Cochlear Implants in Infants and Toddlers

Indexing Metadata/Description

› **Device/equipment:** Cochlear Implants in Infants and Toddlers

› **Synonyms:** Bionic ear; assistive listening device; implants, cochlear in infants and toddlers; auditory prosthesis in infants and toddlers; prosthesis, auditory in infants and toddlers; cochlear prosthesis in infants and toddlers; prosthesis, cochlear in infants and toddlers

› **Area(s) of specialty:** Hearing Impairment

› **Description/use:** A cochlear implant (CI) is a small, partially implanted electronic device that provides a person with a profound hearing loss a representation of sound from the environment.\(^1\) A CI contains components that are located outside of a person’s body (external portion) as well as parts that are implanted (implanted portion). The external portion includes a microphone, a sound processor, and a transmitter system, which transmits speech signals to the receiver-stimulator. The external portion is removable and is powered by batteries, much like traditional hearing aids. The implanted portion includes a receiver-stimulator, which converts information from the transmitter into electric impulses, and an electrode system. The electrode system, which is threaded through the cochlea, transports the electrical impulses to the auditory nerve. The external and internal systems are held next to each other by an implanted magnet which works through the person’s skin.\(^1,2,3\)

› **Indications:** Children who are permanently profoundly deaf or have severe, bilateral hearing impairment are typically good candidates for CIs; however, hearing threshold is only one factor when considering a child’s candidacy for a CI.\(^1,4,61\)

• Although the United States Food and Drug Administration (FDA) has approved multichannel CIs for implantation in children as young as 12 months of age, many children younger than 12 months have received CIs off-protocol\(^5,6\)

– Authors of a review of the literature found that evidence of the auditory and linguistic benefits of CI implantation outweigh the risks in children under age 12 months with an experienced surgeon and pediatric anesthesiologist\(^7\)

– In a retrospective chart review study conducted in Australia, researchers found that better communication development was associated with early implantation. Subjects in the study were 35 children implanted between 6 and 12 months of age and 85 children implanted between 13 and 24 months of age. Outcome measures of language, speech production, and speech perception were taken at 1, 2, 3, and 5 years post implantation; subjects who had been implanted prior to 12 months of age performed significantly better than those who received the CI after 12 months of age.\(^45\)

– Implantation after 12 months of age is common among children with progressive hearing loss because they often are not candidates for CI at the time their hearing impairment initially is diagnosed. Children with known progressive hearing loss should be monitored frequently to determine when they are appropriate candidates for implantation.\(^56\)

• Children with the following diagnoses might be candidates for a CI:

– Sensorineural hearing loss\(^3\)

– Auditory neuropathy\(^3\)
- Meningitis followed by hearing loss
- Otosclerosis
- Inner ear malformations

- Authors of a systematic review of the literature on cochlear implantation for children with unilateral hearing loss published in 2016 reported that CIs for these children might be an effective treatment for improving quality of life and speech perception; however, they were unable to draw firm conclusions due to the variability in study methodologies and the heterogeneity of study findings and outcome measures. There were only 5 articles that met criteria for this systematic review and the studies had small sample sizes.

- The following risks are associated with CIs
- Risks associated with receiving general anesthesia
- Facial nerve could become damaged during surgical implantation
- Meningitis could occur (especially in individuals with inner ears that are abnormally formed)
- Cerebrospinal fluid could leak
- Perilymph fluid could leak
- Skin wound could become infected
- Blood or fluid could collect at the site of surgery
- Dizziness or vertigo could develop
- Tinnitus
- Taste sensation could be disturbed
- The ear could become numb
- The body could reject the implant, leading to localized inflammation and a reparative granuloma (localized inflammation that occurs if the body rejects the implant)
- Sounds might be perceived differently than prior to surgery
- Residual hearing will be lost
- Long-term effects are unknown and uncertain
- Hearing abilities following surgery vary among patients; results might be disappointing
- Language comprehension following surgery varies among patients; results might be disappointing
- Certain medical procedures cannot be performed following CI:
  - MRI (an MRI can dislodge the CI or cause the internal magnet to demagnetize; however, two multichannel CIs exist that can undergo 1.5 Tesla MRI imaging in certain conditions)
  - Neurostimulation
  - Electrical surgery
  - Electroconvulsive therapy
  - Ionic radiation therapy
- CI might need to be removed if infection occurs
- CI could fail
- As CI technology improves, the patient might not be able to upgrade to new CI if new external parts are not compatible with existing internal portion
- CI is dependent upon batteries to function
- There is a risk of damaging the CI (e.g., external portions might be damaged if they become wet)
- Patient might need to make lifestyle changes
- Skin might become irritated due to placement of external portions
- Strange sounds might be heard when in the presence of magnetic fields (e.g., near airport passenger screening machines)
There is a large amount of opposition to CI use in the Deaf community. Many members of this community believe that CI use is not culturally responsive in that it prevents CI users from being complete members of the community, implies that deafness is a disability (a belief that the Deaf community adamantly opposes), and implies that Deaf persons are of lesser worth.  

CPT codes
- 92601 cochlear implant follow-up exam (under age 7)
- 92602 reprogramming of cochlear implant (under age 7)
- 92507 aural rehabilitation

ICD-10 codes
- Z96.21 cochlear implant status

(Codes are provided for the reader’s reference, not for billing purposes)

G-Codes
- **Spoken Language Comprehension G-code set**
  - G9159, Spoken language comprehension functional limitation, current status at therapy episode outset and at reporting intervals
  - G9160, Spoken language comprehension functional limitation, projected goal status at therapy episode outset, at reporting intervals, and at discharge or to end reporting
  - G9161 Spoken language comprehension functional limitation, discharge status at discharge from therapy or to end reporting
- **Spoken Language Expressive G-code set**
  - G9162, Spoken language expression functional limitation, current status at therapy episode outset and at reporting intervals
  - G9163, Spoken language expression functional limitation, projected goal status at therapy episode outset, at reporting intervals, and at discharge or to end reporting
  - G9164, spoken language expression functional limitation, discharge status at discharge from therapy or to end reporting

Reimbursement: CIs are now recognized as standard treatment for severe-to-profound sensorineural hearing impairment, and most insurance companies cover the cost of implantation.  
- As of 2004, Medicare, Medicaid, the Veterans Benefits Administration (see [http://www.benefits.va.gov/benefits/](http://www.benefits.va.gov/benefits/)), and over 90% of all commercial health plans cover CIs in the United States.
- CI centers will often take the responsibility of obtaining authorization from the patient’s insurance company prior to proceeding with surgery.

<table>
<thead>
<tr>
<th>G-code Modifier</th>
<th>Impairment Limitation Restriction</th>
</tr>
</thead>
<tbody>
<tr>
<td>CH</td>
<td>0 percent impaired, limited or restricted</td>
</tr>
<tr>
<td>CI</td>
<td>At least 1 percent but less than 20 percent impaired, limited or restricted</td>
</tr>
<tr>
<td>CJ</td>
<td>At least 20 percent but less than 40 percent impaired, limited or restricted</td>
</tr>
<tr>
<td>CK</td>
<td>At least 40 percent but less than 60 percent impaired, limited or restricted</td>
</tr>
<tr>
<td>CL</td>
<td>At least 60 percent but less than 80 percent impaired, limited or restricted</td>
</tr>
<tr>
<td>CM</td>
<td>At least 80 percent but less than 100 percent impaired, limited or restricted</td>
</tr>
<tr>
<td>CN</td>
<td>100 percent impaired, limited or restricted</td>
</tr>
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Indications for device/equipment

› Children who are profoundly deaf, have a significant, permanent hearing loss, or have another type of hearing disorder (as listed above) are candidates for a CI\(^1\)\(^3\)\(^5\)

  • Although children with cognitive impairment and developmental disabilities can benefit from a CI, it is more difficult to anticipate outcomes and there might be significantly greater risks associated with surgery for these children\(^8\)\(^45\)\(^60\)

According to current FDA guidelines, in order to receive a CI a child must have a significant hearing loss, display a lack of auditory development even when using hearing aids, and be medically fit for surgery\(^9\)

  • 95% of deaf children are born to parents with normal hearing; CI implantation can improve parent-child bonding between parents with normal hearing and their hearing-impaired infants\(^2\)

› Prior to being approved for a CI, a child must undergo audiological, otologic, ophthalmologic, physical, auditory skill, and language assessments\(^4\)\(^5\)

› Prior to receiving a CI, the expectations of the child’s family should be evaluated to confirm that they are realistic. Additionally, prior to implantation, the family should receive information on the requirements for programming and aural rehabilitation following implantation. Characteristics displayed by family members that may hinder progress following implantation should be identified and addressed if necessary\(^4\)

Guidelines for use of device/equipment

› The FDA regulates the development and manufacturing of CIs available in the United States. CIs must be deemed safe and effective prior to FDA approval\(^2\)

› Typically, a child’s family/caregiver and surgeon work collaboratively to determine which type of CI best meets the patient’s needs\(^5\)

› A CI can be implanted unilaterally or bilaterally; however, a growing body of evidence regarding CI implantation supports early, bilateral implantation of CIs in congenitally deaf children in order to achieve the best auditory and linguistic outcomes\(^10\)\(^11\)\(^12\)\(^24\). In children with asymmetrical hearing loss, unilateral implantation might be performed initially with a hearing aid used on the less impaired ear\(^51\). In some cases, the unilateral CI is sufficient, but a second CI might be necessary in cases in which optimal outcomes are not achieved or hearing in the better ear worsens over time\(^51\)

  • Despite the fact that evidence supports early implantation, there are sometimes barriers to early implantation of CI in children with hearing loss. Researchers in Australia found that, in a sample of 417 children who received a CI before the age of 3, children with stable (nonprogressive) hearing losses, higher socioeconomic status, and those who had been fitted for hearing aids earlier in life were more likely to have cochlear implantation at an earlier age compared to children without these characteristics\(^58\)

Contraindications/Precautions to device/equipment

› There are two primary questions that must be addressed when considering cochlear implantation in a child: 1) Do the benefits of a CI outweigh the risks associated with this type of intervention? 2) Will CIs provide greater benefit to the child and facilitate communication significantly more than traditional amplification (e.g., hearing aids)?\(^4\)

› A child’s family must make an ongoing commitment of time and resources following implantation. Is the child’s family able to make this commitment?\(^4\)

In order to determine whether a child is a good candidate for a CI, input from professionals in several fields is required. An audiologist and CI surgeon will be the primary professionals involved in the decision-making process; however, speech-language pathologists (SLPs), aural rehabilitation specialists, psychologists, social workers, occupational therapists, and/or early intervention specialists are often among the professionals involved in preimplantation assessment as well\(^5\)\(^9\)

Cochlear nerve deficiency (seen on MRI) is a marker of very poor outcomes with CI\(^13\)

› There is an increased, life-threatening risk of bacterial meningitis in patients with CIs; the U.S. Centers for Disease Control and Prevention (CDC) and FDA recommend fully immunizing persons who receive CIs against bacterial meningitis\(^2\)

  • Children implanted with CIs that have a positioner were at greatest risk; however, the only model with a positioner was withdrawn from the market in 2002\(^2\)

› The SLP should carefully review all audiological and medical records before evaluating or treating a child at any stage in the implantation process (i.e., before or after implantation)
A child and family’s cultural and linguistic background should be considered to determine the appropriateness of examination and therapy materials.

**Examination**

> **Contraindications/precautions to examination**
> - Assessments conducted prior to implantation should be conducted by SLPs who have experience in postimplantation management and rehabilitation.
> - During a preimplantation assessment, the family should be presented with alternative rehabilitative options (e.g., use of hearing aids or signed languages).
> - When assessing a young child, both prior to cochlear implantation and following implantation, developmentally appropriate practices must be used. This may include dividing assessment time into multiple sessions and relying heavily on parental report for an accurate picture of the child’s skills.
> - SLPs must review all medical and audiological evaluation reports prior to conducting a speech-language assessment.
> - During a speech-language evaluation, it is of utmost importance to be aware of and sensitive to a child’s level of frustration.
> - Prior to surgery, the child’s family/caregivers should receive counseling on the potential risks and benefits of implantation.
> - Prior to surgery, the patient’s family/caregivers should be educated on the components of a CI and the daily responsibilities of CI use.
> - Prior to surgery, the SLP should discuss the range of possible speech and language outcomes with the child’s family/caregivers.

> **History**
> - **History of present illness/injury**
>   - **Mechanism of injury or etiology of illness:** Document the etiology and history of the patient’s hearing loss. In cases with congenital absence of neural foramina and acoustic tumors, auditory nerve innervation may not be possible and thus a CI is inappropriate. In cases of hearing loss resulting from structural abnormalities, if a CI is determined to be appropriate electrode placement must be carefully considered and/or adjusted. Decisions such as these will be made by the CI surgeon.
>   - **Course of treatment:** A trial period with a hearing aid is highly recommended; the child’s response to the hearing aid during this period will help to determine whether the child is a good candidate for CI. A child’s willingness to tolerate wearing a hearing aid could indicate a likelihood of success in wearing of external CI components. However, a child who is unwilling to wear a hearing aid could still be appropriate for a CI in order to access auditory information.
>   - **Medical management:** Document previous use of hearing aids and/or other amplification devices (e.g., FM systems) and examine audiograms (if available) that illustrate the patient’s pure tone thresholds with and without the use of amplification.
>   - **Medications for current illness/injury:** Determine what medications (if any) physician has prescribed; are they being taken?
>   - **Diagnostic tests completed:** Usual tests to determine candidacy for a CI are described below. In addition, a child’s speech, language, and auditory skills should be assessed regularly following implantation to determine if the child is meeting developmental benchmarks.
>     - High-resolution computerized tomography (HRCT), CT, and/or MRI should be conducted by the appropriate medical professional in order to evaluate accessibility of the cochlea, identify or rule out any structural abnormalities of the inner ear, assess the temporal bone, and assess cochlear nerve integrity.
>     - Audiologic evaluation to determine:
>       - Presence and degree of hearing loss
>       - Preoperative hearing status, including a complete audiogram, air and bone conduction thresholds, and speech perception testing (when age appropriate)
>       - Patient’s previous use of amplification systems, such as hearing aids, and results of amplification use
>       - Need for trial period of a less permanent type of amplification (i.e., hearing aids) in patients who have not previously used amplification.
- Physical examination to determine:
  - State of general physical health\(^{(4,5)}\)
  - Ability to tolerate general anesthesia\(^{(2,4-5)}\)
  - Ability to participate in postimplantation training\(^{(5)}\)
  - Physical state of the patient’s cochlea and inner ear\(^{(2,5)}\)
  - Visual skills/acuity: can the child use vision to support language development as his or her auditory skills develop?
    - Does the child display visual abnormalities associated with congenital sensorineural deafness (e.g., strabismus)?
    - Does the child have visual symptoms that are associated with other conditions (e.g., Usher syndrome, rubella retinopathy)?\(^{(4)}\)

- Psychological evaluation to assess:
  - Parental expectations regarding outcomes following implantation\(^{(4)}\)
  - Patient and family/caregiver’s ability to cope with having a CI\(^{(2)}\)
  - Patient and family/caregiver’s ability to complete needed follow-up care\(^{(5)}\)
  - Patient and family/caregiver’s ability to incorporate CI into daily living\(^{(5)}\)

- General eligibility considerations:
  - Patient has been identified with a severe-to-profound sensorineural hearing loss in both ears\(^{(1)}\)
  - Patient receives limited benefit from hearing aids and expectations are that communication would benefit from a CI\(^{(3,5)}\)
  - No medical or radiological contraindications exist\(^{(3,5)}\)
  - Patient’s family/caregivers are highly motivated to attend programming and rehabilitation appointments\(^{(2)}\)
  - Patient’s family/caregivers display appropriate expectations regarding potential outcomes\(^{(2)}\)
  - Patient’s family/caregivers have or can develop necessary support networks to use a CI on a daily basis\(^{(5)}\)

– Previous therapy: Document whether patient has had speech therapy, occupational therapy, physical therapy, or other early intervention services for this or other conditions and what specific treatments were helpful or not helpful

– Psychosocial status: Communication is a significant aspect of psychosocial development in children with hearing impairment\(^{(15)}\)
  - In a study conducted in Denmark, researchers reported that children with hearing impairment who had access to early sign language or oral communication did not have an increased prevalence of psychosocial difficulties when compared to normal-hearing peers\(^{(15)}\)
  - Additionally, the researchers found increased psychosocial difficulties in children who had other disabilities in addition to the hearing loss\(^{(15)}\)
  - In a 5-year prospective follow-up study conducted in Finland with 18 unilaterally implanted children, parents reported a major increase in spoken language and socioemotional development 1–2 years after implantation; by 5 years postimplantation, two thirds of the parents reported that they perceived their children to be as independent as their normal-hearing peers\(^{(16)}\)
  - Parents of children with profound hearing loss can experience grief and depression\(^{(11)}\)
  - Document information related to psychosocial status. Inquire about symptoms of depression and anxiety in the child, parents, and caregiver(s) and refer to psychiatric professional for intervention as appropriate\(^{(11)}\)

– Barriers to learning
  - Are there any barriers to learning? Yes__ No__
  - If Yes, describe _________________________

• Medical history
  – Past medical history: Obtain a complete medical history (including information on mother’s pregnancy and child’s birth) as well as a complete developmental history
  – Previous history of same/similar diagnosis: Document etiology of hearing loss, history of hearing loss, and history of amplification use.\(^{(5)}\) Document developmental delays in the areas of speech, language, gross motor skills, fine motor skills, and cognition
  – Comorbid diagnoses: Ask patient’s family/caregivers about other problems the patient is experiencing, including visual deficits, developmental delays, and other diagnoses that might affect communication
- **Medications previously prescribed:** Obtain a comprehensive list of medications prescribed and/or being taken (including OTC drugs)
- **Other symptoms:** Ask patient’s family/caregivers about other symptoms the patient is experiencing

**Social/occupational history**
- **Patient’s goals:** Document what the family/caregivers hope to accomplish in speech therapy
- **Vocation/avocation and associated repetitive behaviors, if any:** Will the patient and parent/caregiver be able to attend follow-up appointments to complete CI programming and aural rehabilitation? Does the patient participate in recreational/social activities? Does the patient attend daycare or preschool? Does the patient receive early intervention services?
- **Functional limitations:** Patient’s family/caregivers must be aware of potential for CI to interact with electronics in the environment (e.g., various types of security systems, cellular phones, computer systems). Do the family/caregivers understand functional limitations associated with having a CI? For example:
  - Patient must avoid static electricity
  - Patient/family/caregiver must remove external components before swimming or bathing
- **Living environment:** With whom does patient live (e.g., caregivers, siblings)? Who will participate in aural rehabilitation program with the patient? Does patient and patient’s family/caregiver have access to transportation for follow-up care? What language(s) are used in the home, daycare, and community? What communication method is used in the home (e.g., oral communication, sign language)? Is the communication method used in the home the same as the method used in the child’s therapy and/or school setting? What is the noise level in the child’s living environment? Children with CIs have considerable difficulty with speech perception tasks in the presence of background noise.

**Communication history:** Assess patient’s history of hearing loss and use of hearing aids and/or other amplification. Did the child’s hearing loss occur before or after he or she acquired some language? Determine what type of communication system patient and family/caregivers primarily use (e.g., oral, total communication, signed languages)

**Relevant tests and measures:** (While tests and measures are listed in alphabetical order, sequencing should be appropriate to patient medical condition, functional status, and setting.) The following areas should be tested/measured prior to implantation in order to establish a child’s baseline skills and then on a regular basis following implantation in order to monitor the child’s progress

**Assistive and adaptive devices**
- Note patient’s previous use of amplification and its effect on patient’s ability to hear and communicate. Determine if both ears have received previous amplification and/or if one ear was more consistently amplified. Ears that have received consistent amplification are associated with better outcomes upon receiving a CI; therefore, if only one ear has received auditory input over time, the cochlear implant surgeon may prefer to implant that ear
- Note if patient uses positioning devices (e.g., for sitting, standing), augmentative and assistive communication (AAC) devices, glasses, or any other type of assistive or adaptive device

**Audiological testing**
- All audiological testing must be done by a certified and licensed audiologist. Specific testing will vary according to the age of the child
- **Infant hearing screening:** Infants in the United States are commonly screened for hearing loss shortly after birth (before leaving the hospital or birthing center). An infant’s hearing can be assessed using otoacoustic emissions testing (OAE) and/or auditory brainstem response testing (ABR)
- Infant hearing screenings can ensure early cochlear implantation. Authors of a retrospective data review study conducted in New Zealand with 123 children who received a CI between March 2003 and August 2014 reported that the median age of implantation for infants who had received a hearing screening shortly after birth was 13 months. The median age of implantation for the children who did not undergo an infant hearing screening was 24 months
- **Toddler/preschool screenings:** If hearing is not screened at birth, the child should be screened at 3 months of age. If a child is at high risk for developing hearing loss (e.g., family history of childhood hearing loss, history of recurrent otitis media), he or she should be rescreened at 6-month intervals until the age of 3 even if no hearing loss is initially identified
- **Otoscopic examination:** Visual examination of the ear canal and tympanic membrane (eardrum)
- **Tympanograms:** Measurement of tympanic membrane mobility
- **Static acoustic measures:** Measures the volume of air in the ear canal (can be used to identify perforated eardrums or blocks in the ventilation of the ear)
Acoustic reflexes: Measures the responses of tiny ear muscles when presented with sound at varying intensities\(^\text{(12)}\).

OAE (otoacoustic emissions): OAEs measure the cochlea’s response to sound stimulation. OAEs can identify blockages in the outer ear canals, fluid in the middle ear, and damage to the cochlea’s outer hair cells\(^\text{(18)}\).

ABR (auditory brainstem response): During ABR testing, electrodes are placed on an infant’s head while sound is presented to his or her ears. The electrodes record the brain’s response (or lack of response) to these sounds. ABRs can be used to identify damage to the cochlea, auditory nerve, and auditory pathways in the brainstem\(^\text{(17)}\).

Pure-tone and speech reception thresholds: A patient’s ability to hear calibrated pure tones and recognize the presence of speech is measured.\(^\text{(18)}\) This may be done using visual reinforcement audiometry (VRA) or conditioned play audiometry (CPA).\(^\text{(2)}\) VRA is primarily used to test children between the ages of 6 months and 2 years. In VRA, children are trained to look towards lights or toys when they hear a sound presented by the audiologist. After training, when the child looks up upon hearing a sound, he or she is rewarded by the presentation of lights or a moving toy. CPA is primarily used on children between the ages of 2 and 3 years and involves training a child to perform an activity (e.g., placing a block in a box) when hearing a sound\(^\text{(17)}\).

Speech perception tests (word and sentence recognition): Speech perception testing will depend upon the age of the child but may include open-set word and sentence recognition or closed-set tasks involving the identification of prosodic features and word identification.\(^\text{(2,9)}\) If age appropriate, speech perception measures should be taken for each ear.

- In a study conducted in Taiwan with Mandarin-speaking children with CIs, researchers found that although there were far fewer tones than speech sounds in Mandarin, tone perception was significantly poorer than speech-sound perception\(^\text{(19)}\).

Evaluation of current amplification: Does the child currently use hearing aids or another form of amplification? How has the patient benefitted from his or her current amplification?\(^\text{(9,2)}\)

Trial use of amplification, if appropriate: If the patient has not previously used amplification, a 3- to 6-month trial period might be recommended\(^\text{(5)}\).

Auditory skills assessment: Prior to receiving the CI, a child should use his or her hearing aids during auditory skills assessment. Assess the child’s ability to 1) pay attention to speech and environmental sounds, 2) integrate auditory perception and speech production (i.e., is the child able to imitate sounds that he or she hears?), 3) assign meaning to the sounds he or she hears, and 4) use hearing to engage in communication exchanges.\(^\text{(4)}\) Examples of auditory skill measures include\(^\text{(9)}\):

- Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS):\(^\text{(20)}\) Results are based upon parent/caregiver report. For use with infants and toddlers. The IT-MAIS assesses the following three areas:
  - Vocalization behavior
  - Alerting to sound
  - Deriving meaning from sound

Speech and language examination:

Speech: Complete a phonetic inventory in order to determine which sounds the child is producing and/or administer an articulation test to identify or rule out articulation errors\(^\text{(9,24)}\).

- Stark Assessment of Early Vocal Development–Revised (SAEVD-R): Instrument used to track vocal development in infants and toddlers from birth to 20 months of age. Includes five hierarchical levels of vocalizations\(^\text{(48)}\).

- Many children with early implantation of a CI will progress through articulation and phonological development in the same manner as their normal-hearing peers.\(^\text{(22)}\) Children with severe-to-profound hearing loss who have early cochlear implantation are able to obtain phonological awareness abilities (which are essential in order to learn to read) similar to those of children with typical hearing abilities\(^\text{(23)}\).

- In a study conducted in the United States with CI implant users aged 9 to 36 months, researchers found that the children developed intonation over a similar amount of time and with a similar pattern to typically developing infants based on the length of hearing experience (i.e., an 18-month-oldchild who has had a CI for 9 months would be expected to have intonation patterns similar to those of a typically developing 9-month-old infant). For children who were slightly older when receiving their CI (> 25 months), intonation development was more rapid than expected (i.e., a 36-month-oldchild with 3 months of CI use might achieve the same intonation developmental level as a typically developing infant does over 6 months of time)\(^\text{(23)}\).
In a study conducted in Poland of severely, prelingually deaf children under the age of 2 years, researchers found that prosodic development was significantly better in children who used CIs than in those who used hearing aids. Even with early cochlear implantation, children with CI have increased risk of vocabulary delays compared to typically developing peers. A child’s language can be assessed through standardized testing or through the collection of language samples. Language assessments that are appropriate for children from birth to age 3 years include:

- Communication and Symbolic Behavior Scales: Assesses prelinguistic skills such as communicative functions, use of gestures, reciprocity, and eye gaze. For use with children from birth through age 4 years
- Rossetti Infant-Toddler Language Scale: Parent report, observation, and elicited behavior are used to assess a child’s communication skills. For use with children from birth to age 3 years or birth through preschool
- MacArthur Index of Communicative Development: Parent report is used to assess language and communication skills in infants and toddlers. For children aged 8–37 months

**Fluency:** Informally assess fluency of speech in order to rule out or identify any atypical disfluencies

**Voice:** Informally assess voice quality in order to rule out or identify any voice disturbances

- Children with CIs have difficulty controlling pitch and loudness of sustained phonation

**Oral mechanism exam and related tests:** Complete an oral motor assessment in order to rule out or identify any oral mechanism dysfunction

**Parent-child communication:** Many parents who are considering a CI for their child may be experiencing feelings of grief, depression, fear, anger, and/or stress due to their child’s hearing loss diagnosis. Evidence exists to suggest that maternal stress and depression can negatively impact a child’s linguistic, emotional, and behavioral development. Additionally, the communication interactions between hearing mothers and their deaf children contain significant disruptions (more maternal control, more rigidity, less joint attention) and lead to poorer communication skills in deaf children. Therefore, it is critical to monitor parent-child interaction styles and refer a family to social work and/or psychology when signs of stress or depression are present

**Setting the cochlear implant processor (programming):** A patient who receives a CI is not able to hear immediately following surgery. Three to 5 weeks after surgery (when the physical healing has taken place), the patient receives the external components of the CI. At that time, the external processor is programmed by the patient’s audiologist. Programming might require several visits to complete, and reprogramming is sometimes necessary if the patient experiences changes in his or her ability to hear over time. Electrodes are assigned specific sound frequencies. During programming, each electrode is programmed for a threshold level (the lowest level at which the patient responds to a sound sensation 100% of the time) and for the most comfortable level of loudness. Other parameters that can be adjusted during programming include global loudness levels and speed of transmission

### Assessment/Plan of Care

#### Contraindications/precautions

- Clinicians should follow the guidelines of their clinic/hospital and what is ordered by the patient’s physician. The summary below is meant to serve as a guide, not to replace orders from a physician or a clinic’s specific protocols
- Once a CI has been implanted, the patient, with the aid of family/caregivers, must heed the following precautions:
  - Avoid static electricity
  - Remove external components before swimming or bathing
- Be aware of the potential for the CI to interact with electronics in the environment (e.g., various types of security systems, cellular phones, computer systems)

#### Diagnosis/need for device/equipment:

An infant or toddler’s family/caregivers work in collaboration with a cochlear implant team to determine the appropriateness of a CI on an individual basis. Team members working with infant and toddler patients might include the CI surgeon, an otologist or otolaryngologist, an audiologist, an SLP, a family counselor/psychologist, an aural rehabilitation specialist, a social worker, an occupational therapist, and/or an early intervention specialist

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(24) Language: Assess the child’s ability to express thoughts, wants, and needs vocally, verbally, gesturally, and/or using signed language. Following implantation, it is important to carefully monitor the child’s vocabulary development.

(25) Even with early cochlear implantation, children with CI have increased risk of vocabulary delays compared to typically developing peers.

(26) A child’s language can be assessed through standardized testing or through the collection of language samples.

(27) Language assessments that are appropriate for children from birth to age 3 years include:

- Communication and Symbolic Behavior Scales: Assesses prelinguistic skills such as communicative functions, use of gestures, reciprocity, and eye gaze. For use with children from birth through age 4 years
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(28) Fluency: Informally assess fluency of speech in order to rule out or identify any atypical disfluencies

(29) Voice: Informally assess voice quality in order to rule out or identify any voice disturbances

(30) Children with CIs have difficulty controlling pitch and loudness of sustained phonation.
Prognosis

Outcomes following a CI are highly variable and a child’s family/caregiver should receive pre-implant counseling to explain this fact and to establish appropriate expectations regarding speech, language, and auditory outcomes.

Factors influencing outcome include the following:

- **Child variables**
  - Length of time that the child has been deaf (shorter periods of deafness lead to more positive outcomes)
  - Age at time of deafness/hearing loss (children who became deaf at later ages typically have more positive outcomes)
  - Age at implantation (children who undergo implantation at younger ages typically have more favorable outcomes)
  - Early implantation (at ages 1–2 years) does not necessarily result in normal language development
  - In a study conducted in the United States of infants with CIs, researchers found that infants with CIs transition from precanonical vocalizations to speech-like babbling rapidly despite initial auditory deprivation prior to receiving a CI. Findings of this study showed a significant decrease in precanonical vocalizations and increase in speech-like babbling after 1 year of CI use.
  - Being cognitively, socially, and motorically mature at the time of implantation (compared with a hearing newborn) can contribute to a rapid pace of vocal development
  - Length of time using implant: children who used a CI for longer periods of time during infancy and very early childhood had more favorable spoken language outcomes
  - In a study conducted in the United States, researchers compared outcomes in 9 English-speaking children who received CIs before 30 months of age at 3, 4, and 5 years postimplantation
  - 9 typically developing peers were used as control subjects
  - Outcome measure was the use of correct tense marking in a story retell task
  - The percent of correct tense marking increased significantly over time for both groups of children (those with CIs and those who were typically developing)
  - As a group, the children with CIs at 3 years postimplantation had similar accuracy of tense marking compared with typically developing children; however, accuracy was significantly lower for the children with CIs at 4 and 5 years postimplantation. There was considerable individual variability among the children with CIs throughout the study; some of the children with CIs performed similarly to typically developing peers at all testing points.
  - Patient’s learning aptitude/nonverbal intelligence
  - Health and structure of the patient’s cochlea
  - Patient’s amount of residual hearing pre-implant
  - Patient’s lip-reading ability

- **Implant variables**
  - Type of speech processor
  - Larger numbers of active electrodes in the CI map lead to better outcomes
  - A precise and accurate map is associated with more positive outcomes
  - Bilateral or unilateral implantation
    - Bilateral implantation is associated with better sound localization and speech recognition in noise
    - In a study conducted in the United States with 36 children who had been implanted either bilaterally or unilaterally at some point between ages 10 and 23 months, researchers found that at 12 months postimplantation, overall vocabulary size did not differ between children with unilateral versus bilateral CIs. The children with bilateral CIs had much more sensitive word-level statistical patterns of spoken language (e.g., words with rare sound sequences and dense neighborhoods) than did those with a unilateral CI. The sensitivity of the children with bilateral CIs was close to that of a child with normal hearing. Because sensitivity to these patterns is associated with the ability to learn new sounds, vocabulary, and sentence structures, researchers concluded that future acquisition of spoken language is likely to be stronger in the children with bilateral CIs.
Rehabilitation program variables
- Mode of communication used in the child’s educational setting(32)
- Enrollment in an oral education program leads to more positive outcomes in auditory and speech skill development(32)
- High level of parental involvement(37,38)
- Parental use of spoken language at home(38)

- It is critical that a child use his or her CI full-time (i.e., during all waking hours except when in water) in order to get the full benefit from the device(14)
- In a longitudinal research study involving 13 children who received CIs prior to 36 months of age and 11 typically hearing children, researchers compared the vocal development of the two groups. Typically hearing children were assessed at 6, 9, and 12 months of age. Children with CIs were assessed at 2, 3, 6, 9, and 12 months post CI activation. Following the conclusion of this study, researchers reported that following implantation young CI users used fewer non-speech-like vocalizations more quickly than typically hearing children and they began to use basic canonical syllables more quickly than younger, typically hearing children. The authors of this study suggest that this rapid development in older children with CIs might be due to motor, cognitive, neurological, and social maturity at time of implantation (when compared to typically developing infants)(50)
- Following study of 12 young children who had received CIs prior to age 3 years and 11 typically developing children, researchers concluded that during the second year of CI use vocal development in CI users continues to occur at a faster rate than in younger, typically developing children(49)

Referral to other disciplines: Patient should be referred to audiology if patient, patient’s family, the SLP, or aural rehabilitation specialist notices changes in hearing levels following implantation and programming(2)

Treatment summary
- Twelve guiding principles have been suggested based upon research findings and clinical experience(37)
- The infant or toddler with a CI must learn to attach meaning to what is heard through his or her CI(37)
- The ultimate goal for children with CIs is that they become competent communicators(37)
- Skills learned in a therapy setting must be transferred into the classroom, home, and other natural settings(37)
- Rehabilitation sessions should integrate goals of speech, language, auditory perception, and pragmatics within an environment that has appropriate social/emotional context(37)
- Almost all children with CIs require a combination of direct teaching of specific auditory skills and incidental learning to acquire spoken language(37)
- A diagnostic teaching approach to CI therapy yields the most benefit. This approach determines what a child can currently do and then challenges the child to perform at the next level of difficulty while identifying factors that facilitate or hinder performance(37)
- Material from the child’s educational program should be used during rehabilitation/therapy sessions(37)
- Music should be integrated into rehabilitation sessions since music perception is closely related to speech and other auditory perception(37)
  - In a study conducted in Taiwan, researchers found that music training helped improve pitch perception in children with CIs who were prelingually deaf(39)
- Infants and toddlers with CIs have unique needs that differ from the needs of older children with CIs(37)
- Established auditory milestones can be used as “red flags” to identify children who are progressing at slower than expected rates(40) These red flags include(14)
  - Child is not using his or her CI full-time (i.e., all waking hours except when in water) 1 month after initial stimulation(14)
  - Child does not display a change in the quality or quantity of his or her vocalizations after 3 months of CI use(14)
  - Child displays skills during audiological testing that he or she does not display in everyday situations(14)
  - Child is not spontaneously alerting to his or her name after 6 months of CI use(14)
  - Child is not responding to environmental sounds after 6 months of CI use(14)
  - Child does not display evidence that he or she is deriving meaning from sound after 12 months of device use(14)
Formal assessment tools might not adequately capture a child’s difficulties in the areas of inference, problem solving, and topic shifts; therefore, they should not be used in isolation\(^\text{(37)}\).

**Parent/caregiver training**

– Since parents/caregivers play a critical role in fostering the speech and language development in infants and toddlers with CIs, aural rehabilitation for this age group should involve the family as a whole\(^\text{(37,40)}\).

– SLPs and/or aural rehabilitation specialists must be able to identify and describe a young child’s communication behaviors to the parent/caregiver\(^\text{(40)}\).

– Parents/caregivers should be taught methods for stimulating developmentally appropriate communication behaviors while they interact with their child\(^\text{(40)}\).

**Considerations for working with infants and toddlers with CIs**

– When working with infants and toddlers, it is critical that therapy activities are centered around the whole family rather than the individual child\(^\text{(14)}\).

– Therapy materials and procedures must be developmentally appropriate for infants and toddlers (e.g., use of real objects rather than pictures of objects during therapy sessions, use of vocabulary and syntax that a young child would understand)\(^\text{(14)}\).

– Infants and toddlers present with behavior and compliance issues not seen in older children\(^\text{(14)}\).

– Therapy sessions should focus on incidental learning rather than didactic instruction\(^\text{(14)}\).

– The home is recognized as the primary language learning environment\(^\text{(14)}\).

– When working with infants it is critical to emphasize the prosody and suprasegmental aspects of speech (e.g., pitch, speech rate, varying loudness levels) rather than focusing on vocabulary or grammar development. The social and emotional content of language is conveyed through prosody and is considered to be the most important linguistic information for an infant to learn\(^\text{(14,37)}\).

– Therapy activities should be play-based and focus on helping parents foster functional communication in everyday situations\(^\text{(37)}\).

**Communication mode:** Families with infants and toddlers with CIs will need to decide on a mode of communication to use with their child. Typically, communication modes used with children with CIs are either oral communication (OC) or total communication (TC).

– **Oral communication:** OC focuses on using speech and audition to communicate. Auditory input is used to monitor the accuracy of an individual’s own speech production and to understand spoken language. OC methods exist along a continuum with varying emphasis placed on the use of auditory versus visual channels to receive spoken information. For example, when using a cued speech mode of communication, a child gains valuable information from hand positions used in conjunction with lip-reading; however, when using an auditory-verbal approach, a child is encouraged to rely exclusively on auditory information\(^\text{(32)}\).

– **Total communication:** TC methods incorporate the use of both signed language and the development of speech and auditory skills. TC methods vary in the extent to which English grammar is emphasized as well in the extent to which speech, audition, and lip-reading skills are emphasized\(^\text{(32)}\).

**Short periods of prelinguistic input (SPPI)**\(^\text{(40)}\)

– The goal of SPPI is to assist children as they progress from precanonical to canonical and postcanonical levels of babbling. It is hoped that the establishment of these early developing phonetic skills will support the development of phonological and lexical skills\(^\text{(40)}\).

– Parents/caregivers/SLP begin by modeling vocalizations that the child is currently able to produce and then model more complex forms of babbling.

– Parent/caregiver models target vocalizations for 1-minute periods 5 times per day, repeating the vocalization once every 5 seconds\(^\text{(40)}\).

– SPPI is performed as vocal play during interactions that should be enjoyable to both the child and parent/caregiver\(^\text{(40)}\).

– SPPI is intended to be used as a strategy in conjunction with interventions focusing on developing meaningful speech and language skills\(^\text{(40)}\).
Although the effectiveness of SPPI has not yet been researched, the use of modeling is an accepted method of teaching speech and language to young children. Additionally, SPPI provides a systematic way to encourage early speech development.

<table>
<thead>
<tr>
<th>Problem</th>
<th>Goal</th>
<th>Intervention</th>
<th>Expected Progression</th>
<th>Home Program</th>
</tr>
</thead>
<tbody>
<tr>
<td>Delayed auditory and language development due to hearing loss</td>
<td>Support parent/family/caregiver’s ability to facilitate child’s auditory and language development</td>
<td><strong>Parent/caregiver training</strong>&lt;br&gt;• Structure therapy sessions to involve the whole family rather than the individual infant/toddler only&lt;sup&gt;(37)&lt;/sup&gt;&lt;br&gt;• SLP will identify and describe child’s communication to parent/caregiver&lt;sup&gt;(40)&lt;/sup&gt;&lt;br&gt;• SLP will teach parent/caregiver strategies to foster language development&lt;sup&gt;(40)&lt;/sup&gt; such as following the child’s lead; emphasizing rhythm, pitch contours, and “motherese”; creating communication temptations for the child&lt;sup&gt;(27)&lt;/sup&gt;</td>
<td>Parent/caregiver will learn strategies to facilitate child’s auditory and language development</td>
<td>Parent/caregiver will use these strategies across communication environments</td>
</tr>
</tbody>
</table>
Poor development of prelinguistic vocalizations due to hearing loss

Improve prelinguistic vocalizations

**Short periods of prelinguistic input (SPPI)**

- Parents/caregivers/SLP begin by modeling vocalizations that the child is currently able to produce and then more complex forms of babbling\(^{(40)}\)
- Parent/caregiver models target vocalizations for 1-minute periods 5 times per day, repeating the vocalization once every 5 seconds\(^{(40)}\)
- SPPI is performed as vocal play during interactions that should be enjoyable to both the child and parent/caregiver\(^{(40)}\)
- SPPI is intended to be used as a strategy in conjunction with interventions focusing on developing meaningful speech and language skills\(^{(40)}\)

Infant/toddler will produce more complex forms of vocal babble

Parent/caregiver will model babble targets during play-based activities throughout the day

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**Desired Outcomes/Outcome Measures**

- Increased sound awareness
- Improved speech recognition
- Increased prelinguistic vocalizations/increased babbling
  - Recording prelinguistic vocalizations and completing phonetic inventories can measure progress in speech development in the early periods following implantation\(^{(40)}\)
  - Tracking changes in babbling behavior (from precanonical, canonical, or postcanonical) can confirm progress in vocal development and indicate progress in speech perception when a child is too young to undergo formal testing\(^{(40)}\)
- Ability to communicate orally
  - Formal and informal testing measures should be completed in the areas of vocabulary, language, articulation, and auditory comprehension 6 months after the CI is activated and then once a year\(^{(2)}\)

**Maintenance or Prevention**

- In order to properly care for a CI
  - Patient should have regular audiological visits to determine any change in hearing ability that would require reprogramming
Family/Caregiver Education

Many parents experience feelings of grief, depression, and stress once hearing loss is diagnosed in their child. Participation in a support group may help address some of these feelings and provide the family/caregivers with additional resources. See Patient Information from the National Institute on Deafness and Other Communication Disorders at https://www.nidcd.nih.gov/health/cochlear-implants.


Information for parents and support for CI users is available on the Cochlear (Australian developer and manufacturer of CIs) website, http://www.cochlear.com/wps/wcm/connect/au/home/understand/understand.

References


10. Graham J, Vickers D, Eyles J, et al. Bilateral sequential cochlear implantation in the congenitally deaf child: evidence to support the concept of a ‘critical age’ after which the second ear is less likely to provide an adequate level of speech perception on its own. Cochlear Implants Int. 2009;10(3):119-141. (R)


