

Current indications for the treatment of deafness with the use of cochlear and brainstem implants in children. New directions and possibilities

Aktualne wskazania do leczenia głuchoty u dzieci z zastosowaniem implantów ślimakowych i pniowych. Nowe kierunki i możliwości

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ABSTRACT:

The publication presents the latest indications for cochlear implantation in children. The principles of early detection of hearing loss in children as well as the detailed methodology and tools for diagnosis of children before cochlear implantation were discussed. Based on a review of publications from recent years, reference was made to the latest trends in implantation, taking into account: electroacoustic stimulation, electronatural stimulation, unilateral deafness, asymmetric sensorineural hearing loss, early implantation in children under 12 months of age and indications for brainstem implants in children with inner ear malformations. The significance of residual hearing in patients with cochlear implants as well as the use of appropriate electrodes for a specific clinical situation were also discussed. These factors may influence future development of speech and hearing.

KEYWORDS:

brainstem implants, cochlear implants, deafness, early implantation, pediatric, qualification

STRESZCZENIE:

W niniejszej publikacji przedstawiono najnowsze wskazania do wszczepienia implantu ślimakowego u dzieci. Omówiono zasady wczesnego wykrywania niedosłuchu u dzieci w Polsce oraz narzędzia stosowane w diagnostyce przed wszczepieniem implantu ślimakowego. Na podstawie przeglądu publikacji z ostatnich lat odniesiono się do najnowszych trendów implantacji, uwzględniając: stymulację elektroakustyczną, stymulację elektronaturalną, jednostronną głuchotę, asymetryczny niedosłuch odbiorczy, wczesną implantację u dzieci poniżej 12. miesiąca życia czy wskazania do implantów pniowych u dzieci z malformacjami ucha wewnętrznego. Poruszono również tematy dotyczące znaczenia zachowania resztek słuchu u pacjentów z implantem ślimakowym oraz zastosowania odpowiednich elektrod do określonej sytuacji klinicznej, co może mieć wpływ zarówno na późniejsze wyniki rozwoju mowy oraz słuchu, jak i na znaczne ułatwienie implantacji w przypadkach trudnych sytuacji anatomicznych.

SŁOWA KLUCZOWE: dzieci, głuchota, implanty pniowe, implanty ślimakowe, kwalifikacja, wczesna implantacja

ABBREVIATIONS

ABIs – auditory brainstem implants
ABR – auditory brainstem response
BAHA – bone-anchored hearing aids
CAEP – cortical auditory evoked potentials

CAP – Category of Auditory Performance
CCD – common cavity deformity
CI – cochlear implant
c-VEMP – cervical vestibular-evoked myogenic potentials
EAS – electroacoustic stimulation
ENG – electronystagmography

ENS – electronatural stimulation
HRCT – High Resolution Computed Tomography
IT–MAIS – Infant-Toddler Meaningful Auditory Integration Scale
MAIS – Meaningful Auditory Integration Scale
MRI – magnetic resonance imaging
PEACH – Parents’ Evaluation of Aural/Oral Performance of Children
PPBSN – Universal Newborn Hearing Screening Program
VHIT – video head impulse test
VNG – videonystagmography
VRA – visual response audiometry

INTRODUCTION

When the first cochlear implant was implanted in 1961 in an adult patient, the indication for its use consisted in complete bilateral deafness [1]. The simple one-electrode cochlear implants used at that time as well as in the 1970s were somewhat flawed and failed to provide satisfactory results in terms of optimum speech discrimination [1].

The advances in implant technology and new reports on their capability to provide better speech recognition and improved quality of life significantly changed the perception of hearing implants over the years [1]. In 1980, the first cochlear aid device was implanted in a pediatric patient [2]. From that time on, indications for use of cochlear implants started to dynamically evolve and their range expanded significantly [3]. Previous limitations such as patient age, inner ear malformation, condition of the middle ear or strict audiological criteria of deep deafness are of much less relevance today. In the last decade, indications for implantation of hearing implants were extended even further, particularly within the developmental age group of patients. This was facilitated by the development of implant systems and electrodes of different length and thickness, dedicated to inner ear malformations, cochlear ossification, or preservation of residual hearing [4].

Confirmation of the possibility to preserve residual hearing upon implantation was associated with electroacoustic and electronatural stimulation which significantly exceeded the typical range of indications for cochlear implants [5]. Clinical usefulness of residual hearing preservation was also confirmed for electric stimulation alone [4]. In recent years, a tendency was also observed to lower the age at implantation to under 12 months due to reports of significant long-term benefits from early intervention in young children [5, 6].

In addition to cochlear implants, which bypass the damaged part of the inner ear and facilitate restoration of hearing due to electrical stimulation of the auditory nerve, novel brainstem implants were also developed which bypass the lower levels of the auditory pathway by stimulating cochlear nuclei directly within the brainstem [1]. The first brainstem device was implanted in an adult patient 1979. Currently, brainstem implants can be used in very young children as well, which constitutes a significant change occurring over the last two decades [7, 8].

PART I COCHLEAR IMPLANTS

Deaf child's path to implantation

In Poland, the Universal Newborn Hearing Screening Program (PPBSN) has been in operation since 2002 [9, 10]. The program is implemented in centers divided into three referral tiers [9]. The first tier consists in neonatology and gynecology/obstetrics departments where otoacoustic emission tests are carried out [9]. The average number of tested children corresponds to 96% of the population of newborns [10]. Children with risk factors (irrespective of the OAE result), children in whom the first result was found to be incorrect, or children who did not undergo the test for various reasons are classified for retest [9, 10]. Retest is usually performed after 3 months at the next tier of the program (otolaryngology and phoniatrics/audiology centers) [9]. Retest usually consists of otoacoustic emission testing and tympanometry with evaluation of the acoustic reflexes; in some centers, auditory brainstem response (ABR) tests are also performed at this stage. The third referral tier consists in specialist centers capable of establishing hearing prostheses, provide surgical interventions, and rehabilitating children with hearing loss [10]. If the results remain incorrect, all the above tests are repeated after another three months and, if the child is pronounced to suffer from sensorineural hearing loss, bilateral hearing aids are usually used. This means that audiological diagnostics of deep congenital hearing loss should be completed by the age of 6 months, which unfortunately is often not the case. Starting from the age of 6 months, the child may undergo behavioral tests to assess their subjective reactions to sounds possibly enhanced by visual stimuli (visual response audiometry, VRA). These test are used to verify the results of objective exams. From that moment on, the child remains under the care of the speech and language therapists who observe the development of speech and hearing. If profound hearing loss is confirmed in subsequent studies and no adequate development of speech and hearing is observed, the child is usually classified for cochlear implant placement at the age of about 12 months.

Audiological Evaluation

Comprehensive audiological evaluation is required in all patients undergoing qualification for implant placement. In children the principle of „cross-check” as proposed by Jerger and Hayes in 1976 is used, meaning that mutually verifiable objective and subjective tests are carried out. Multiple repetition of complementary studies makes it possible to reliably evaluate children in whom the detection of hearing loss is generally much more difficult than in adults. The list of most common exams used at pediatric hearing implant placement centers is provided in Tab. I.

Current indications for cochlear implant placement in a pediatric patient:

1. Congenital or early acquired (i.e. in the prelingual period) sensorineural hearing loss preventing normal development of speech and hearing;

Tab. I. List of subjective and objective exams used as part of qualification for implant placement.

SUBJECTIVE EXAMINATIONS	OBJECTIVE EXAMINATIONS
behavioral audiometry: behavioral observational audiometry (BOA) or visual reinforcement audiometry (VRA) – in younger children aged 8–24 months	impedance audiometry including tympanometry and acoustic reflex measurements
toy audiometry – in children aged 18–48 months	otoacoustic emissions (OAE) – distortion product otoacoustic emissions (DP-OAE)
pure-tone audiometry thresholds (without hearing aids)	auditory brainstem responses (ABR) – examination of the auditory evoked potential
pure-tone audiometry thresholds with hearing aids in free sound field – both ears	auditory steady state responses (ASSR) – auditory evoked steady state potentials auditory potentials of steady state – currently considered one of the standards for diagnosing hearing loss a young child prior to the cochlear implant placement
speech audiometry with hearing aids	
speech audiometry without hearing aids	
speech discrimination as examined with hearing aids, in a free sound field of 65 dB – both ears examined individually and together	

- Children with sensorineural loss in the postlingual period in whom speech discrimination as assessed with the hearing aids using a verbal test appropriate to their age with a stimulus presented at 65 dB (the presented sound intensity levels range from 55 to 65 dB in different clinics) is lower than 50% (ears are examined independently and together) – in cases of documented lack of speech development, children with speech discrimination of < 50% in the more affected ear and < 60% in the less affected ear may also be qualified for implantation;
- Inappropriate development of speech and hearing despite correct prosthetization using the hearing aids is the most important criterion for qualifying a child for implant placement. In the event of insufficient speech and hearing, each patient should be qualified in an individual manner.

Otoneurological Evaluation

An important element of qualification for cochlear implant placement consists in the diagnostics of the vestibular organ. This is particularly important with the view of avoiding the bilateral vestibular areflexia following the placement of the second cochlear implant which might have a very adverse effect on the quality of life and the functioning of the child [11]. For this reason, sequential implantation and verification of the vestibular organ function prior to the placement of the second implant is a safer option in children qualified for bilateral implantation [11]. The purpose of the examination is to determine if no vestibular areflexia developed following the first implantation as observed in about 10% of patients undergoing implantation [12–14]. The second implant may improve the perception of sounds and the understanding of speech in silence [15]. Despite the fact that bilateral implantations are carried out at some centers, it is safer not to recommend

simultaneous bilateral implantations in children due to the risk of bilateral vestibular areflexia [11]. The most important component of diagnostic examinations consists in the assessment of vestibulo-ocular reflexes using electronystagmography (ENG), videonystagmography (VNG), and video head impulse test (VHIT). These tests allow the selective evaluation of the function of semicircular canals. Their limitation consists in a difficulty in their implementation in young children.

Videonystagmographic examination is performed with the patient's eyes open requiring voluntary cooperation. In young or non-cooperating children who shut their eyes and make it difficult to carry out the VNG test, ENG may be used instead as the test allows to record eyeball movements regardless of whether the eyes are open or shut. Another vestibular function test that is possible and more feasible in children is the investigation of cervical vestibular-evoked myogenic potentials (c-VEMP).

Specialist Consultations

As part of the qualification process, children should be consulted by a neurologist, an ophthalmologist, a psychologist, and a speech therapist. The criterion to be verified by a psychologist is whether the child/parents are sufficiently motivated for cooperation and rehabilitation and whether their expectations regarding the treatment outcomes are adequate. An ophthalmologist assesses the child for potential vision defects, examines the eye fundus to exclude retinitis pigmentosa which may be present in some congenital syndromes involving hearing and vision impairment. Patients with significant amblyopia or blindness are a special group of children in whom hearing is the primary information channel and development-supporting organ. Implantation is particularly important in these patients. Speech therapist's assessment is usually complemented by scores measured using the Meaningful Auditory Integration Scale (MAIS), Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) for children up to 3 years of age, and Category of Auditory Performance (CAP) [16]. In children in whom implantation is planned before the age of 12 months, diagnostic examinations are complemented by the Parents' Evaluation of Aural/Oral Performance of Children (PEACH) [17]. These scales facilitate an objective diagnosis as well as the monitoring of children's development following implantation.

Imaging Studies

Once audiological criteria have been established, qualification of a patient for the placement of hearing aid implants, involves imaging examinations [18]. These are necessary to assess whether the basic conditions required for implantation are being met, including the presence of implantable space the presence of the cochlear nerve capable of transmitting acoustic stimuli to the higher levels of the auditory pathway [18]. In addition, imaging scans allow to determine whether any additional factors exist which might impede or prevent implantation of the device, complicate the surgical procedure or cause post-operative complications [18].

Tab. II. Eligibility criteria for cochlear implant placement in children – summary of indications.

1	Sensorineural hearing loss preventing optimum development of speech and hearing in both pre- and postlingual deafness – at present, there is no lower limit of age, but the implantation must be performed after hearing impairment is diagnosed in the child in a rational manner in the child and after attempts at stimulations are made using the hearing aids.
2	Children with sensorineural loss in the postlingual period in whom speech discrimination as assessed with the hearing aids using a verbal test appropriate to their age with a stimulus presented at 65 dB is lower than 50% (ears are examined independently and together).
3	Children with profound sensorineural hearing loss following meningitis – immediate implantation!
4	Absence of speech development despite well-suited hearing aids, intensive rehabilitation and hearing therapy used for a minimum period of 3–6 months as documented by the opinions of speech therapists and psychologists.
5	No medical or radiological contraindications (specialist consultations, MRI, HRCT of the temporal bone).
6	Child/parents being sufficiently motivated for cooperation and rehabilitation and their expectations regarding the treatment outcomes being adequate.

Most frequently, these include [18]:

1. Obliteration of labyrinthine fluid spaces – usually due to ossification of scala tympani in the course of meningitis;
2. Congenital malformations, including internal ear malformations observed in approximately 20% to 30% of patients with profound hearing loss;
3. Congenital cholesteatoma;
4. Unfavorable anatomical variations, such as protruding sigmoid sinus, abnormal course of the facial nerve, difficult access to the round window.

Imaging studies facilitate prediction of numerous surgical difficulties and may be crucial for the choice of the surgical access, operated ear, or electrode [18]. The protocols regarding the choice of appropriate imaging studies vary from one site to another and may include HRCT of temporal bones and MRI as complementary studies. Performing both studies in each case is the safest option for both the patient and the surgeon. The protocols for detailed evaluation of imaging studies at the authors' center were presented in other publications [18, 19].

Attempted Prosthesis. The Benefits Of Hearing Aids

The absence of speech development despite well-suited hearing aids, intensive rehabilitation and hearing therapy used for a minimum period of 3–6 months as documented by the opinions of speech therapists and psychologists, is an indication for cochlear implant placement. Low-frequency stimulation of the cochlea prior to the implantation of a cochlear implant is considered beneficial for further development of speech and hearing; prosthesis stage should not be missed out prior to implant placement even in cases of deafness [20].

The summary of current indications for the placement of a cochlear implant (CI) is provided in Tab. II.

The final decision regarding patient's eligibility if made by the qualification committee, usually made up of an otosurgeon, an audiologist, and a speech and language therapist. The eligibility criteria for implantation of the second cochlear implant in children as practiced at the authors' center are set out in another publication [11].

Cochlear implants – new directions and perspectives

Preservation Of Residual Hearing

Initially, cochlear implants were used in the treatment of total bilateral deafness, and residual hearing in patients with implanted devices was outside the scope of audiologists' interests [1]. On the basis of observations of patients in whom residual hearing was observed even with implants in place, a hypothesis was raised at the end of the 1990s suggesting the possibility of preserving residual hearing; a number of studies followed to confirm the benefits of residual hearing preservation in both adults and children [4, 21, 22, 23]. Preservation of residual hearing in patients is associated with numerous benefits including better discrimination and development of speech, better understanding of speech in noisy and difficult acoustic conditions, better perception of music and a more natural perception of sounds and speech [21, 15, 22]. In small children, particularly infants, the nature of audiological diagnostics prevents full and accurate determination of residual hearing. Therefore, in every small patient, the procedural technique and the type of electrodes used should allow for preservation of potential residual hearing for a possible use being made of it in the future. Loss of residual hearing is attributed to many factors, such as acoustic trauma during the surgery (drilling), mechanical injury caused by the insertion of the electrode (damage to the osseous spiral lamina, damage to the basal membrane, rupture of the periosteum of the scala tympani), damage at the molecular level, fluid shock wave being generated within the cochlea upon electrode insertion, osteoneogenesis, or fibrosis caused by foreign bodies, bone dust, or blood [24, 25, 13].

To avoid the loss of residual hearing, surgical techniques and implant technology are still subject to improvement and modifications [26, 27]. Currently, the main factors considered to influence preservation of residual hearing include [26–28]:

- characteristics of the electrodes (length of the electrode inserted into the cochlea, its diameter and stiffness);
- surgical implantation technique (cochleostomy vs round window approaches, electrode insertion rate);
- pharmacological treatment (antibiotics, glucocorticosteroids) in the perioperative period.

Two types of electrodes can be differentiated in relation to their location and position within the cochlea (Fig. 1.). Both types of electrodes have their advantages and disadvantages:

- straight or lateral wall electrodes – aligned along the external wall of the cochlea. These electrodes have

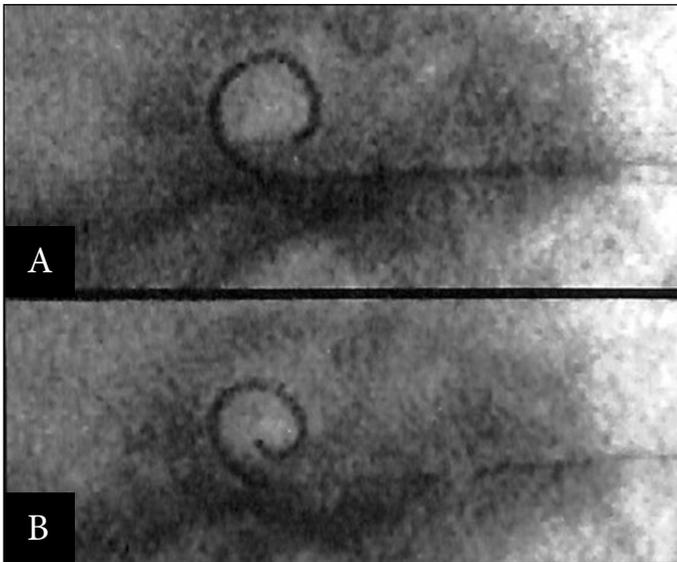


Fig. 1. Straight (A) and perimodiolar (B) electrodes.

been traditionally used since the onset of implantation practice [28, 29];

- perimodiolar electrodes – over time, electrodes aligned along the internal wall of the cochlea were also introduced [27].

Perimodiolar electrodes provide focused stimulation and precise propagation thereof due to the close proximity of the auditory nerve, theoretically allowing for lower current consumption and better discrimination of speech as compared to straight electrodes [30, 31]. Preservation of residual hearing is possible when using both perimodiolar and straight electrodes [32]. The first meta-analysis by Santa Maria et al. (2014) revealed that straight electrodes were not superior to the perimodiolar ones [27]. A more recent meta-analysis from 2019 revealed a statistically significant difference between straight and perimodiolar electrodes after one month from implantation, favoring the use of straight electrodes for preservation of residual hearing ($p < 0.001$) [26].

The length of the electrodes is another factor affecting preservation of residual hearing and the hearing quality. Initially, the use of long electrodes was considered to lead to the loss of residual hearing [27, 28]. This short electrodes were used in patients with partial deafness and residual hearing to provide stimulation within the basal turn of the cochlea [28]. As the research into the preservation of residual hearing was progressing, numerous papers were published regarding the potential to preserve residual hearing following implantation of full-length electrodes [23, 27, 28]. In the case of residual hearing loss, the longer-length electrodes have an advantage over the short electrodes by providing stimulation over a longer section of the cochlea [23]. This may be particularly important in children whose implants should work for many years. According to the meta-analysis, the use of long electrodes does not reduce the percentage rates of residual hearing preservation [26, 27].

The speed of electrode insertion is also important. As the electrode is introduced to the scala tympani, a fluid shock wave is generated

resulting in a significant, insertion rate-dependent change in fluid pressure within the cochlea [24]. This may result in the loss of residual hearing [24]. Slower insertion of the electrode ensures better preservation of residual hearing [22, 24, 27].

Another questionable factor related to the preservation of residual hearing consists the choice of access to the scala tympani, namely whether better preservation of residual hearing can be achieved by means of cochleostomy, round window approach, or the enlarged round window approach. According to the most recent analysis there are no statistically significant differences between these approaches [26]. An important factor in peri- and intraoperative period is administration of steroids which facilitate better preservation of residual hearing [28]. The procedure is performed according to the soft surgery procedure which includes:

- absence of perilymph suctioning,
- careful manipulation during implantation,
- very slow and gentle insertion of the electrode,
- intraoperative use of glucocorticosteroids [3].

Preservation of residual hearing after implant placement ensures better discrimination of speech and hearing quality. Additional benefits in this group of patients may be provided by electroacoustic stimulation in which the electric component is accompanied by the acoustic component [22].

NEW STRATEGIES FOR THE TREATMENT OF PARTIAL DEAFNESS. THE BROADENING OF AUDIOLOGICAL CRITERIA

EAS – electroacoustic stimulation

The concept of electroacoustic stimulation (EAS) was introduced in 1999 [33]. It consists of acoustic stimulation of the residual hearing at low frequencies combined with the use of a cochlear implant and electrical stimulation at higher frequencies [22, 33].

A lot of evidence and study data was published to support long-term benefits of EAS in children and adults [22]. The most important include [22]:

- better discrimination in speech (single syllable tests) in silence as compared to the hearing aid alone,
- better hearing in difficult acoustic conditions – noise, reverberation,
- better localization of sounds,
- better perception of music.

An electroacoustic system consists of a cochlear implant and a hearing aid integrated within a single device. Intracochlear electrode of the device is used to provide electrical stimulation to the acoustic nerve endings while the sound processor provides additional acoustic stimulation via the inserted earmold [34]. Today, most companies manufacture implants which facilitate electroacoustic stimulation if residual hearing is detected even a long time after the implant has been installed, offering the possibility of the

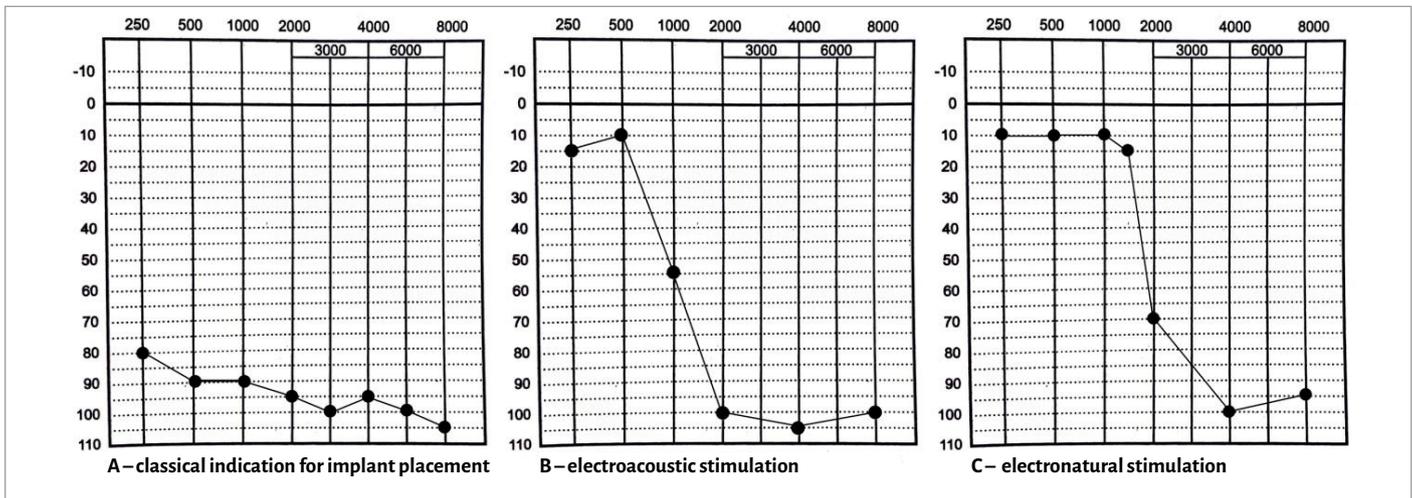


Fig. 2. Examples of audiograms in patients qualified for cochlear implants: profound sensorineural loss—classical indication for implant placement (A), electroacoustic stimulation (B), and electronatural stimulation (C).

acoustic component being attached to the implant. The acoustic component can also be disconnected in the case of residual hearing loss, leaving the patient to electric stimulation alone [23]. In this situation, the length of the implant electrode is important. If the electrode is long enough, electrical stimulation will provide good discrimination of speech. In the past, when short electrodes (11 mm) were used for electroacoustic stimulation and when the loss of residual hearing occurred afterwards, some patients required reimplantation with longer electrodes (> 16 mm), since the short electrodes used for electrical stimulation did not allow for adequate discrimination of speech and satisfactory hearing [35].

Ens—Electronatural Stimulation

Another extension of the indications to the cochlear implant placement consists in electronatural stimulation of patients with partial deafness, i.e. normal or near-normal hearing in the frequency range of 125–1500 Hz with simultaneous profound hearing loss at higher frequencies [36]. The concept of electronatural stimulation makes use of preserved natural hearing in low and medium frequencies and of the cochlear implant within the high frequency range [36]. The frequency ranges for EAS and ENS are compared in Fig. 2. It should be highlighted that the eligibility criteria for electronatural stimulation broadly exceed the classical indications for cochlear implantation. Notably, if hearing loss occurs following the implantation, which is always possible, patient may experience a deterioration in hearing quality compared to the pre-implantation condition. The candidate for implantation should present with markedly restricted ability to discriminate speech.

Single-Sided Deafness And Cochlear Implants In Children

Initially, single-sided deafness was considered to require no intervention [37] as the child could develop the ability to speak and hear relatively well. However, there is no doubt that bilateral hearing offers a number of advantages. At present,

solutions for use in SSD patients include CROS-type hearing aids or bone-anchored hearing aids (BAHA) [14]. The devices work by transmitting the sound from the deaf ear to the hearing ear to make the subject aware of the sounds emitted from the deaf side [14]. However, this cannot be considered “real” binaural hearing as it facilitates the hearing of sounds emitted from the deaf side but fails to provide the real benefits of binaural hearing with regard to discrimination of speech in noisy conditions, elimination of acoustic head shadow, and better sound localization abilities [14]. The cochlear implant has become another potential option for intervention in this group of patients, and it is the only solution allowing patients with single-sided deafness to take experience the benefits of binaural hearing [38]. It should be pointed out that cochlear implants are not a standard treatment of single-sided deafness. The use of cochlear implants to treat single-sided deafness in children remains a matter of dispute, and the clinical experience in this age group is much lower than that in adult patients [38]. It is known that deprivation of acoustic stimulation during the development of the auditory pathway leads to irreversible changes within the cerebral cortex. The period of the greatest plasticity of the cerebral cortex, referred to as the critical period, falls at about the age of 3 to 5 years, with the plasticity gradually diminishing starting from the age of 7 [38]. A large group of SSD patients may be highly reluctant to use cochlear implants due to the marked differences between electric and natural stimulation. Failure to accept this difference and the need to adapt to its use may lead to some children rejecting the use of the cochlear implant [37, 14]. It is suggested that these results depend on the etiology of deafness in children, being the better the shorter is the duration of deafness and the earlier the placement of the implant in both acquired and congenital deafness. However, decisions should be taken on a case by case basis [37].

The early fitting of the implant allows the significant plasticity of the child’s nervous system to be used to their advantage, offering a better chance for accepting the cochlear implant in congenital or early-acquired single-sided deafness and more successful rehabilitation of patients [39].

Currently, the authors' center provides care to three children in whom implant was used to treat congenital single-sided deafness. The implants were placed at the ages of 14 and 16 months, and are used by the children every day to provide them with benefits of binaural hearing.

The Age Of The Child At Implantation. The Early Intervention Strategy

An increasing number of reports raise the subject of early qualification of children for cochlear implant placement [40, 41]. It has been known for many years that the earlier the child receives the implant, the better the speech and hearing results [42, 43]. For many years, the standard of conduct recommended by the National Institute of Health, the Joint Committee on Infant Hearing, and the American Academy of Pediatrics was to diagnose hearing disabilities before the age of 3 months, to prosthetize and rehabilitate the patient using a hearing aid at the age of 6 months and, if no benefits are observed from the use of the hearing aid, to place a cochlear implant at the age of about 12 months.

However, recent reports suggest that significantly better results are obtained in terms of speech and hearing development in children subjected to earlier implantation between the ages of 6 and 12 months [42–44].

Early qualification for implant placement requires earlier diagnostic examinations ensuring reliable results in children only a few months old. Implantation must not be allowed in children capable of functioning well with the use of hearing aids. Diagnostic examinations should be appropriately extended to include, in addition to standard methods consisting of traditional examinations and early prosthetization attempts, the assessment of aided (speech-evoked) cortical auditory evoked potentials (CAEP) and assessments using the PEACH scale. These examination provide additional confirmation of profound hearing loss in this group of patients. Lowering the age at implantation presents no problem in patients with genetically confirmed deafness with all family members being deaf.

The PEACH scale is an age-unrestricted questionnaire to be completed by the patient's parents, assessing a total of 12 hearing parameters in a child (Tab. III.).

Therefore, in cases of genetically confirmed deafness as well as in children in whom no doubts are raised by the qualification committee regarding the profundity of their hearing loss, lowering the implantation age to less than 12 months should be striven for as it ensures better development of speech and hearing compared to implantation performed after the age of 12 months [40–44].

Electrode Selection Depending On Clinical Conditions

Accurate preoperative analysis, including that of the nature of hearing loss and imaging studies, facilitates appropriate cochlear implant electrode being selected for a particular clinical

conditions [19]. The first case in which a different electrode may be beneficial is the post-inflammatory early-stage ossification of the cochlea when the connective tissue forms within the inner ear spaces [19, 45]. In such cases, thicker and thus more rigid electrodes may be preferred [19, 45]. Cochlear implant manufacturers offer a wide assortment of electrodes appropriate for varying clinical settings. In the aforementioned case, rigid, stylet-based electrodes such as CI612 (Cochlear™) are preferred by the authors [45]. The stylet which enhances the rigidity of the electrode facilitates penetration of adhesions and soft obstacles within the cochlea. Advanced oblitative lesions within the cochlea frequently prevent implantation and increase the risk of the expensive electrode being damaged by the surgeon. In cases of labyrinthine obliteration, preliminary use of a depth tester gauge is an extremely useful solution preventing unnecessary loss of the implant electrode [19]. The depth tester gauge looks like the actual electrode and allows the surgeon to check whether the fluid spaces within the cochlea are not obliterated before the actual implantable electrode is inserted.

Another clinically relevant problem consists in the malformations of the inner ear [19, 46]. In some malformations, the space available for implantation is limited, as in the case of cochlear hypoplasia, or has the form of a joined space consisting of the vestibule and the cochlea with nerve fibers located on the periphery, as in the case of common cavity deformity (CCD) [47]. Proper electrode selection can significantly facilitate implantation in these difficult otological settings [19, 46].

The third important clinical condition consists in residual hearing being preserved in the patient. Initially, straight electrodes and shorter electrodes were considered beneficial in preservation of residual hearing [32]. This outlook, however, has changed over the years. Many studies have now confirmed that optimum electrodes to be used in these cases are small-diameter, delicate-structured, and appropriately long electrodes (ensuring effective stimulation of the cochlea in the case of residual hearing loss). Effective preservation of residual hearing is possible when using either perimodiolar or straight electrodes [32]. CI632 (Cochlear™) or CI612 (Cochlear™) are the example electrodes of this type. It is worth highlighting that electrodes facilitating preservation of residual hearing are now offered by all implant manufacturers.

PART II BRAINSTEM IMPLANTS

In some patients, cochlear implants are either ineffective, or their placement is impossible for various reasons [7, 32, 48]. In such cases, solution can be provided by auditory brainstem implants (ABIs) which provide the patients with a new opportunity to enter the world of sounds [7, 47]. Their operation is based on direct electrical stimulation of the cochlear nuclei regions – brainstem implants provide a bypass of the auditory pathway consisting of the cochlea and the cochleovestibular nerve [48]. The first brainstem implant to be used in a child was implanted in Italy in 2001 [48]. Few of the children with profound deafness are potential candidates for ABI placement; currently, there are about 400 pediatric users of these implants worldwide [49].

Tab. III. The PEACH scale. Each question is scored on a scale of 0 (never) to 4 (always).

HOW OFTEN DOES THE CHILD EXPERIENCE	HOW DIFFICULT IS IT FOR THE CHILD TO
0 – never – 0%	0 – very difficult – 0%
1 – rarely – 1–25%	1 – difficult – 1– 25%
2 – never – 26–50%	2 – neutral – 1–50%
3 – frequently – 1–75%	3 – easy – 51–75%
4 – never – 76–100%	4 – very easy – 76–100%

PRELIMINARY QUESTIONNAIRE		
In the past week:	YES	NO
Has the child been healthy?		
Has the child been wearing the hearing aid/cochlear implant speech processor?		
Has the hearing aid/cochlear implant speech processor been working properly?		

WHAT WERE THE RESPONSES/REACTIONS OF THE CHILD TO THE FOLLOWING SITUATIONS DURING THE PAST WEEK – PLEASE TICK ONE OF THE ANSWERS	HOW OFTEN DOES THE CHILD EXPERIENCE:	HOW DIFFICULT IS IT FOR THE CHILD TO:
1 How often does the child wear the hearing aid/cochlear implant speech processor?		
2 How often does the child complain of sounds being too loud?		
3 Does the child react when one of the parents calls them when they're not seeing them (turns their head, responds)?	In quiet conditions	
4	In noisy conditions	
5 Does the child follow simple instructions?	In quiet conditions	
6	In noisy conditions	
7 How often does the child start a conversation or participate in a conversation?	In quiet conditions	
8	In noisy conditions	
9 Does the child listen carefully when someone reads to them? When the child is listening to music/watching cartoons, do they keep up with the narrative and try to understand it?		
10 How often does the child understand what is being said to them in a car/on a bus/on a train?		
11 How often does the child recognize the voices of others without seeing their faces?		
12 How often does the child respond to sounds other than voices ?		

RESULT	RESULT (%)	RESULT – EASE OF HEARING (%)
In quiet conditions	A (add up the scores for questions 3+5+7+9+11)/20×100	
In noisy conditions	B (add up the scores for questions 4+6+8+10+12)/20×100	
Overall	C (A + B)/2	

Initially, the main indication for brainstem implant consisted in acquired deafness caused by bilateral damage to the cochleovestibular nerves in the natural history of neurofibromatosis type 2 [48]. The first international consensus on the use of brainstem implants in children and non-NF2 patients, established in 2011, was based on the multicenter experience in 61 centers and permitted the use of brainstem implants also in cases of congenital malformations of the inner ear, including cochlear nerve aplasia/hypoplasia and acquired causes such as cochlear ossification in the course of meningitis [7].

The 2016 consensus identified 3 groups of radiological indications for the brainstem implant placement [7, 8]:

1. Well-defined congenital indications:

- Complete labyrinthine (Michel) aplasia;
- Cochlear aplasia;
- Cochlear aperture aplasia;
- Cochlear nerve aplasia;

2. Possible congenital indications:

- Hypoplastic cochlea with hypoplastic cochlear aperture;
- Common cavity and incomplete partition of type I according to Sennaroglu, with or without the cochlear nerve;
- Unbranched cochleovestibular nerve;
- The hypoplastic cochlear nerve (radiologically defined as nerve having 50% of the size of a normal nerve or the diameter smaller than that of the facial nerve);

3. Acquired indications:

- Cochlear ossification in the course of meningitis – complete ossification of the cochlea in CT, complete absence of visible labyrinthine fluid spaces;
- bilateral transverse fractures of the temporal bone with damage to the cochleovestibular nerve;
- Otosclerosis with massive cochlear destruction.

Obliteration of labyrinthine fluid spaces according to the 2011 and the 2016 consensus [7, 8]:

1. In cases when fluid spaces are seen in T2-weighted MR images, cochlear implants should be the first therapeutic choice;
2. When complete ossification of the cochlea is observed in CT scans, and complete absence of visible fluid spaces is observed in MRI scans, the chances for effective CI placement are low and the patient should be qualified for the placement of a brainstem implant.

Child's Age

In line with current recommendations, brainstem devices should be implanted in children aged at least 12 months, preferably

Tab. IV. Categories of auditory performance (CAP).

HEARING-RELATED BEHAVIORS (WITHOUT LIP READING)	
Lack of awareness of ambient sounds and voices	0
Awareness of ambient sounds	1
Response to the sounds of speech	2
Ability to identify ambient sounds	3
Ability to discriminate between speech sounds without lip reading	4
Ability to understand common expressions such as „open the door“ without lip reading	5
Ability to understand conversations without lip reading	6
Speaking over the phone with a known speaker	7
Ability to follow a conversation in a group of people in a room with reverberation or background, for example in a classroom or restaurant	8
Speaking over the phone with an unknown speaker in an unpredictable context	9

before the age of 2 years and not later than at the age of 3 years [7, 8, 50]. In the event of inner ear malformations and favorable anatomical conditions, brainstem implant may be the second line treatment when cochlear implant has shown to be ineffective [8]. When the decision is made to use the cochlear implant in the first line, the device should be implanted at approximately 1 year of age [8]. Next, the patient should be followed up and his development should be evaluated [8]. If no benefits are observed, the decision regarding the placement of a brainstem implant should be made before the patient is 24 months old. The delay in the decision is associated with poorer expected results of the brainstem implant placement [8].

The Benefits Of Brainstem Implant

The effects of treatment with brainstem implants are usually inferior to those of CIs, with rehabilitation being longer and more difficult [8, 51]. Speech development is delayed and worse compared to children with cochlear implants, with complementary use of sign language and lip reading being quite frequent [8]. However, brainstem implants facilitate an improvement in the quality of life in a large part of patients. A low percentage of patients experience no benefits from the placement of the brainstem implant, and a vast majority of patients willingly use the device. The outcomes depend on a number of factors, such as the age at implantation, the number of active electrodes, and comorbidities, and may differ significantly ranging from the perception of the sound though improved lip reading to the understanding of speech [8, 51, 50].

Good, and even very good outcomes are possible in some patients. In children with prelingual deafness, inner ear malformations and comorbidities, Pure Tone Audiometry of 30–60 dB HL can be achieved in most cases [8]. The assessment of the child's response to sounds after the implantation can be based on the categories of auditory performance which provide a good indicator for the assessment of hearing and speech development in patients with hearing implants (Tab. IV.).

In most children with brainstem implants, the CAP scores are at approximately 4–5 [8]. Better CAP scores of up to 8 are sometimes observed in some implant users. The results are better in patients in whom the procedure was carried out earlier, i.e. below the age of 2 years, preferably at the age of 1–1.5 years, in non-NF2 patients as opposed to NF2 patients, in patients with postlingual hearing loss, and in patients with no comorbidities [8, 51]. About 50% of children with ABI will use speech as the primary means of communication. About 15% will be able to talk over the phone and 15% will only be capable of the perception of sound. At the ESPCI conference in Bucharest, Manrique et al. presented the results of speech and hearing rehabilitation in children with brainstem implants: 3 out of 24 patients had CAP scores of > 6, 2 out of 24 patients had CAP score of 1, and the CAP scores of the other 19 children ranged between 2 and 5 [52]. All children used their implants willingly and presented with improved rehabilitation of speech and hearing as well as with improved quality of life [52].

Identification of the family expectations, increasing the awareness regarding rehabilitation opportunities and expected results, and highlighting the importance of long and tedious rehabilitation is an important aspect to be raised during the discussion with the family of the child to be qualified for brainstem implant placement. Currently, the authors' center provides care to one child with a brainstem implant placed before the age of 2 due to major bilateral congenital malformation of the inner ear (cochlear aplasia with vestibular enlargement and type I cochlear hypoplasia). Clear responses to sounds of different frequencies were observed following implantation; currently, the patient undergoes extensive rehabilitation of hearing and speech.

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