Chapter 10
Cochlear Implants

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The average age of identification of deaf or hard-of-hearing newborns has decreased over the last 20 years from approximately 30 to 48 months to 6 months or less.

Introduction

With the implementation of universal newborn hearing screening (UNHS) programs in the Unites 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 for infants and young children. Candidacy considerations will be discussed in another chapter of this publication.

NCHAM National Center for Hearing Assessment and Management Utah State University™

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 the radio frequency (RF) 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.

Placement closer to the modiolus requires less current to achieve a response from the auditory nerve and in turn requires less power for loudness. This placement may also produce less channel interaction. Post-CI hearing thresholds are thought to be better when the electrodes are closer to spiral ganglion cells due to more localized current flow. One way to get an electrode array to lie closer to the modiolus is to insert a pre-curved array. However, not all available electrode arrays are pre-curved.

Lateral wall electrodes are thought to be less traumatic for cochlear structures in the scala media. A recent focus of the CI manufacturers is attaining atraumatic insertion of the electrode array. Some arrays are touted as more atraumatic than others. If the basilar membrane or spiral lamina are not damaged (or infection does not occur), electrodes can be inserted without causing a significant loss of auditory neurons. A straight electrode array may cause trauma to the cochlea during insertion, but this is certainly not the case in all instances.

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The goal is to reduce damage to the cochlea during insertion. Less cochlear damage may correlate with better CI performance.

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 (EAS) 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 incoroporates 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 that 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

electrode contact. Early devices had relatively slower stimulation rates (250 pps or less), but current devices can deliver up to as many as 5,000 pps. Higher rates (above 2,000 pps) improve the representation of temporal information by providing finer amplitude variations through greater control of the rate and population of nerves excited. While there is much research to demonstrate consistent improvements in patient performance with rates >2,000, there is little research to support that rates above 2,000 pps provide better speech recognition. The optimal stimulation rate varies on an individual basis.

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 RF. The RF coil and its signal also serve as the power supply for the internal stimulator.

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Telemetry offers the opportunity to record evoked potentials by stimulating nerve fibers to elicit compound action potentials.

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.

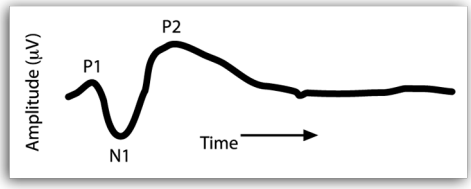
In the absence of both T and C/M measures, the map may be created using live voice. This method is more commonly used for adult patients with previous hearing experience. Upper limits are often set by increasing stimulation levels to the patient's most comfortable listening level while listening to live 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

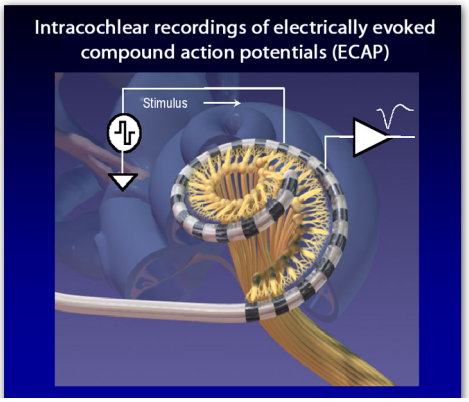
Telemetry is the exchange of information from the external components of the CI through a transcutaneous link (RF waves) to the internal components. Bidirectional exchange of information allows transmission of data from the implanted components to the external coil and speech processor. Telemetry can 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.*



Intracochlear recordings of electrically ECAPs.

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

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.

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.

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! More detailed information regarding candidacy can be found in the appropriate chapter of this publication.

Auditory Neuropathy Spectrum Disorder (ANSD)

Although none of the CI manufacturers specifically discuss patients with 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 (see *Table 1* for the CI components of three manufacturers).

Photo courtesy of Cochlear Americas

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, including:

- The best possible speech perception and production.
- Academic success.
- Emotional adjustment.
- Social competence.
- Occupational preparation.
- Be equipped to lead a healthy, productive life.

Table 1
CI Components of Three Manufacturers

MED-EL

Advanced Bionics Corporation

Cochlear Corporation

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