Pediatr Pol 2024; 99 (2): 136-142
DOI: https://doi.org/10.5114/polp.2024.140854
Submitted: 01.01.2024, Accepted: 16.02.2024, Published: 27.06.2024

PEDIATRIA Polska
Polish Journal of PAEDIATRICS

REVIEW PAPER

Hearing impairment in children – otolaryngologist's perspective, causes, diagnosis, rehabilitation and hearing prosthetics

Kamila Bała, Karolina Dorobisz

Department of Otolaryngology, Head and Neck Surgery, Wrocław Medical University, Wrocław, Poland

ABSTRACT

Hearing impairment in children is a significant issue worldwide, including Poland, as it hinders their proper development. Early hearing prosthetics ensure language and social development comparable to their healthy peers. Proper diagnosis is essential to achieve this goal. In Poland, the Universal Newborn Hearing Screening Program allows for the rapid detection of children with hearing impairment and early therapeutic intervention. Bilateral sensorineural hearing loss is the primary indication for cochlear implantation. Patient assessment, surgery, and rehabilitation are carried out in highly specialized centers. Hearing impairment is also a concern for an older age group of children, where the causes are primarily inflammatory. In this case, parents, pediatricians, family doctors, speech therapists, and teachers play a significant role in suspecting hearing impairment in children. It is crucial to effectively and promptly include all children with hearing impairment in the program, especially those with risk factors, where delayed onset hearing loss may occur.

KEY WORDS:

speech development, cochlear implant, sensorineural hearing loss.

INTRODUCTION

Hearing, as one of the senses, is a vital element in every individual's functioning. Its proper operation is necessary for a child to achieve developmental milestones at the right time. Hearing impairment represents a significant problem among children. Worldwide, it affects approximately 0.2% of newborns, 0.4% of infants, 1% of children aged 1–4, 1.5% of those aged 5–9, 1.7% of those aged 10–14, and 1.9% of those aged 15–19 [1]. Early hearing prosthetics using hearing aids or cochlear implants are crucial for children with sensorineural hearing loss [2]. Cochlear implants are among the most significant achievements in medicine and technology [3]. In Poland, the program for treating total deafness and profound hearing loss with cochlear implants began in 1992 [4]. A significant milestone in Polish oto-surgery was on 16th July 1992, when Professor Henryk Skarżyński performed the first cochlear implantation in Poland [3]. Since then, more than 12,500 cochlear implants have been implanted at the Institute of Physiology and Pathology of Hearing [5]. Approximately 800 patients undergo cochlear implantation annually across the country [6]. This paper aims to discuss the issue of hearing impairment and the possibilities of treatment through cochlear implantation.

HEARING DIAGNOSIS

The Universal Newborn Hearing Screening Program (UNHSP) in Poland has significantly contributed to early detection of hearing-impaired children. Poland was the first of the 9 countries in the world that conduct universal hearing screening covering over 90% of the population [7]. Before the establishment of the program, hearing

ADDRESS FOR CORRESPONDENCE:

Kamila Bała, Department of Otolaryngology, Head and Neck Surgery, Wrocław Medical University, Wrocław, Poland, e-mail: kamila.bala@wp.pl

Source: https://www.nfz-szczecin.pl/lquil_przesiewowe_badania_sluchu_u_noworodkow.htm

FIG. 1. A blue certificate and a yellow certificate

screening was only conducted on newborns from the risk group for hearing impairment, and studies showed that the presence of hearing loss risk factors was observed in half of the cases [8]. Moreover, 90% of persistent sensorineural hearing impairments in children aged 0–5 are either congenital or acquired during the perinatal period [8]. These data indicated that newborn hearing screening should encompass the entire newborn population and be performed immediately after birth in the neonatal unit. Currently, within the first 3 days of a child's life in the neonatal ward, a screening test using otoacoustic emissions (OAE) is performed. In the case of a normal result and no risk factors, the patient receives a blue certificate, as presented in Figure 1, which is included in the child's health booklet. Children with abnormal OAE results, risk factors for hearing loss as presented in Table 1, or no OAE screening are issued a yellow certificate, also shown in Figure 1. Children with a yellow certificate are referred for retesting with OAE, followed by auditory brainstem response (ABR) testing for a precise objective assessment of hearing. Based on audiological examinations, patients qualify for hearing prosthetics using hearing aids. If conventional hearing aids do not yield the desired results according to the assessment criteria, cochlear implantation is proposed to the patient and their parents.

According to the UNHSP report for the year 2022, 266 870 (96%) live-born children in Poland were covered by the program [9]. Parents of 22 194 children (8.32%) received a yellow certificate along with program leaflets and referrals to the second level of the UNHSP. Data from levels II and III in 2022 show that diagnostic evaluations were completed for 8 762 children (39.5%), and 177 were confirmed to have hearing impairment [9]. In 2021, 43% of children participated in the second level of the program, where abnormal hearing was diagnosed in 343 patients [10]. The data suggest a declining trend in child participation in subsequent hearing diagnostic stages. The most common type of hearing impairment in children examined at level II is bilateral sensorineural hearing loss, which, if undetected at an early developmental stage, can hinder proper child development. With moderate hearing impairment, speech development is delayed, and profound hearing loss results in no speech development. Speech is a primary means of communication, and its disruption lowers the patient's and their family's quality of life [11].

TABLE 1. Risk factors for hearing impairment [13]

1.	Intrauterine infections from the TORCH group: toxoplasmosis, syphilis, viral hepatitis type B (hepatitis B), rubella, cytomegalovirus, herpes viruses, and the Zika virus
2.	Ototoxic drugs: aminoglycoside antibiotics administered for longer than 5 days
3.	Prolonged neonatal intensive care exceeding 5 days
4.	Extracorporeal membrane oxygenation (ECMO) usage
5.	Birth asphyxia
6.	Hyperbilirubinemia requiring exchange transfusion
7.	Craniofacial anomalies
8.	Genetic syndromes associated with hearing loss
9.	Chemotherapy or head injury requiring hospitalization
10.	Confirmed perinatal or postnatal meningitis or encephalitis
11.	Family history of permanent childhood hearing loss
12.	Parental concerns regarding hearing, speech, language, developmental delays

Additionally, a summary of 12 years of hearing screening programs among first and sixth-grade students attending primary schools in Warsaw showed that abnormal hearing test results were obtained for 25,559 children (13.3%). This demonstrates that hearing impairment is also a concern for an older age group, with primarily inflammatory causes—chronic inflammation of the nasal mucosa leading to Eustachian tube obstruction and subsequent chronic effusive otitis media [12].

The following audiological examinations are used for hearing diagnosis: subjective: pure-tone audiometry (from age 3), speech audiometry (from age 6–7), behav-

FIG. 2. Cochlear implantation centers

ioral audiometry (under age 3), and objective tests, including ABR, OAE, and impedance audiometry.

Failure to undertake diagnostic and therapeutic measures in these cases leads to learning difficulties, emotional disturbances, interpersonal problems, and delays in socio-cultural development [12].

IMPLANT PROGRAM

A cochlear implant is an electronic device surgically implanted in the inner ear to transmit and process sounds into electrical signals, which are then directly conveyed to the auditory nerve. Treating patients with a cochlear

implant involves a team of specialists working in the centers presented in Figure 2. The pediatric team responsible for the assessment for cochlear implantation, surgery, and rehabilitation comprises otolaryngology surgeons, pediatric anesthesiologists, audiologists, psychologists, speech therapists, clinical engineers and deaf educators [14]. Together, they make decisions regarding the eligibility or ineligibility of patients for cochlear implantation based on the indications for implantation and the latest scientific research in this field. The primary indications for cochlear implantation include bilateral deafness, bilateral residual hearing, bilateral profound sensorineural hearing loss and a lack of speech development despite appropriately fitted hearing aids and intensive rehabilitation for a minimum of 3–6 months [15]. Extended indications encompass bilateral cochlear implantations and implantations in cases of unilateral deafness [15]. Another crucial criterion is the absence of medical and radiological contraindications to cochlear implantation [16]. To determine this, a high-resolution computed tomography scan of the temporal bones and/or magnetic resonance imaging of the head is performed before the planned surgery. These tests allow for the early detection of congenital inner ear abnormalities (e.g., absence of the auditory nerve) as well as obliteration of fluid spaces in the vestibular system, often caused by ossification of the cochlea during meningitis [17]. Such abnormalities serve as contraindications to cochlear implantation, and in cases of hearing loss following meningitis, urgent implantation is necessary due to developing complications [16, 17]. Criteria for cochlear implant eligibility also take into account the child's or parents' sufficiently high motivation to cooperate and appropriate expectations regarding treatment outcomes [16]. The assessment committee makes the final decision based on previous examinations and observations. After being found eligible for cochlear implant surgery, the patient undergoes preparation so that implantation can occur before the child reaches 12 months of age [18]. Studies on the auditory system in young children have shown that cochlear implantation before the first year of life enables patients to acquire auditory and verbal skills during the period of greatest plasticity in the central nervous system [18]. Before a patient embarks on the diagnostic-therapeutic-rehabilitative path of cochlear implant qualification presented in Table 2, they must be referred by a physician whose knowledge of the indications can aid in their daily clinical practice.

ELIGIBILITY CRITERIA LIMITATIONS

Based on evidence-based practice, it is emphasized that cochlear implantation in children should be performed as early as possible to facilitate their language development [20]. Studies suggest that early implantation can prevent long-term speech language deficits in children [21]. A 10-year observation of two groups of children who received implants before or after 12 months of age showed that after 5 years, all younger children who received implants at an earlier age had speech understandable to an average listener, compared to 67% of children who received implants later [22]. Additionally, this study found significantly better results in the grammatical development of the younger group [22]. In children with single-sided deafness, where the goal is to provide bilateral stimulation for the development of binaural hearing, a meta-analysis showed that cochlear implantation improved speech perception in noise and quiet, as well as sound localization [23, 24]. Furthermore, the average age at implantation in individuals not using implants was statistically significantly higher than the age at implantation in those using implants regularly or intermittently [24]. In addition to the benefits for the patient, the decision for early implantation is influenced by safety studies. In a study involving 136 patients aged 3.6–11.9 months, patients were divided into those under 9 months and those 9–11 months old [25]. There were no significant differences in the frequency or severity of adverse events between the two groups at the time of surgery [25]. To emphasize the importance of early fitting of a hearing aid, which has a wearing period of 3–6 months before cochlear implantation, the current guidelines of the Joint Committee on Infant Hearing state that hearing screening should be conducted by 1 month of age, hearing loss or deafness identification should occur by 3 months of age, and therapeutic intervention by 6 months of age [26].

COLLABORATION AMONG PEDIATRICIANS, FAMILY DOCTORS, SPEECH THERAPISTS, NEUROLOGISTS, PSYCHIATRISTS, AND OTOLARYNGOLOGISTS

Hearing loss is an interdisciplinary problem. In a study conducted in Israel involving children aged 2–12 years referred for hearing assessment for various reasons, it was found that if a child was referred to a doctor due to suspected hearing loss, the likelihood of an abnormal audiometric result was 54% [27]. Parents, doctors, speech therapists and teachers were among those who raised concerns about hearing loss [27]. Pediatricians and family doctors have direct contact with children and their parents during vaccinations, conservative treatment or periodic developmental assessments [28]. They should verify parents' observations about a child's hearing impairment, delayed speech development and speech defects. Chronic otitis media with effusion is a common cause of conductive hearing loss. In this case, parents of older children may report that the child understands speech well over the phone, hears better in noise than in silence, and that increasing the volume of the television or radio improves their understanding of speech [29]. The most common causes of sensorineural hearing loss are genetic and acquired factors [30]. In the interview, parents may report

TABLE 2. The diagnostic-therapeutic-rehabilitative path of cochlear implant

Preoperative procedures [14]	Perioperative procedures [14]	Postoperative procedures [14]	Rehabilitation and assessment after surgery [14]
Medical examinations • examination by an otolaryngologist/ audiologist • referral, if necessary, for MRI, CT scans, X-rays, assessment of vestibular function, genetic consultation, ophthalmological examination, pneumococcal vaccination • discussion of all preoperative and postoperative risks associated with the procedure • obtaining parental consent for the procedure Audiological examination • otoscopic examination • tonal audiometry • otoacoustic emissions • auditory brainstem response and/or auditory steady-state response • impedance audiometry • discrimination of sounds in quiet and in noise • questionnaire for parents regarding the child's hearing-related behaviors • examination and assessment for hearing aids • videonystagmography and rotary chair testing to evaluate head dizziness Assessment of the benefits of hearing aids • fitting the child with bilateral hearing aids to provide the broadest possible range of sound perception Assessment of communication methods • assessment of the child's speech reception and expression abilities based on age through observation and testing procedures Psychological support for the family • referral to a psychologist/psychiatrist Family support and education • support organizations • gathering information about supporting organizations, charities, self-help associations, and devices and services for individuals with hearing impairments	Surgeon's responsibilities • making every effort to preserve the internal structures of the cochlea and any residual hearing in the child while protecting the facial nerve from damage (utilizing intraoperative facial nerve monitoring) • employing surgical techniques that are consistent with the latest knowledge and medical technology in the field • considering performing intraoperative or postoperative imaging to assess the position of the implant capsule and/or electrode • monitoring the child's progress during the postoperative period and addressing any surgical or medical issues that may arise in connection with the implant Stages of the procedure [19] • incision behind the ear and creation of a skin-muscle flap • mastoidectomy • posterior tympanotomy • cochleostomy • securing the internal portion of the implant to the bone • suturing the skin	Before discharging the child from the hospital, parents should • receive written information about the proper wound/ ear care and pain management • receive written instructions on what to do in case of medical or surgical issues • be aware of the arrangements regarding follow-up appointments and further procedures • receive guidelines on the safety and hygiene of using the cochlear implant, along with written manufacturer recommendations on safety Starting and adjusting the speech processor after the operation • as soon as the postoperative wound is sufficiently healed	After the operation, the child must be examined by an otolaryngologist and should have the opportunity for additional consultations if needed The child should have access to long-term care (annual medical consultations, implant and speech processor function checks) The number of (re) habilitation sessions offered should be sufficient to achieve optimal benefits from the implant Parents and children must have easy access to the cochlear implant center for (re)habilitation and counseling when needed Regular measurements should be taken at intervals to monitor changes in audiologic results, speech perception, language and speech development, as well as the child's academic achievements

CT – computed tomography, MRI – magnetic resonance imaging, X-rays – radiation

that the child can hear but not understand, that there has been a decline in speech comprehension in noise, and an unpleasant sensation of very loud sounds in the ear with hearing loss [29]. During the visit, attention should be paid to the yellow certificate in the child's health booklet, as the presence of risk factors for hearing loss (Table 1) in a child may lead to delayed-onset hearing loss [31]. Therefore, if the child is not yet under the care of level II and III reference centers of the UNHSP, they should be referred as soon as possible to complete audiological diagnostics. Before referring a patient to a speech therapist for delayed speech development and speech defects, hearing diagnostics should be carried out in a laryngological outpatient clinic. In some cases, children should also be evaluated by a psychologist, neurologist, and sometimes a child psychiatrist. In searching for the causes of this disorder, hearing loss, intellectual disability or autism spectrum disorders should be taken into account [32].

After excluding hearing loss, the child can start intensive speech therapy [32].

PARENTS' PERSPECTIVE

Effective collaboration with the parents of a patient is a crucial element in the process of cochlear implant candidacy assessment, as well as treatment and rehabilitation. It is important to understand the parents' expectations, their education, involvement, and logistical possibilities related to the family's place of residence. A study assessed parents' expectations regarding communication, speech hearing, and speech development of their children based on a survey before implantation and 1, 2, and 3 years after implantation [33]. The results indicated that preoperative expectations were met or exceeded in each time frame in a given category [33]. Parents' satisfaction with cochlear implants is also influenced by factors related to their place of residence. Studies in Poland have shown that as many as 74% of respondents live more than 100 km away from the implantation center, and 52% of families need more than 3 hours to reach the implantation center [34]. Consequently, 56% of respondents express a willingness to use the services of a center located closer to their place of residence [34]. One solution to this problem is remote fitting and adaptation of the speech processor, also known as 'telefitting'. It involves specialists from implanting facilities using the Internet to organize a teleconference, enabling both audio and visual communication with the patient and supporting staff in collaborating clinics across the country [35]. Additionally, remote desktop software makes it possible to take control of a distant computer, facilitating measurements and adjustments to the speech processor [35]. A study involving 20 cochlear implant users who underwent remote fitting showed that 90% of the respondents found this procedure not difficult; for most, the audio and video quality was good, and 85% of the individuals wanted to use online fitting again [36]. This solution saves time and reduces the costs associated with travel and work absence, and children do not have to miss school. The awareness of the consequences of early implantation in a child is influenced by the education of the parents, especially the child's mother. Research in California indicated that an increase of 1 percentage point in the number of mothers who had completed high school was correlated with a 5 percentage point increase in the number of children aged 2 and younger receiving implants [37]. It is worth noting that among the reasons for not using cochlear implants in children, the lack of adequate family support was mentioned [24].

CONCLUSIONS

The implant program, including patient assessment, treatment, and rehabilitation, is a complex and lengthy process that offers children with bilateral deafness a chance for proper speech development. In the observed situation of insufficient coverage of high-risk children with hearing screening, knowledge of its principles enables physicians to educate parents. It is also important to understand and appreciate the family factors that influence the course of implantation. The efforts of specialists can influence parents' decisions to seek treatment, providing children with a chance for proper speech development in the future.

DISCLOSURES

1. Institutional review board statement: Not applicable.
2. Assistance with the article: None.
3. Financial support and sponsorship: None.
4. Conflicts of interest: None.

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