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Original Research Article

High resolution temporal bone computerized tomography in paediatric sensorineural hearing loss prior to cochlear implantation

Suhail Rafiq[1]*, Fahad Shafi[2], Ajaz Mohiuddin[2], Sajjad A. Dar[1]

[1]Department of Radio diagnosis and Imaging, Government Medical College, Srinagar, Jammu and Kashmir, India
[2]Department of Radio diagnosis and Imaging, Sher-i-Kashmir Institute of Medical Sciences, Soura, Srinagar, Jammu and Kashmir, India

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***Correspondence:**
Dr. Suhail Rafiq,
E-mail: suhailrafiq777@gmail.com

ABSTRACT

Background: Congenital sensorineural hearing loss (SNHL) is one of the most common birth defects with incidence of approximately 1:1000 live births. Imaging plays an important role in the work up of cochlear implant candidates not only to identify inner ear congenital and acquired abnormalities or cochlear nerve anomalies but also to detect temporal bone abnormalities or variations that may alter surgical approach. Preoperative evaluation of cochleovestibular anatomy is an important component of the cochlear implant evaluation. The objective of the study was high resolution computerized tomography (HRCT) assessment of congenital ear anomalies before cochlear implantation.
Methods: This prospective study was conducted in the Department of Radio diagnosis and Imaging, GMC, Srinagar. 24 children in the age group of 1 to 12 years with unidentified causes of bilateral SNHL were subjected to HRCT over a period of 7 months from January to July 2019.
Results: Eighteen patients had normal radiological scans and 6 had congenital anomalies. We had one each case of common cavity, bilateral labrynthine aplasia, incomplete partition type 1, Mondini's deformity with dilated vestibular duct, Internal auditory canal stenosis and bilateral large vestibular aqueduct. Out of 18 patients without congenital anomaly, two had Korner's septum and giant jugular bulb which were important for operating surgeon.
Conclusions: HRCT temporal bone is superior at identifying the bony labyrinth, including enlarged vestibular aqueduct and caliber of internal auditory canal. HRCT temporal bone should be supplemented by magnetic resonance imaging especially for cochlear nerve assessment. It is the initial imaging modality of choice for assessment of congenital SNHL.

Keywords: High resolution computerized tomography, SNHL, Vestibular aqueduct

INTRODUCTION

Congenital sensorineural hearing loss is one of the most common birth defects with incidence of approximately 1:1000 live births.[1] Cochlear implants (CIs) are a well-accepted treatment for severe-to-profound sensorineural hearing loss patients who are refractory to conventional hearing augmentation.[2,3] Imaging plays an important role in the work up of CI candidates not only to identify inner ear congenital and acquired abnormalities or cochlear nerve anomalies but also to detect temporal bone abnormalities that may be encountered during surgery that may alter surgical approach.[3,4] Some

variations are potential surgical hazards that may lead to problems during the surgery and may alert the surgeon regarding potential surgical dangers and complications.[5] Both computed tomography (CT) and magnetic resonance imaging (MRI) should be used as they delineate, in different manners, cochlear and middle ear anatomy as well as other anatomical variants.[5,6]

Sensorineural hearing loss has a variety of causes and can be generally classified according to genetic and nongenetic etiologies. Cochleovestibular anomalies are common amongst pediatric cochlear implant candidates because they often correlate with sensorineural hearing loss. Approximately 20% of patients with congenital hearing loss have a radiographically identifiable morphological abnormalities of inner ear.[7] The remaining 80% of the cases of congenital malformations are membranous malformations in which bony architecture of the inner ear is normal and the pathology is at the cellular level. In the latter patient group, the result of radiological investigations of the inner ear falls within the normal limits.[8] In general, inner ear malformations can be associated with a wide range of hearing sensitivity.[8] As a general rule, however, the more severe is the deformity, the worse is the hearing.[8] It is generally accepted that first trimester inner ear malformations may be diagnosed by high resolution computerized tomography (HRCT). Inner ear malformations occurring beyond this time are thought to involve the membranous labyrinth only, thus not identifiable by HRCT.[8] Therefore, preoperative evaluation of cochleovestibular anatomy is an important component of the cochlear implant evaluation.

Aim and objective

The aim and objective of the study was radiological assessment of congenital ear anomalies before CI.

METHODS

This prospective study was conducted in the Department of Radiodiagnosis and Imaging, Government Medical College, Srinagar. Total number of 24 children in the age group of 1 to 12 years with unidentified causes of bilateral sensorineural hearing loss, after undergoing historical and audiological evaluation were analysed radiologically over a period of 7 months from January to July 2019. Each patient underwent high resolution CT scanning of temporal bone. HRCT of the temporal bone was performed on a 256-slice volume in axial plane. The original isometric volume data was used to obtain coronal images.

Inclusion criteria

Age group 1-12 years, children with pure sensorineural hearing loss and no history of any otolaryngological surgery were included in this study.

Exclusion criteria

Refusal to participate in the study and children with mixed or conductive hearing loss were excluded.

The collected data was analyzed using Microsoft excel 2010 and presented in number and percentages.

RESULTS

Out of the 24 patients of which 14 were males (58.3%) and 10 were females (41.6%) (Figure 1). Mean age of patients in our study was 2.5 years with maximum number of patients in age group of 1-5 years.

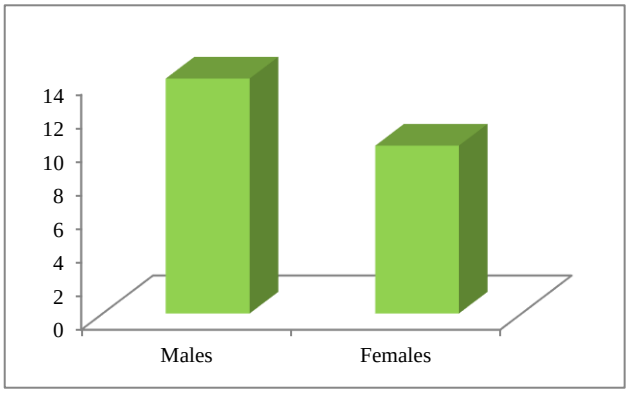

Figure 1: Distribution of patients according to gender basis.

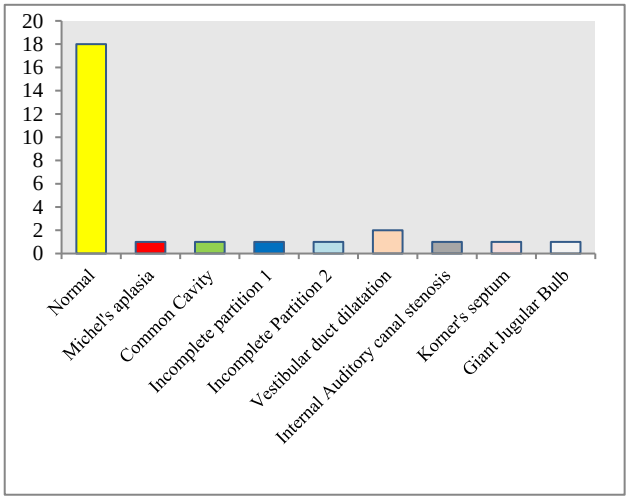

Figure 2: Incidence of different pathologies/ variations in HRCT temporal bone.

Number of 18 patients (75%) in our study had normal radiological scans and 6 had congenital anomalies. Incidence of different pathologies/ variations in HRCT Temporal bone is shown by Figure 2.

Common cavity was present in one patient (4.1%) (Figure 3).

Bilateral labrynthine aplasia (Michel's anomaly) was present in one patient (4.1%). Incomplete partition type 1 was present in one patient (4.1%).

One patient had Mondini's deformity (incomplete partition type 2) with triad of abnormal cochlea with only 1.5 turns, enlarged vestibule and enlarged vestibular duct (4.1%) (Figure 4).

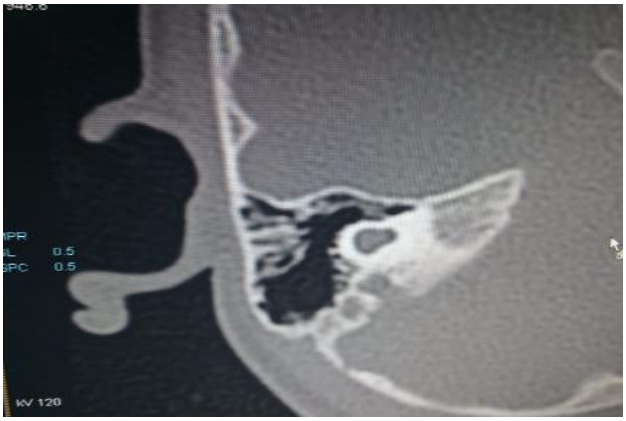

Figure 3: Axial HRCT image shows evidence of common cavity replacing cochlea, vestibule and semicircular ducts.

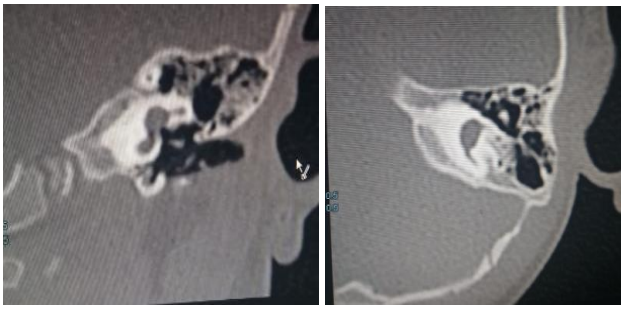

Figure 4: Coronal and axial HRCT image revealing evidence of only one and half turns of cochlea along with dilated vestibule suggestive of Mondini's deformity.

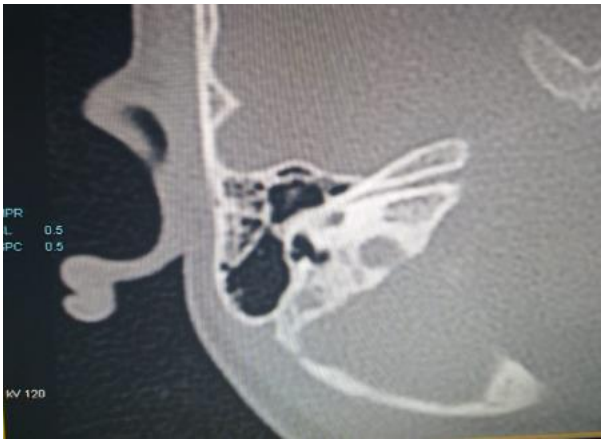

Figure 5: Axial HRCT image reveals evidence of hypoplastic/atretic internal auditory canal.

One patient had internal auditory canal stenosis (4.1%) (Figure 5).

Bilateral large vestibular aqueduct (VA) (Figure 4) was present in 1 patient (4.1%) and was also associated with one case of unilateral Mondini's anomaly. So, overall vestibular aqueduct was most common finding in our study.

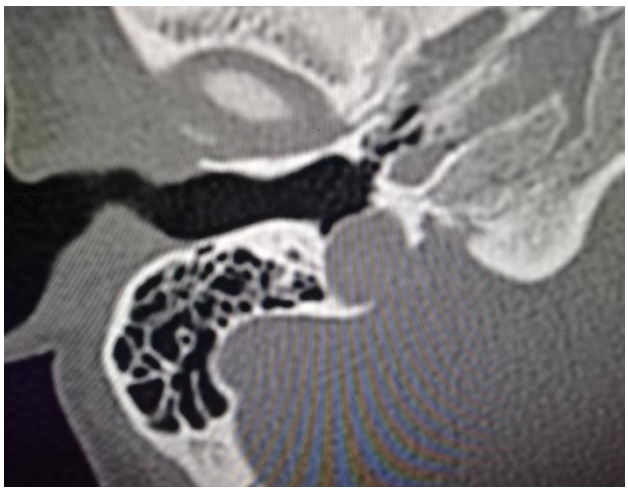

Figure 6: Axial HRCT image revealing evidence of giant high riding jugular bulb.

Out of 18 patients without congenital anomaly, two had anomalies which were important for operating surgeon. These included Korner's septum and giant jugular bulb.

DISCUSSION

Radiographic imaging of the temporal bone is invaluable to the otological surgeon contemplating cochlear implantation.[9] Initial radiological evaluation of the cochlear implant candidate is typically performed with high resolution CT scanning. HRCT scanning technology has increased the ability to assess the structures within the petrous pyramid.[10] An HRCT scan of the temporal bone reveals four major features: inner ear malformations, the patency of cochlear coils, the position of the jugular bulb (which if high enough may reach up to the level of the round window), and the presence of the retrocochlear and infracochlear air cells which may be mistaken for round window niche.[11] HRCT can analyze the direction of insertion of the electrode array to minimize the risk of misplacement and by assessing the malformation preoperatively we can minimize the trauma to the vital structure.

Arrest of development during the 3rd week results in aplasia of the labyrinth, known as Michel's anomaly. Incomplete partition type-I (cystic cochleovestibular malformations) is a malformation involving the cochlea and vestibule. In a case of IPI, a cystic dilated vestibule accompanied the cystic, empty cochlea. This pathology represents a form of common cavity that is one step more

organized and differentiated than common cavity.[12] The cochlear duct has 1 and 1/2 turns by seventh week of gestation. Arrest in this development results in the classical Mondini dysplasia. The VA narrows from the fifth to the eighth week and it will be wide in ears with arrested development.[9,13] Several reports have speculated on the cause of a sudden drop in hearing in patients with large VA; two of these possible causes are reflux of hyperosmolar fluid from the endolymphatic sac to the inner ear and rupture of membranous labyrinth or perilymph fistula due to direct transmission of intracranial pressure to the inner ear through the enlarged VA.[14,15]

Congenital internal auditory canal stenosis which implies absence or hypoplasia of the cochlear nerve is a relatively rare disorder.[16] This malformation comprises 12% of congenital temporal bone anomalies.[9,17] A narrow internal auditory canal or internal auditory canal stenosis is defined as a canal of only 2 mm or less in diameter on HRCT.[18-20] The aetiology of congenital internal auditory canal stenosis is believed to be secondary to aplasia or hypoplasia of the vestibulocochlear nerve.[19,21]

Korner's septum divides the mastoid process into a superficial squamous portion and a deep petrous portion. It may mislead the surgeon to the mastoid antrum during surgery.[4]

Jugular bulb variations are common, the roof of a normal jugular bulb lies either at or slightly below the level of the external auditory canal floor and is separated from the middle ear cavity by the thin bony sigmoid plate.[22,23] The average width of jugular bulb is 1 cm.[23] A jugular bulb larger than 1 cm is called a giant or mega jugular bulb.[23]

75% of patients had normal HRCT scans and 25% had congenital anomaly. Jallu et al in their study had normal radiological scans in their study in 72.5% patients consistent with our figures.[24] Bilateral large VA was the most common finding in their study. In our study also vestibular duct dilatation was the most common anomaly accounting for 8.3% cases. High percentage of normal HRCT scans could be attributed to the fact that membranous labyrinth anomalies are not assessed by HRCT.

As compared to HRCT, MRI in measuring the transverse diameter of the cochlear nerve and in identifying early ossification of labyrinth and soft tissue anomalies in the inner ear the most important being presence or absence of cochlear nerve.[25]

CONCLUSION

Radiological imaging as an integral component of evaluation of Sensorineural hearing loss. HRCT temporal bone is superior at identifying the bony labyrinth, including enlarged VA and caliber of internal auditory canal. HRCT temporal bone should be supplemented by MRI especially for cochlear nerve assessment.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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