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Cochleair Implantation and Electrophysiological Testing in Cochlear Nerve Hypoplasia

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Abstract | **Original Research Article**

Background: Cochlear nerve hypoplasia (CNH) is a congenital anomaly characterized by underdevelopment or absence of the cochlear nerve and represents an important cause of congenital profound sensorineural hearing loss. Cochlear implantation (CI) in patients with CNH remains controversial due to the reduced number of functional nerve fibers, which may limit electrical signal transmission and result in variable auditory outcomes. Accurate preoperative evaluation of both anatomical and functional integrity of the auditory pathway is therefore essential, and a multimodal approach is required, as no single test can reliably predict cochlear implant benefit. ***Materials and Methods:*** We report two pediatric cases of bilateral prelingual profound deafness who underwent comprehensive preoperative assessment. Evaluation included behavioral audiometry, auditory brainstem response (ABR) testing, high-resolution computed tomography (CT) of the temporal bone, and magnetic resonance imaging (MRI) with parasagittal views of the internal auditory canal to assess cochlear nerve morphology. Functional assessment consisted of intraoperative promontory stimulation, electrically evoked auditory brainstem responses (EABR), and cochlear implant telemetry with impedance measurements and electrically evoked compound action potentials (ECAPs). Cochlear implantation was performed using a perimodiolar electrode array. ***Results:*** Both patients demonstrated bilateral profound sensorineural hearing loss with absent ABR wave V responses at maximal stimulation. Imaging revealed severe cochlear nerve hypoplasia or aplasia associated with inner ear malformations, including internal auditory canal stenosis and vestibular abnormalities, while cochlear morphology was preserved. Despite these findings, promontory testing elicited reproducible low-amplitude responses at high stimulation thresholds, indicating residual neural excitability. Intraoperative EABRs confirmed the presence of electrically evoked responses with reduced amplitude and prolonged latency. Acceptable impedance values and recordable ECAPs were obtained, suggesting effective electrical stimulation of remaining neural elements. Postoperative auditory-verbal rehabilitation was initiated in both children, with ongoing follow-up. ***Conclusion:*** Cochlear implantation in patients with cochlear nerve hypoplasia remains challenging but is not absolutely contraindicated. A comprehensive multimodal assessment is crucial to identify residual auditory nerve function and guide individualized clinical decision-making. Careful counseling is necessary to establish realistic expectations regarding auditory and language outcomes.

keywords: Cochlear nerve hypoplasia, Cochlear implantation, Profound sensorineural hearing loss, Promontory stimulation test.

INTRODUCTION

Cochleair nerve hypoplasia (CNH) is a congenital anomaly characterized by the underdevelopment or absence of the cochlear nerve. It is increasingly recognized as a cause of congenital profound sensorineural hearing loss.

Cochlear implantation (CI) in these patients remains controversial due to the anatomical and functional limitations of the auditory nerve, which may result in suboptimal auditory outcomes. Therefore, preoperative identification of neural structures through imaging and electrophysiological testing is critical to determine candidacy and predict the effectiveness of implantation. However, no single test is sufficient, and a comprehensive approach is required to guide clinical decision-making.

MATERIALS AND METHODS

This case series involves two pediatric patients with bilateral prelingual profound deafness. A thorough preoperative assessment was conducted, including:

- Behavioral audiometry to confirm the severity of the hearing loss.
- Auditory Brainstem Responses (ABR) to evaluate the presence or absence of neural responses to acoustic stimuli.
- High-resolution computed tomography (CT) of the temporal bone to assess cochlear morphology, internal auditory canal (IAC), and semicircular canals.
- MRI with parasagittal sequences focused on the internal auditory canal to evaluate the cochlear nerve and detect any hypoplasia or aplasia.
- Intraoperative promontory testing using surface electrode stimulation and recording of electrically evoked auditory brainstem responses (EABR) to assess the functional integrity of the auditory pathway.
- Intraoperative cochlear implant telemetry, including impedance measurements and electrically evoked compound action potentials (ECAPs).

RESULTS

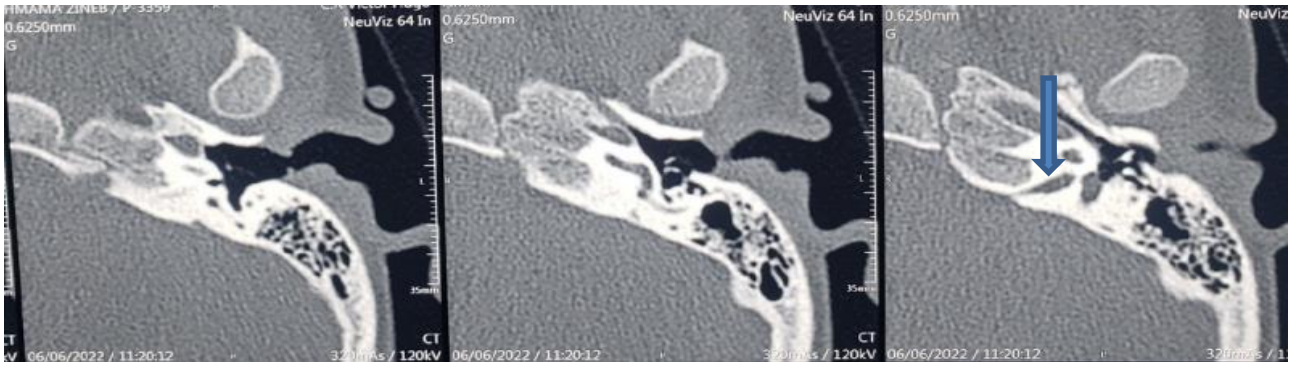
The first case concerned a 3-year-old boy with a medical history of meningitis at the age of 8 months. He presented with prelingual bilateral profound hearing loss, which was suspected at 9 months due to a lack of response to sound. The second case involved a 5-year-old girl, the only child of a consanguineous first-degree union. She had a history of neonatal jaundice and presented with delayed speech development, poor auditory responsiveness, and social disengagement. Both children underwent a thorough audiological evaluation. Behavioral audiometry confirmed profound bilateral deafness in both cases. Auditory Brainstem Response (ABR) testing revealed the absence of wave V bilaterally at 100 dB, consistent with severe cochlear nerve dysfunction. Tympanometry was Type A in both children.

Temporal bone CT scans showed significant inner ear malformations, including hypoplasia of the cochlear nerve canal, internal auditory canal (IAC) stenosis or atresia, hypoplasia of the lateral semicircular canals, and vestibular dilation. High-resolution MRI with parasagittal views of the IAC further confirmed severe cochlear nerve hypoplasia or aplasia, with thinning or absence of the nerve bundle, while cochlear morphology appeared preserved.

Despite these anatomical anomalies, a promontory test was performed under general anesthesia to assess functional neural integrity. In both patients, the test was positive, with reproducible but low-amplitude electrically evoked responses recorded at high stimulation thresholds. These findings indicated some degree of residual neural excitability, though limited. Intraoperatively, electrically evoked auditory brainstem responses (EABRs) were present at high current levels, consistent with the promontory test results. These waveforms, although small in amplitude, confirmed partial but functional conduction through the cochlear nerve.

Cochlear implantation was performed on the right ear in Case 1 and the left ear in Case 2. A perimodiolar electrode array was chosen to ensure optimal proximity to the residual nerve fibers. Intraoperative impedance testing showed acceptable values across all electrodes. Electrically Evoked Compound Action Potentials (ECAPs) were successfully recorded at several electrode sites, suggesting effective neural stimulation despite radiological evidence of hypoplasia.

Postoperatively, both children were enrolled in intensive auditory-verbal rehabilitation. Long-term follow-up is ongoing to monitor auditory perception and language acquisition, which will ultimately determine the functional benefit of cochlear implantation in these complex neuro-otological profiles.



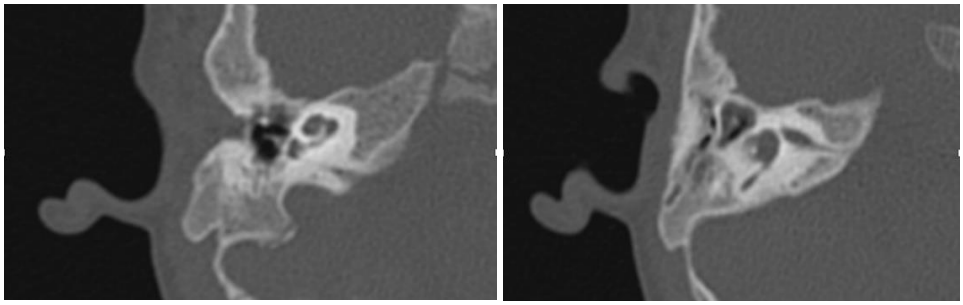


Figure 1,2: CT scan of the temporal bone, axial cut, left and right side (hypoplasia of the cochlear nerve canal, internal auditory canal (IAC) stenosis or atresia

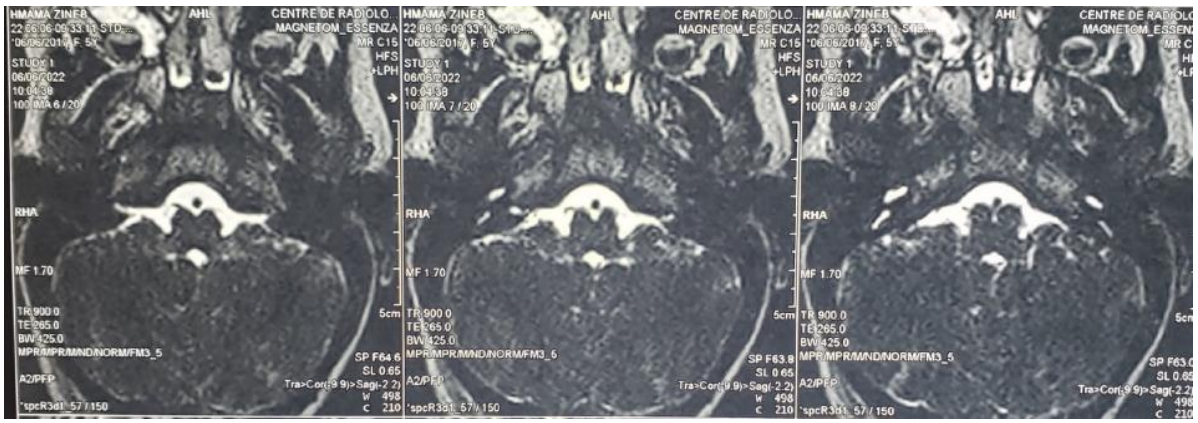


Figure 3: MRI of the temporal bones, axial cut: atresia of the internal auditory canal on the left side, with hypoplasia of the vestibulocochlear nerve on the right

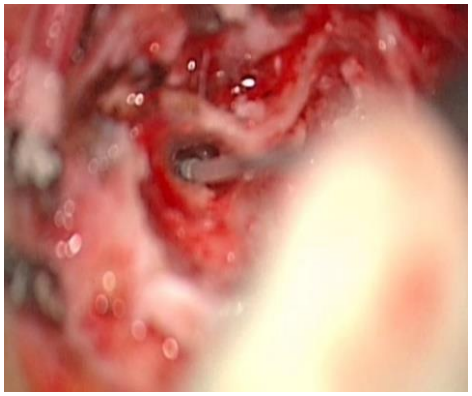


Figure 4: Electrocochleography probe insertion throught the round window

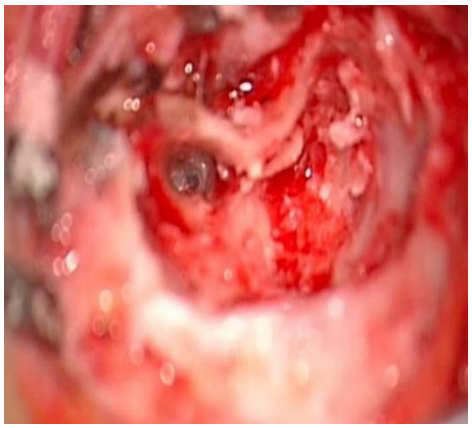


Figure 5: Operative vieuw after round window identification throught transmastoid posterior tympanotomy, left ear

DISCUSSION

Cochleair nerve hypoplasia (CNH) represents a significant challenge in the management of prelingual deafness. The reduced density of functional nerve fibers often limits the effectiveness of the electrical stimulation delivered by cochlear implants.

Imaging, particularly 3T MRI with parasagittal reconstructions, allows for fine anatomical evaluation of the cochlear nerve and the internal auditory canal and allows its classification. The classification of cochlear nerve hypoplasia, as proposed by *Sennaroglu et al.,* (2011), distinguishes several types based on the anatomy of the nerve and its involvement in deafness. This classification is crucial for assessing cochlear implantation possibilities. (table 1)

Type	Affected nerve on imaging	Remarks
I	Cochleovestibular nerve	The labyrinth may be normal or dysplastic, the internal auditory canal is stenotic
IIa	Cochlear branch with labyrinth dysplasia	Labyrinth dysplasia ranges from a minor dysplasia, like in case 3, to a common cavity
IIb	Cochlear branch with normal labyrinth	
III ?	Vestibular branch	Was anticipated not to exist as an isolated aplasia and has not been reported so far

The promontory test, while often used to assess the cochlear nerve's ability to respond to electrical stimulation, may not provide normal responses in cases of severe CNH. This results in low amplitude responses, sometimes absent, indicating nerve dysfunction even when the nerve is anatomically present.

In addition, intraoperative EABR recordings also showed low amplitude responses with prolonged latency, further supporting the hypothesis of abnormal development of the nerve or its pathways. These atypical electrophysiological profiles are frequently observed in children with CNH and should not be confused with typical responses. Furthermore, chronic stimulation of a hypoplastic nerve does not appear to induce normal maturation of the auditory brainstem and may even promote abnormal development of certain neural pathways.

It is also important to note that the abnormal response cannot be predicted by the severity of the hypoplasia of the nerve or the internal auditory canal. This unpredictability complicates the estimation of the expected benefits. In the literature, although initial improvement in auditory perception may be observed, children with CNH typically do not achieve the same outcomes as those with an intact auditory nerve. This is further complicated by associated syndromes, such as CHARGE syndrome, which can worsen the prognosis.

Therefore; clear and realistic information must be provided to families regarding expectations for auditory and language development. The benefit-risk ratio of implantation should always be evaluated on a case-by-case basis

CONCLUSION

Cochleair implantation in patients with cochlear nerve hypoplasia remains a challenging but not contraindicated procedure. These observations underscore the importance of a multimodal and personalized preoperative evaluation. However; a favorable radiological profile, even with atypical electrophysiological responses, does not constitute an absolute contraindication to implantation.

Declarations

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Ethics Approval and Consent to Participate Informed consent was obtained from all individual participants included in the study.

Consent to Publish The authors affirm that human research par- ticipants provided informed consent for publication of the images in all Figures.

Informed Consent Written informed consent was obtained from all subjects (patients) in this study.

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