Original article:

Internal auditory canal diameter in children with congenital sensorineural hearing loss

Dr. Ayshath Shamseena[1], Dr. Devdas Acharya[2], Dr. Arafat M. Haris[3], Dr. Ravichandra G.[4]

[1] Dept. Of Radiology, Yenepoya Medical College, Mangalore

[2] H.O.D., Dept. Of Radiology, Yenepoya Medical College, Mangalore

[3] Assistant Professor, Dept. Of Radiology, Yenepoya Medical College, Mangalore

[4] Associate Professor, Dept. Of Radiology, Yenepoya Medical College, Mangalore

Corresponding author : Dr. Ayshath Shamseena

Abstract

Aim – The aim of this study was to evaluate the predictive value of the diameter of the internal auditory canal (IAC) on computed tomogram for the aplasia or hypoplasia of the cochlear nerve in children with congenital sensorineural hearing loss (SNHL).

Materials and Methods – A retrospective study of 49 patients diagnosed with congenital sensorineural hearing loss was done using computed tomography (CT) and magnetic resonance images (MRI). All patients were candidates for possible cochlear implantation. To reduce motion artefacts, some of the children were studied in sedation. The children were between the age group of 2 to 12 years.

Results – Cochlear nerve was not visualised in 6 out of 8 patients with narrow IAC. However, the cochlear nerve was visualised in 2 patients with narrow IAC. P value was found to be < 0.001 and is statistically significant.

Conclusion – A hypoplastic IAC is an indicator of a hypoplastic/aplastic cochlear nerve and a contraindication for cochlear implantation. Temporal bone CT can be used as the modality of choice in initial investigation of children with sensorineural hearing loss and the IAC canal diameter on CT can be used to select patients with SNHL who should undergo further evaluation with MR imaging.

Keywords – Internal auditory canal diameter, sensorineural hearing loss, cochlear nerve

Introduction

Children suffering from hearing loss often present to otolaryngologists and paediatrician at an early age. Early diagnosis of the cause of sensorineural hearing loss (SNHL) and its correction are important in speech and language development for young children [1]. Cochlear implantation is an effective treatment for selected patients with SNHL [2]. Preoperative evaluation of paediatric cochlear implant candidates consist of assessment of a functioning cochlear nerve.

The present study was done to assess whether the internal auditory canal (IAC) diameter on computed tomography (CT) can predict the presence of the cochlear nerve.

Methods and Materials

Subjects

CT and MRI images of 49 patients with SNHL were retrospectively evaluated which were obtained during the period December 2014 –May 2015 at our centre. The patients were between the ages of 2 to 12 years.

Statistical Method

Descriptive statistical analysis of the IAC diameter and the visualisation of cochlear nerve were done. A diameter of less than 3mm was considered stenotic. The mean IAC diameter, the frequency of

IAC stenosis and absence of cochlear nerve were calculated for categorical data and P value was determined for statistical significance.

Imaging

Noncontrast temporal bone CT and MRI were performed as part of preoperative workup of all the patients in our study population. CT scanning was performed on a 16-channel multidetector CT scanner (BrightSpeed; GE Healthcare). The images were acquired and reconstructed in an axial plane at 0.625 mm thickness using a bone algorithm. The diameter of the internal auditory canal was measured on the coronal images in each ear along the inner margin of its bony wall at the mid section. High resolution MRI with CISS T2-weighted volumetric pulse sequences (3DT2) was also done in all the patients to visualise the cochlear nerve. Some of the children were sedated to reduce motion artefacts during the imaging process.

Observation and Results

A total of 49 patients were included in the study. 55.1% patients were females whereas 44.9% were males. Patients between ages 2 to 12 years were included. The mean age was found to be 6.67 years. The mean IAC diameter was found to be 4mm. Out of the 49 cases included in the study, 8 were found to have internal auditory canal diameter equal to or below 3mm, that is, 16.3% subjects had narrow internal auditory canal. Cochlear nerve was not visualised in 6 out of 8 narrow IACs, however in 2 patients who had narrow IAC the cochlear nerve was visualised. P value (<0.001) was found to be significant.

	Frequency	Percent
Females	27	55.1
Males	22	44.9
Total	49	100.0

Table.1. Number of males and females in the study.

	Frequency	Percent
yes	43	87.8
no	6	12.2
Total	49	100.0

Table.2. Frequency of presence and absence of cochlear nerve.

		Cochlear nerve		
		No	Yes	Total
IAC	<=3	6 75.0%%	2 25.0%	8 100.0%
	>3	0 .0%	41 100.0%	41 100.0%
Total		6 12.2%	43 87.8%	49 100.0%

Table.3. Frequency of presence of cochlear nerve and its correlation with the IAC diameter.

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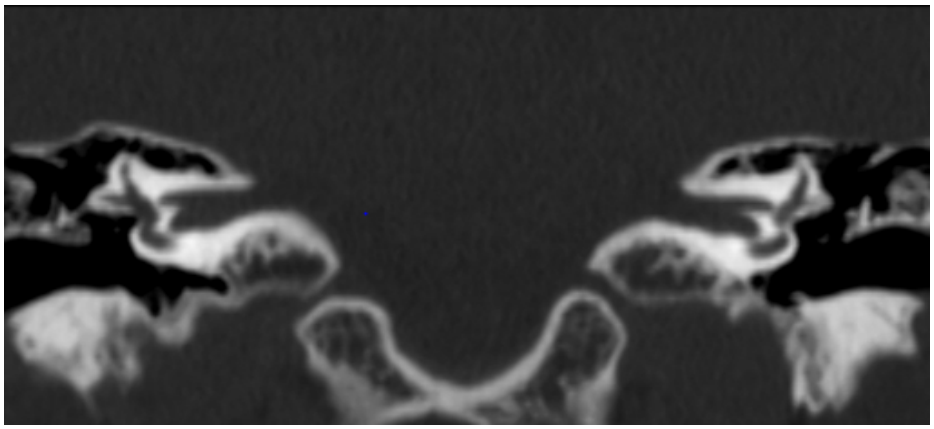

Fig. 1a. Coronal thin section temporal bone CT showing normal IAC in a 5 year old female patient with bilateral SNHL. The diameter of the IAC was found to be 5.1mm on the right side and 5mm on the left side.

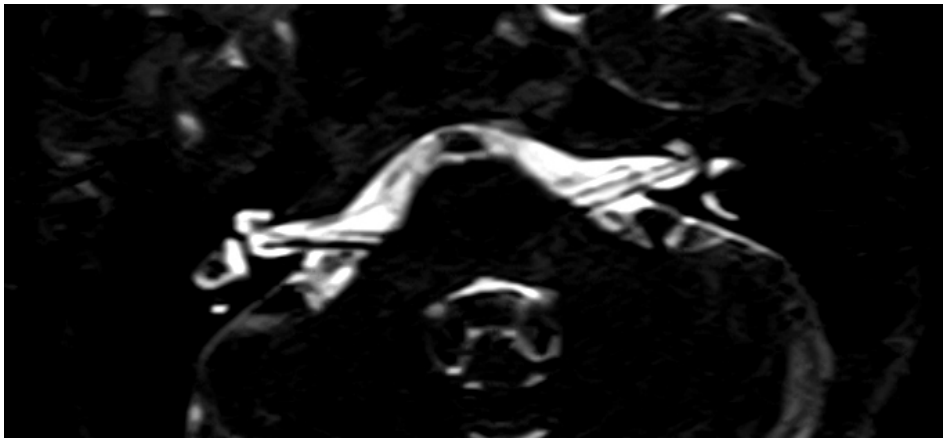

Fig. 1b. Axial 3DT2 thin section MR images in the same patient showing normal cochlear nerve bilaterally.

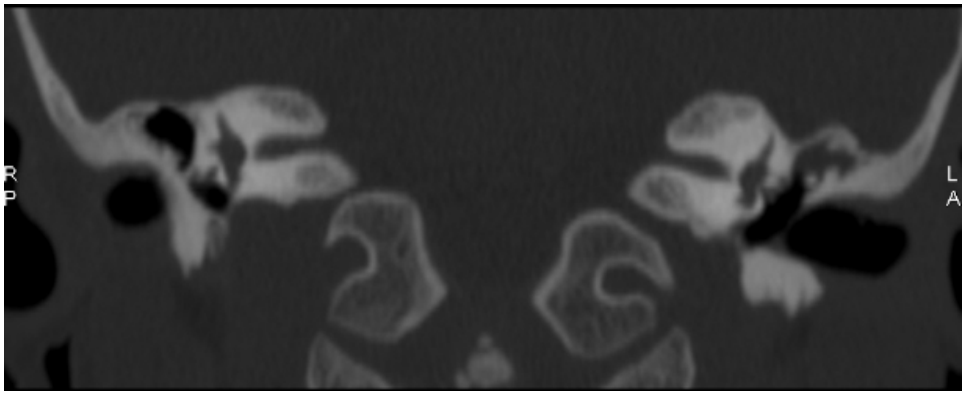

Fig. 2a. Bilateral hypoplastic IAC canal in a 7-year-old boy with bilateral SNHL on coronal, thin-section CT scan. The diameter was 2.4mm bilaterally.

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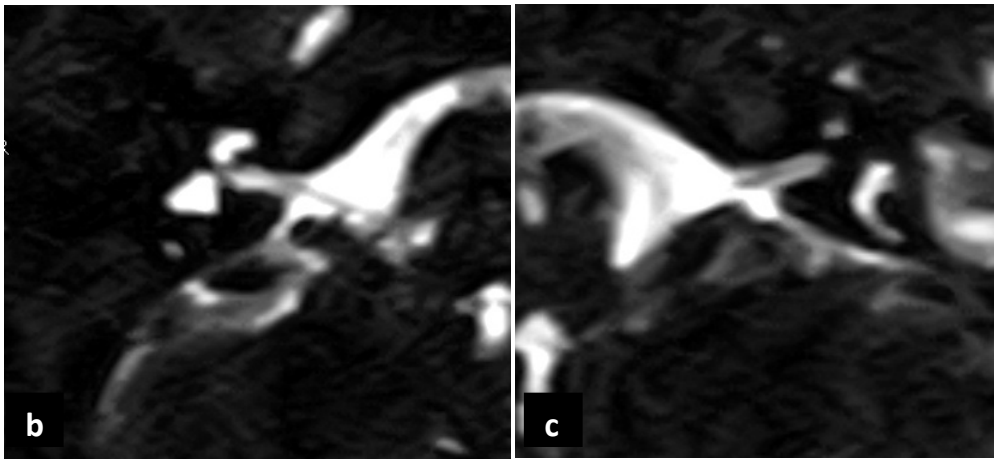

Fig. 2b,c. Absence of the cochlear nerve noted on 3DT2 thin MRI sections bilaterally. b. Right. c. Left

Discussion

Cochlear nerve deficiency can be associated with absence or hypoplasia of the cochlear nerve. A hypoplastic cochlear nerve is considered a relative contraindication for cochlear implantation. Some studies have shown that a reduction in the diameter of the cochlear nerve may be due to reduced number of spiral ganglion cells [3]; although the residual cochlear nerve fibers may still be effective, the percentage of functioning nerve fiber is uncertain and needs to be evaluated [4]. Absolute contraindication for cochlear implantation is an absent cochlear nerve. Lack of a visible cochlear nerve on MRI has been shown to be related to poor outcome of cochlear implantation [5].

CT and MRI are the imaging modalities considered for preoperative evaluation of cochlear implant. High-resolution CT has been recommended by many authors as the initial imaging modality for the work -up of children with SNHL [6, 7]. CT may provide good resolution for abnormalities of the bony labyrinth, IAC, ossicular chain, facial nerve canal, and jugular bulb, and can assist in the planning of the operative route [8]. Detection of and direct visualization of the cochlear nerve is better done with MRI [9, 10], but has a higher cost and a longer examination time.

Some authors have suggested that there may be a correlation between cochlear nerve deficiency and size of the IAC [11-13], while others have suggested that the correlation is unreliable [14, 15].

Embryonic development of the inner ear begins with the formation of the otic vesicle into a vestibular pouch and a cochlear pouch (the future utricle and saccule), which occurs between the fourth and fifth gestational week in humans [6, 16, 17]. The cochlear duct begins to extend from the cochlear pouch late in the fifth gestational week, further developing into cochlear turns in the seventh gestational week. The semicircular canals develop from the vestibular pouch in the seventh gestational week. The developing inner ear elicits a trophic effect on cochlear neurons [9]. Disturbance of this neurotrophic effect may lead to cochlear nerve hypoplasia

The development of IAC occurs by inhibition of cartilage formation at the medial side of the otic vesicle during fetal life. This inhibition requires the presence of the vestibulocochlear nerve. A canal will not be formed in the absence of a cochlear nerve [18]. The presence of the vestibulocochlear nerve is essential for the formation of the IAC, but survival and promotion of the nerve seems to require the presence of a growth factor from the

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otic vesicle [19]. Previous studies have suggested that an inhibitory substance may be induced by the presence of the vestibulocochlear nerve [18, 20]. Glastonbury et al. further suggested that the degree of vestibulocochlear nerve hypoplasia may be related to the severity of IAC stenosis [11]. The results of the present study indicate that IAC stenosis is a strong statistically significant positive predictor of cochlear nerve deficiency, which supports the developmental theory of the cause-effect relationship between the cochlear nerve and IAC.

Shelton et al [12] proposed that the aplastic IAC revealed by CT did not contain a cochlear nerve and was a contraindication to cochlear implantation. With high resolution MR imaging, it is possible to show that these patients indeed lack the cochlear nerve.

Congenital sensorineural hearing loss caused by IAC stenosis cannot be rehabilitated by hearing aids. Indication for cochlear implantation needs to be determined in bilateral IAC stenosis. Hearing rehabilitation and supportive treatment using a vibrotactile device can be applied and an auditory brainstem implant may be considered in such patients.

Conclusion

Temporal bone CT can be used as the modality of choice in initial investigation of children with sensorineural hearing loss as a hypoplastic IAC is an indicator of a hypoplastic/aplastic cochlear nerve which is a contraindication for cochlear implantation. Hence, IAC canal diameter on CT can be used to select patients with SNHL who should undergo further evaluation with MR imaging.

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