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Congenital Internal Auditory Canal Stenosis

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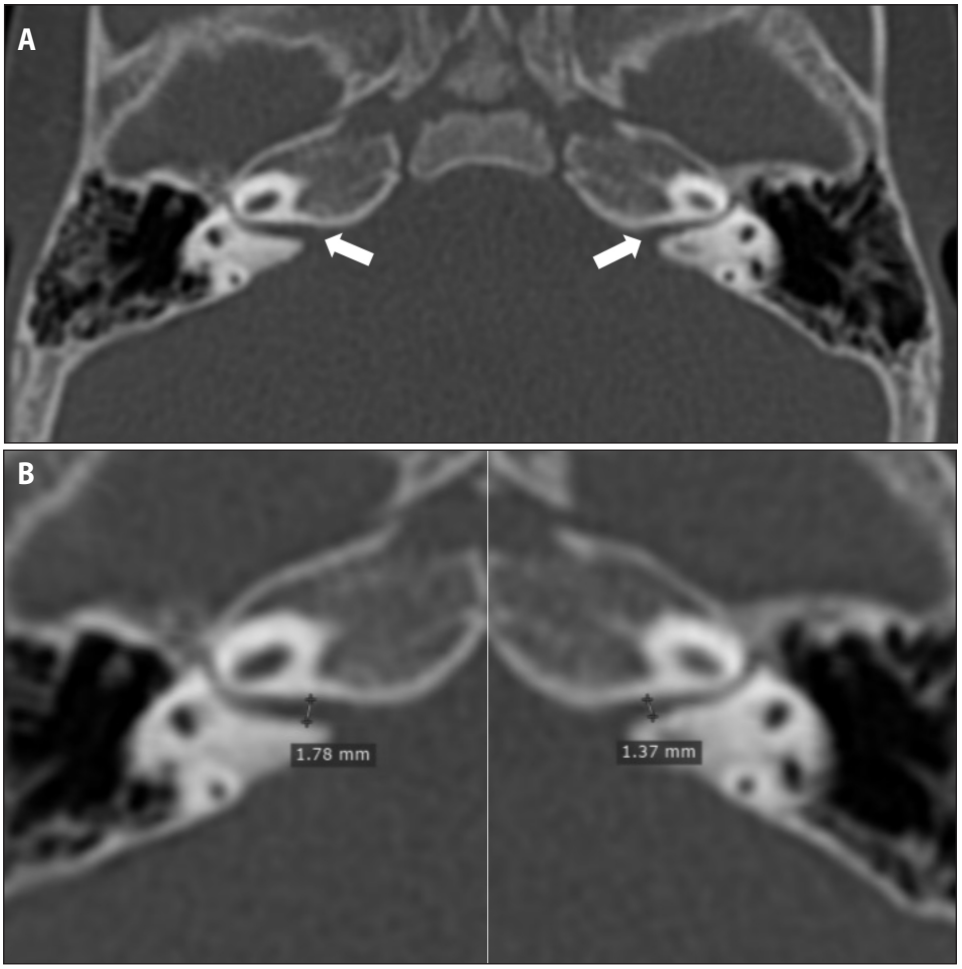
A 3-year-old boy underwent evaluation for possible cochlear implantation. He had failed a neonatal otoacoustic emission (OAE) hearing screen. A combined auditory brainstem response/ auditory steady-state response (ABR/ASSR) test battery confirmed the presence of a severe hearing loss on the right and a profound hearing loss on the left. No Joint Committee on Infant Hearing (JCIH) risk factors for early childhood hearing loss[1] were identified. Rehabilitation via hearing aid amplification and auditory-verbal speech therapy was unsuccessful. Computerized tomographic (CT) imaging of the temporal bone was performed to identify the presence of any inner ear abnormalities. No abnormalities of the cochlea, vestibule and semicircular canals on both sides were identified by the radiologist. The internal auditory canals were described as "fairly symmetrical without widening", and the study was officially reported as an "unremarkable study of the temporal bones".



Figures 1A and **1B.** Temporal Bone CT, axial plane, level of internal auditory canal. 1A shows the internal auditory canals (white arrows), which are nearly symmetrical but seemingly narrow when viewed subjectively. 1B Magnified views show the actual width of the internal auditory canals when measured using the DICOM imaging software length measurement tool. The width of the IAC is 1.78 mm on the right and 1.37 mm on the left. These are both below the cutoff value of 2 mm for the normal width of the IAC.

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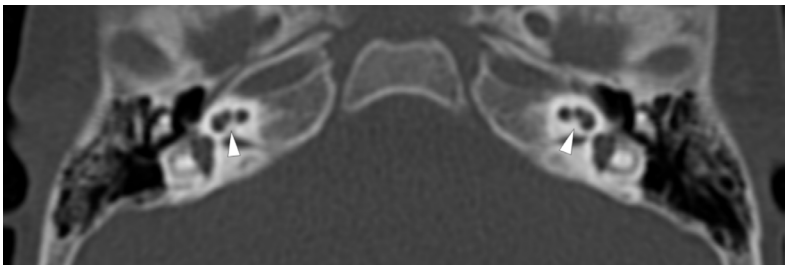

Figure 2. Temporal Bone CT, axial plane, mid-modiolar level. The bony cochlear nerve canals which are normally visually evident openings at the fundus of the IAC through which the cochlear nerve passes to enter the cochlea are both filled by bone.

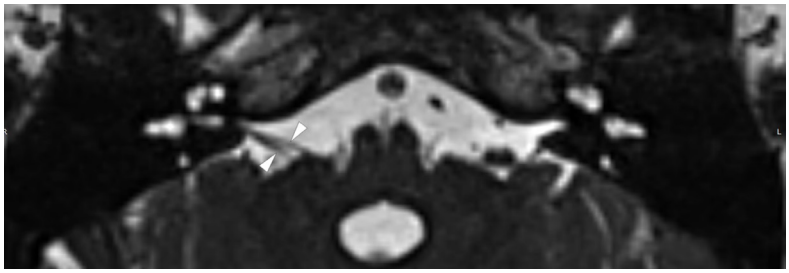

Figure 3. Temporal Bone MRI, axial plane, 3D CISS sequence, level of internal auditory canal. Two cranial nerves (white arrows) can be seen on the right side, originating from the brainstem, traversing the cerebellopontine angle cistern, and entering into the internal auditory canal. No similar structures are identifiable on the left side.

Independent review of the CT imaging revealed the presence of seemingly narrow internal auditory canals (IAC) on both sides. *(Figure 1A)* The width of the IACs on the axial plane were measured by drawing a perpendicular line starting from the posterior wall of the IAC, 2 mm inside the posterior lip of the internal auditory meatus, and ending on the anterior canal wall, as described by McClay *et al*.[2] Measurements taken utilizing the length measurement tool in the DICOM imaging software (RadiAnt DICOM Viewer, Version 2024.1, Medixant) indicated an IAC width of 1.78 mm on the right (with severe hearing loss) and 1.37 mm on the left (with profound hearing loss). *(Figure 1B)* These measurements confirmed the presence of bilateral internal auditory canal stenosis, a diagnosis defined by a canal of 2 mm or less on high-resolution CT.[3]

Furthermore, bilateral stenosis of the bony cochlear nerve canal (also known as cochlear aperture stenosis) was visually identified. *(Figure 2)*

Given the association of these two findings with cochlear nerve deficiency, a high-resolution three-dimensional T2-weighted MRI sequence was performed. The three-dimensional constructive interference in steady state (3D-CISS) sequence clearly showed the presence of two cranial nerves in the cerebellopontine angle cistern entering into the right internal auditory canal, but no similarly located neural structures on the contralateral side. This indicated the presence of a cochlear nerve deficiency on the side with profound hearing loss. *(Figure 3)*

Radiologically identifiable abnormalities of the inner ear and cochlear nerve are relatively common causes of pediatric sensorineural hearing loss, comprising up to 42% of cases.[4] Although stenosis of the internal auditory canal is among the less common abnormalities seen, it was observed in 12% of 98 ears of patients with congenital sensorineural hearing loss in a series by Jackler *et al*.[5] As such, its presence should be ascertained in patients with congenital hearing loss. Since the internal auditory canals can be bilaterally stenosed, mere visual determination of any canal size asymmetry as was initially done in this case will not suffice. Actual measurement of the internal auditory canal diameter is necessary.

Internal auditory canal stenosis has been associated with other abnormalities particularly hypoplasia or aplasia of the cochlear nerve and stenosis of the bony cochlear nerve canal.[6] Since all of these factors individually and collectively have an impact on hearing rehabilitation with external hearing devices and cochlear implants, identifying these conditions using the appropriate imaging study becomes necessary. As such, a clinician must accept the reality that both CT and MRI studies may be warranted in an individual patient, as these studies provide complementary information on the status of the hearing pathway.

REFERENCES

1. Joint Committee on Infant Hearing. Year 2019 Position Statement: Principles and guidelines for early hearing detection and intervention programs. Journal of Early Hearing Detection and Intervention. 2019;4(2): 1-44. DOI: https://doi.org/10.15142/fptk-b748.
2. McClay JE, Tandy R, Grundfast K, Choi S, Vezina G, Zalzal G, *et al.* Major and minor temporal bone abnormalities in children with and without congenital sensorineural hearing loss. *Arch Otolaryngol Head Neck Surg*. 2002 Jun;128(6):664-71. DOI: 10.1001/archotol.128.6.664; PubMed PMID: 12049561.
3. Sakina MS, Goh BS, Abdullah A, Zulfiqar MA, Saim L. Internal auditory canal stenosis in congenital sensorineural hearing loss. *Int J Pediatr Otorhinolaryngol*. 2006 Dec;70(12):2093-7. DOI: 10.1016/j.ijporl.2006.08.001; PubMed PMID: 16996619.
4. Simons JP, Mandell DL, Arjmand EM. Computed tomography and magnetic resonance imaging in pediatric unilateral and asymmetric sensorineural hearing loss. *Arch Otolaryngol Head Neck Surg*. 2006 Feb;132(2):186-92. DOI: 10.1001/archotol.132.2.186; PubMed PMID: 16490877.
5. Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis. *Laryngoscope*. 1987 Mar;97(3 Pt 2 Suppl 40):2-14. DOI: 10.1002/lary.5540971301; PubMed PMID: 3821363.
6. Tahir E, Bajin MD, Atay G, Mocan BÖ, Sennaroğlu L. Bony cochlear nerve canal and internal auditory canal measures predict cochlear nerve status. *J Laryngol Otol*. 2017 Aug;131(8):676-683. DOI: 10.1017/S0022215117001141; PubMed PMID: 28566097.