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# Major Internal Auditory Canal Anomalies Candidate for Cochlear Implantation: A Case Report

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**Abstract** | **Case Report**

Congenital profound deafness is often linked to significant anomalies of the inner ear and auditory nerve pathways, necessitating detailed imaging prior to cochlear implantation. We present the case of an 8-year-old female patient with bilateral severe hearing loss who underwent magnetic resonance imaging of the brain and temporal bones as part of the preoperative workup. Imaging revealed marked bilateral stenosis of the internal auditory canals with absence of identifiable cochleovestibular nerve fibers, aberrant courses of the facial and vestibular nerves, and enlargement of the vestibular structures, notably the vestibules and lateral semicircular canals, more evident on the right side. No abnormalities were detected in the brain parenchyma, midline, or brainstem. This report highlights the complex anatomical variations encountered in such cases and discusses their critical impact on surgical decision-making and cochlear implant feasibility.

**Keywords:** Internal Auditory Canal Anomalies, Congenital Profound Deafness, Cochlear Implantation, Auditory Nerve Malformations, Temporal Bone MRI, Inner Ear Malformations.



## INTRODUCTION

Profound congenital deafness represents a significant clinical challenge, often resulting from a spectrum of inner ear and auditory nerve abnormalities. Accurate identification of these malformations is essential before cochlear implantation, as anatomical variations can considerably influence surgical feasibility and outcomes. Imaging techniques, particularly magnetic resonance imaging (MRI) of the temporal bones and brain, play a pivotal role in the preoperative evaluation by providing detailed visualization of the auditory pathways and surrounding structures.

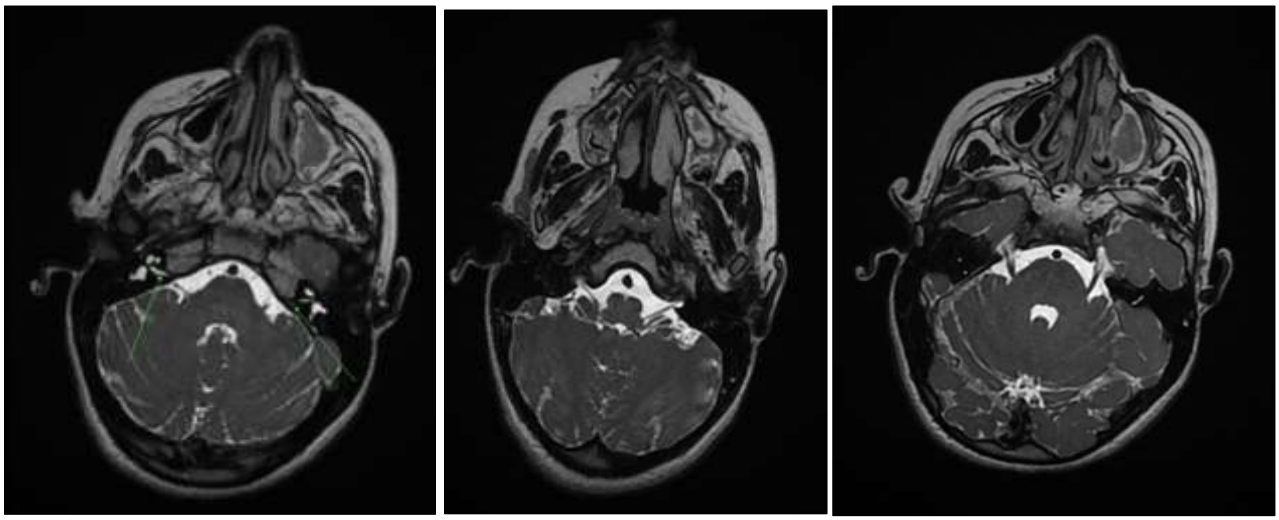
In pediatric patients, such as the case presented here, the coexistence of severe bony labyrinthine anomalies and nerve abnormalities further complicates implant planning. Understanding these variations is crucial for tailoring surgical approaches and improving prognostic predictions. This report aims to describe a rare combination of internal auditory canal stenosis, cochleovestibular nerve agenesis, and labyrinthine dilatation in an 8-year-old child, highlighting the diagnostic and therapeutic challenges faced in cochlear implantation candidates.

## OBSERVATION

An 8-year-old girl with bilateral profound sensorineural hearing loss was referred for evaluation as a candidate for cochlear implantation. The patient's medical history was unremarkable, and no syndromic features were identified. Due to the severity of hearing impairment, a detailed imaging workup was performed.

Magnetic resonance imaging (MRI) of the brain and temporal bones revealed significant bilateral narrowing of the internal auditory canals. The cochleovestibular nerve bundles were not visualized on either side, indicating probable nerve aplasia or severe hypoplasia. Additionally, the facial and vestibular nerves exhibited ectopic origins with abnormal courses. The vestibular labyrinth demonstrated notable dilatation, particularly affecting the vestibules and lateral semicircular canals, with greater prominence on the right. The brain parenchyma, brainstem, and midline structures appeared normal, with no evidence of additional intracranial abnormalities.

These anatomical findings posed considerable challenges to cochlear implantation candidacy, necessitating multidisciplinary discussion regarding the feasibility and potential outcomes of the procedure.



- Important rétrécissement bilatéral des conduits auditifs internes avec absence de visualisation des paquets acoustico-faciaux au niveau de leur trajet cisternal.
- Naissance et trajet ectopique des nerfs faciaux et vestibulaires.
- Dilatation vestibulaire bilatérale.
- Dilatation bilatérale des canaux semi circulaires latéraux qui sont raccourci et d'aspect vésiculaire du côté droit.

## DISCUSSION

Profound congenital deafness frequently arises from a wide range of malformations affecting the inner ear and auditory nerve structures. These anomalies can significantly influence the candidacy for cochlear implantation, which relies on the presence of a functional cochleovestibular nerve and a patent internal auditory canal to ensure effective electrical stimulation and auditory rehabilitation.

In this case, the imaging findings of bilateral internal auditory canal stenosis combined with absence of visible cochleovestibular nerves represent a severe form of inner ear malformation. The ectopic positioning of the facial and vestibular nerves further complicates the anatomical landscape. Such nerve anomalies reduce the likelihood of successful implant function and increase the risks associated with surgery.

Additionally, the dilatation of the vestibular labyrinth, including the vestibules and lateral semicircular canals, suggests abnormal inner ear development and may reflect compensatory or secondary changes related to nerve absence. The normal appearance of central nervous system structures indicates that the anomalies are confined to the peripheral auditory apparatus.

The preoperative imaging assessment is thus indispensable not only for diagnosis but also for surgical planning and prognostication. It informs the multidisciplinary team about the complexity of the anatomy, guides patient and family counseling, and may influence the choice of alternative auditory rehabilitation strategies, such as auditory brainstem implants, when cochlear implantation is contraindicated.

This case highlights the critical role of high-resolution MRI in revealing complex inner ear and nerve abnormalities in pediatric patients with profound deafness and underscores the importance of individualized assessment in optimizing therapeutic approaches.

## CONCLUSION

Accurate imaging is vital to detect complex inner ear anomalies in children with profound deafness. Severe nerve and canal malformations may limit cochlear implant options, highlighting the need for tailored surgical planning and alternative therapies.

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