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RESEARCH ARTICLE

Role of Imaging in congenital inner ear anomalies in children with severe -profound sensory neural hearing loss

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ABSTRACT

Introduction

Sensory neural hearing loss (SNHL) is quite a significantly burdened cause of childhood disability; the estimated prevalence is 1 in 2000 neonates and 6 in 1000 children present with SNHL by 18 years of age. High-resolution computed tomography (HRCT) of the temporal bone and magnetic resonance imaging (MRI) of the temporal region and brain provide invaluable evaluation and characterization of inner ear structures and their anomalies. Radiological imaging also plays a major role in cochlear implantation with regard to intraoperative monitoring, postoperative evaluation and also in research and experimental techniques. Imaging the complete intracranial as well as extracranial auditory pathway of the implant candidate is necessary to screen for morphological conditions that will preclude or complicate the implantation process including final outcome in terms of development of speech. In this study we have analysed the inner ear congenital anomalies and their prevalence which we encountered while imaging congenitally deaf and mute children to decide their candidature for cochlear implantation and also there is a paucity of literature in this regard in Indian context for this reason we conducted this study.

Methodology

This prospective study was conducted at a tertiary care center in central India, which is also a designated center for cochlear implant surgery. All children attending the outpatient department with hearing loss in the age group of 1–7 years were screened, and those who had severe to profound sensorineural hearing loss [>90 dB] were selected as subjects. All patients were subjected to an audiological assessment after ear examination, HRCT, and MRI of the temporal bone.

Result

The total number of children analyzed was 210 with congenital SNHL. The number of radiologically abnormal cases with one or more anomalies as per HRCT and MRI temporal bone along with congenital SNHL was 35 Out of all inner ear anomalies observed, it was noticed that cochlear anomalies bear most of the burden. We observed that nearly 60% of children have one or the other cochlear anomaly present, whereas facial nerve anomalies were the least observed, i.e., only 8.57%. Among various abnormalities of vestibule, nearly 53% had dilated vestibule whereas it was hypoplastic in 13%.

Conclusion

With the appropriate and timely use of imaging studies and understanding the diagnostic yield of HRCT and MRI of temporal bone, it is possible to understand and clearly find out the exact cause of hearing loss in children, which can be further utilized to plan and manage the children for various options available for management of their hearing.

Introduction

SNHL is quite a significantly burdened cause of childhood disability; the estimated prevalence is 1 in 2000 neonates and 6 in 1000 children present with SNHL by 18 years of age[1]. This is to be diagnosed and identified early, as impairment of hearing has quite a significant impact on various aspects of development like speech and language, social and emotional and academic achievements as well[2].

In terms of embryological development, inner ear development starts as early as the 3rd week of intrauterine life. Out of all congenital anomalies of the inner ear, only 20–30% will appear significantly on radiological imaging[3]. With the advancement in technology in the current scenario, there are three main treatment options available for rehabilitation of hearing: hearing aids, cochlear implants (CI), and auditory brain stem implants (ABI). Convincingly, the cochlear implant has gained a lot of popularity in recent years and is said to be an effective modality to provide hearing for congenital severe to profound SNHL. High-resolution computed tomography (HRCT) of the temporal bone and magnetic resonance imaging (MRI) of the temporal region and brain provide invaluable evaluation and characterization of inner ear structures and their anomalies. HRCT is an excellent informative assessment tool to diagnose congenital anomalies of the bony labyrinth, which include osseous details as well as variant anatomy, whereas MRI is used to look in the membranous labyrinth, vestibulocochlear nerve-free fluid spaces, and any other brain pathology to good effect[5].

NORMAL INNER EAR ANATOMY

The inner ear consists of a membranous (endolymphatic) labyrinth containing the functional sensory epithelium, surrounded by an osseous (bony) labyrinth with an interposed perilymphatic labyrinth. The membranous labyrinth consists of the utricle, saccule, semicircular ducts, cochlear duct (scala media), and endolymphatic duct and sac. Radiologically shown via Figure 6, there are various classifications of congenital inner ear malformation that describe the same in different ways. The 2002 classification given by Sennaroglu and Saatci[6] says.

COCHLEAR MALFORMATIONS

- Michel's deformity: complete absence of all cochlear as well as vestibular structures
- Cochlear aplasia: complete absence of only cochlea
- Common cavity deformity: the common cystic cavity replaces the cochlea and the vestibule
- Cochlear hypoplasia: The dimensions of the cochlea and vestibule are smaller than normal
- Incomplete Partition Type 1 (IP-I): The cochlea lacks a modiolus and is accompanied by a large cystic vestibule. (Figures 2 and 5)
- Incomplete partition type 2 (IP-II), also known as Mondini malformation: The cochlea consists of 1.5 turns, in which the middle and apical turns join to form an apex of cystic aspect with the dilated vestibule and the enlarged vestibular aqueduct.

In this study, we have analyzed the inner ear congenital anomalies and their prevalence that we encountered while imaging congenitally deaf and mute children to decide their candidacy for cochlear implantation.

Methodology

This prospective study was conducted at a tertiary care center in central India, which is

also a designated center for cochlear implant surgery. All children attending the outpatient department with hearing loss in the age group of 1–7 years were screened, and those who had severe to profound sensorineural hearing loss [>90 dB] were selected as subjects. A total of 210 congenital SNHL patients were selected from the Department of Otorhinolaryngology and Head and Neck Surgery at Sri Aurobindo Institute of Medical Sciences and Post Graduate Institute, Indore, India, as subjects for the study. Parents and guardians of these children were explained clearly about the study and those who were willing to participate in it; their consent was recorded on a consent form, duly signed by them. A total of 210 such children were available for the study. Children above 7 years of age with middle ear or tympanic membrane pathology were excluded from our study. All patients were subjected to an audiological assessment, ear examination, HRCT, and MRI of the temporal bone. Images were analyzed for the presence of normal structure as well as any anomalies present in the inner ear. Various expected vestibular malformations include Michel malformation, common cavity, absence of vestibule, and hypoplastic/dilated vestibule. Among the superior semicircular canal malformations, expected outcomes could be an absent canal, a hypoplastic canal, or a dilated semicircular canal. Even malformations of the internal auditory canal are not uncommon, like an absent, narrow, or dilated internal auditory canal. Vestibular and cochlear aqueduct findings observed could be dilated or normal cochlear vestibule and aqueduct, as well as the presence or absence of the nerves (facial and cochlear), the relative internal auditory canal (IAC), and any associated other inner ear abnormalities. Data was entered in Microsoft Excel and analyzed using SPSS software version 2.0.

Observations and result

The total number of children analyzed was 210 with congenital SNHL. The number of radiologically abnormal cases with one or more anomalies as per HRCT and MRI temporal bone along with congenital SNHL was 35.

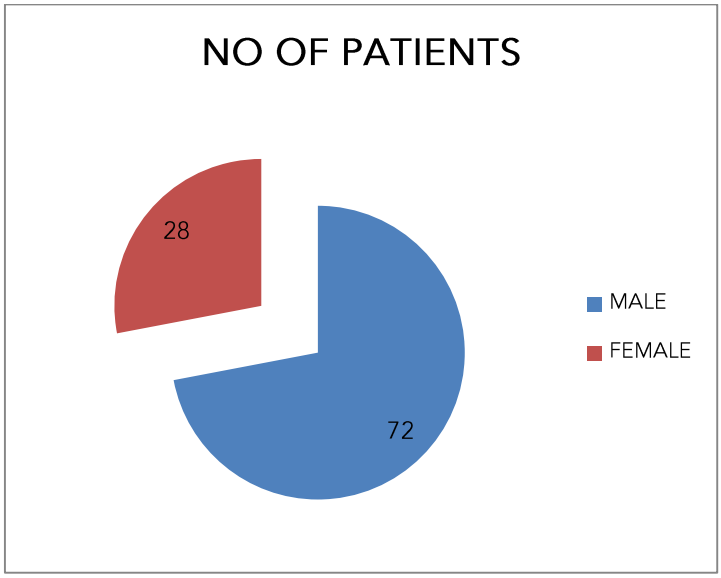

FIGURE 1 Male-to-female ratio of children with congenital anomalies among the study population

Out of a total of 35 children, males were 25 and females were 10. So male predominance was maintained with male: female is 2.5:1(Figure 1).

Out of all inner ear anomalies observed, it was noticed that cochlear anomalies bear most of the burden. We observed that nearly 60% of children have one or the other cochlear anomaly present, whereas facial nerve anomalies were the least observed, i.e., only 8.57%. (Tables 1 and 2).

Table 1- Various Inner Ear Abnormalities Observed

Abnormality	No of patients	Percentage
Cochlear abnormality	21	60%
Vestibular abnormality	15	42.8%
Vestibular aqueduct	16	45.71%
Semicircular canal	14	42.85%
Cochlear nerve	15	42.85%
Facial nerve	3	8.57%
Endolymphatic duct	8	22.85%
Internal auditory canal	11	31.42%

Table 2- Cochlear Abnormality (n=35)

Abnormal	21	60%
Normal	14	40%
Total	35	100%

Table 3: Distribution of patients according to cochlear abnormality (n = 21)

Cochlear hypolasia	7	33%
Cochlear aplasia	3	14%
Cochlear dysplasia	2	9.5%
Common cavity	5	23.8%
Cystic cochlea	4	19.04%

Among various cochlear anomalies cochlear hypoplasia was seen in most of the cases which is around 33%, in 23.8% children cochlea and vestibule were seen as common fused cavity also complete absence of cochlea known as cochlear aplasia(Figure-) was seen in 3 out of 35 children who have inner ear abnormalities (Table 3).

Table 4- Distribution of patients according to vestibular abnormality

Dilated vestibule	8	53.33%
Hypolpasia	2	13.33%
Cystic	1	6.66%
Absent	1	6.66%

Among various abnormalities of vestibule, nearly 53% had dilated vestibule whereas it was hypoplastic in 13% (Table 4). Semicircular canal anomalies were also observed, most of the children had lateral semicircular canal abnormalities i.e 60%, nearly 6 patients out of 35, out of 6, 2 children had absent LSCC whereas 3 had a hypoplastic one. (Table 5).

Table 5- LSCC (Lateral Semicircular Canal)

Lscc absent	2	33%
Lscc hypoplastic	3	50%
Lscc dilated	1	17%
Total	6	100%

30% of patients were present with PSCC abnormalities, and only 10% of cases had SCCC abnormalities. Cochlear nerve abnormality, 80% of cases have cochlear nerve absent. Internal auditory canal anomalies: 50% of cases have a narrow IAC, and 37% of cases present with a dilated IAC endolymphatic duct abnormality. Only 24% of cases have a dilated endolymphatic duct, and 76% of cases do not have an abnormality.

In our study, the total number of patients with congenital SNHL was 150, out of which 25 (17%) had an inner ear abnormality. In 2002, Mafong et al.[7] determined that 39 % of children with sensorineural hearing loss (SNHL) had an abnormality on either a temporal bone HRCT scan or an MRI. Clinically, children show unilateral or bilateral sensorineural hearing loss right from birth itself, which represents around 20% of cochlear malformations and is seen in imaging studies as cystic cochlea and the dilated vestibule.[8] It originates as early as the fifth week of intrauterine life and affects the development of the internal structures of the cochlea. There are different degrees of anomalies; the mild degree presents the malformation only at the level of the cochlea with vestibule and normal semicircular ducts and sometimes absence of modiolus, unlike the more advanced degree of anomalies, in which the vestibule-cochlear apparatus and horizontal semicircular cannal are globularly enlarged, which then form an outline. Radiological findings are detected by an HRCT scan[9].

In 2015, Huang B et al.[10] had a total number of patients with congenital SNHL of 1012, of which only 9% had inner ear abnormalities. In 2016, Dagkiran et al.[11] had a total of 600 patients with congenital SNHL, of which 20% had inner ear malformations. Overall, as per the literature, cochlear aplasia accounts for 5% of cochlear malformations, as embryonic development stops at the end of the third week of gestation and is usually of unknown

etiology[8,9]. Cochlear hypoplasia represents 12% of cochlear malformations and is due to an alteration in the development of the cochlear canal during the sixth week of gestation. In cochlear malformation, we found the incidence of cochlear hypoplasia is 33% in most of the patients; cochlear aplasia is 13% (Figure 4). We also found 7% cochlear dysplasia, 27% of patients have a common cavity, and 20% have a cystic cavity (Figure 3). In 1987, Jackler et al.[12] found that the common cavity is 26%, cochlear aplasia is 1%, cochlear hypoplasia is 15%, and cystic cavity is in 55% of cases. In 2001, Casselman JW et al.[13] found cochlear aplasia in 3% of patients. In 2012, Joshi V et al.[14] found anomalies in 18% of patients with cochlear dysplasia (10.2%). In our study, we found 44% of vestibular abnormalities, in which most of the patients have a dilated vestibule, i.e., 55%, 18% of patients have a common cavity, and 9% of patients have vestibule hypoplasia, cystic, and absent vestibule. In 2012, Joshi V et al[14] found vestibular dysplasia was 10.2%. In 2015, Wu W et al.[15] found only 3% vestibular abnormality. In the present study, 48% of patients have vestibular aqueduct abnormalities (Figure 7), of which 75% have dilated vestibular aqueducts and 25% have absent vestibular aqueducts. In 201, Muhammed et al.[16] observed wide vestibular aqueducts in 16 ears of 136 (11.7%). In 2002, a study conducted by Sennaroğlu et al.[17] found that wide Vestibular Aqueduct was observed in 22.2%, and in another study, they observed it in 34%.

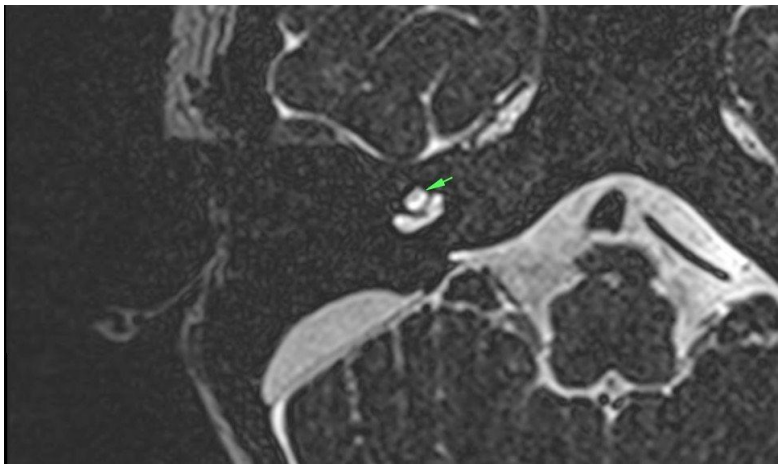

Figure 2: Axial section in MRI temporal bone showing ONE AND A HALF TURN COCHLEA (INCOMPLETE PARTITION 2) (arrow)

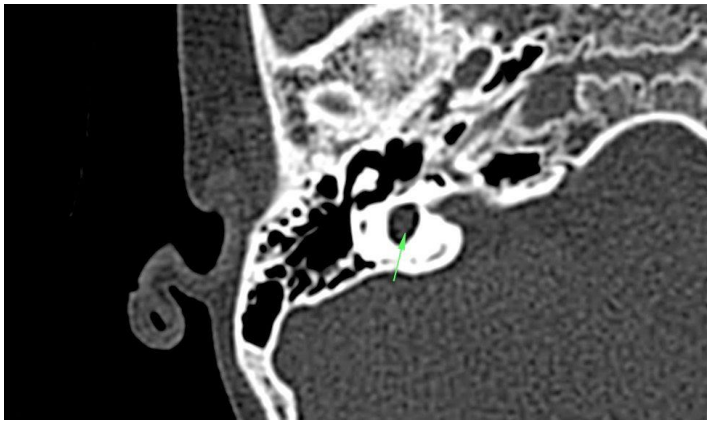

Figure 3: Axial section of the HRTC temporal bone showing the common cavity (arrow)

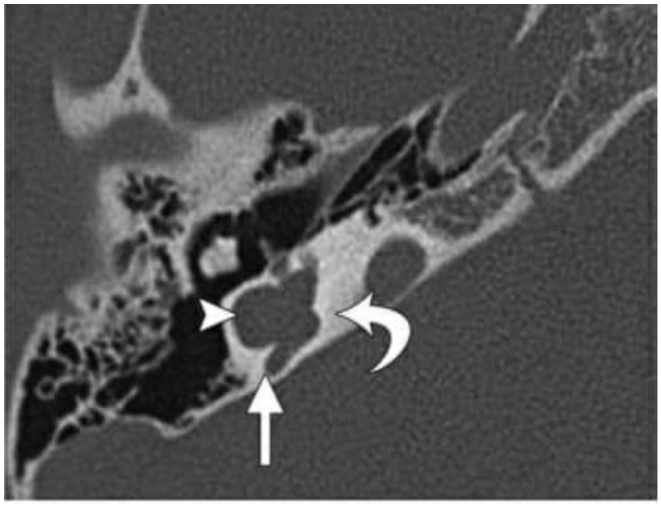

Figure-4 Cochlear Aplasia

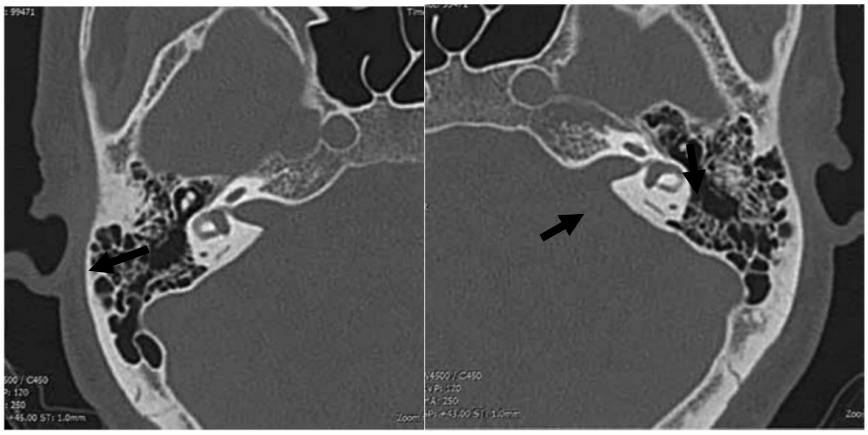

Figure 5. a) Right (b) Left ear Incomplete Partition Type 1 (IP-1). The arrow, the malformation is only in the cochlea. Since there is no interscalar septum or modiolus, the vestibule and the semicircular ducts are normal[18].

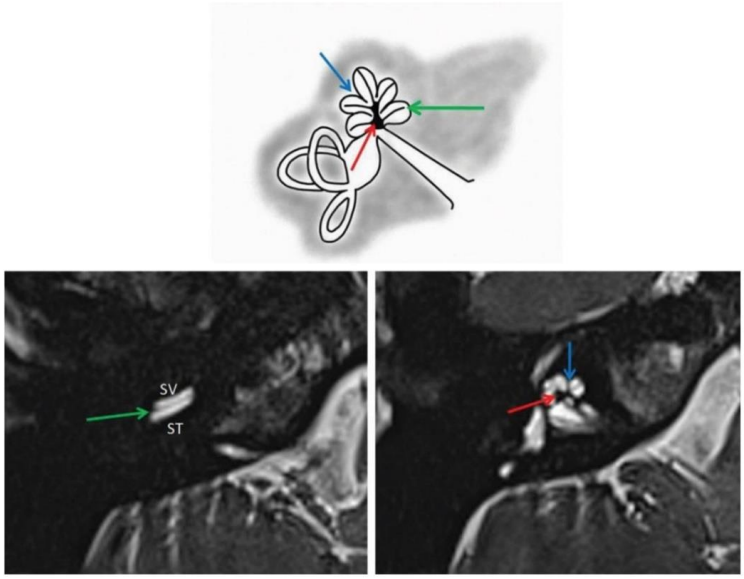

Figure 6 (A) Schematic diagram of normal inner ear (B)(C) Axial 3D T2W SPACE images shows osseous spiral lamina of basal tum of cochlea separating the scala vestibuli (SV) and scala tympani (ST), interscalar septum between middle and apical turn of cochlea (blue arrow), and central modiolus (red arrow)[19]

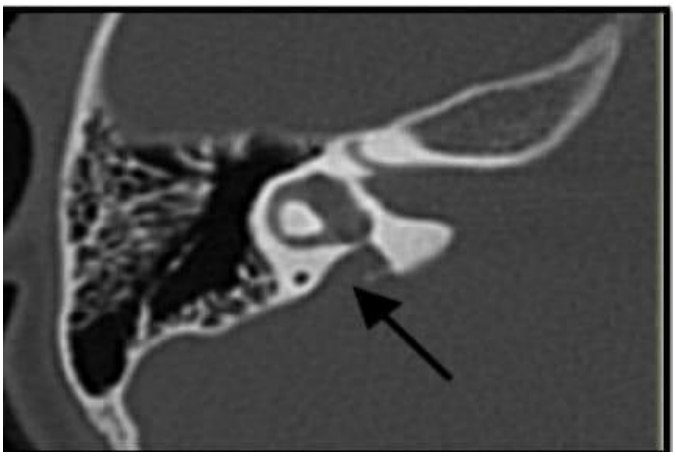

Figure 7- Enlarged vestibular aqueduct

Conclusion

With the appropriate and timely use of imaging studies and understanding the diagnostic yield of HRCT and MRI of temporal bone, it is possible to understand and clearly find out the exact cause of hearing loss in children, which can be further utilized to plan and manage the children for various options available for management of their hearing, including candidacy for cochlear implantation and also appropriate planning for the same. Both imaging techniques precisely delineate inner ear malformations with complementary information and are used together in the preoperative planning for cochlear implantation in children as well as adults. Most children who present with congenital SNHL do have one or more vestibulo-cochlear malformations. Although it is beneficial to understand the site and cause of hearing loss, this still has no prognostic or preventive value.

Conflict of Interest:

None

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