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CASE STUDY

Anatomical and clinical aspects and outcomes of bilateral cochlear implantation in cochlear hypoplasia type IV – a case report

Anatomiczne i kliniczne aspekty oraz wyniki obustronnej implantacji ślimakowej w hipoplazji ślimaka typu IV – studium przypadku

Authors' Contribution:
A – Study Design
B – Data Collection
C – Statistical Analysis
D – Manuscript Preparation
E – Literature Search

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ABSTRACT:

Cochlear hypoplasia is a congenital inner ear malformation (IEM) characterized by a reduced external cochlear dimension, usually accompanied by an abnormal internal architecture. Type IV cochlear hypoplasia is a cochlea with hypoplastic middle and apical turns. It may occur along with dislocation of the facial nerve, associated with semicircular abnormalities, less clearly marked promontory, or stapedial fixation. Such patients can present a broad spectrum of audiological test results, from sensorineural or mixed mild to profound hearing loss. The above anatomical changes may be responsible for intraoperative difficulties during cochlear implantation. In the studied case, a 6-month-old patient was diagnosed with an inner ear malformation – cochlear hypoplasia type IV on both sides. Computed tomography with multiplanar and 3D reconstruction was performed to analyze the middle and inner ear anatomy in detail. Both types of imaging reconstruction helped decide which cochlear implant electrode to choose. Perimodiolar-positioned cochlear implant electrode was found to be the most suitable choice. The patient underwent sequential bilateral cochlear implantation with expected incomplete electrode array insertion on both sides. First repeatable auditory responses were observed 2 months after the second implant activation. Good parental cooperation with therapists and adequately defined developmental goals in the presented patient allowed the multidisciplinary team to take advantage of the child's intellectual abilities and choose a suitable communication method; however, the patient's auditory responses were obtained slowly. The final auditory results cannot be predicted in inner ear malformations due to abnormal anatomical structure and, thus, heterogeneous innervation within the deformed cochlea. The programming of the sound processor must be individual in each case, based on the child's behavior observation and, if possible, objective test results. Patients with cochlear malformations usually require higher stimulation intensities to obtain sound sensations than patients with a typical cochlear structure.

KEYWORDS: cochlear hypoplasia, cochlear implantation, congenital malformations, hearing loss, inner ear, speech therapy

STRESZCZENIE:

Hipoplazja ślimaka to wrodzona wada rozwojowa ucha wewnętrznego charakteryzująca się zmniejszonym zewnętrznym wymiarem ślimaka, któremu zwykle towarzyszy jego nieprawidłowa architektura wewnętrzna. Hipoplazja typu IV zdefiniowana jest jako ślimak z hipoplastycznym środkowym oraz szczytowym zakrętem. Może jej towarzyszyć zaburzony przebieg nerwu twarzowego, nieprawidłowości budowy kanałów półkolistych, mniej wyraźnie zaznaczone promontorium lub fiksacja strzemiączka. Pacjenci z taką patologią mogą prezentować szerokie spektrum wyników badań audiologicznych, od niedosłuchu odbiorczego do mieszanego oraz stopnia od niewielkiego do głębokiego. Powyższe zmiany anatomiczne mogą być odpowiedzialne za trudności śródoperacyjne podczas wszczepienia implantu ślimakowego w przypadku niedosłuchu głebokiego. W opisywanym przypadku u 6-miesięcznej pacjentki rozpoznano wadę rozwojową ucha wewnętrznego – obustronną hipoplazję ślimaka typu IV. Tomografia komputerowa z rekonstrukcją wielopłaszczyznową oraz 3D została wykorzystana w celu szczegółowej analizy anatomii ucha środkowego i wewnętrznego. Oba rodzaje rekonstrukcji obrazowej były pomocne w podjęciu decyzji o wyborze elektrody implantu ślimakowego. Elektroda perimodiolarna została uznana za najodpowiedniejszą w powyższym przypadku. Pacjentka została poddana sekwencyjnemu, obustronnemu wszczepieniu implantu ślimakowego ze spodziewanym, niepełnym wprowadzeniem elektrody do ślimaka po obu stronach. Pierwsze powtarzalne reakcje słuchowe zaobserwowano po 2 miesiącach od aktywacji drugiego implantu. Dobra współpraca rodziców z terapeutami i odpowiednio określone cele rozwojowe

w przedstawionym przypadku pozwoliły multidyscyplinarnemu zespołowi na wykorzystanie możliwości intelektualnych dziecka i dokonanie wyboru odpowiedniej metody komunikacji, jednakże odpowiedzi słuchowe u pacjentki uzyskiwano powoli. Ostateczne audiologiczne rezultaty wszczepienia implantów ślimakowych są trudne do przewidzenia w przypadku wad rozwojowych ucha wewnętrznego ze względu na nieprawidłową budowę anatomiczną, a zatem niejednorodność unerwienia w zdeformowanym ślimaku. Programowanie procesora dźwięku musi być przeprowadzane indywidualnie w każdym przypadku oraz oparte na obserwacji zachowania dziecka i, jeśli to możliwe, na obiektywnych wynikach badań. Pacjenci z wadami rozwojowymi ślimaka wymagają większej intensywności stymulacji w celu uzyskania wrażeń dźwiękowych niż pacjenci z typową budową ślimaka.

SŁOWA KLUCZOWE: hipoplazja ślimaka, implantacja ślimakowa, niedosłuch, terapia słuchowo-językowa, ucho wewnętrzne, wady wrodzone

ABBREVIATIONS

IEM – inner ear malformation
CH – cochlear hypoplasia
MPR – MultiPlanar Reconstruction

INTRODUCTION

Cochlear hypoplasia is the congenital inner ear malformation (IEM) characterized by a reduced external cochlear dimension, usually accompanied by an abnormal internal architecture. Type IV cochlear hypoplasia is a cochlea with hypoplastic middle and apical turns. It may occur along with dislocation of the facial nerve, associated with semicircular abnormalities, less clearly marked promontory, or stapedial fixation [1, 2]. Such patients can present a broad spectrum of audiological test results, from sensorineural or mixed mild to profound hearing loss. The above anatomical changes may be responsible for intraoperative difficulties during cochlear implantation.

CASE PRESENTATION

Patient description and clinical aspects

A 6-month-old girl was admitted to the department of otorhinolaryngology to extend the hearing loss diagnosis found in hearing screening tests after birth. The family history of hearing loss was negative. The patient was born at 41 weeks gestation by cesarean section with a score of 10 on the Apgar scale. In general pediatric assessment, a set of congenital defects was found, including narrowing of the branches of pulmonary arteries, umbilical hernia, skin aplasia in the right knee area, decreased muscle tone, and laryngomalacia. Otoscopy revealed a normal tympanic membrane, preceded by a wide, typical external auditory canal. An ABR showed bilateral profound sensorineural hearing loss accompanied by type A tympanogram and absent stapedius muscle reflexes on both sides. The patient started using hearing aids bilaterally at the age of 4 months.

Anatomical aspects of cochlear malformation

Computed tomography of the temporal bones was ordered to complement the diagnostics. It revealed bilateral cochlear malformations with preserved basilar turn and hypoplastic, anteriorly and medially located middle and apical turns (Fig. 1.). The deformity was classified as type IV cochlear hypoplasia [2]. In addition, there were no lateral semicircular canals on either side, with fully developed anterior and posterior semicircular canals (Fig. 2.). To accurately assess the inner ear's anatomy, we used the MultiPlanar Reconstruction (MPR) option to inspect the malformation details using 2D presentations (RadiAnt DICOM Viewer 2022.1 64-bit Medixant, Poznan, Poland). In addition, complementing the computed tomography, we performed 3D reconstruction in the Mimics Innovation Suite 24.0 program (Materialise, Belgium). Fig. 3., 4. show 2D presentations and 3D reconstruction of a normal ear to compare it to the congenital inner ear anomaly shown in Fig. 1., 2. for easier visual pathology identification.

In addition, referring to the work of Pamuk et al. [3], specific measurements of the malformed cochlea in the presented case were performed. In the described case, 3 measurements were possible: basal turn length, basal turn maximum height, and mean cochlear duct lateral wall length. The remaining measurements, cochlear canal midscalar and lateral wall length were impossible to perform due to the disturbed internal architecture of the cochlea and the lack of certainty as to the quality of their performance. The measurement results are presented in Tab. I. The CT scans with measurements of the right cochlea in the presented case are shown in Fig. 5. for a better understanding of methodology.

In the presented patient, both types of imaging reconstruction helped decide which cochlear implant electrode to choose. Perimodiolar-positioned cochlear implant electrode was found to be the most suitable choice. Cochlear sequential implantations on both sides were performed 10 months apart, with the first surgery taking place at the age of 10 months. Electrode placement in the malformed cochlea was possible through standard surgical access using posterior tympanotomy. On the right side, the electrode was inserted through cochleostomy, and on the left side, through the round window. The maximum electrode insertion was up to 10 electrode contacts on both sides, as predicted preoperatively. The postoperative period was uneventful, and the patient was dismissed for further rehabilitation.

Logopedic and psychological aspects

At first counseling for the first implant, the patient did not present any auditory reactions or pre-communication behaviors. She did not maintain eye contact, did not share the field of attention, and did not reach for objects with her hands. Frequent "freezing" moments in space were observed, and active communication with the child was impossible. For 6 months after the first cochlear implantation, no changes in the behavior or auditory progress of the child were observed. However, after 6 months, the first auditory responses appeared. The girl began to pay attention to the sounds of musical instruments, although these reactions were not stabilized. 1 year

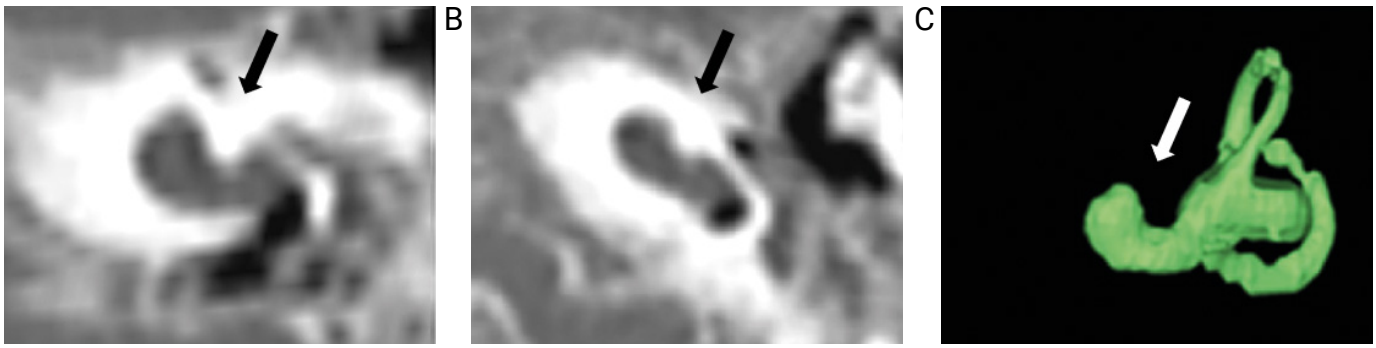

Fig. 1. Malformation of the cochlea in the presented case, shown in the planes selected for the most precise visualization in 2D presentation (A, B) and its (C) 3D reconstruction. Preserved basilar turn and hypoplastic, anteriorly and medially located middle and apical turns are indicated with arrows–cochlear hypoplasia type IV.

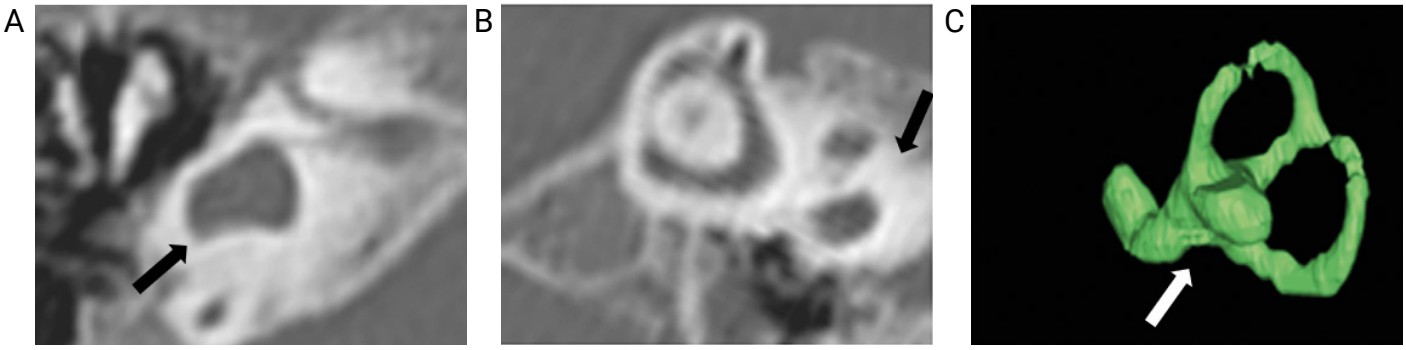

Fig. 2. Malformation of the semicircular canal in the presented case, shown in dedicated planes in 2D presentation (A, B) and its (C) 3D reconstruction. The absent lateral semicircular canal is indicated with arrows. Anterior and posterior semicircular canals are fully developed.

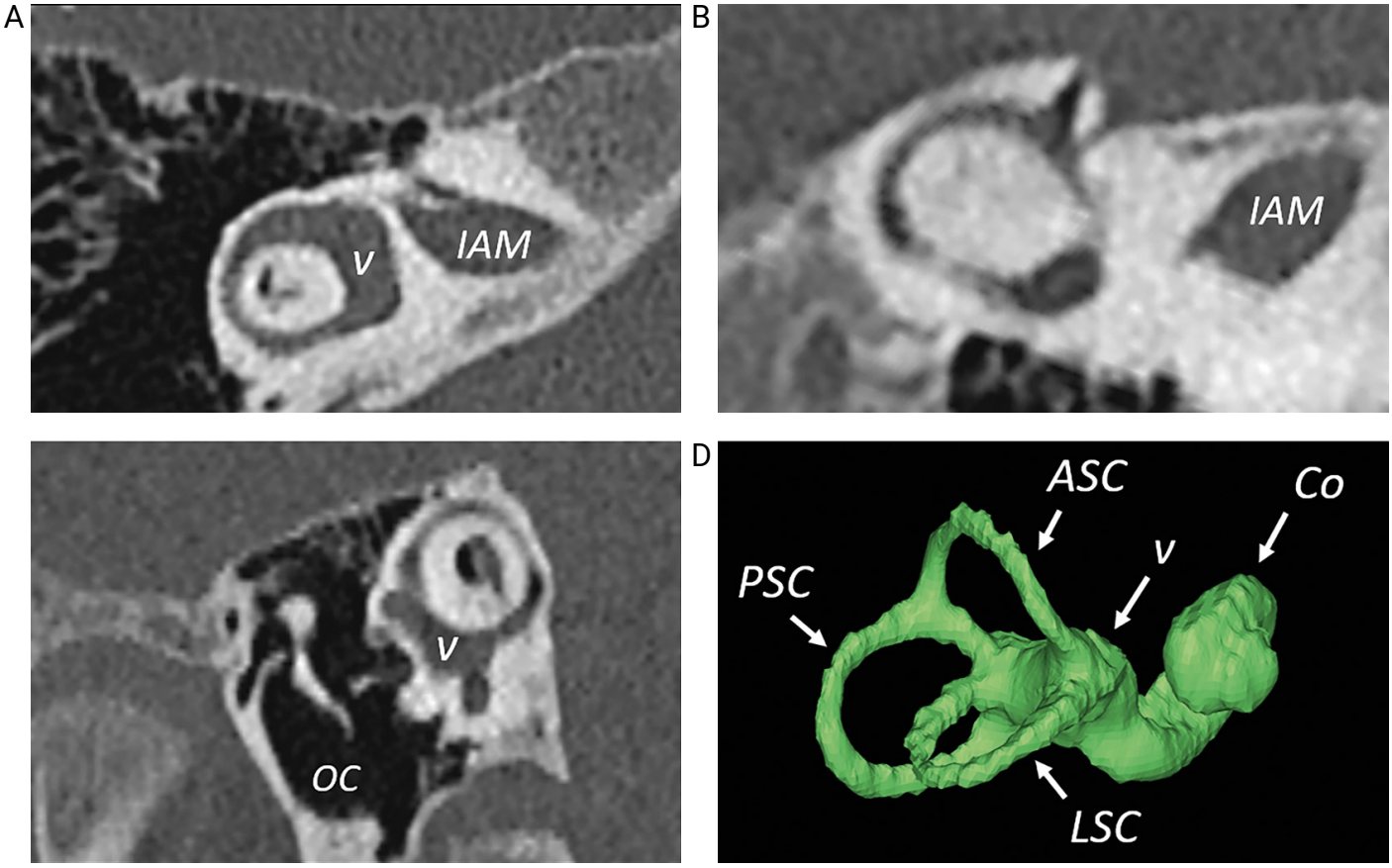

Fig. 3. Normal ear as reference shown with a set of multiplanar images of normal vestibular labyrinth presenting semicircular canals in the 3 reference planes (A, B, C) and its (D) 3D reconstruction. (A) shows the lateral semicircular canal in the axial plane, revealing a typical "signet ring appearance". (B) shows the plane of the anterior semicircular canal (as described by Pöschl). (C) shows the plane of the posterior semicircular canal with visible common crus. (D) shows 3D reconstruction. The markings in the panels present as follows: V – vestibule, IAM – internal acoustic meatus, OC – ossicular chain, PSC – posterior semicircular canal, ASC – anterior semicircular canal, LSC – lateral semicircular canal, Co – cochlea.

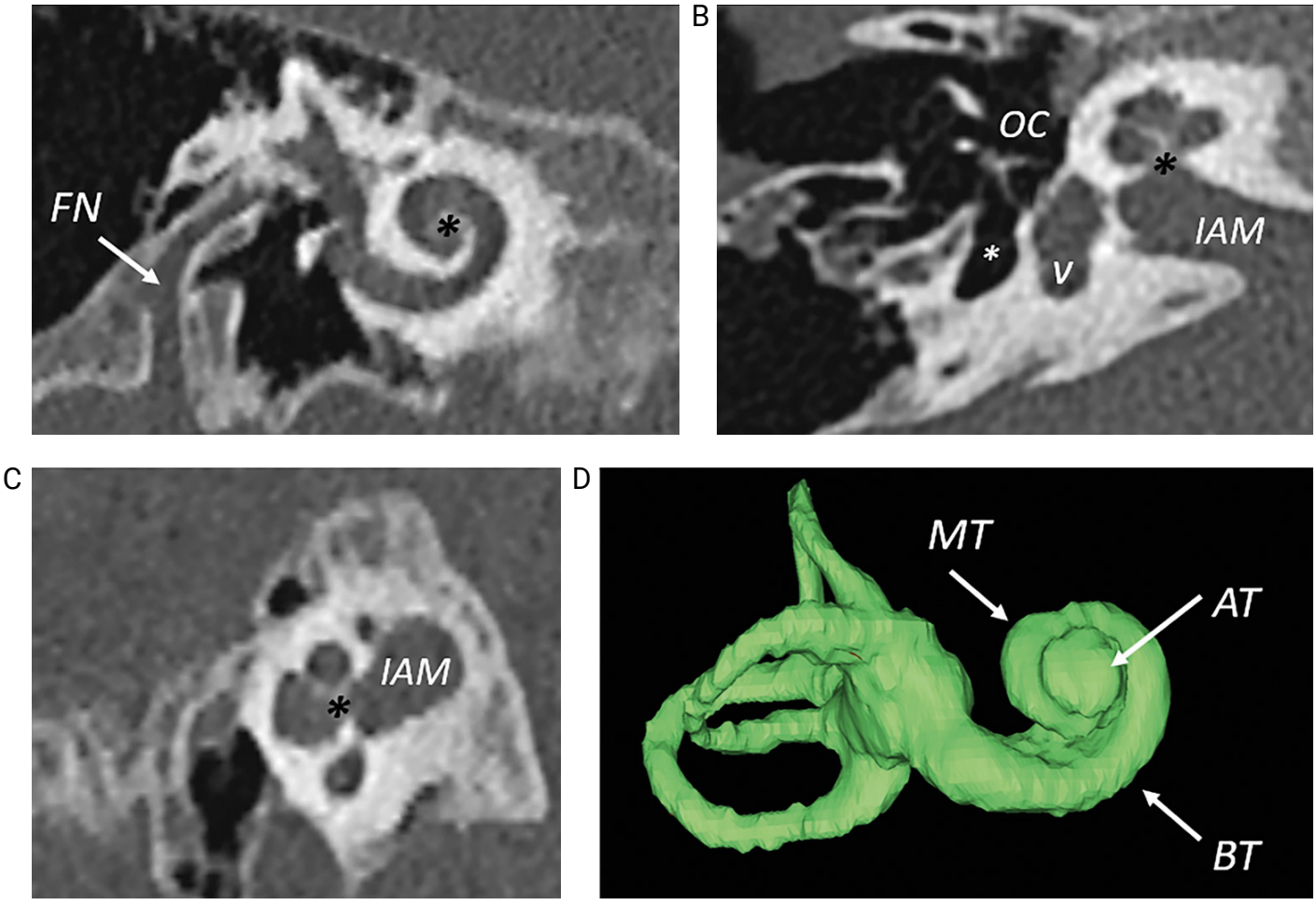

Fig. 4. Normal ear as reference shown with a set of multiplanar images of the normal cochlea and its 3D reconstruction. (A) shows the longitudinal section of the cochlea – the basal, middle, and apical turns around the modiolus (marked with a black "asterisk"). (B) shows the modified axial plane – the modiolus (marked with a black "asterisk") and all 3 cochlea turns; the tympanic cavity is marked with a white "asterisk". (C) shows the perpendicular plane to the modified axial-modiolus (marked with a black "asterisk") and all 3 turns of the cochlea. (D) shows 3D reconstruction. The markings in the panels present as follows: FN – facial nerve, BT – basal turn, MT – meddle turn, AT – apical turn.

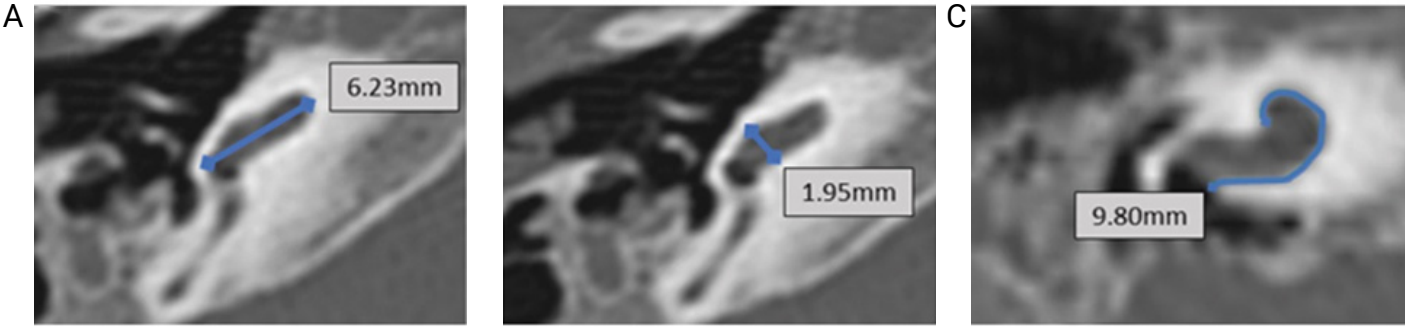

Fig. 5. Computed tomography axial reformatted images showing measurements (mm) of the right cochlea as described by Pamuk et al. [3]. (A) presents the measurement of the basal turn length (6.23 mm), (B) the basal turn maximum height (1.95 mm), and (C) the mean cochlear duct lateral wall length (9.80 mm).

after the first surgery, the first pre-communication behaviors were recorded. The child began to accost the people in the environment with her voice and pulled out her hand for toys.

1 year after the implantation of the first implant, the parents came to the department for second implant counseling for their daughter. During the process, information was provided about the potentially small benefits resulting from the use of bilateral implantation treatment. Due to the significantly delayed overall development of the child, no significant progress was expected in verbal communication. The parents continued her auditory education and logopedic rehabilitation. Due to the lack of progress in rehabilitation using the auditory-verbal method, the total communication method was introduced, with particular attention paid to gesture and sign language. Total communication focused on finding and using suitable communication methods in this case. It aimed to help the patient form connections and interactions with parents and therapists and, with time, ensure information exchange.

Tab. I. Results of the performed measurements of the malformed cochlea in the presented case as described by Pamuk et al. [3].

	Basal turn length (mm)	Basal turn maximum height (mm)	Cochlear duct outer wall mean length (mm)
Right cochlea	6.23	1.95	9.80
Left cochlea	5.62	2.01	10.2

After 5 months, the girl with 2 implants constantly reacted to all sounds of musical instruments. A labile but frequent reaction to her spoken name appeared. She actively communicated with her mother by pointing her finger at objects, perfectly reading the emotional state, and enforcing her needs by crying. When asked, the patient understood and pointed to several parts of the body and animals (onomatopoeia) when their sound was mimicked. At that point, several words in active speech and the passive dictionary appeared.

Postoperative processor fitting strategies

During the activation of the first implant in the right ear, 10 electrodes (el 13–22, the ones that were inserted during surgery) were activated, and the impedances of electrodes were correct. The neural response telemetry (NRT) measurements showed the patient's lack of reaction during the measurements and no neural response in automatic and manual analysis. The patient did not respond behaviorally to thresholds' determination for effective stimulation. 4 programs were written in the processor, varying in intensity – the parents were recommended to observe the child's reaction. During subsequent follow-up visits (1, 2, 3, 4, and 6 months after the activation), the intensities of electric stimulation were gradually increased – without unambiguous responses to the stimulation of individual electrodes and in "live mode" to any sounds up to 8 months after activation.

12 months after activating the first implant (right ear), the implant in the second ear (left) was activated. The parameters during activation and the procedure were analogous to the implant in the right ear. Adjustments were made based on the manually determined thresholds of the auditory nerve response; the records from individual electrodes were differentiated in terms of potentially present neural responses – electrodes that differed in recording (no response versus potentially present responses) were distinguished. On this basis, 12 consecutive electrodes were distinguished (11–22). The approximate settings of the C threshold below the NRT threshold were inspected for the child's response – both to the activation/deactivation of stimulation and the observation of the response to sounds. No reactions were observed at the moment; however, carefully observing the child's reactions was recommended to the parents. At subsequent follow-up visits, NRT inconclusive responses to stimulation were observed. After 2 months, an attempt to determine the thresholds was repeated, and the thresholds were manually determined. 11 effective stimulating electrodes were obtained (no response from electrode 21 was observed; however, it was left in the active form). 2 months after the activation of the processor in the second ear (left), repeatable auditory responses were observed in the first implanted ear (right). Due to the progress in rehabilitation and the first successful designated indicative hearing threshold determination by play audiometry, along with the help of assessment of the hearing threshold by a speech therapist, the thresholds of high-frequency stimulation were raised. However, careful attention was paid not to exceed the comfortable hearing thresholds. Further rehabilitation and observation of the child's reaction to sounds were recommended.

DISCUSSION

Cochlear hypoplasia (CH) was fully described for the first time by Sennaroglu et al. [1], who distinguished 4 types of this anomaly. Apart from the anomalies described in the presented case, this defect may be accompanied by an aberrant course of the tympanic segment of the facial nerve requiring modification of the surgical technique or intraoperative facial nerve monitoring. Moreover, stapes fixation may also be present in patients with CH types III and IV, and these patients may benefit from stapedotomy [2]. In addition, malformations of the semicircular canals may be observed. Li et al. [4] described the case of a patient with CH type IV with a sigmoid sinus obscuring the facial recess such that a posterior tympanotomy or retro-facial approach was impossible to perform, forcing a trans-meatal approach assisted by an endoscope. In our patient's CT scans, the course of the facial nerve and sigmoid sinus location seemed normal (typical), which did not require any modification of the surgical approach to the middle. An approach through atromastoidectomy and posterior tympanotomy was performed. However, the absence of the lateral semicircular canal, which serves as an easy referring point approaching the middle ear safely, made it a little more difficult to identify the position of the facial nerve during surgery and perform posterior tympanotomy. For an experienced otosurgeon, this small difficulty is quite easy to overcome, as it was in our case. Eventually, the surgery did not cause any complications.

In the study by Pamuk et al. [3], morphometric measurements carried out among cochleas with type IV hypoplasia showed no significant differences in basal turn length between CH type IV patients and the control normal group (unlike in CH types I–III). However, the basal turn maximum height differed significantly when comparing each type of hypoplastic cochleas to the normal group. Moreover, the mean mid-modiolar height was significantly different between those with CH type I and those with types II–IV. The measurements in our patient were partly consistent with Pamuk's results. Basal turn length and maximum height were within the ranges for type IV hypoplasia. However, the mean cochlear duct lateral wall length was significantly smaller and corresponded to the ranges found in type I hypoplasia. More research is needed on how to measure in detail the curve in the case of malformations with a more difficult or unclear internal cochlear architecture.

Incomplete cochlear implant electrode insertion in patients with inner ear malformations is reported in the literature more often than in patients without one. In Farhood et al. [5] study, complete insertion was seen in 81.8% of all IEMs compared with non-IEM cohorts, which achieved 98%. In Melo et al. [5] study, incomplete electrode

insertion was necessary for 3.8% of patients with cochlear hypoplasia. In Adunka et al. [6] and Isaiah et al. [7] studies, 4.9% and 6.4% of patients with IEMs had incomplete electrode insertion, respectively. In our case, incomplete electrode insertion was an expected complication during electrode selection before surgery. The choice in our case was the perimodiolar cochlear implant electrode, which, due to its construction idea, adapts to the shape of the modiolus, and thus is positioned closer to it, providing more focused stimulation with a lower current level, which results in more focused neural stimulation and lowers current consumption [8–10]. The cochlear hypoplasia type IV mainly involves the absence of the middle and apical turns, with relatively well-preserved basal turn. The choice of the perimodiolar electrode to fully exploit basal turn potential seemed reasonable. As mentioned before, the problem of incomplete electrode array insertion was expected. Above all, it was easy to overcome by activating only a certain number of contacts inserted in the cochlea. The electrodes left outside were not activated, which is fully reasonable.

In the statistical analysis demonstrated by Melo et al. [5] and Bille et al. [11], the children without inner ear malformations did not achieve statistically significantly better scores than children with one. However, the researchers noted that there were no children in the study group with significant comorbidities or mental retardation, which is considered to negatively influence the outcome of cochlear implantation. Black et al. [12] pointed out 2 main prognostic factors in the systematic literature review: medical/surgical and speech/language. In the first category, negative prognostic factors include, among others, severe malformations of the inner ear, which include hypoplasia and aplasia. An isolated enlarged vestibular aqueduct is considered the best prognosis, while cochlear nerve hypoplasia presents the worst prognosis of all IEMs [7]. In the second group, amongst prognostic factors, the cognitive delay, especially in the coincidence with the motor delay, seems to cause slower speech perception development skills [12–14]. In the presented case, apart from the developmental defect of the cochlea, the main obstacle in achieving satisfactory results of implantation seemed to be a delay in cognitive development. The absence of eye contact and visual interest in the environment, difficulties in sharing the field of attention, "freezing" moments in space, and slower overall perception were observed. These noticeably changed after rehabilitation with cochlear implants; however, the child's development still deviated from the age-appropriate norm.

The final auditory results cannot be predicted in inner ear malformations due to abnormal anatomical structure and, thus, heterogeneous innervation within the deformed cochlea. The programming of the sound processor must be individual in each case, based on the child's behavior observation and, if possible, objective test results, i.e., measurements of electrically evoked potentials from the distal part of the auditory nerve [15]. Regardless of the possibility of objective evaluation of the stimulation thresholds or their absence, the patient's behavior assessment should be used to determine the effective stimulation thresholds; in such cases, the experience of the person programming the implant processor and assessing the child's reactions is extremely important [16, 17]. In our case, based on the observations of parents and speech therapists, the intensities of electric stimulation were gradually increased, especially the thresholds of high-frequency stimulation. Patients with cochlear malformations usually require higher stimulation intensities to obtain sound sensations than patients with a typical cochlear structure [18].

CONCLUSIONS

As shown in the presented case, patients with inner ear malformations require a more individual approach to surgery and rehabilitation and higher stimulation intensities to obtain sound sensations. In the studied case, a 6-month-old patient was diagnosed with an inner ear malformation – cochlear hypoplasia type IV. CT with multiplanar and 3D reconstruction was performed to analyze the middle and inner ear anatomy in detail. The patient underwent sequential bilateral cochlear implantation with expected incomplete electrode array insertion on both sides. 10 electrode contacts in the first implant and 12 in the second were activated, confirmed with impedance measurements. However, the neural response telemetry (NRT) showed no response either in automatic or manual analysis. First repeatable auditory responses were observed 2 months after the second implant activation. Good parental cooperation with therapists and adequately defined developmental goals in the presented patient allowed the multidisciplinary team to take advantage of the child's intellectual abilities and choose the suitable communication method; however, the patient's auditory responses were obtained slowly.

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Competing interests: The authors declare that they have no competing interests.

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