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.

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The author declared that this represents original material, that the manuscript has been read and approved by the author, that the requirements for authorship have been met by the author, and that the author believes that the manuscript represents honest work.

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FROM THE VIEWBOX

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. 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.

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. 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, and this has significant negative implications in relation to rehabilitation with external hearing devices and cochlear implants.

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