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ROLE OF IMAGING IN CHILDREN WITH PROFOUND HEARING LOSS TO ESTABLISH CANDIDATURE OF COCHLEAR IMPLANT

Radiology

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ABSTRACT

Introduction: Sensory neural hearing loss is major cause of deafness which can be treated with cochlear implant . Evaluation of Inner ear structural anomaly on HRCT while 7th -8th nerve complex and cochlea on MRI help to decide candidacy for cochlear implant **Methodology:** The study was conducted prospectively from June 2023 to may 2024 at our institute , which is a tertiary care centre in Gujarat, - designated centre for cochlear implant surgery. Children of deafness in the outpatient department and full filling the inclusion criteria were included in the study. HRCT temporal bone and MRI cochlea with 7th and 8th nerve was done with CSIS and Space sequences. **Result:** We examined total 100 with congenital SHL. The number of abnormal findings were in 18 cases , out of these - 8 cases could be diagnosed on HRCT and rest with help of MRI . It was observed that Cochlear anomaly was the most common cause of congenital SNHL and nerve complex anomaly was least. **Conclusion:** With timely and appropriate use of imaging , it is possible to diagnose earliest and with cochlear implant , we can provide better quality of life and achieve all mile stones timely.

KEYWORDS

Cochlea, High resolution CT, Temporal, Sensory neural hearing loss, Cochlear implant.

INTRODUCTION

Interrupted development of the inner ear during the first trimester is the principal cause of Sensory neural hearing loss in children and of childhood disability, with the estimated prevalence is one in 2000 neonates and 6 in 1000 children presented with SNHL by the age 18[1].

With awareness and availability of resources now up to 1 years of age, most of the Cases are diagnosed and identified. Hearing has a significant role in in various aspects of brain 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, but unfortunately it can not be diagnosed on fetal ultrasound.

With the advancement in treatment 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 as an effective modality to provide hearing for congenital severe to profound SNHL[3,4].

For 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, where MRI is used to look in the membranous labyrinth, vestibulocochlear nerve-free fluid spaces, and any other associated brain pathology[1,2]

Inner Ear Anatomy:

The inner ear includes the membranous labyrinth within the bony labyrinth (otic capsule). **Membranous labyrinth contains** the utricle, saccule, cochlear duct and the membranous channels within the semicircular canals and the endolymphatic duct.

Bony labyrinth contains the cochlea, the vestibule, the semicircular canals and cochlear and vestibular aqueducts; these structures can be well demonstrated by high resolution CT scan and MRI.

Congenital abnormalities of inner ear:

Sennaroglu classifications[5]

1. Michel deformity (complete labyrinth aplasia)
2. Rudimentary otocyst.
3. Cochlear aplasia
4. Common cavity
5. Cochlear hypoplasia
6. Incomplete partition of cochlea
7. Enlarged vestibular aqueduct

Michel Aplasia: Also known as complete labyrinthine aplasia, rare anomaly refers to the total aplasia of the inner ear. [5]Michel aplasia may be associated also with skull base and vascular anomalies like . petrous bone hypoplasia, platybasia and aberrant jugular bulb veins[6]

Cochlear Aplasia: complete absence of the cochlea is a rare anomaly . Dense otic bone is seen at the anatomical site of the cochlea. Cochlear nerve canal and cochlear nerve are absent. Cochlear promontory is hypoplastic and flattened[7]

Common Cavity Malformation: Absence of the normal differentiation between the cochlea and vestibule replaced by cystic structure[8]. Confluence of the cochlea, vestibule and horizontal SCC in a cystic cavity variable in size with no internal architecture[9].

Incolmplete Partition Type 1[10]: Also called cystic cochleovestibular malformation.

Features include absent modiolus, absent interscalar septum and wide (most common) or normal cochlear nerve canal.

Incomplete Partition Type Ii (classic Mondini Malformation): The Mondini abnormality consists a triad[11]. Abnormal cochlea with Only 1.5 turns (instead of the normal 2.5 turns), Enlarged vestibule with normal semicircular canals , Enlarged vestibular aqueduct.

Enlarged Vestibular Aqueduct Syndrome: Enlargement of the vestibular aqueduct is the most frequently seen osseous abnormality in patients with SNHL. A commonly accepted imaging criterion for an enlarged vestibular aqueduct is a calibre of 1.5 mm or greater in the midportion of the aqueduct. Enlargement of the vestibular aqueduct may also be suspected if the aqueduct is larger in calibre than the normal lateral semicircular canal[12].

7. Superior Semicircular Pathology: Semicircular Canal Dysplasia : Dysplasia of the lateral SCC is a common type of inner ear malformation. On imaging , short, broad cystic space confluent with the vestibule seen.

Semicircular Canal Aplasia : SCC aplasia is less common than SCC

dysplasia. It is usually associated with cochlear anomalies.[13]

Internal Auditory Canal: The internal auditory canal should be symmetrical in any one person. Difference in size of more than 2 mm as compared to opposite side is significant. The facial nerve and vestibulocochlear nerve passes through it.

Methodology

This prospective study was conducted at secondary care centre in Gujarat , which is designated centre for cochlear implant surgery. All children attending the outpatient department with hearing loss in the age group of 1 to 7 years were screened, and those who had severe to profound sensory neural hearing loss, greater than 90 dB were elected as subjects. A total of 100 congenital SNHL patients were selected as a subjects for the study. Consent of Parents and guardians of the children were explained clearly about the study and those who were willing to participate in their consent were recorded on a consent form, duly signed by them. A total of 100 such children were included in the study. Children above seven years of age with middle year or tympanic membrane pathology or brain abnormality where excluded from our study. HRCT and MRI of the temporal bone were done with 0.75 mm slice thickness and axial , coronal and saggital reconstruction for all the patients. Image were analysed for the presence of normal structure as well as any anomalies present in the inner ear.

OBSERVATIONS AND RESULT

The total number of children analysed was 100 with congenital SNL. The number of radiological abnormal cases with one or more anomalies as per high resolution computed tomography and MRI temporal bone along with congenital SNHL was 18.

Out of a total of 18 children, males were 10 and females were 8. Out of all inner ear anomalies observed, it was noticed that most common nearly 55% was cochlear anomaly and least common 7.5 % of facial nerve anomaly.

Out of 10 cochlear abnormality , 4 of cochlear hypoplasia, 2 were cochlear aplasia , 3 were common cavity and 1 were of cystic cochlea.

Among various abnormalities of vestibule, 5 had dilated vestibule, 2 had hypoplastic vestibule and 1 had cystic vestibule.

In Lateral semicircular canal anomalies , most of the children had lateral semicircular canal, abnormality, among which 1 had absent LSCC and 2 had hypoplastic LSCC.

2 patients were present with PSCC and superior SCC abnormalities. 5 patients had Internal auditory canal anomalies. 82 % of cases do not have an abnormality.

Associated brain abnormality was seen in 3 cases which predicts poor outcome after cochlear implantation

Table 1 Various inner ear abnormality observed.

Abnormality	Number of patients	Percentage
Cochlear abnormality	10	55%
Vestibular abnormality	8	42 %
Vestibular aqueduct	7	40%
Semicircular canals	7	37 %
Cochlear nerve	7	37 %
Fascial nerve	1	7.5 %
Internal auditory canal	5	30%

Table 2 Distribution of patients according to the cochlear abnormality

Cochlear hypoplasia	4	40 %
Cochlear aplasia	2	20 %
Cochlear dysplasia	0	0 %
Common cavity	3	30 %
Cystic cochlea	1	10%

Table 3 Distribution of patients according to the vestibular abnormality

Dilated vestibule	5	62 %
hypoplasia	2	25 %
Cystic	1	12.5 %
Absent	0	0%

Table 4 Lateral Semicircular canals

Absent	1	33%
Hypoplastic	2	66%
Dilated	0	00%

CONCLUSION

With timely use of imaging , we can clearly delineate inner ear malformation which can further guid us in treatment planning and management of children wit profound hearing loss. Both Imaging studies are used together to precisely delineate inner ear malformations. Although it is beneficial to understand the site and cause of hearing loss, this still has no prognostic or preventive value.

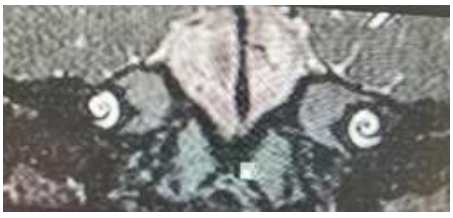

Figure 1. MRI CISS image show normal cochlear 2.5 turns

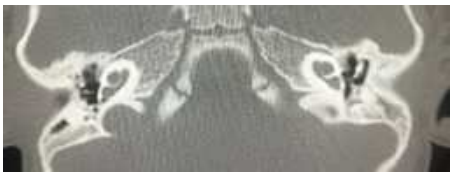

Figure 2. HRCT Temporal bone, cochlea shows 1.5 turns with incomplete bony partition between middle and apical turn forming cystic apex and dilated vestibule – Incomplete partition defect

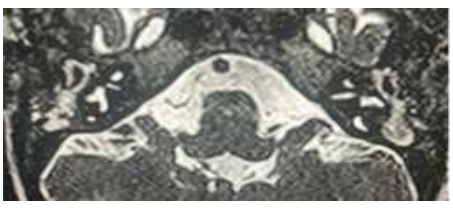

Figure 3. MRI CISS image show hypoplastic bilateral lateral semicircular canal

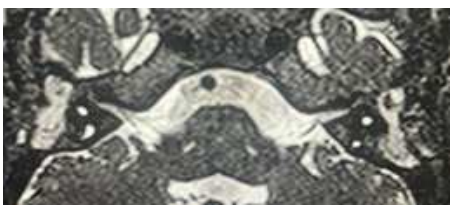

Figure 4. MRI CISS image shows narrowing of bilateral internal auditory canal

Figure 5: MRI CISS image shows right sided mondini dysplasia

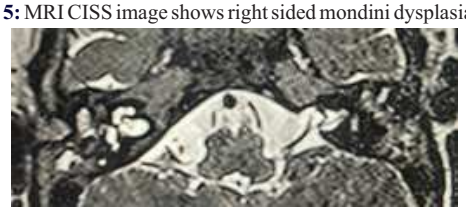

Figure 6. MRI CISS image shows complete labyrinthine aplasia with mild dilatation of IAC on left side

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