

Preoperative dual imaging evaluation of profound sensorineural hearing loss in patients for cochlear implantation

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Abstract

Background: Profound sensorineural hearing loss (SNHL) may be the result of major inner ear structural malformations, and cochlear implantation remains the only viable treatment option. High-resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) are indispensable for optimum preoperative implant workup and thus play a vital role in patient selection, pre-implantation counseling, and surgical management.

Aim and Objectives: The aim of this study is to evaluate patients with profound SNHL for cochlear implantation preoperatively on both HRCT and MRI and to compare imaging findings in both modalities.

Materials and Methods: This longitudinal prospective study was conducted in the Department of Radiology of a tertiary care-based hospital in North India. A total of 45 patients (90 temporal bones) with clinically diagnosed bilateral profound SNHL were included in the study. Patients with a previous history of temporal bone injury were excluded from the study. All cases were evaluated on both 128 slice Philips computed tomography (CT) machine and 1.5 Tesla Siemens Magnetom MRI scanner. Each temporal bone was systematically analyzed for anatomical and structural abnormalities.

Results: Both high-resolution CT and MRI played vital roles in the workup of patients with profound SNHL for cochlear implantation and allowed accurate assessment of critical inner ear abnormalities. Cochlear malformations (30%) were responsible for the majority of structural abnormalities in this study with Type II incomplete partition (8.9%) being the most common. Cochlear nerve deficiency was seen in 20 cases (22.2%) and was diagnosed only on MRI. Similarly, early fibrosis and abnormal signal intensity were also detected only on MRI, which were missed on CT.

Conclusions: Both high-resolution CT and high magnet MRI complement each other and reduce the chances of missing critical findings, which are crucial for surgical management and planning. Thus, it is advisable to perform dual imaging with both modalities wherever and whenever possible, to offer maximum information to treating surgeon preoperatively.

Keywords: Cochlear nerve, computed tomography, magnetic resonance imaging, sensorineural hearing loss

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INTRODUCTION

Profound sensorineural hearing loss (SNHL) may result from aberrance or arrest in the development of cochlear apparatus and/or eight nerve or due to abnormalities in the higher auditory centers of the brain. The cause may be hereditary, genetic, posttraumatic, post-infectious, or idiopathic.^[1-5]

At present, cochlear implantation is the only viable treatment option available for patients with profound SNHL in both ears who failed to benefit from hearing aids. Preoperative imaging of temporal bone is mandatory in such cases to rule out major structural malformations and thus plays a vital role in decision making, surgical planning, and prognosis.^[3,4,6,7]

Although imaging in the majority of such patients may reveal normal findings, with the advent of high-end sophisticated equipment the increasing numbers of abnormalities are being detected. Both high-resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) can be performed in these patients to assess complex inner ear anatomy. Conventionally, computed tomography (CT) has been the imaging modality of choice for the initial workup of patients with bilateral SNHL, but recently, the utility of MRI is also being increasingly acknowledged.^[1,3,4,8-10]

Both these imaging modalities have their own merits and demerits. HRCT with thin slices, and bony algorithm superbly depicts intricate osseous anatomy, middle ear aeration, and mastoid pneumatization, which are poorly demonstrated on MRI. MRI on other hand, allows the excellent depiction of cisternal and meatal segments of the eight nerve (vestibulocochlear nerve) and fluid-filled membranous labyrinth, which cannot be reliably assessed on CT.^[1,3,4,9,11,12]

Preoperative diagnostic workup for cochlear implant candidates varies from institute to institute with no clear cut consensus on imaging protocols. Some centers routinely perform dual imaging with both CT and MRI in such patients, while others prefer either CT or MRI alone depending on patient’s affordability and availability of equipment.^[3,11,12]

With this background, the present study was undertaken to evaluate patients with profound SNHL for cochlear implantation preoperatively on both HRCT and MRI. We also compared imaging findings in both modalities and tried to find out the better modality among the two.

MATERIALS AND METHODS

This longitudinal prospective study was conducted in the Department of Radiology of a tertiary care-based hospital in North India. Institutional Ethical research approval was taken prior to the commencement of the study. Before enrollment, written and informed consent were obtained from all the patients or their guardians (in case of minor). Patients of all ages and gender were included in this study. A total of 45 patients referred from the otolaryngology department of the hospital with clinically diagnosed bilateral profound SNHL were enrolled. Detailed clinical history and examination were performed in all cases. Patients with a previous history of temporal bone injury were excluded from the study.

All cases were evaluated with both 128 slice Philips CT machine and 1.5 Tesla Siemens Magnetom MRI scanner. Sedation was required in the majority of pediatric patients to avoid motion artifacts and to obtain high-quality diagnostic scans. In sedated patients, MRI was performed first, followed later by CT.

Three-dimensional (3D) constructive interface steady state sequence in axial plane was used for demonstration of cisternal segment of the eighth nerve and in the oblique sagittal plane perpendicular to internal acoustic meatus for optimal demonstration of “four dot” sign of all the nerve bundles in internal auditory canal (IAC) (facial nerve anterosuperior, cochlear nerve antero-inferiorly, superior and inferior divisions of vestibular nerve posteriorly). In addition, 3D maximum intensity projection reconstruction was done for the evaluation of cochlea [Figure 1]. A screening section of the brain with an axial T2-weighted image (T2WI) (TR 4000 ms, TE 80 ms) was also taken.

Various parts of each temporal bone (cochlea, vestibule, semicircular canals, vestibular and cochlear aqueducts,

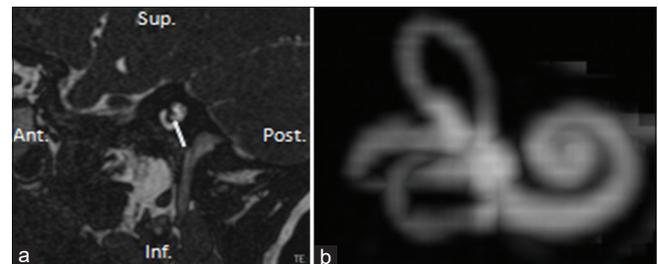


Figure 1: Normal eight nerve and fluid filled membranous labyrinth on high resolution magnetic resonance imaging (a) oblique sagittal three-dimensional constructive interface steady state images show normal four dot sign of nerve bundles in internal auditory canal with facial nerve antero-superiorly, cochlear nerve antero-inferiorly (white arrow), superior and inferior divisions of vestibular nerve posteriorly (b) magnetic resonance maximum intensity projection image shows normal cochlea, vestibule, and semicircular canals

cochlear nerve, and IAC) were systematically evaluated on both CT and MRI scans and findings were recorded in predesigned proforma. The inner ear malformations were categorized as per the classification proposed by Jackler *et al.*^[2] and Sennaroglu and Saatci.^[13] Incidental findings of various important anatomical variations (high riding/dehiscent jugular bulb), the status of the middle ear, and mastoid aeration were also recorded.

RESULTS

A total of 90 temporal bones of 45 patients were evaluated in this study.

The study population included patients ranging from the age of 2 months to 18 years with a mean age 3.6 years. Majority of patients belonged to the pediatric age group, with 40 (89%) of patients below the age of 5 years. Twenty-six (58%) of the patients were males and the remaining (42%) were females.

Comparative analysis of various malformations on both HRCT and MRI is shown in Tables 1-3.

Cochlear malformations were the overall most common structural abnormalities (27, 30%) detected in our study. Type II incomplete partition (Mondini’s malformation) constituted the most common (8, 8.9%) cochlear pathology [Figure 2]. Two cases with nonspecific sclerosis and 1 case of late stage Labyrinthitis ossificans were diagnosed only on CT. 1 case of fibrotic Labyrinthitis ossificans, and 4 cases with abnormal signal intensity were detected only on MRI [Table 1]. We also found two cases with Michel’s aplasia [Figure 3], four cases with common cavity malformation [Figure 4] and 5 cases with type 1 incomplete partitioning [Figures 5 and 6].

Thirteen (14.4%) cases on CT and 15 (16.6%) cases on MRI had vestibular abnormalities. Both of the modalities detected an almost equal number of abnormalities with few exceptions. One case with focal sclerosis was picked up only on CT, while focal fibrosis (1 case) and abnormal signal intensity (2 cases) were detected only on MRI [Table 2].

In 71 (78.9%) cases, no abnormality was detected in semicircular canals (SCCs) on CT, while on MRI, 66 (73.4%) cases were normal. Both modalities performed equally in the detection of aplasia of SCC (2 cases), while MRI performed better than CT in diagnosing dysplastic malformations of SCC (18 vs. 15). Focal fibrosis in 1 case and abnormal signal in 3 cases was identified only on MRI, while 2 cases with focal sclerosis were diagnosed only on CT [Table 2].

Twenty (22.2%) cases with the Deficient cochlear nerve were diagnosed only on MRI. In 8 (8.9%) cases, the cochlear

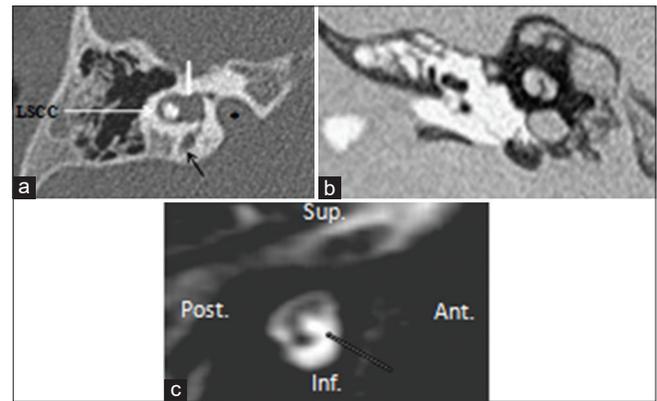


Figure 2: Type II incomplete partition (Mondini deformity) (a) Axial computed tomography image show dilated vestibule (white arrow) and vestibular aqueduct (thin black arrow) with widening of internal auditory canal (*) and dysplastic lateral semicircular canal (b) coronal computed tomography inverted image show cystic cochlear apex with only 1.5 turns (c) oblique sagittal three-dimensional magnetic resonance constructive interface steady state image shows absent cochlear nerve in the internal auditory canal (thick black arrow)

Table 1: Distribution of cochlear abnormalities (malformations) on both high resolution computed tomography and magnetic resonance imaging in temporal bones

Distribution	High resolution CT (n=90), n (%)	MRI (n=90), n (%)
Normal	65 (72.3)	63 (70)
Abnormal	25 (27.7)	27 (30)
Abnormalities		
Michel’s malformation	2 (2.2)	2 (2.2)
Isolated cochlear aplasia/hypoplasia	3 (3.3)	3 (3.3)
Common cavity malformation	4 (4.4)	4 (4.4)
Incomplete partition Type I	5 (5.6)	5 (5.6)
Incomplete partition Type II (Mondini’s malformation)	8 (8.9)	8 (8.9)
Labyrinthitis ossificans		
Fibrotic stage	0	1 (1.1)
Ossified stage	1 (1.1)	0
Uncategorized		
Sclerosis	2 (2.2)	0
Signal loss	0	4 (4.4)

MRI – Magnetic resonance imaging; CT – Computed tomography

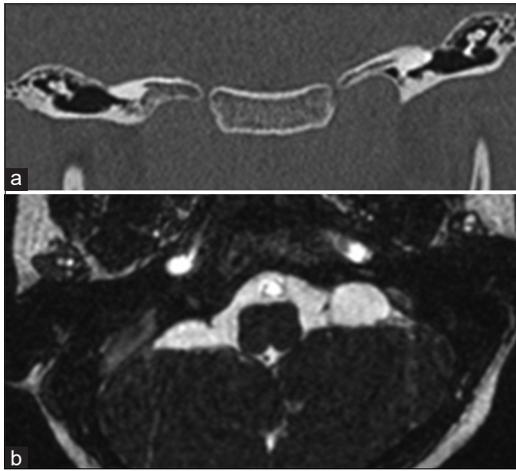


Figure 3: Bilateral Michel's aplasia (a) coronal high-resolution computed tomography images show complete absence of inner ear structures bilaterally with dense sclerotic bone underneath the cochlear promontory and hypoplastic petrous bones (b) Axial constructive interface steady state magnetic resonance images shows bilateral severely atretic internal auditory canal with visualization of single hypoplastic seventh nerve in cerebellopontine angle cistern

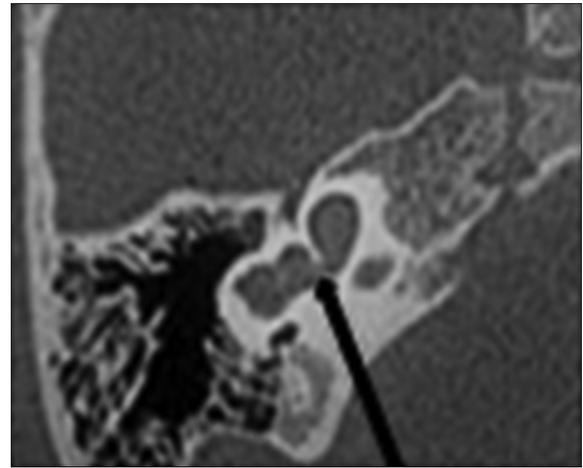


Figure 4: Right common cavity deformity. Confluent cochlea and vestibule forming a cystic cavity (thick black arrow) seen on axial high resolution computed tomography with no internal architecture and absent semicircular canals

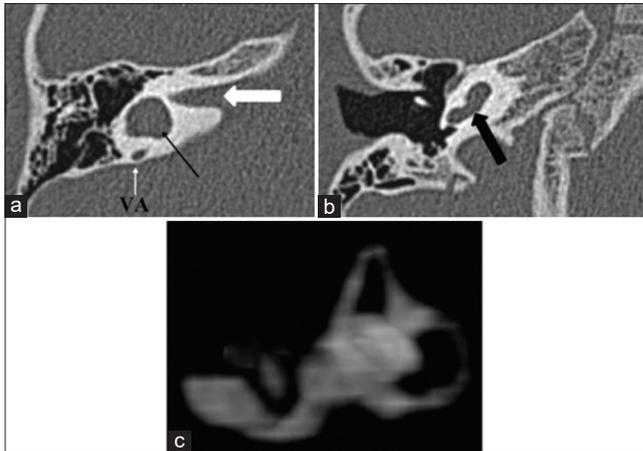


Figure 5: Cystic cochleo-vestibular malformation. (a and b) High resolution computed tomography axial images shows cystic dilated vestibule (thin black arrow) with widened basal cochlear turn (thick black arrow). Internal auditory canal (thick white arrow) and vestibular aqueduct are normal in size. (c) Magnetic resonance maximum intensity projection image show enlarged vestibule with dysplastic dilated basal turn of cochlea with nonvisualization of middle and apical turns with absent modiolus

nerve was not seen while in 12 (13.3%) of cases, it was hypoplastic. In 9 (10%) cases, vestibular aqueduct (VA) was hypoplastic and in 5 (5.6%) cases, it was enlarged [Table 3].

In 8 cases with deficient cochlear nerve associated hypoplasia of IAC was seen. One case with widened IAC was also observed. Modiolus was absent in 10 (11.1%) cases and was only partially seen in 6 (6.7%) cases [Table 3].

Only CT allowed accurate assessment of mastoid aeration in cases with structural abnormalities. Out of 27 cases with

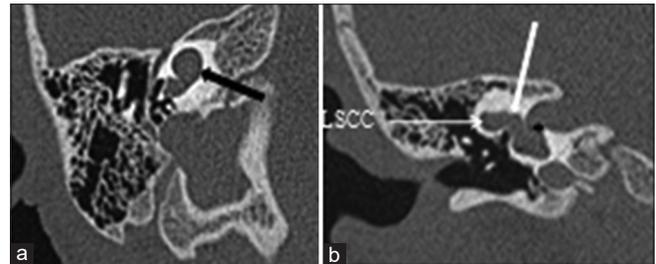


Figure 6: Cochlear dysplasia (Type I incomplete partition). High resolution computed tomography (a) axial and (b) coronal images show dilated cystic basal cochlear turn (thick black arrow) with absent modiolus, enlarged vestibule (thick white arrow) and malformed lateral semicircular canal. Internal auditory canal is also widened with partially dehiscent cribriform plate (*)

cochlear malformations 7 (26%) had under-pneumatized mastoids and 5 had completely sclerosed mastoids. In 15 cases, associated middle ear disease was also seen. We also found few anatomical variations on HRCT in the cases with positive malformations, which include high riding jugular bulb (2 cases), dehiscent jugular bulb (1 case), and dehiscent facial nerve canal (1 case). Abnormal white matter signal intensities were also detected in 4 cases on screening axial sections of MRI.

DISCUSSION

SNHL is considered one of the major causes of pediatric disability, with almost 20% of cases associated with inner ear malformations.^[10,14] Early diagnosis, as well as management, is very crucial because it is well recognized that any delay in diagnosis adversely affects the development of language and speaking skills with overall impaired academic, emotional and social growth of the child.^[9,15] SNHL results from the malfunctioning of the

Table 2: Distribution of vestibular and semicircular canal abnormalities (malformations) on both high resolution computed tomography and magnetic resonance imaging in temporal bones

Distribution	High resolution CT (n=90), n (%)	MRI (n=90), n (%)
Vestibule		
Normal	77 (85.6)	75 (83.4)
Aplastic	2 (2.2)	2 (2.2)
Malformed	10 (11.1)	10 (11.1)
Others		
Focal sclerosis	1 (1.1)	0
Focal fibrosis	0	1 (1.1)
Signal loss	0	2 (2.2)
Semicircular canals		
Normal	71 (78.9)	66 (73.4)
Aplastic	2 (2.2)	2 (2.2)
Malformed	15 (16.7)	18 (20)
Others		
Focal sclerosis	2 (2.2)	0
Focal fibrosis	0	1 (1.1)
Signal loss	0	3 (3.3)

MRI – Magnetic resonance imaging; CT – Computed tomography

Table 3: Distribution of other abnormalities of inner ear on both high resolution computed tomography and magnetic resonance imaging in temporal bones

Distribution	High resolution CT (n=90), n (%)	MRI (n=90), n (%)
Cochlear nerve		
Normal	0	70 (77.8)
Deficient		
Aplastic	0	8 (8.9)
Hypoplastic	0	12 (13.3)
VA (endo-lymphatic duct)		
Normal	76 (84.4)	76 (84.4)
Hypoplastic	9 (10)	9 (10)
Enlarged	5 (5.6)	5 (5.6)
Internal auditory meatus		
Normal	81 (90)	82 (91.1)
Hypoplastic	8 (8.9)	7 (7.8)
Widened	1 (1.1)	1 (1.1)
Modiolus		
Normally present	74 (82.2)	74 (82.2)
Partially present	6 (6.7)	6 (6.7)
Absent	10 (11.1)	10 (11.1)

MRI – Magnetic resonance imaging; CT – Computed tomography; VA – Vestibular aqueduct

inner ear or conditions affecting cochlear nerve or central auditory centers in the brain.^[5,16]

Currently, electrical stimulation of the cochlear nerve by cochlear implantation is the treatment of choice for children with profound SNHL in whom the routine amplification methods fail to provide clinically significant improvement.^[17-20] Cross-sectional imaging forms an integral part of preoperative workup in such cases. CT and/or MRI are routinely ordered by the otolaryngologists to ascertain the potential cause of hearing loss, to preoperatively define the anatomy of the petrous bones, and to identify the structural malformation which may contraindicate implant surgery.^[10,19,21]

In the present study, all the cases of profound SNHL were evaluated on both imaging modalities. Majority of patients (89%) belonged to the age group of 0–5 years. With increasing parental awareness, most of the patients who are deaf and mute now present early for the clinical evaluation, a fact which is well depicted in our study and correlates with previous studies^[1,5,14] as well.

Males were more commonly affected in our study with a male:female ratio of 1.4. Our observations match with Cremers *et al.*^[22] and Bamiou *et al.*^[23] as they also reported male preponderance in their respective studies. Researchers are still not able to find out any convincing explanation for this male predominance, but possible reasons could be the higher genetic susceptibility of males and preferential male referral to the higher tertiary care centers.

Numerous classification systems have been developed to categorize patients with inner ear malformations based on distinctive imaging patterns.^[4,10,15,16,24] The most widely accepted classification is the one proposed by Jackler *et al.*^[2] and Sennaroglu and Saatci.^[13] They proposed that dysplasia of inner ear is a result of aberrance/arrest in the process of development during different stages of organogenesis and categorize them into following types: Complete labyrinthine aplasia, cochlear aplasia, cochlear hypoplasia, common cavity malformation and incomplete partitioning which was further divided into type I (cystic cochleovestibular dysplasia) and type II (classic Mondini abnormality).^[2,13,16] This framework is most useful in describing and understanding labyrinthine dysplasias.

Type II incomplete malformations were more common than type I incomplete malformations in this study. The differentiation between the two malformations is important from the surgical point of view as there is an increased likelihood of cerebrospinal fluid leakage during implantation in type I malformation.^[3] Both CT and MRI performed well in the identification of these malformations in our study.

Isolated Mondini dysplasia was the third-most common abnormality in the study of Bamiou *et al.*^[23] who retrospectively evaluated 116 patients of SNHL on CT. The dilated VA was the most frequently detected abnormality in their study. Chaturvedi *et al.*^[14] prospectively evaluated 30 patients with deafness on CT, of which 15 subsequently underwent MRI also. The incidence of cochlear malformations in their study was only 6.6%. The most frequently observed malformation in the study by Digge *et al.*^[1] was SCC abnormalities (89/144, 62%) followed by cochlear malformations (39/144, 27%). They

evaluated 72 patients (144 temporal bones) prospectively on both CT and MRI, and Mondini's deformity was the most common cochlear pathology in their study. Westerhof *et al.*^[25] used dual modalities to evaluate 21 children (42 ears) with congenital deafness and detected 99 malformations. Mondini variants and fusion of SCC with vestibule were two most frequent imaging findings seen in 12 of 42 ears each. Thus it can be observed that the incidence of inner ear malformations vary from study to study largely owing to differences in the sample size, study design, study population, racial, and genetic factors.

The abnormalities of SCC were more commonly seen than the vestibular anomalies in our study. Both CT and MRI performed fairly well in the identification of these malformations with few exceptions. Focal areas of sclerosis with increased density were picked up only on CT and not on MRI, while focal fibrotic lesions and abnormal signal intensity were detected only on MRI. Our findings are in concordance with the previous studies.^[1,23] Abnormalities of SCC and vestibule were frequently encountered in patients with SNHL in the study by Bamiou *et al.*^[23] where malformations of SCC were the second most commonly detected abnormality (7.75%). Digge *et al.*^[11] in their study also found CT superior to MRI in diagnosing ossified/sclerosing lesions while fibrotic lesions were detected only on MRI.

The presence of cochlea and the cochlear nerve are the two absolute requirements for the cochlear implantation.^[14,20] Even in patients with small hypoplastic cochlear nerve, cochlear implantation can still be done, although the prognosis is guarded. The term cochlear nerve deficiency encompasses both hypoplastic as well as absent cochlear nerve and is usually associated with atretic IAC. The morphology of the bony IAC is best depicted on CT, but the cochlear nerve *per se* cannot be directly visualized on CT. Further, the normal-sized IAC and completely normal inner ear on CT still does not rule out the possibility of the absent cochlear nerve.^[26] MRI is the only reliable modality which allows direct assessment of the cochlear nerve within the cerebellopontine angle cistern and IAC. Oblique sagittal sections perpendicular to the longitudinal plane of IAC allow excellent demonstration of all four major nerve bundles within the canal.^[1,3,4,26] In this study, we found a deficient cochlear nerve in 20 cases, which was diagnosed only on MRI. Of these in 8 (8.9%) cases, it was totally absent while in rest 12 (13.3%), it was hypoplastic. Thus, we found MRI superior to CT in the detection of the cochlear nerve. Our findings are in agreement with the previous research. Nine out of 21 pediatric cases in the study by Westerhof *et al.*^[25] had rudimentary or absent cochlear nerve, which

was diagnosed only on MRI. Komatsubara *et al.*^[27] in their study found that in cases with narrow (<1.5 mm) IAC on CT, the detection of cochlear nerve deficiency was achieved better by MRI. Ellul *et al.*^[28] retrospectively evaluated 31 cochlear implant candidates and concluded that MRI is more accurate than CT in the assessment of the presence and size of vestibulocochlear nerve.

Bacterial meningitis with resultant labyrinthitis ossificans constitutes one of the major causes of acquired SNHL in pediatric patients. The onset of the disease process is usually variable and may manifest months or years after the initial insult. Early stages of labyrinthitis ossificans are characterized by postinflammatory fibrosis, while ossification represents the end stage disease process. Early diagnosis and referral before the onset of cochlear obstruction are essential for successful cochlear implantation in these cases. MRI is the first-line modality for diagnosing early-stage labyrinthitis ossificans where the loss of normal T2 hyperintensity of labyrinthine fluid suggests fibrosis. CT, however, cannot distinguish fibrosis from normal labyrinth fluid, but it has a definitive role in later stages characterized by dense ossification. MRI on the other hand cannot differentiate between fibrotic and mineralized obstruction as both appear hypointense on T2WI.^[14,29] We found two cases of labyrinthitis ossificans in our study, one in the early stage, which was detected on MRI and other in the late stage, which was picked up only on CT. Digge *et al.*^[11] also encountered similar situation in their study where two cases of labyrinthitis ossificans with early fibrosis were identified on MRI and other 2 cases with late-stage ossification were detected on CT. Chaturvedi *et al.*^[14] missed 2 cases with subtle early cochlear ossification on HRCT in their study, which were subsequently detected on MRI. They thus emphasized that all the cases with a high index of suspicion (postmeningitic deafness) for cochlear obstruction must undergo preoperative MRI to rule out early labyrinthitis ossificans.

In our study, we observed that the status of middle ear pneumatization, mastoid aeration, and anatomical variants were depicted accurately only on CT and MRI was of little help. The pathologies of mastoid and middle ear result in conductive hearing loss and not sensorineural. However, nevertheless, the assessment of anatomy and status of aeration of mastoids and tympanic cavity preoperatively plays a vital role in planning surgical roadmap for cochlear implantation, which necessitates partial mastoidectomy. Underpneumatized or sclerosed mastoids limit the exposure of the middle ear cavity. Similarly, few anatomical variants such as high riding jugular bulb or dehiscent jugular bulb, may interfere with the placement of cochleostomy.^[3,4]

Chaturvedi *et al.*^[14] found CT superior to MRI in detecting high riding jugular bulb and tracing aberrant course of the facial nerve. Bhavana and Kumar^[6] found HRCT useful in evaluating middle ear bony anatomical variations and mastoid pathologies. Thus, it can be concluded that only CT can provide reliable presurgical information about variant ear anatomy and pneumatized structures.

Selecting ideal preoperative imaging in cochlear implant candidates has always been a field of intense debate with divided opinions among the researchers. The earlier work focused primarily on the utility of HRCT in the evaluation of congenital SNHL. Various studies reported a substantial yield of CT (ranging from 7% to 30%) in detecting abnormalities pertaining to the causes of SNHL.^[23] CT of petrous bones brilliantly demonstrates osseous anatomy, bony landmarks, mineralized, and aerated structures.^[3,4,23] However, the inability to demonstrate the cochlear nerve directly constitutes one of the major limitations of the CT.^[1,3,6] On the other hand, owing to the excellent soft-tissue resolution, MRI is far better than CT in the demonstration of neural structures.^[26] MRI can also provide additional information regarding the status of fluid in the membranous labyrinth and coexisting brain abnormalities.^[6,14,30] While few authors^[28] suggest MRI as the sole imaging modality in the preimplant workup, we in our study found that neither CT nor MRI alone is sufficient enough and combination of both modalities offer the best imaging algorithm in patients with SNHL which is in agreement with most of the previous studies.^[1,5,25,30]

Limitations of the study

The sample size in our study is relatively small, which may limit the statistical significance. No attempt was made in our study to ascertain the etiology of deafness.

CONCLUSIONS

The addition of both multislice CT and high strength MRI to preoperative imaging workup in patients with bilateral profound SNHL for cochlear implantation is justified as none of the modality in isolation is capable enough to address all the issues. Both modalities complement each other and reduce the chances of missing critical and crucial findings. Thus, it is recommended to perform dual imaging with both high-resolution CT and high magnet MRI, wherever and whenever possible, to offer maximum information to managing surgeon preoperatively, thereby helping inappropriate patient management.

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Conflicts of interest

There are no conflicts of interest.

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