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Review article

COCHLEAR IMPLANTATION IN CHILDREN WITH HEARING IMPAIRMENT

Abstract

A cochlear implant is a surgically implanted device used for hearing rehabilitation of adults and children with severe to profound sensorineural hearing loss and poor speech discrimination who gain limited benefit from conventional hearing aids. There is growing evidence that early application of a cochlear implant in children affected by profound hearing loss is of the greatest importance for the development of adequate auditory performance and language skills.

The indication for cochlear implantation in children is bilateral sensorineural hearing loss > 80 dB determined on the basis of hearing tests. In congenital deafness, after completing diagnosis, the cochlear implant should be placed at the age of 12 months.

When the desired outcome of cochlear implantation is to develop listening and spoken language skills, intensive speech and language therapy is necessary. Although services differ based on each child's current level of performance, it is recommended that children receive auditory-based therapy after implantation to maximize benefit from the cochlear implant. Whatever approach is selected, rehabilitation after cochlear implantation is a long process. Parents and family members have a big role in development of spoken language of the child.

Keywords: *COCHLEAR IMPLANT, CHILDREN, EARLY INTERVENTION*

Introduction

A cochlear implant (CI) is a surgically implanted device used for hearing rehabilitation of adults and children with severe to profound sensorineural hearing loss and poor speech discrimination who gain limited benefit from conventional hearing aids (Deep et al., 2019). The first single channel cochlear implant was introduced in 1972. Over 1000 people were implanted from 1972 to the mid 1980s including several hundred children. This early single channel device was well tolerated and provided many users with significant speech reading enhancement. The first multi-channel cochlear implant system was introduced in 1984 (ASHA, 2004).

There is growing evidence that early application of a CI in children affected by profound congenital hearing loss is of greatest importance for the development of adequate auditory performance and language skills (Colletti

et al., 2005). Childhood hearing loss not only affects speech and language development, but also cognitive, social, and emotional development (Albert, 2007). Sensorineural hearing loss affects 1 to 3 of every 1000 children born in developed countries. The rate is probably higher in the developing countries (Papsin and Gordon, 2007). Despite the differences in the definition of permanent hearing loss in childhood, most retrospective studies have shown a prevalence of 1.1 to 1.7 per 1000 children (Davis, Davis and Mencher, 2009). By etiology, 50% of children with congenital sensorineural hearing loss were influenced by genetic factors. Of these, 15% have syndromic and 35% have non-syndromic hearing impairment. The most common form of syndromic hereditary sensorineural hearing loss is Pendred's syndrome (Smith, Bale and White, 2005).

Early hearing loss intervention via cochlear implantation has a positive effect on the speech and language development of children. A potential sensitive period exists for implantation before 12 months of age. These outcomes support the recent trend toward early cochlear implantation in pre-lingually deaf children (May-Mederake, 2012). Cochlear implants can provide effective auditory stimulation and enable early auditory development of children with profound hearing loss. Children implanted very early (up to 12 months) develop faster than children implanted between 12 and 24 months of age. Similarly, children with residual hearing before implantation do better than children who did not have the benefits of the hearing aids (Lorens, Obrycka and Skarzynski, 2021). For children with post lingual single-sided deafness, CI surgery presents the only opportunity to restore binaural hearing abilities (Arndt et al., 2015).

Universal newborn hearing screening for early identification of children with hearing loss

Screening for hearing loss in newborns is based on two concepts. First, a critical period exists for optimal language skills to develop, and earlier intervention produces better outcomes. Second, treatment of hearing defects has been shown to improve communication. Automated auditory brainstem response (AABR) and otoacoustic emissions (OAE) test and are tests used for screening (Wrightson, 2007). Screening protocols at the first stage of the Universal newborn hearing screening (UNHS) programs can be classified into four categories:

1. *AABR only* – both neural and cochlear hearing losses are detected using one type of technology;
2. *OAEs only* – do not detect neural hearing losses;
3. *OAE followed by AABR* – when the OAE is not passed, OAE screening is completed on both ears first, AABR is only done for those newborns that do not pass the OAE screen. If one or both ears do not pass

the AABR, the infant is referred for outpatient diagnostic testing;
4. *Both AABR and OAE* – newborns must pass both an OAE and an AABR screening. The newborn who fails one or both screening in one or both ears is referred for outpatient diagnostic testing (Wroblewska-Seniuk et al., 2017).

A study of assessment of hearing screening programs across 47 countries or regions found established NHS programs in 42 countries. Five countries did not have screening programs: Albania, Kosovo, Malawi, Rwanda and Montenegro. NHS programs in India varied and were not nationwide. North Macedonia and Malta had selective, not nationwide NHS programs. Universal NHS but not nationwide was found in the following countries: Romania, Bosnia and Herzegovina, Serbia, Moldova, Czechia, China, Greece and Italy. Thirty-one country had nationwide universal NHS programs: Latvia, Lithuania, Faroe Islands, Estonia, Belgium, Russia, Austria, Croatia, Poland, Luxembourg, Spain (Autonomous community Valencia), Bulgaria, Portugal, Slovakia, Slovenia, Switzerland, Spain (Asturias), Hungary, Cyprus, Israel, Denmark, Belgium, France, Germany, Iceland, Netherlands, Sweden, Finland, Ireland, England (UK), and Turkey (Bussé et al., 2021).

A survey on the global status of newborn and infant hearing screening showed that 38% of the world's population were reported to have no/minimal screening, 33% reported screening of more than 85% of the babies. A total of 158 countries provided information. Average age at diagnosis of permanent childhood hearing loss (PCHL) was 4.6 months for screened children and 34.9 months for non-screened children. Average age at start of intervention was 6.9 months for screened children and 35.2 months for non-screened children. Methods used for screening included OAE in 57% of countries, AABR in 11%, and two-step OAE-AABR in 30%. On average, 4.5% of the infants failed the screening and 17.2% of those children were reported as lost-to-follow-up. The prevalence of PCHL identified in hearing screening programs ranged from 0.3-15.0 per 1000 infants with a median of 1.70 (Neumann et al., 2020). Early diagnosis and intervention in hearing loss are crucial in order to provide access to sound and increase the likelihood of spoken language development in pre-lingual deaf children. Children referred via UNHS are referred and implanted at a younger age (Gabriel et al., 2020).

The way the cochlear implant works

The cochlear implant consists of external parts and implanted device. A microphone, processor and transmitter are worn externally to control the implanted internal device. The parts of the CI and the way the implant works are displayed in Figure 1:

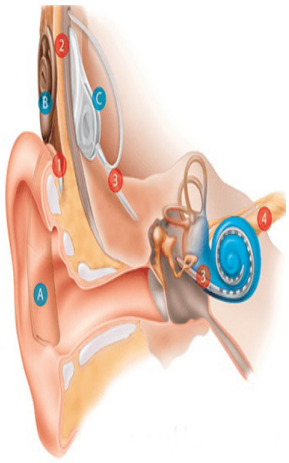

Figure 1. *Parts of the cochlear implant and the way of working*

1. A microphone captures the sound and a sound processor (A) analyses and encodes sound into digital code.
2. The sound processor transmits the digitally coded sound through the coil (B) to the implant (C) just under the skin.
3. The implant converts the digitally coded sound to electrical signal and sends them along the electrode array, which is positioned in the cochlea.
4. The implant's electrodes stimulate the cochlea's hearing nerve fibers, which relay the sound signals to the brain to produce hearing sensations (MedStar Health, 2022).

While the cochlea has several thousand inner hair cells responsible for detection of sounds and stimulation of the auditory nerve, most cochlear implants have around 22 electrodes, and therefore sound quality is inevitably different, due to the loss of the detail that the fine structures of the cochlea provide in normal, auditory hearing. A child with CI therefore must learn the patterns of sound that are stimulated by the implant's electrodes, and understand how to interpret them (Jaffer, 2017).

Indications for cochlear implantation

The treatment with a cochlear implant is the gold standard in therapy of patients with profound hearing loss or deafness and the medical indications for this treatment has continuously been broadened (Loth et al., 2022). The indication for cochlear implantation in children is bilateral sensorineural hearing loss > 80 dB determined on the basis of hearing tests, after approx-

imately 6-month rehabilitation with the use of hearing aids. In congenital deafness, after completing diagnosis, the cochlear implant should be placed at the age of 12 months (except for children with congenital malformations requiring another specialist examination). In some children with significant hearing loss, one must remember about the maturation of the auditory pathway, which may lead to improvement of hearing at a further period of time, which is why they require close monitoring and rehabilitation with repeated objective tests. After stabilization of response, the decision about implantation should be made, which should take place in the second year of life at the latest. In the case of asymmetric hearing loss, the inferior ear should be implanted. In children, unilateral implantation should be combined with prosthetisation of the other ear with hearing aid (Szyfter et al., 2019).

Cochlear implantation should be considered a priority for children at risk of hearing loss progression in the better hearing ear. Children with single-sided deafness due to bacterial meningitis should be implanted promptly (Park et al., 2022).

Cochlear implant candidacy

The pure tone threshold criteria for cochlear implantation vary across manufacturers, but most cochlear implant teams in the United States will consider a patient as a candidate for a CI of any of the manufactures providing CIs when the patient meets indications for one of the devices. For *Cochlear Nucleus* cochlear implants the Food and Drug Administration (FDA) approved guidelines indicate that candidates with moderate to profound low-frequency hearing loss and severe to profound mid- to high-frequency loss should be considered for cochlear implantation. The criterion does not clearly define the frequencies that make up the low-, mid-, and high-frequency ranges. *Advanced Bionics* indications call for a severe to profound bilateral sensorineural hearing loss, whereas *MED-EL* calls for a pure-tone average (500, 1000, and 2000 Hz) of 70 dB HL or poorer (Wolfe, 2020).

American Cochlear Implant Alliance Task Force recommended a 50/70+ guideline for referral to pediatric CI candidacy. That is, clinicians should refer pediatric patient for evaluation if they meet any of the following criteria: appropriately selected word recognition scores <50% correct; unaided pure-tone thresholds > 70 dB HL; or poor functional performance, limited progress in language or auditory development, or poor quality of life (Warner-Czyz et al., 2022). Referral for consideration of cochlear implant has evolved over the years. Referral criteria for cochlear implants for children under 5 years were the following:

- Bilateral sensorineural hearing loss of > 90 dB HL at 2 kHz and 4 kHz;
- Click ABR thresholds at > 90 dB;

- No minimum age of referral;
- Children with additional need will always be considered;
- Parental consent for referral obtained.

Referral criteria for children 5 years and over were:

- Children with sudden onset or progressive hearing loss;
- Bilateral sensorineural hearing loss of > 90 dB HL at 2 kHz and 4 kHz;
- Children whose primary form of communication is speech;
- Children with additional needs will always be considered;
- Parental consent for referral obtained (Owens et al., 2006).

Contraindications for cochlear implantation

There are absolute and relative contraindications for cochlear implantation. Contraindications may include:

- Deafness due to lesions of the eight cranial nerve or brain stem;
- Chronic infections of the middle ear and mastoid cavity or tympanic membrane perforation;
- Cochlear aplasia as demonstrated on CT scans;
- Certain medical conditions that preclude cochlear implant surgery (eg. specific hematologic, pulmonary, and cardiac conditions);
- The lack of realistic expectations regarding the benefits of cochlear implantation and/or a lack of strong desire to develop enhanced oral communication skills (Megerian, 2022).

According to American Cochlear Implant Alliance Task Force Guidelines for clinical assessment and management of cochlear implantation in children with single-sided deafness, cochlear implantation to address single-sided deafness in an ear with cochlear nerve deficiency is contraindicated. Accurate diagnosis of nerve deficiency is important because it is present in almost half of children with single-sided deafness. Therefore, high resolution 3D magnetic resonance imaging (MRI) of the internal auditory canals is recommended rather than computer tomography alone (Park et al., 2022).

Preoperative diagnostics for indication of cochlear implantation

Preoperative diagnostics should include the following:

- Ear inspection, paracentesis if needed and tympanostomy tubes;
- Subjective audiometry (behavioral observation audiometry, play audiometry);
- Objective audiometry (otoacoustic emissions, impedance audiometry, objective determination of the hearing threshold in high and low frequencies by means of brainstem evoked response audiometry – BERA, slow brainstem response – SN10, frequency following

response – FFR, and amplitude modulation following response – AMFR, electrocochleography, and electrically brainstem evoked response audiometry – EBERA for functional testing of the hearing nerve);

- Imaging in cases of proven hearing loss (high-resolution computer tomography – CT, cone beam tomography – CTB of the temporal bone, assessment of the bony structures of the temporal bone, sizing of the cochlea);
- High-resolution magnetic resonance imaging – MRI of the temporal bone (evaluation of the inner ear structures, identification of the hearing nerve aplasia, hypoplasia);
- MRI of the central hearing pathway (neoplasms, developmental disorders, trauma sequelae);
- Functional imaging with positron emission tomography – PET, functional magnetic resonance imaging – fMRI, and near-infrared spectroscopy – NIRS for identification of an increased activity in the area of the auditory cortex under electrical stimulation;
- Genetic diagnostics;
- Pedagogical assessment of the status of hearing and speech development;
- Neuropediatric examinations (Lenarz, 2017).

Candidacy test batteries should include age-appropriate behavioral assessment and cross-check, spatial hearing assessment in the child's everyday listening conditions, and relevant subjective questionnaires. Recorded aided word recognition testing with contralateral masking should be completed preoperatively if the child uses traditional amplification and/or if required by insurance (Park et al., 2022). Visual reinforcement audiometry (VRA) may be used to measure threshold-level responses in infants and children between the developmental ages of 6 and 30 months (Wolfe, 2020). In order to be able to differentiate the beneficial information obtained from MRI and high-resolution CT, some authors suggested a combination of the diagnostic imaging tools with audiological tests for the evaluation of the integrity of the hearing nerve (Arndt et al., 2015).

Cochlear implant surgery

The cochlear implantation comprises stepwise drilling of the temporal bone followed by insertion of electrodes into the scala tympani (Frendø et al., 2021). Surgery begins with administration of general anesthesia. Hair is shaved above and behind the ear, the skin is prepared with an antiseptic solution, and sterile drapes are placed around the ear. A post auricular incision is made, and a well is created in the skull behind the mastoid bone to accommodate the receiver-stimulator portion of the internal device. The

surgeon drills through the mastoid air cells and removes bone between the tympanic membrane and the facial nerve until the round window and the cochlear promontory are visualized. An opening (cochleostomy) is made into the basal turn of the scala tympani just anterior to or through the round window, and the electrode array is inserted into the scala tympani. If a ground electrode is attached to the receiver, it is then placed under the temporalis muscle. The receiver-stimulator is placed and secured into the well behind the mastoid, the incision is closed, and a pressure dressing is placed over the ear for 24 hours. Some surgeons may forego the use of the well and instead place the implant receiver in a tight pocket of skin behind the mastoid (Zwolan, 2015).

In implanting the electrode array and the receiver-stimulator package, great care must be taken, as there are more nerves and vessels concentrated in a small area of the temporal bone than elsewhere in the body. The mastoid bone is partly filled with air cells which provide space for the placement of the receiver-stimulator package and lead wires. Nevertheless, just behind the mastoid air cells, the skull often needs to be drilled down to the dural lining of the brain to accommodate the package without it protruding too far above the surface of the skull, and so producing a bulge. Partial removal of the air cells provides a route from behind the ear to the middle ear, and thence to the inner ear. The skin must be closed over the receiver-stimulator package thus not leaving a path for the entry of infection (Clark, 2003).

Some special considerations are needed when implanting young children because surgical intervention with this age group requires specific knowledge of temporal bone anatomy and the impact of skull growth on the implanted device. CI surgery typically lasts between 2 and 5 hours depending on the surgeon's experience, the device selected, and the complexity of the anatomy encountered in each patient (Zwolan, 2015).

Risks from the cochlear implant surgery

Cochlear implantation has the same risks as other procedures conducted under general anesthesia and those of other surgeries of the middle or inner ear. Risks include a remote possibility of infection, temporary or permanent facial paralysis on the operated side, mild temporary taste disturbances, tinnitus, and vertigo. In traditional CI surgery, one may expect loss of any residual hearing in the implanted ear, as well as mild pain and numbness at the side at the incision following the surgery. Complications require an adequate management that must be controlled by the cochlear implant surgeon. Continuous improvement of the surgical technique led to a relevant reduction of the complication rates (Lenarz, 2017).

CI recipients must avoid various medical/surgical procedures that could damage the implanted device or the functioning auditory nerve fibers that transmit the electrical signal to the brain. Some procedures can cause

excessive magnetic and electromagnetic interference, which may result in demagnetization of the internal magnet, displacement of the device, and/or disruption of the device electronics. There are commercial CI devices manufactured with a removable internal magnet and may be preferable for patients who are expected to need MRI in the future. There is also CI device which does not have a removable magnet, but is approved for use with MRI at a maximum strength of 1.5 Tesla (Zwolan, 2015).

Counseling for families

Counseling for families should emphasize the importance of neuroplasticity and thus the potential advantage for a younger age at implantation to improve outcomes. Counseling should include a discussion of the importance of post implant listening therapy, full-time use, reasonable expectations, and audiologic follow-up. Counseling for families considering single-sided deafness and CI should include information about developmental disadvantages of single-sided deafness such as inability to develop spatial hearing in the absence of bilateral input, resultant difficulty with localizing sound and hearing in noise, and listening fatigue (Park et al., 2022).

Post implantation evaluation

Post activation test batteries completed at regular intervals should include regular assessment of unaided hearing, validation of audibility from the CI, isolated single-word recognition using direct audio input (DAI), spatial hearing assessment with and without the CI, and relevant subjective questionnaires (Park et al., 2022).

Med-El Corporation used EARS (evaluation of auditory responses to speech) protocol to track the progress of children who use CIs. This collection of tests has varying degrees of difficulty and is appropriate for children aged 1 to 18 years with differing levels of auditory experience and linguistic development. This test battery emphasizes the development of auditory skills (detection, discrimination, identification, recognition, and comprehension) and is administered using only an auditory input (Mendel, 2008).

Rehabilitation of children with cochlear implants

Cochlear implant activation usually takes place 4 weeks after surgery. At that time, the patient will be fitted with the external parts of the device, which requires programming of the external speech processor by the cochlear implant audiologist. During this process, the patient will begin to hear the first sounds generated by the implant (MedStar Health, 2022).

The speech-language pathologist focuses on developing listening skills to facilitate language acquisition. The teacher of the deaf complements the speech-language pathologist by focusing on language through academic development (Soman et al., 2012). The successful hearing rehabilitation of patients receiving CIs is a multi-stage process consisting of a large number of necessary individual steps. These include audiological evaluation, surgery, fitting of the audio processor, hearing training (rehabilitation) and lifelong follow-up of the implanted patient. A study that aimed to identify currently existing CI-related Clinical Practice Guidelines (CPGs) and registries in Europe found CPG in 16 (38%) of the countries. Forty-two countries of the European continent, including the Great Britain, Russia and Turkey were screened. In terms of the population of Europe, close to 80% of people live in countries where a CPG already exists. Macedonia was noted as a country without CPG and CI Registry (Loth et al., 2022).

When choosing rehabilitation method, there are three main pathways that can be adopted:

1. Auditory Verbal Therapy – in this approach the emphasis is placed on the auditory skills development of the child with the implant. Children are encouraged to learn to listen and develop spoken language without the need to rely on sign language or lip reading. The primary goal is to develop spoken language and integration into the hearing community. The academic development goals for the children are to develop their skills so that they are mainstreamed.

2. Auditory Oral Approach – this method encourages children to develop spoken language through use of both verbal and non-verbal means, including lip reading. Development of spoken language and communication skills are necessary for their integration into the hearing community. Academically, the aim is to encourage development of skills that will ensure successful mainstreaming. The language development using this method is through early, consistent and successful use of their cochlear implants accompanied by lip reading.

3. Total Communication – this approach encourages using all means to communicate. The child is exposed to a formal sign language, finger spelling, natural gestures, lip reading and spoken language. The idea of this approach is to communicate and teach in any manner that works and the child is encouraged to use a combination of communication methods e.g. speech and sign. The primary goal is to provide an easy and less restrictive communication method for the children and their parents, family, friends and peers. It is important to understand that therapy, whatever approach is selected, is a long process, and parents and family members have a big role in development of spoken language of the child (Jaffer, 2017).

Conclusion

The cochlear implant is one of the most successful prosthetic devices developed to date. Cochlear implants are the treatment of choice for auditory rehabilitation of sensory hearing loss. They restore the function of inner hair cells by transforming the acoustic signal into electrical stimuli for activation of auditory nerve fibers. The cochlear implant has given back to many children what they lost or given them what they never had. Many children are happy to have it, but many are waiting for it in the "world of silence".

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