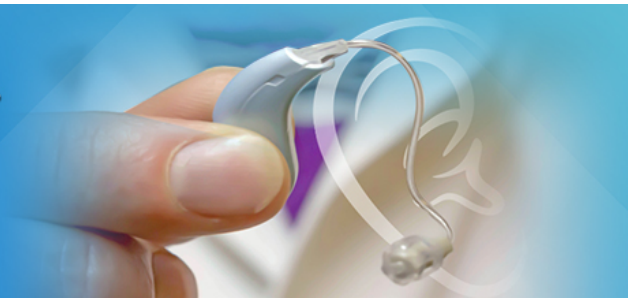
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Audiological and radiological profile of bilateral large vestibular aqueduct syndrome with Mondini dysplasia in a pediatric patient: A case report

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Abstract

Introduction: Bilateral Large Vestibular Aqueduct Syndrome (LVAS) is a congenital inner ear malformation frequently associated with *Mondini cochlear* dysplasia and is a well recognized cause of fluctuating sensorineural hearing loss in children. Early identification through radiological and audiological correlation is essential to guide timely rehabilitative decisions, including hearing aid fitting and cochlear implant candidacy.

Case presentation: A 6-year old female presented with bilateral hearing impairment and a history of hearing deterioration in the left ear over approximately six months. Previous audiometry documented profound hearing loss in both ears. The child had received intravenous steroid treatment for left sided sensorineural hearing loss, with only temporary improvement reported. A history of third-degree parental consanguinity was documented. MRI and high-resolution CT of the petrous temporal regions demonstrated bilateral enlarged vestibular aqueducts measuring 2.3 mm on the right and 2.0 mm on the left, with hypoplasia of the apical and middle portions of the cochlea, described radiologically as Mondini malformation. Pure tone audiometry performed on 23 June 2026 indicated severe mixed hearing loss bilaterally. Tympanometry revealed type A tympanogram in both ears, while distortion-product otoacoustic emissions were absent bilaterally. Auditory brainstem response testing indicated moderately severe hearing loss bilaterally, with wave V responses obtained till 80dBnHL. A bilateral hearing-aid trial using Oticon Zircon 2 RIC devices demonstrated aided responses within the speech spectrum except at 4kHz, with an aided sound-field average of approximately 25 dBHL and a speech discrimination score of 80%. The audiological recommendations included hearing-aid fine-tuning, frequency transposition, periodic hearing evaluation, auditory training, and ENT review. The ENT team documented bilateral cochlear implantation, followed by audio-verbal therapy, as the management plan

Conclusion: This case highlights the importance of correlating audiological and radiological findings in a child with progressive bilateral hearing loss, and underscores the role of comprehensive serial Audiological monitoring in guiding the transition from hearing aids to cochlear implantation in children with LVAS and Mondini malformation.

Keywords: Mondini dysplasia, large vestibular aqueduct, consanguinity, *Cochlear* implantation, hearing aid, auditory training

Introduction

Large Vestibular Aqueduct Syndrome (LVAS) is one of the most common radiologically identifiable inner ear malformations associated with childhood sensorineural hearing loss. It was first described radiologically by Valvassori and Clemis in 1978 [1], who reviewed 3,700 temporal bone tomograms and identified 50 patients with a vestibular aqueduct exceeding 1.5 mm in diameter at the midpoint, a threshold that remain in clinical use today (Valvassori *et al.*, 1978) [1]. LVAS represents a spectrum of arrested inner ear embryogenesis, ranging from an isolated enlarged aqueduct to more complex cochleovestibular malformations, and it has been suggested that the underlying defect arises from early arrest in the development of the endolymphatic duct and sac (Jackler *et al.*, 1989) [2]. LVAS is frequently associated with incomplete partition anomalies of the cochlea, most notably Mondini dysplasia (incomplete partition type II, IP-II) in which the cochlea demonstrates a hypoplastic apical turn (Sennaroglu *et al.*, 2017) [3].

Children with LVAS commonly present with congenital or early-onset hearing loss that may be stable or progressive, and sensorineural or mixed in nature (Jackler *et al.*, 1989; Sennaroglu *et al.*, 2017) [2, 3]. A characteristic feature is sudden or stepwise deterioration in hearing following relatively minor head trauma, barotrauma, or febrile illness, a phenomenon well documented in the pediatric LVAS literature (Levenson *et al.*,

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1989) [4]. Bilateral involvement is reported in a substantial proportion of cases and carries implications for early rehabilitative planning (Valvassori *et al.*, 1978) [1].

A significant proportion of enlarged vestibular aqueduct cases, both syndromic and non-syndromic are attributable to biallelic or monoallelic mutations in the SLC26A4 (pendrin) gene, which also underlies Pendred syndrome; genotype has been shown to correlate with the severity of hearing loss and the presence of associated cochlear malformation (Albert *et al.*, 2006; Huang *et al.*, 2023) [5, 6]. Diagnosis relies on a combination of high resolution CT of the temporal bone and MRI, which together characterize both the bony aqueduct dimensions and the associated cochleovestibular dysplasia, alongside a comprehensive serial Audiological test battery to document the trajectory of hearing loss and guide amplification versus implantable decisions.

This case is reported to illustrate the radiological and audiological progression of bilateral LVAS with Mondini deformity in a young child, highlighting the value of serial audiological monitoring in guiding rehabilitative decisions.

Case presentation

A 6 year old female child was referred for detailed Audiological evaluation with a complaint of hearing loss in both ears. Her mother reported that the child had profound hearing loss in the right ear since 4 years of age, following recurrent high fever, for which she was fitted with a hearing aid (Oticon, Zircon 2 RIC). She also reported to have objective vertigo after waking up. Over the preceding 6 months, the child had developed a progressive, sudden-onset hearing loss in the left ear. She was administered intravenous steroids for 4 days for the left ear, following which only temporary improvement in hearing thresholds was noted. No other significant medical history was reported.

Birth history revealed an uneventful pregnancy, full term normal delivery, normal birth weight (3 kg) and immediate birth cry. Family history was notable for third degree consanguinity between the parents; however, there was no significant history related to hearing loss. On clinical examination the tympanic membranes were intact bilaterally, and the oral cavity, oropharynx and external nasal framework were within normal limits. According to the caregiver's history, the child's responses to name calls and loud sounds were inconsistent, and she relied predominantly on visual cues and lip-reading for communication.

Radiological evaluation MRI brain and CT of the petrous temporal region were performed on 6 February 2026 imaging revealed bilateral large bony vestibular aqueducts with hypoplastic apical turn and middle third of the cochlea (right 2.3 mm, left 2.0 mm), consistent with bilateral large vestibular aqueduct syndrome with Mondini deformity, more marked on the right side. Slightly prominent vestibules were noted bilaterally. The superior, lateral and posterior semicircular canal appeared normal except for mild dehiscence of the posterior semicircular canal. Adenotonsillar hypertrophy was also noted. A mildly thinned corpus callosum with periventricular hyperintensities was noted, suggestive of a mild perinatal insult. No vascular abnormality or intracranial mass lesion was identified. The 5th, 7th and 8th cranial nerve complexes appeared normal bilaterally on MR neurogram.

Previous audiological evaluation was done at Harshini hospital, Madurai on 13/01/2026 & 16/06/2026 showed profound hearing loss in both ears on pure tone audiometry, and wave V up to 90 dBnHL (Severe to Profound loss) on ABR. A detailed Audiological test battery was carried out in MERF-ISH Pure tone audiometry performed on 23 June 2026 using piano inventis revealed both ears severe mixed hearing loss (Figure 1). Tympanometry performed using MAICO Touch type M134H, revealed type A tympanogram bilaterally with no indication of middle ear pathology (Figure 2). Ipsilateral and contralateral acoustic reflex were absent. Otoacoustic emission was performed using IHS Duet Jr system, which revealed that DPOAEs were absent bilaterally, suggestive of outer hair cell dysfunction in both ears (Figure 3). Electrophysiological assessment was carried out using IHS Duet Jr system, Auditory brainstem response showed that wave V could be obtained till 80dBnHL using click stimuli at a repetition rate of 19.3/s in rarefaction polarity. Both ears showed moderately severe hearing loss (Figure 4). A bilateral hearing aid trial was conducted using Oticon Zircon 2 RIC devices. Aided sound-field responses were reported to fall within the speech spectrum except at 4kHz (Figure 5). The audiological record documented an aided sound-field average of approximately 25 dBHL and a speech discrimination score of 80%. The audiological recommendations were to continue using the existing hearing aids with appropriate fine-tuning and frequency transposition, undergo periodic hearing evaluation every three months, participate in structured auditory training, and attend ENT review. The ENT team also documented bilateral cochlear implantation as a management plan. The final rehabilitation decision should be described according to the treating team's documented assessment and subsequent follow-up.

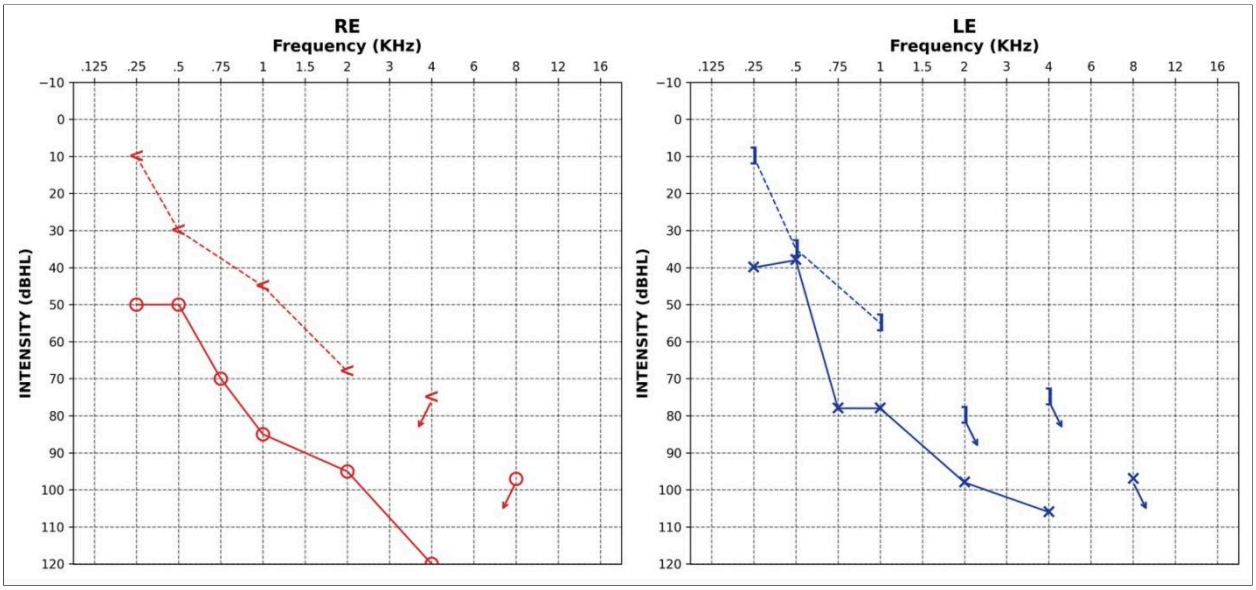


Figure 1. Pure tone Audiogram threshold of right and left ear

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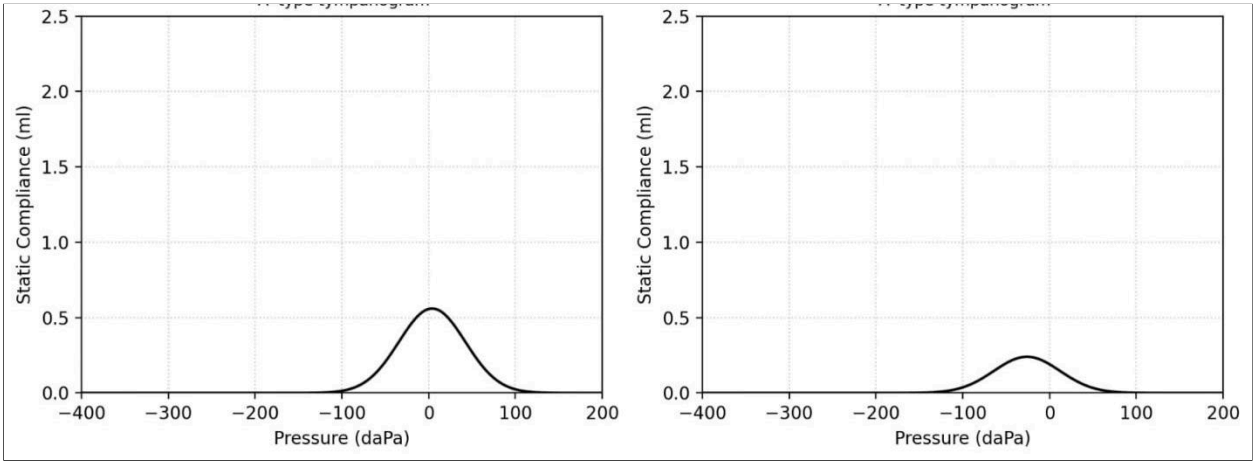


Figure 2. Tympanogram of the right and left ear

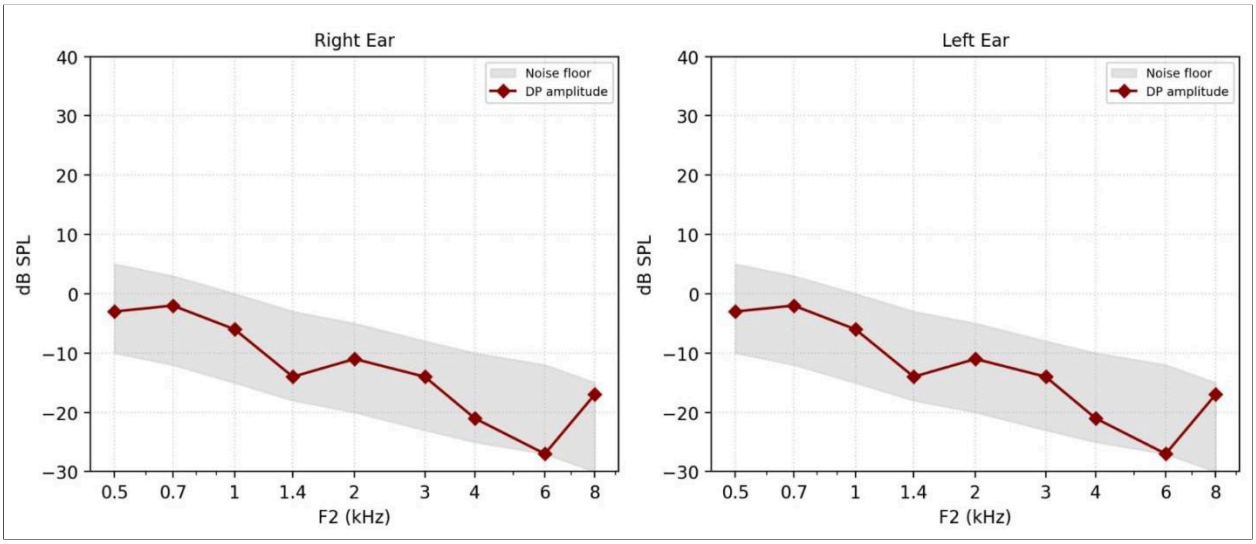


Figure 3. DPOAE responses of the right and left ear

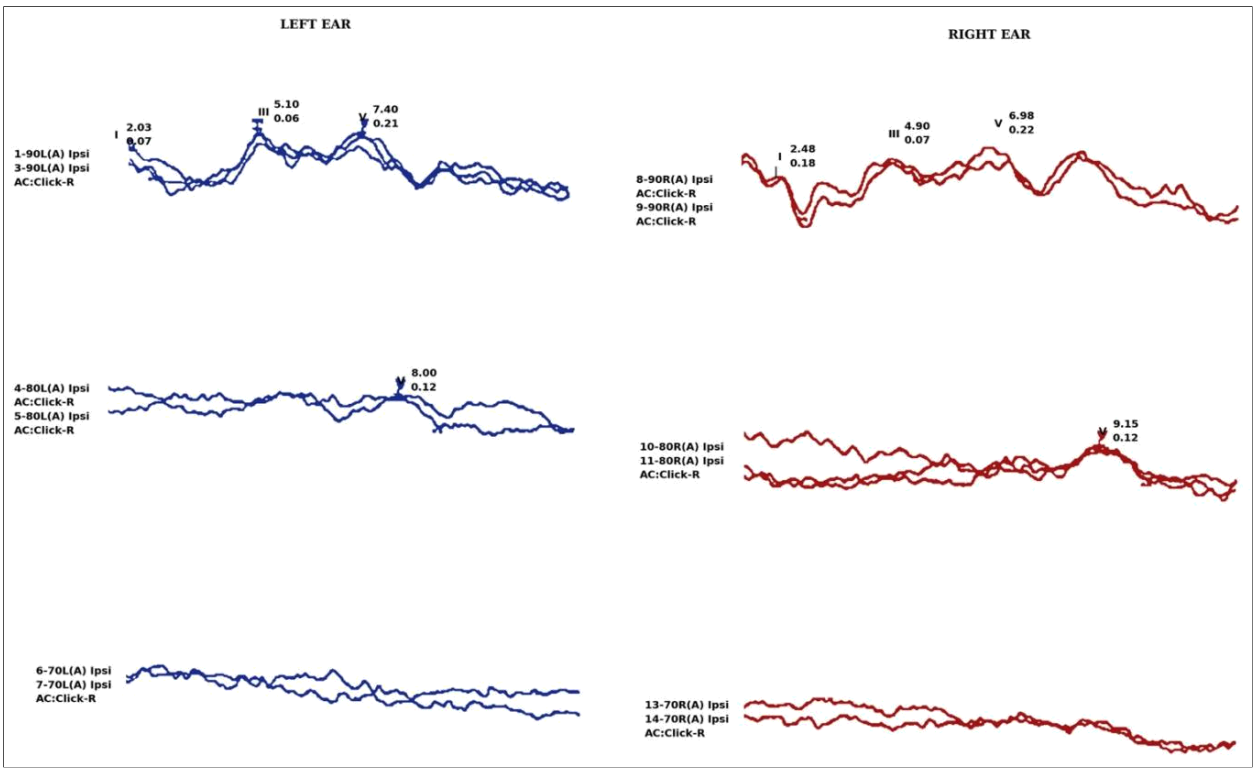


Figure 4. Auditory brainstem response (ABR) of the right and left ear (Threshold estimation)

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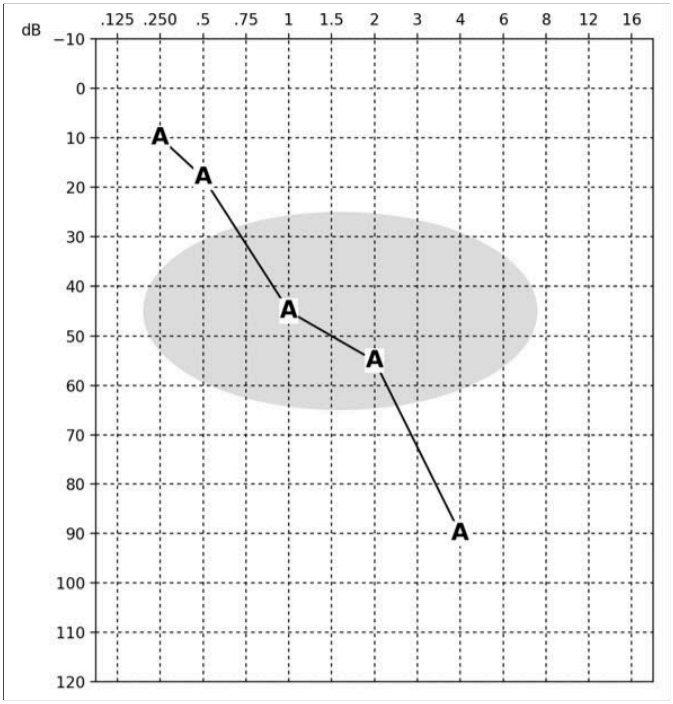


Figure 5. Aided response of bilateral (Oticon Zircon 2 RIC)

Discussion

This case demonstrates the classic radiological and audiological features of bilateral large vestibular aqueduct syndrome with associated *Mondini cochlear* dysplasia, presenting with a progressive pattern of hearing loss in a young child, with more marked radiological involvement on the right.

The child's history of profound right sided hearing loss following a febrile illness at 4 years of age, followed by a subsequent progressive decline in left sided hearing over 6 months, is consistent with the well documented tendency of LVAS ears to undergo sudden or stepwise deterioration in response to relatively minor physiological stressors such as fever, minor trauma or barotrauma (Jackler *et al.*, 1989; Levenson *et al.*, 1989) [2, 4]. This is thought to reflect abnormal transmission of intracranial or cerebrospinal fluid pressure through the dilated vestibular aqueduct to the perilymphatic and endolymphatic spaces of the inner ear.

Radiologically, the finding of bilateral enlarged vestibular aqueducts (right 2.3 mm, left 2.0 mm) with hypoplastic apical and middle turns of the cochlea, more pronounced on the right, is in keeping with the spectrum originally, described by (Valvassori and Clemis) [1] in which enlarged aqueducts co-ocuur with cochlear hypoplasia in a subset of cases (Valvassori *et al.*, 1978; Sennaroglu *et al.*, 2017) [1, 3].

Audiologically, the combination of absent DPOAEs bilaterally and elevated pure tone thresholds is consistent with cochlear based hearing loss involving outer hair cell dysfunction, while the presence of reproducible ABR wave V, at elevated thresholds together with normal 7th and 8th nerve complexes on MRI, argues against a significant retrocochlear component. The transition in PTA findings from bilateral profound loss on earlier ABR testing to a bilateral severe mixed loss on later testing may reflect a combination of true audiometric fluctuation, a well-recognized feature of LVAS (Jackler *et al.*, 1989) [2], together with inherent variability in behavioural threshold testing in a young child.

Although aided thresholds fall within the speech spectrum except at 4kHz however the child's everyday functional benefit from hearing aids appeared limited, as reflected by inconsistent responses to name calls and loud sounds and continued reliance on visual cues and lip reading. Together with the progressive hearing loss in the left ear, this is relevant to the ENT team's documented plan for bilateral cochlear implantation followed by audio-verbal therapy. *Cochlear* implantation in ears with EVA and associated Mondini/incomplete partition type II malformation has been shown in systematic reviews to produce favourable audiological and speech outcomes, broadly comparable to implantation outcomes in ears without inner ear malformations, although intraoperative CSF/perilymph gusher is reported more frequently in this group (pooled incidence approximately 27-49% depending on the presence of associated cochlear malformation) (Alahmadi *et al.*, 2022; Benchetrit *et al.*, 2022; Alahmadi *et al.*, 2023) [7, 8, 9]. This risk should be anticipated and appropriately managed intraoperatively and gusher occurrence has not been shown to adversely affect eventual speech perception outcomes (Benchetrit *et al.*, 2022) [8].

The history of third-degree parental consanguinity in this case raises the possibility of an autosomal recessive genetic contribution. Mutations in SLC26A4 are the most commonly identified genetic cause of both syndromic (Pendred syndrome) and non syndromic enlarged vestibular aqueduct, and are more frequently identified in patients with associated cochlear malformation and more severe hearing loss (Albert *et al.*, 2006; Huang *et al.*, 2023) [5, 6]. Genetic counseling and testing were not pursued in this case, representing a limitation, and would be a valuable addition to future evaluation of such children, both for prognostic counseling and family planning purposes.

This case reinforces the importance of a multidisciplinary approach, combining radiological, audiological and where possible genetic evaluation, in child presenting with progressive or asymmetric bilateral hearing loss, which can enable timely and appropriate rehabilitative intervention.

Limitations

The history of right-ear hearing-loss onset was inconsistent across the available records (documented as since birth, since 4 years of age and for 2 years in different notes). Differences were also noted between earlier and subsequent audiological findings, limiting interpretation of the hearing-loss progression. Genetic testing and long term rehabilitation outcome data were unavailable. As a single case report, the findings cannot be generalized.

Patient consent and ethics

Written informed consent for publication of this case report, including clinical details and images, was obtained from the parents/legal guardians of the child.

Conclusion

This case highlights the importance of correlating audiological and radiological findings in children with bilateral large vestibular aqueduct syndrome and Mondini dysplasia. Serial audiological monitoring, hearing-aid optimization and multidisciplinary follow-up are essential for rehabilitation planning and cochlear implant candidacy evaluation.

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This study received no financial support.

Author's contribution

Author 2 & Author 3 contributed to data collection, evaluation and manuscript preparation. Author 1 & Author 4 supervised the study and reviewed the manuscript.

Conflict of interest

The authors declare no conflict of interest.

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