ORIGINAL RESEARCH PAPER
Volume - 15 | Issue - 04 | April - 2026 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

COCHLEAR IMPLANTATION IN A BILATERAL PROFOUND HEARING LOSS CHILD WITH LEFT APLASTIC AND RIGHT HYPOPLASTIC INTERNAL AUDITORY CANAL: A CASE REPORT

Otorhinolaryngology

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KEYWORDS

Summary of Case report:
This case report describes a 3.5-year-old female presenting with pre lingual deafness with a history of consanguineous parents. Previous use of conventional hearing aids provided no benefit. Audiological evaluation confirmed bilateral profound hearing loss. Behavioral observation audiometry revealed no responses and otoacoustic emissions were absent bilaterally. Aided audiometry helped significant in-patient ear selection for cochlear implant as minimal responses were observed in left ear and absent responses in right ear. Brainstem evoked response audiometry showed absent wave V in both ears, consistent with profound sensorineural hearing loss. Radiological assessment was crucial for management. MRI revealed a hypoplastic right internal auditory canal with single nerve bundle where individual nerves could not be identified while the left cochlear nerve was not identifiable but with a narrow cochlear aperture. High-resolution CT confirmed severe stenosis of the right internal auditory canal and non-visualization of the labyrinthine segment and first genu of the facial nerve while 5 mm width internal auditory canal was noted on the left with narrow aperture. Cochlear implantation was performed in the left ear via standard mastoidectomy and facial recess approach, with full electrode insertion into the scala tympani using a Cochlear™ Nucleus® 7 (CP1002) implant. The postoperative course was uneventful. At three months follow-up, the child demonstrated meaningful auditory improvement, reflected in favorable Category of Auditory Performance and Meaningful Auditory Integration Scale scores. This case highlights the importance of thorough audiological and radiological assessment, individualized surgical planning, and careful preoperative evaluation to achieve successful cochlear implantation outcomes in children with complex auditory anatomy.

List of Abbreviations

Sr. No.	Abbreviation	Full Form
1	BOA	Behavioural Observation Audiometry
2	BERA	Brainstem Evoked Response Audiometry
3	CAP	Category of Auditory Performance
4	CI	Cochlear Implant
5	CSF	Cerebrospinal Fluid
6	CT	Computed Tomography
7	HRCT	High-Resolution Computed Tomography
8	IAC	Internal Auditory Canal
9	MAIS	Meaningful Auditory Integration Scale
10	MRI	Magnetic Resonance Imaging
11	NRT	Neural Response Telemetry
12	SNHL	Sensorineural Hearing Loss
13	TEOAE	Transient Evoked Otoacoustic Emissions
14	VRA	Visual Reinforcement Audiometry

INTRODUCTION

Hearing is a vital sensory modality essential for speech, language development, and social interaction. Hearing impairment is often described as a "silent disability," as it frequently remains undetected during early childhood, leading to delayed speech and language acquisition. Unlike visual impairment, hearing loss is less apparent and is often underestimated despite its profound impact on quality of life. In India, hearing loss is the second most common cause of disability, affecting approximately 63 million individuals (6.3% of the population). Data from the National Sample Survey Organization indicate that nearly 291 persons per lakh population suffer from severe-to-profound hearing loss, with a significant proportion comprising children aged 0–14 years, resulting in long-term educational, social, and economic consequences (1).

Pediatric sensorineural hearing loss (SNHL) has a multifactorial etiology, with genetic causes accounting for nearly 50% of cases, followed by acquired and idiopathic causes (2). Congenital SNHL is frequently associated with abnormalities of the inner ear and auditory pathways. Radiological studies have demonstrated that approximately 20% of congenital SNHL cases are associated with bony inner ear malformations, while the remaining cases are attributed to membranous labyrinthine abnormalities with normal bony architecture (3). Anomalies involving the internal auditory canal (IAC) and cochleovestibular nerve complex, although rare, are clinically significant due to their implications for auditory rehabilitation and prognosis (1,4).

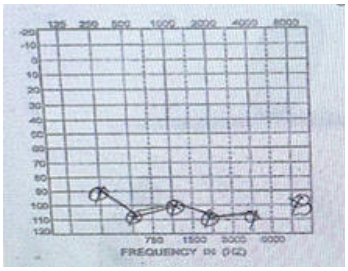
Cochlear implantation (CI) is an established and effective modality for auditory rehabilitation in children with congenital severe-to-profound SNHL who derive minimal or no benefit from conventional hearing aids. Cochlear implants bypass damaged sensory hair cells and directly stimulate the auditory nerve, resulting in significant improvement in auditory perception and speech development (5,6). Advances in surgical techniques and a better understanding of inner ear malformations have broadened the indications for cochlear implantation; however, implantation in children with cochlear nerve deficiency or IAC anomalies remains challenging, with variable postoperative outcomes (6,7).

Detailed preoperative radiological assessment using high-resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) is essential for evaluating inner ear anatomy, cochlear nerve integrity, and surgical feasibility in such complex cases (3,7). The presence of cochlear nerve deficiency in a child with bilateral profound hearing loss poses a unique clinical challenge with respect to ear selection, surgical planning, and prognostication. Reporting such cases contributes to improved understanding of anatomical variations, radiological–clinical correlation, and outcomes of cochlear implantation in children with complex auditory anomalies.

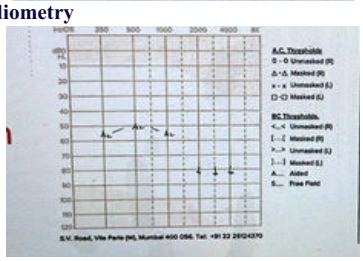
Case Report

A 3.5-year-old female child was brought by her parents with complaints of delayed speech and language development and poor response to sound since early childhood. The child was born at full term through normal delivery and had no history of perinatal complications, neonatal intensive care unit admission, ototoxic drug exposure in mother, meningitis, or recurrent ear infections. There was no significant past medical or surgical history. The parents reported a history of consanguineous marriage. The child had previously been fitted with conventional hearing aids; however, there was no improvement in auditory responsiveness or speech development.
Audiological evaluation revealed findings consistent with profound hearing impairment. Behavioural observation audiometry (BOA) revealed no responses suggestive of bilateral severe-to-profound hearing loss. Transient evoked otoacoustic emissions (TEOAE) were absent in both ears. Pure tone audiometry was suggestive of bilateral profound hearing loss so aided audiometry was done using visual reinforcement audiometry (VRA) with warble tones in the sound field, that demonstrated minimal response levels in the aided condition in the left ear, while no responses were observed in the right ear. Brainstem evoked response audiometry (BERA) showed poor wave morphology and repeatability, with absence of wave V in both ears, suggestive of

bilateral profound sensorineural hearing loss. Clinically and with evidence shown by the patient's mother, it was concluded that patient has better hearing in left so decision of better ear to be operated was taken.

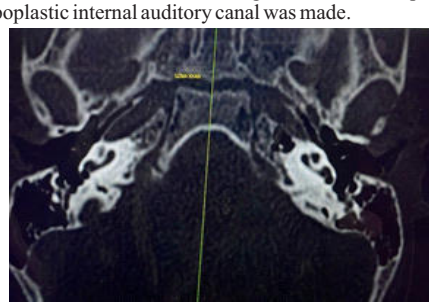


Pure tone audiometry



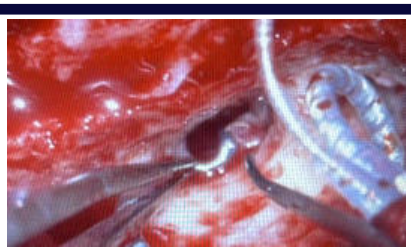
Aided audiogram

Radiological evaluation was carried out as part of cochlear implant candidacy assessment. Magnetic resonance imaging (MRI) of the brain and inner ear revealed mild asymmetric thinning of the cisternal segments of the right seventh and eighth cranial nerves with a hypoplastic right internal auditory canal and effacement of the cerebrospinal fluid (CSF) space within the right internal auditory canal (IAC) which limited visualization of the individual cranial nerves so only a single nerve bundle was visualized in IAC. On the left side, the facial nerve, superior vestibular nerve, and inferior vestibular nerve were identified separately within the internal auditory canal while the left cochlear nerve was not visualised and the left cochlear aperture appeared narrow. High- resolution computed tomography (HRCT) of the temporal bone, performed under sedation using 0.6-mm slice collimation on a 128-slice scanner, demonstrated severe stenosis of the right internal auditory canal. The labyrinthine portion and first genu of the bony facial nerve were not visualized on the right side. Left internal auditory canal width measuring about 5 mm with narrow aperture , with normal cochlear structure and facial nerve bony course. Based on clinical, audiological, and radiological findings, a diagnosis of bilateral profound sensorineural hearing loss with Left Aplastic and Right Hypoplastic internal auditory canal was made.

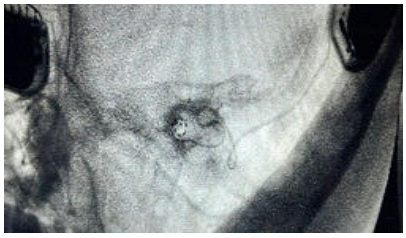


HRCT of bilateral temporal bone and inner ear

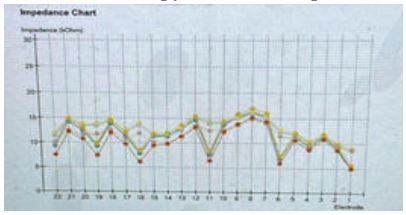
After detailed counselling regarding prognosis and available management options, the child was planned for cochlear implantation in the left ear. Under general anaesthesia, the child was positioned supine with the head turned to the opposite side. A postauricular incision was made, and a musculoperiosteal flap was elevated. Standard mastoidectomy was performed, followed by a facial recess posterior tympanotomy approach to access the middle ear and round window niche, with careful preservation of the facial nerve using intra-operative facial nerve monitoring was done. The cochlea was targeted with the aim of entering the scala tympani. After gentle exposure and opening of the endosteum, the electrode array was slowly inserted into the scala tympani to achieve full insertion. A bony well was drilled in the squamous temporal bone to accommodate the receiver–stimulator, with meticulous handling of the dura. The receiver– stimulator was secured in the implant well, and the ground electrode was placed beneath the temporalis muscle. Wound closure was performed in layers after achieving hemostasis, and a pressure dressing was applied.



Surgical image showing cochlear electrode being implanted

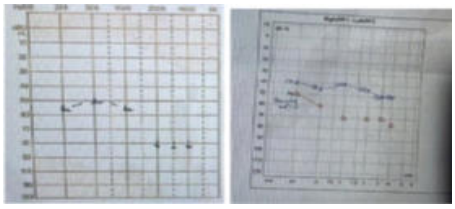


Intra operative 3D fluoroscopy of cochlear implant



Impedance History Report

A Cochlear™ Nucleus® 7 (CP1002) implant was used for the left ear which was confirmed intra operatively using C arm machine, impedance telemetry testing and nerve response telemetry (NRT) all of which suggested normal Cochlear implant placement and functionality. The postoperative period was uneventful, and facial nerve function remained intact. Switch-on and auditory rehabilitation were initiated after adequate wound healing. At three months post-implantation, auditory performance assessment revealed a Category of Auditory Performance (CAP) score of 4, indicating understanding of some spoken words with additional visual cues. The Meaningful Auditory Integration Scale (MAIS) score was 34 out of 40, and the music perception score was 9 out of 72.



Pre and post-operative audiologram

Overall, the child showed good adaptation to the implant and active participation in auditory–verbal therapy sessions. The parents were advised to continue intensive speech and language stimulation at home, ensure regular follow-up, and initiate appropriate schooling to maximize auditory and language outcomes.

CONCLUSION

This case demonstrates that cochlear implantation can yield meaningful auditory outcomes in children with bilateral profound hearing loss and cochlear nerve deficiency . Careful preoperative evaluation and appropriate ear selection according to audiological assessment and patient's clinical hearing status becomes crucial. Detailed radiological assessment and structured postoperative rehabilitation were key to achieving satisfactory auditory performance, highlighting the importance of individualized management in anatomically complex cases.

REFERENCES

1. Nair G, Vadivu S, Sampathkumar R, Pradhananga RB, Kameswaran M. A study of anomalies of cochlea and cochlear nerve in children with congenital profound hearing loss–an Indian perspective. J Sci Res Rep. 2017;13:1–11.
2. Abdullah AB, Zaki FB, Rosla LB. Congenital sensorineural hearing loss with anomalous vestibular nerve complex: a rare radiological finding. Indian J Otol.

2018;24(1):71–73.
3. Agarwal SK, Singh S, Ghuman SS, Sharma S, Lahiri AK. Radiological assessment of the Indian children with congenital sensorineural hearing loss. Int J Otolaryngol. 2014;2014:808759.
4. Huy PT. Efficacy of cochlear implantation in cochlear nerve deficiency children–A single center study. American Journal of Otolaryngology. 2024 Nov 1;45(6):104428.
5. Swain SK. Cochlear deformities and its implication in cochlear implantation: a review. Int J Res Med Sci. 2022;10(10):2339–2344.
6. Arumugam SV, Nair G, Paramasivan VK, Goyal S, Murali S, Kameswaran M. A study of outcome of pediatric cochlear implantation in patients with cochleovestibular nerve deficiency. J Int Adv Otol. 2020;16(2):147–152.
7. Hiremath SB, Biswas A, Mndebele G, Schramm D, Ertl-Wagner BB, Blaser SI, Chakraborty S. Cochlear implantation: systematic approach to preoperative radiologic evaluation. Radiographics. 2023;43(4):e220102.