Chapter 13
Cochlear Implants

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Introduction

With the implementation of universal newborn hearing screening (UNHS) programs in the United States and the rapid increase in the number of states and birthing facilities conducting UNHS, the average age of identification of deaf or hard-of-hearing (D/HH) newborns has decreased over the last 20 years from approximately 30 to 48 months to 6 months or less. Although infants and young children are being identified earlier, those who are D/HH will likely fall behind their hearing peers in language, cognition, and social-emotional development. D/HH infants who receive intervention before 6 months maintain language development commensurate with their cognitive abilities through the age of 5 years.

Intervention in the forms of hearing aids (HAs), FM systems, and/or cochlear implants (CIs) are the single most important component to help the hearing-impaired child access sound. When fitted appropriately, they will, in most instances, enable the child to maximize their use of residual hearing. If the child is receiving appropriate aural rehabilitation, speech and language can develop at or near an age-appropriate pace.

No assistive device will enable a D/HH child to perform normally in all listening situations. HAs and CIs for children should make speech audible at a comfortable level and provide as many acoustic cues as possible without over-amplifying any sounds, especially loud sounds. Reception of soft speech is particularly important for incidental language learning (which accounts for a very large portion of overall language learning), self-monitoring of speech, and ease of communication in various real-world listening environments.

There is always a need to make evidence-based clinical decisions, but the pace of technological innovation in HAs and CIs has begun to exceed that of supporting research. Today’s advanced features and styles of HAs (noise reduction, directional microphones, receiver-in-the-ear (RITE), open-canal, etc.) are being fitted on children. In the absence of research to support the outcomes of such fittings, every audiologist who fits devices on children and infants has the responsibility to verify those fittings. The same can be said for audiologists who program and maintain the settings/map on a child’s CI; verification and validation of its performance is mandatory.

This chapter will provide an overview of CIs (including components, candidacy, and outcomes with regard to speech/language) for infants and young children.
CIs: The Basics

CIs have electrodes that are placed in the cochlea to stimulate the eighth nerve (nVIII). These electrodes produce electrical currents that induce compound action potentials in nVIII fibers, which are then transmitted to the brain for interpretation. CIs bypass damaged or missing outer hair cells in the cochlea that would normally code sound.

All CIs, regardless of manufacturer, have common components, but there are many variations in the methods used to process sounds, transmit information to the internal implant, and stimulate the electrodes. There are numerous electrode arrays available from each of the manufacturers, including a shortened array used with hybrid CIs (see below).

Internal Components

Implanted components must be biocompatible and not lead to long-term adverse tissue damage.

Receiver-Stimulator

One of the internal components is called the receiver-stimulator, sometimes known as the internal coil, which is implanted in a flattened or recessed portion of the skull—posterior to and slightly above the pinna. This receives power and decodes instructions from the speech processor. It converts the electrical signal into a digital code and converts again to electrical pulses, which are delivered to the electrodes in the cochlea. It receives stimulus information via radio frequency (RF) transmission from the external coil housed in the headpiece. This method of coupling is called a transcutaneous link.

Electrode Arrays

Multichannel devices have up to 22 active electrodes. Research has indicated more electrodes typically result in better speech perception. However, this is not a one-to-one relationship, as many individuals achieve very good speech perception without the use of all the electrodes in their array. An electrode array stimulates residual auditory nerve fibers along the modiolus and in nVIII. CI electrodes are designed for placement in the scala tympani of the cochlea. Keeping the electrodes relatively close to the spiral ganglion cells is best for localized stimulation of the auditory nerve. Different electrodes ideally stimulate different subpopulations of cochlear neurons. Electrode arrays try to mimic the tonotopic organization of cochlea. Neurons near the base of the cochlea (first turn) respond to high-frequency sounds, and neurons in the apex of the cochlea respond to low-frequency sounds.
Successful placement depends heavily upon the skill of the surgeon and whether the electrode array is being inserted via a cochleostomy or through the round window. To ensure appropriate placement of the electrode array, insertion tools are used in the majority of cases. Manufacturers offer multiple electrode array designs, lengths, and features. New electrode arrays appearing on the market are straight, slimmer, have a flexible tip, are shorter, or have any combination of these attributes. The goal is to reduce damage to the cochlea during insertion. Less cochlear damage may correlate with better CI performance (i.e., better speech perception).

A shorter electrode array intended specifically for partial insertion is now available for those patients with normal or moderate low-frequency (up to 500 Hz) hearing and severe hearing loss (70 dB or greater) from 1000 Hz. This electrode array is intended to allow the patient to use electric and acoustic stimulation in the same ear and attempts to preserve low-frequency residual hearing. Patients can then use their natural low-frequency hearing with mid- to high-frequency electrical stimulation. Even persons with low-frequency hearing that would benefit from amplification [hearing aid(s)], but have very poor mid- to high-frequency hearing, may benefit from this hybrid CI (CI and HA in same ear). Please refer to the information on hybrid CIs below for more information.

Double electrode arrays, designed for the ossified cochlea, can be used on children who are post-meningitic. There are also shorter arrays that can be used for post-meningitic ossified cochleae.

Current CIs:
- Are compatible with FM units.
- Have directional or multiple microphones
- Incorporate Bluetooth technology.
- Can be connected to iPods, MP3 players, computers, phones, televisions, gaming systems.
- Have almost limitless ways to program "the maps" through the speech processor.

Stimulating Electrodes

There are two electrode stimulation modes. Each incorporates intricate processes that vary by manufacturer.

**Bipolar.** In a bipolar mode of stimulation, one intracochlear electrode is stimulated with reference to another nearby intracochlear electrode. Current flows between a pair of electrodes, with one serving as the ground electrode. Research has indicated more electrodes typically result in better speech perception. However, this is not a one-to-one relationship, as many individuals achieve very good speech perception without the use of all the electrodes in their array.

**Monopolar.** Monopolar stimulation means that each electrode is stimulated with reference to a ground electrode, which is remote from the cochlea. This remote electrode can be on the internal device or on the end of a silastic tube that extends from the internal receiver/stimulator. The latter design is called a ball electrode and is designed for placement under the temporalis muscle. The monopolar stimulation strategy is often used in CI maps, because the amount of current required to elicit perceptible stimulation is less than in bipolar, which increases battery life. All contemporary CIs use monopolar stimulation as the default mode.

Rate of Stimulation

Current CIs deliver trains of biphasic electrical pulses to the electrode array and contacts within the cochlea. The rate of stimulation defines the number of electrical current pulses per second (pps) that may be delivered to an individual
Manufacturers devote a great deal of attention to developing new and improved processing schemes. Often the new schemes can be incorporated into existing processors via a software update—otherwise processor replacement is necessary.

**External Components**

**Microphone.** The microphone, which is typically housed in/on the speech processor, is a device for picking up and processing incoming sound. It senses pressure variations in a sound field and converts them into electrical variations. These electrical signals are typically sent to a preamplifier to improve the signal-to-noise ratio, providing a boost in the higher frequencies. The microphone has a broad frequency response but minimizes responses to low-frequency vibrations, such as those produced by head and body movements. All manufacturers offer multiple microphones, increasing the selectivity of the directional pattern to aid speech understanding speech in noisy situations. Directional microphones emphasize sounds in front of the microphone and suppress sounds emanating from other directions. All three manufacturers have multiple microphone options available to:

- Reduce wind noise.
- Enhance localization.
- Assist with speech understanding in background noise.

All manufacturers have programs/features to allow the microphone(s) to be self-adjusting to the listener's environment. The microphone sends this modified signal to the external speech processor.

**Speech processor.** The speech processor of a CI uses sound from the microphone to create a set of electrical stimuli for the electrodes. The received signal is analyzed by a digital signal processor (DSP) to separate the input according to intensity, frequency, and time domains, which will be represented at the nVIII. Manufacturers devote a great deal of attention to developing new and improved processing schemes. Often the new schemes can be incorporated into existing processors via a software update—otherwise processor replacement is necessary. Replacement of the internal components is rarely, if ever, necessary to utilize new speech-processing schemes. The speech processor takes the processed electrical signal and transmits it via a cord to the headpiece. The speech processor is powered by batteries—either standard or rechargeable. Typical battery life is greater than 12 hours for a body-worn processor and usually somewhat less for a behind-the-ear processor.

**Headpiece.** The headpiece houses the external coil of the CI and is held in place over the internal receiver/stimulator (internal coil) with magnets. The headpiece transmits the electrical signal, after converting it to an electromagnetic signal, to the internal receiver-stimulator via radio frequency (RF). The RF coil and its signal also serve as the power supply for the internal stimulator.
Creating a Map: The Basics

Two psychophysical measures are used to create a program or map: thresholds (T levels) and comfort/maximum levels (C levels or M levels, depending on manufacturer). Ts are minimal stimulation levels—or the softest sound that can be reliably identified by the patient 100% of the time. C/Ms are maximum stimulation levels—the loudest sound that can be listened to comfortably for a sustained period of time. Obtaining these two measures for each electrode is desirable, although current CI software allows for one or both of these measures to be foregone. For children (and adults, when measured), methods of determining these levels are similar to those used for diagnostic audiology. For children, this could be visual reinforcement audiometry, conditioned play audiometry, or the typical “raise your hand” voluntary responses.

In the absence of both T and C/M measures, the map may be created using live voice. This method is more commonly used for adult patients with previous hearing experience. Upper limits are often set by increasing stimulation levels to the patient’s most comfortable listening level while listening to live speech. For infants, very young children, or individuals who cannot respond behaviorally, evoked stapedial reflex threshold (ESRT) testing is highly recommended to set upper-stimulation levels. Telemetry can also be used to assist in the creation of a map.

Telemetry

Telemetry is the exchange of information from the external components of the CI through a transcutaneous link (RF waves) to the internal components. Bidirectional exchange of information allows transmission of data from the implanted components to the external coil and speech processor. Telemetry can provide information about the status of the implanted receiver, impedances of implanted electrodes, and voltages of unstimulated electrodes. It also offers the opportunity to record evoked potentials by stimulating nerve fibers to elicit compound action potentials. Voltage generated by an active electrode can be measured to help determine the state of the cochlea in that region. Measurement of electrode impedances is a routine procedure done immediately after implantation, as well as during every subsequent visit where programming or reprogramming of the CI is necessary.

Telemetry is called something different by each CI manufacturer. Neural Response Telemetry (NRT) is the term used by Cochlear Corporation, Neural Response Imaging (NRI) is the term used by Advanced Bionics Corporation, and Auditory Neural Response Telemetry (ANRT) is the term from MED-EL Corporation. For purposes of this chapter, all will be referred to as telemetry.

Using telemetry, compound action potentials of the nVIII can be generated, which is an indication of how much neural activity stimulation is causing. This information can be used to estimate threshold and comfort/maximum stimulation levels. Evoked compound action potentials (ECAPs) can provide an objective and noninvasive measure of neural function. The ECAP produces a waveform, usually with 2 peaks and 1 major trough labeled P1, N1, and P2. ECAPs are stimulated on multiple electrodes. Each electrode will have a threshold established by eliciting multiple ECAPs using a threshold-seeking method. This information is used to assist in creating a map for the patient. Research has demonstrated the ECAP thresholds often fall somewhere between Ts and M/Cs, usually closer to the M/C levels.

ECAP waveform. The amplitude of the ECAP defined as the voltage difference between N1 and P2.
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**Bilateral CIs**

The number of bilateral CI users worldwide is increasing. This is not unexpected. We are born with two ears, and we hear better listening with both. Bilateral CIs can be provided in the same surgery (simultaneous) or sequentially (two separate surgeries). Simultaneous implants are usually considered for patients who receive no benefit from acoustic amplification or have had meningitis. A concern with simultaneous implantation is keeping the patient under anesthesia for a prolonged period of time. Sequential implantation is best for children under the age of 8. Research has demonstrated that recipients older than 8 find integrating two implants more difficult, unless they have been wearing an HA on the other ear.

There are multiple advantages to hearing with two CIs. Some of the benefits include:

- Better localization of sound—hearing in "surround sound."
- Better hearing of speech in background noise.
- Binaural summation (sound is louder with two ears).
- Decreases impact of head-shadow effect.
- Keeps the auditory pathways stimulated—“use it or lose it.”
- Listening with less effort (less tiring, improved concentration).
- Improved music appreciation.

Subjective reports indicate that overall quality of life improved with two implants when recipients compared themselves when using only one CI. Some research indicates that wearing a CI on one ear and a HA on the other provides some of the benefits mentioned above.

Many studies have been done with adult bilateral CI recipients in controlled environments as well as in everyday listening situations. Little research has been completed with children who are implanted bilaterally. A few studies on children have been done in controlled environments, not in the “real world.” However, there is no reason to believe that the benefits afforded adults with bilateral implants would not also be available to children with two CIs.

The current standard of care for newly identified infants and children with hearing loss is to recommend bilateral implants when all other candidacy criteria have been met.

**Hybrid CIs**

The purpose of a hybrid CI is to provide electrical stimulation to the nVIII for high-frequency sound input while preserving the low-frequency residual hearing of the user. Hybrid CI arrays are shorter and narrower than conventional electrode arrays. These electrode arrays are designed for lateral wall placement as opposed to modiolar hugging. The external sound processor of a hybrid system contains an acoustic component to deliver amplification for the lower frequencies. Some users wear an in-the-ear hearing aid with a conventional CI to amplify the lower frequencies, although this is less common.

Cochlear Corporation and MED-EL offer hybrid CIs, but only Cochlear Corporation has FDA approval for use in the U.S. Recipients must be 18 years of age or older. However, children have been implanted with a hybrid CI successfully in Europe.
Candidacy

Determination of candidacy for a CI requires assessment of patient suitability based on many factors. Critical information that must be understood by all potential recipients or their family is that a CI is a communication device and not a cure for hearing loss. Preoperative expectations significantly shape postoperative satisfaction!

Communication disorders, especially in children, require a multifaceted rehabilitation program. Processing, speech, language, cognition, and attention are among the areas that need to be addressed. Regardless of manufacturer, CI users overall perform equally well on measures of speech perception. For adults, three CI companies support and recommend a minimum speech test battery (MSTB, 2011). The MSTB materials consist of an audio CD with the:

- AZBio sentences presented in quiet and noise.
- CNC word test.
- BKB-SIN test.

The recommended presentation level for adults receiving the MSTB materials is 60 dBA, which is comparable to the level of conversational speech. The same level is recommended for children who are undergoing speech perception testing.

Table 1 shows these three manufacturers’ CI candidacy criteria.

Auditory Neuropathy Spectrum Disorder (ANSD)

Cochlear implantation in some patients with ANSD has been successful. For some children with ANSD, mild gain amplification and/or FM units have proven successful. It must be noted that each child with ANSD must be treated on an individual basis. Recommendations for amplification in the form of HAs, FM system, CIs, or any combination thereof must be made on a case-by-case basis. A full discussion of ANSD is beyond the scope of this chapter. For further information and references regarding ANSD, see the appropriate chapter in this publication.

The Role of Amplification in CI Candidacy for Children

All three CI manufacturers recommend that prior to implantation for infants and young children, no benefit from appropriately fitted HAs nor progress in auditory skills with amplification must be demonstrated. “Appropriately fitted” is defined using aided speech perception performance. Tests of speech perception and parent questionnaires are used in pre-implant assessment. All companies agree that if a child is post-meningitic, the hearing aid trial, as well as speech perception testing, may be waived due to the threat of ossification.

Manufacturer candidacy and the subsequent U.S. Food and Drug Administration (FDA) guidelines were created to protect the patient from any unnecessary invasive procedures. These guidelines are regularly modified and not always adhered to strictly. Research has demonstrated conclusively that the younger a child is implanted, the greater
the chances of success in multiple areas, such as academics, self-esteem, speech perception and production, etc. Speech perception performance in CI users overall has exceeded any expectations of the audiology profession. Bilateral CIs have significantly improved these scores. These very positive outcomes have swayed pediatric audiologists at times to recommend a CI prior to completion of hearing aid trial. The exceptional performance CI users are exhibiting creates a sense of urgency to implant young children with a severe-to-profound hearing loss, bypassing an extended trial with amplification.

Summary

Fitting HAs and/or CIs on infants and young children is a vital function of being a pediatric audiologist. It is critical that all audiologists working with children have exceptional knowledge of both HAs and CIs. Pediatric audiologists have a responsibility to ensure that all assistive devices are appropriately fitted and maintained. It must be the ultimate goal for each child to receive maximal benefit from their assistive technology and be equipped to lead a healthy, productive life, including:

- The best possible speech perception and production.
- Academic success.
- Emotional adjustment.
- Social competence.
- Occupational preparation.

It must be the ultimate goal for each child to receive maximal benefit from their assistive technology and be equipped to lead a healthy, productive life.
**Table 1**
Manufacturers’ CI Candidacy Criteria

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<th>MED-EL</th>
<th>Advanced Bionics Corporation</th>
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**A CI is designed for:**
- Children with severe to profound sensorineural hearing loss in both ears. Age at implantation may be as young as several months, depending on individual circumstances and local practices.
- Individuals who receive little or no benefit from HAs.
- Individuals with access to education and rehabilitation follow-up programs.
- Individuals with no contraindication preventing surgery.

To gain the best benefit from a CI, it is important for children to have the full support of their family and to participate in rehabilitation programs. This is vital to ensure that your child obtains the best hearing experience using the implant. If a family cannot commit to participation in a rehabilitation program, alternative options to a CI may need to be considered.

**Real-world experiences that might suggest the need for a CI evaluation.**

**With HAs:**
- Cannot understand most phone conversations.
- Difficulty understanding conversations in groups or noisy places.
- Rely heavily on speech reading.
- Limited social, educational, or professional life options.

**Adults**
These are the CI candidacy requirements. However, determination of CI candidacy is based ultimately on the clinical judgment of the CI team and whether or not a CI would be in an individual’s best interest.
- Severe-to-profound sensorineural hearing loss in both ears.
- Limited benefit from HAs noted by a score on sentence tests of equal to or less than 50% in the ear being considered for implantation.
- An adult who is 18 years of age or older with no medical contraindications.

**Pediatrics**
Similar to candidacy in adults, the determination for a CI is based ultimately on the clinical judgment of the CI team and the best interests of the child.
- The child is 12 months of age or older and experiencing a profound bilateral sensorineural hearing loss.
- The child needs to have completed a trial period with appropriately fitted HAs, unless otherwise medically indicated.
- Limited benefit from amplification needs to be established based on age-appropriate measures of auditory and speech development.

**Real-world experiences that might suggest the need for a CI evaluation.**

**With HAs:**
- Delayed or lack of speech and language development.
- Rarely responds to name.
- Lack of social interaction with children and adults.
- Emphasis on auditory development in child’s education or therapy environment.
Infants (12 to 23 months)
- Profound bilateral sensorineural hearing loss.
- No progress in auditory skill development with HAs and intervention. (Limited benefit is defined as lack of progress in the development of simple auditory skills in conjunction with appropriate amplification and participation in intensive aural habilitation over a 3- to 6-month period. It is recommended that limited benefit be quantified on a measure, such as the Meaningful Auditory Integration Scale or the Early Speech Perception test.)
- No medical contraindications.
- High motivation and appropriate expectations from family.

Children (25 months to 17 years/11 months)
- Severe-to-profound sensorineural hearing loss in both ears.
- Multisyllabic Lexical Neighborhood Test (MLNT) scores of 30% or less in best-aided condition (children, 25 months to 4 years/11 months).
- Lexical Neighborhood Test (LNT) scores of 30% or less in best-aided condition (children, 5 years to 17 years/11 months).
- Lack of progress in the development of auditory skills.
- No medical contraindications.
- High motivation and appropriate expectations (both child when appropriate and family).
- Pre- or post-linguistic onset of hearing loss.

Adults
Nucleus CIs are intended for use in individuals 18 years of age or older who have bilateral pre-, peri-, or post-linguistic sensorineural hearing impairment and obtain limited benefit from appropriate binaural HAs.
- Moderate-to-profound bilateral sensorineural hearing loss.
- < 50% sentence recognition in ear to be implanted (aided).
- < 60% in best listening condition—binaural (aided).
- No medical contraindications.
- A desire to be part of the hearing world.
References


