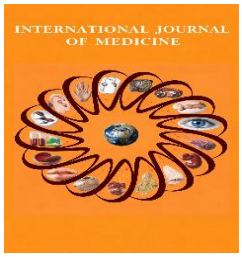

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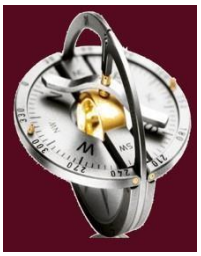

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Original Research

Evaluation of Prelingually Deaf Children by CT and MRI Temporal Bone for Deciding Cochlear Implant Candidacy

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Abstract

Background: Prelingual deafness affects 1.2 to 5.7 per 1000 live births in India and represents a significant cause of sensory impairment requiring early intervention. High resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) play complementary roles in evaluating temporal bone anatomy and determining cochlear implant candidacy by assessing bony and membranous labyrinth structures, cochlear nerve status, and identifying contraindications. ***Methods:*** A retrospective study was conducted from January 2023 to June 2024 at District McGann Hospital, Karnataka, evaluating 32 prelingually deaf children referred for cochlear implantation under the government cochlear implant scheme. All patients underwent HRCT temporal bone using a 128-slice CT scanner and MRI cochleogram with brain imaging using a 1.5 Tesla machine. Standardized imaging & reporting protocols were followed with detailed evaluation of bony labyrinth, membr anous labyrinth, internal auditory canal, cochlear nerve, and anatomical variants. ***Results***: Among 32 patients (19 males, 13 females; age range 2-6 years), normal imaging findings were observed in 25 patients (78.1%). Inner ear malformations were identified in 4 patients (12.5%), including bilateral cochlear hypoplasia, bilateral cochlear aplasia, Mondini malformation, and bilateral enlarged vestibular aqueduct. Isolated white matter abnormalities suggestive of cytomegalovirus infection or meningitis were found in 3 patients (9.4%). Additional pathologies included acute and chronic otomastoiditis in 5 patients (15.6%). Anatomical variants such as deep sigmoid groove, high jugular bulb, and low-lying tegmen were noted in 3 patients (9.4%). ***Conclusion***: Comprehensive imaging evaluation using HRCT and MRI is essential for determining cochlear implant candidacy, identifying anatomical variants, detecting inner ear malformations, and providing crucial preoperative information to surgeons for optimal surgical planning.

Keywords: Prelingual Deafness, Cochlear Implant, Temporal Bone Imaging, High-Resolution CT, MRI Cochleogram.

Introduction

Prelingual deafness refers to severe to profound hearing loss that occurs before the acquisition of speech and language skills, typically before two years of age. This condition represents one of the most common sensory deficits in childhood, with an estimated incidence ranging from 1.2 to 5.7 per 1000 live births in India.[1] The critical period for language development occurs within the first three years of life, making early identification and intervention paramount for optimal neurodevelopmental outcomes. Children with prelingual deafness face signi ficant challenges in speech development, educational achievement, and social integration if left untreated.[2]

The etiology of prelingual deafness is multifactorial, encompassing genetic factors, congenital infections, birth complications, ototoxic medications, and anatomical malformations of the inner ear. Approximately 50-60% of prelingual deafness cases have a genetic basis, while the remaining cases result from acquired causes such as cytomegalovirus infection, meningitis, hypoxic-ischemic encephalopathy, and kernicterus. The majority of prelingual deafness involves sensorineural hearing loss, which results from dysfunction of the cochlea or auditory nerve, rendering conventional hearing aids ineffective in severe to profound cases.[3]

Cochlear implantation has emerged as the gold standard treatment for children with severe to profound sensorineural hearing loss who do not benefit from conventional amplification. A cochlear implant is an electronic medical device that bypasses damaged hair cells in the c ochlea and directly stimulates the auditory nerve, enabling sound perception. The success of cochlear implantation depends critically on the integrity of the auditory pathway, particularly the presence and functionality of the cochlear nerve and spiral ganglion cells. Early implantation, ideally before 18 months of age, maximizes auditory cortex plasticity and facilitates normal speech and language development.[4].

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The preoperative radiological evaluation of children being considered for cochlear implantation is essential and serves multiple purposes. First, imaging helps identify anatomical variants and inner ear malformations that may contraindicate surgery or necessitate modified surgical

approaches. Second, it provides crucial information about cochlear patency, round window accessibility, facial nerve course, and the relationship of vital structures such as the sigmoid sinus and jugular bulb to the surgical field. Third, imaging assesses the status of the cochlear nerve, which is the most critical factor determining implant success. Finally, radiological evaluation helps identify treatable causes of hearing loss and detects associated intracranial abnormalities.[5]

HRCT of the temporal bone provides excellent visualization of the bony labyrinth, including the cochlea, vestibule, semicircular canals, internal auditory canal, and ossicular chain. HRCT is superior for evaluating bony anatomy, detecting inner ear malformations, assessing cochlear dimensions and patency, identifying labyrinthitis ossificans, and detecting anatomical variants that may complicate surgery.[6]
MRI complements CT by providing superior soft tissue resolution and is essential for evaluating the membranous labyrinth, cochlear nerve, brain parenchyma, and detecting fluid signal abnormalities within the inner ear structures. MRI is also valuable for detecting white matter abnormalities associated with congenital infections and identifying associated central nervous system malformations.[7]

The classification of inner ear malformations has evolved significantly with advances in imaging technology and genetic understanding. The Sennaroglu classification system categorizes inner ear malformations based on the embryological timing of developmental arrest. Complete labyrinthine aplasia represents the most severe form, resulting from arrest at the third week of gestation. Common cavity malformation occurs from arrest at the fourth week, while cochlear aplasia and hypoplasia result from developmental disruption at 4-5 weeks. Incomplete partition types I, II (classic Mondini malformation), and III occur from disruptions at 5-7 weeks of gestation. Enlarged vestibular aqueduct syndrome, one of the most common inner ear malformations associated with sensorineural hearing loss, results from developmental abnormalities affecting the endolymphatic duct and sac.[8]

The successful implementation of cochlear implant programs requires a multidisciplinary approach involving audiologists, otolaryngologists, radiologists, speech therapists, and psychologists. Radiologists play a pivotal role in this team by providing accurate anatomical information that guides patient selection, surgical planning, and prognostic counseling. Understanding the normal anatomy, common variants, and pathological findings on temporal bone imaging is essential for radiologists reporting these studies. Standardized reporting templates and systematic evaluation protocols ensure comprehensive assessment and effective communication of relevant findings to the surgical team.[9]
In the Indian context, government-sponsored cochlear implant schemes have significantly expanded access to this life-transforming technology for economically disadvantaged families. The Karnataka government's cochlear implant scheme, under which this study was conducted, represents an important public health initiative addressing childhood hearing impairment. Such programs underscore the importance of establishing efficient diagnostic pathways and ensuring quality radiological evaluation to optimize patient outcomes and resource utilization.[10]
This retrospective study aimed to systematically evaluate the imaging findings in prelingually deaf children referred for severe SNHL, document the spectrum of inner ear malformations and anatomical variants encountered, and assess the role of HRCT and MRI in determining implant candidacy and surgical planning.

AIMS AND OBJECTIVES

The primary aim of this study was to evaluate the intricate anatomy of the inner ear structures in prelingually deaf children using standardized HRCT and MRI.

The study further aimed to evaluate the membranous labyrinth and cochlear nerve status using MRI sequences including T2-weighted sagittal oblique images to assess the four-nerve configuration in the distal internal auditory canal and axial T2-DRIVE sequences to demonstrate cochlear nerve presence and caliber. Brain screening using diffusion-weighted imaging and FLAIR sequences was performed to detect white matter abnormalities and associated central nervous system pathology that could impact implant outcomes.

An important objective was to determine the etiology of congenital deafness by identifying inner ear malformations, evidence of previous infections, and anatomical abnormalities that could explain the hearing loss. The study aimed to document the spectrum and frequency of inner ear malformations encountered in the study population and compare these findings with published literature from similar populations.

The research sought to identify absolute and relative contraindications to cochlear implantation based on imaging findings, thereby assisting the multidisciplinary team in patient selection and counselling.

Materials and Methods

Study Design and Setting

This retrospective observational study was conducted at the Department of Radiodiagnosis, District McGann Hospital, Karnataka, India, over an 18-month period from January 2023 to June 2024. District McGann Hospital is an empanelled center under the Government of Karnataka Cochlear Implant Scheme, which provides comprehensive evaluation and cochlear implantatio n services to economically disadvantaged children with severe to profound sensorineural hearing loss. The study was conducted in accordance with institutional ethical guidelines and principles of the Declaration of Helsinki.

Study Population and Sample Size

The study included 32 prelingually deaf children who were referred to the Department of Radiodiagnosis for preoperative imagi ng evaluation as part of the cochlear implant candidacy assessment protocol. The sample consisted of all consecutive patients meeting the inclusion criteria during the study period, representing a convenience sampling method. The demographic characteristics of the study population were systematically recorded including age, gender, laterality of hearing loss, family history of deafness, history of consanguinity, antenatal and perinatal complications, neonatal intensive care unit admission, and history of ototoxic drug exposure or meningitis.

Inclusion Criteria

Children were included in the study if they were diagnosed with prelingual sensorineural hearing loss defined as severe to profound hearing impairment occurring before two years of age prior to speech and language acquisition. All included patients had undergone comprehensive audiological evaluation including otoacoustic emissions testing, which showed absent responses bilaterally indicating cochlear dysfunction. Brainstem evoked response audiometry demonstrated absent or severely abnormal waveforms with thresholds exceeding 90 decibels indicating profound hearing loss. All patients had completed a mandatory three-month trial with appropriately fitted hearing aids without significant benefit as determined by audiological and speech therapy assessment. Only patients referred by the otolaryngology department with confirmed sensorineural hearing loss and deemed potential candidates for cochlear implantation based on clinical evaluation were included in the study.

Exclusion Criteria

Children with postlingual deafness defined as hearing loss occurring after speech and language development were excluded from the study as these patients represent a different clinical entity with distinct prognostic implications. Patients with pure conductive hearing loss as determined by audiological testing were excluded since conductive hearing loss typically responds to medical management or middle ear surgery rather than cochlear implantation. Children with severe cognitive impairment or developmental delay that would preclude benefit from cochlear implantation were excluded. Patients with active middle ear infections or mastoiditis requiring medical treatment were temporarily excluded until complete resolution of infection was documented.

Imaging Protocol

All patients underwent both MRI of the inner ear and brain and HRCT of the temporal bone as per the institutional protocol for cochlear implant evaluation.

The feed-and-sleep technique was used for neonates and infants.The procedure was performed under short IV sedation or general anesthesia for toddlers and young children with help of anesthesiologists.MR compatible pulseoximeter & workstation was kept available during the procedure for emergency situations.

MRI was performed first using a 1.5 Tesla MRI scanner with a dedicated head coil. The imaging protocol included diffusion-weighted imaging sequence for detection of cholesteatoma, acute ischemic lesions and abscesses. FLAIR sequences in the axial plane were acquired for optimal detection of white matter abnormalities and periventricular lesions. High-resolution T2-weighted fast spin echo sequences in axial and coronal planes provided anatomical detail of brain structures. Three-dimensional heavily T2-weighted DRIVE sequences were acquired in the axial plane for detailed visualization of the membranous labyrinth, cranial nerves in the internal auditory canal, and cerebellopontine angle cisterns. Oblique sagittal T2-weighted images perpendicular to the long axis of the internal auditory canal were obtained on both sides to assess the four-nerve bundle configuration including the facial nerve, cochlear nerve, superior vestibular nerve, and inferior vestibular nerve in the lateral internal auditory canal.

HRCT was performed next using a 128-slice multidetector CT scanner with patients positioned supine with head extended and immobilized to minimize motion artifact. Axial images were acquired from the external auditory canal to the petrous apex using submillimeter collimation (0.625 mm) with the following technical parameters: tube voltage 120 kilovolt peak, tube current 200-250 milliampere seconds adjusted based on patient size, rotation time 0.5 seconds, and pitch factor 0.8. Images were reconstructed using a bone algorithm with slice thickness of 0.625 mm and increment of 0.3 mm to provide overlapping slices for high-quality multiplanar reconstructions. Coronal and sagittal reformations were generated perpendicular and parallel to the long axis of the temporal bone respectively using a dedicated workstation. All images were reviewed using bone window settings optimized for temporal bone evaluation.

Image Analysis and Measurements

All imaging studies were systematically reviewed by two radiologists with five and ten years of experience in temporal bone imaging respectively. Disagreements were resolved by consensus after joint review. A standardized reporting format was used to ensure comprehensive evaluation of all relevant anatomical structures and uniform documentation of findings. For HRCT evaluation, mastoid pneumatization was assessed and classified as well-pneumatized, partially pneumatized, or sclerotic based on the volume of air cells and degree of bony sclerosis. Middle ear cavity was examined for presence of fluid, soft tissue opacification, or ossicular abnormalities. Round window orientation was documented as normal, obliterated, or abnormally angled, as this information is critical for electrode insertion.

Specific cochlear measurements were performed on axial HRCT images perpendicular to the cochlear axis. Cochlear height was measured as the maximum vertical dimension from the base to the apex of the cochlea in the plane perpendicular to the modiolus. Cochlear base width designated as parameter A was measured as the maximum horizontal dimension of the basal turn of the cochlea in the same plane. Cochlear duct length was calculated using the validated formula: cochlear duct length equals 4.16 multiplied by cochlear base width minus 2.7, expressed in millimeters. This calculated parameter helps in selection of appropriate electrode length. The number of cochlear turns was counted, with normal cochlea displaying 2.5 to 2.75 turns. Modiolus, interscalar septum of cochlea, cochlear aqueduct and vestibule, vestibular aqueduct were evaluated for the width.

The internal auditory canal was evaluated in both axial and coronal planes. The width of the internal auditory canal was measured at mid canal. Normal internal auditory canal diameter ranges from 3 to 8 mm, with diameters less than 2 mm considered stenotic and greater than 10 mm considered dilated. The cochlear aperture, which is the bony opening through which the cochlear nerve enters the base of the cochlea, was identified at the anteroinferior aspect of the fundus of the internal auditory canal and its size was assessed. Distance from facial nerve recess to annulus & distance from vertical facial nerve to mastoid cortex were measured for surgical guidance.

Anatomical variants that could influence surgical approach or increase operative risk were systematically documented. Tegmen height was measured as the vertical distance from the superior aspect of the external auditory canal to the floor of the middle cranial fossa. Low-lying tegmen (less than 10 mm) increases risk of dural exposure or injury during mastoidectomy. The sigmoid sinus was evaluated for position and configuration. Types of sigmoid sinusweredetermined based on the extension of the sigmoid plate beyond the axis of the posterior SCC, the tympanic segment of the facial nerve and the malleal-incudal axis. Jugular bulb position was assessed in relation to the floor of the middle ear cavity.

MRI evaluation focused primarily on assessment of the cochlear nerve and detection of associated brain abnormalities like white matter hyperintensities, calcifications, microcephaly or posterior fossa cysts. The cochlear nerve was visualized on oblique sagittal T2-weighted images through the internal auditory canal and on axial three-dimensional T2-DRIVE sequences. Normal cochlear nerve appeared as a distinct hypointense linear structure in the anteroinferior quadrant of the lateral internal auditory canal, typically measuring 1 to 2 mm in diameter and equal in caliber to the facial nerve. Cochlear nerve hypoplasia was diagnosed when the nerve diameter was less than 50% of the ipsilateral facial nerve diameter. Cochlear nerve aplasia was diagnosed when no distinct cochlear nerve could be identified despite optimal imaging technique.

Systematic documentation of the findings and all measurements were done on films and given to the patient for immediate reference for the clinician. Digital images through DVD were also given to the patients.

Statistical Analysis

Patient demographic data, clinical history, audiological test results, and imaging findings were systematically recorded in a structured database using Microsoft Excel. Categorical variables including gender distribution, presence or absence of inner ear malformations, normal versus abnormal imaging findings, anatomical variants, and mastoid pathology were expressed as frequencies and percentages. Continuous variables including age, cochlear dimensions, internal auditory canal width, and other measured parameters were expressed as mean ± standard deviation with range. The chi-square test was used to analyze associations between categorical variables such as gender and presence of inner ear malformations. Independent samples t-test was used to compare continuous variables between groups. A p-value of less than 0.05 was considered statistically significant. Statistical analysis was performed using SPSS software version 23.0.

Results

Demographic Characteristics

A total of 32 prelingually deaf children who underwent comprehensive imaging evaluation for cochlear implant candidacy were included in this retrospective analysis. The study cohort comprised 19 males (59.4%) and 13 females (40.6%), yielding a male-to-female ratio of 1.46:1. The age distribution of patients ranged from 2 years to 6 years with a mean age of 3.4 ± 1.2 years. The majority of patients were in the 3-year age group (n=17, 53.1%), followed by the 4-year age group (n=8, 25.0%). Two patients (6.3%) were 2 years old, four patients (12.5%) were 5 years old, and only one patient (3.1%) was 6 years old at the time of evaluation. Bilateral severe to profound sensorineural hearing loss was documented in all 32 patients (100%) based on comprehensive audiological evaluation including otoacoustic emissions testing showing absent responses bilaterally and brainstem evoked response audiometry demonstrating thresholds exceeding 90 decibels in both ears. All patients had undergone a three-month trial with appropriately fitted bilateral hearing aids without significant auditory or speech development benefit as assessed by audiologists and speech therapists.

Etiological Classification Based on Imaging Findings

The imaging evaluation revealed diverse etiological patterns among the study population. Normal imaging findings on both HRCT and MRI were observed in 25 patients (78.1%), indicating no detectable structural abnormality of the temporal bone, inner ear structures, cochlear nerve, or brain parenchyma that would explain the hearing loss. These cases were presumed to represent genetic or idiopathic sensorineural hearing loss and were declared eligible for cochlear implantation based on normal anatomical prerequisites.

Inner ear malformations were identified in 4 patients (12.5%) representing significant structural developmental abnormalities that influenced cochlear implant candidacy and surgical approach. The specific malformations included one patient with bilateral cochlear hypoplasia characterized by reduced cochlear height (2.1 mm on the right and 2.3 mm on the left compared to normal range of 3.5 -5.0 mm) and reduced number of cochlear turns (1.25 turns bilaterally instead of normal 2.5-2.75 turns) associated with bilateral cochlear nerve hypoplasia with cochlear nerve diameter measuring less than 50% of the facial nerve diameter on both sides. This patient was deemed a poor candidate for standard cochlear implantation due to the combined cochlear and nerve hypoplasia and was declared ineligible.

A second patient demonstrated bilateral cochlear aplasia with complete absence of cochlear structures bilaterally, associated with hypoplastic internal auditory canals measuring 1.8 mm on the right and 2.1 mm on the left (normal range 3-8 mm), bilateral cochlear nerve aplasia with no identifiable cochlear nerve on high-resolution MRI sequences, and hypoplastic vestibules measuring 2.5 mm in maximum dimension compared to normal range of 4-5 mm. This patient was declared ineligible for cochlear implantation due to absence of cochlea and cochlear nerve.

The third patient with inner ear malformation presented with bilateral Mondini malformation characterized by normal basal tur n but confluent cystic middle and apical turns, enlarged vestibular aqueducts measuring 2.8 mm on the right and 3.1 mm on the left at the midpoint (normal less than 1.5 mm), and preserved bilateral cochlear nerves of normal caliber. This patient was considered a suitable candidat e for cochlear implantation with anticipated good outcomes given the presence of one normal cochlear turn and intact cochlear nerves bilaterally.

The fourth patient demonstrated bilateral enlarged vestibule, vestibular aqueduct & endolymphatic sac as an isolated finding. The vestibules appeared mildly enlarged measuring 5.3 mm on the right and 5.2 mm on the left (normal 4-5 mm). The vestibular aqueduct measuring 4.4 mm on the right and 2.0 mm on the left at the midpoint (normal less than 1.5 mm). Dilated endolymphatic sac noted measuring 8.5mm on right and 4.6mm on left in transverse diameter. The cochlear morphology was otherwise normal. This patient was declared eligible for cochlear implantation with counselling regarding the underlying anatomical variant and potential for progressive hearing loss.

Isolated brain white matter abnormalities without inner ear structural malformations or cochlear nerve abnormalities were detected in 3 patients (9.4%). These patients demonstrated scattered hyperintense foci in the periventricular white matter, subcortical white matter, and centrum semiovale on FLAIR, suggesting previous insults such as congenital cytomegalovirus infection or neonatal meningitis. The pattern of white matter lesions was characterized by bilateral symmetric distribution predominantly in the periventricular regions with sparing of the corpus callosum and basal ganglia. Two of these patients had documented history of neonatal intensive care unit admission for prematurity-related complications, while one patient had maternal history suggestive of first -trimester infection. All three patients with brain white matter abnormalities had normal inner ear anatomy, normal cochlear nerve size bilaterally, and were declared eligible for cochlear implantation with counselling provided to families regarding potential additional developmental challenges beyond hearing loss.

Additional Pathological Findings

Mastoid and middle ear pathology that could influence surgical planning and timing was identified in 5 patients (15.6%). Three patients demonstrated acute otomastoiditis characterized by complete or partial opacification of mastoid air cells with fluid signal on MRI and soft tissue attenuation replacing normal pneumatization on HRCT, associated with middle ear effusion and thickened inflamed mucosa. These patients were treated with appropriate antibiotic therapy for 4-6 weeks followed by repeat HRCT to document resolution of acute infection before proceeding with cochlear implantation. Two patients showed chronic otomastoiditis with sclerotic mastoids characterized by marked reduction in pneumatization, and chronic inflammatory changes in the middle ear cavity with mucosal thickening. In these two cases, the contralateral ear with better pneumatization was selected for cochlear implant electrode placement to facilitate easier surgical access and reduce operative time and complications.

Anatomical Variants

Anatomical variants of the temporal bone that could impact surgical approach or increase operative risk were documented in 3 patients (9.4%). One patient demonstrated bilateral high jugular bulbs extending above the level of the inferior annulus of the tympanic membrane and projecting into the hypotympanum, which potentially limited surgical access to the round window niche. The jugular bulb heigh t measured 3.2 mm above the inferior tympanic annulus on the right side and 2.8 mm on the left side. The left ear with slightly lower jugular bulb position was selected for implantation with intraoperative precautions to avoid injury to the jugular bulb during round window approach.

One patient showed bilateral Type IV deep sigmoid grooves with marked anterior extension of the sigmoid sinus into the mastoid cavity, significantly limiting the available working space for posterior tympanotomy approach. The distance from the sigmoid sinus to the posterior external auditory canal measured only 8 mm on the right side and 10 mm on the left side (normal range 12-18 mm), reducing the safe surgical corridor for mastoidectomy and facial recess approach. The left ear was selected for cochlear implantation with anticipated modified surgical approach and careful dissection to avoid sigmoid sinus injury.

One patient demonstrated bilateral low-lying tegmen with tegmen height measuring 7 mm on the right and 8 mm on the left (normal range greater than 10 mm), indicating reduced vertical dimension between the external auditory canal and the floor of the middle cr anial fossa. This anatomical variant increased the risk of inadvertent dural exposure during cortical mastoidectomy. The surgical team was specifically informed about this finding to ensure careful dissection of the tegmen with avoidance of excessive superior extension during mastoidectomy. The left side with slightly greater tegmen height was selected for implantation.

Cochlear Measurements

Among the 28 patients with normal cochlear anatomy (excluding the 4 patients with inner ear malformations), detailed morphometric analysis was performed. The mean cochlear height was 4.2 ± 0.4 mm (range 3.5-4.9 mm) on the right side and 4.3 ± 0.4 mm (range 3.6-5.0 mm) on the left side. The mean cochlear base width measured 8.8 ± 0.6 mm (range 7.8 -9.9 mm) on the right and 8.9 ± 0.5 mm (range 7.9-9.8 mm) on the left. Using the formula for calculated cochlear duct length (cochlear duct length = 4.16 × cochlear base width - 2.7), the mean calculated cochlear duct length was 33.9 ± 2.5 mm (range 29.7-38.5 mm) on the right and 34.3 ± 2.1 mm (range 30.2-38.1 mm) on the left. These values were within normal ranges reported in published literature and indicated adequate cochlear dimensions for standard electrode insertion. The mean internal auditory canal width measured 4.8 ± 0.7 mm (range 3.5 -6.2 mm) on the right and 4.9 ± 0.6 mm (range 3.6-6.4 mm) on the left, confirming normal internal auditory canal dimensions in patients with normal anatomy. There was no statistically significant difference in cochlear measurements between right and left sides ($p > 0.05$ for all parameters).

Cochlear Implant Candidacy Determination

Based on comprehensive imaging evaluation, 30 patients (90.6%) were declared eligible for cochlear implantation. This group included 25 patients with completely normal imaging findings, 3 patients with brain white matter abnormalities but normal temporal bone anatomy and cochlear nerve, 1 patient with bilateral enlarged vestibular aqueduct with otherwise normal cochlear anatomy and preserved co chlear nerves, and 1 patient with bilateral Mondini malformation and preserved cochlear nerves. Both Mondini malformation and enlarged vestibular aqueduct are considered compatible with cochlear implantation with appropriate electrode selection and surgical modifications.

Three patients (9.4%) were declared ineligible for standard cochlear implantation. These included one patient with bilateral cochlear hypoplasia and cochlear nerve hypoplasia who was counselled about poor expected outcomes and another patient with complete bilateral coc hlear aplasia and cochlear nerve aplasia representing absolute contraindications. Theses represented absolute or relative contraindications to cochlear implantation as there is no functional auditory nerve pathway to transmit electrical signals from the implant to the brainstem. These two patients were referred for consideration of auditory brainstem implantation or alternative rehabilitation strategies.

Statistical Analysis of Key Variables

Statistical analysis revealed no significant association between gender and presence of inner ear malformations ($\chi^2 = 0.29$, $p = 0.59$). Among male patients, 2 out of 19 (10.5%) had inner ear malformations compared to 2 out of 13 females (15.4%). Similarly, there was no significant association between age group and presence of anatomical abnormalities ($\chi^2 = 2.18$, $p = 0.70$). The presence of brain white matter abnormalities showed no significant correlation with history of neonatal intensive care unit admission, although the small sample size limited statistical power for this analysis (Fisher's exact test, $p = 0.08$). Comparison of cochlear dimensions between male and female patients revealed no statistically significant differences in mean cochlear height (males 4.2 ± 0.4 mm vs. females 4.3 ± 0.3 mm, $p = 0.43$), cochlear base width (males 8.8 ± 0.5 mm vs. females 8.9 ± 0.6 mm, $p = 0.62$), or calculated cochlear duct length (males 34.0 ± 2.3 mm vs. females 34.2 ± 2.4 mm, $p = 0.78$).

Parameter	Category	Number	Percentage
Gender	Male	19	59.4%
	Female	13	40.6%
Age (years)	2	2	6.3%
	3	17	53.1%
	4	8	25.0%
	5	4	12.5%
	6	1	3.1%
Laterality	Bilateral SNHL	32	100%
Hearing Aid Trial	Failed 3-month trial	32	100%

Table 1: Demographic Distribution of Study Population (N=32)

Category	Subcategory	Number	Percentage
Normal Imaging	No structural abnormality detected	25	78.1%
Inner Ear Malformations	Bilateral cochlear hypoplasia with CN hypoplasia	1	3.1%
	Bilateral cochlear aplasia with CN aplasia	1	3.1%
	Bilateral Mondini malformation	1	3.1%
	Bilateral enlarged vestibular aqueduct	1	3.1%
	Total IEM	**4**	**12.5%**
Brain Abnormalities	White matter lesions (CMV/meningitis)	3	9.3%
Total with Etiology Identified		**7**	**21.9%**

Table 2: Etiological Classification Based on Imaging Findings (N=32)

CN = Cochlear Nerve; IEM = Inner Ear Malformation; CMV = Cytomegalovirus

Finding	Number	Percentage	Clinical Significance
Acute Otomastoiditis	3	9.4%	Required antibiotic treatment before CI
Chronic Otomastoiditis/Sclerotic Mastoids	2	6.3%	Contralateral ear selected for CI
High Jugular Bulb	1	3.1%	Limited RW access, modified approach
Type IV Deep Sigmoid Groove	1	3.1%	Limited surgical corridor, careful dissection
Low-lying Tegmen	1	3.1%	Risk of dural exposure, cautious mastoidectomy
Total Anatomical Variants	**3**	**9.4%**	Influenced surgical planning
Total Mastoid Pathology	**5**	**15.6%**	Affected timing or side selection

Table 3: Additional Pathological Findings and Anatomical Variants (N=32)

CI = Cochlear Implant; RW = Round Window

Parameter	Right Ear (Mean ± SD)	Range (Right)	Left Ear (Mean ± SD)	Range (Left)	p-value
Cochlear Height (mm)	4.2 ± 0.4	3.5-4.9	4.3 ± 0.4	3.6-5.0	0.38
Cochlear Base Width (mm)	8.8 ± 0.6	7.8-9.9	8.9 ± 0.5	7.9-9.8	0.52
Calculated CDL (mm)	33.9 ± 2.5	29.7-38.5	34.3 ± 2.1	30.2-38.1	0.46
IAC Width (mm)	4.8 ± 0.7	3.5-6.2	4.9 ± 0.6	3.6-6.4	0.58
Number of Cochlear Turns	2.6 ± 0.2	2.5-2.75	2.6 ± 0.2	2.5-2.75	1.00
Facial Recess Width (mm)	3.4 ± 0.6	2.5-4.8	3.5 ± 0.5	2.6-4.6	0.62

Table 4: Cochlear Morphometric Measurements in Patients with Normal Anatomy (N=28)

CDL = Cochlear Duct Length; IAC = Internal Auditory Canal; SD = Standard Deviation

Cochlear Nerve Status	Right Ear	Left Ear	Bilateral	Total Patients	Percentage
Normal CN	30	30	30	30	93.7%
CN Hypoplasia	1	1	1	1	3.1%
CN Aplasia	1	1	1	1	3.1%
Four-nerve bundle clearly visualized	30	30	30	30	93.7%
Abnormal IAC configuration	2	2	2	2	6.1%

Table 5: Cochlear Nerve Assessment on MRI (N=32)

CN = Cochlear Nerve; IAC = Internal Auditory Canal

Category	Subcategory	Number	Percentage	Rationale
Eligible for CI	Normal imaging	25	78.1%	Normal anatomy and CN
	Brain WM abnormalities only	3	9.3%	Normal temporal bone and CN
	Enlarged vestibular aqueduct	1	3.1%	Normal CN and cochlear turns
	Mondini with normal CN	1	3.1 %	Compatible malformation
	Total Eligible	**30**	**93.7%**	
Ineligible for CI	Cochlear + CN hypoplasia	1	3.1%	Poor expected outcomes
	Cochlear + CN aplasia	1	3.1%	Absolute contraindication
	Total Ineligible	**2**	**6.3%**	
Alternative Options	Auditory brainstem implant	2	6.3%	For CN aplasia cases

Table 6: Final Cochlear Implant Candidacy Determination (N=32)
CI = Cochlear Implant; CN = Cochlear Nerve; WM = White Matter

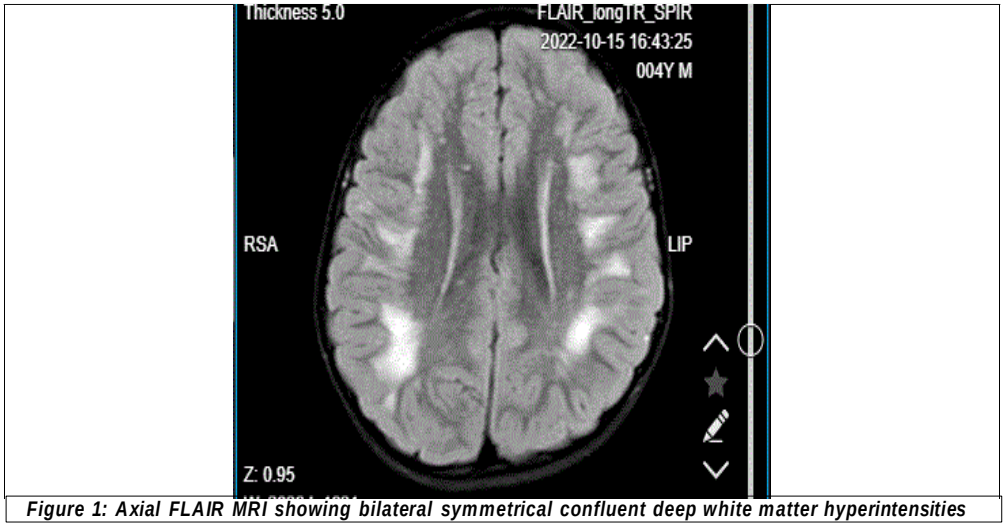

Figure 1: Axial FLAIR MRI showing bilateral symmetrical confluent deep white matter hyperintensities

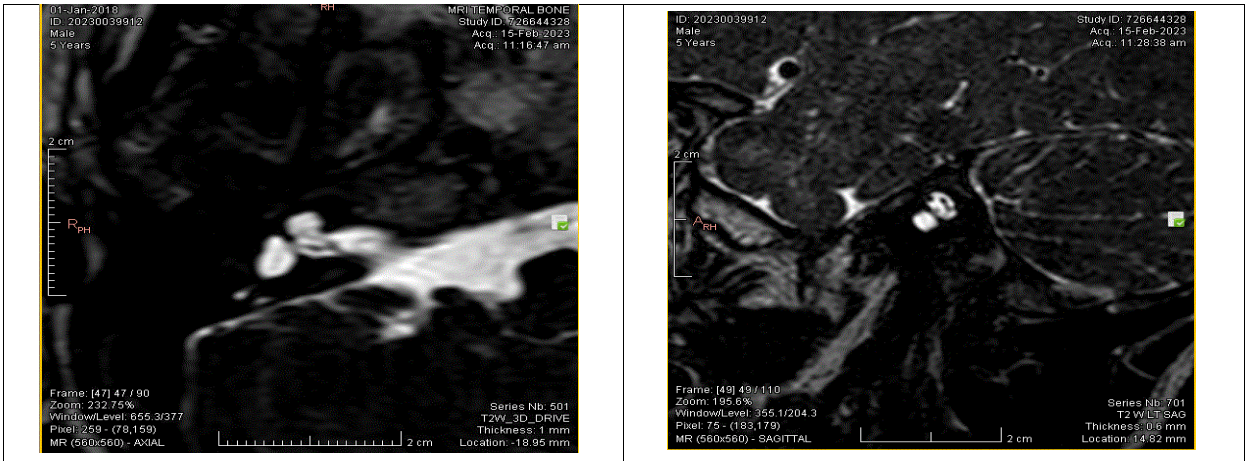

Figure 2: Axial T2 & sag oblique T2 showing hypoplastic cochlea with hypoplastic cochlear nerve

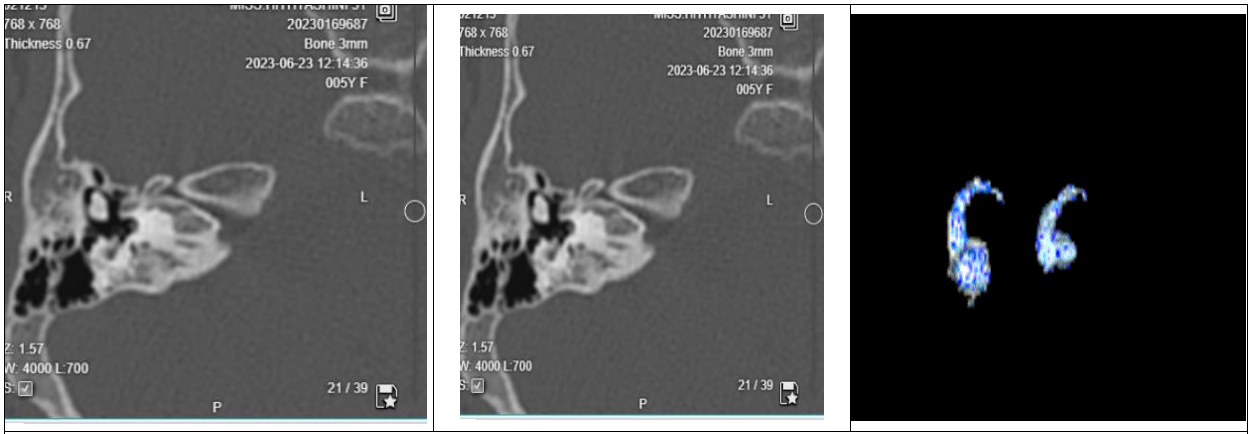

Figure 3: HRCT showing complete cochlear aplasia with stenotic Internal auditory canal. 3 D image showing hypoplastic vestibular apparatus with absent cochlea

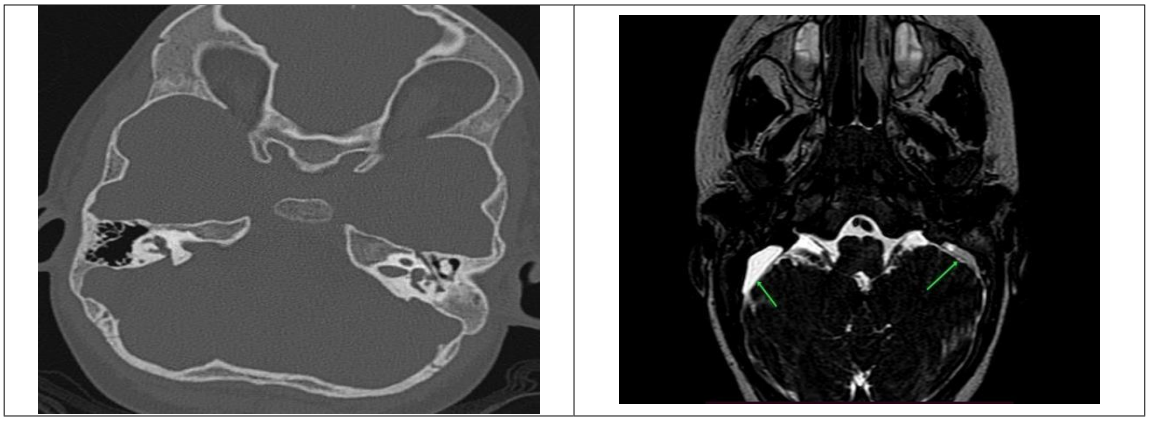

Figure 4: Axial HRCT & Axial MR showing enlarged vestibular aqueduct and endolymphatic sac on both sides (right>left)

Discussion

This retrospective study systematically evaluated the temporal bone imaging findings in 32 prelingually deaf children referred for cochlear implant candidacy assessment under the Government of Karnataka Cochlear Implant Scheme. The comprehensive imaging protocol combining HRCT for bony labyrinth assessment and MRI for membranous labyrinth, cochlear nerve, and brain evaluation provided crucial information for patient selection, surgical planning, and prognostic counselling. The complementary roles of these two imaging modalities were clearly demonstrated throughout the study, with HRCT offering superior visualization of cochlear anatomy, ossicular chain, facial nerve canal, and anatomical variants, while MRI excelled in detecting cochlear nerve abnormalities, brain parenchymal lesions, and membranous labyrinth pathology.[11]

The demographic distribution in this study showed a male predominance with a male-to-female ratio of 1.46:1, which is consistent with several published studies reporting slightly higher prevalence of congenital hearing loss in males.[12] However, this gender difference did not reach statistical significance in our cohort and likely reflects natural variation rather than a true gender predisposition. The age distribution demonstrated that the majority of patients presented at 3 years of age, which is later than the currently recommended optimal implantation age of 12-18 months. This delay in presentation reflects ongoing challenges in early hearing screening, delayed diagnosis, and socioeconomic barriers to accessing tertiary care facilities in developing countries.[13] The importance of early implantation has been consistently demonstrated in literature, with children implanted before 18 months showing significantly better speech and languag e outcomes compared to those implanted after 3 years due to the critical period of auditory cortex plasticity.[14]

The most significant finding in this study was that 78.1 % of patients demonstrated completely normal imaging on both HRCT and MRI, indicating no detectable structural abnormality to explain the profound hearing loss. This high proportion of normal imaging findings is consistent with several large series reporting 60-80% normal imaging in prelingual deafness cohorts.[15,16] These cases are generally attributed to genetic causes of sensorineural hearing loss, particularly mutations in genes encoding connexins, otoferlin, an d other proteins essential for cochlear function. Common genetic causes such as mutations in GJB2 gene encoding connexin 26 and GJB6 gene encoding connexin 30 account for approximately 30-50% of autosomal recessive non-syndromic hearing loss but produce no detectable structural abnormality on imaging.[17] The normal imaging findings in these cases are reassuring from a cochlear implant perspective, as they indicate normal cochlear anatomy suitable for electrode insertion, preserved cochlear nerve, and absence of associated brain abnormali ties. Multiple studies have demonstrated excellent cochlear implant outcomes in children with normal temporal bone imaging, with speech perception and language development outcomes comparable to or better than those with structural abnormalities.[18]

Inner ear malformations were identified in 12.5% of patients in this study, which falls within the 10-25% incidence range reported in most large series from developed countries.[19,20] The four inner ear malformations identified in this study represented different points on the spectrum of cochlear developmental abnormalities. Bilateral cochlear hypoplasia, characterized by reduced cochlear size and fewer than two complete turns, results from developmental arrest at approximately 5 weeks of gestation and is frequently associated with cochlear ner ve hypoplasia or aplasia. The combination of cochlear and cochlear nerve hypoplasia in one of our patients represented a relative contraindication to standard cochlear implantation, as outcomes are significantly poorer when the cochlear nerve diameter is less than 50% of the facial n erve diameter. The most severe malformation encountered was bilateral cochlear aplasia with complete absence of cochlear structures, stenotic internal auditory canals, and bilateral cochlear nerve aplasia. This constellation of findings represents an absolute contraindication to cochlear implantation as there is no cochlear structure to accommodate an electrode and no functional cochlear nerve to transmit elect rical signals to the auditory brainstem. These patients are potential candidates for auditory brainstem implantation, which involves placing an electrode array directly on the cochlear nucleus in the brainstem, bypassing both the cochlea and cochlear nerve. However, auditory brainstem implant outcomes are generally poorer than cochlear implant outcomes with most patients achieving only environmental sound awareness and limited open-set speech recognition.

The identification of bilateral Mondini malformation in one patient represented an important finding with significant implications for surgical planning. Mondini malformation, more accurately termed incomplete partition type II, is characterized by normal basal turn but cystic confluence of the middle and apical turns and is frequently associated with enlarged vestibular aqueduct. Despite the structural abnormality, cochlear implantation can be successfully performed in Mondini malformation cases with generally favourable outcomes, particularly when the cochlear nerve is normal and at least one complete cochlear turn is present for electrode placement. However, several surgical considerations are important including risk of cerebrospinal fluid gusher due to abnormal communication between the subarachnoid space and inner ear through an enlarged cochlear aperture, potential difficulty with electrode insertion due to abnormal cochlear architecture, and increased risk of postoperative meningitis requiring pneumococcal vaccination.

The identification of bilateral enlarged vestibule, vestibular aqueduct and endolymphatic sac as an isolated finding in one patient represents a significant clinical entity. Enlarged vestibular aqueduct syndrome is one of the most common radiologically identifiable inner ear malformations associated with sensorineural hearing loss, accounting for approximately 1-10% of paediatric hearing loss cases in various series. The vestibular aqueduct is considered enlarged when it measures greater than 1.5 mm at the midpoint or greater than 2.0 mm at the operculum on HRCT. The hearing loss associated with enlarged vestibular aqueduct is typically progressive and fluctuating, often triggered by minor head trauma. The pathophysiology is believed to involve abnormal pressure transmission from increased cerebrospinal fluid pressure through the enlarged aqueduct to the membranous labyrinth. Cochlear implantation in patients with isolated enlarged vestibular aqueduct and normal cochlear nerve has been shown to be safe and effective, with outcomes comparable to patients with normal anatomy. However, families should be counselled about the underlying anatomical variant and the potential for progressive hearing loss even after implantation.

The detection of isolated brain white matter abnormalities in 9.4% of patients without inner ear structural malformations or cochlear nerve abnormalities represents an important finding with implications for counselling and prognostic expectations. The pattern of periventricular and subcortical white matter hyperintensities on FLAIR sequences in these three patients was suggestive of previous insult during the perinatal or early postnatal period. Congenital cytomegalovirus infection is the most common non-genetic cause of sensorineural hearing loss in children, affecting approximately 0.5-0.7% of live births with 10-15% of infected infants developing hearing loss. The hearing loss in ganglion neurons rather than structural malformations, explaining the normal temporal bone imaging. Associated brain white matter abnormalities occur in 20-40% of symptomatic congenital cytomegalovirus infection cases and may be accompanied by microcephaly, ventriculom egaly, intracranial calcifications, and cerebellar hypoplasia. Similarly, bacterial meningitis during infancy can cause sensorineural hearing los s through inflammatory damage to cochlear structures and may produce white matter injury through associated encephalitis or vascular complications. The identification of mastoid and middle ear pathology in 15.6% of patients highlights the importance of temporal bone imaging in detecting conditions that may influence surgical timing or approach. Acute otomastoiditis in three patients required antibiotic therapy and resolution of infection before proceeding with cochlear implant surgery to minimize the risk of implant infection and ensure optimal surgical field visualization. Cochlear implant surgery in the presence of active middle ear or mastoid infection significantly increases the risk of device infection, potentially necessitating explanation with devastating consequences for the patient. Chronic otomastoiditis and sclerotic mastoids in two patients presented different challenges, as the dense bony sclerosis and reduced pneumatization increase surgical difficulty, prolong operative time , and may necessitate more extensive drilling during mastoidectomy. In such cases, selecting the contralateral ear with better pneumatization when possible facilitates easier surgical access and potentially reduces complication rates.

The documentation of anatomical variants in 9.4% of patients underscores the importance of detailed preoperative imaging anal ysis and communication of these findings to the surgical team. High jugular bulb, present in 3.1% of our patients, can significantly complicate round window approach for electrode insertion. When the jugular bulb extends above the level of the inferior tympanic annulus, it m ay obscure visualization of the round window niche and limit the angle of electrode insertion, potentially necessitating alternative approaches such as extended round window or cochleostomy anterior to the round window.

Deep sigmoid groove with anterior extension into the mastoid cavity, classified as Type IV in one patient, significantly narrows the surgical corridor for posterior tympanotomy approach. The normal distance from the sigmoid sinus to the posterior external auditory canal ranges from 12-18 mm, and when this distance is reduced to less than 10 mm, as seen in our Type IV case, the safe working space for drilling the facial recess and inserting the electrode is markedly limited. Low-lying tegmen, present in one patient, increases the risk of inadvertent dural exposure or injury during cortical mastoidectomy, as the reduced vertical dimension between the external auditory canal and m iddle cranial fossa floor leaves less margin for error during superior dissection.

The cochlear morphometric measurements in patients with normal anatomy demonstrated dimensions consistent with published normative data from Indian and international populations. The mean cochlear height of approximately 4.2-4.3 mm and mean cochlear base width of 8.8-8.9 mm in our study are comparable to values reported in previous studies of Indian patients and fall within the normal ranges established by multiple international studies. The calculated cochlear duct length averaging 33.9-34.3 mm provides important information for electrode selection, as cochlear implant electrodes are available in various lengths ranging from 20 mm to 31.5 mm for standard electrodes. Selecting an appropriate electrode length that matches the individual patient's cochlear duct length optimizes electrode positioning with the goal of achieving full insertion without trauma to the delicate cochlear structures or excessive coiling in the basal turn.

The assessment of cochlear nerve status using high-resolution three-dimensional T2-weighted MRI sequences represented a critical component of the preoperative evaluation. The cochlear nerve was normal in 93.7 % of patients in our study, which is consiste nt with reported incidences of cochlear nerve abnormalities ranging from 5-15% in various series of children with congenital sensorineural hearing loss. The ability to confidently identify the cochlear nerve as a distinct structure separate from the facial nerve on oblique sagittal images through the internal auditory canal and to assess its caliber relative to the facial nerve is essential for prognostic counselling. Multiple studies have established that cochlear nerve diameter less than 1.0 mm or less than 50% of the facial nerve diameter is associated with poor cochlear implant outcomes, while complete cochlear nerve aplasia represents an absolute contraindication to cochlear implantation.

The overall cochlear implant eligibility rate of 93.7% in this study is encouraging and demonstrates that comprehensive imagi ng evaluation successfully identifies the small proportion of patients with anatomical contraindications while confirming suitability for the vast majority. This high eligibility rate is consistent with other series reporting 85-95% of prelingually deaf children being suitable candidates for cochlear implantation following imaging evaluation. The 6.3% of patients deemed ineligible due to cochlear and cochlear nerve abnormalities highlights the critical importance of preoperative imaging, as proceeding with surgery in these cases would result in poor outcomes, unn ecessary operative risk, and significant financial costs.

This study had several limitations that should be acknowledged. The retrospective design limited the ability to control for a ll potential confounding variables and precluded standardized long-term follow-up to correlate imaging findings with actual surgical observations and postoperative outcomes. The relatively small sample size of 32 patients, while representative of the caseload at a single center over 18 months, limited the statistical power to detect associations between variables and meant that rare malformations and variants were not encountered. The lack of genetic testing in the majority of patients with normal imaging prevented definitive etiological diagnosis in 78. 1% of the cohort. The absence of postoperative imaging follow-up precluded assessment of electrode position and detection of delayed complications. Future prospective studies with larger sample sizes, inclusion of genetic testing, correlation with intraoperative findings, and long-term follow-up of cochlear implant outcomes would provide valuable additional insights.

Conclusion

This retrospective study demonstrates that comprehensive preoperative imaging evaluation using HRCT and MRI plays an indispensable role in determining cochlear implant candidacy in prelingually deaf children. The complementary nature of these two modalities provides complete assessment of temporal bone anatomy, inner ear structures, cochlear nerve status, and associated brain abnormalities. The identification of normal imaging in 78.1% of patients confirms that the majority of prelingual deafness cases are suitable for cochlear implantation with favourable anatomical prerequisites. The detection of inner ear malformations in 12.5% and cochlear nerve abnormalities in 6.3 % allows appropriate counselling regarding surgical approach, electrode selection, and expected outcomes.

The study emphasizes the importance of systematic evaluation using standardized reporting protocols to ensure that all relevant anatomical structures and potential variants are assessed and communicated to the multidisciplinary cochlear implant team. The critical role of MRI in detecting isolated cochlear nerve aplasia despite normal cochlear anatomy underscores the necessity of including both CT and MRI in the standard preoperative evaluation protocol. Identification of anatomical variants such as high jugular bulb, deep sigmoid groove, and low-lying tegmen in 9.4% of patients provides crucial information for surgical planning and complication avoidance. Detection of mastoid pathology requiring treatment before surgery in 15.6% of cases demonstrates the value of imaging in optimizing surgical timing. The high overall eligibility rate of 93.7% validates the screening process while the identification of absolute contraindications in 6.3 % of patients prevents futile surgical procedures in cases where outcomes would be poor. Radiologists must maintain high levels of expertise in temporal bone imaging and remain updated on evolving classification systems for inner ear malformations, emerging genetic correlations, and refinements in surgical techniques that influence the relevance of specific imaging findings. Effective communication between radiologists and otolaryngologists through standardized reporting templates and multidisciplinary conferences ensures optimal utilization of imaging information in clinical decision-making. As cochlear implant technology continues to advance and government-sponsored programs expand access to this transformative intervention, the role of comprehensive imaging evaluation will remain central to successful patient selection and outcome optimization in prelingual deafness management.

DECLARATIONS

Patient Consent

Informed written consent for the imaging procedures was obtained from the parents or legal guardians of all patients at the time of clinical evaluation. As this was a retrospective analysis of imaging data acquired as part of routine clinical care, the requirement for additional informed consent for research participation was waived by the Institutional Ethics Committee with maintenance of strict patient confidentiality throughout the study.

Ethical Approval

This study was conducted in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments. The study protocol was reviewed and approved by the Institutional Ethics Shimoga institute of medical sciences, Shivamogga, Karnataka, India.

Conflict of Interest

The authors declare that they have no conflicts of interest, financial or otherwise, related to this research. No funding was received fo r conducting this study from any commercial or non-commercial entities.

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Author Contributions

Dr. Madhu S Revatagaon contributed to data collection, image analysis, literature review , and manuscript preparation. Dr. Uma Pandurangi suggested the concept of research work, supervised the study design, provided expert guidance on image interpretation & structured reporting format, and critically revised the manuscript. Dr. Shruthi S contributed to case referral, literature review, statistical analysis, and manuscript review.Dr. Sadananda S B contributed to data collection, image analysis and preliminary reporting. All authors read and approved the final manuscript.

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