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Congenital Unilateral Hearing Loss from Cochlear Nerve Canal Stenosis

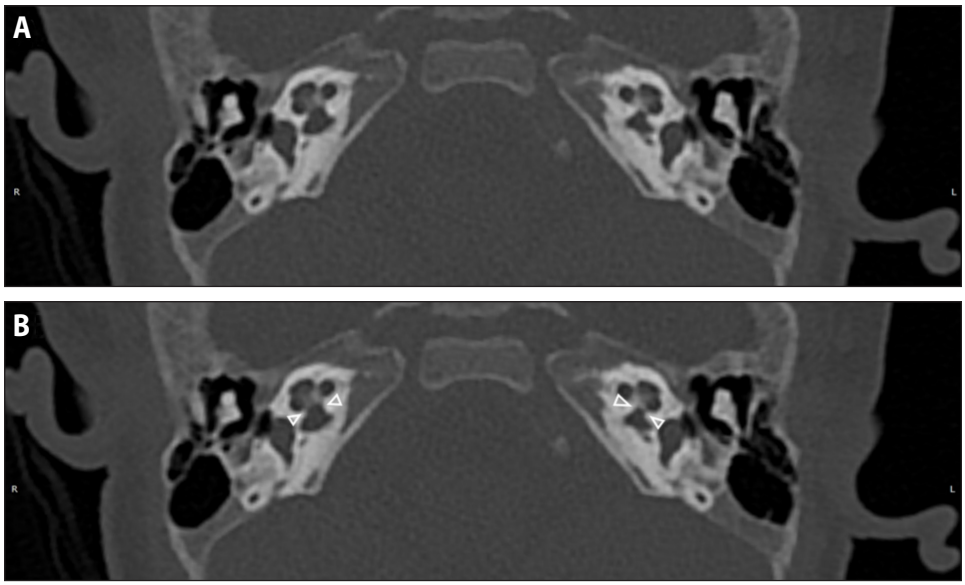
Nathaniel W. Yang, MD[1,2]

[1]*Department of Otolaryngology-Head and Neck Surgery College of Medicine - Philippine General Hospital University of the Philippines Manila*

[2]*Department of Otolaryngology -Head and Neck Surgery Far Eastern University - Nicanor Reyes Medical Foundation Institute of Medicine*

A 2-month-old baby girl presented with a failed neonatal otoacoustic emission (OAE) hearing screening in the left ear. Combined Auditory Brainstem Response/Auditory Steady-State Response (ABR/ASSR) testing confirmed the presence of a unilateral left moderate to severe hearing loss. No Joint Committee on Infant Hearing (JCIH) risk factors for early childhood hearing loss[1] were identified. She subsequently underwent computed tomography (CT) of the temporal bones to determine the presence of any inner ear malformation. No abnormalities of the internal auditory canal, cochlea, semicircular canals and ossicles were noted by the radiologist, and the study was officially reported as a "normal temporal bone CT scan."

Independent review of the CT imaging revealed the presence of a visually apparent disparity in the width of the cochlear nerve canals. *(Figure 1)* Measurement of the cochlear nerve canal width in the axial plane parallel to the infraorbitomeatal line[2] using the length measurement tool in the DICOM imaging software (RadiAnt DICOM Viewer, Version 2024.1, Medixant) indicated a cochlear nerve canal width of 2.18 mm on the right and 1.02 mm on the left. *(Figure 2)* Applying the suggested cutoff point of 1.2 mm as described by Lin *et al*.,[2] we identified the presence of left cochlear nerve canal stenosis as the etiology of the congenital unilateral hearing loss.



Figures 1A and **1B.** Temporal Bone CT, axial plane, mid-modiolar level. **1A.** shows the inner ear structures without any markings in order to provide the reader with an unobscured view of the relevant structure and its surrounding elements. **1B.** shows the cochlear nerve canals on each side bracketed by the paired unfilled white triangles. They are short bony canals between the base of the cochlea and the fundus of the internal auditory canal. The right cochlear nerve canal is visibly larger than the left. Determination of canal stenosis will depend on actual measurement of the canal width.

Correspondence: Dr. Nathaniel W. Yang
Department of Otolaryngology – Head and Neck Surgery
University of the Philippines Manila
Ward 10, Philippine General Hospital, Taft Avenue
Ermita, Manila 1000
Philippines
Phone: (632) 8526 4360
Fax: (632) 8525 5444
Email: nwyang@up.edu.ph

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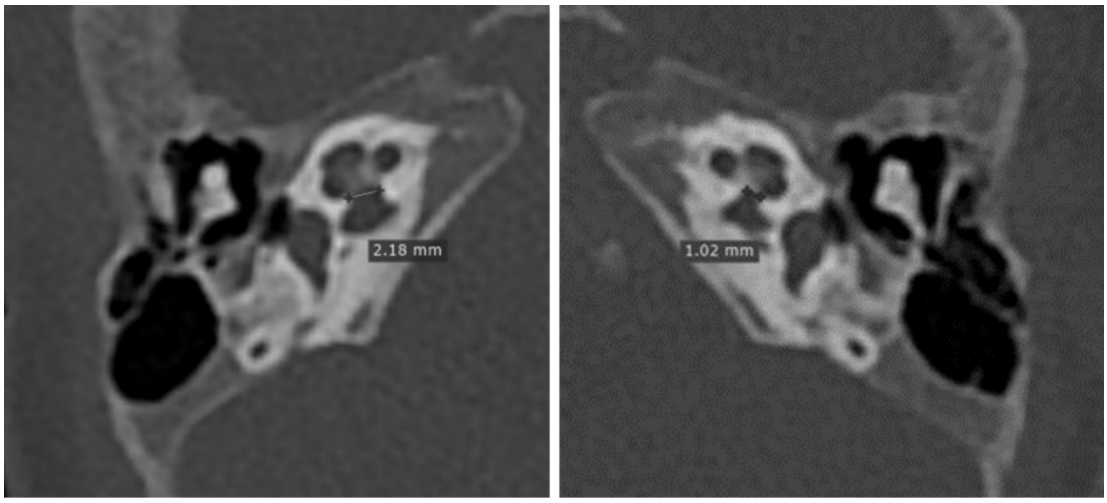

Figure 2. Magnified view of the cochlear nerve canals from *Figure 1*, which are mid-modiolar sections in an axial plane parallel to the infraorbitomeatal line. The width of each canal is determined using the DICOM imaging software length measurement tool. The width of the cochlear nerve canal is 2.18 mm on the right and 1.02 mm on the left.

The cochlear nerve canal, which has also been referred to as the bony canal for the cochlear nerve (BCNC), cochlear aperture, and cochlear fosette, is the bony transition point between the internal auditory canal and the cochlear modiolus. A relationship between a hypoplastic cochlear nerve canal and congenital sensorineural hearing loss was first suggested by Fatterpekar *et al.* in 2000.[3] Subsequent studies confirmed the association between cochlear nerve canal stenosis and sensorineural hearing loss that ranges from near-normal to profound, with a statistically significant relationship between the degree of hearing loss and the degree of stenosis.[4]

Various cutoff points to define stenosis of the cochlear nerve canal have been identified in the medical literature. These cutoff points range from 1.2 mm to 1.7 mm when the canal width is measured in the axial plane.[2] This particular case demonstrated clear-cut evidence of cochlear nerve canal stenosis, as it satisfied the smallest cutoff criteria (< 1.2 mm) seen in the medical literature.

The identification of cochlear nerve canal stenosis as the cause of congenital sensorineural hearing loss is important not only from a diagnostic point, but also from a prognostic perspective. Cochlear nerve deficiency has been noted to be highly prevalent among pediatric patients with cochlear nerve canal stenosis,[5] and this has significant negative implications in relation to rehabilitation with external hearing devices and cochlear implants.

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. Lim CH, Lim JH, Kim D, Choi HS, Lee DH, Kim DK. Bony cochlear nerve canal stenosis in pediatric unilateral sensorineural hearing loss. *Int J Pediatr Otorhinolaryngol*. 2018 Mar;106:72-74. DOI: 10.1016/j.ijporl.2018.01.015; PubMed PMID: 29447896.
3. 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 Apr;215(1):243-6. DOI: 10.1148/radiology.215.1.r00ap36243; PubMed PMID: 10751494.
4. Wilkins A, Prabhu SP, Huang L, Ogando PB, Kenna MA. Frequent association of cochlear nerve canal stenosis with pediatric sensorineural hearing loss. *Arch Otolaryngol Head Neck Surg*. 2012 Apr;138(4):383-388. DOI: 10.1001/archoto.2012.237; PubMed PMID: 22508622.
5. Dorismond C, Smetak MR, Perkins EL, Foust AM, Sarma A, Virgin FW Jr. High prevalence of cochlear nerve deficiency in pediatric patients with cochlear aperture stenosis. *Otolaryngol Head Neck Surg*. 2024 Apr 12. DOI: 10.1002/ohn.774; PubMed PMID: 38606641.