IMAGING STUDY & SURGICAL PERSPECTIVE OF COCHLEAR IMPLANTEES

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Abstract

Introduction: *A cross-sectional prospective clinical study was done to evaluate the inner ear anomalies in candidates undergoing cochlear implantation.*

Materials and Methods: *Study was carried out over a period of 07 years (Jan 2013 to Dec 2019) at Cochlear Implant Centre, Combined Military Hospital Dhaka. A total 206 deaf candidates, both pre and post lingual, were included in convenient sampling method.*

Result: *The sample consisted of 206 candidates, of whom 117 (56.80%) were female and 89 (43.20%) were male. All candidates had severe to profound bilateral deaf-ness. 182 (88.35%) of them were pre and 24 (11.65%) were in post lingual group.*

All patients diagnosed audiologically as profound deafness underwent HRCT scan of temporal bone with 3D reconstruction of cochlea and MRI of internal auditory canal & brain. Radiological examination revealed that 36 (17.47%) candidates had ear anomalies. Mondini deformity 14 (06.80%), cochlear ossification 11 (5.34%), large vestibular aqueduct 04 (1.94%), high jugular bulb 04 (1.94%) and anteriorly placed sigmoid sinus 03 (1.45%) were noted.

During intervention, 42 (20.39%) candidates had anomalous intra operative findings. Among them CSF gusher 19 (9.23%), ossification of basal turn 12 (5.83%), isolated rotated Cochlea 02 (0.97%), high jugular bulb 04 (1.94%), anteriorly placed sigmoid sinus 03 (1.45%), and very high facial nerve 02 (0.97%) were observed.

During surgery, electrode insertion difficulty due to high CSF gusher was experienced in19 (9.23%) cases, full length of electrode insertion was not possible due to ossified basal turn 12 (5.83%), difficult to find out round window membrane due to rotated cochlea 02 (0.97%), prevented direct visualization of the round window due to high jugular bulb 04 (1.94%), difficult to approach round window due to very high facial nerve 02 (0.97%). and approach to round window membrane was compromised due to anteriorly placed sigmoid sinus 03 (1.45%). Most of these difficulties were effectively managed during surgery.

Conclusions: *This study highlights the importance of preoperative radiological scanning in the assessment of patients undergoing cochlear implantation. It provides vital information on cochlear status and in ruling out non cochlear causes where cochlear implantation is not feasible. Detection of anatomical abnormalities with appropriate evaluation, specially imaging should be mandatory in every patient undergoing cochlear implantation.*

Key words: *Cochlear Implant, Mondini dysplasia, Vestibular aqueduct.*

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Introduction:

Background: Hearing is an essential sense for development of speech which is crucial for verbal communication and personality development. Hearing impairment is one of the most frequent sensory deficits in human beings and it is the second commonest form of disability in Bangladesh producing substantial social and economic costs because of its effect on child health development and education.[1]

Cochlear implantation has now become established Worldwide as an effective means of hearing rehabilitation in severe to profound deaf individuals. As the indications of Cochlear implantation have expanded, the Cochlear implant surgeon faces challenging situations. Anatomical abnormality can pose a formidable challenge during surgery. In multi-handicapped children, surgery and rehabilitation can be extremely difficult. In patient with congenital hearing loss the anatomical abnormalities are estimated as 20%.[2,3] According to present literature, IEMs are classified into eight distinct groups. 1. Complete Labyrinthine Aplasia (Michel Deformity) 2. Rudimentary Otocyst, 3. Cochlear aplasia, 4. common cavity, 5. Cochlear hypoplasia, 6. Incomplete partition of the cochlea, 7. Enlarged vestibular aqueduct, 8. Cochlear aperture abnormalities.[4,5,6]

Most common congenital anatomical abnormalities that needs to be addressed prior cochlear implantation include Mondini's deformity, Large vestibular aqueduct syndrome, narrow internal auditory canal or hypoplasia of eighth cranial nerve, common cavity deformity, anomalous course of facial nerve and common acquired anatomical abnormalities include post meningitis labyrinthine ossificans, otosclerotic involvement of cochlea, post mastoidectomy cavity.[7]

Mondini's deformity accounts for 55% of all inner ear malformation and 5% of cause of sensorineural hearing loss unassociated with any syndrome. Mondini's deformity is described by Carlo Mondini in 1791, is characterized by a normal basal turn, middle and apical turns being replaced by cyst like cavity. The cochlea develops only 1.5 turns and partial or complete lack of interscalar septum and modiolar hypoplasia occurs. Normal cochlea measures about 8-10 mm vertically but in Mondini's the cochlea measures 5-6 mm. Development of organ of corti and auditory neural population is variable. Normal basilar development is the key to an imaging diagnosis. It is associated with semicircular canal, vestibule, and endolymphatic duct / sac deformity in 20% of cases [7]. Patient with the Mondini's deformity are at increased risk of developing recurrent meningitis or perilymphatic fistula as these patient often have associated with large vestibular aqueduct as well as large endolymphatic duct or sac. Hence, in any child with sensorineural deafness or two unexplained attack of meningitis, a CT scan study of temporal bone is mandatory. CT axial and coronal views may shows the cystic appearing cochlea with absence of modiolas / osseous spiral lamina usually present at the level of the apical and middle turns of the cochlea. Magnified coronal CT image at the level of vestibule shows the presence of normal basal turn. MRI with high resolution T2 weighted image (fast spine echo) shows an incomplete partition deformity with the absence of intra cochlear septum.[1, 3, 10]

The normal **vestibular aqueduct** is a bony canal extending from the medial wall of the vestibule to the posterior cranial fossa. It houses the endolymphatic duct, a small artery and vein. Normal diameter is 0.4 t0 1 mm. It is considered large when it measures greater than 1.5 mm in width at the midpoint between the common crus and the external aperture. Typically it is bilateral in 55-94% of cases. Children are usually born with normal or mild impaired hearing that gradually deteriorates over a period of years to profound sensorineural hearing loss. Radiological technique for assessing an enlarged vestibular aqueduct include a high resolution CT axial view at and above the level of internal auditory canal. MRI demonstrate dilated endolymphatic duct and sac.[7,10]

Narrow internal auditory canal may indicate a failure of eighth nerve development when a patient has normal facial function and an axial CT scan shows an internal auditory canal less than 2 to 2.5 mm or less in diameter, it is likely

that the bony canal transmits only the facial nerve. In such cases cochlear implantation is contraindicated. Again a hypoplastic internal auditory canal can contain the vestibulocochlear nerve with abnormal function been considered as a relative contraindication to cochlear implantation.[1, 3]

Anomalous facial nerve course usually associated with other congenital external and middle ear abnormalities. In congenital atresia, in 20% of these patient, the facial nerve will have an abnormal course and is therefore liable to surgical damage. To avoid such surgical complications, early identification of facial nerve is mandatory. The most abnormal anomalous course is in the anterior epitympanum, when the nerve enters the tympanic cavity. Congenital dehiscence of fallopian canal in the horizontal portion of facial nerve may occur. The nerve may displaced inferiorly to encroach on the stapes and the oval window. Rarely the nerve lies below the stapes. In approximately 15%, the descending portion of the facial nerve is displaced laterally and anteriorly. Anomaly is commoner in the mastoid segment than in the tympanic segment.[10]

Labyrinthine ossificans is a typical finding in deafness due to meningitis especially bacterial but may occur after trauma, chronic otitis media, labyrinthitis, otosclerosis, syphilis, autoimmune disease. It starts 4-8 weeks after meningitis and may continue upto 30 years. The incidence of hearing loss after meningitis has been reported as 5-35% .Ossification is nearly always most severe in the region of the round window and proximal scala tympani in the basal turn, adjacent to the opening of the cochlear duct. The middle and apical turns are less commonly affected, and the scala vestibule is often spared. If profound hearing loss occurs after meningitis, implantation is advised as early as possible. If residual hearing is present, radiological monitoring for 6-9 months is recommended. Complete obstruction may occur, more commonly in children and adults and has been reported in 3%.The CT Scan appearance of Labyrinthine ossification is that of diffuse or localized to basilar turn of the cochlea or round window niche, unilateral if tympanogenic and bilateral on meningogenic.[1,3,7]

Preoperative identifications of these abnormalities is crucial. In that perspect imaging of the temporal bone (CT scan / MRI) is mandatory prior to cochlear implantation to help the surgeon anticipate possible intraoperative problems.[12]

Materials and methods

A cross sectional prospective clinical study underwent at Cochlear implant center, CMH Dhaka over a period of 07 years from 1st Jan 2013 to 31st Dec 2019. A total 206 Cochlear implant candidates who reported with severe to profound deafness were included in this study. Pre lingual candidates below 06 years and post lingual deaf who lost their hearing within 10 years were included in this study group. All syndromic patients were excluded from this study.

Methods and data collection procedure

This is a hospital based cross sectional study on 206 patients. Samples were selected by convenient method. Hearing loss was confirmed by clinical and audiological investigations.

Assessment of patient includes the general & ENT examination, and any syndromic signs. Investigations includes the audiological assessment e.g. Behaviour Observation Audiometry (BOA), Tympanometry, SRT, Brainstem Evoked Response Audiometry (BERA), Auditory Steady State Response (ASSR), Otoacoustic Emissions (OAE), Aided audiogram, speech and language evaluation, CAEP etc. Patients having bilateral severe to profound sensorineural hearing loss were included in study group.

All candidates for possible cochlear implantation, underwent HRCT of temporal bone and MRI of internal acoustic canal & brain. CT scan were performed in the axial orientation using multi slice light speed with a slice thickness of 0.625mm. Coronal and sagittal reconstructions were performed. All MRI scans were performed on a 1.5 TESLA MRI scanner using an 8-channel head coil and heavily T2 weighted sequence. To reduce motion artifacts, the children were studied under sedation. All

printed CT and MRI were evaluated by Cochlear Implant surgeon & a senior radiologist.

All candidates underwent cochlear implantation, post aural incision with an anteriorly/superiorly based Palva musculoperiosteal flap was raiised. A classical cortical mastoidectomy was done. Implant wells created and tie down holes/subperiosteal pocket was made. Standard posterior tympanotomy done and the round window niche identified. Finally electrode was inserted through round window approach or a limited cochleostomy in some cases. Intra op telemetry was done in all cases.

Data analysis

Data for socio-demographic and clinical variables were obtained from all participants by the use of a pre- designed and easily understandable questionnaire. After collection of all data, these data were checked, verified for consistency and edited for finalized result. After editing and coding, the coded data directly entered into the computer by using SPSS version 16. Data cleaning validation and analysis was performed using the SPSS/PC software and graph and chart by MS excel. The result was presented in tables in proportion.

Quality assurance strategy

The proper data collection procedure & for maintaining good quality a work manual was made. Then a sample size was selected & a standard questionnaire was made. The questionnaire was pretested & ensured that the respondents are able to understand the questionnaire and answered accordingly.

Ethical measures

The study protocol and consent form were approved by the ethical committee of Combined Military Hospital, Dhaka. Risk, benefits and potential complications were explained to all the patients/ guardian and written informed consents were obtained prior to data collection. They had the freedom to withdraw from the study at any time. Collected all data were checked very carefully to identify any error. Data processing work consisted of registration of schedules, editing, coding and computerization, preparation of tables, analysis and matching were done by researcher.

Observation and Results

Table I

Age distributions of the cochlear implant candidates (n=206)

Age Group (Years)	No of Patient	Percentage
Prelingual	182	88.35%
01-03 (123)		
04-06 (59)		
Postlingual adult	24	11.65%
Total	206	100 %

The highest number of candidates selected were below 03 years age group.

Table II

Hearing status of the cochlear implant candidates (n=206)

Degree of Hearing loss	No of Patient	Percentage
Severe to profound	18	08.74 %
Profound	188	91.26 %
Total	206	100 %

The series shows that 188 (91.26%) had been suffering from profound deafness. Remaining 18 (8.74%) were severe to profound deaf.

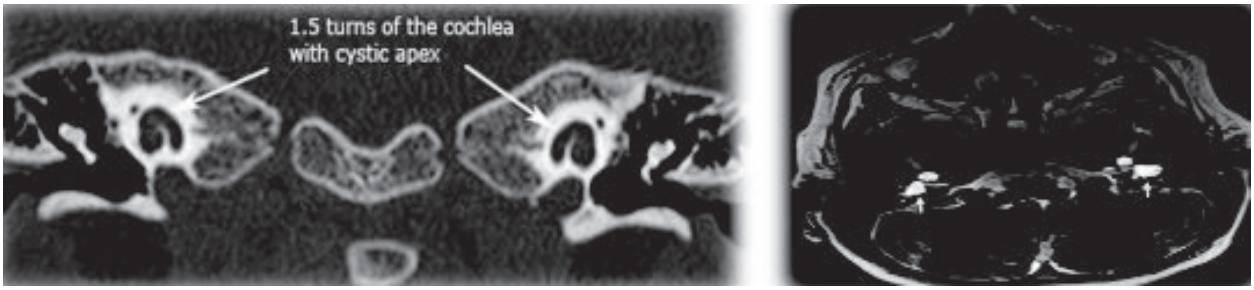

Fig.- *Mondini deformity*

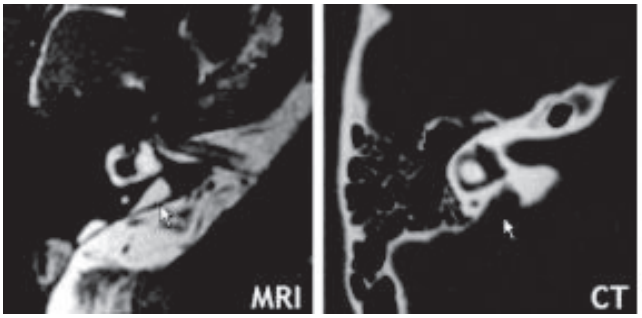

Fig: *LVAS*

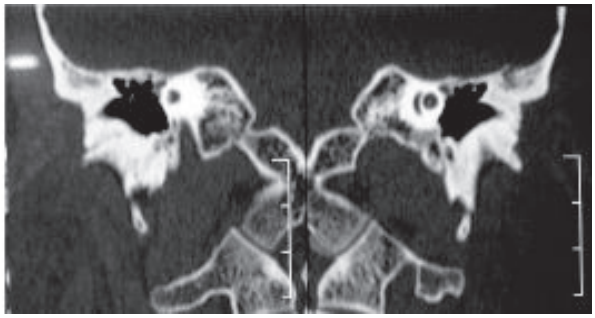

Fig: *Ossified cochlea*

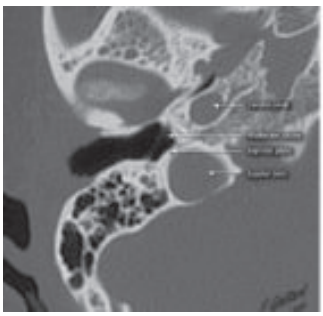

Fig: *High jugular bulb*

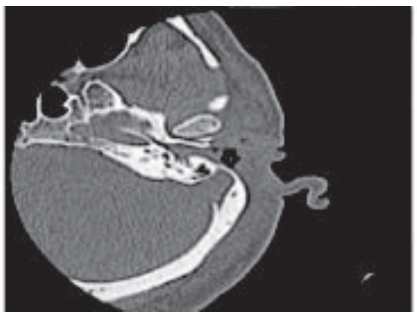

Fig: *Anteriorly placed sigmoid sinus*

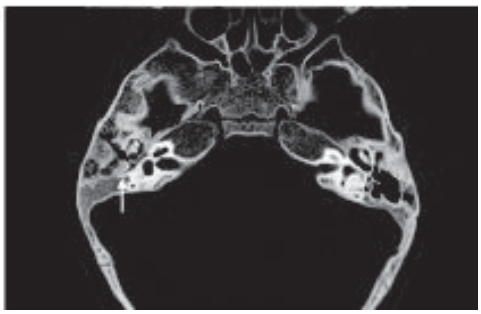

Fig: *High facial nerve*

Table III

Radiological findings among cochlear implant candidates (n=206)

Imaging temporal bone	No of Patient	Percentage
Anomalous	36	17.47 %
Mondini deformity (14) Ossified cochlea (11) -Post meningitis (08) -Otosclerosis(03) Large vestibular aqueduct (04) High jugular bulb (04) Anterior sigmoid sinus (03)		
Normal	170	82.53 %
Total	206	100 %

Anomalous radiological findings were present in 36 (17.47%) cases.

Table IV

Intra-operative findings and difficulties encountered during implantation (n=206)

Intra-operative finding	Intra-operative difficulties	No of Patient	Percentage
CSF gusher	Electrode insertion	19	09.23%
Ossification of basal turn	Full length of electrode insertion was not possible.	12	05.83%
Isolated rotated cochlea	To find out round window membrane.	02	0.97%
High jugular bulb	Prevented direct visualization of the round window.	04	1.94 %
Anteriorly placed sigmoid sinus	Approach to round window membrane was compromised.	03	1.45 %
High facial nerve	Difficult to approach the RW,needs cochleoetomy	02	0.97 %
Normal findings	No difficulties	164	79.61 %
Total	206	100 %	

Table shows 42 (20.39%) cases anomalous intra operative findings with difficulties encountered during surgery.

Table V

Number of electrode insertion among the candidates (n=206)

Electrode insertion	No of Patient	Percentage
Complete	194	94.17 %
Incomplete		
11 pair out of 12 pair(9)		
10 pair out of 12 pair(3)	12	05.83 %
Total	206	100 %

Table revealed 12 (05.83%) cases electrode insertion was incomplete.

Result:

The sample consisted of 206 Cochlear Implant candidates, of them 182 (88.35%) were prelingual and 24 (11.65%) were post lingual group. All patient diagnosed audiologically as severe to profound deafness underwent HRCT scan of temporal bone and MRI of internal auditory canal & brain. Radiological examination revealed 36 (17.47%) out of 206 (100%) candidates had inner ear anomalies. Mondini deformity 14 (06.80%), cochlear ossification 11 (05.34%), large vestibular aqueduct 04 (1.94%), high jugular bulb 04 (1.94%) and anteriorly placed sigmoid sinus 03 (1.45%) were noted.

During intervention, 42 (20.39%) out of 206(100%) candidates had anomalous intra operative findings. Electrode insertion difficulty due to high CSF gusher 19 (09.23%) of which 14 (06.80%) cases were Mondini deformity and another 04 (01.94%) were large vestibular aqueduct. Excessive CSF expression was experienced during intervention in 01 case which was missed to diagnose radiologically prior to surgery. Full length of electrode insertion was not possible due to ossified basal turn in 12 (05.83%) cases of which 01 case developed recent ossification undiagnosed during pre op imaging. Difficult to find out round window membrane due to rotated cochlea was observed in 02 (0.97%) patients, those needed high cochleostomy. Direct visualization of the round window membrane was prevented due to high jugular bulb 04 (01.94%), difficult to approach round window due to very high facial nerve 02 (0.97%). and approach to round window membrane was compromised due to anteriorly placed sigmoid sinus in 03 (1.45%) case. Most of these difficulties were effectively managed during surgery.

Discussion:

A prospective clinical study was conducted at the Cochlear Implant Centre, Combined Military Hospital Dhaka. All Prelingual deaf before 06 years of age and Postlingual deaf who lost their hearing within 10 years were enrolled for study. Detailed history, clinical examination and audiological evaluation was carried out meticulously. HRCT of temporal bone to see the cochlear architecture and to exclude inner ear malformations and MRI of internal auditory canal & brain to see the viability and dimensions of cochlear nerve & exclusion of intracranial lesions are routinely practice in our center.

Present study illustrates, imaging findings in patients with SNHL and focused all anomalies of the inner ear .Most of the inner ear anomalies detected by imaging study was Mondini deformity, labyrinthine ossification, large vestibular aqueduct, hypoplastic internal acoustic canal (Excluded from our study) high jugular bulb and anteriorly placed sigmoid sinus. Among them, the most common abnormality was Mondini's deformity 14 (06.80%). In a study by Woodford TJ, Roberts GR et al. of computed tomography findings in 105 patients, 2 cases of Mondini deformity were bilateral. Phelps claims it is usually unilateral, while Jackler et al, found a Mondini deformity in 41 ears of 25 patients.[2, 15] In patient with Mondini deformity & LVAS formed electrode was inserted through the round window approach. The round window was sealed with fascial graft and muscle supported by tissue glue.

Johannes P. Westerhof et al., studied congenital malformations of the inner ear and the vestibulo-cochlear nerve in children with SNHL. Twenty one children (42 inner ears) were studied with high resolution MR and helical CT examinations who were candidates for cochlear implants. They identified 99 malformations. Mondini abnormality and Mondini variants were 12 out of 42.[14]

Another study by Han DM et al. among 300 patients who received multi-channel cochlear implants from 1996 to 2002 in Beijing Tongren Hospital, 15 patients were diagnosed with Mondini malformation. A retrospective analysis was performed dealing with the surgical techniques. Gusher was found more common than the normal cochlear implantation, most of them were serious. The electrodes was inserted in the "cochleostomy" in full length of 13 Patients, 2 pairs of electrodes remains outside of "cochleostomy" in 2 patients. They concluded, Multi-channel cochlear implantation could be performed safely in patients with Mondini malformation. The primary outcome for patients with Mondini malformation are similar to those with normal cochlear structure following the multi-channel cochlear implantation.

In present study, 12 (05.83%) cases of labyrinthine ossificans were observed. In a study of ossified cochlea by Thomas J Balkany, Philip Bird et al. the result and degree of ossification revealed that obstruction of the fluid space of the cochlea is not uncommon in candidates for cochlear implantation. Previously it was thought to be a contraindication for cochlear implantation. Of 235 patient underwent implantation, 22(9%) had some degree of ossification. They reported that children and adults with partially ossified cochleas underwent complete electrode insertion and hearing results similar to those with normal cochleas. Patient with totally ossified cochleas achived some open-set word recognition, however insertion depths and word recognition scores were reduced in comparison with those who had partially or nonossified cochleas. Balkany et al. reported 14% of abnormal bone formation within the scala tympani in their 135 patients with implants.[15,16] Luxford and House identified ossification in 34% of their initial 128 children with implants· Bath AP et al. predicted the patency of the cochlea in children undergoing cochlear implantation by using HRCT scans. High degree of accuracy (87%) in predicting a patent cochlea was achieved by CT in patients who had normal inner ears. Degree of ossification was considerably under-estimated (15% of cases) causing major difficulties at the time of surgery. Even though HRCT provides great clarity and fine detail, it has its limitations.

All cases of ossificans were detected by HRCT scan of temporal bone. Among 08 (03.88%) developed deafness following meningitis and 03 (01.45%) following otosclerosis. During intervention ossified cochlea was noticed in a pre-lingual deaf child who had recent history of viral fever. Extended cochleostomy was done and ossified spongy bone was drilled off from the scala tympani. Near total insertion was achieved with 11 pair out of 12 pair in 09 cases and 10 pair out of 12 pair in 03 cases

Gibson WPR, Brown C et al reported on their study of 100 children who underwent implantation. 32 become profoundly deaf as a result of meningitis. Hemophilus influenza type B and streptococcus pneumonia were the main causes of meningitis in the majority of cases.10 of the 32 children who had meningitis had ossification of cochlea. Of these 10 children, 3 had scala vestibule insertion, 3 had partial insertion, and 5 had inlay, drill-out insertion.

In our study, 04 (1.94%) candidate presented with congenital bilateral profound hearing loss who's imaging (CT, MRI) revealed bilateral large vestibular aqueduct. In a study by Slattery WH, Luxford WM of 10 patients with congenital inner ear malformations, 2 had enlarged vestibular aqueducts. A cerebrospinal fluid leak occurred in 4 of the 10 cases.

Study by Bamiou DE et al. computed tomography findings in bilateral SNHL. They studied only with CT in comparison to our study, where both CT and MRI were used for evaluation. Dilated vestibular aqueduct was the most common CT abnormality in their series (10 of 116 cases) [16]. Okumura et. al., identified 13(7.18%) patients with LVA out of a group of 181 who had sensorineural hearing loss (SNHL) of unknown etiology. Half of the 13 patients with LVA had experienced sudden hearing loss. Similarly, Callison and Horn found a 5% prevalence of LVA in their clinical population. LVA is generally bilateral and almost always leads to some degree of progressive or fluctuating hearing loss. Hearing loss is often reported following head injury (Smith & Van

Camp, 2006). According to the literature, LVA is the most common malformation of the inner ear associated with SNHL. It can occur in isolation or in combination with other malformations of the cochlea.[15, 17]

Another study by Claros Pedro et al. found 11 patients (1.3%), six boys and five girls with EVA who underwent cochlear implant surgery in there center during the 22-year study period, out of a total of 827 implanted. EVA was bilateral in 10 cases, isolated anatomical finding in 7 cases, and associated with other malformations in 4 cases. Cochlear implantation was bilateral in 5 cases and unilateral in 6 cases.[18]

Giovanni Bianchin et al, reported 278 consecutive cochlear implant recipients, including children and adults and evaluated between January2000 to December 2015 and found eight patients with enlarged vestibular aqueduct. Cerebrospinal fluid gusher occurs in five subjects with enlarged vestibular aqueduct.[19,20]

In our study, 02 (0.97%) candidates had an anomalous high facial nerve. In a study by Raine CH, Hussain SSM. Out of 42 implant surgeries, 5 cases had facial nerve anomaly which prevented direct visualization of the round window. In 4 cases, the nerve was anterior and 1 case, there was an obvious congenital bifurcation. Facial nerve monitoring was done using Neurosign 100. In 4 cases, where the nerve was anterior, a curve probe was used to locate the round window and the cochleostomy was performed. In the 5th case, where the nerve was bifurcate, the nerve was decompressed and mobilized and then the cochleostomy was performed.[14]In our study, lumen of cochlea was not encountered at the usual site in 02 (0.97%) cases who had normal cochlea in radiology, those were diagnosed as rotated cochlea during intervention. A study by Muhaimeed H.A.S. on 12 patients in which difficult cochleostomy was faced intraoperatively in 1 patient (number 2), whose preoperatively measured BTA was 65° (more obtuse than the normal range), predicting a rotated cochlea with difficult cochleostomy.[16] They concluded that the possibility of a rotated cochlea should be borne in mind when difficult cochleostomy is encountered.

Another study by Mikhail et al. on 25 patients that underwent cochlear implantation at Tanta Cochlear Implant Centre, Otolaryngology, Head & Neck surgery department, Tanta University, that had been done from April 2015 to December 2015. All cases had cochlear implantation using standard trans-mastoid technique. Following review of the computed tomography images of the two patients in whom cochlear implant was difficult. It appeared that cochlea is rotated that means the orientation of the basal turn of the cochlea relative to the midline was more obtuse or more acute than in other patients.[14]

Conclusion:

Cochlear implantation is an established method of rehabilitation for bilateral severe to profound sensorineural hearing loss unresponsive to most powerful conventional amplification. As the criteria for implantation have expanded, challenging situations are being increasingly encountered. The ideal imaging algorithm in children with bilateral profound SNHL still appears to be contentious. Neither CT nor MRI of the brain and temporal bones appears to be adequate as the sole imaging modality of choice in all children.

But using both MRI and HRCT modalities judiciously in selected cases can fill each other's lacunae. The two modalities in combination can provide the cochlear implant surgeon the information which is surgically relevant. This is especially true in congenital hearing loss. A good number of patient undergoing cochlear implantation have been reported to have anatomical abnormalities. The presence of such abnormalities have a significant effect on surgical technique employed and is associated with higher risk of complications also adversely affect the hearing outcome. Thus, it helps surgeon to choose best ear for cochlear implantation. So based on our study, both modalities are preferred preoperatively in cochlear implant patients rather than single modality.

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All data and material are available.

Competing interests:
All of the authors declare that they have no competing interest.

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