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Imaging In Congenital Sensorineural Hearing Loss – Incidence Of Cochlear Nerve Abnormalities Associated With Narrow/Stenotic Cochlear Fossette

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I. Introduction

One of the most common birth defects is Sensorineural hearing loss with incidence of approximately 1:1000 live births (1). Cochlear implantation is the only U.S. Food and Drug Administration–approved treatment for children with marked sensorineural hearing loss. The auditory benefits of this procedure that range from simple sound detection to substantial word understanding.(2) The presence of a functioning cochlear nerve fiber is a crucial issue in the cochlear implant candidates. The purpose of this study was to determine whether a narrow /stenotic cochlear fossette/aperture/nerve canal could serve as a useful and dependable predictive factor for the presence of cochlear nerve deficiency.

II. Materials and Methods

Subjects - A prospective study based on imaging of pediatric patients diagnosed with congenital sensorineural hearing loss was done over a period of 3 months from December 2014 to February 2015. Imaging modalities consisted of both CT and MRI. The children included in the study were between 2 to 12 years of age. A total of 55 children underwent CT and MRI after complete audiological examination and investigations including pure tone audiometry and brainstem evoked response audiometry. Out of this, 6 children who had other structural abnormalities of the ear like Mondini's dysplasia were excluded and only those children with no other bony abnormalities of the ear were included in the study.

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 (MDCT) 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 cranial nerve canal was measured on the axial images in each ear along the inner margin of its bony walls at its mid portion on an axial image through the base of the modiolus. 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.

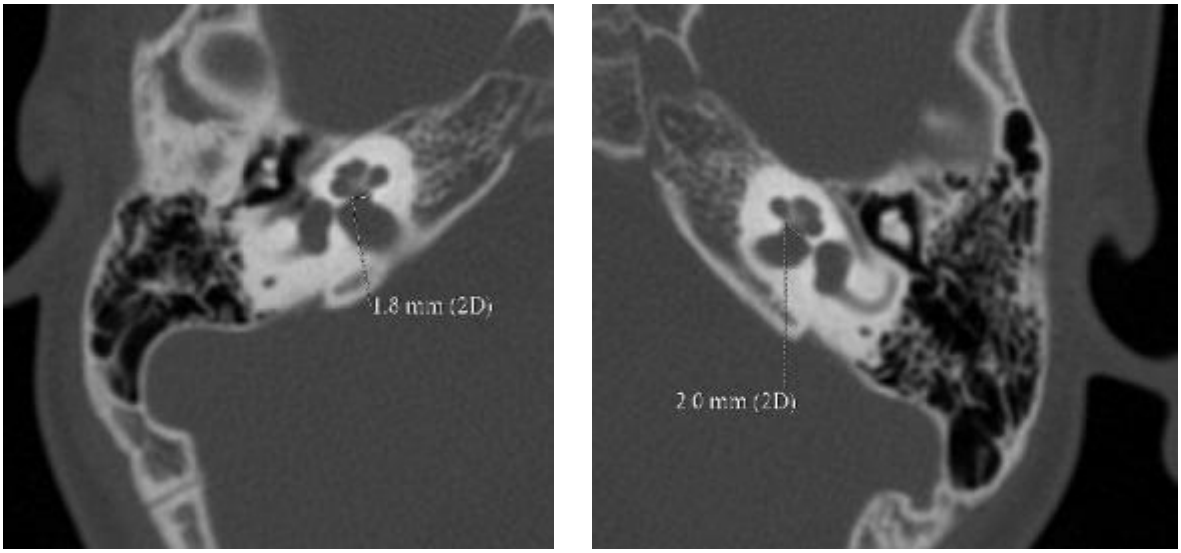


Fig.1.a. Normal bony canal for the cochlear nerve in a 6-year-old boy who underwent a temporal bone examination for SNHL. Transverse, thin-section CT scan through the midportion of the modiolus shows a

normal bony canal for the cochlear nerve. The canal width was found to be 1.8mm on the right side (a) and 2.0mm on the left side (b).

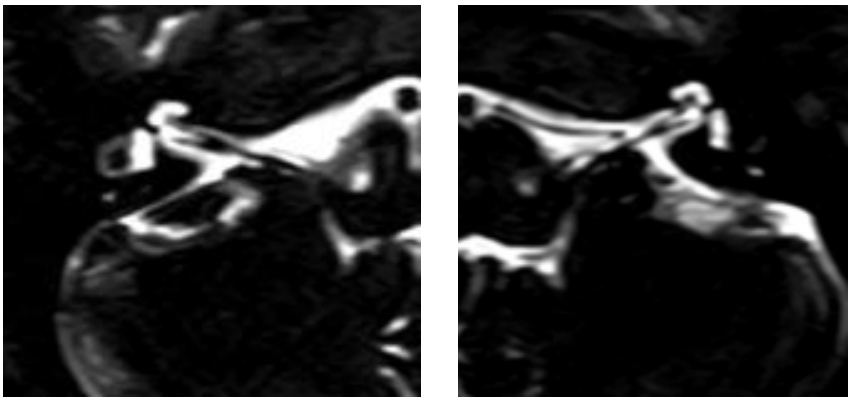


Fig. 1.b. 3DT2 MR images in the same patient demonstrating the bilateral cochlear nerves entering the cochlear canal, right side(a) and left side (b).

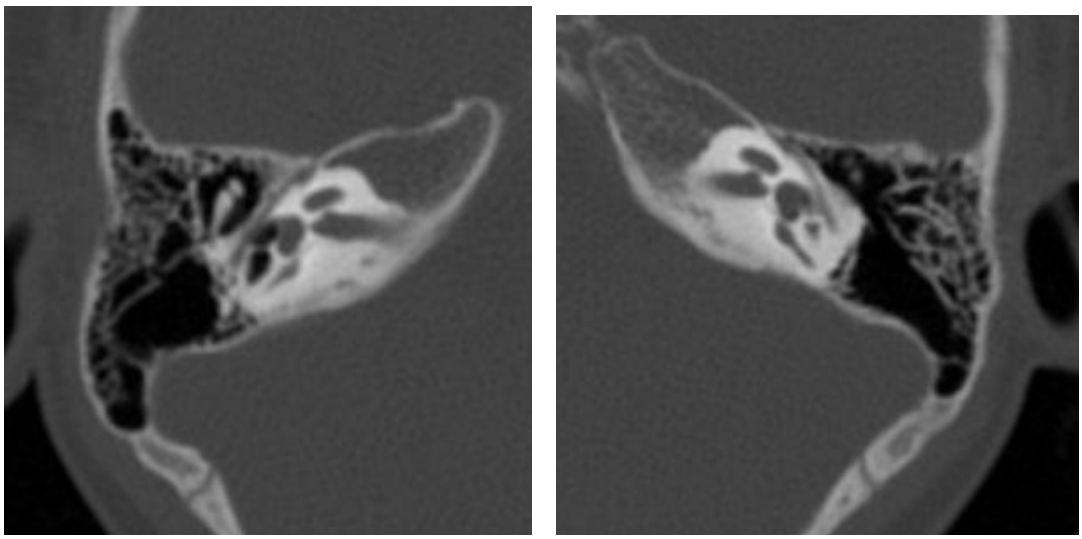


Fig.2. Hypoplastic bony canal for the cochlear nerve in a 7-year-old boy with SNHL. Transverse, thin-section CT scan through the midportion of the modiolus shows a hypoplastic bony canal for the cochlear nerve. The canal width was found to be 0.6mm bilaterally.

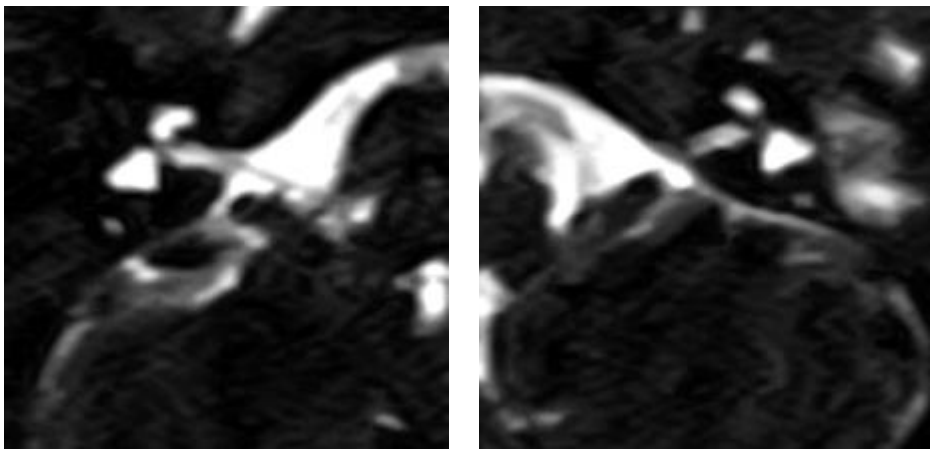


Fig. 2. In the same patient, there is absence of the cochlear nerve bilaterally on MRI 3DT2 sequence.

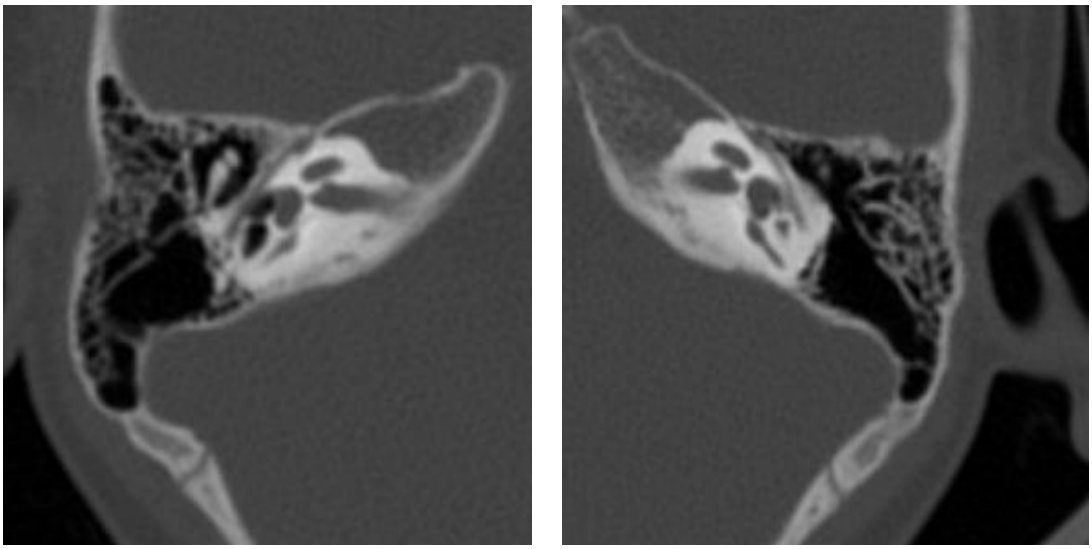


Fig. 3. 5 year old female child with SNHL showing bilateral stenotic cochlear nerve canal.

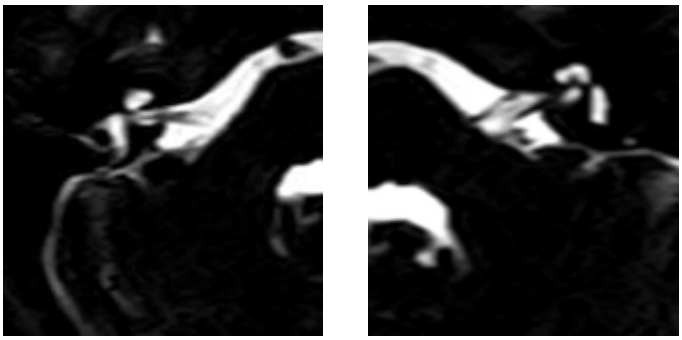


Fig. 3. In the same child, there is non visualisation of the cochlear nerve on both sides.

Results – The statistical analysis was done with the help of Student's T test. The total number of female patients included was 27 (55.1%). The mean age was found to be 6.67 with a standard deviation of 3.16. The mean (SD) cochlear nerve canal diameter was 1.67 (0.17) mm in patients with normal cochlear nerve and in patients with absent cochlear nerve, the mean (SD) diameter was found to be 0.79 (0.39) mm. The p value was found to be significant (<0.001).

Statistics

	Age (in yrs)
Mean	6.67
Std. Deviation	3.158

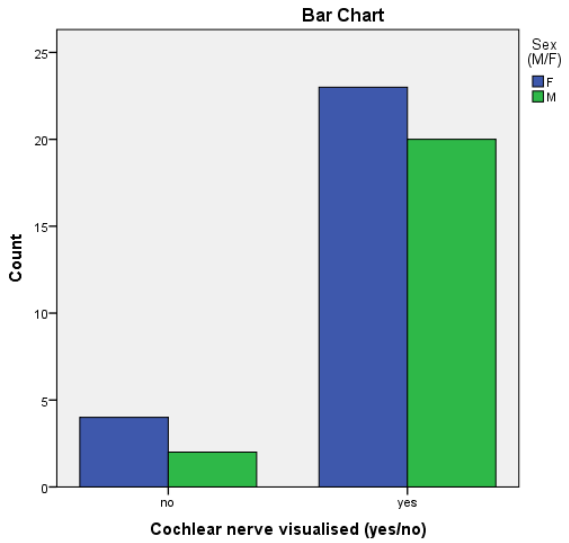
Sex (M/F)

		Frequency	Percent
Valid	F	27	55.1
	M	22	44.9
	Total	49	100.0

Cochlear nerve visualised (yes/no)

		Frequency	Percent
Valid	No	6	12.2
	Yes	43	87.8
	Total	49	100.0

Crosstab					
			Sex (M/F)		Total
			F	M	
Cochlear nerve visualised (yes/no)	no	Count	4	2	6
		% within Sex (M/F)	14.8%	9.1%	12.2%
	yes	Count	23	20	43
		% within Sex (M/F)	85.2%	90.9%	87.8%
Total		Count	27	22	49
		% within Sex (M/F)	100.0%	100.0%	100.0%



III. Discussion

Children with congenital sensorineural hearing loss (SNHL) require a complete analysis at the earliest. Diagnosis of the cause of SNHL and its correction are important in speech and language development for young children.(3) Cochlear implantation is considered an effective treatment for pediatric patients with congenital SNHL. The presence of a functioning cochlear nerve becomes an important issue in the preoperative assessment of pediatric cochlear implant candidates.(4) Cochlear nerve deficiency implies an absence or either a hypoplastic cochlear nerve. A hypoplastic cochlear nerve is a relative contraindication while the absence of the cochlear nerve is considered an absolute contraindication for cochlear implantation. Narrow or stenotic cochlear canal are associated with a hypoplastic or aplastic cochlear nerve. Narrow cochlear canal on computed tomography (CT) and lack of visualization of cochlear nerve on magnetic resonance imaging (MRI) has been shown to be related to poor outcome of cochlear implantation.

In this study, high resolution temporal bone CT was used to measure the cochlear aperture diameter whereas 3DT2 MRI high resolution sequence was used to identify the cochlear nerve. In all patients with HRCT temporal bone demonstrating a stenotic cochlear nerve canal, MR imaging revealed cochlear nerve hypoplasia/aplasia. However, in patients with mildly narrow (above 1.4mm) cochlear nerve canal on CT, the cochlear nerve was visualized on MRI.

Our study has demonstrated that congenital SNHL pediatric patients with very small or stenotic cochlear nerve canal were associated with aplasia /hypoplasia of the cochlear nerve.

Such a correlation may be helpful in identifying patients who may not benefit from cochlear implantation. Supportive treatment and hearing rehabilitation using a vibrotactile device can be applied and an auditory brainstem implant may be considered.

The exact cause of the smaller bony canal for the cochlear nerve in patients with SNHL is unclear. Labyrinthine development commences at approximately 3 weeks' gestation with the formation of the otic placode that will become the otic vesicle. The spiral organ of Corti develops from the cochlear duct and along with the fibers from the spiral ganglia together form the cochlear nerve at 7 weeks. The IAC is formed at the medial aspect of the otic vesicle by inhibition of cartilage formation which requires the presence of the vestibulocochlear nerve. A canal will not be formed in the absence of the nerve (6).

A study by Fatterpekar et al speculated that any anomaly in development of the otic vesicle may inhibit normal production of nerve growth factor resulting in excessive neuronal degradation and prevention of normal growth of the developing cochlear nerve(7).

Casselman et al have speculated in their study that just as a normal cochlear nerve is required for the developing internal acoustic meatus for attaining normal adult dimensions, it is likely that the bony cochlear nerve canal also requires a similar stimulus for its normal development.(8)

Glasgonbury et al found deficient cochlear nerves in patients with profound SNHL at high-spatial-resolution, T2-weighted, fast spin-echo magnetic resonance (MR) imaging performed before cochlear implantation.(9)

The study conducted by Fattepekar et al revealed that the cochlear apertures of patients with SNHL were significantly smaller then patients having normal hearing using temporal bone CT.(7)

The mean width of cochlear nerve canal was found to be 2.58mm in a study conducted by Stjernholm and Muren in 117 silicone temporal bone casts. From these measurements, abnormally narrow and wide CNC defined as less than 1.4mm and more than 3mm correspondingly.(10)

Similar studies conducted by Adunka et al and Miyakasa et al also concluded that identifying a narrow cochlear aperture can indeed indicate hypoplasia or aplasia of the nerve that runs through it, especially in children with profound hearing loss.(11,12) A more recent study by Pagarkar et al have also come to similar conclusions.(13)

IV. Conclusion

The findings of the present study suggest that CN deficiency is not an unusual cause of congenital hearing loss. In majority of the children with congenital sensorineural hearing loss the cochlear aperture was found to be normal in size on HRCT of the temporal bone with visualization of the cochlear nerve on MRI. However, in those children with a significantly narrow or stenotic cochlear aperture, the cochlear nerve was not visualized on MRI indicating that the nerve is either hypoplastic or aplastic. Hence, detection of a narrow/stenotic cochlear canal on HRCT of the temporal bone can be used a s a predictor in diagnosing cochlear nerve hyoplasia/aplasia and should prompt for an MRI to look for the presence of cochlear nerve especially in children with hearing loss in the severe to profound range and cochlear implantation is being considered.

References

[1]. M. B. St. Martin and B. E. Hirsch, "Imaging of hearing loss," Otolaryngologic Clinics of North America, vol. 41, no. 1, pp. 157–178, 2008.

[2]. JosephY. Young, MD Maura E. Ryan, MD Nancy M. Young, MD, "Preoperative Imaging of Sensorineural Hearing Loss in Pediatric Candidates for Cochlear Implantation", RadioGraphics 2014; 34:E133–E149, Published online 10.1148/rg.345130083.

[3]. Osberger MJ. Cochlear implantation in children under the age of two years: candidacy considerations. Otolaryngol Head Neck Surg 1997; 117: 145-149

[4]. Makhdoum MJ, Snik FM, van de Broek P. Cochlear implantation in deaf children. Ann Saudi Med 1997; 17: 533-539

[5]. Larsen WJ. Human Embryology. 2nd ed. New York, NY: Churchill Livingstone; 1997:385–411.

[6]. McPhee JR, Van De Water TR. Epithelial-mesenchymal tissue interactions guiding otic capsule formation: the role of the otocyst. J Embryol Exp Morphol 1986;97:1–24.

[7]. Fatterpekar GM, Mukherji SK, Alley J, Lin Y, Castillo M. Hypoplasia of the bony canal for the cochlear nerve in patients with congenital sensorineural hearing loss: initial observations. Radiology. 2000;215(1):243-246, PubMed.

[8]. Casselman JW, Offeciers EF, Govaerts PJ, et al. Aplasia and hypoplasia of the vestibulocochlear nerve: diagnosis with MR imaging. Radiology 1997; 202:773–781.

[9]. Glastonbury CM, Davidson HC, Harnsberger HR, Butler J, Kertesz TR, Shelton C. Imaging findings of cochlear nerve deficiency. AJNR Am J Neuroradiol. 2002;23(4):635-643, PubMed.

[10]. Stjernholm C, Muren C. Dimensions of the cochlear nerve canal: a radioanatomic investigation. Acta Otolaryngol. 2002;122(1):43-48, PubMed.

[11]. Adunka OF, Jewells V, Buchman CA. Value of computed tomography in the evaluation of children with cochlear nerve deficiency. Otol Neurotol. 2007;28(5):597-604, PubMed.

[12]. Miyasaka M, Nosaka S, Morimoto N, Taiji H, Masaki H. CT and MR imaging for pediatric cochlear implantation: emphasis on the relationship between the cochlear nerve canal and the cochlear nerve. Pediatr Radiol. 2010;40(9):1509-1516, PubMed.

[13]. Pagarkar W, Gunny R, Saunders DE, Yung W, Rajput K. The bony cochlear nerve canal in children with absent or hypoplastic cochlear nerves. Int J Pediatr Otorhinolaryngol. 2011;75(6):764-773, PubMed.

[14]. Eyal E. Russo, MDa, Spiros Manolidis, MDb, "Cochlear nerve size evaluation in children with sensorineural hearing loss by high-resolution magnetic resonance imaging", American Journal of Otolaryngology–Head and Neck Medicine and Surgery 27 (2006) 166– 172.

[15]. Yen-Ling Lin[1], Che-Ming Wu[2], Cheng-Hong Toh[1,4], Mun-Ching Wong[1,4], Sheung-Fat Ko[3,4], Kar-Wai Lui[1,4], Shu-Hang Ng[1,4], "Magnetic Resonance Imaging Evaluation for Predictive Factors of Cochlear Nerve Deficiency in Pediatric Cochlear Implant Candidates", J Radiol Sci June 2013 Vol.38 No.2.

[16]. D E Bamiou, P Phelps, T Sirimanna, "Temporal bone computed tomography findings in bilateral sensorineural hearing loss", Arch Dis Child 2000;82:257–260.

[17]. Girish M. Fatterpekar, MD, Amish H. Doshi, MD, Mohit Dugar, MD, Bradley N. Delman, MD, Thomas P. Naidich, MD, Peter M. Som, MD, "Role of 3D CT in the Evaluation of the Temporal Bone", RadioGraphics 2006; 26:S117–S132. Published online 10.1148/rg.26si065502.