREVIATIONS

HRCT	–	High Resolution Computed Tomography
MRI	–	Magnetic Resonance Imaging
SNHL	–	Sensorineural Hearing Loss
CI	–	Cochlear Implant
VA	–	Vestibular Aqueduct
LVA	–	Large Vestibular Aqueduct
LVAS	–	Large Vestibular Aqueductal Syndrome
CSF	–	Cerebrospinal Fluid
SCCD	–	Semicircular Canal Dehiscence
STD	–	Spontaneous Tegmen Defect
IAC	–	Internal Auditory Canal
IAM	–	Internal Auditory Meatus
EAC	–	External Auditory Canal
RW	–	Round Window
RWM	–	Round Window Membrane
FN	–	Facial Nerve
MPFN	–	Mastoid Portion Of Facial Nerve
PT	–	Posterior Tympanotomy
CA	–	Cochlear Aqueduct
SCC	–	Semicircular Canal
IP	–	Incomplete Partition
HJB	–	High Jugular Bulb

PRE-OPERATIVE HIGH RESOLUTION CT &MRI IMAGING FINDINGS IN CHILDREN WITH BILATERAL PROFOUND SENSORYNEURAL HEARING LOSS

CASE PROFORMA

1. Name: 2. Age: years 3.Sex: Male/Female

4. District of origin:

5. Religion: Hindu/Christian/Muslim/Others

6. Education of parents:

Father: Illiterate/Primary schooling/Secondary/High school/

High secondary/Degree/Others

Mother: Illiterate/Primary schooling/Secondary/High school/

High secondary/Degree/Others

7. Occupation of parents:

Father: Unemployed/Unskilled worker/Semiskilled worker/

Skilled worker/business/Professionals/Others

Mother: Home maker/ Unskilled worker/Semiskilled worker/

Skilled worker/business/Professionals/Others

8. Income: /Month

9. Degree and type of hearing loss:

10. Hearing aid use: Duration:

11. Developmental milestones: Normal/delayed

11. HRCT TEMPORAL BONE & MRI BRAIN WITH INTERNAL AUDITORY MEATUS SCREENING-FINDINGS:

EAR STRUTURES		RIGHT	LEFT
Mastoid	Pneumatization		
	Cortical thickness		
Facial recess			
Facialnerve course			
IAM	Size		
	Facial nerve		
	Vestibulocochlear nerve		
Inner ear	Cochlear turns		

	Scala chambers		
	Modiolus		
	Vestibule		
	Semicircular canals		
	Endo/Perilymphatic fluid		
	Bony labyrinth		
	Cochlear aqueduct		
	Vestibular aqueduct		
Middle ear	Ossicles		
	Aeration		
Other findings	Carotid artery path		
	Sigmoid sinus		
	Jugular bulb		
External auditory canal			

Signature of Investigator:

Date:

PATIENT CONSENT FORM

Title of the project:

"PRE-OPERATIVE HIGH RESOLUTION CT&MRI IMAGING FINDINGS IN CHILDREN WITH BILATERAL PROFOUND SENSORYNEURAL HEARING LOSS"

Institution: Upgraded Institute of Otorhinolaryngology,

Madras Medical College & Rajiv Gandhi Govt. General Hospital.

Chennai- 600 003

Name: Date:

Age : IP No:

Sex : Project patient No:

The details of the study have been provided to me in writing and explained to me in my own language.

I confirm that I have understood the above study and had the opportunity to ask questions.

I understood that my participation in the study is voluntary and that I am free to withdraw at any time, without giving any reason, without the medical care that will normally be provided by the hospital being affected.

I agree not to restrict the use of any data or results that arise from this study provided such a use is only for scientific purposes.

I have been given an information sheet giving details of the study.

I fully consent to participate in the above study.

Name of the subject Signature date

Name of the investigator Signature

S.NO	NAME	AGE	SEX	R MASTOID PNEUMATIZATION	L MASTOID PNEUMATIZATION	R FACIAL RECESS	L FACIAL RECESS	R SIGMOID SINUS	L SIGMOID SINUS	R JUGULAR BULB	L JUGULAR BULB	R RW NICHE	L RW NICHE	R COCHLEA	L COCHLEA	R VESTIBULE	L VESTIBULE	R COC AQUEDUCT	L COC AQUEDUCT
1	Fasila	2 yrs	F	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
2	Radhika	2 yrs	F	Absent	Absent	Narrow	Narrow	Anterior	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
3	Alagujeevitha	1.3 yrs	F	Absent	Absent	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Absent	IP-1	Hypoplastic	Dilated	Aplasia	Normal	Normal
4	Keerthasri	2 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
5	Yaswanth	3 yrs	M	Absent	Present	Narrow	Narrow	Anterior	Normal	High	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
6	Jeevitha	2.6 yrs	F	Absent	Present	Narrow	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
7	Karthiga	2 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
8	Rajesh	2 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
9	Shivani	2 yrs	F	Present	Present	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
10	Manikandan	4 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
11	Balaji	5 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
12	Sarathy	2 yrs	M	Absent	Absent	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	IP-1	IP-1	Dilated	Dilated	Normal	Normal
13	Tharun	1.8 yrs	M	Present	Present	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
14	Rishitha	2.6 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
15	Dinesh	1.2 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
16	Megarajan	3 yrs	M	Present	Present	Wide	Wide	Normal	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
17	Santhosh	2 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
18	Pradeep	3 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
19	Thirumalai	5 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	Absent	Absent	michel"s Aplasia	michel"s Aplasia	michel"s Aplasia	michel"s Aplasia	Absent	Absent
20	Priyadharshini	4 yrs	F	Present	Present	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
21	Dhanusri	2 yrs	F	Present	Present	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
22	Durga	2 yrs	F	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
23	Rakshka	2 yrs	F	Absent	Absent	Narrow	Narrow	Anterior	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
24	Thiruvasan	3 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
25	Kathiravan	2 yrs	M	Absent	Absent	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
26	Nusrathbegam	3 yrs	F	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
27	Dhansika	5 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
28	Nishanthi	4 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
29	pugalendhi	2 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
30	Iniya	3.6 yrs	F	Absent	Absent	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
31	Astika	2 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
32	Bhavashree	2 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
33	Vinothini	5 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
34	Prasanna	3 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
35	Manisha	3 yrs	F	Present	Present	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
36	Sasikala	2 yrs	F	Present	Present	Narrow	Narrow	Normal	Normal	Normal	Normal	Absent	Present	michel"s Aplasia	IP-2	michel"s Aplasia	Dilated	Absent	Normal
37	Vignesh	2 yrs	M	Present	Present	Wide	Wide	Anterior	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
38	Evaneswaran	2 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
39	Mounish	2.1 yrs	F	Absent	Absent	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
40	Srinarayana nambi	5 yrs	M	Absent	Present	Narrow	Narrow	Normal	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal

S.NO	NAME	AGE	SEX	R MASTOID PNEUMATIZATION	L MASTOID PNEUMATIZATION	R FACIAL RECESS	L FACIAL RECESS	R SIGMOID SINUS	L SIGMOID SINUS	R JUGULAR BULB	L JUGULAR BULB	R RW NICHE	L RW NICHE	R COCHLEA	L COCHLEA	R VESTIBULE	L VESTIBULE	R COC AQUEDUCT	L COC AQUEDUCT
41	Harsha	3 yrs	F	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
42	Rahul	4 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
43	Prathap	3 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
44	Thara	3 yrs	F	Absent	Absent	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
45	Yogesh	3 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
46	Radhika	4 yrs	F	Absent	Absent	Narrow	Narrow	Anterior	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
47	Sadana	1.6 yrs	F	Absent	Absent	Wide	Wide	Anterior	Anterior	Normal	Normal	present	Present	IP-1	IP-1	Dilated	Dilated	Normal	Normal
N	Srivignesh	2 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	High	High	Absent	Absent	Common cavity	IP-1	Common cavity	Dilated	Normal	Normal
49	Srihari	2 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
50	Sagaya sandana	2 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
51	Kalyana sundaram	2.4 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
52	Om prakash	3 yrs	M	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
53	Mukil	1.6 yrs	M	Absent	Absent	Narrow	Narrow	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
54	Dharani	1.6 yrs	F	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
55	Muthulakshmi	3 yrs	F	Present	Present	Wide	Wide	Anterior	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
56	Harish	3 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
57	Diwakar	4 yrs	M	Present	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	IP-1	Common cavity	Dilated	Common cavity	Normal	Normal
58	Manikandan	4 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
59	Sadana	2 yrs	F	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
60	Moushika	3 yrs	F	Absent	Absent	Narrow	Narrow	Anterior	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
61	Gopika	2.2 yrs	F	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
62	Anbu	2 yrs	M	Present	Present	Wide	Wide	Anterior	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
63	Yashini	2 yrs	F	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
64	Rajina devi	2 yrs	F	Absent	Absent	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
65	Yuvasri	3 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
66	Jagadeesh	2'7 yrs	M	Absent	Absent	Narrow	Narrow	Anterior	Anterior	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
67	Mithra	1 yr	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
68	Shiya sri	2 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Absent	IP-2	Michel"s aplasia	dilated	Michel"s aplsia	Normal	Absent
69	Tamil murugan	5 yrs	M	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal
70	Sophika	5.8 yrs	F	Present	Present	Wide	Wide	Normal	Normal	Normal	Normal	present	Present	Normal	Normal	Normal	Normal	Normal	Normal

S.NO	R VESTI AQUEDUCT	L VESTI AQUEDUCT	R IAM	L IAM	R COC NERVE	L COC NERVE	R LABY OSSIFICANS	L LABY OSSIFICANS	DIAGNOSIS	SURGERY	INSERTION	SCORE
1	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3
2	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	PR Cochleostomy	5
3	Normal	Absent	Normal	Narrow	Hypoplasia	Absent	Absent	Absent	IP-1 R Side,Hypoplastic R Cochlear nerve,Hypoplastic cochlea L side,Aplasia of L Cochlear nerve	R CI	RW Insertion	9
4	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
5	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	L CI	RW Insertion	4
6	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	L CI	RW Insertion	2
7	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
8	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	1
9	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	4
10	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
11	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3
12	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	IP-1 B Sides	R CI	PR Cochleostomy	7
13	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	4
14	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	0
15	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	1
16	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
17	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	1
18	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	PR Cochleostomy	2
19	Absent	Absent	Normal	Normal	Present	Present	_	_	Michel"s aplasia	NOT DONE		_
20	Normal	Normal	Narrow	Narrow	Aplasia	Aplasia	Absent	Absent	B/L COCHLEAR NERVE APLASIA	NOT DONE		_
21	Normal	Normal	Narrow	Narrow	Aplasia	Aplasia	Absent	Absent	B/L COCHLEAR NERVE APLASIA	NOT DONE		_
22	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3
23	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	6
24	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3
25	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	5
26	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	1
27	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	0
28	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
29	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
30	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	PR Cochleostomy	5
31	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
32	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	0
33	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
34	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
35	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
36	Absent	Normal	Normal	Normal	Aplasia	Present	_	Absent	Michel"s aplasia R side,IP-2 L side,cochlear nerve aplasia R side	NOT DONE(not willing)		
37	Normal	Normal	Narrow	Narrow	Aplasia	Aplasia	Absent	Absent	Narrow IAM both side,B/L Cochlear nerve aplasia	NOT DONE		_
38	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3
39	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	5
40	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3

S.NO	R VESTI AQUEDUCT	L VESTI AQUEDUCT	R IAM	L IAM	R COC NERVE	L COC NERVE	R LABY OSSIFICANS	L LABY OSSIFICANS	DIAGNOSIS	SURGERY	INSERTION	SCORE
41	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	1
42	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	1
43	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
44	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3
45	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3
46	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	5
47	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	IP-1 B Sides	R CI	PR Cochleostomy	4
N	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	Common cavity R side,IP-1 L side	planned for CI on L side		8
49	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
50	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
51	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	0
52	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3
53	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	5
54	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	1
55	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	0
56	Enlarged	Enlarged	Normal	Normal	Present	Present	Absent	Absent	B/L Enlarged endolymphatic sac	R CI	RW Insertion	3
57	Normal	Normal	Normal	Normal	Present	Aplasia	Absent	Absent	IP-1 R side,common cavity L side,cochlear nerve aplasia L side	planned for CI on R side		6
58	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	0
59	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	1
60	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	5
61	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	PR Cochleostomy	3
62	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
63	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	3
64	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	1
65	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
66	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	PR Cochleostomy	5
67	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	0
68	Enlarged	Absent	Normal	Narrow	Present	Aplasia	Absent	_	IP-1 R Side,Michel"s aplasia L Side,cochlear nerve aplasia L Side	planned for CI on R side		6
69	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	2
70	Normal	Normal	Normal	Normal	Present	Present	Absent	Absent	NORMAL	R CI	RW Insertion	0

INSTITUTIONAL ETHICS COMMITTEE
MADRAS MEDICAL COLLEGE, CHENNAI 600 003

EC Reg.No.ECR/270/Inst./TN/2013
Telephone No.044 25305301A
Fax: 011 25363970

CERTIFICATE OF APPROVAL

To
Dr.R.Kalaimani
Post Graduate in MS ENT
Upgraded Institute of Oto Rhinolaryngology
Madras Medical College & RGGGH
Chennai 600 003

Dear Dr.R.Kalaimani,

The Institutional Ethics Committee has considered your request and approved your study titled **"PRE-OPERATIVE HIGH RESOLUTION CT & MRI IMAGING FINDINGS IN CHILDREN WITH BILATERAL PROFOUND SENSORYNEURAL HEARING LOSS" NO. 24122016.**

The following members of Ethics Committee were present in the meeting hold on **14.12.2016** conducted at Madras Medical College, Chennai 3

1.Dr.C.Rajendran, MD.,	:Chairperson
2.Dr.M.K.Muralidharan,MS.,M.Ch.,Dean, MMC,Ch-3	:Deputy Chairperson
3.Prof.Sudha Seshayyan,MD., Vice Principal,MMC,Ch-3	: Member Secretary
4.Prof.B.Vasanthi,MD., Prof.of Pharmacology.,MMC,Ch-3	: Member
5.Prof.A.Rajendran,MS, Prof. of Surgery,MMC,Ch-3	: Member
6.Prof.N.Gopalakrishnan,MD,Director,Inst.of Nephrology,MMC,Ch	: Member
7.Prof.Baby Vasumathi,MD.,Director, Inst. of O & G	: Member
8.Prof.K.Ramadevi,MD.,Director,Inst.of Bio-Che,MMC,Ch-3	: Member
9.Prof.R.Padmavathy, MD, Director,Inst.of Pathology,MMC,Ch-3	: Member
10.Prof.S.Mayilvahanan,MD,Director, Inst. of Int.Med,MMC, Ch-3	: Member
11.Tmt.J.Rajalakshmi, JAO,MMC, Ch-3	: Lay Person
12.Thiru S.Govindasamy, BA.,BL,High Court,Chennai	: Lawyer
13.Tmt.Arnold Saulina, MA.,MSW.,	:Social Scientist

We approve the proposal to be conducted in its presented form.

The Institutional Ethics Committee expects to be informed about the progress of the study and SAE occurring in the course of the study, any changes in the protocol and patients information/informed consent and asks to be provided a copy of the final report.

24.12.16

Member Secretary – Ethics Committee

MEMBER SECRETARY
INSTITUTIONAL ETHICS COMMITTEE
MADRAS MEDICAL COLLEGE
CHENNAI-600 003

PLAGIARISM CERTIFICATE

URKUND

Urkund Analysis Result

Analysed Document: Kalaimani FINAL DISSERTATION - Copy.docx (D31546089)
Submitted: 10/22/2017 7:48:00 PM
Submitted By: drkalainkl@gmail.com
Significance: 12 %

Sources included in the report:

thesis.docx (D30836726)
https://clinicalgate.com/congenital-malformations-of-the-inner-ear/
http://famona.tripod.com/ent/cummings/cumm152.pdf
https://entokey.com/inner-ear-malformations-and-implantation/
https://www.emaze.com/@AZWLCTWO/inner-ear-malformations?_escaped_fragment_=

Instances where selected sources appear:

50

sensorineural hearing loss where the pathology is generally in the cochlea or retrocochlear region(In the 8th nerve or brain)2. In cases of bilateral SNHL, the radiological imaging may be abnormal in 6.8% to 12.8% and in up to 30% of children3. It is vital to obtain a good quality high resolution CT and MRI of temporal bone in patients with SNHL with accurate technical parameters. Most centers are now equipped with multi slice spiral CT scanners, which is essential as reconstructions can be obtained in any plane with isotropic voxels in high end CT scanners without loss of resolution. MRI should be obtained on a high tesla strength magnet, 1T (tesla) or higher. In our institute, 64 slice spiral CT and 1.5T MRI machine are used2.

AIMS AND OBJECTIVES 1. To pre-operatively evaluate the cochlea-vestibular anomalies, middle ear abnormalities, cochlear duct patency, presence of cochlear nerve, course of facial nerve and other abnormalities 2. To know the type of pneumatization of the mastoid

3. Finally to make surgical decisions regarding candidacy for surgery, side selection and surgical technique in cochlear implantation.

INCLUSION CRITERIA:

• Children aged between one to six years • Bilateral profound sensorineural hearing loss with <90dB in better ear • No appreciable benefit with digital hearing aid

• Motivated parents for surgery

• No medical contraindications

EXCLUSION CRITERIA: • Children aged more than six years • Benefit with digital hearing aid

• Parents not willing for cochlear implant surgery

REVIEW OF THE LITERATURE

Robert K.Jackler et al (1987) explained the incidence of congenital inner ear malformations. 80 to 90 % of the inner ear anomaliesare limited to the membranous labyrinth .10 to 20 % of inner ear anomalies are associated with radiographically detectable malformations, in which cochlear malformations are most common. But many cases demonstrate abnormalities in more than one portion of the inner ear4.