

ROLE OF HRCT AND 3D MRI IN CHILDREN WITH CONGENITAL SENSORINEURAL HEARING LOSS

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ABSTRACT

Background: Congenital hearing loss sensorineural variety is one of the most common birth defects. Imaging is very important to determine the underlying pathology of such cases. **Objective:** To assess the various cochlea vestibular bony and nerve anomalies by HRCT scan of temporal bone and MRI scan with 3D reconstruction images of inner ear. **Methodology:** A total of 140(280 ears) children with congenital deafness (88 males and 52 females), between 1st January 2012 to 30th September 2014 were included in this cross sectional study. All patients were assessed radiologically by HRCT scan of temporal bone and MRI with 3D scan of inner ear. This study was conducted at Al Razi Health Care Hospital, Lahore and Sheikh Zayed Medical College/Hospital, Rahim Yar Khan. The frequency of cochlear, vestibular aqueduct anomalies were noted. The data was entered and analyzed by using SPSS version 15. **Results:** We found various congenital anomalies of bony labyrinth and vestibulocochlear nerve in this study. Out of 280 inner ears we found 37 (13.2%) anomalous inner ears. Out of these 37 inner ears 25 (67%) has cochlear anomaly, 30 (83.1%) has anomalous vestibule, 19 (52.2%) has abnormal vestibular aqueduct and 9 (24.9%) has abnormal cochlea vestibular nerves. **Conclusion:** The study showed that most common ear anomaly in congenital sensorineural loss was cochlear vestibular, internal auditory canal anomalies. This study shows the outcome of preoperative inner ear assessment by radiological imaging mainly HRCT of temporal bone and MRI with 3D reconstruction

Keywords: MRI, HRCT, Hearing loss, Congenital

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INTRODUCTION

Sensorineural hearing loss is an important congenital anomaly, while imaging is one of the frequent modality applied in an attempt to determine an underlined pathology in such cases.¹ Incidence of sensorineural hearing loss is approximately 1: 1000 live births. Bony malformations of inner ear are usually best assessed by HRCT scan of temporal bone while on the other hand MRI scan provides better visualization of membranous labyrinth and status of vestibulocochlear nerves.²

In cases of congenital hearing loss the most common CT scan abnormality is unilateral or bilateral dilated vestibular aqueduct (LVA). Bony inner ear malformations are fairly uncommon as compared to congenital membranous anomalies. Vestibulocochlear congenital anomalies may be classified as follows;¹ Michel deformity,² cochlear aplasia,³ common cavity deformity,⁴ and cochlear hypoplasia.⁵ Further classification includes,⁶ Incomplete partition type I and incomplete partition type II (Mondini deformity).⁷ Vestibular malformations include: Michel deformity, common cavity, absent vestibule, and dilated vestibule.⁸ Semicircular canal malformations,

may be absent, hypoplastic, or enlarged.

Radiological investigations provide information on type of abnormality, of middle ear and vestibular system.⁷ These are best demonstrated by HRCT scan of the temporal bone with axial and coronal sections. MRI is another important tool to diagnose the presence or absence of nerves in the IAC and cochlear fluids.⁹ In patients with sensorineural hearing loss, cochlear implants are method of choice for auditory rehabilitation. Radiological imaging plays main role in cochlear implantation and with regard to preoperative evaluation and post-operative follow up.⁸

This study was conducted to assess the types of cochlear vestibular bony or nerve anomalies by HRCT and 3D MRI scan of inner ear.

METHODOLOGY

This cross sectional study was undertaken in the Department of Radiology at Alrazi Healthcare Hospital, Lahore and Sheikh Zayed Medical College Rahim Yar Khan, from 1st January 2012 to 30th September 2014. We evaluated a total of 140 children (males: 88, females: 52) of age range of 01–12 years, with bilateral congenital severe to profound sensorineural hearing loss. All patients has

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congenital deafness and sent to us for radiology imaging. All patients were candidates for possible cochlear implantation and all the patients underwent HRCT and MRI examination of the temporal bone and inner ear. The patients included in the study were selected on the basis of following inclusion and exclusion criteria. **Inclusion criteria:** Children who were congenitally deaf. **Exclusion criteria:** Children who were not congenitally deaf and developed hearing loss after some acquired cause.

Radiology Imaging Protocol: HRCT of temporal bone with special emphasis on inner ear structures were performed in the axial orientation using DSCT with a slice thickness of 0.3 mm. All MRI scans were performed on a 1.5T MRI scanner (Siemens) using an 8-channel head coil and the SPACE and CISS (heavily T2 weighted) sequence.

Image Analysis: CT and MRI images were evaluated for malformations. Different parts of inner ear were studied for Cochlea, vestibule, semicircular canals, and internal auditory canal along with vestibulocochlear nerve were focused upon. The malformations were classified using new classification of inner ear malformations based on CT and MRI given by Sennaroglu and Saatci.⁴ Data was entered and analyzed in SPSS version 15.

RESULTS

A total of 140 children (280 ears) within the age group of 01 to 12 years with bilateral congenital sensorineural hearing loss were radiologically evaluated with HRCT of temporal bone and MRI of inner ear. Out of 140 children, 121 children were normal and 19 children (37 inner ears) were found to be congenitally abnormal. All the 19 children have bilaterally abnormal inner ear except for 1 child who has unilateral abnormal ear.

Cochlear Anomalies

Out of 37 abnormal inner ears, in 25 (67%) cochlea was found to be abnormal. In 4 (16%) inner ears, cochlea has no turn or only a bony mass without any turn was visualized so it was classified as incomplete partition type-I (IP-I). In 15 (60%) inner ears, cochlea was of incomplete partition type-II (IP-II), means Mondini deformity, in this type 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 vestibular aqueduct. In 6 (24%) of cases cochlea was classified under the common cavity as there was cystic cavity representing the cochlea and vestibule, without showing any differentiation into cochlea and vestibule. (Figure I,II,III) (Table I)

Figure I: Single cochlear turn

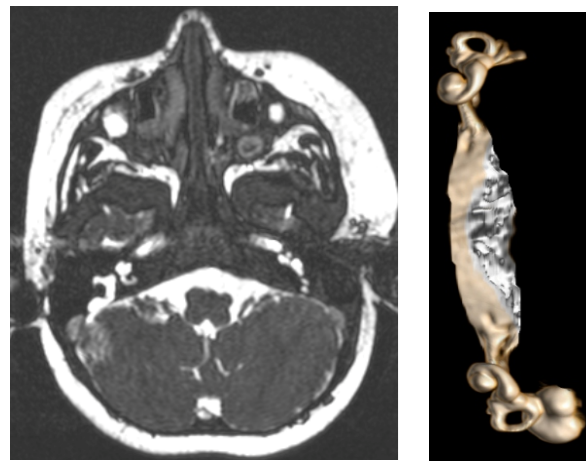
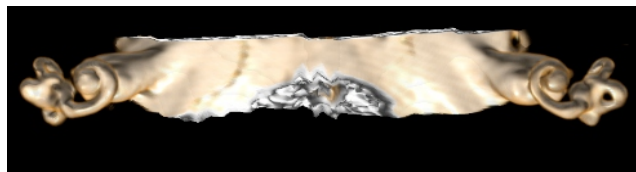


Figure II: Normal two and half turns of cochlea and semicircular canal



Vestibular Anomalies

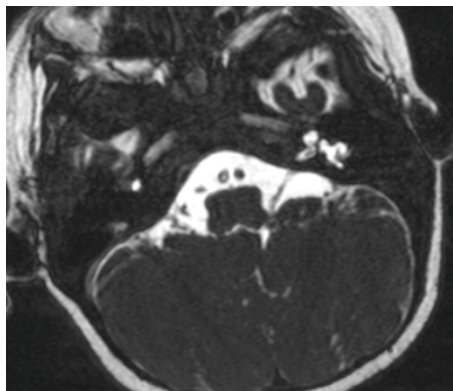
Vestibular anomalies were the second anomalies found. Out of 37 abnormal inner ears in 19 (51.1%) inner ears vestibule was found abnormal. In 15 (79%) inner ears vestibule was dilated and in the rest 4 (21%) it was aplastic or hypoplastic.

Semicircular Canal Anomalies: In 9 (42.9%) out of 21 malformed inner ears, lateral semicircular canals were found to be aplastic or hypoplastic and in 5 (24%) inner ears lateral semicircular canals were dilated. Superior semicircular canal was aplastic or hypoplastic in 6 (28.5%) cases and dilated in 1 (4.7%) cases.

Vestibular Aqueduct Anomalies: In 19 out of 37 (52.2%) of abnormal inner ears the vestibular aqueduct was found to be abnormal. (Table II)

Internal Auditory Canal (IAC) Anomalies: In 12 out of 37 (32.4%) of abnormal inner ears the internal auditory canal was found to be abnormal.

Figure I: Complete absence of all cochlea and vestibular structure on RT Side & Cochlea and hypoplasia on left side.



Status of Vestibulocochlear Nerves

In all cases where IAC was malformed, vestibulocochlear nerves were also malformed except for 1 case where IAC was dilated but nerves were visualized.

Table I: Frequency of ear Anomalies among examined ears

Type	Number	Percentage
Cochlea	25/280	9
Vestibule	19/280	6.7
Semicircular canals	09/280	3.2
Interneal auditory canal	12/280	4.2

Table II: Distribution of anomalies among abnormal ears (n=37)

Types	Number	Percentage
Cochlea	25	67.5
Vestibule	19	51.3
Semicircular canals	21	56.71
Interneal auditory canal	12	32.4

DISCUSSION

This study showed that HRCT and MRI scans were able to reveal similar morphological findings of malformed inner ears, except for vestibulocochlear nerves which were better appreciated on MRI scan. The knowledge of these malformations is important for cochlear implants.³ MRI scan delivers additional information that is needed in the preoperative work-up of patients. The fluid filled spaces of the normal cochlea are necessary for the insertion of the electrode array of the cochlear implantation. This can be clearly

visualized with MRI scan by using a 3D T2-weighted fast SE sequence.⁵

Our imaging studies not only demonstrated the causes of congenitally sensorineural hearing loss but also illustrated the other anomalies of hearing system. The majority of our patients demonstrated multiple anomalies of the inner ear. We have classified the anomalies according to the latest classification of congenital inner ear malformations given by Sennaroglu and Saatci in 2002.⁴

Incomplete partition type-I (cystic cochleovestibular malformations) is a malformation involving the cochlea and vestibule. In a case of IP-I, a cystic dilated vestibule accompanied the cystic, empty cochlea is usually seen.⁹ This pathology represents a form of common cavity that is one step more organized and differentiated than common cavity. Vestibule was grossly enlarged and the vestibular aqueduct was also dilated.¹⁰ Varsha M. Joshi et al discribed frequency of cochlear anomalies as, type -I in complete partition 6%, type-II incomplete partition 50% and common cavity deformity in 25% among abnormal ear. Result of our study are comparable with few variables which demonstrates is complete.

The malformation of incomplete partition type II (Mondini malformation) represents cochlea in which only the basal part of the modiolus is present. This is the type of cochlea originally described by Carlo Mondini and together with a minimally dilated vestibule and large vestibular aqueduct it constitutes the triad of the Mondini deformity.

CONCLUSION

The study showed that most common ear anomaly in congenital sensorineural loss was cochlear vestibular and internal auditory canal anomalies. This study shows the importance of preoperative inner ear assessment by radiological imaging mainly HRCT of temporal bone and MRI with 3D reconstruction.

Conflict of interest

The authors have declared no conflict of interest.

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