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THE STUDY OF HRCT AND MRI TEMPORAL BONE FINDINGS IN PEDIATRIC PATIENTS WITH CONGENITAL SENSORINEURAL HEARING LOSS

Radiodiagnosis

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ABSTRACT

Introduction -The incidence of Congenital Sensorineural hearing loss (SNHL) is approximately 1: 1000 live births. SNHL is either due to disorders of the inner ear or cochleovestibular cranial nerve. Radiological evaluation is necessary to detect or rule out causes of SNHL. Also, the treatment of SNHL is predominantly determined by the etiology of hearing loss.

Aim & objective- Radiological assessment of various congenital inner ear malformations in pediatric age group patients with sensorineural hearing loss in a tertiary care centre.

Material & Methods-This is a prospective study conducted between 1 January 2018 and 1 June 2019 in Department of Radiodiagnosis, SAIMS, Indore & included all paediatric patients (93 children), who came for HRCT and MRI temporal bone imaging with the clinical diagnosis of congenital sensorineural hearing loss (SNHL) / for evaluating congenital sensorineural hearing loss (SNHL).

Results- In our study out of 93 paediatric patients , no significant radiological abnormality were detected in 68 patients (73.11%), however, 25 patients (26.88%) had various congenital anomalies of the inner ear and vestibulocochlear nerve.

Most commonly affected organ was cochlea. Among these 25 patients only cochlea was involved in 7(28%), both Cochlea and semicircular canal in 4(16%), and Cochlea and vestibular aqueduct in 1 patient(4%). Isolated vestibular aqueduct dilatation was found in 8 (32%) patients. Isolated semicircular canal involvement and cochleovestibular nerve abnormality were seen in 3 (12%) and 2 (8%) patients respectively.

Conclusion- In this study, imaging has helped us to detect various inner ear malformations in children with congenital sensorineural hearing loss. It is helpful for preoperative planning and preparation for cochlear implant in a tertiary care centre.

KEYWORDS

Temporal bone imaging, inner ear malformations, congenital deafness

INTRODUCTION

The incidence of Congenital sensorineural hearing loss is approximately 1: 1000 live births, one of the most common birth defects [1]. Sensorineural hearing loss (SNHL) is either due to disorders of the inner ear or retrocochlear diseases. Radiological evaluation (HRCT and MRI imaging) is necessary to detect or rule out causes of SNHL, that may be congenital, inflammatory, infectious or tumour pathology. In all patients posted for cochlear implantation, HRCT and MRI of the temporal bones are baseline investigations, necessary for preoperative planning and parental councilling. MRI provides accurate information of the membranous labyrinth and the cranial nerves. Evaluation of facial nerve and internal auditory canal (IAC) is better done on HRCT; however the presence of nerves in the IAC and cochlear fluids can be diagnosed on MRI [2].

The membranous malformations are present in approximately 80% of the cases of congenital sensorineural hearing loss and the pathology is at the cellular level. However inner ear malformations are present approximately only 20% of the cases of congenital sensorineural hearing loss [3].

Malformations of the Inner Ear -

The inner ear congenital malformations may be considered in two broad categories (5):

(a) Only membranous labyrinth malformations
(b) Malformations involving both the osseous and the membranous labyrinth (malformed otic capsules).

Membranous Labyrinth Malformations

The classification of these abnormalities are not radiologically useful, as their differentiation requires histopathologic examination. (5)

Malformations involving both the Osseous and the Membranous Labyrinth

The radiological diagnosis of these malformations significantly helps in both the clinical management and the prognosis (6).

Congenital anomalies of Vestibule and cochlea can be classified as follows.

(1) Complete labyrinthine aplasia (Michel deformity): there is complete absence of all cochlear and vestibular structures.
(2) Cochlear aplasia: the cochlea is completely absent.
(3) Common cavity deformity: there is a cystic cavity representing the cochlea and vestibule but without showing any differentiation into cochlea and vestibule.
(4) Cochlear hypoplasia: the cochlea and vestibule are separate from each other but their dimensions are smaller than normal.
(5) Incomplete partition type I: the cochlea is lacking the entire modiolus and cribriform area resulting in a cystic appearance. This is accompanied by a large cystic vestibule.
(6) Incomplete partition type II (Mondini deformity): the cochlea consists of 1.5 turns in which the middle and apical turns coalesce to form a cystic apex, accompanied by a dilated vestibule and enlarged VA.
(7) Vestibularmalformations: They include Michel deformity, common cavity, absent vestibule, and dilated vestibule.
(8) Semicircular canal malformations: They are absent, hypoplastic, or enlarged.
(9) Internal auditory canal malformations: They are absent, narrow, or enlarged.

Cross sectional Radiological modalities gives informations regarding the type of malformation of middle ear, inner ear (Vestibule and cochlea) as well as also about the presence or absence of the vestibulocochlear nerve

Aim & objective - Radiological assessment of various congenital inner ear malformations in pediatric age group patients with sensorineural hearing loss in a tertiary care centre.

MATERIALAND METHODS

This was a prospective study conducted between 1 January 2018 and 1 June 2019 in department of Radiodiagnosis in Sri Aurobindo Institute of Medical Sciences, Indore and included all paediatric patients (93 children), came for HRCT and MRI temporal bone imaging with clinical diagnosis of congenital Sensorineural hearing loss (SNHL).

Inclusion criteria: Children who were congenitally deaf with clinical diagnosis of SNHL were included in study after taking consent from the parents.

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Exclusion criteria
Children with conductive and acquired causes of deafness were excluded from the study.

Clearance from the ethical committee was taken.

Imaging Protocol
HRCT
HRCT scans are performed on a True 64-slice volume CT scanner (Somatom definition AS, Siemens) in a straight axial plane. Axial CT are acquired in 0.6mm slice thickness and reconstructed into magnified axial and coronal images, 0.3mm interval, using bone algorithm

MRI
MRI scans are performed on 1.5-T MR machine (Siemens Magnetom Symphony 1.5T MRI System with 18-channel). Sedation is used in most of the patients. MRI was done using axial 3D SPACE sequence with slice thickness 0.5mm for cochlear nerve and inner ear structures. A routine Axial T2 FLAIR sequence was also obtained in all patients for screening of the brain.

RESULTS -
In our study out of 93 paediatric patients, no significant radiological abnormality were detected in 68 patients (73.11%), however 25 atients (26.88%) had various congenital anomalies of inner ear and vestibulocochlear nerve.

Total 66 females were having SNHL out of 93 children, among these 9 female cases were having abnormal radiological findings, however 47 female cases were with normal radiological findings. Only 37 cases were male, among these abnormal radiological findings were found in 16 cases and no radiological findings in 21cases.

Table / Figure -1

Ear malformations	Patients (Percentage)
semicircular canal	*3(12%)*
Isolated cochlea	*7(28%)*
Cochlea+ semicircular canal	*4(16%)*
Cochlea+ vestibule aqueduct	*1(4%)*
vestibule aqueduct	*8(32%)*
Cochlear nerve	*2(8%)*
Total	*25*

Out of these 25 patients, most common anomaly was cochlear anomaly in 11 patients, followed by anomalous vestibule aqueduct dilatation in 9 patients (isolated - 8=32%). Abnormal isolated semicircular canal and cochleovestibular nerve were seen in 3 (12%) and 2 (8%) patients respectively.

DISCUSSION-
In our study, total 26.88% of cases had inner malformations which is approximately same incidence as in other studies like 20% (Sennaroglu and Saatciin2001 [7]), 23% (Abdullah et al. in 2003 [8]), 30% (Ma et al. in 2008 [9]) and 31% (McClay et al. in 2008 [10]), however there was lower incidence (14.2%) in Indian study conducted by Agarwal S K et al 2014(11).

Multiple malformations of the inner ear were seen in our study in congenitally sensorineural hearing loss patients. Latest classification of congenital inner ear malformations were given by Sennaroglu and Saatci in 2002 [7].

In our study we found following malformations involving both the Osseous and the Membranous Labyrinth –

Complete Labyrinthine Aplasia (Michel aplasia) —
this is the most severe form of inner ear deformity. This malformation is very rare; only 1 out of 25 patients (4%) had this inner ear malformation (Table/Figure-2). However in most of the studies its incidence was approximately 1 % (12).

It occurs as the results of developmental arrest of the otic placode in 3rd week of fetus (13–16).

Other associated abnormalities - Hypoplasia of the petrous bone, absence of the round and oval windows, and flattening of the medial wall of the middle ear cavity are characteristic features. The characteristic finding of complete labyrinthine aplasia is flattened medial wall of the middle ear. This helps in its differentiation from labyrinthitis ossificans (15, 16).

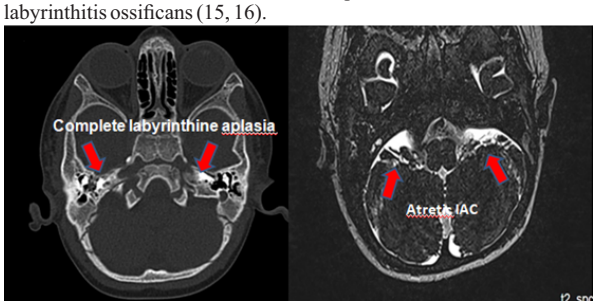

Table / Figure -2 Complete labyrinthine aplasia (internal auditory canal - IAC)

Common cavity cochlear malformation -
In our study we found 2 patients out of 25 patients (8%) with Common cavity cochlear malformation malformation on either side. In Agarwal SK et al 2014 its incidence was around 20.5%.(11).

In this malformation there is absence of the normal differentiation between the cochlea and vestibule. Most of the time with this malformation, the semicircular canals are also malformed, but sometimes may be normal. A developmental arrest in the 4th week of fetal life results this type of malformation (17, 18).

In figure CT and MR images show a common cavity cyst, which is formed by confluence of the cochlea and vestibule with no internal architecture (Fig3).

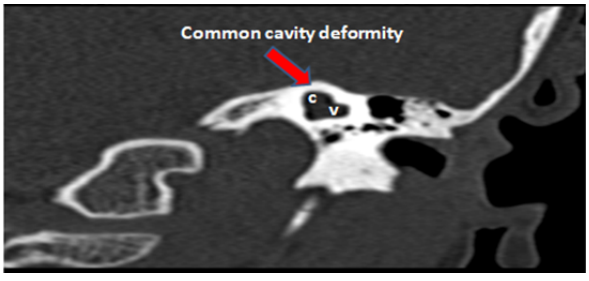

Table / Figure -3 Common cavity cochlear malformation (cochlea –C, vestibule –V)

Type II incomplete partition cochlear malformation-
Incomplete partition type-II (Mondini deformity) was found in 5 among 25 (41%) malformed patients. In other studies IP-II incidence was 15% (Sennaroglu and Saatci in 2002 [7]), 22% (Westerhof et al 2001 [19]) and 11.5% (Agarwal SK et al 2014). (11).

There is approximately 1.5 turns of middle and apical turns of cochlea coalesce to form a cystic apex (fig-5), however basal cochlear turn appears normal. This is associated with a dilated vestibule and enlarged vestibular aqueduct. This is the most common type of cochlear malformation.

Most of the time, this type of malformation has been described as a triad- a cochlea with cystic apex and a normal basal turn, enlarged vestibular aqueduct and vestibule and normal semicircular canals. Distinct scalae tympani and vestibuli, the interscalar septal defects and the absence of the osseous spiral lamina findings are seen on MRI inner. Type II incomplete partitition occurs due to a developmental arrest in the 7th week of fetal life (7,13,18).

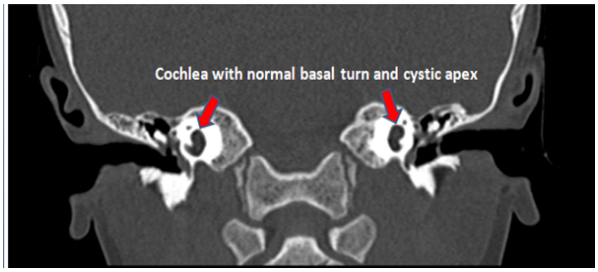

Table / Figure -4 Type II incomplete partition cochlear malformation(Mondini deformity)

Cochlear hypoplsia- this malformation was present in 2 patients out 25 patients(8%). In this CT and MR images shows a small cochlea in the form a small bud arising from the vestibule. The cochlea may have only one turn or a partial turn. The vestibule and semicircular canals may or may not be normal (7–9). Cochlear hypoplasia occurs usually during the 6th week of fetal life (7, 13,14).

Vestibular aqueduct / Endolymphatic duct and sac malformation- In 9 patients (36%) dilated vestibular aqueduct, or a dilated endolymphatic duct and sac were found, however 8 patients were having isolated findings. One patient had dilated endolymphatic duct and sac, along with cochlear malformations.(fig-6). In most of the patients it is bilateral. However in Agarwal SK et al 2014 it's incidence was high (56.4%) (11).

In most of the patients (~ 90%) it is bilateral, but may be asymmetric. Also it is slightly more common in females.

Enlargement of the osseous vestibular aqueduct is seen on HRCT; however at MR imaging enlargement of the endolymphatic duct and sac is diagnosed (17).

If the diameters of the endolymphatic duct and sac exceed diameter of the adjacent ascending portion of the posterior semicircular duct (diameter of approximately 1.5 mm) on thin-section T2-weighted gradient-echo images, than these are usually considered enlarged. However on HRCT, the vestibular aqueduct is diagnosed as dilated, when its diameter, midway between the common crus and the external aperture is greater than 1.5 mm.

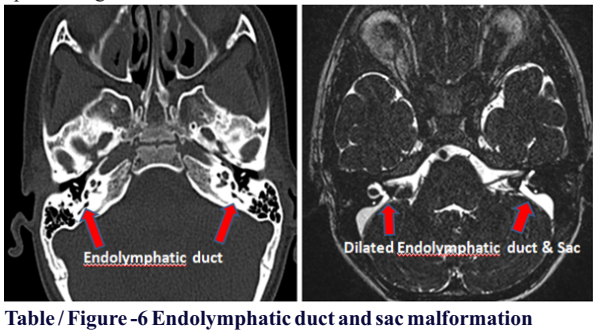

Table / Figure -6 Endolymphatic duct and sac malformation

Cochlear Nerve Anomalies—
In our study we found isolated 2 cases out of 25 cases(8%) with one case associated with Complete Labyrinthine Aplasia (Michel aplasia) had cochlear nerve anomaly. However there was 29.4% incidence of vestibulo-cochlear nerve anomalies in study of Agarwal SK et al 2014(11).

Four major nerves of the IAC - the facial, cochlear, superior vestibular, and inferior vestibular nerves are best seen on MR Oblique sagittal images in a plane perpendicular to the long axis of the IAC (20).

The completely normal IAC and labyrinth may not rule out absence nerve (21). That's why, MR imaging is necessary for accurate assessment of the cochlear nerve.

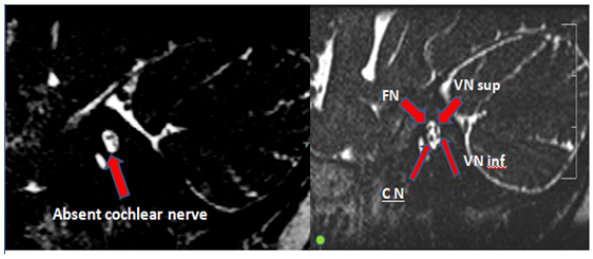

Table / Figure-7 Absent Cochlear Nerve (CN- cochlear nerve, FN- facial nerve, VN- vestibular nerve, sup.- superior , inf.- inferior)

Semicircular Canal Anomalies – Total 7 (26.9%) out of 25 malformed inner ears have semicircular canal anomalies. In some patients semicircular canals were found to be aplastic or hypoplastic. In 4 patients (16%) Semicircular Canal Anomalies are also associated cochlear malformations; however isolated semicircular canal involvement was seen in 3 patients.

In extensive malformations, the canals (one or more) may be completely absent as in our case (table/Figure-4). However, Dysplasia of the semicircular canals is far more common than aplasia. Semicircular canal anomalies may be associated with CHARGE syndrome (coloboma, heart anomalies, choanal atresia, retardation of growth and development, and genital and ear anomalies- Absence of all semicircular ducts), and Waardenburg syndrome as well as Alagille syndrome (Isolated aplasia of the posterior semicircular duct) (22).

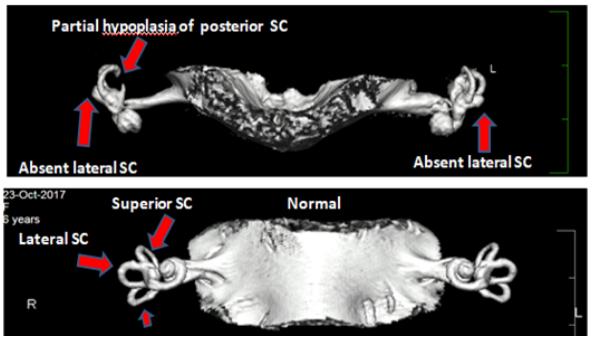

Table / Figure -5 Semicircular Canal (SC) Anomalies

Cochlear aplasia and Type I incomplete partition cochlear malformation
In our study we did not get any patient having radiological findings of Cochlear aplasia as well as Type I incomplete partition cochlear malformation.

Vestibular anomalies - isolated Vestibular malformation is rarely seen (13, 17).

LIMITATION:

1) In few patients cochlear nerve was not well evaluated on MRI in cases of the nerve is too slender to discern or because its fibers parallel those of the vestibular nerve and are inseparable from it and in these patients —may not do well after cochlear implantation and should undergo evaluation by an experienced audiologist before the decision about implantation was made(22,23)
2) In those cases where the cochlear nerve is not seen on MRI, it is possible that the nerve is so thin that it is beyond the resolution of a 1.5 T MRI scanner; alternatively, it may be that the cochlear nerve fibers traverse along the vestibular nerve and hence are not detected on MRI. These patients should undergo a hearing aid trial and periodical audiological evaluation by expert audiologists/therapists before a decision is taken on their candidacy for cochlear implant surgery.
3) CT has limitation for the identification of an enlarged vestibular aqueduct and MRI is gold statndard for the same

CONCLUSION

Inner ear malformations are found in 20-30 % of patients with severe or profound sensorineural hearing loss. HRCT and MRI are baseline investigations that need to be done prior to cochlear implant surgery. Both these modalities provide exquisite anatomical details and information. HRCT& MRI are complementary modalities. Preoperative HRCT offers the advantage of visualizing the bony structures as well as any coexistent middle or external ear anomalies and important anatomic variants. On the other side MR imaging provides definitive information about the integrity of the cochlear nerve and the fluid-filled spaces of the inner ear. Both modalities together provide a detailed view of the morphology of the inner ear and the operative field to the surgeon. Identification of inner ear malformations help in preoperative planning of cochlear implant patients. These have a significant impact on cochlear implant surgery and its outcome. These radiological findings may alert the surgeon to possible complications.

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