

The role of preoperative imaging for auditory implants in children

Authors' Contribution:

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G – Funds Collection

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ABSTRACT:

Introduction: Preoperative imaging, besides audiological evaluation, plays a major role in evaluation of candidacy for auditory implants, and in particular cochlear implants. It is essential to assess whether the basic criteria necessary for implantation are met. Diagnostic imaging is crucial not only in determining candidacy, but also determining the feasibility of cochlear implantation as it allow to anticipate surgical difficulties which could preclude or complicate the implantation of the device. The aim of the study is to present the protocol for the evaluation of preoperative imaging studies with particular focus on the factors potentially affecting clinical decisions in children qualified for cochlear implantation.

Material and method: Preoperative imaging studies of 111 children performed prior to cochlear implantation were analyzed: high-resolution computed tomography (HRCT) of temporal bones and MRI. The assessment was made according to the presented protocol.

Results: Pathologies and anomalies identified during the assessment of preoperative imaging studies significantly altered clinical decisions in 30% of patients. In the study group, in 17% of patients inner ear malformations were identified. 2.7% of children were disqualified from a cochlear implantation due to severe congenital inner ear malformations. 9% of the patients have had bacterial meningitis. In 50% of them difficulties related to complete or progressive cochlear ossification occurred. In 4.5% of patients less common surgical approaches other than mastoidectomy with a posterior tympanotomy were applied.

Discussion: Preoperative imaging allow for the identification of significant pathologies and anomalies affecting qualification decisions and further treatment. HRCT and MRI are complementary to each other for preoperative imaging. The two modalities in combination allow accurate and optimal evaluation of the anatomical structures prior to implantation. Inner ear malformations and cochlear ossification following meningitis are relatively frequently encountered in children qualified for a cochlear implant.

KEYWORDS:

auditory brainstem implant, cochlear implant, cochlear ossification, imaging studies, inner ear malformations

ABBREVIATIONS

ABI – Auditory Brainstem Implant
CC – Common cavity
CH – Cochlear hypoplasia
EVA – Enlarged vestibular aqueduct
HRCT – High-resolution computer tomography
IAC – internal auditory canal
IP – Incomplete partition
MRI – Magnetic Resonance Imaging

INTRODUCTION

The contemporary qualification of patients for auditory implants, and in particular cochlear implants, after establishing audiological criteria, is associated with preoperative imaging [1–4].

Diagnostic imaging is essential in assessing the feasibility of cochlear implantation, it allow to answer the question whether the basic criteria necessary for implantation are met: the presence of an implantable space to which an electrode can be inserted, and whether there is a cochlear nerve that will allow transmission of the impulses to further levels of the auditory pathway [1]. Besides answering a series of questions regarding the diagnosis and implant candidacy, preoperative imaging help determine additional factors that could complicate surgery or the postoperative management, or preclude or complicate the implantation [1–3].

One of the major reasons possibly complicating cochlear implantation is obliteration of the labyrinth fluid spaces. The most common cause of luminal obstruction, especially in the pediatric population, is postmeningitic labyrinthitis ossificans [1]. Deafness is complication of meningitis in 2–11% of children [1].

The best method of assessing the ossification of the labyrinth is MRI in the heavily T2-weighted sequence [1, 3].

Congenital inner ear malformations are another relatively frequent problem constraining the possibility of implantation.

In some cases, there is no implantable space, or there is no auditory nerve, therefore some inner ear malformations are a contraindication to cochlear implantation [5, 6]. Thus, in some children with inner ear malformations, cochlear implant will not be effective or cochlear implantation possible, and the auditory brainstem implant will be the only method of treatment. The international consensus on auditory brainstem implantation in children and non-neurofibromatosis type 2 patients from 2011 distinguished 3 groups of radiological indications for a brainstem implantation. It classified 4 inner ear malformations as “well-defined congenital indications”, and included several malformations in the group of “possible congenital indications” [6]. In children with severe inner ear malformations a particularly detailed and individual radiological and clinical assessment of the patient is required in order to ensure the best possible treatment and rehabilitation options for the child.

Inner ear malformations are present in 20–30% of patients with profound hearing loss [7]. Many of them are patients with severe or profound bilateral hearing loss who qualify for cochlear implantation. In numerous such cases, cochlear implantation is possible, but can often be very challenging due to considerable surgical difficulties, such as: 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) and other difficult to predict situations, as well as the need to use various surgical approaches [5, 1, 8]. While it is easy to identify obvious inner ear malformations such as cochlear aplasia, many inner ear malformations may be overlooked or misclassified by visual assessment of computed tomography [9, 10]. For all these reasons, a precise assessment of the inner ear in imaging studies is essential.

In addition to the above-mentioned challenges, surgery may be hampered by numerous other congenital and acquired pathologies and anomalies, such as unfavorable anatomical variants, e.g. an abnormal course of the facial nerve or a protruding sigmoid sinus [1, 11].

Protocols for the evaluation of preoperative imaging studies vary by center and may include the following complementary tests: HRCT of temporal bones and/or MRI. In our center, children always undergo HRCT of temporal bones and MRI of temporal bones, which may often need to be performed in pharmacological sleep due to the limited cooperation of children during examination.

The aim of the study is to present the protocol for the evaluation of preoperative imaging studies used in our center and to present the results of evaluation, with particular focus on the factors potentially affecting clinical decisions in children qualified for cochlear implantation.

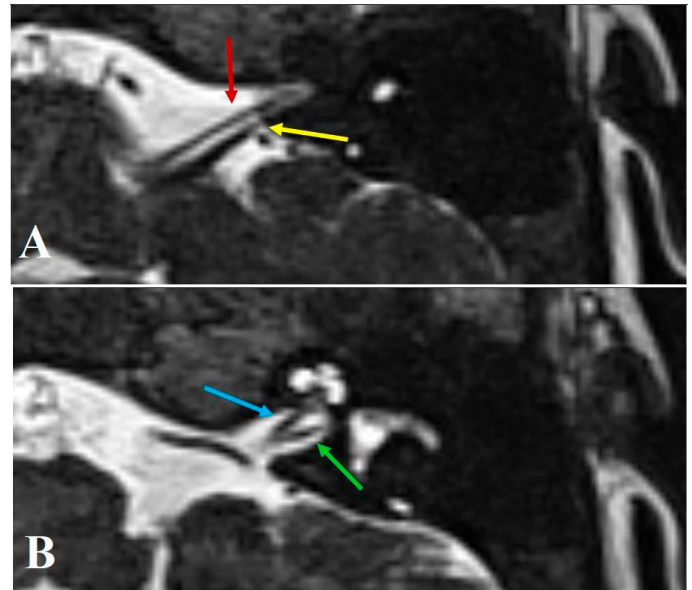


Fig. 1. Axial plane, nerves visible in T2 weighted sequence. (A) facial (red arrow) and superior vestibular (yellow arrow); (B) cochlear (blue arrow) and inferior vestibular (green arrow).

MATERIAL AND METHODS

The study was conducted in accordance with the ethical requirements for human studies and the Declaration of Helsinki. Bioethics Committee approval number: KB62/2018.

Medical records of patients hospitalized from 01/01/2010 to 01/06/2020 in one cochlear implant center, who were qualified or underwent cochlear implantation were retrospectively reviewed. In all patients preoperative HRCT and MRI of temporal bones were performed before cochlear implantation as a part of qualification process. Patients with no available imaging studies were excluded from the study group.

Preoperative imaging studies: HRCT of temporal bones and MRI were analyzed using RadiAnt DICOM Viewer software. 111 children – 41 girls and 70 boys were included in the study group. Age at examination ranged from 4 months to 17 years.

The protocol for the evaluation of preoperative imaging studies included:

Assessment of HRCT of temporal bones:

1. Pneumatization of the mastoid, the position of the sigmoid sinus and dura of the middle cranial fossa; mastoid air cell opacification; mastoid pneumatization has been classified as:
 - ◆ **sclerotic** – no pneumatization, non-pneumatized areas are filled with compact bone,
 - ◆ **diploic** – partial pneumatization, areas devoid of pneumatization are filled with bone marrow,
 - ◆ **pneumatic mastoid** – air cells are large and numerous;
2. Cortical thickness of the mastoid;
3. Bony intra-temporal courses of the facial nerve, superior and inferior vestibular nerves, cochlear nerve;

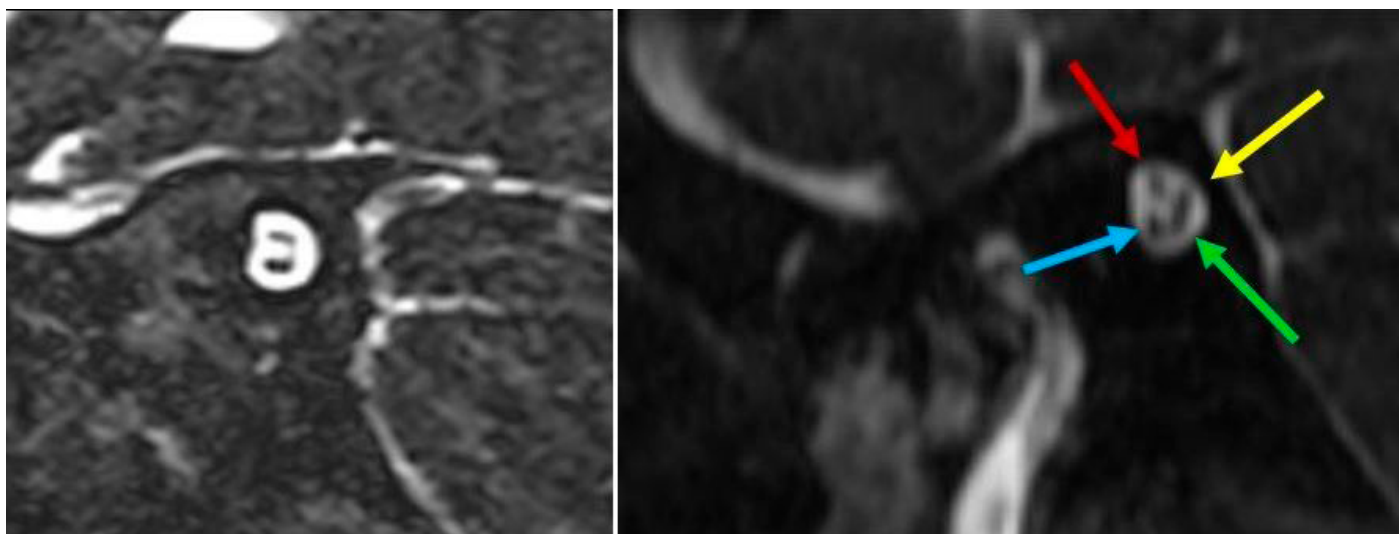


Fig. 2. Sagittal cross-section: facial, cochlear, superior and inferior vestibular nerves visible in the T2 weighted sequence (see Fig. 1.).

4. The facial nerve course through the temporal bone: in the meatal, labyrinthine, tympanic and mastoid segment – is there any anomaly, is the nerve dislocated, exposed, whether there is a risk of damage during the surgery [12];
5. Assessment of vascular structures: carotid artery (whether or not it is exposed), sigmoid sinus (whether it is protruding), jugular bulb (whether it is high-riding or exposed);
6. Bony labyrinth to exclude pathologies such as Paget's disease, otosclerosis, post-inflammatory narrowing of the round window niche;
7. Inner ear:
 - The cochlea, including: general outline of the cochlea, number of turns, internal architecture: presence of a modiolus and spiral lamina, possible obliteration of the cochlear fluid spaces;
 - Round and oval window. The structure of the round window (whether it is present, non-narrowed, whether there is good access to the window);
 - Vestibule: general outline, general shape of the semicircular canals, presence of fistulas;
 - Internal auditory canal (IAC) – it is considered narrow when its midpoint diameter is <2 mm, which can be accompanied by a hypoplastic or aplastic cochlear nerve [12];
 - Bony cochlear nerve canal (cochlear aperture) – when its width in axial scanning showing the modiolus and the internal auditory canal is less than 1.4mm, it is considered hypoplastic [5]. It may also be aplastic when there is solid bone between the cochlea and the IAC [5];
 - The vestibular aqueduct – in terms of broadening – the aqueduct is considered enlarged if its width halfway along its length in the axial scan exceeds 1.5 mm [5];
8. Tympanic cavity – the presence of inflammatory changes, cholesteatoma;
9. The presence of additional pathologies;
10. **MRI of the head** – MRI is superior to CT in the evaluation of soft tissues, allows for an accurate assessment of the brain, auditory pathway, membranous labyrinth and neural structures of the temporal bone [1, 3, 4]. T2- weighted sequence enables the assessment of the labyrinth fluid spaces, which is particularly important in terms of assessing luminal obstruction.

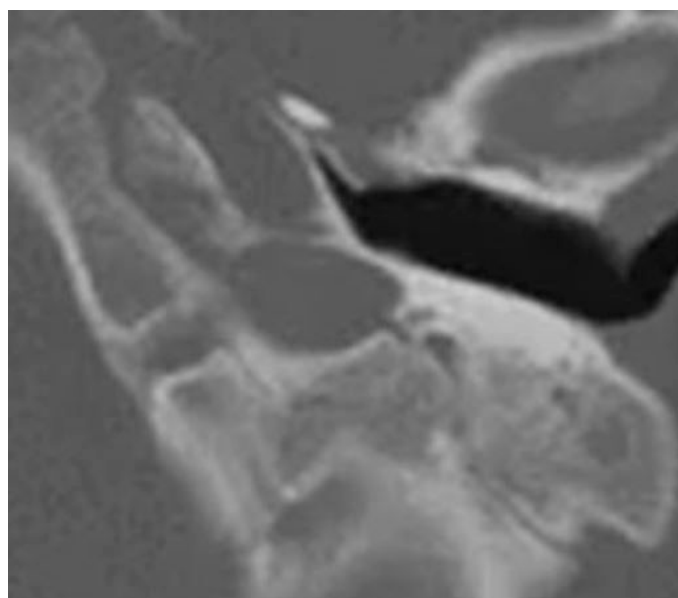


Fig. 3. Complete labyrinthine aplasia.

11. Assessment in the T2 weighted sequence in axial and sagittal planes enables the evaluation of VII and VIII nerves (Figs. 1. and 2.).

Preoperative imaging MRI of temporal bones was performed in the T2-weighted FSE and CISS, 3DT1VIBE, DWI HASTE sequences in frontal and transverse planes, and additionally T2 weighted sagittal sequence and T2, FLAIR of the brain in transverse planes.

According to the protocol, the following elements were assessed:

- general development of: cochlea, vestibule and semicircular canals,
- image of the labyrinth fluid spaces,
- spiral lamina and modiolus,
- symmetry of the scala tympani and scala vestibuli – internal auditory canals their width,

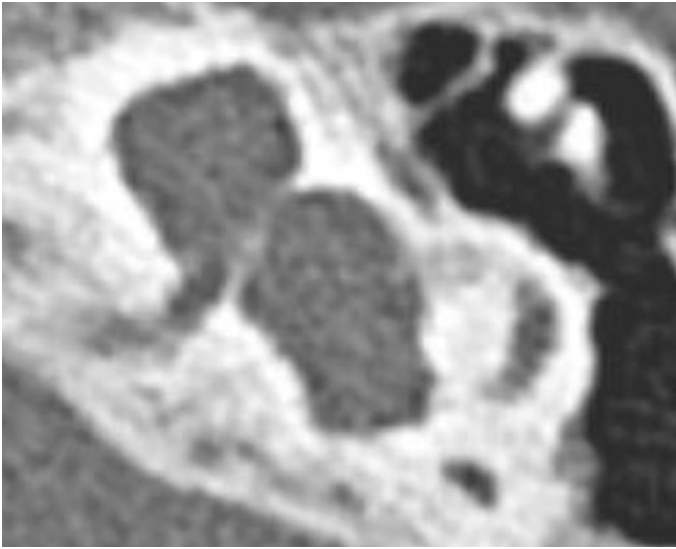


Fig. 4. Cochlear incomplete partition.

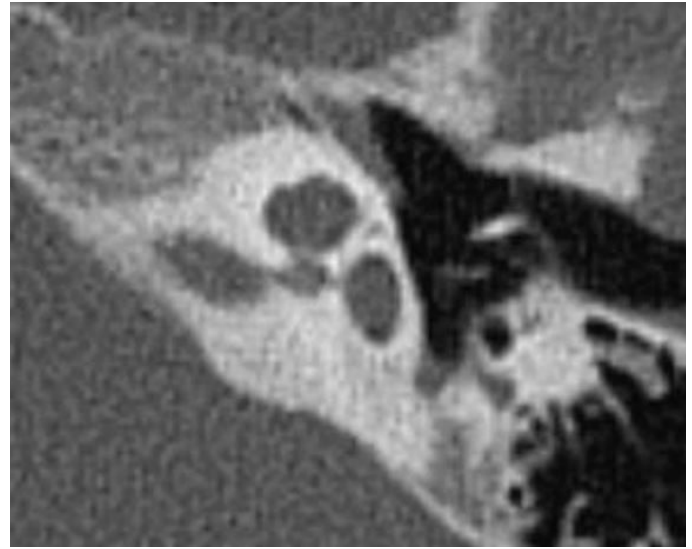


Fig. 5. Cochlear hypoplasia.

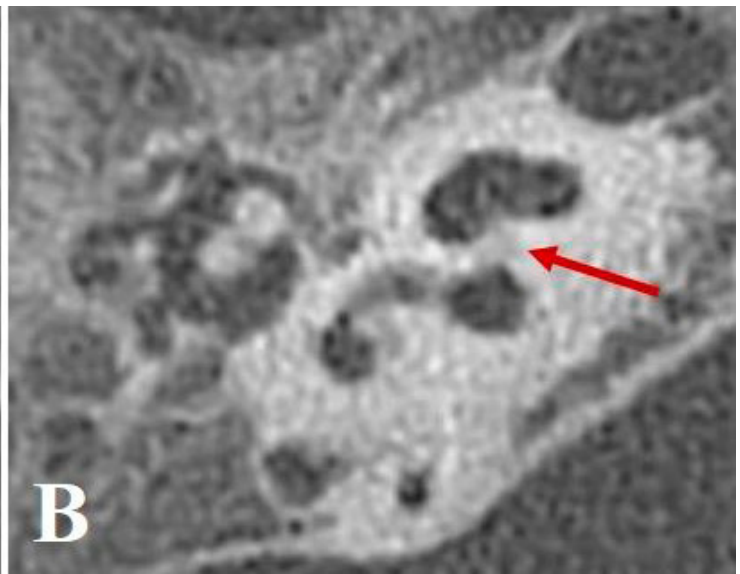
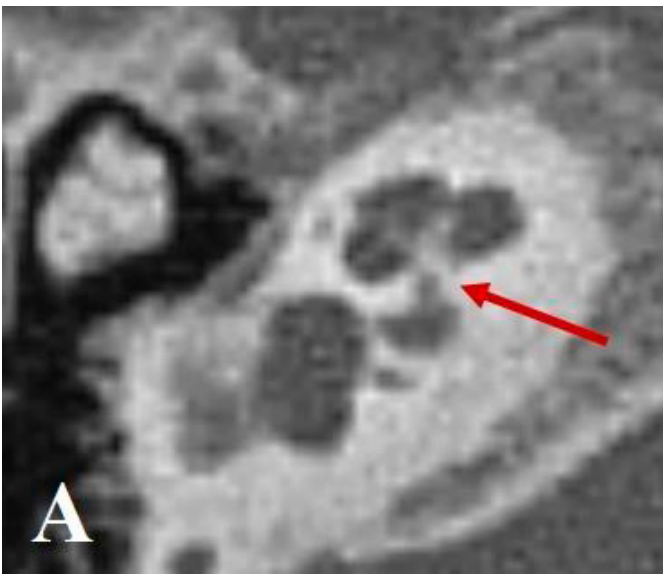


Fig. 6. Hypoplasia (A) and aplasia (B) of the nerve entry into the cochlea.

- VII and VIII nerves in the cerebellopontine angle and IAC, with particular focus on cochlear nerves,
- pyramids of temporal bones,
- location of the sigmoid sinuses and jugular bulbs,
- screening of brain.

The identified inner ear malformations are classified according to the Sennaroğlu classification (2017) [5].

Inner ear anomalies are classified into eight groups as follows:

1. Complete labyrinthine aplasia (Michel deformity),
2. Rudimentary otocyst,
3. Cochlear aplasia,
4. Common cavity (CC),
5. Cochlear hypoplasia (CH), including 4 subtypes: CH-I to CH-IV,
6. Incomplete partition of the cochlea (IP), including 3 subtypes: IP-I-III,
7. Enlarged vestibular aqueduct (EVA),
8. Cochlear aperture abnormalities.

RESULTS

Inner ear malformations

In the analyzed group, major inner ear malformations according to Sennaroğlu classification were found in 19 (17%) patients in 35 ears – in 2 patients malformations were unilateral, 1 patient had a cochlear implant in the contralateral ear. 7 out of 8 inner ear malformations described in this classification were found. The only inner ear malformation that was not present in analyzed group was the rudimentary otocyst. Identified malformations included:

- 2 complete labyrinthine aplasias,
- 1 cochlear aplasia,
- 1 common cavity,
- Cochlear hypoplasia – all subtypes:
 - 1 Cochlear hypoplasia type I (CH-I),
 - 1 Cochlear hypoplasia type II (CH-II),
 - 4 Cochlear hypoplasias type III (CH-III),
 - 3 Cochlear hypoplasias type IV (CH-IV),

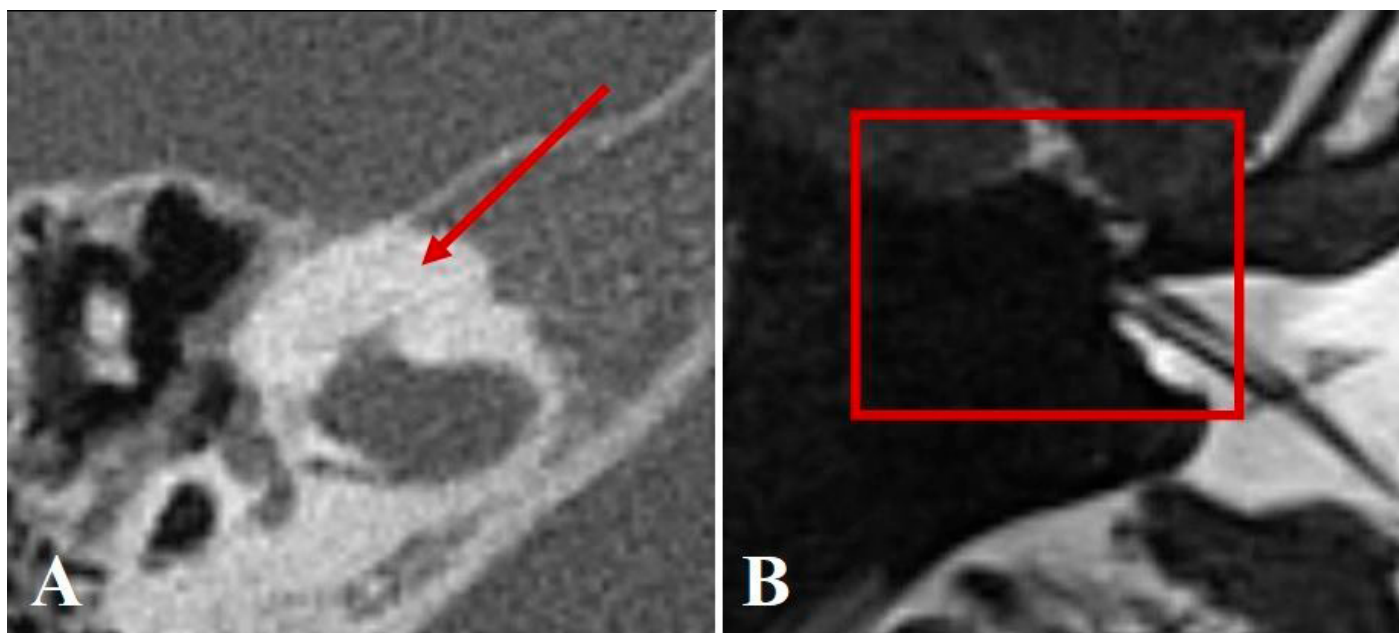


Fig. 7. Complete ossification of the cochlea in the course of meningitis – CT (A) and MRI (B).

- ♦ Incomplete partition – all subtypes:
 - 1 Incomplete partition type I (IP-I),
 - 2 Incomplete partition type II (IP-II),
 - 8 Incomplete partition type III (IP-III),
- ♦ 6 Enlarged vestibular aqueducts,
- ♦ 5 Cochlear aperture abnormalities – including 3 hypoplasias and 2 aplasias. Additionally, one patient with bilateral cochlear hypoplasia type III (CH-III) had bilateral aplasia of the bony cochlear nerve canal.

The figures below show the identified inner ear malformations (Figs. 3.–6.).

OBLITERATION OF THE LABYRINTH FLUID SPACES

In the analyzed group of patients, 10 (9%) had bacterial meningitis, and 7 (6.3%) of them qualified for cochlear implantation. 1 (0.9%) patient with unilateral complete labyrinthine ossification was not qualified for cochlear implantation due to the lack of any changes in the contralateral ear and normal hearing, and was subjected to further regular follow-up. Two (1.8%) children presented too late with complete ossification of the cochlea (Fig. 7A., B.).

Eighteen children (16%) had: unilateral/bilateral secretory otitis media (14 children – 12.6%), ventilation tubes (2 children – 1.8%), cholesteatoma (2 children – 1.8%).

CHOLESTEATOMA

Was found in 2 (1.8%) patients: In one girl (0.9%) congenital cholesteatoma was found during sequential second side cochlear implantation (Fig. 8.).

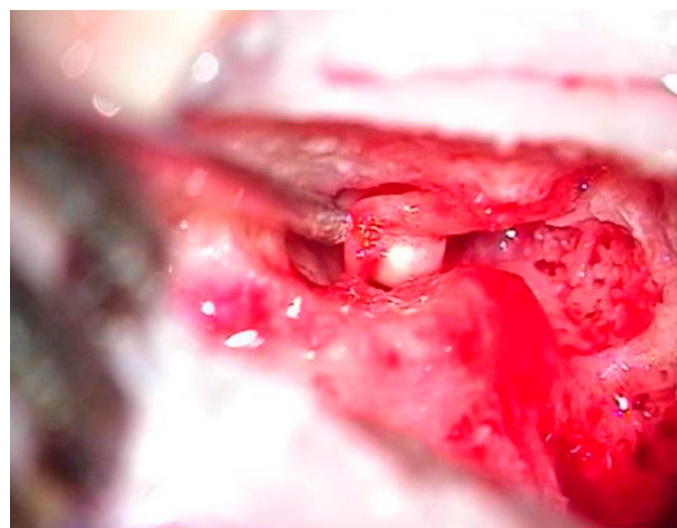


Fig. 8. Congenital cholesteatoma identified intraoperatively during placement of a cochlear implant.

In 1 (0.9%) patient with bilateral profound hearing loss operated due to cholesteatoma, because of confirmed recurrence of cholesteatoma and planned cochlear implantation, lateral petrosotomy was performed, cholesteatoma was removed and the external auditory canal was closed (blind sac procedure).

OTITIS MEDIA

Fluid or inflammatory changes in the middle ear were found in 14 (12.6%) patients during the surgery or in HRCT. In 6 (5.4%) children, the changes were unilateral, including 4 (3.6%) children in the implanted ear, and in 2 (1.8%) in the non-implanted ear. In 5 (4.5%) children the changes were bilateral. In 2 children (1.8%), no fluid or inflammatory changes were found intraoperatively. Moreover, 3 patients had inflammatory changes detected intraoperatively, which were not identified in imaging studies.

Tab. I. The impact of pathologies identified in imaging studies on clinical decisions.

IDENTIFIED PATHOLOGY	NUMBER OF PATIENTS	MANAGEMENT
INNER EAR DEFECTS – 19 (17%) patients		
1. Congenital defects of the inner ear which are a contraindication to the implant – 3 patients (2.7%)		
Complete bilateral aplasia of the labyrinths (8-year-old child)	1 (0,9%)	Disqualification from placement of a cochlear implant
Cochlear aplasia with dilated vestibule of the left ear and cochlear hypoplasia of the right ear, no communication of any of those spaces with the nerve (12-month-old child)	1 (0,9%)	Disqualification from placement of a cochlear implant Qualified for a brainstem implant – awaiting implantation
Cochlear hypoplasia with aplasia of the cochlear aperture (8-year-old child)	1 (0,9%)	Disqualification from placement of a cochlear implant
2. Congenital defects of the inner ear, in which implantation is possible – 16 patients (14.4%)		
Common cavity – right ear, CH-II cochlear hypoplasia, left ear, spaces communicate with the nerve, condition after attempt of double implantation in the ear with a common cavity	1 (0,9%)	Modification of surgical access – “Banana” cochleostomy – via the mastoid Selection of electrode: CI24RE(ST) (Cochlear™), Cochlear Limited with full rings
Bilateral cochlear hypoplasia CH-III, residual mastoid, protruding and lateralizing sigmoid sinus, and a highly placed jugular bulb	1 (0,9%)	Removal of the posterior wall with preservation of the duct, skin and eardrum which was moved aside with a downward shift of the jugular bulb covering the round window
Bilateral cochlear hypoplasia CH-IV	1 (0,9%)	A 4-month-old child was qualified for placement of a cochlear implant during imaging tests, awaiting implantation. Planned use of a shorter electrode
CH-IV cochlear hypoplasia and incomplete partition IP-I	1 (0,9%)	Qualified for placement of a cochlear implant into the ear with IP-I Selection of electrode: CI512 (Cochlear™), Cochlear Limited
Bilateral incomplete partition IP-II	1 (0,9%)	Qualified for placement of a cochlear implant Selection of electrode: CI512 (Cochlear™), Cochlear Limited
Bilateral incomplete partition IP-III	2 (1,8%)	Qualified for placement of a cochlear implant Selection of electrode: CI512 (Cochlear™), Cochlear Limited
Bilateral incomplete partition IP-III, anteriorly shifted facial nerve	2 (1,8%)	Modification of surgical access – retrofacial approach Selection of electrode: CI512 (Cochlear™), Cochlear Limited
Enlarged vestibular aqueduct	4 (3,6%)	Qualified for placement of a cochlear implant
Defects of the bony cochlear nerve canal	3 (2,7%)	Qualified for placement of a cochlear implant
OBLITERATION OF THE LABYRINTH FLUID SPACES – 10 patients (9%)		
Complete cochlear ossification found in CT and MRI in T2 weighted sequence	2 (1,8%)	Disqualification from placement of a cochlear implant
Correct image of the labyrinthine fluid spaces, no cochlear obliteration was identified in imaging studies	1 (0,9%)	The patient was qualified for cochlear implantation, the spaces of the inner ear were intraoperatively filled with connective tissue, which made insertion of the electrode impossible. Cochlear implantation was abandoned.
Correct image of the labyrinthine fluid spaces, no cochlear obliteration was identified in imaging studies	1 (0,9%)	During placement of the implant into the left ear, we found extensive inflammatory changes in the spaces of the middle ear – insertion was postponed by 4 weeks. Follow-up MRI revealed obliteration of the inner ear on the previously operated side and the implant was placed in the right ear.
Unilateral complete cochlear ossification, correct hearing of the opposite ear	1 (0,9%)	Disqualification from placement of a cochlear implant.
The imaging studies revealed partial post-inflammatory changes – segmental obliteration of the signal of fluid space	2 (1,8%)	The patients were qualified for cochlear implantation. Before inserting the correct electrode into the cochlea, we used the depth gauge Contour Advance (Cochlear™), Cochlear Limited Selection of electrode: CI512 (Cochlear™), Cochlear Limited
Correct image of the labyrinthine fluid spaces, no cochlear obliteration was identified in imaging studies	3 (2,7%)	The patients were qualified for cochlear implantation Selection of electrode: CI512 (Cochlear™), Cochlear Limited
INFLAMMATORY CHANGES – 2 patients (1.8%)		
Congenital cholesteatoma found intraoperatively during second implant placement in sequential mode	1 (0,9%)	The lesion was completely removed and a cochlear implant was placed
Bilateral profound sensorineural hearing loss, status following right ear surgery due to cholesteatoma, recurrence of cholesteatoma was diagnosed	1 (0,9%)	Due to otorrhea and the presence of cholesteatoma, a lateral petrosectomy was performed, the cholesteatoma was removed and the external auditory canal was closed (blind sac procedure). After 12 months, a cochlear implant was inserted into the operated ear.
OTHER CASES – 2 patients (1.8%)		
The presence of a ventriculoperitoneal shunt	1 (0,9%)	Modification of implant location – the implant was placed vertically above the auricle to bypass the valvular system
Status after BAHA implantation	1 (0,9%)	Prior to cochlear implantation, the BAHA implant bracket was removed, modification of the location of cochlear implant

Additionally, 2 children (1.8%) had ventilation tubes: one of the patients had bilateral ventilation tubes placed 5 months before implantation, and one boy had a ventilation tube in the implanted ear.

OTHER CASES

In 1 (0.9%) patient, due to the presence of a ventriculoperitoneal shunt, the implant was placed vertically above the auricle to bypass the valve system.

In 1 (0.9%) patient with bilateral sensorineural hearing loss, acquired on the right and congenital on the left, a cochlear implant was implanted in the right ear, in which a BAHA implant had previously been implanted, and the titanium implant was removed prior to cochlear implantation.

Pathologies and anomalies identified during the assessment of preoperative imaging studies that significantly altered clinical decisions are presented in Tab. I.

DISCUSSION

Preoperative imaging is essential in evaluation of candidacy for auditory implants and preparation for the surgery [1, 3–5]. Diagnostic imaging allow not only to make a decision about cochlear or brainstem implantation, but also play a role in choosing surgical approach, operated ear, cochlear implant electrode, as well as help predict numerous surgical challenges [1, 3–5].

QUALIFICATION FOR COCHLEAR IMPLANTATION

In some patients, cochlear implantation may not be possible or cochlear implant may not be effective [5, 6, 13]. The auditory brainstem implant (ABI) is the only option for hearing rehabilitation in this group of patients, but only a few children will qualify for such treatment. In 2015, less than 200 children have received ABI worldwide [14].

Children with inner ear malformations are the first group of patients in which diagnostic imaging will allow to decide on the type of auditory implant. Current indications for ABI include two groups of pathologies identified in preoperative imaging [6, 15]:

1. It is not possible to insert the implant electrode in implantable cavity in proximity to stimuable neural elements;
2. There is no neural connection between the inner ear and further levels of the auditory pathway.

According to the international consensus on ABI in children 4 inner ear malformations: complete labyrinthine aplasia, cochlear aplasia, cochlear nerve aplasia and cochlear aperture aplasia are classified as “well-defined congenital indications” for ABI [6]. Other inner ear malformations, including cochlear hypoplasia with cochlear aperture hypoplasia, common cavity and incomplete partition type I with or without cochlear nerve, as well as unbranched

cochleovestibular nerve and hypoplastic cochlear nerve (defined as a nerve that is 50% of the size of a normal nerve or which the size is less than the contralateral normal cochlear nerve or ipsilateral normal facial nerve) may be a potential indication for an ABI and should be considered on a case by case basis [6]. Each of these patients should undergo detailed radiological and audiological evaluation. What is more, in some cases of inner ear malformations, children may become candidates for ABI after trial with cochlear implant. In such cases children who underwent cochlear implantation should be followed up for several months in terms of development of behavioral and objective auditory performances. When there is no sufficient progress with a CI, a decision to go ahead with an ABI should be done as early as possible [6, 13].

Three patients (2.7%) had severe inner ear malformations which changed decisions about cochlear implantation:

1. 8-year-old child with bilateral complete labyrinthine aplasia,
2. 8-year-old child with congenital bilateral cochlear hypoplasia type III (CH-III) with cochlear aperture aplasia.

None of these children qualified for a cochlear implant.

3. in a child with cochlear aplasia with a dilated vestibule in the left ear and cochlear hypoplasia type I in the right ear. In preoperative diagnostic imaging, there was no communication of these spaces with the cochlear nerve. During behavioral audiological examinations no responses were observed. The child was qualified for ABI and is awaiting surgery.

Another important problem that can be encountered during radiologic evaluation of the cochlear implant candidate is obliteration of the labyrinth fluid spaces. Inflammatory processes (such as chronic otitis media, meningitis, rubella, mumps, other viral infections), abnormalities of bone metabolism (Paget’s disease, otosclerosis), other diseases (granulomatosis with polyangiitis, Cogan’s syndrome) or trauma, may ultimately result in luminal obstruction either by ingrowth of fibrous scar tissue or pathologic neoossification [1]. The origin most commonly encountered in pediatric cochlear implant candidates is postmeningitic labyrinthitis ossificans [1]. The cochlear aqueduct is believed to be the main route of spread of the inflammatory process into the inner ear in cases of meningitis [1, 3]. Some degree of cochlear neoossification may be encountered intraoperatively in as many as 80% of patients deafened by meningitis [1, 16, 17]. Cochlear ossification is nearly always most severe in the region of the round window and proximal scala tympani in the basal turn, middle and apical turns are less commonly affected and the scala vestibuli is often spared [1]. The best method of assessing the ossification of the labyrinth is MRI in the T2 weighted sequence [1, 3].

In our center, children with profound hearing loss with early signs of cochlear obliteration after meningitis are qualified for urgent cochlear implantation. According to the consensus on auditory brainstem implantation in children from 2011 and 2016, if the cochlea seems to be patent on T2 MRI (MRI show visible fluid spaces), cochlear implant should be the first treatment option [13, 6]. However, when complete ossification with white cochleas on computed tomography with absolute no cochlear duct signal on MRI is

found the chance of an successful cochlear implantation is limited and an ABI may be a preferred option in this situation [13, 6].

9% of the patients have had bacterial meningitis. In 50% of them labyrinthine ossification following meningitis occurred.

In the study group, 10 (9%) patients had bacterial meningitis, and 7 out of 10 qualified for cochlear implantation. Additional difficulties occurred in 2 out of 7 children qualified for cochlear implantation. In 1 of these 2 children, preoperative imaging did not show cochlear ossification. An unsuccessful cochlear implantation was attempted with the use of Cochlear™ Contour Advance Depth Gauge. Intraoperatively, ossification was not yet found, but both the scala tympani and the scala vestibuli were filled with connective tissue, which made Depth Gauge insertion impossible. Electrode insertion was not attempted. Second child was qualified for cochlear implantation 2 months after the bacterial meningitis. During the cochlear implantation, extensive inflammatory changes within the left middle ear were found. A mastoidectomy with posterior tympanotomy was performed and inflamed tissues were removed. Due to active inflammatory process of the middle ear cochlear implantation was postponed by 4 weeks. MRI performed before the implantation revealed obliteration of the inner ear on the previously operated side and contralateral ear was implanted.

5 out of 7 children who qualified for cochlear implantation received a cochlear implant without any additional complications.

In 3 out of 10 children decision not proceed with cochlear implantation was taken. 1 girl had unilateral complete obliteration of the cochlea and good hearing in the opposite ear. 2 children presented too late with complete cochlear ossification. One of them had pneumococcal meningitis around the age of 1. At the age of 3.5 years, he presented to our center with a bilateral hearing loss of 50/60 dB and was fitted with hearing aids. Within 6 months, the hearing loss progressed. Imaging studies were performed and complete ossification of the cochleas was diagnosed. The child was disqualified from a cochlear implantation. The decision to implant a child who does not meet audiological criteria and who can hear relatively well in hearing aids is not easy, but just like in this situation, the delayed implantation led to complete cochlear ossification and bilateral deafness with no chance for successful placement of cochlear implant.

In 10 children who had bacterial meningitis, only 5 did not experience additional difficulties. 5 out of 10 children had a complete or progressive cochlear ossification.

ASSESSMENT OF ANATOMICAL ANOMALIES AND FACTORS THAT CAN COMPLICATE THE SURGERY

In addition to assessing the feasibility of cochlear implantation, preoperative imaging is necessary to assess pathologies that could complicate or increase the degree of difficulty of the surgery, such as unfavorable anatomical variants or anomalies or inflammatory changes [1, 11]. The most common unfavorable anatomical variants include: poor pneumatization of the mastoid,

protruding sigmoid sinus, a high-riding jugular bulb, displaced carotid artery, lowered middle cranial fossa, exposed facial nerve or facial nerve with an abnormal course, absence of/hypoplasia of the round window [1, 3, 4, 11, 18].

Both the anteriorly displaced sigmoid sinus (found in 1.6% of patients), the lowered dura of middle cranial fossa and the sclerotic, poorly developed mastoid may hinder surgical access and posterior tympanotomy [1, 3]. In the study group, anteriorly displaced sigmoid sinus was found in 5 (4.5%) patients. A high-riding jugular bulb may be present in 6% of the general population [1]; in the study group, it was found in 7 patients (6.3%). There are a number of criteria for diagnosis: if the superior margin of the jugular bulb lies at or above the level of the floor of the ipsilateral IAC, or if it lies above the level of the basal turn of the cochlea, it is considered as a “high-riding jugular bulb” [4]. There is a risk of damaging the high-riding jugular bulb during the surgery and massive hemorrhage [1]. Very rarely, a jugular bulb may overlie the round window niche or promontory or be exposed [1]. In the described group of patients, this happened in 1 (0.9%) patient – during the surgery the bulb was exposed and shifted to obtain a good access to the round window.

Internal carotid artery – assessment include the horizontal and vertical segment of the artery in the temporal bone to exclude its abnormal course or defects in the bony wall of artery. The aberrant carotid artery enters the posterior portion of the middle ear through the enlarged inferior tympanic canaliculus, continues forward along the promontory, and enters the horizontal portion of the carotid canal.

Abnormal course of the facial nerve – an exposed or anteriorly displaced facial nerve is associated with a higher risk of damage [3]. An abnormal course of the facial nerve is more common in patients with inner ear malformations [1, 5, 12]. The conventional technique for cochlear implantation is the mastoidectomy with posterior tympanotomy approach. Posterior tympanotomy is the route to perform cochlear implantation via facial recess. The major boundaries of the posterior tympanotomy are the facial nerve (posteriorly), chorda tympani (anteriorly) and fossa incudis (superiorly). In cases when the facial nerve is severely anteriorly displaced it may be difficult to perform a posterior tympanotomy broad enough to obtain good visualization of the round window through the facial recess. Therefore, possibility to visualize round window via facial recess approach should be assessed. The round window is not visible through the facial recess in 7 to 24% of patients during cochlear implantation due to anatomical variations of the facial nerve and cochlear anomalies [18]. Many studies have demonstrated the usefulness and correlation of HRCT of temporal bones with the possibility of visualizing the round window [18].

Narrow internal auditory canal –when the IAC is narrow hypoplastic or aplastic cochlear nerve should be suspected [3, 5].

It should be mentioned that anatomical anomalies and variants are more common among patients with inner ear malformations [5]. In such cases, a detailed evaluation of imaging studies is essential.

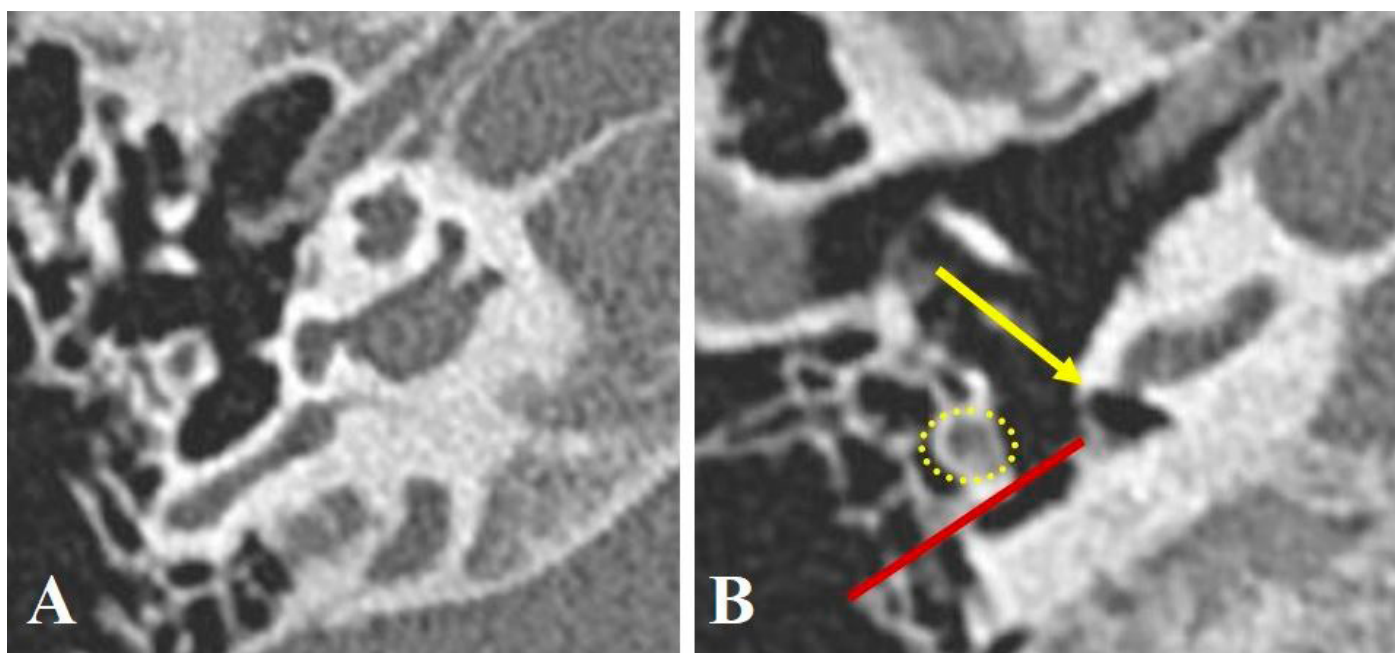


Fig. 9A., B. Patient with IP-III congenital defect, the implant was placed retrofacially (facial nerve—yellow outline, round window—yellow arrow).

Cochlear implantation in patients with inflammatory processes of the middle ear, especially chronic otitis media can present a significant challenge to the otologic surgeon. The incidence of chronic otitis media in patients receiving CI is relatively rare, and ranges from 2.2 to 10.9% [19]. Preoperative imaging in this group of patients allow to assess extend of the disease and may provide additional information about the possible luminal obstruction. Initially, cochlear implantation was contraindicated in patients with otitis media because of the risk of further complications [20].

Currently, cochlear implantation in patients with chronic otitis media, especially in bilateral otitis media or if the patient has one-sided otitis media but the other ear is unsuitable for device implantation may be a relatively effective method of treatment [21, 20, 19]. However, the risk of complications is higher [20, 21]. There are several ways to deal with cochlear implant candidates with chronic otitis media, including cochlear implantation via the middle cranial fossa approach or performing a subtotal petrosectomy with closure of the external auditory canal [19, 22–24]. Controversies still exists in the literature regarding the choice of staged operation or single stage operation in active inflammation, indications for lateral petrosectomy and the surgical approaches [19]. According to Hellingman et al. most centers prefer two-stage procedures for implantation in patients with active chronic otitis media or cholesteatoma, with an interval of 3–6 months between disease eradication and cochlear implantation [20]. In active chronic otitis media subtotal petrosectomy may be performed in the first stage, followed by cochlear implantation in a second procedure once the ear is considered healthy [20]. A single stage operation can be a good alternative for patients with inactive chronic otitis media (tympanic membrane perforations without otorrhea, chronic adhesive otitis).

In the analyzed group, in 1 (0.9%) patient lateral petrosectomy was performed. In a boy with bilateral profound hearing loss and

cholesteatoma lateral petrosectomy was performed when the recurrence of the cholesteatoma was observed. The cholesteatoma was removed and the external auditory canal was closed (blind sac procedure). After 12 months, he received cochlear implant in the operated ear. One girl (0.9%) had cholesteatoma identified intraoperatively [25]. She was diagnosed with bilateral profound hearing loss of genetic origin. She underwent bilateral sequential implantation, with her first CI at 12 months of age. The second implantation was performed two years after the first CI surgery. The diagnostic imaging performed before the first cochlear implant surgery excluded middle ear pathology. In our clinic, in cases of sequential implantation, we do not routinely re-image the temporal bones if the first examination showed no pathology. Intraoperatively, after opening the facial recess, pearl-appearing congenital cholesteatoma was identified between the facial ridge, incudostapedial joint, and cochleariform process. To remove the pathology en bloc, the ossicles were disarticulated and the cholesteatoma was removed together with the incus. All tissues contacting the cholesteatoma were vaporized superficially with a diode laser and the cochlear implant was inserted via an extended round window approach. No recurrence occurred.

SURGICAL APPROACH

Posterior tympanotomy was first described by Jansen in 1957 [26, 17]. Currently, the mastoidectomy with posterior tympanotomy is the standard approach for cochlear implantation. A surgical approach with the advantage of avoiding mastoidectomy is the “suprameatal approach” proposed by Kronenberg in 2004 [27], which involves tympano-meatal flap elevation to expose the middle ear, drilling of the groove into the outer attic wall posterosuperior to the chorda tympani and lateral to the body of the incus, and connecting it to the tunnel drilled above the external auditory canal [28]. Several other techniques that can be used when it is not possible to visualize the

round window by posterior tympanotomy, such as combined transmastoid/transcanal approach, transcanal endoscopic approach, retrofacial approach, sacrifice of the chorda tympani nerve with extension of the facial recess, canal wall down mastoidectomy with overclosure of the ear canal, and facial nerve mobilization were described in the world literature [18, 24].

The retrofacial approach was first described by Pulec in 1996 [29, 18]. He described drilling the dense bone bordered by the facial nerve anteriorly, posterior semicircular canal posteriorly, and lateral semicircular canal superiorly [18]. The retrofacial approach may not be feasible in the presence of protruding sigmoid sinus, high jugular bulb, facial nerve anomalies, and highly elevated posterior fossa plates [18]. Yilmazer et al. presented 3 criteria for assessing HRCT of temporal bones, retrofacial approach is feasible in cochlear implantation when the following 3 criteria are met:

1. the round window – sigmoid sinus line is posterior to the facial nerve,
2. the jugular bulb is not obstructing the round window by extending just lateral to and above the level of the round window,
3. the distance between the posterior semicircular canal and the facial nerve is greater than 3 mm [18].

In some patients, all the above-mentioned surgical approaches may also be useless.

In cases of patients with chronic suppurative otitis media, unstable mastoid cavities with recurrent otorrhea, partially ossified cochleae, and in some cases of inner ear malformations, the middle fossa approach has been described in the literature as a good alternative in this group of patients [24]. Cochlear implantation through the middle fossa approach was described by Coletti, and in Poland by Szyfter et al. [23, 30].

In the case of some inner ear malformations, such as the common cavity or cystic forms of cochlear hypoplasia, the use of classic mastoidectomy with posterior tympanotomy may sometimes be impossible due to anomalies in the course of the facial nerve or difficulties in identifying the round window [31]. Canal-wall-down mastoidectomy or direct approach to the cavity through a transmastoid labyrinthotomy may be good alternative in this group of patients [32]. Various techniques to insert the electrode into the cavity were described, including the double posterior labyrinthotomy technique, single-slit cochleostomy or “banana” cochleostomy [33–35].

Another major problem that may require a different surgical approach is cochlear ossification, where progressive obliteration of the scala tympani may prevent standard full-length electrode insertion [16, 17]. The literature describes various surgical techniques that enable the cochlear implantation in such cases, including partial insertion, double-array insertion, retrograde cochlear implantation, full insertion into the scala vestibuli, drill-out procedure or radical cochleostomy procedure [16, 17, 36].

In our center, less common surgical approaches were used in 5 patients (4.5%):

- 2 retrofacial approaches in patients with bilateral incomplete partition type III-IP-III (Fig. 9.);
- Canal-wall-down mastoidectomy with preservation of the external auditory canal skin and tympanic membrane, which was moved aside with a downward shift of the high-riding jugular bulb obscuring the round window in a patient with bilateral cochlear hypoplasia CH-III;
- “Banana” cochleostomy – via the mastoid – in one patient with a common cavity [34];
- Lateral petrosectomy in the above-described patient with chronic otitis media.

ELECTRODE CHOICE

Preoperative imaging studies play important role in choosing the appropriate electrode array and determining its optimal length. In some inner ear malformations, such as cochlear hypoplasia, the available implantable space is smaller, or represents one common cavity of the vestibule and cochlea with nerve fibers located peripherally, as in the common cavity [5].

The common cavity (CC) is the first inner ear malformation where a different type of electrode may be required. HRCT shows a single, ovoid or round chamber, representing cochlea and vestibule that connects to the IAC, most often in the center [5]. This cavity has cochlear and vestibular neural structures, however, the exact location of neural tissue is unclear; it is likely to be located at the periphery and in the antero–inferior part of the cavity [5, 34, 35].

During implantation in CC, it is beneficial to use the straight full-banded electrode arrays that come into contact with the outer wall, enabling optimal electric stimulation [34, 25]. Examples of such electrodes are: CI24RE(ST) (Cochlear™), Cochlear Limited and MED-EL™ FORM [34, 37]. The dimensions of implantable space may vary; therefore, it is necessary to determine the length of the electrode before the implantation which is made by measuring the cavity diameter on HRCT of temporal bones. Establishing the radius (r) of the common cavity allows to define the appropriate length of the electrode, which should be $l = 2\pi r$, where r is the radius of the cavity [34]. The research group involved 1 (0.9%) patient with this malformation, who underwent cochlear implantation using a straight electrode CI24RE (ST) (Cochlear™), Cochlear Limited [34].

Another inner ear malformation, in which proper planning of the procedure and selection of electrode are required, is cochlear hypoplasia. In this malformations, there is a clear differentiation between cochlea and vestibule, external dimensions are less than those of a normal cochlea and various internal architecture deformities can be found [5]. Due to the smaller dimensions of the cochlea and various abnormalities in its internal architecture with narrower fluid spaces, shorter and thinner electrodes are recommended. An example of electrodes used in this malformation are the Nucleus STR24K and MED-EL™ FORM19 [31, 37].

Tab. II. Comparison of the usefulness of HRCT of temporal bones and MRI.

HRCT	MRI
<ul style="list-style-type: none"> · Assessment of bony structures, · Mastoid – pneumatization, · Tympanum – auditory ossicles, round window, · Bony labyrinth: bony sheath of the labyrinth outline, general structure of the cochlea, modiolus, bony cochlear nerve canal, · Course of nerve VII. 	<ul style="list-style-type: none"> · Labyrinthine fluid spaces – signs of obliteration, · Inner ear canal and the nerves running in it, · Cerebellopontine angle, · Further course of the auditory pathway in CNS, · Assessment of the encephalon.

In case of small, cystic hypoplastic cochleas, the use of straight electrodes with full rings is also beneficial; it is possible to perform a single-slit or a “banana” cochleostomy [34].

Another important inner ear malformation is incomplete cochlear partition (IP). In this malformation the use of the following electrodes were reported: CI24RE, CI24RE(ST), CI24RST, CI24M, CI24RCS, CI612 (Cochlear™), Cochlear Limited, Clarion HiRes 90 K, MED EL™ FORM24 and FORM19 [38, 19]. For IP-I and IP-III electrodes with complete rings should be preferred to stimulate as much neural tissue as possible [31]. In IP-II, the basal part of the modiolus is normal, therefore all kinds of electrodes – perimodiolar and straight can be used [31]. In the study group, in 6 (5.4%) patients with IP we decided to use a perimodiolar electrode CI512 (Cochlear™), Cochlear Limited. This electrode is held straight prior to insertion by platinum arrow stylet located inside the electrode array. The stylet is gradually removed as the electrode is inserted into the cochlea and placed in the perimodiolar position. Incomplete partition can be associated with a wide communication between the cochlea and the internal auditory canal [5]. In such cases, it is easy to insert the electrode array into the IAC. When the electrode is inserted via standard posterior tympanotomy or retrofacial approach IAC is running along the line of insertion. The use of a perimodiolar electrode, which take up its perimodiolar shape as the stylet is removed, allowed for a more controlled insertion. In none of the patients electrode misplacement into the IAC occurred.

In case of post-inflammatory cochlear ossification, thicker and therefore stiffer electrodes may be preferred. Of the two perimodiolar electrodes: CI532 (Cochlear™), Cochlear Limited and CI512 (Cochlear™), Cochlear Limited, some centers prefer the CI512 when intrascalar resistance may be expected during electrode insertion [40]. In the study group, the CI512 electrode was used in all patients who received a cochlear implant after meningitis.

In the case of expected luminal obstruction, a depth gauge is an extremely valuable tool to protect against unnecessary loss of the implant electrode. An example of one is Contour Advance Depth Gauge (Cochlear™), Cochlear Limited. It looks like a cochlear implant electrode. During the surgery, before the electrode array insertion, it allows to check that the labyrinthine fluid spaces are not obliterated and the cochlear implant electrode can be inserted.

HRCT AND MRI – COMPLEMENTARY TESTS

HRCT and MRI are complementary to each other for preoperative imaging. The two modalities in combination allow accurate and optimal evaluation of the anatomical structures prior to implantation (Tab. II). HRCT of temporal bones is superior to MRI in the evaluation of bony structures, course of the canal of facial nerve, anatomy of facial recess, tympanic cavity and mastoid, and otic capsule [1, 3]. In turn, MRI demonstrates higher applicability in the assessment of neural structures (especially in the T2-weighted sequence) and labyrinthine fluid spaces; it also allows the assessment of the central nervous system [1, 3].

MRI does not involve additional exposure of children to radiation, but often requires general anesthesia due to limited cooperation and relatively long examination time.

The main important questions that can be answered by preoperative imaging studies before cochlear implantation:

- is it possible to insert the electrode into the cochlea – is there an implantable space available (inner ear malformation or cochlear ossification)?
- is there a cochlear nerve that will allow the transmission of stimuli to further levels of the auditory pathway?
- are there any congenital anomalies or anatomical variations that could significantly complicate or preclude implantation?

CONCLUSIONS

Pathologies and anomalies identified during the assessment of preoperative imaging studies significantly altered clinical decisions in 30% of patients. The results confirm that preoperative imaging is an essential element of qualifying patients for auditory implant.

Diagnostic imaging studies can substantially affect clinical decisions – with regard to preoperative candidacy evaluation and subsequent patient management, including disqualification from a cochlear implant, selection of surgical approach or appropriate electrode, or qualification for an auditory brainstem implant.

In the study group, in 17% of patients inner ear malformations were identified. 2.7% of children were disqualified from a cochlear implantation due to severe congenital inner ear malformations. 9% of the patients have had bacterial meningitis. Of those, 50% experienced difficulties related to complete or progressive cochlear ossification. In 4.5% of patients less common surgical approaches other than mastoidectomy with a posterior tympanotomy were applied.

HRCT and MRI are complementary to each other and in combination allow accurate and optimal evaluation of the substantial anatomical structures prior to implantation.

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