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ORIGINAL ARTICLE

Inner ear malformations – overview, current classifications, challenges in management

Wady wrodzone ucha wewnętrznego – klasyfikacja, przegląd, problemy kliniczne

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

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ABSTRACT:

Introduction: Some patients with profound sensorineural hearing loss have inner ear malformations. Initially, those were believed to be a contradiction to cochlear implantation. However, with the advance of cochlear implant surgery and technology, it has become possible to treat this group of patients. However, cochlear implantation in the case of inner ear malformations can be associated with numerous surgical difficulties and possible complications.

Purpose: The paper aims to present: (1) modern classification of inner ear malformations, (2) surgical and clinical challenges related to all types of malformations, and (3) cases of inner ear malformations identified in the authors' centers.

Material and methods: Of 111 children enrolled for cochlear implantation in three clinical centers, a group of patients with malformations of the inner ear was selected. We analyzed: preoperative imaging studies of patients performed prior to cochlear implantation, hearing tests, surgical protocols of cochlear implantation, results of intraoperative measurements and intraoperative imaging studies. We discuss what is currently believed to be the leading classification of inner ear malformations.

Results: In 19/111 (17%) children, we diagnosed severe inner ear malformations in 35 ears. In 13/19 (68%) patients the malformations were bilateral and symmetrical, while 6/19 (32%) patients had different malformations in the right and left ear or one-sided malformation. All inner ear malformations described in the classification were found, except for rudimentary otocyst. The most common are: cochlear hypoplasia, incomplete partition, and enlarged vestibular aqueduct..

Discussion: Severe inner ear malformations are a major diagnostic and clinical challenge in children qualified for cochlear implantation. They can preclude or considerably complicate cochlear implantation and postoperative care. Inner ear malformations are found in imaging studies even in 20 to 30% of patients with profound hearing loss.

KEYWORDS: brainstem implant, cochlear implant, imaging tests, inner ear malformations

STRESZCZENIE:

Wprowadzenie: U części pacjentów z głębokim niedosłuchem odbiorczym stwierdzane są wady ucha wewnętrznego. Początkowo uważano je za przeciwskazanie do wszczepienia implantu ślimakowego. Jednakże wraz z rozwojem chirurgii i technologii implantów ślimakowych możliwe stało się leczenie również tej grupy pacjentów. Wszczepienie implantu w przypadku wad ucha wewnętrznego może jednak wiązać się z wieloma trudnościami chirurgicznymi i możliwymi komplikacjami.

Cel: Celem pracy jest przedstawienie: (1) współczesnej klasyfikacji wad ucha wewnętrznego, (2) problemów chirurgicznych i klinicznych związanych z poszczególnymi wadami, a także (3) przypadków wad ucha wewnętrznego stwierdzonych w ośrodkach autorów.

Materiał i metody: Ze 111 dzieci kwalifikowanych do wszczepienia implantu ślimakowego w trzech ośrodkach klinicznych wyłoniono grupę pacjentów, u których stwierdzono wady ucha wewnętrznego. Przeanalizowano: badania obrazowe pacjentów wykonane przed założeniem implantu, badania słuchu, protokoły operacyjne ze wszczepienia implantu ślimakowego, wyniki pomiarów śródoperacyjnych oraz śródoperacyjne badania obrazowe. Omówiono klasyfikację wad ucha wewnętrznego uznawaną obecnie za wiodącą.

Wyniki: U 19/111 (17%) dzieci stwierdzono duże wady ucha wewnętrznego w 35 uszach. U 13/19 (68%) pacjentów wada była obustronna i symetryczna, u 6/19 (32%) pacjentów zaobserwowano inne wady w uchu prawym i lewym lub wadę jednostronną. Stwierdzono wszystkie z głównych wad wyróżnionych w klasyfikacji, poza resztkową otocystą. Najczęstsze z nich to: hipoplazja ślimaka, niepełny podział ślimaka i poszerzony wodociąg przedsionka.

Dyskusja: Duże wady wrodzone ucha wewnętrznego są istotnym problemem diagnostycznym i klinicznym u dzieci kwalifikowanych do wszczepienia implantu ślimakowego. Mogą one uniemożliwiać bądź znacząco utrudniać wszczepienie implantu ślimakowego i opiekę pooperacyjną. Duże wady stwierdzane są w badaniach obrazowych nawet u od 20 do 30% pacjentów z głębokim niedosłuchem.

SŁOWA KLUCZOWE: badania obrazowe, implant pniowy, implant ślimakowy, wady ucha wewnętrznego

ABBREVIATIONS

CA – cochlear aplasia
CC – common cavity
CH – cochlear hypoplasia
CLA – complete labyrinthine aplasia
CT – computed tomography
EVA – enlarged vestibular aqueduct
FN – facial nerve
IAC – internal auditory canal
IP – incomplete partition
MRI – magnetic resonance imaging
RO – rudimentary otocyst
ZOMR – meningitis

INTRODUCTION

Inner ear malformations are diagnosed both in patients with normal hearing and in those with varying levels of hearing loss [1] and are often associated with profound sensorineural hearing loss [1]. Such defects occur in 20 to 30% of patients with profound hearing loss [1, 2]. Initially, inner ear malformations were considered a contraindication to cochlear implantation [1, 3, 4], but with the emergence of the first reports of successful implantations and satisfactory patient outcomes, as well as the development of surgery and cochlear implant technology, the treatment of this group of patients has become possible [1, 3, 5]. It should be noted that the results of hearing and speech rehabilitation in children with inner ear malformations are worse than in the population of children with profound hearing loss but with proper inner ear formation [3]. Implantation in the case of inner ear malformations may be associated with numerous surgical difficulties and possible complications [1, 3, 6]. Common issues include: problems with access to the cochlea, identification of the implantable space, abnormal course of the facial nerve, abundant cerebrospinal fluid leak (Gusher), risk of misplacement of cochlear implant electrode (internal auditory canal, hypotympanum, vestibule), incomplete insertion of the electrode or incorrect positioning of electrode in the inner ear (electrode tip fold-over) [1, 6, 7]. In some circumstances, it may be necessary to use less common surgical approaches [1, 6]. For malformations in which the available implantable space is smaller, such as in cochlear hypoplasia or the common cavity, it may be necessary to select the appropriate electrode of an optimal length [1, 6]. The first cochlear turn develops by eight weeks of gestation, the second by about ten weeks, and the remaining half of the turn by week 25 [8]. Each malformation occur at different stages of development of the inner ear. The first 5-element classification of congenital inner ear malformations, based on embryogenesis and used for many years, was presented by Jackler in 1987 [9], while a new, modernized classification was presented by Sennaroğlu in 2017 [1]. In classification proposed by Sennaroğlu et al. inner ear anomalies are classified into eight major groups [1]. Currently, his classification is recognized as the leading one; it is also widely used in recent publications on malformations of the inner ear. A new classification presented by the same author in 2020 additionally addresses the morphology of the semicircular canals and the course of the facial nerve in the temporal bone in: the internal auditory canal, labyrinthine, tympanic and mastoid segments [10].

The purpose of this work is to present: (1) congenital malformations of the inner ear, (2) their frequency and surgical and clinical problems related to individual malformations, and (3) malformations of the inner ear found at the authors' centers. To our knowledge, a detailed overview of the inner ear malformations as well as classification of malformations have not been discussed in the Polish literature so far.

MATERIAL AND METHODS

The study was conducted in accordance with the standards of research ethics for human studies and the Declaration of Helsinki. Bioethics Committee approval number: KB62/2018 (Consent of the Bioethics Committee at the Polish Mother's Memorial Hospital – Research Institute).

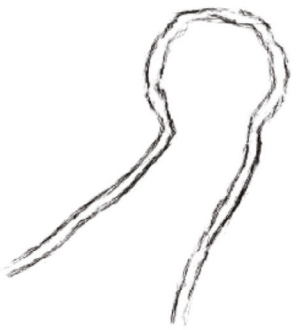
Retrospective analysis of medical records of 111 patients who had been qualified or undergone cochlear implantation from January 1, 2010 to July 1, 2020 in three clinical institutions was performed. Study group included patients with diagnosed inner ear malformations (in all patients preoperative imaging studies were analyzed by radiologists and otolaryngologists).

The inclusion criteria for the study group were:

- availability of preoperative imaging studies: HRCT of temporal bones and MRI in the hospital data collection system;
- inner ear malformation found upon qualification.

The exclusion criterion from the study group was the lack of available imaging studies.Imaging studies were evaluated using RadiAnt DICOM Viewer software.

Tab. I. Subtypes of cochlear hypoplasia and incomplete partition.

COCHLEAR HYPOPLASIA			
			
CH-I	**CH-II**	**CH-III**	**CH-IV**
· ovoid cochlea · no modiolus and spiral lamina.	· normal external outline of cochlea, · defects of modiolus and spiral lamina.	· the internal and external outline are similar to that of a normal cochlea fewer turns, short modiolus and spiral lamina.	· well-developed basal turn, · middle and apical turns are severely hypoplastic.
INCOMPLETE PARTITION			
			
IP-I	**IP-II**	**IP-III**	
Complete absence of modiolus and spiral lamina	Apical part of the modiolus and the corresponding interscalar septa are defective	Spiral lamina present, no modiolus	

The group comprised 19/111 (17%) patients qualified for the study, including 5 girls and 14 boys, who had imaging studies done at the age of 10 months to 8 years.

Data of patients were collected regarding: age, gender and tests performed before and after cochlear implantation, including: preoperative imaging studies, hearing tests (pure tone audiometry, evoked auditory brainstem responses or auditory steady-state evoked potentials – ASSR), surgical protocol of cochlear implantation, results of intraoperative measurements – electrode impedances, NRT and intraoperative imaging tests: X-ray of the temporal bone in a modified Stenvers view and a transorbital view. 3D spatial reconstruction of the inner ear was made using the 3D Slicer Freeware software on the basis of HRCT of temporal bones.

The identified inner ear malformations are classified according to the Sennaroğlu classification from 2017 [1]. A detailed description of the major inner ear malformations identified by this classification is provided below.

Complete labyrinthine aplasia (CLA, Michel anomaly)

Complete labyrinthine aplasia, first described in 1863, is a rare inner ear malformation which frequency is reported in the literature between 1 and 6% [9, 11]. It is formed before the third gestational

week, at the stage of otic placode [11]. The malformation usually occurs bilaterally, although unilateral cases have also been described [11]. However, the other ear is usually not formed properly as well [11]. This malformation involves the absence of: the cochlea, vestibule, semicircular canals, vestibular and cochlear aqueducts [1]. The petrous part of the temporal bone and the otic capsule can be hypoplastic or aplastic, which forms a basis for dividing the defect into three subtypes [1]. There may also be a characteristic flattening of the cochlear promontory [11]. The IAC can consists only of the facial canal [10]. The further course of the facial nerve can be identified in the temporal bone, but most often is abnormal [1, 10]. The auditory ossicles may be properly formed, although defects of the stapes are often described as a feature of CLA (the medial surface of the stapes footplate arises from the otic capsule) [1, 11].

Rudimentary otocyst (RO)

This defect involves a small spherical structure – representations of the otic capsule, which does not communicate with the internal auditory canal [1]; it may be accompanied by residual semicircular canals [1]. Rudimentary otocyst represents a defect "between" aplasia of the labyrinth, in which the structures of the inner ear are completely missing, and the common cavity that communicates with the internal auditory canal.

Cochlear aplasia (CA)

Cochlear aplasia develops around the 5th week of gestation [12]. This malformation is associated with the absence of the cochlea. The vestibule is located in a typical location and can have varied shapes, hence two types of this malformation are distinguished [1]:

1. Cochlear aplasia with normal labyrinth, in which there is a properly developed vestibule and semicircular canals;
2. Cochlear aplasia with dilated vestibule (CADV).

Common cavity (CC)

The common cavity comprises approximately 26% of inner ear malformations and is considered the second most common inner ear malformation; it was first described in 1838 [9, 13]. This malformation occurs around the 4th week of gestation, at the otocyst stage, before the division into: cochlea, vestibule and semicircular canals [14]. The anomaly involves the emergence of a common cavity – a single, spherical structure representing cochlea and vestibule [1]. It communicates with the IAC, the IAC usually enters the cavity at its center, which promotes meningitis [1, 13]. The risk of meningitis in patients with major defects of the inner ear can be up to 40% [15]. The common cavity may be accompanied by variously shaped or rudimentary semicircular canals. In the peripheral part of the cavity it contains the neural tissue of the vestibular and cochlear nerves – the "cochleovestibular nerve" [1]. That said, the number of nerve fibers is usually small [1].

Cochlear hypoplasia (CH)

Cochlear hypoplasia accounts for about 15 to 23.5% of ear malformations and occurs between the 6th and 8th, and in the case of subtype IV, after the 10th week of gestation [12, 16]. It may occur in: CHARGE syndrome (CHD7 mutation), bronchopulmonary dysplasia, Waardenburg syndrome and Down syndrome [16], and can be both unilateral or bilateral [12]. In the case of asymmetric inner ear malformations it most often coexists with IP-I [12]. Other common inner ear malformations include: CC, CLA, CA [12]. In cochlear hypoplasia, both the cochlea and the vestibule are formed, but the external dimensions of the cochlea are smaller [1]. There are various degrees of defects in the internal structure of the cochlea, which is the basis for the division into the following 4 subtypes [1]:

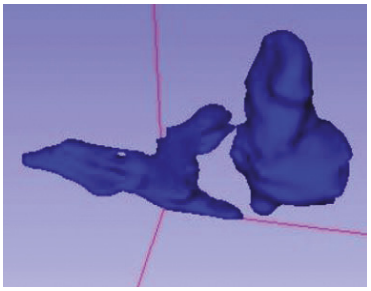
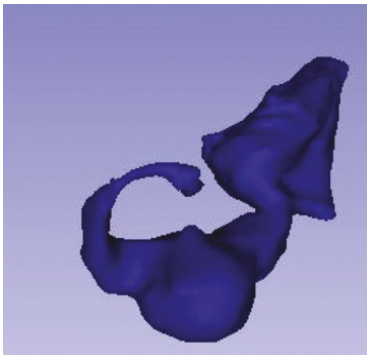
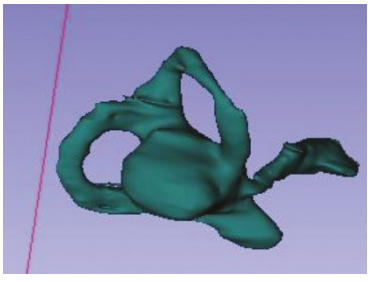
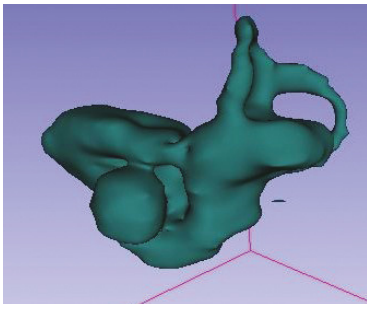
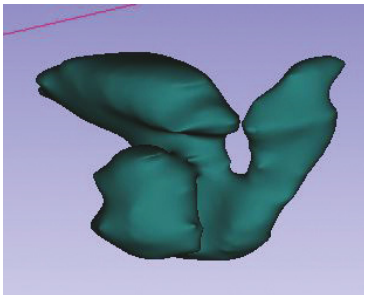
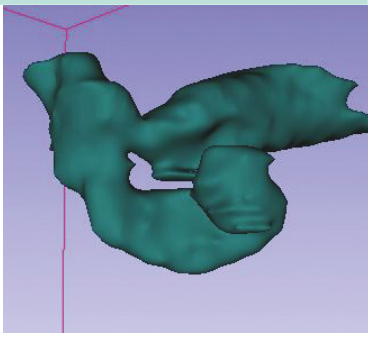
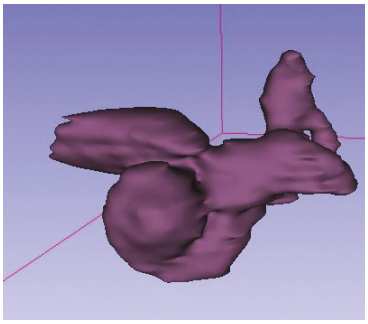
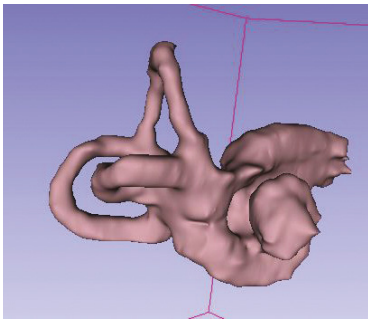
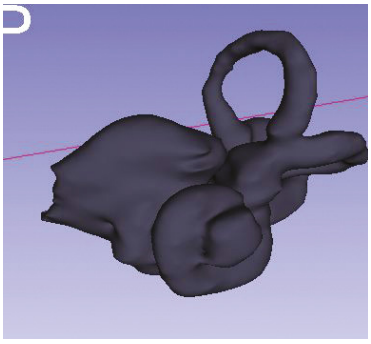
1. Cochlear hypoplasia type 1 (CH I) – a Bud-like cochlea As with any hypoplasia, the cochlea has smaller dimensions. It is spherical, small, and its internal architecture is completely deformed: it does not have a modiolus and a spiral lamina [1]. It can communicate broadly with the IAC [12];
2. Cochlear hypoplasia type II (CH-II) – cystic hypoplastic cochlea. The external outline of the cochlea is normal, but its internal architecture is not, with defective modiolus and interscalar septa [1]. There may be a broad communication with the IAC and the risk of gusher associated with it. This inner ear malformation can coexists with: enlarged vestibular aqueduct, dilated vestibule, stapes footplate defects and recurrent meningitis [1, 12];
3. Cochlear hypoplasia type III (CH-III) – cochlea with <2 turns. The cochlea has smaller dimensions, but correct external and internal structure: short modiolus and spiral lamina. The cochlea has fewer turns [1]. The vestibule and semicircular canals are hypoplastic [12];
4. Cochlear hypoplasia type IV (CH-IV) – a cochlea with a hypoplastic middle and apical turns. Only the basal turn is well developed; the remaining turns are very hypoplastic [1]. This defect arises after the 10th week of gestation, when the basal turn is fully developed and formation of the other turns is inhibited [12].

Incomplete partition (IP)

Inincomplete partition, similarily to cochlear hypoplasia, there is a clear differentiation between cochlea and vestibule with various internal architecture defects [1]. However, the external dimensions of the cochlea are within normal limits [1]. Due to defects in the modiolus and the spiral lamina, there are three types of incomplete partition [1]:

1. Incomplete partition I (IP-I) – characterized by the complete absence of a modiolus and a spiral lamina [1]. This malformation occurs between the 4th and 5th week of gestation [17]. During implantation, there may be a leak of cerebrospinal fluid caused by a defect between the cochlea and IAC, or a defect in the stapes footplate, which may also lead to recurrent meningitis. Additionally, the vestibule can be dilated;
2. Incomplete partition type II (IP-II) – in this subtype, there is no modiolus and no corresponding interscalar septa in the apical part of the cochlea [1]. IP-II coexisting with EVA and dilated vestibule is called Mondini triad [1]. It occurs in Pendred's syndrome (mutation SLC26A);
3. Incomplete partition III (IP-III) – a defect first described in 1971 [18, 19]. IP-III occurs in X-linked hearing loss in patients

Tab. II. Inner ear malformations found in the study group.

MALFORMATION	NUMBER OF EARS WITH MALFORMATION	3D RECONSTRUCTION
Complete labyrinthine aplasia	2	
Cochlear aplasia	1	
Common cavity	1	
Cochlear hypoplasia type I	1	
Cochlear hypoplasia type II	1	
Cochlear hypoplasia type III	4	
Cochlear hypoplasia type IV	3	
Incomplete partition type I	1	
Incomplete partition type II	2	
Incomplete partition type III	8	
Enlarged vestibular aqueduct	6	
Abnormalities of the cochlear aperture	5 + 2	

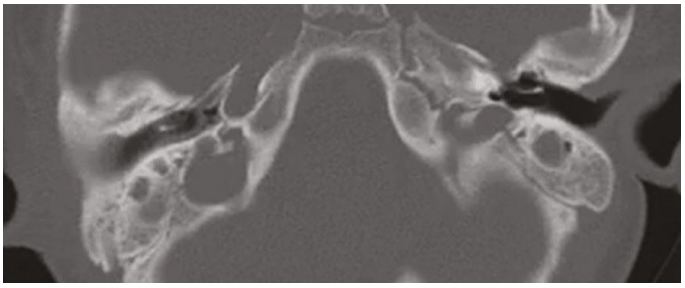


Fig. 1. Bilateral complete labyrinthine aplasia.

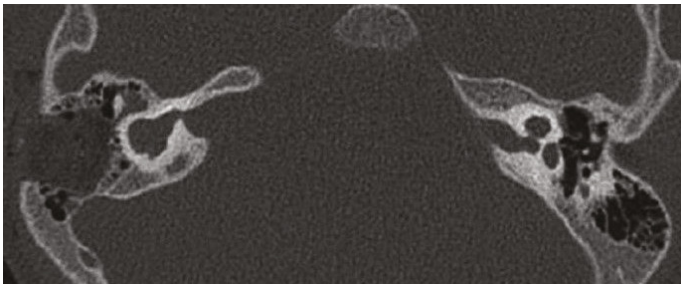


Fig. 3. Common cavity – right ear, cochlear hypoplasia of the left ear.

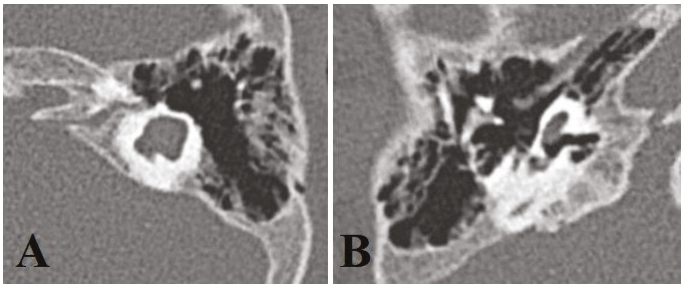


Fig. 2. (A) CADV of the left ear; and (B) cochlear hypoplasia type I of the right ear.

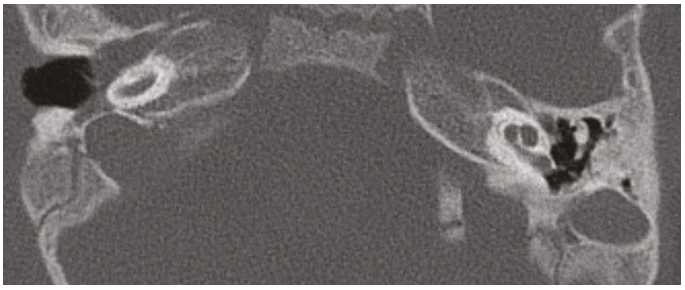


Fig. 4. Bilateral cochlear hypoplasia type III.

with mutations of the POU3F4 gene, and less frequently in COL4A6 mutations [16, 20]. Interscalar septa arepresent, but the modiolus is completely absent [1]. There is often an enlarged, dilated bulbous IAC and incomplete separation of the cochlea from the IAC. Therefore, there is a high risk of a cerebrospinal fluid leak (gusher) and placement of an electrode into the inner ear canal. The stapes may be immobilized [16].

A comparison of the subtypes of cochlear hypoplasia and incomplete partition is presented in Tab. I.

Enlarged vestibular aqueduct (EVA)

Enlarged vestibular aqueduct is considered the most common inner ear malformation in children with hearing loss. It is found in up to 15% of patients, and was first described in 1978 [21–23]. The malformation involves: a properly formed cochlea, vestibule and semicircular canals [1]. The vestibular aqueduct is enlarged – the EVA diagnostic criterion allows for the diagnosis of enlargement if the midpoint between posterior labyrinth and operculum is > 1.5 mm [1]. Previous values reported by various authors ranged from 1 to 4 mm [24]. IP-II is the most common concomitant inner ear malformation [24].

Malformations of the cochlear aperture (cochlear aperture abnormalities)

This defect concerns the "cochlear aperture" – cochlear fossette, or bony cochlear nerve canal that transmits the cochlear nerve from the cochlea to IAC [1]. It can be assessed in axial sections of HRCT, preferably in the so-called mid-modiolar view, which is the most important section to evaluate internal architecture of cochlea [1]. The mid-modiolar viewshows the modiolus, the basal and middle turn and the cochlear aperture [1]. 2 types of cochlear aperture abnormalities can be distinguished [1]:

1. Hypoplasia – when the width of the cochlear aperture in the mid-modiolar view <1.4 mm. It is more common in CHARGE syndrome [16];
2. Aplasia – when there is no communication between the IAC and the cochlea, and there is solid bone between them. In such cases, MRI should be obtained to evaluate VIIIth nerve.

Morphology of the semicircular canals and facial nerve course anomalies

The Sennaroglu classification from 2020 distinguishes four possible morphologies of semicircular canals: normal, hypoplastic, aplastic and dilated [10]. The facial nerve (FN) is assessed in four parts: in the meatal, labyrinthine, tympanic, and mastoid segments [10]. This classification has been published recently; it is not yet widespread; therefore, it will not be discussed in this paper.

RESULTS

Major inner ear malformations were diagnosed in 19/111 (17%) children in 35 ears. Seven of the eight major inner ear malformations distinguished in the Sennaroglu classification were observed [1]. The only malformations that was not found was rudimentary otocyst. The identified malformations are presented in Tab. II.

In 16/19 (84%) patients, the malformation was bilateral. In 3/19 (16%) cases, the defect was found in one ear: in two it was unilateral – unilateral enlarged vestibular aqueduct and unilateral hypoplasia of the cochlear aperture, and one patient with enlarged vestibular aqueduct had previously had cochlear implantation in the opposite healthy ear.

In 13/19 (68%) patients, the defect was symmetrical, and 3/19 (16%) patients had other defects in the right and left ear: common cavity

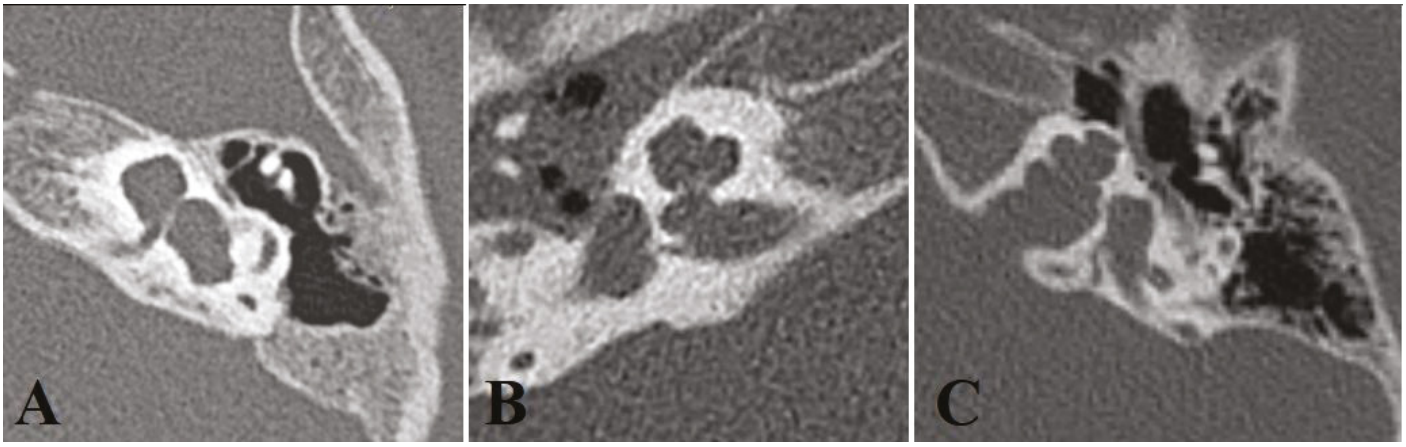


Fig. 5. Incomplete partition: (A) type I, (B) type II, (C) type III.

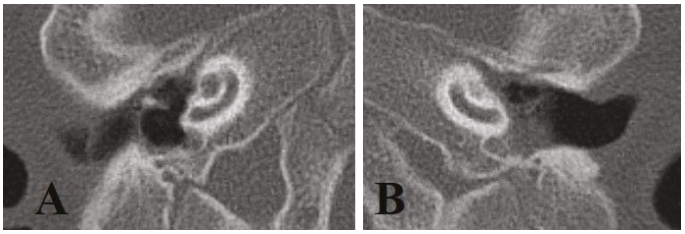


Fig. 6. (A, B) Bilateral cochlear hypoplasia type IV.

with cochlear hypoplasia type II, incomplete partition type I with cochlear hypoplasia type IV and cochlear aplasia with dilated vestibule with cochlear hypoplasia type I. Additionally, one girl with cochlear hypoplasia type 3 (CH III) had bilateral aplasia of the cochlear aperture.

Complete labyrinthine aplasia was found in one boy – bilaterally (Fig. 1.).

Cochlear aplasia was found in one girl (Fig. 2A., B.), who had cochlear aplasia with a dilated vestibule (CADV). In HRCT of the temporal bones, this cavity did not communicate with the IAC; the MRI also showed no communication of CADV with the nerve. The defect was asymmetrical – type I cochlear hypoplasia was found in the other ear, also without communication with the nerve visible on MRI.

A common cavity was found in one girl (Fig. 3.). This cavity communicated with the IAC; MRI revealed communication of the cavity with the nerve. The defect was asymmetrical – cochlear hypoplasia type II was found in the left ear.

Both type I and type II cochlear hypoplasia was found in one ear. These malformations occurred as an asymmetric malformation with other anomalies in the opposite ear. Both patients are described above.

Bilateral cochlear hypoplasia type III was found in two patients (Fig. 4.). Fig. 5. shows a hypoplastic cochlea in the left ear with fewer turns, a short modiolus and a spiral lamina, as well as the first turn of the right cochlea.

Cochlear hypoplasia type IV was found in three ears: bilaterally in one boy (Fig. 6.) and in one ear of a girl with IP-I defect in the other ear.

Incomplete partition type I (Fig. 5A.) was found in one ear of the above described girl with type IV cochlear hypoplasia in the other ear. One boy had incomplete partition type II – the malformation was bilateral (Fig. 5B.). Incomplete partition type III is the most common inner ear malformation identified in the study group, and occurred in 4 patients; in all of them it was bilateral (Fig. 5C.).

The enlarged vestibular aqueduct was the second most common inner ear malformation identified in the studied group. It was found in six ears in four patients, and in two patients it was bilateral (Fig. 7A.).

Abnormalities of the cochlear aperture were found in five ears of three patients. One child had unilateral hypoplasia, while in the other children the malformation was present on both sides (bilateral aplasia and bilateral hypoplasia). Additionally, one girl with bilateral CH-III had bilateral cochlear aperture aplasia, therefore, cochlear aperture abnormalities were found in seven ears. Both subtypes of this anomaly are presented in Fig. 7.

DISCUSSION

In the study group, 7 out of the 8 major inner ear malformations described in the Sennaroğlu classification (2017) were found [1]. 16/19 (84%) patients were qualified for cochlear implantation, whereby 3/19 (16%) patients with severe malformations such as labyrinthine aplasia, cochlear aplasia and bilateral CH-III with bilateral cochlear aperture aplasia were disqualified from cochlear implantation.

Among the described children, there was one case of bilateral complete labyrinthine aplasia in an 8-year-old child (Fig. 1.). The basic conditions necessary for cochlear implantation are: the presence of an implantable space where an electrode can be placed; and of a cochlear nerve, which will allow the signal to be transmitted to further levels of the auditory pathway [7]. The first international consensus on auditory brainstem implantation in children and non-neurofibromatosis type 2 patients from 2011, as well as the subsequent consensus on inner ear malformations, classified this malformation as a well-defined congenital indication for an auditory brainstem implant (ABI) [25, 26].

However, the child had reported too late to be a candidate for an auditory brainstem implant. Ideally ABI surgery should be done before the age of 2, maximum 3 [26]. In this case, it would be

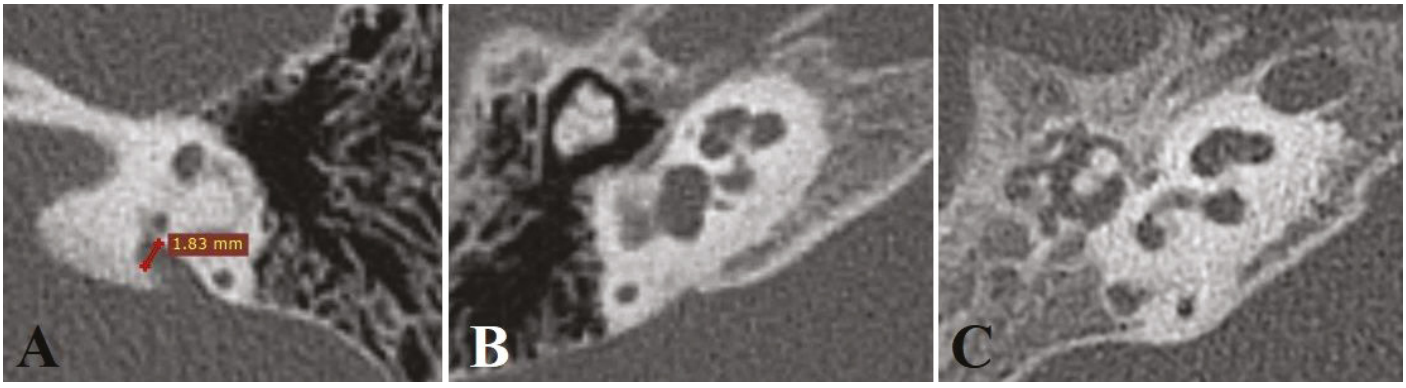


Fig. 7. (A) Enlarged vestibular aqueduct, (B) cochlear aperture hypoplasia and (C) cochlear aperture aplasia.

inadvisable to expose the child to the procedure due to expected poor cochlear implantation effectiveness.

Rudimentary otocyst, the only malformation which was not found at the authors' centers, is also defined as well-defined congenital indication for an auditory brainstem implant [1].

Another malformation found in the study group, which was qualified as a well-defined congenital indication, is cochlear aplasia [25, 26]. However, there are two similar inner ear malformations in the form of a spherical structure visible at the site of the cochlea and vestibule, whose differentiation can be difficult – cochlear aplasia with a dilated vestibule and a common cavity. These two malformations vary in the possibility of cochlear implantation. In CADV there are no nervous structures and cochlear implantation will not provide auditory perception [1]. In common cavity, neural tissue can be present and some patients will benefit from a cochlear implant.

Cochlear aplasia with a dilated vestibule was observed in a 10-month-old girl with cochlear hypoplasia type I in the other ear (Fig. 2.). However, neither any of these spaces communicated with the nerve, nor was there any response to sound during the behavioral assessment, with or without hearing aids. The girl was disqualified from a cochlear implant; she received auditory brainstem implant.

The fourth major malformation – a common cavity – was initially considered a contraindication for cochlear implantation until the report in 1987 by Jackler et al. of the first successful implantation in this malformation [9]. The main surgical difficulties in CC are related to: surgical approach and access to the implantable space, the risk of placing the electrode in the internal auditory canal and inadvertent facial nerve stimulation, profuse leakage of the cerebrospinal fluid (gusher) and the possibility of meningitis as well as atypical course of the facial nerve [27, 28]. The incomplete insertion of the electrode may occur [29]. During implantation, it is possible to use classic mastoidectomy with posterior tympanotomy approach, canal-wall-down mastoidectomy, or direct approach to the cavity through a transmastoid labyrinthotomy [28]. However, it should be borne in mind that in CC the promontory and the round window may not be formed properly or be difficult to identify via standard mastoidectomy with posterior tympanotomy [30]. Transmastoid labyrinthotomy , described by McElveen in 1997 may be good alternative in this group of patients. It allows not only to avoid injury of the facial nerve, the course of which may be abnormal, but also to use many techniques for inserting an electrode into the cavity [30, 5]. Another problem encountered in this malformation, apart from the use of less common surgical access, may be the need for a different implantation technique. Many implantation techniques have been developed to facilitate electrode placement in the CC. Fishman et al. have used fluoroscopy during the procedure [31]. Beltrame et al. described the use of a double labyrinthotomy: performing two cochleostomies and inserting a special, custom-made electrode with inactive tip pulled out by a second cochleostomy [27]. Xia et al., instead of a double labyrinthotomy, proposed access through a single slit and placing a precurved electrode in the cavity, which shortened the time of the procedure [32]. A similar technique is banana cochleostomy [33]. The purpose of all of the above is to prevent a malinsertion of the implant in the internal auditory canal [27, 33].

When implanting in CC, it is preferable to use straight electrodes with full bands due to the unknown location of the neural tissue [33]. The use of a straight array with concentric bands that come into contact with the outer wall of the CC enables the best contact with the nerve fibers and optimal electric stimulation in these unpredictable situations. Examples of such electrodes are: CI24RE(ST) (Cochlear™), Cochlear Limited or MED-EL™ FORM [33, 17]. The optimal length of the electrode is determined by measuring the diameter of the cavity [33]. The use of full-banded straight electrodes and some implantation techniques (Banana, single-slit) may be useful in other cystic malformations of the inner ear, such as cochlear hypoplasia [33].

In the study group, CC was found in one patient. The girl had cochlear hypoplasia type II in the left ear with a hearing loss at the level of 70 dB, and common cavity in the right ear with a hearing loss at the level of 100 dB (Fig. 3.). The child had previously had two unsuccessful attempts of cochlear implantation on the right side via standard mastoidectomy with posterior tympanotomy. The first trial of inserting the electrode through the round window-like structure was unsuccessful, resulting in hypotympanic cell insertion. During the second cochlear implantation, the trial of insertion through the round window-like structure was performed. However, this trial was abandoned due to the presence of a hanging facial nerve and strong stimulation with the risk of facial damage. At the authors' center, the electrode of the cochlear implant was placed via "banana cochleostomy" [33]. Straight electrode CI24RE (ST) (Cochlear™), Cochlear Limited with full bands was used [33]. There were no complications or gusher during the procedure, and auto-NRT was obtained from 21 out of 22 electrodes.

Outcomes of children with CC vary greatly. Some of them may even obtain results comparable to patients without inner ear malformations [27]. That said, overall, the outcomes of CC patients are worse than of those without inner ear malformation [32, 29]. For some patients with CC, the benefits of the cochlear implant and the results of speech and hearing rehabilitation may turn out to be unsatisfactory [1, 27]. In such cases, an auditory brainstem implant may be an effective solution [14]. When the decision is made to apply a cochlear implant first, the surgery should be performed around the age of 1 year [25, 26]. If there is no benefit from the cochlear implant, the decision to perform brainstem implantation should be made before 24 months of age, up to a maximum of 3 years, as delaying this decision is associated with worse expected outcomes [26].

Patients with another major malformation of the inner ear – cochlear hypoplasia – have a different degree of hearing loss, from normal hearing to profound hearing loss [1, 12]. Most have sensorineural hearing loss (79.1%), some mixed hearing loss (18.52%), and a minority are affected by conductive hearing loss (2.47%) [12]. The conductive component is associated with the fixation of the stapes, the footplate of which develops from the otic capsule [12]. Mixed and conductive hearing loss are most common in CH-III and CH-IV [1, 12]. In these patients, stapes surgery, such as stapedotomy, may be an effective treatment [1]. For people with mild to moderate hearing loss, the use of hearing aids may be sufficient. Patients with profound hearing less may be candidates for a cochlear implant [1, 12]. Implantation in the small cavity requires smaller, more delicate electrodes [34]. Examples of the used electrodes include: Nucleus STR24K and MED-EL™ FORM19 [4, 17]. In the case of concomitant hypoplasia or aplasia of the VIII nerve, an auditory brainstem implant is also a possible treatment option [26]. Decisions in such cases are often difficult and should be made case-by-case based on imaging and audiological testing.

All subtypes of cochlear hypoplasia were found in the study group (Fig. 2.–5.):

CH-I in the above-described patient with CADV in the opposite ear (Fig. 2.); CH-II in a girl with CC in the opposite ear with a hearing loss of 70 dB in a hypoplastic ear, (Fig. 3.); bilateral CH-III in one patient with bilateral sensorineural hearing loss (Fig. 4.). In this case, we found: small, sclerotic mastoid (mastoid antrum only), anteriorly displaced sigmoid sinus and a high riding jugular bulb. Surgical access was modified appropriately: the posterior wall of the ear canal was removed, maintaining the continuity of the skin of the canal and the eardrum, then the malleus was removed, which was followed by identification of the facial nerve canal, the stapes and the round window obscured by the high riding jugular bulb [34]. Auto-NRT was obtained from 14 of the 22 electrodes. Bilateral type IV hypoplasia was found in a 6-month-old boy who was qualified for cochlear implantation (Fig. 6.). He is currently using hearing aids and awaiting implantation. One-sided hypoplasia occurred in a girl with an asymmetric malformation and IP-I of the opposite ear which was implanted.

Another major inner ear malformation to be discussed is incomplete partition. In terms of the formation of the cochlea and vestibule with different variants of internal structure anomalies, it is somewhat similar to cochlear hypoplasia – however, theexternal dimensions of the cochlea are normal, unlike in hypoplasia.

Patients with IP are good candidates for a cochlear implant [1]. The most common complication of cochlear implantation is cerebrospinal fluid leak, which occurs in up to 63.6% of procedures [35, 36]. According to Shi et al., cerebrospinal fluid leakage in inner ear malformations occurs most often in: IP-III, IP-I and CC [37]. This leak may be minor and is called oozing, or profuse – the so-called gusher. Several techniques to manage this complication have been described, such as: reverse Trendelenburg position, sealing of the cochleostomy and posterior tympanotomy, placement of a lumbar drain, or even lateral petrosectomy with obliteration of the cavity and the Eustachian tube with fat, with occlusion of the external auditory canal (blind sac procedure) [4, 35, 37]. In addition, some authors use electrodes, such as MED EL™ FORM, which end with a cone-shaped silicone element (CSF SEAL) to prevent cerebrospinal fluid leakage [17]. Others suggest performing a larger cochleostomy that is easier to seal[4].

In IP straight electrodes are useful. as the following electrodes were reported to be successfully used in patients with IP: CI24RE, CI24RE(ST), CI24RST, CI24M, CI24RCS, CI612 (Cochlear™), Cochlear Limited, Clarion HiRes 90 K, MED EL™ FORM24 and FORM19 [35, 38]. For IP-I and IP-III, the electrodes with complete rings may provide better stimulation [4]. In IP-II, the basal part of the modiolus is present, and it is also possible to use a perimodiolar electrode [4]. The outcomes of children with each subtype of this malformation are good and may be comparable with children with a normal inner ear [38, 39, 19].

In the studied group, all types of incomplete partition were found, whereby IP-III was the most common (Fig. 5.). The first patient with an asymmetric defect – IP-I and CH-IV is described above. A CI512 electrode cochlear implant (Cochlear ™), Cochlear Limited was implanted into the ear with IP-I. There was an intense leakage of the cerebrospinal fluid intraoperatively. Auto-NRT was obtained from 11 electrodes, and control X-ray showed incomplete insertion. The child is now 1.5-years-old, uses the implant willingly, and is developing proper hearing and speech.

Bilateral IP-II was found in one boy, who also had intense leakage of cerebrospinal fluid intraoperatively. The course of postoperative treatment was complicated by turbulent vestibular symptoms on the day of surgery, which withdrew within the next 48 hours. Immediately after the procedure, correct results of electrode impedance measurements were obtained, but auto-NRT was not obtained. Bilateral IP-III was found in four patients (Fig. 4.). All of them received a Contour Advance CI512 electrode (Cochlear ™), Cochlear Limited. In two patients, retrofacial approaches were used. All patients developed intense gusher, and two of them required a lumbar drain and underwent observation in the Department of Anesthesiology and Intensive Care for 4 days. In the remaining two patients, the gusher resolved after the cochleostomy and posterior tympanotomy were sealed. A control X-ray of the temporal bone was performed in all patients in a modified Stenvers view and transorbital view, which confirmed the correct position of the electrodes in the cochlea. Two out of four patients had normal electrode impedances and

auto-NRT measurements from most electrodes, in one patient impedance measurements were normal, but auto-NRT was not obtained.

The management of patients with an enlarged vestibular aqueduct depends on the degree of hearing loss. Most patients have bilateral inner ear malformation and progressive bilateral hearing loss [23, 24]. The extent of the hearing loss (from normal hearing to profound hearing loss) and its type (stable, fluctuating, progressive) in this defect are diverse, although some authors believe that there is a relationship between the width of IAC and the degree of hearing loss [23, 40]. Ascha et al. claim that each millimeter increase in vestibular aqueduct size above 1.5 mm was associated with an increase of 17.5 dB in speech reception threshold and a decrease of 21% in word recognition score [23].

In the study group, EVA was found in four patients (Fig. 7A.). Two patients with bilateral defect developed progressive hearing loss – both boys received a cochlear implant at the age of 3 and 4. One boy with a unilateral defect and profound hearing loss in this ear received an implant at the age of 24 months. In another 15-year-old patient with unilateral EVA, before implantation, the remaining hearing was found at the level of: 55, 65, 80 and 80 dB HL respectively for the frequencies: 125, 250, 500 and 1000 Hz, which was confirmed in long-term follow-up (last follow up visit – 2 years after implant placement). None of the patients experienced any difficulties or complications during cochlear implantation. Results of intraoperative measurements: electrode impedances and auto-NRT were normal, and control X-ray of the temporal bone confirmed the correct location of the electrode. Moreover, in the study group, cochlear aperture abnormalities were found in five ears (Fig. 7B., C.). One patient had bilateral aplasia, another one patient had bilateral hypoplasia, and one child had unilateral hypoplasia. They received cochlear implants, which they currently use.

Diagnostic imaging is an extremely important element of qualification for a auditory implant[34]. At the authors' centers, two imaging studies are performed each time – HRCT of the temporal bones and MRI [34]. HRCT and MRI are complementary to each other for preoperative imaging. HRCT of temporal bones is superior to MRI in the evaluation of bony structures. In turn, MRI demonstrates higher applicability in the assessment of neural structures and labyrinthine fluid spaces; it also allows the assessment of the central nervous system and the auditory pathway, including the cochlear nuclei [34]. In some inner ear malformations in which the HRCT of the temporal bones shows an implantable space, there may be no communication of these spaces with the nerve – in such cases, the cochlear implant will not be effective. Thus, MRI is essential to evaluate the cochlear nerve, which may be aplastic or hypoplastic, or may not communicate with the implantable space. The two modalities in combination allow accurate and optimal evaluation of the anatomical structures of the temporal bone prior to implantation and assessment of whether the basic conditions necessary for the implantation of an auditory implant have been met [7, 34].

Accurate clinical and radiological evaluation of the patient with both imaging modalities is particularly important in patients with inner ear malformations. In the case of some malformations – defined by the international consensus on auditory brainstem implantation in children and non-neurofibromatosis type 2 patients as "possible congenital indications" (cochlear hypoplasia, CC, IP-I, hypoplasia or aplasia of the cochlear nerve) – it will allow to make a decision about qualification or disqualification from a cochlear or auditory brainstem implant [26].

CONCLUSIONS

Inner ear malformations are an extremely significant diagnostic and clinical challenge in children qualified for cochlear implantation. In the vast majority of cases, cochlear implantation is possible, but patients with a malformation require a thorough preoperative assessment and planning of the procedure, as well as an appropriate selection of electrodes. Severe malformations may prevent or significantly hinder cochlear implantation and postoperative care. Inner ear malformations are found in imaging studies in 20% to 30% of patients with profound hearing loss.

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