ORIGINAL RESEARCH ARTICLE

Role of CT & MRI in Evaluating Etiology of Congenital Hearing Loss

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How to cite this article: Mythri Nag, Shyam Shankar, Chandan Giriyappa, Venkatesha B K. Role of CT & MRI in evaluating etiology of congenital hearing loss. International Journal of Contemporary Medicine Surgery and Radiology. 2022;8(1):A12-A14.

A B S T R A C T

Aims and objectives: To identify and analyze diagnostic yield of CT and MRI in detecting abnormalities of the external ear, middle ear, inner ear and cochlear nerve in cases of congenital hearing loss.

Material and methods: The study design was a two year observational clinical correlation study to compare the detection rates of specific diagnostic findings between CT and MRI. The imaging features and final diagnosis were documented in a structured case record form, separately.

Results: Overall sensitivity of detection of abnormalities in CT & MRI is 44.44%, specificity is 37.50%, Positive predictive value is 28.57%, Negative predictive value is 54.55% and accuracy is 40.00%. CT & MRI provides additional information in the diagnosis of patients with congenital hearing loss.

Conclusion: HRCT Temporal bone and MRI are complementary to each other and provide exquisite vital anatomical details and information. Hence they are now considered as baseline investigations and are mandatory prior to definitive surgery.

Keywords: Hearing Loss, Sensory Neural Hearing Loss, Conductive Hearing Loss, HRCT Temporal Bone.

INTRODUCTION

Congenital hearing loss is a major cause of childhood disability worldwide. Hearing loss can be classified as conductive hearing loss (CHL), sensorineural hearing loss (SNHL), or mixed hearing loss (MHL). Disruption of the transmission of sound waves from the outside environment to the cochlea will result in CHL. It can be caused by a range of developmental, congenital or acquired pathology to the external or middle ear.

Sensorineural hearing loss (SNHL) refers to deafness secondary to conditions affecting the inner ear, internal acoustic canal, cerebellopontine angle, and or vestibulocochlear nerve. Congenital SNHL is one of the most common birth defects with incidence of approximately 1:1000 live births.[1] Most inner ear malformations arise when formation of the membranous labyrinth is interrupted during the first trimester of pregnancy.[2] This may be due to inborn genetic error or as a result of teratogenic exposure during the period of inner ear organogenesis between the fourth and eighth week of gestation.

Aims and objectives

1. To identify and analyze diagnostic yield of CT and MRI in detecting abnormalities of the external ear, middle ear, inner ear and cochlear nerve in cases of congenital hearing loss.
2. To compare the detection rates of specific diagnostic findings (cochlear anomalies, enlarged vestibular aqueduct, cochlear nerve abnormalities, and brain findings between CT and MRI.

Joseph Y. Young et al (2014) performed a study on Preoperative Imaging of SNHL in Paediatric Candidates for Cochlear Implantation and concluded that MR imaging may depict relevant abnormalities of the membranous labyrinth CT is particularly helpful for evaluating the bone mastoid and middle-ear.[15]

MATERIALS AND METHODS

The study was conducted in the department of Radio-diagnosis, Shri Shamanur Shivashankarappa medical science and research center, Davangere over a period of two years from September 2019 to September 2021.

Study Design

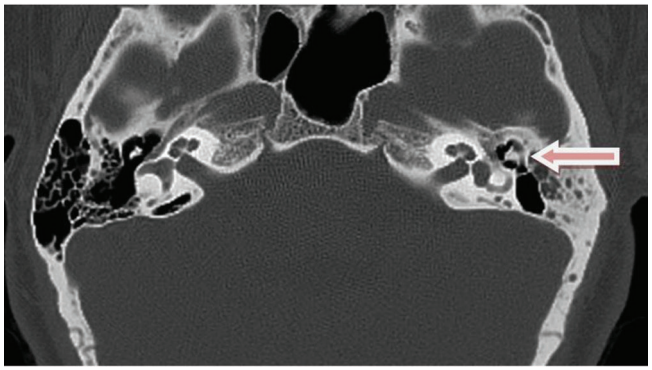
The study design was a two year observational clinical correlation study to compare the detection rates of specific diagnostic findings (cochlear anomalies, enlarged vestibular aqueduct, cochlear nerve abnormalities, and brain findings) between CT and MRI.

Study Period and duration:

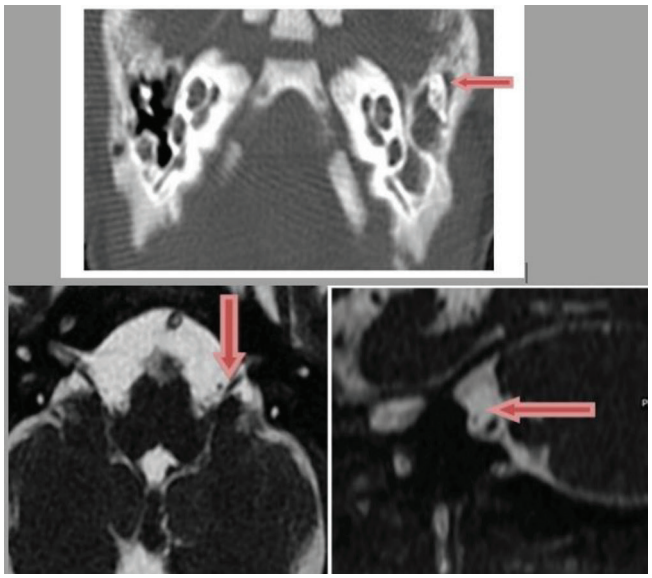
Source of data

All patients with clinical congenital hearing loss referred from Department of ENT at SSIMS and RC hospital, Davangere, after satisfying the inclusion criteria after undergoing audiological evaluation (brainstem evoked response audiometry and /or oto-acoustic Emission) will be studied after obtaining written informed consent.

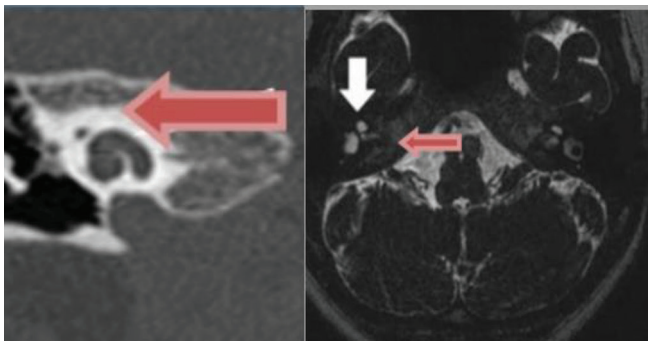
Sample size: A total of 25 patients fulfilling the selection criteria were studied.



Case-1: Left microtia, EAC Atresia, malformed ossicles. poorly pneumatised mastoid and normal inner ear



Case-2: Left anotia with EAC atresia and facial nerve aplasia

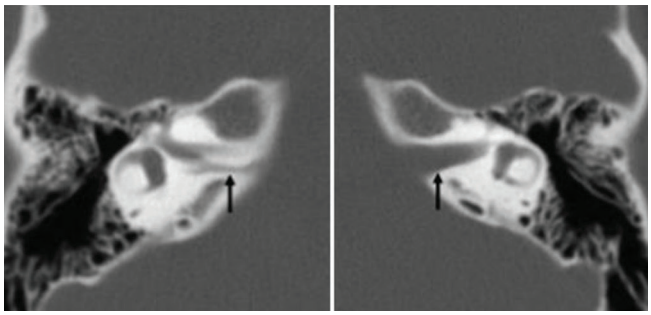


Case-3: Mondini dysplasia; Cochlea showing less number of coils

Type of study: Prospective study

Inclusion criteria

1. Age-0 to 15 yrs
2. Congenital hearing loss
3. Children with unilateral/bilateral hearing loss.
4. Children with mixed hearing loss.
5. Children with CHL.
6. Hearing evaluation suggestive of severe to profound SNHL.



Case-4: Narrow IAC; Infero-posterior portion of right internal auditory canal is smaller than contralateral side

Exclusion criteria

1. No definite evidence of congenital onset.
2. Sudden onset hearing loss.
3. Children with known history of trauma and infections preceding hearing loss.

Data collection

Once a patient fulfilled the inclusion criteria for this study he / she was administered the predesigned / pretested proforma. Demographic characteristics of the study population such as age, sex were obtained through an interview.

Confirmation and categorization of hearing loss was done with pure tone Audiometry (PTA), Brainstem Evoked response Audiometry (BERA) and/ or Otoacoustic Emissions (OAE). Patients will be undergoing CT and MRI of the bilateral temporal bones.

HRCT of the temporal bone of all cases was taken in both axial and coronal section. In all cases, 0.625 mm thickness contiguous axial sections in bony algorithm with 1mm thickness contiguous axial and coronal reformations. The following observations were made on both sides on CT: a) Pinna, b) External auditory canals (EAC), c) Middle ear cleft, d) Ossicles, e) Pneumatisation of Mastoid, f) Facial nerve canals, g) Jugular bulbs, h) Sigmoid sinus plate, i) Internal auditory meatus (IAM) and j) Cochlea.

MRI covering bilateral petrous temporal bones of all cases in axial and coronal planes was taken. 0.8mm thickness 3D-FIESTA sequences covering bilateral petrous temporal bones in axial and coronal planes. In addition, screening sequences of Brain was performed.

RESULTS AND STATISTICAL ANALYSIS

The study group consists of 13 male and 12 female patients. In our study of 25 patients, 8 cases had congenital CHL and 17 cases had congenital SNHL. Out of 8 patients with congenital CHL, 2 patients had anotia, 6 patients had microtia, 6 patients had atresia, 7 patients had narrow middle ear cleft, 3 patients had malformed ossicles and 5 patients had poor mastoid pneumatisation.

Out of 17 patients with congenital SNHL, 1 patient had Mondini's malformation, 2 patients had enlarged vestibular aqueduct and 1 patient had high riding jugular bulb, 1 patient had narrow IAM, 1 patient had facial nerve hypoplasia and 3 patients had brain abnormalities.

In all the 25 patients, the facial canal, cochlear nerves,

ISSN (Online): 2565-4810; (Print): 2565-4802 | ICV 2019: 98.48 |

sigmoid sinus plates, vestibule and semicircular canals are anatomically normal in radiological studies.
P=0.434, Not Significant, Fisher Exact Test
Sensitivity=44.44
Specificity=37.50
PPV=28.57
NPV=54.55
Accuracy=40.00

DISCUSSION

In our study, two imaging modalities, HRCT temporal bone and MRI brain with cochlea are performed.
A total of 25 patients were selected for our study. All patients with audiologically proven congenital hearing loss, both conductive and sensorineural types between the ages of one to fifteen years were included.
This study shows that CT better outlines the external ear, middle ear and bony abnormalities of inner ear. It is useful in pre-operative evaluation of abnormalities of external and middle ear and pre-operative assessment of malformation which helps the surgeon in deciding on the direction of electrode array to minimize the risk of misplacement and minimizing the trauma to vital structures.
MRI provides better delineation of cochlea, semicircular canal, and nerve abnormalities. It allows evaluation of the anatomy of the cochlear nerve within the IAM in cases of congenital SNHL.
Our study shows that imaging studies in congenital SNHL patients should not only focus just on the cochlear nerve or cochlea. One of our patient demonstrated hypoplastic facial nerve. In patients with cochleo-vestibular malformation should always anticipate aberrant course or shape of the facial nerve.
Preoperative radiological imaging of temporal bone and brain helps in planning preoperatively the type of surgery, determination of the side of cochlear implantation and surgical approaches for cochlear implantation.

CONCLUSION

Audiological evaluation is helpful in differentiating type of hearing loss. i.e.,Conductive, sensorineural and mixed type. However, the etiology cannot be established. Imaging plays a key role in evaluation of etiology and further line of management.
Our study demonstrated that preoperative HRCT and MRI together is more accurate in detecting cochlear malformations, ossicles, jugular bulb and cochlear nerve. Preoperative imaging before definitive surgery is important and should be done with ideal standards. This helps in selecting patients for ear reconstruction and cochlear implantation and also for preparing surgeons to anticipate complications and the best approach to avoid them.

Source of Support: Nil; **Conflict of Interest:** None

Submitted: 10-01-2023; **Accepted:** 14-02-2023; **Published online**: 30-03-2023