



Radiological Study of Congenital Anomalies in Profoundly Deaf Children

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Abstract

Introduction: Sensorineural hearing loss (SNHL) is a major cause of childhood disability worldwide, with an estimated prevalence of 1 in 2000 neonates and 6 in 1000 children by 18 years of age. Imaging plays an important role in the evaluation of congenital sensorineural hearing loss. It provides vital preoperative information about the Temporal bone and its anomalies, the vestibulocochlear nerve, and brain.

Aims & Objective: To study and evaluate the causes of congenital sensorineural hearing loss through radiological screening (CT/MRI) with following objectives (a) To find the incidence of cochlear malformations, vestibular semicircular canal, Vestibular and cochlear aqueduct malformations and (b) To find the incidence of Cochlear nerve deficiency, endolymphatic duct, Internal auditory canal abnormality.

Methods: All patients with congenital sensorineural hearing loss who visited study center were included who fulfilled the selection criteria. A total of 150 congenital SNHL patients were selected from the Department of Otorhinolaryngology and head and neck surgery at Sri Aurobindo Medical College and Post Graduate Institute, Indore. All patients were subjected to audiological assessment, ear examination, CT and MRI temporal bone.

Results: In 150 patients we found 25 (17%) patients had inner ear abnormality. Males were 72% and 28% were female, aged between 1 to 7 years.

Conclusion: Inner ear malformations were found in 17% of patients with congenital sensorineural hearing loss. High-resolution CT and MR imaging plays an important role in the evaluation of congenital hearing loss by providing crucial information about the inner ear, vestibulocochlear nerve, and brain.

Keywords: Congenital sensorineural hearing loss, inner ear abnormalities, computed tomography, magnetic resonance imaging.

Introduction

Sensorineural hearing loss (SNHL) is a multifaceted condition with profound medical and social effects. SNHL is a major cause of childhood disability worldwide, with an estimated prevalence of 1 in 2000 neonates and 6 in 1000 children by 18 years of age.^[1] Early diagnosis and

treatment of SNHL in children is critical because it is well recognized that a delay in identification of hearing impairment can adversely affect speech and language development, academic achievement, and social and emotional development.^[2] Imaging plays an important role in the evaluation of congenital sensorineural hearing loss.

It provides vital preoperative information about the Temporal bone and its anomalies, the vestibulocochlear nerve, and brain. In patients with congenital sensorineural hearing loss (SNHL) abnormalities of the inner ear that cannot be detected by conventional imaging techniques. High-resolution temporal bone computed tomography (CT) has been the first line imaging study of choice obtained by pediatric otolaryngologists in the workup of children with all types of SNHL, including symmetric, asymmetric, and unilateral SNHL.^[3,4,5,6]

Adults with asymmetric or unilateral SNHL often undergo gadolinium-enhanced magnetic resonance (MR) imaging of the brain with attention to the internal auditory canals to evaluate for retrocochlear abnormalities.^[7,8] In contrast to adults, children with unilateral or asymmetric SNHL rarely have acoustic neuromas, and therefore the indications for MR imaging are less clear.^[8]

Congenital malformations of the inner ear are rare anomalies; they can be identified on imaging with HRCT and/or MRI in about 20% of patients with congenital sensorineural hearing loss.^[9] Bony abnormality of the inner ear can be detected in 10%–25% of such patients by conventional computed tomography (CT).

High-resolution computed tomography (CT) and magnetic resonance (MR) imaging provide excellent delineation of the intricate anatomy of the inner ear. Narrow internal auditory canal (IAC) with duplication is a rare congenital disorder that is usually associated with other inner ear, middle or external ear abnormalities.^[10] A narrow IAC can be diagnosed when the diameter of IAC is less than 2 mm as seen on high-resolution CT (HRCT) of the temporal bone.^[11] This malformation comprises 12% of congenital temporal bone anomalies.^[9,12] It has been thought that this anomaly is caused by aplasia or hypoplasia of the vestibulocochlear nerve that results in ipsilateral congenital sensorineural hearing loss.^[10,11,13] CT depicts the minute details of osseous structures, and MR imaging allows

visualization of the fluid-filled spaces and the vestibulocochlear nerve. Together, these complementary modalities can aid in decision making about the best management strategy by facilitating the identification and characterization of inner ear malformations and any associated neurologic abnormalities.

Aim

To study and evaluate the causes of congenital sensorineural hearing loss through radiological screening (CT/MRI).

Objectives

- To find the incidence of cochlear malformations.
- To find the incidence of vestibular malformations.
- To find the incidence of Malformations of semicircular canals.
- To find the incidence of Vestibular and cochlear aqueduct malformations.
- To find the incidence of Cochlear nerve deficiency.
- To find the incidence of endolymphatic duct abnormality.
- To find the incidence of Internal auditory canal abnormality.
- To find the incidence of Facial nerve abnormality.

Material and Methods

Research Design: Retrospective and prospective (observational study).

Place of Study: The study was conducted in the Department of Otorhinolaryngology and Head and Neck Surgery at Sri Aurobindo Medical College and Post Graduate Institute, Indore (M. P.).

Study Duration: The duration of study was two year i.e. from January 2014 to December 2015.

Sampling: All patients with congenital sensory neural hearing loss according to inclusion-exclusion criterion were selected.

Study Tools

1. History

2. Examination
3. Routine investigations
4. Audiometry
5. Computed tomography temporal bone
6. MRI temporal bone

Inclusion Criteria

1. Patient attending the ENT OPD at Sri Aurobindo Institute Of Medical Sciences, Indore with complaint of unable to hear and speak since childhood.
2. Patients of age group 1-7 years
3. Patients with severe to profound and profound sensorineural hearing loss [>90 db]
4. Patient whose parents were willing to provided their voluntary written informed consent for letting their child participate in the study

Exclusion Criteria

1. Patient with middle ear pathology
2. Patient with tympanic membrane perforation
3. Uncooperative patient
4. Patient with age more than 7 years
5. Patient whose parents were not willing to provided their voluntary written informed consent for letting their child participate in the study

Methodology

The patients who presented with sensorineural hearing loss, their parents were explained clearly about the study and their willingness to allow their child participate in the study was recorded in a consent form, voluntarily signed by them. All the study related procedures were conducted after obtaining the written voluntary informed consent.

A total of 150 congenital SNHL patients were selected from the Department of Otorhinolaryngology and Head and Neck surgery at Sri Aurobindo Institute of Medical Sciences and Post Graduate Institute, Indore as patients for the study.

History recording and data collected when the analysis was performed. History in terms of

antenatal, postnatal and birth history for infection, injury, immunization, consanguinity were collected. All the patients were subjected to audiological assessment, ear examination, CT and MRI of temporal bone. In all subjects complete general, systemic and ENT examination was done. Images were evaluated for presence of normal structure as well as for any anomaly. Present or absent of the nerves (facial, cochlear), relative internal auditory canal (IAC), and any associated inner ear abnormalities.

Statistical Analysis

Data were entered in Microsoft Excel and analysed using SPSS software. The data was calculated in the form of numbers and percentages. The final results were presented in the form of tables and graphs.

Results

In our study we had included 150 patients with congenital sensorineural hearing loss, of which were found abnormal findings in 25 (17%) of the patients. There was a male preponderance in comparison to the females with a Male:Female ratio of 2.57 : 1.

Of the 25 abnormal patients, in 60% cochlear abnormalities were present, in 44% vestibular abnormality, semicircular canal and cochlear nerve abnormalities was present. In 48% vestibular aqueduct abnormalities [Fig. 1], in 32% IAC abnormality, in 24% endolymphatic duct and in 8% facial nerve anomalies were present. [Table 1]

Of the 15 (60%) cases of cochlear abnormalities, most of the patients 33% had cochlear hypoplasia, 13% had cochlear aplasia, 27% had cochlear dysplasia, 20% had cystic cochlea and only 7% had cochlear dysplasia. [Fig. 2]

Of the 11 (44%) patients with vestibular abnormalities, most of the patients had dilated vestibule i.e. 55%, 18% patients had common cavity, 9% patients had vestibule hypoplasia, cystic and absent vestibule. [Fig. 3]

12 (75%) patients had dilated vestibular aqueduct and 25% have absent vestibular aqueduct.

Of 10 (40%) patients with semicircular canal abnormalities, 30% patients had PSCC abnormalities and only 10% cases had SCCC abnormalities.

Table 1: Distribution of patients according to abnormality

Abnormality	No of patients	Percentage
Cochlear abnormality	15	60%
Vestibular abnormality	11	44%
Vestibular aqueduct	12	48%
Semicircular canal	10	44%
Cochlear nerve	11	44%
Facial nerve	2	8%
Endolymphatic duct	6	24%
Internal auditory canal	8	32%

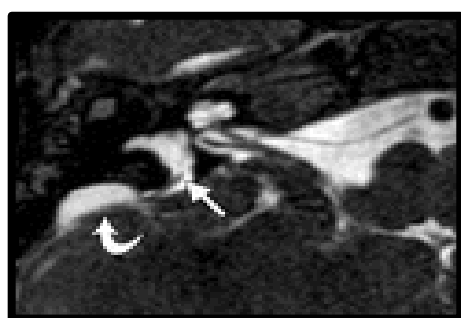


Fig. 1: Axial MR images show marked dilatation of the endolymphatic duct (straight arrow) and sac (curved arrow) in comparison with the normal posterior semicircular canal

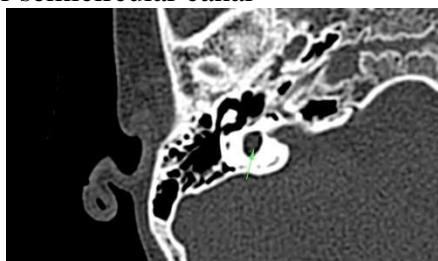


Fig. 2 : Common cavity of cochlea

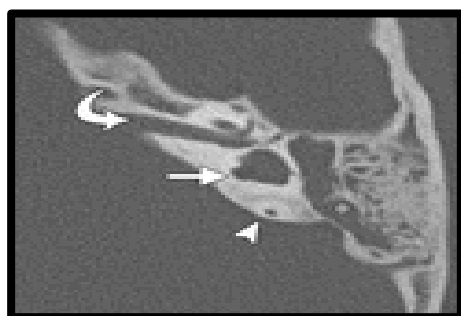


Fig. 3: Axial CT image shows a stenotic IAC (curved arrow), mildly dilated vestibule with a stunted lateral semicircular canal (straight arrow), and middle ear and mastoid air cell effusion. The posterior semicircular canal (arrowhead) appears normal.

Discussion

In our study, we found 25 (17%) patients with inner ear abnormalities. While study done by Mafong et al^[14] found a higher percentage (39%) and Huang et al^[15] found a very low percentage of inner ear abnormalities (9%). While Dagkiran et al^[16] found an incidence similar to our study (20%).

Male preponderance was seen in our study in comparison to females with a male:female ratio of 2.57 : 1. Similarly Halpin et al^[17] also found male preponderance.

We found 60% of the patients with cochlear abnormality on CT findings. Jackler et al found 76% cochlear abnormalities, while Reilly et al^[18] found only 4% abnormality in their study. Dagkiran et al^[16] found cochlear abnormalities in 13% of their series.

With regard to cochlear malformations, we found cochlear hypoplasia in 33% patients, cochlear aplasia in 13%, cochlear dysplasia in 7%, common cavity in 27% patients and cystic cavity in 20% patients. Jackler et al found common cavity in 26%, cochlear aplasia in 1%, cochlear hypoplasia in 15% and cystic cavity in 55% cases. Casselman et al^[19] found cochlear aplasia in 3% of their patients, Joshi et al^[20] found anomalies in 18% with most common as cochlear dysplasia in 10.2% patients.

Vestibular abnormality was seen in 44% patients, of which, dilated vestibule was the most common (55%), followed by common cavity (18%) and vestibule hypoplasia, cystic and absent vestibule in 9% patients. Joshi et al^[20] found vestibular dysplasia in 10.2% patients and Wu et al^[15] found 3% vestibular abnormalities.

48% patients had vestibular aqueduct abnormality, of which, 75% patients had dilated vestibular aqueduct and 25% had absent vestibular aqueduct. Sennaroglu et al^[21] found VA in 22.2% in one study and 34% in another study.

Of the 25 cases of anomalies, 40% patients had semicircular canal abnormalities, of which 50% of patients had hypoplastic lateral semicircular canal,

33% patients had LSCC absent and 17% patients had dilated LSCC.

In the present study, 30% patients had PSCC abnormalities and 10% patients had SCCC abnormalities. Joshi et al^[19] found semicircular canal dysplasia in 10.2% patients. Dagkiran et al^[16] found posterior semicircular canal malformation in 7.3% patients, lateral semicircular canal in 7.4% and superior semicircular canal aplasia / hypoplasia in 5.9% patients.

Facial nerve abnormalities were seen in 8% of the patients and cochlear nerve abnormalities absence in 80% patients. Dagkiran et al^[16] found cochlear nerve absence in 15.4% patients.

32% patients had internal auditory canal abnormality, of which 50% had narrow internal auditory canal and 37% patients had dilated internal auditory canal. Dagkiran et al^[16] found dilatation of internal acoustic canal in 30.9% patients.

Endolymphatic duct abnormalities were seen in 24% of the patients of the present study. Koesling et al^[22] found 10% patients with enlarged endolymphatic duct.

Conclusion

Inner ear malformations were found in 17% of patients with congenital sensorineural hearing loss. High-resolution CT and MR imaging play an important role in the evaluation of congenital hearing loss by providing crucial information about the inner ear, vestibulocochlear nerve, and brain. Both modalities precisely and accurately delineate the inner ear anatomy and malformations. They are often complementary and are used together in the preoperative evaluation of pediatric candidates for cochlear implantation. Most of the patients of congenital SNHL are found with cochlear malformations. Preoperative high-resolution CT offers the advantage of visualizing any coexistent middle or external ear anomalies and important anatomic variants, and MR imaging provides definitive information about the integrity of the cochlear

nerve and the fluid-filled spaces of the inner ear. These radiological instruments aid in determining contraindications and predicting intraoperative difficulties. Identification of the inner ear abnormality has a substantial effect on clinical decision making, prognosis, and management strategy.

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