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ORIGINAL ARTICLE

Prevalence of Temporal Bone Malformations in Prospective Cochlear Implant Candidates

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Abstract Severe to profound Sensorineural Hearing Loss is a challenging medical problem, particularly if this condition is associated with an inner ear anomaly. This case series studies the prevalence of inner ear anomalies among 76 consecutive prospective cochlear implant candidates who presented to our tertiary care hospital over 2 years. Inner ear anomalies were identified in 11 cases with a prevalence rate of 14%. Narrow Internal Auditory Canal (IAC) is the most common inner ear anomaly (5) followed by Mondini (3) and Globular vestibule (3). Combined CT and MRI play an important role in the preoperative assessment of inner ear anomalies which may affect not only the decision to perform the Implant procedure and the prognosis but also the choice of implant and surgical technique.

Keywords Sensorineural hearing loss · Temporal bone malformation · CT and MRI

Introduction

Membranous malformations account for 80% of congenital hearing loss cases. The bony abnormalities of the temporal bone can be radiologically demonstrated in 20% of cases. This group with bony anomalies present surgical challenges and also affect decision making in the management. SNHL can result from a combination of Temporal bone abnormalities that may involve the labyrinth, internal auditory canal, cerebellopontine angle, brain stem, or auditory pathways.

Cochleovestibular anomalies are a variety of congenital anomalies, the severity of which depends on the time at which an insult occurs during embryogenesis. These anomalies have been classified by Jackler1 et al. [1] in 1987 which was modified by Sennaroglu's in [2] (Table 1). Starting at the 3rd week of gestation, an insult during each subsequent week, up to the end of the 7th week, results in a distinct inner ear abnormality. Total labyrinthine aplasia (Michel) occurs if the embryogenesis is insulted as early as the 3rd week and Incomplete partition (IP) type 2 (Mondini) is seen if it is at the 7th week. The common cavity has been described as one of the common malformations which results if the embryogenesis is halted in the 4th week. Incomplete partition anomalies represent a group of cochlear malformations, where there is a clear differentiation between cochlea and vestibule. The most common Incomplete partition type is Type 2 (Classical Mondini), IP type 1 is a more severe anomaly resulting in a cystic cochleovestibular malformation. IP type III cochlear malformation is the type of anomaly present in X-linked deafness, which was described by Nance et al. Sennaroglu and Bajin [3] added Isolated Enlarged Vestibular Aqueduct (EVA) and Cochlear aperture abnormalities to the above types of Inner Ear Malformations.

Isolated EVA with normal cochleovestibular structures in prospective cochlear implant candidates is less common, but EVA is found associated with other Cochleovestibular anomalies like in Mondini. Cochlear aperture abnormalities are usually seen with Internal auditory canal malformations. Narrow IAC (midpoint less than 2.5 mm) is seen on HRCT should be evaluated with a thickness of the 8th nerve on MRI. The hypoplastic or aplastic cochlear nerve on MRI has an important bearing on the management of congenital

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Table 1 Modified sennaroglu's classification

Time of Insult (week)	Malformation	Features
Third	Labyrinthine aplasia (Michel deformity)	Complete absence of cochlea and vestibule
Third-to-fourth	Cochlear aplasia	Complete absence of cochlea; vestibule present
Fourth	Common cavity	Single cystic cavity representing cochlea and vestibule, without any differentiation
Fifth	Cystic cochleovestibular anomaly (IP-I)	Cystic-appearing cochlea lacking entire modiolus and cribriform area; large cystic vestibule
Sixth	Cochlear hypoplasia	Cochlea and vestibule are separate but are smaller than normal; hypoplastic. Cochlea resembles a small bud off the IAC
Seventh	Incomplete partition type II (IP-II)	Cochlea consists of 1.5 turns, in which middle and apical turns coalesce to form a cystic apex; vestibule and VA may be enlarged

deafness cases. CT and MR imaging often play a complimentary role in its evaluation and preoperative assessment. Imaging has an important role in deciding candidacy for the feasibility of implant, providing realistic preoperative counselling and predicting postoperative outcomes.

Methods and Materials

The study aims to determine the prevalence of inner ear anomalies and the frequency of different anomaly types among preoperative cases for cochlear implantation.

This is a Prospective observational study of all patients who presented with severe to profound SNHL between June 2017 to May 2019 in KIMS HOSPITALS SECUNDERABAD. All subjects underwent combined CT of the temporal bone and MRI. CT scan was done on a 128-slice scanner. CT was done using a multi-detector scanner, 0.625 mm axial scans of the temporal bone were acquired using a high-resolution bone technique. The scans were retrospectively targeted for the right and left sides, and subsequently reconstructed in the coronal plane, again targeting the right and left sides individually, as well as the entire skull base. MRI was performed on a 3 Tesla MR system. MRI technique was done with selected sequences. Both axial and coronal images were reviewed in the bone window and 0.8 mm thin slices on HRCT of the temporal bone. 3D volume-rendered images of the inner ear structures were also assessed by post-processing on the workstation.

Mixed, moderate hearing loss cases were excluded. Patients with acquired causes of SNHL like meningitis leading to labyrinthitis ossificans were excluded.

Results

Total 76 cases during the study were reviewed. Maximum patients in our study belonged to 1–3 year age group constituting 70% of the total sample and 87% of the study population was less than 5 years of age (Table 2). Overall study population constituted 47 (62%) males and 29 (38%) females.

Table 2 Age-wise distribution

	Frequency	Percentage
1–3 years	53	70
4–5	13	17
> 6 years	10	13

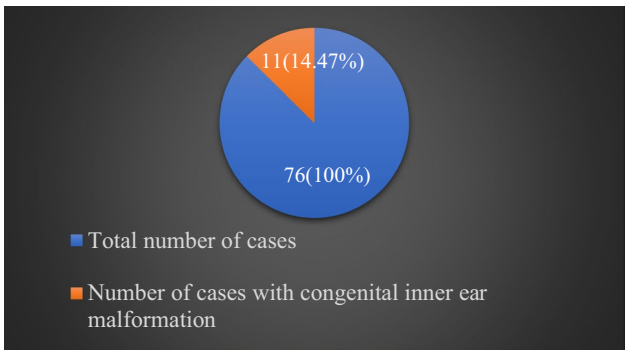

Fig. 1 Percentage of number of cases with congenital inner ear malformations

Malformations were identified in 11 cases (Fig. 1). 5 cases had Narrow IAC which was the most common anomaly followed by IP-2 and Globular vestibule (3). The EVA was seen in 2 patients, 2 had cochlear dysplasia, 1 had IP type 1and 1 patient had common cavity. Michel deformity was found in one case (Fig. 2). Some of these patients had multiple anomalies, so the total number of malformations was 18 (Figs. 3, 4, 5, 6, 7).

7 of these 11 patients with malformations had cochlear implant surgery in our hospital.

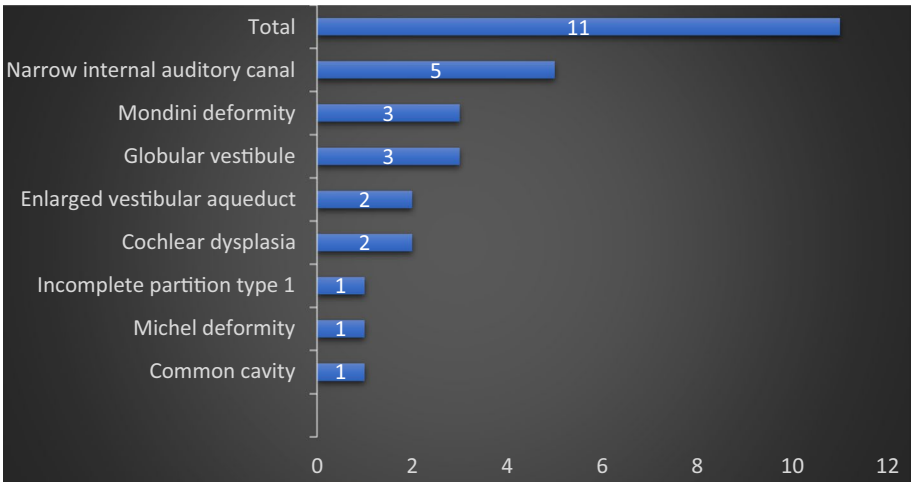

Fig. 2 Congenital inner ear malformations

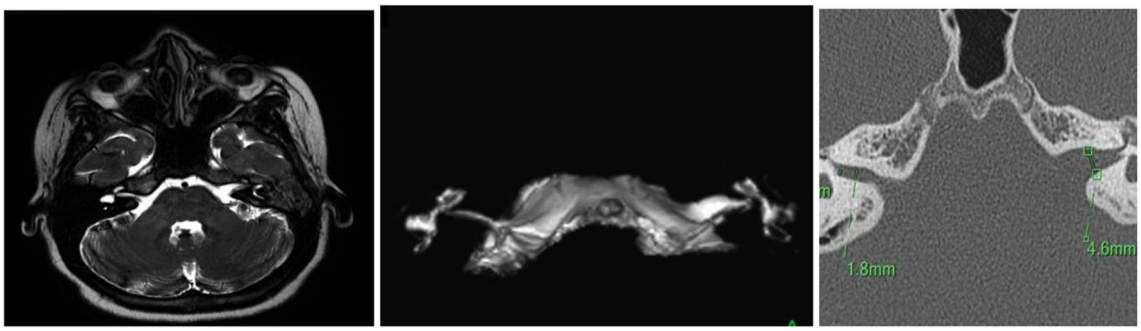

Fig. 3 A case of 4 years old male child with bilateral profound SNHL with right narrow IAC, associated with Hypoplastic auditory nerve

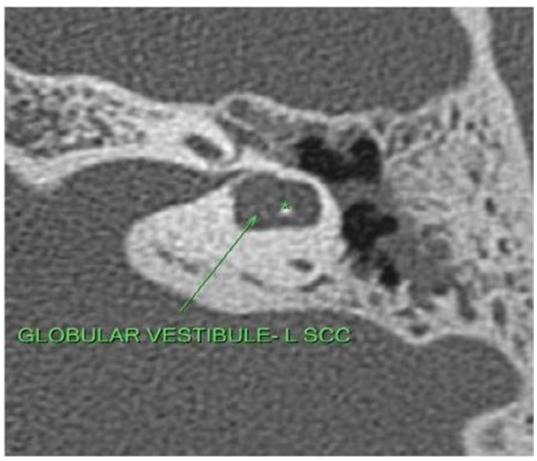

Fig. 4 8 year old female with bilateral profound SNHL showing Globular vestibule on the left side

Discussion

Cochleovestibular and other temporal bone anomalies are not uncommon. The prevalence of these anomalies has been reported by various studies to be ranging from 3 to 40%. The individual frequencies of various malformations have also been widely different in published literature. In our study, the incidence among prospective cochlear implant candidates with bilateral severe to profound SNHL has been 14.47% and the Narrow IAC is found to be the most common followed by globular vestibule and Incomplete partition type 2.

In the present series, 47 males and 29 females indicated no significant gender bias associated with Temporal bone anomaly. Most of our subjects were less than 5 years as shown by age distribution since the majority of our implantees were young children. The youngest was 1 year old and the oldest was 67 with only 10 cases above 6 years of age.

International literature shows a highly variable prevalence of Temporal bone anomalies. Mocan et al. [4] from Turkey in a study involving 481 cases found malformations in 187 (38%) with Incomplete partition being the most common (39%) followed by cochlear hypoplasia (8%). Aldhafeeri and Alnasi [5] in a paper published in 2016 noted 24 anomalies in 316 Cochlear implant candidates with EVA in 8 (33.3%) and Mondini deformity in 7 (29.1%), cochlear hypoplasia was seen in only 1 (4.1%).

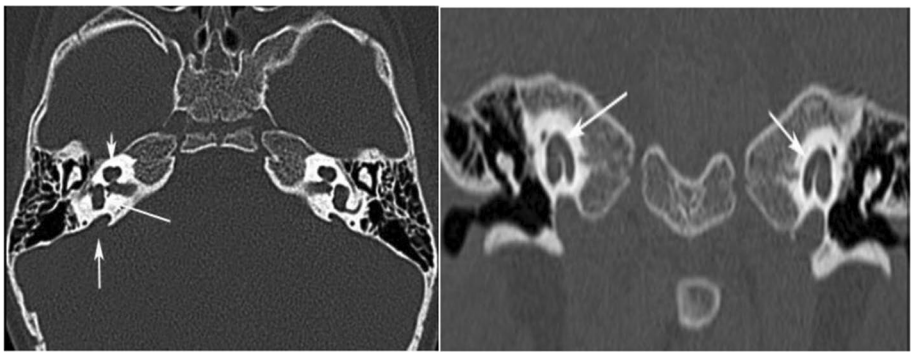

Fig. 5 8 year old female with bilateral severe SNHL showing IP type 2 anomaly. CT is showing a cystic apex with an enlarged vestibular aqueduct and 1.5 turns of cochlea

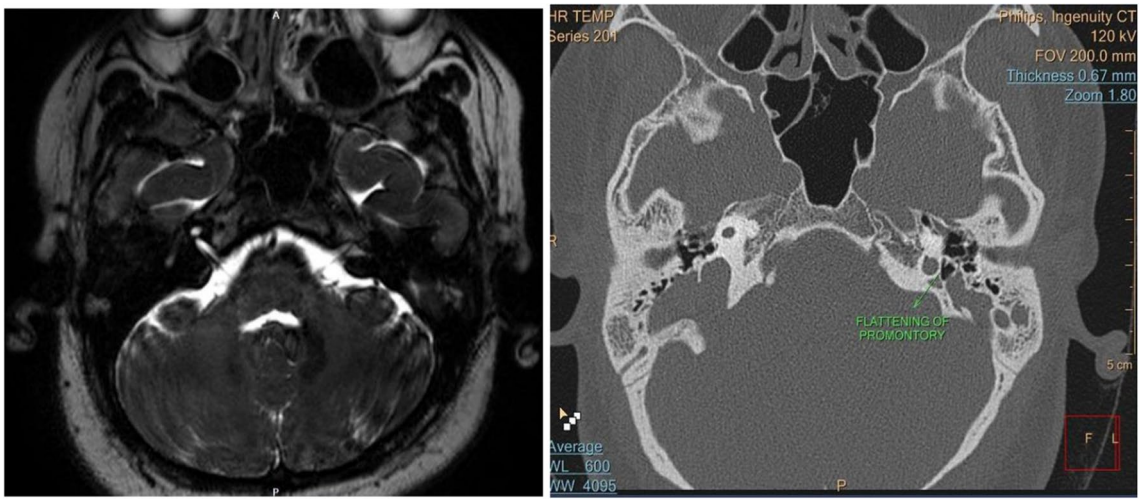

Fig. 6 18-year-old female with bilateral profound SNHL, a case of Labyrinthine aplasia (Michel deformity) on the left side. The patient was advised to go for ABI since the Cochlear Implant was contraindicated

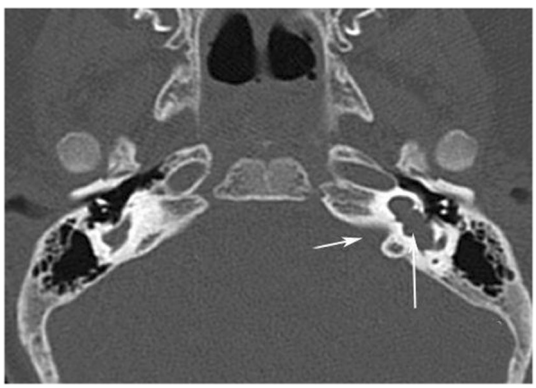

Fig. 7 A 2 year old male child with bilateral profound SNHL, case of Common cavity on the left side, there is no partition between vestibule and cochlea

In an article published in 2019, Klarov et al. [6] from Russia identified Inner ear malformations in 16 of 165 patients (9.7%). IP type 2 (34.3%) was common followed by IP type 3 (18.7%) and Internal auditory canal abnormalities (12.5%). Ahmed et al. [7] noted 10% as the frequency of Inner ear malformations (48/481) in cases with bilateral SNHL. In their series, complete labyrinthine aplasia was commonest followed by cochlear hypoplasia.

Higher prevalence of temporal bone malformation has been reported by Masuda and Usui [8] in 2019 (24.3%), Chinese study by Sun et al. [9] (30.69%).

An Indian study by Agarwal et al. [10] reported a prevalence of 14.28% which is very similar to the present series. In their series, Incomplete partition was the most common anomaly. whereas in the present study Narrow IAC was the commonest followed by IP type 2.S Masuda et al. [11] in 2013 reported a Narrow IAC, 32 (46.4%) as the most common anomaly like in the present study.

Many researchers focussed on cochleovestibular anomalies which did not include Internal auditory canal and nerve malformations, which explains the low reporting of Internal auditory canal anomalies. There is a wide

Table 3 Different studies and incidence of inner ear malformations

Study	Incidence of inner malformation	Common inner ear malformation
Mocan et al. [4], Turkey	187/481–38%	Incomplete partition Type 1–20% Incomplete Partition Type 2–19% Cochlear Hypoplasia-12%
Agarwal [10], India	40/280–14.28%	Incomplete partition type 2–41% Common cavity-20% Incomplete type 1–11.5%
Aldhafeeri and Alsanosi [5], Saudi Arabia	24/316–7.5%	Large Vestibular Aqueduct-33% Semi-circular canal dysplasia-33% Incomplete partition Type-29%
Klarov et al. [6], Russia	16/165–9.7%	Incomplete Type 2-34.3% Incomplete Type 3-18.7% Internal auditory meatus anomaly-12.5% Enlarged Vestibular Aqueduct-12.5%
Ahmed et al. [7], Pakistan	48/481–10%	Complete Labyrinthine aplasia-18.8% Semicircular canal dysplasia-8.3%
Present Study	11/76–14.47%	Narrow Internal Auditory canal-45% Globular Vestibule-27% Incomplete Partition Type 2–27%

variation in total prevalence (3–40%) as well as frequencies of individual malformations (Table 3).

Conclusion

CT and MRI play an important role in the pre-op assessment of Cochlear Implant candidates. This helps in candidate selection, predicting intra op problems, choosing appropriate surgical techniques and electrode array. It also helps to counsel parents regarding prognosis.

The incidence of Temporal bone malformation in the present study is 14.47% with the Narrow IAC as the most common deformity followed by Incomplete partition type 2 and Globular vestibule. The reported prevalence of temporal bone malformation and frequency of individual anomalies is widely variable. Further studies with a larger sample size are needed to establish the prevalence of Temporal bone malformations in our population.

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Declarations

Conflict of interest All authors declared that have no conflict of interest.

Ethical approval Ethical approval was obtained by institutional ethics committee.

References

1. Jackler RK, Luxford WM, House WF (1987) Congenital malformations of the inner ear, a classification based on embryogenesis. Laryngoscope 97:2–14
2. Sennaroglu L, Saatci I (2002) A new classification for cochleovestibular malformations. Laryngoscope 112:2230–2241
3. Sennaroglu L, Bajin MD (2017) Classification and current management of inner ear malformations. Balkan Med J 34(5):397–411
4. Mocan BO, Sanverdi E, Orhan N, Sarac S, Sennaroglu L (2011) Prevalence and relative frequencies of congenital inner ear abnormalities in patients evaluated with temporal bone CT for sensorineural hearing loss Type: Scientific paper. European Soc Radiol 1262:1–13.1
5. Aldhafeeri AM, Alsanosi AA (2016) Prevalence of inner ear anomalies among cochlear implant candidates. Saudi Med J 37(10):1096–1100
6. Klarov LA, Barashkov NA, Teryutin FM, Romanov GP, Popov MM, Solovev AV, Luginov NV, Tobokhov AV, Kotlyarov PM (2020) The spectrum and frequency of inner ear anomalies in patients with congenital hearing impairment in Yakutia. J Radiol Nucl Med 101(2):90–102
7. Ahmed J, Saqulain G, Khan MIJ, Kausar M (2020) Prevalence & features of inner ear malformations among children with congenital sensorineural hearing loss: A Public Cochlear Implant Centre Experience. Pak J Med Sci 36(7):1511
8. Masuda S, Usui S (2019) Comparison of the prevalence and features of inner ear malformations in congenital unilateral and bilateral hearing loss. Int J Pediatr Otorhinolaryngol 125:92–97
9. Sun B, Dai P, Zhou C (2015) Study on 2,747 cases of inner ear malformation for its classification in patient with sensorineural hearing loss. Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi 29(1):45–47
10. Agarwal SK, Singh S, Ghuman SS, Sharma S, Lahiri AK (2014) Radiological assessment of the Indian children with congenital

sensorineural hearing loss. Int J Otolaryngol 58:55898. https://doi.org/10.1155/2014/808759
11. Masuda S, Usui S, Matsunaga T (2013) High prevalence of inner-ear and/or internal auditory canal malformations in children with unilateral sensorineural hearing loss. Int J Pediatr Otorhinolaryngol 77(2):228–232