American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2026; 27: e954074
DOI: 10.12659/AJCR.954074

Received: 2026.05.11
Accepted: 2026.07.27
Available online: 2026.09.01
Published: 2026.XX.XX

Cochlear Implantation in a Child With Arnold–Chiari Syndrome, Severe Inner Ear Malformations, and Cochlear Nerve Hypoplasia: A Case Report

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Financial support: None declared
Conflict of interest: None declared

Patient: Female, 18-month-old
Final Diagnosis: Severe inner ear malformations • Arnold-Chiari syndrome
Symptoms: cochlear nerve hypoplasia • hearing loss
Clinical Procedure: —
Specialty: Otolaryngology

Objective: Rare disease

Background: Arnold–Chiari syndrome is a congenital malformation of the posterior cranial fossa associated with neurological and otoneurological abnormalities, including hearing loss of variable severity. Cochlear implantation in patients with complex central nervous system malformations remains challenging due to uncertain auditory outcomes. This study aimed to evaluate the feasibility and early outcomes of cochlear implantation in a child with Arnold–Chiari syndrome and severe bilateral inner ear malformations.

Case Report: A 1.5-year-old girl with Arnold–Chiari malformation, hydrocephalus, spina bifida, and profound bilateral sensorineural hearing loss was evaluated for cochlear implantation. Imaging studies revealed severe bilateral inner ear malformations, including cochlear hypoplasia, accompanied by multiple intracranial abnormalities. Despite early hearing aid use, auditory development remained limited. The patient underwent left-sided cochlear implantation with a MED-EL SYNCHRONY 2 device using a standard electrode array. Intraoperatively, significant cerebrospinal fluid leakage ("gusher") occurred during cochleostomy and was successfully controlled using autologous tissues and tissue adhesive. Twelve months after implantation, follow-up audiological evaluation demonstrated improved behavioral auditory responses and increased responsiveness to environmental sounds, as reported by the parents.

Conclusions: Cochlear implantation may provide meaningful auditory benefit in selected patients with Arnold–Chiari syndrome despite severe inner ear and central nervous system abnormalities. Comprehensive preoperative imaging and multidisciplinary assessment are essential for surgical planning and prognosis evaluation. Further studies involving larger patient cohorts are required to better define prognostic factors influencing cochlear implantation outcomes in this population.

Keywords: Arnold–Chiari Malformation • Cochlear Implantation • Hearing Loss
Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/954074

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Introduction

Arnold–Chiari syndrome comprises a group of congenital structural abnormalities of the posterior cranial fossa characterized by downward displacement of the cerebellar structures and brainstem toward the spinal canal [1,2]. The first description of this malformation was provided in 1883 by the Scottish physician John Cleland, who studied children with spinal dysraphism [3]. However, it was Julius Arnold and Hans von Chiari who later investigated these malformations in detail and described the mechanisms underlying their development [4,5].

This disorder results in abnormal cerebrospinal fluid flow and compression of neural structures at the craniovertebral junction. Depending on the extent and nature of the displacement, several types of malformations can be distinguished [6]:

- Type 0 – characterized by syringomyelia without hindbrain herniation or cerebellar tonsillar descent;
- Type 1– the most common form, typically diagnosed in adults; characterized by downward displacement of the cerebellar tonsils leading to impaired cerebrospinal fluid circulation, without association with neural tube defects;
- Type 1.5 – a more advanced variant of Type I malformation;
- Type 2 – a pediatric form commonly associated with hydrocephalus and spina bifida; characterized by herniation of the cerebellar vermis and the fourth ventricle into the spinal canal, frequently accompanied by myelomeningocele;
- Type 3 – similar to Type II, but associated with an occipital encephalocele and/or high cervical abnormalities instead of myelomeningocele;
- Type 3.5 – shares features of Type III, with the presence of an occipitocervical encephalomyelocele and communication with the upper gastrointestinal tract;
- Type 4– characterized by severe cerebellar hypoplasia associated with occipital encephalocele;
- Type 5 – complete absence of the cerebellum.

The clinical presentation of the disease is heterogeneous and encompasses a broad spectrum of neurological symptoms, including headaches, balance disturbances, sensory deficits, and bulbar symptoms. Increasing attention has also been directed toward the coexistence of otoneurological manifestations, including hearing disorders of varying type and severity [7,8].

Hearing loss associated with Arnold–Chiari syndrome may have a multifactorial etiology. The pathophysiology of this phenomenon is thought to involve both the mechanical impact of displaced posterior fossa structures on the brainstem and cranial nerve nuclei, as well as disturbances in cerebrospinal fluid circulation that can affect the function of auditory pathway structures [9,10]. In some cases, coexisting inner ear abnormalities or disturbances in neural conduction involving the vestibulocochlear nerve have also been reported [11,12]. Consequently, some patients develop hearing loss that progressively worsens, leading to significant impairment in quality of life and social functioning.

In cases where conventional methods of hearing rehabilitation, such as hearing aids, fail to provide sufficient improvement in sound perception, cochlear implantation may be considered [13]. Cochlear implants are an established treatment modality for profound sensorineural hearing loss and deafness, and the indications for cochlear implantation are being progressively expanded to include new patient populations, including individuals with congenital malformation syndromes and rare disorders [14,15]. This technology is widely used in both pediatric and adult patients with various etiologies of hearing loss [16,17]. However, in patients with central nervous system disorders, including abnormalities of the posterior cranial fossa, the effectiveness of cochlear implantation may be more difficult to predict due to potential damage to the central components of the auditory pathway [18].

The aim of the present study was to evaluate whether cochlear implantation was technically feasible and associated with early auditory benefit in a child diagnosed with Arnold–Chiari syndrome despite severe inner ear malformations and central nervous system abnormalities, based on the available clinical and audiological data. The study focused specifically on surgical feasibility and short-term auditory outcomes rather than on broader conclusions regarding long-term treatment effectiveness.

Case Report

Medical History and Clinical Interview

A 1.5-year-old girl was admitted to the Audiology and Phoniatrics Clinic for comprehensive diagnostic evaluation and assessment of cochlear implantation feasibility. Prenatal examination during the 19th week of gestation revealed hydrocephalus and spina bifida. Following birth, the patient required respiratory support with nasal continuous positive airway pressure (nCPAP). Physical examination demonstrated multiple congenital abnormalities, including open lumbosacral myelomeningocele, macrocephaly, widened cranial sutures, enlarged fontanels, and clubfoot deformity. Surgical repair of the myelomeningocele was performed on the first day of life. Due to severe psychomotor delay, the child was unable to walk and could only roll from side to side. She remained under continuous neurosurgical care.

Newborn hearing screening and subsequent follow-up screening tests yielded abnormal bilateral results. The first auditory brainstem response (ABR) examination, performed at the

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age of 4 months, demonstrated absent bilateral responses to click stimuli. Hearing aids were introduced at the age of 5 months; however, auditory development remained limited, raising concerns regarding the effectiveness of conventional auditory rehabilitation.

Given the coexistence of severe neurological abnormalities and suspected complex auditory pathway involvement, particular attention was paid to preoperative assessment and realistic expectation-setting prior to cochlear implantation qualification.

Physical examination

Oral cavity: Oral mucosa was pink and moist. Tongue mobility was within normal limits. Salivary gland duct openings were without pathological changes.

Throat: Palatine arches were symmetrical and non-hyperemic. Soft palate mobility was preserved. Tonsils were symmetrical, not enlarged, without purulent exudate.

Nose: Nasal passages were patent.

Ears: The right ear tympanic membrane was intact and dull in appearance; the left ear tympanic membrane was intact and dull in appearance.

Speech and Language Therapy Consultation

The patient participated in auditory-verbal therapy twice weekly and received motor rehabilitation using the Vojta and Bobath methods.

Behavior during examination: The child was calm, sitting on her mother's lap, manipulating objects with her hands, and establishing only occasional eye contact with the therapist.

Auditory responses and speech comprehension: The child demonstrated sporadic and uncertain auditory responses to suddenly occurring environmental sounds. During the visit, responses to musical instruments (drum, tambourine, and triangle) were assessed. Auditory reactions included sound searching and turning toward the source of the sound; however, the responses were difficult to evaluate reliably. The child showed greater interest in visual and movement-related stimuli.

Speech production: Speech development remained at the vocalization stage, with the emergence of syllables such as "mama" and "ba-ba." The child responded to changes in facial expressions of the speaking person. Increased vocal activity was observed mainly in situations of discomfort or when attempting to attract the caregiver's attention. Alternative and augmentative communication (AAC) strategies were being introduced.

Psychological Consultation

The patient attended the consultation accompanied by her mother. During the examination, the child appeared calm and cheerful, lying in the stroller and observing the surroundings. She established eye contact with the examiner. According to the mother, the patient was socially responsive, smiled frequently, and communicated her needs primarily through crying. In interactions with others, visual input appeared to be the dominant communication channel.

Psychomotor development was delayed due to spina bifida. The child remained under multidisciplinary specialist care and participated in continuous intensive rehabilitation, including early developmental support programs. According to the mother, the patient had been making developmental progress and demonstrated good cooperation during therapy sessions.

The child used hearing aids regularly, and no difficulties related to device use were reported. The patient's mother had no prior knowledge regarding cochlear implants; however, she actively asked questions concerning relevant issues and received the information calmly and attentively.

Imaging Tests

High-resolution computed tomography (CT) of the temporal bones demonstrated severe bilateral inner ear malformations. On the right side, the findings were consistent with extreme cochlear hypoplasia (with possible aplasia), cystic enlargement of the vestibule, and severe dysplasia of the semicircular canals. On the left side, cochlear hypoplasia with partial preservation of the basal turn, enlargement of the vestibule, dysplasia of the lateral semicircular canal, and enlargement of the vestibular portion of the vestibular aqueduct were identified. The oval and round windows, as well as the middle ear structures, were preserved bilaterally. Narrowing of the right internal auditory canal was observed. Based on the osseous anatomy, CT findings suggested severe hypoplasia (or aplasia) of the right cochlear nerve and possible hypoplasia of the left cochlear nerve; however, definitive assessment of the cochlear nerves was performed using MRI. The inner ear anatomy indicated an increased risk of an intraoperative cerebrospinal fluid "gusher".

Magnetic resonance imaging (MRI) of the head confirmed the presence of complex congenital central nervous system malformations, including hypoplasia of the corpus callosum, marked enlargement of the ventricular system with a normally functioning ventriculoperitoneal shunt, hypoplasia of the right cerebellar hemisphere and cerebellar vermis, and features of Arnold–Chiari malformation with herniation of the left cerebellar tonsil into the spinal canal. MRI also confirmed bilateral congenital

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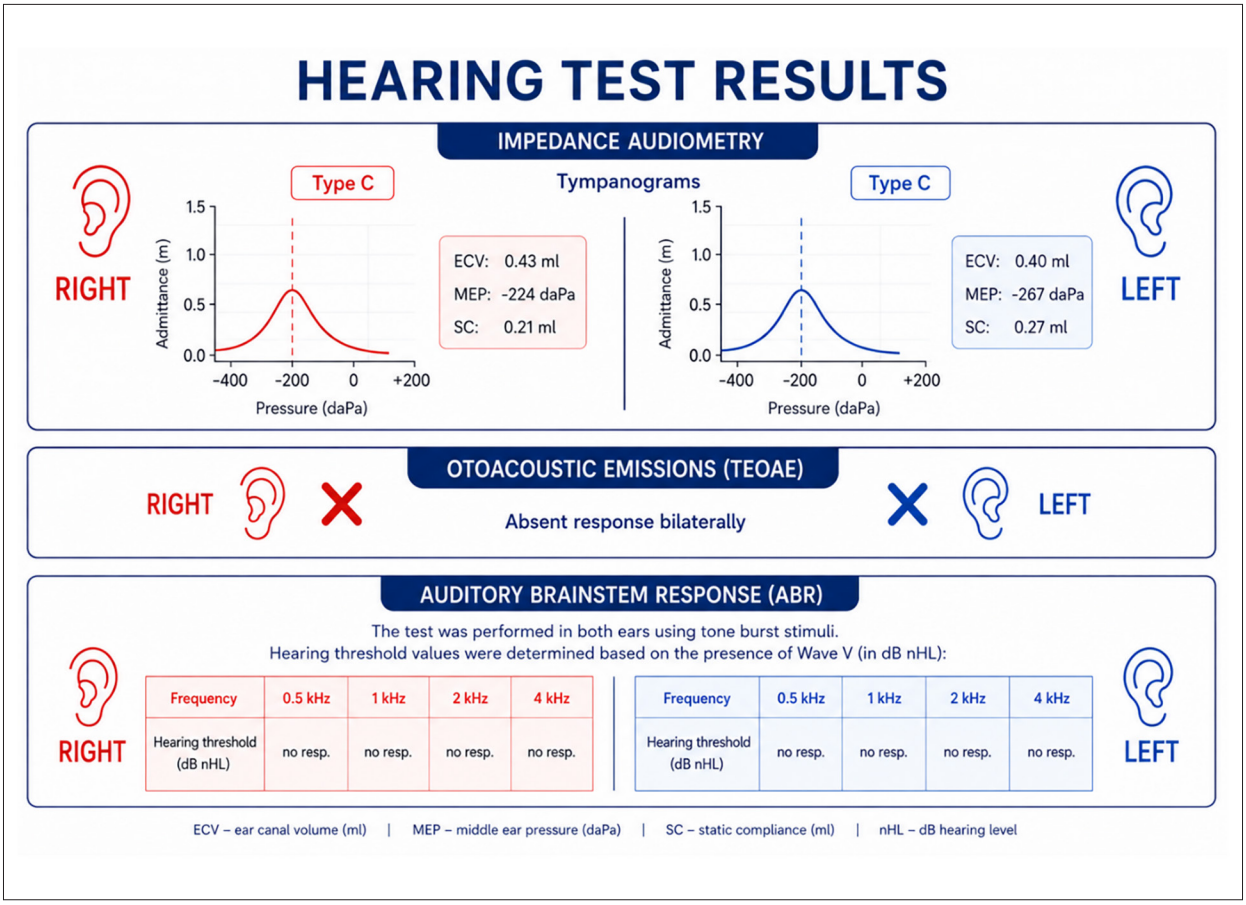

Figure 1. Preoperative audiological assessment. Impedance audiometry demonstrated bilateral type C tympanograms, indicating negative middle ear pressure. Transient evoked otoacoustic emissions (TEOAE) were absent bilaterally. Auditory brainstem response (ABR) testing using tone-burst stimuli revealed no detectable Wave V responses at 0.5, 1, 2, or 4 kHz in either ear, consistent with profound bilateral sensorineural hearing loss.

inner ear malformations. Direct visualization of the cranial nerves demonstrated hypoplasia of the facial (VII) and vestibulocochlear (VIII) nerves on the right side. On the left side, both the facial and vestibulocochlear nerves appeared normal and were visualized throughout their course within the internal auditory canal, indicating preserved cochlear nerve anatomy on the implanted side. No abnormalities were identified within the cerebellopontine angles.

Audiological Evaluation

During hospitalization for audiological assessment, the patient underwent a series of examinations aimed at evaluating the degree of hearing loss. The results of the audiological tests are presented in **Figure 1**.

Impedance audiometry revealed bilateral type C tympanograms, indicating negative middle ear pressure in both ears. Transient evoked otoacoustic emissions (TEOAE) were absent bilaterally. Auditory brainstem response (ABR) testing performed using tone burst stimuli demonstrated no detectable Wave V responses at 0.5, 1, 2, and 4 kHz in either ear, consistent with profound bilateral hearing loss.

Surgical Treatment

In September 2024, the patient underwent cochlear implantation of the left ear using a MED-EL SYNCHRONY 2 device with a Standard electrode array. The implantation procedure was performed according to Skarżyński's 6-step method [19]. An extended approximately 3-cm "S"-shaped postauricular incision was performed. A musculoperiosteal flap was elevated. With meticulous preservation of the intact periosteum, the mastoid process was opened using a chisel, and a cortical bone plate was harvested. Atticoantromastoidectomy and posterior tympanotomy were subsequently performed using cutting and diamond burrs. The mastoid was well pneumatized.

The size of the posterior tympanotomy was approximately 3 × 4 mm. Visualization of the round window niche was limited.

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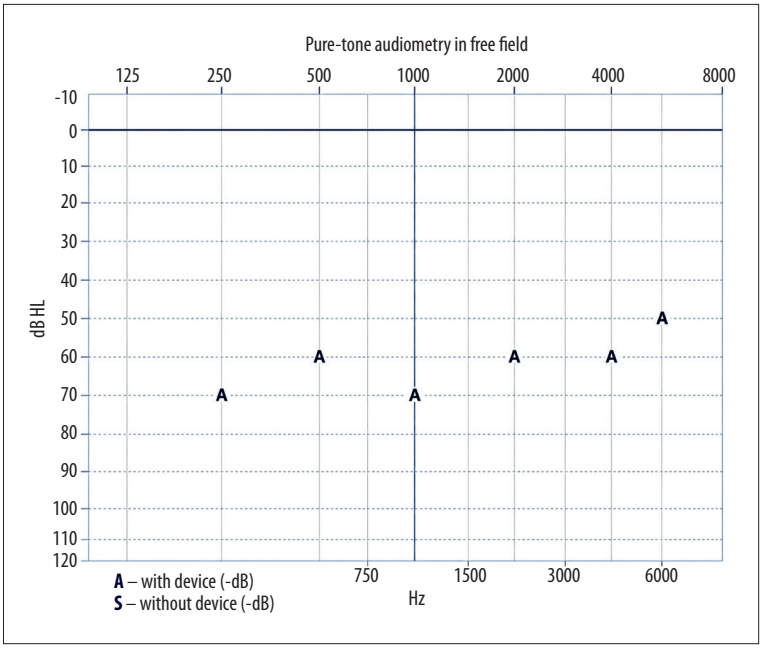

Figure 2. Twelve-month postoperative aided free-field pure-tone audiometry performed using Behavioral Observation Audiometry (BOA) and Visual Reinforcement Audiometry (VRA). Functional hearing thresholds ranged from approximately 50 to 70 dB HL across frequencies from 250 to 8000 Hz, demonstrating improved auditory access with the cochlear implant. Filled circles represent aided hearing thresholds obtained during behavioral testing.

Extensive drilling of the lateral bony overhang was required to expose the round window membrane. The condition of the round window could not be assessed.

A cochleostomy was performed using a burr. A relatively profuse outflow of fluid from the cochlea occurred. The active electrode array of the implant (all channels) was inserted completely, initially manually and subsequently with forceps, encountering slight resistance. The electrode was sealed with fragments of soft tissue fascia and Tissucol fibrin glue. The cerebrospinal fluid leak subsided initially but subsequently appeared from the vestibular side. The region of the oval window was sealed using fragments of fat, fascia, and periosteum with tissue adhesive, resulting in cessation of the fluid leak.

Within the area of the posterior tympanotomy approach, the electrode was secured to avoid compression of the facial nerve canal and the wall of the external auditory canal. The mastoid cavity surrounding the electrode was filled with antibiotic-soaked Spongostan, displacing the electrode away from the posterior canal wall. The mastoid opening was closed with the previously harvested bone plate and tissue adhesive.

The internal implant processor was fixed on the surface of the temporal bone within a periosteal-fascial pocket. A continuous suture was placed in the periosteum to seal the tissues overlying the implant, followed by a second layer of interrupted inverted sutures. The skin was closed with a continuous suture and an external dressing was applied [20,21].

Audiological Tests After Operation

Twelve months after surgery, the patient attended a follow-up visit at the Department of Implants and Auditory Perception. The parents reported no problemss related to the implant and observed that the child had been responding increasingly better to environmental sounds.

Aided free-field audiometry was performed using Behavioral Observation Audiometry (BOA) and Visual Reinforcement Audiometry (VRA). Functional hearing thresholds ranged from approximately 50 to 70 dB HL across frequencies from 250 to 8000 Hz (**Figure 2**). Compared with the preoperative audiological assessment, in which no auditory brainstem responses were detected and behavioral auditory responses were minimal, the postoperative findings indicated improved access to sound with the cochlear implant. Given the patient's severe developmental delay, these results should be interpreted cautiously, as the assessment was based on behavioral measures supported by parental observations.

Discussion

Patients with Arnold–Chiari syndrome and coexisting auditory system abnormalities constitute a particularly challenging clinical group. The present case adds confirmatory evidence that cochlear implantation can still provide early auditory benefit even in the presence of highly unfavorable combined peripheral and central nervous system abnormalities. The main contribution of this case report is the demonstration of surgical feasibility and early postoperative auditory improvement despite severe bilateral

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inner ear malformations, cochlear nerve hypoplasia, hydrocephalus, and multiple intracranial abnormalities. In addition, the case highlights the importance of careful preoperative imaging assessment and surgical preparedness in patients at increased risk of intraoperative cerebrospinal fluid leakage ("gusher").

The literature emphasizes that the effectiveness of cochlear implantation in children with congenital inner ear malformations is more variable than in patients without coexisting anatomical anomalies [22,23]. A particularly important prognostic factor remains the presence and degree of development of the cochlear nerve. Hypoplasia or aplasia of the eighth cranial nerve may limit the effectiveness of electrical stimulation; however, it does not constitute an absolute contraindication to cochlear implantation [24,25]. In the present case, despite hypoplasia of the auditory nerves, improvement in sound perception and behavioral responses to auditory stimuli was observed following activation of the cochlear implant system. This observation is consistent with previous reports indicating that even partially preserved auditory pathway structures can enable functional auditory benefits [26].

Comparable findings have been reported in patients with other severe inner ear malformations. Studies involving common cavity deformity, cochlear hypoplasia, and incomplete partition anomalies demonstrated that meaningful auditory benefit can be achieved despite markedly abnormal cochlear anatomy, although postoperative outcomes are generally more variable than in patients with normal inner ear structures [27]. Papsin et al [28] and Sennaroğlu et al [27] reported improved auditory perception and environmental sound awareness in implanted children with major cochleovestibular malformations, emphasizing that residual neural integrity rather than anatomical appearance alone is a key determinant of postoperative performance. The early auditory improvements observed in the present patient are therefore consistent with outcomes reported in other complex inner ear malformations, despite the additional presence of central nervous system abnormalities.

An important clinical issue in patients with severe inner ear malformations is the increased risk of intraoperative cerebrospinal fluid leakage ("gusher") [29]. In the presented case, profuse fluid outflow from the cochlea occurred during cochleostomy and was successfully controlled using autologous tissues and tissue adhesive. Similar complications have been repeatedly reported in patients with cochlear anatomical abnormalities and enlargement of the vestibular aqueduct [30,31]. This case therefore additionally supports the technical feasibility of cochlear implantation in anatomically high-risk patients when appropriate surgical preparation and experience are ensured.

Another noteworthy aspect of this case is the coexistence of Arnold–Chiari malformation and severe bilateral inner ear abnormalities. Although hearing loss and vestibular symptoms have been described in patients with Chiari malformations, the association between Arnold–Chiari syndrome and major congenital inner ear malformations remains poorly understood [32]. It has been hypothesized that abnormalities affecting neural tube formation, hindbrain development, and cerebrospinal fluid dynamics during early embryogenesis can contribute to the simultaneous occurrence of posterior fossa anomalies and malformations of the developing otic structures. However, current evidence is insufficient to determine whether this combination represents a distinct developmental spectrum or merely a coincidental coexistence of rare congenital abnormalities. Further clinical and genetic studies are needed to clarify the underlying mechanisms [33,34].

From a practical perspective, this case highlights several important considerations for clinical decision-making in medically complex children. Comprehensive preoperative imaging and evaluation of cochlear nerve status are essential not only for surgical planning and anticipation of gusher risk, but also for realistic counseling regarding expected postoperative outcomes. In patients with severe neurological and anatomical abnormalities, conventional speech-language outcomes may develop slowly or remain limited [35,36]. Nevertheless, even modest postoperative improvements—such as increased responsiveness to environmental sounds and improved behavioral auditory reactions observed in the present patient—may represent clinically meaningful benefit for both the child and the family. Therefore, outcome assessment in this population should be multidimensional and should include functional behavioral changes and quality-of-life aspects in addition to standard audiological measures [37].

Conclusions

Cochlear implantation was technically feasible in this child with Arnold–Chiari syndrome despite severe inner ear malformations and complex central nervous system abnormalities. Early postoperative follow-up demonstrated improvement in auditory responsiveness and behavioral reactions to sound, although long-term auditory and language outcomes remain uncertain. This case highlights the importance of comprehensive preoperative imaging, careful patient selection, multidisciplinary assessment, and cautious interpretation of postoperative benefit in medically complex children. Further studies involving larger patient cohorts are required to better define prognostic factors influencing cochlear implantation outcomes in this population.

Limitations

The main limitation of the present study is that it describes a single case with a relatively short postoperative follow-up

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period. Further studies involving larger groups of patients with Arnold–Chiari syndrome and coexisting inner ear malformations are necessary to better identify prognostic factors influencing the effectiveness of cochlear implantation.

Institution Where Work Was Done

The work was done in the Institute of Physiology and Pathology of Hearing, Warsaw, Poland.

Informed Consent Statement

All study participants, or their legal guardian, provided informed written consent prior to study enrollment.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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