



Kansas City

An Independent Licensee of the Blue Cross and Blue Shield Association

Cochlear Implants

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Policy

Blue Cross and Blue Shield of Kansas City (Blue KC) will provide coverage for cochlear implants when it is determined to be medically necessary because the criteria shown below are met.

When Policy Topic is covered

Unilateral or bilateral cochlear implantation of a U.S. Food and Drug Administration (FDA) -approved cochlear implant device may be considered **medically necessary** in patients age 12 months and older with bilateral severe-to-profound pre-or postlingual (sensorineural) hearing loss defined as a hearing threshold of pure-tone average of 70 dB (decibels) hearing loss or greater at 500 HZ (hertz), 1000 HZ and 2000 Hz, and have shown limited or no benefit from hearing aids.

When Policy Topic is not covered

Cochlear implantation as a treatment for patients with unilateral hearing loss with or without tinnitus is considered **investigational**.

Cochlear implantation with a hybrid cochlear implant/hearing aid device that includes the hearing aid integrated into the external sound processor of the cochlear implant, including but not limited to the Nucleus® Hybrid™ L24 Cochlear Implant System, is considered **investigational** (see Considerations section).

Upgrades of an existing, functioning external system to achieve aesthetic improvement, such as smaller profile components or a switch from a body-worn, external sound processor to a behind-the-ear (BTE) model, are considered **not medically necessary**.

Considerations

Bilateral cochlear implantation should be considered only when it has been determined that the alternative of unilateral cochlear implant plus hearing aid in the contralateral ear will not result in a binaural benefit; i.e., in those patients with hearing loss of a magnitude where a hearing aid will not produce the required amplification.

In certain situations, implantation may be considered before 12 months of age. One scenario is post-meningitis when cochlear ossification may preclude implantation. Another is cases with a strong family history, since establishing a precise diagnosis is less uncertain.

Hearing loss is rated on a scale based on the threshold of hearing. Severe hearing loss is defined as a bilateral hearing threshold of 70–90 dB, and profound hearing loss is defined as a bilateral hearing threshold of 90 dB and above.

In adults, limited benefit from hearing aids is defined as scores 50% correct or less in the ear to be implanted on tape-recorded sets of open-set sentence recognition. In children, limited benefit is defined as failure to develop basic auditory skills, and in older children, $\geq 30\%$ correct on open-set tests.

A post-cochlear implant rehabilitation program is necessary to achieve benefit from the cochlear implant. The rehabilitation program consists of 6 to 10 sessions that last approximately 2.5 hours each. The rehabilitation program includes development of skills in understanding running speech, recognition of consonants and vowels, and tests of speech perception ability.

Contraindications to cochlear implantation may include deafness due to lesions of the eighth cranial nerve or brain stem, chronic infections of the middle ear and mastoid cavity or tympanic membrane perforation. The absence of cochlear development as demonstrated on computed tomography scans remains an absolute contraindication.

In 2003, CPT established a range of codes (92601–92606) to define a variety of postoperative evaluative and therapeutic services related to cochlear implants. Codes 92601 and 92603 describe postoperative analysis and fitting of previously placed external devices, connection to cochlear implant, and programming of the stimulator. Codes 92602 and 92604 describe subsequent sessions for measurement and adjustment of the external transmitter and reprogramming of the internal stimulator.

Hybrid cochlear implant devices that include a hearing aid integrated into the external sound processor of the cochlear implant are considered investigational because the available evidence does not demonstrate that hybrid devices improve outcomes compared with standard cochlear implants.

Extensions of the speech therapy benefit, if necessary, will be given individual consideration by the Medical Director.

Description of Procedure or Service

Cochlear implant is a device for individuals with severe-to-profound hearing loss who only receive limited benefit from amplification with hearing aids. A cochlear implant provides direct electrical stimulation to the auditory nerve, bypassing the usual transducer cells that are absent or nonfunctional in deaf cochlea.

Background

The basic components of a cochlear implant include both external and internal components. The external components include a microphone, an external sound processor, and an external transmitter. The internal components are implanted surgically and include an internal receiver implanted within the temporal bone and an electrode array that extends from the receiver into the cochlea through a surgically created opening in the round window of the middle ear.

Sounds that are picked up by the microphone are carried to the external sound processor, which transforms sound into coded signals that are then transmitted transcutaneously to the implanted internal receiver. The receiver converts the incoming signals to electrical impulses that are then conveyed to the electrode array, ultimately resulting in stimulation of the auditory nerve.

Regulatory Status

Several cochlear implants are commercially available in the U.S. and are manufactured by Cochlear Corporation, Advanced Bionics, and the Med El Corporation. Over the years, subsequent generations of the various components of the devices have been approved by the U.S. Food and Drug Administration (FDA), focusing on improved electrode design and speech-processing capabilities. Furthermore, smaller devices and the accumulating experience in children have resulted in broadening of the selection criteria to include children as young as 12 months. The labeled indications from the FDA for currently marketed implant devices are summarized below.

FDA Approved Cochlear Implant Systems

Manufacturer and Currently Marketed	Advanced Bionics® HiResolution Bionic Ear System (HiRes 90K)	Cochlear® Nucleus 5	Med El® Maestro (Sonata or Pulsar)
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Cochlear Implants			
Predecessor Cochlear Implants	Clarion Multi-Strategy or HiFocus CII Bionic Ear (P940022)	Nucleus 22, 24, Freedom with Contour (P840024)	Combi 40+ (P000025)
Indications	<p>Adults:</p> <ul style="list-style-type: none"> • ≥ 18 years of age • Post-lingual onset of severe to profound bilateral sensorineural hearing loss (≥ 70 decibels (dBs)) • Limited benefit from appropriately fitted hearing aids, defined as scoring $\leq 50\%$ on a test of open-set Hearing in Noise Test (HINT) sentence recognition <p>Children:</p> <ul style="list-style-type: none"> • 12 months to 17 years of age • Profound bilateral sensorineural deafness (>90dB) • Use of appropriately fitted hearing aids for at least 6 months in children 2 to 17 years of age or at least 3 months in children 12 to 23 months of age. • Lack of benefit in children <4 years of age is defined as a failure to reach developmentally-appropriate auditory milestones (e.g., spontaneous response to name in quiet or to environmental sounds) measured using the Infant-Toddler Meaningful Auditory Integration Scale or Meaningful Auditory Integration Scale or $<20\%$ correct on a simple open-set word recognition test 	<p>Adults:</p> <ul style="list-style-type: none"> • ≥ 18 years old • Pre- or post-lingual onset of moderate to profound bilateral sensorineural hearing loss • $\leq 50\%$ sentence recognition in the ear to be implanted • $\leq 60\%$ sentence recognition in the opposite ear or binaurally <p>Children 25 months to 17 years 11 months:</p> <ul style="list-style-type: none"> • Severe to profound bilateral sensorineural hearing loss • Multi-syllabic Lexical Neighborhood Test (MLNT) scores of $\leq 30\%$ in best-aided condition in children 25 months to 4 years 11 months • Lexical Neighborhood Test (LNT) scores of $\leq 30\%$ in best-aided condition in children 5 years to 17 years and 11 months • Lack of progress in the development of 	<p>Adults:</p> <ul style="list-style-type: none"> • ≥ 18 years old • Severe to profound bilateral sensorineural hearing loss (≥ 70dB) • $\leq 40\%$ correct Hearing in Noise test (HINT) sentences with best-sided listening condition <p>Children:</p> <ul style="list-style-type: none"> • 12 months to 18 years with profound sensorineural hearing loss (≥ 90dB) • In younger children, little or no benefit is defined by lack of progress in the development of simple auditory skills with hearing aids over a 3-6 month period • In older children, lack of aided benefit is defined as $<20\%$ correct on the MLNT or LNT depending upon the child's cognitive ability and linguistic

	<p>(Multisyllabic Lexical Neighborhood Test) administered using monitored live voice (70 dB SPL [sound pressure level])</p> <ul style="list-style-type: none"> • Lack of hearing aid benefit in children >4 years of age is defined as scoring <12% on a difficult open-set word recognition test (Phonetically Balanced-Kindergarten Test) or <30% on an open-set sentence test (HINT for Children) administered using recorded materials in the soundfield (70 dB SPL) 	<p>auditory skills</p> <p>Children 12 months to 24 months:</p> <ul style="list-style-type: none"> • Profound sensorineural hearing loss bilaterally • Limited benefit from appropriate binaural hearing aids • Lack of progress in the development of auditory skills 	<p>skills</p> <ul style="list-style-type: none"> • A 3-6 month trial with hearing aids is required if not previously experienced
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HINT: Hearing in Noise Test; IT-MAIS: Infant-Toddler Meaningful Auditory Integration Scale; LNT: Lexical Neighborhood Test; MAIS: Meaningful Auditory Integration Scale; MLNT: Multisyllabic Lexical Neighborhood Test; PBK: Phonetically Balanced-Kindergarten; SPL: sound pressure level.

^a Cochlear Ltd. voluntarily recalled the Nucleus CI500 range in September 2011 for device malfunction in the CI512 implant. The external Nucleus 5 sound processor is not a part of the recall. Advanced Bionics HiRes90K was voluntarily recalled in November 2010 and given FDA-approval for re-entry to market the device in September 2011.

In March 2014, FDA approved the Nucleus® Hybrid™ L24 Cochlear Implant System (Cochlear Corporation, Centennial, CO) through the premarket approval process.⁽¹⁾ This system is a hybrid cochlear implant and hearing aid, with the hearing aid integrated into the external sound processor of the cochlear implant. It is indicated for unilateral use in patients aged 18 years and older who have residual low-frequency hearing sensitivity and severe to profound high-frequency sensorineural hearing loss, and who obtain limited benefit from appropriately fit bilateral hearing aid. The electrode array inserted into the cochlea is shorter than conventional cochlear implants. According to the FDA's premarket approval notification, labeled indications for the device include:

- Preoperative hearing in the range from normal to moderate hearing loss (HL) in the low frequencies (thresholds no poorer than 60 dB HL up to and including 500 Hz).
- Preoperative hearing with severe to profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz ≥ 75 dB HL) in the ear to be implanted.
- Preoperative hearing with moderately severe to profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz ≥ 60 dB HL) in the contralateral ear.
- Consonant-Nucleus-Consonant (CNC) word recognition score between 10% to 60% (inclusively) in the ear to be implanted in the preoperative aided condition and in the contralateral ear equal to or better than that of the ear to be implanted but not more than 80% correct.

Other hybrid hearing devices have been developed but do not have FDA approval, including the Med El® EAS Hearing Implant System.

While cochlear implants have typically been used unilaterally, in recent years, interest in bilateral cochlear implantation has arisen. The proposed benefits of bilateral cochlear implants are to improve

understanding of speech in noise and localization of sounds. Improvements in speech intelligibility may occur with bilateral cochlear implants through binaural summation; i.e., signal processing of sound input from 2 sides may provide a better representation of sound and allow one to separate out noise from speech. Speech intelligibility and localization of sound or spatial hearing may also be improved with head shadow and squelch effects, i.e., the ear that is closest to the noise will be received at a different frequency and with different intensity, allowing one to sort out noise and identify the direction of sound. Bilateral cochlear implantation may be performed independently with separate implants and speech processors in each ear or with a single processor. However, no single processor for bilateral cochlear implantation has been approved by the FDA for use in the U.S. In addition, single processors do not provide binaural benefit and may impair sound localization and increase the signal-to-noise ratio received by the cochlear implant.

Rationale

This policy was originally created in 1995 and was updated regularly with searches of the MEDLINE database. The most recent literature search was performed through April 4, 2014. The following is a summary of the key literature to date. Unless otherwise noted, the policy refers to traditional cochlear implants (ie, not hybrid cochlear implant/hearing aid systems such as the Nucleus Hybrid.)

Cochlear Implantation for Bilateral Hearing Loss

Cochlear Implantation in Adults: Unilateral Stimulation

Cochlear implants are recognized as an effective treatment of sensorineural deafness, as noted in a 1995 National Institutes of Health Consensus Development conference, which offered the following conclusions(2):

- Cochlear implantation has a profound impact on hearing and speech reception in postlingually deafened adults with positive impacts on psychological and social functioning.
- Prelingually deafened adults may also benefit, although to a lesser extent than postlingually deafened adults. These individuals achieve minimal improvement in speech recognition skills. However, other basic benefits, such as improved sound awareness, may meet safety needs.
- Training and educational intervention are fundamental for optimal postimplant benefit.

The effectiveness of cochlear implants has been evaluated in several systematic reviews and technology assessments, both from the U.S. and abroad. In 2009, Bond et al(3) authored a technology assessment in the United Kingdom to investigate the clinical and cost-effectiveness of unilateral cochlear implants (using or not using hearing aids) and bilateral cochlear implants compared with a single cochlear implant (unilateral or unilateral plus hearing aids) for severely to profoundly deaf children and adults. The clinical effectiveness review included 33 papers, 2 of which were randomized controlled trials (RCTs) (deaf children, n=1513; adults, n=1379). They used 62 different outcome measures and overall evidence was of moderate to poor quality. (The authors' summary of the effectiveness of bilateral cochlear implants and cochlear implants in children are summarized in the following appropriate sections.) The authors concluded, "Unilateral cochlear implantation is safe and effective for adults and children and likely to be cost-effective in profoundly deaf adults and profoundly and prelingually deaf children."

Bond et al additionally published 2 systematic reviews on the clinical and cost-effectiveness of unilateral cochlear implants, first focusing on children in 2009(4) and subsequently focusing on adults in 2010.(5) Both reviews were conducted with a literature review that identified 1580 titles and abstracts on cochlear implants. In the 2010 review, the authors identified 9 studies that met their inclusion criteria addressing implantation in adults.(5) The authors found the studies available were methodologically weak and too heterogeneous to perform a meta-analysis. However, they concluded there is sufficient, consistent evidence demonstrating positive benefits with unilateral cochlear implants in severely to profoundly hearing impaired adults when compared with acoustic hearing aids or no hearing support.

In January 2009, the National Institute for Health and Care Excellence (NICE) released technology appraisal guidance 166, *Cochlear Implants for children and adults with severe to profound deafness*.⁽⁶⁾

This guidance was based on this technology assessment report by Bond et al.⁽³⁾

The NICE guidance includes the following recommendations:

1. "Unilateral cochlear implantation is recommended as an option for people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids.
2. For purposes of this guidance, severe to profound deafness is defined as hearing only sounds that are louder than 90 dB HL [hearing level] at frequencies of 2 and 4 kHz without acoustic hearing aids. For adults, adequate benefit from acoustic hearing aids is defined for this guidance as a score of 50% or greater on Bamford-Kowal-Bench (BKB) sentence testing at a sound intensity of 70 dB SPL [sound pressure level].
3. Cochlear implantation should be considered for adults only after an assessment by a multidisciplinary team. As part of the assessment, implant candidates should also have had a valid trial of an acoustic hearing aid for at least 3 months (unless contraindicated or inappropriate)."

In April 2011, a technology assessment was completed by the Tufts Evidence-based Practice Center for the Agency for Health Care Research and Quality (AHRQ) on the effectiveness of cochlear implants in adults.⁽⁷⁾ This assessment examined 22 studies with 30 or more patients and concluded that while the studies reviewed were rated as poor to fair quality, unilateral cochlear implants are effective in adults with sensorineural hearing loss. Pre- and postcochlear implant scores on multisyllable tests and open-set sentence tests demonstrated significant gains in speech perception regardless of whether a contralateral hearing aid was used along with the cochlear implant. Additionally, the assessment found generic and disease-specific health-related quality of life (QOL) improved with unilateral cochlear implants. However, the available evidence was insufficient to draw conclusions on improvements in open-set sentence test scores (ie, >40% and ≤50% or >50% and ≤60%), and any relationship between preimplantation patient characteristics and outcomes (eg, age, duration of hearing impairment, Hearing in Noise Test [HINT] scores and pre- or postlinguistic deafness).

In 2013, Gaylor et al published an update to the AHRQ technology assessment.⁽⁸⁾ Sixteen (of 42) studies published through May 2012 were of unilateral cochlear implants. Most unilateral implant studies showed a statistically significant improvement in mean speech scores, as measured by open-set sentence or multisyllable word tests; meta-analysis of 4 studies revealed a significant improvement in cochlear-implant relevant QOL after unilateral implantation (standard mean difference: 1.71; 95% confidence interval [CI], 1.15 to 2.27). However, these studies varied in design, and there was considerable heterogeneity observed across studies.⁽⁸⁾ Similarly, a 2012 systematic review of 11 studies by Bittencourt et al also concluded cochlear implants improved hearing outcomes over conventional hearing aids in patients with severe to profound postlingual deafness.⁽⁹⁾

In October 2011, Berrettini et al published results of a systematic review of cochlear implant effectiveness in adults.⁽¹⁰⁾ Included in the review were 8 articles on unilateral cochlear implants in advanced age patients. All of the studies reported benefits with cochlear implantation, despite advanced age at time of implant (age 70 years or older). In 6 studies, results were not significantly different between younger and older patients. However, 2 studies reported statistically significant inferior perceptive results (eg, HINT and consonant nucleus consonant test) in older patients. This systematic review also examined 3 studies totaling 56 adults with prelingual deafness who received unilateral cochlear implants. The authors concluded unilateral cochlear implants provided hearing and QOL benefits in prelingually deaf patients, but results were variable.

Cochlear Implantation in Adults: Bilateral Stimulation

While use of unilateral cochlear implants in patients with severe to profound hearing loss has become a well-established intervention,⁽⁷⁾ bilateral cochlear implantation is becoming more common. Many

publications have reported slight to modest improvements in sound localization and speech intelligibility with bilateral cochlear implants, especially with noisy backgrounds but not necessarily in quiet environments. When reported, the combined use of binaural stimulation improved hearing by a few decibels or percentage points.

The 2009 Bond et al technology assessment on the clinical and cost-effectiveness of cochlear implants made the following conclusions on the evidence related to bilateral cochlear implants in adults.(3) The strongest evidence for an advantage from bilateral over unilateral implantation was for understanding speech in noisy conditions. The comparison of bilateral with unilateral cochlear implants plus an acoustic hearing aid was limited by small sample sizes and poor reporting. The authors concluded, "There are likely to be overall additional benefits from bilateral implantation, enabling children and adults to hold conversations more easily in social situations."

In January 2009, the NICE technology appraisal guidance noted above (6) indicates:

1. Simultaneous bilateral cochlear implantation in adults is recommended as an option in people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids *and* are blind or have other disabilities that increase their reliance on auditory stimuli as a primary sensory mechanism for spatial awareness.
2. Sequential bilateral cochlear implantation is not recommended as an option for people with severe to profound deafness.
3. For purposes of this guidance, severe to profound deafness is defined as hearing only sounds that are louder than 90 dB HL at frequencies of 2 and 4 kHz (Hz) without acoustic hearing aids. Adequate benefit from acoustic hearing aids for adults is defined for this guidance as a score of 50% or greater on Bamford-Kowal-Bench (BKB) sentence testing at a sound intensity of 70 dB SPL [sound pressure level].
4. Cochlear implantation should be considered for adults only after an assessment by a multidisciplinary team. As part of the assessment, implant candidates should also have had a valid trial of an acoustic hearing aid for at least 3 months (unless contraindicated or inappropriate)."

Crathorne et al published an update of the NICE systematic review in 2012.(11) The objective was to conduct a systematic review of the clinical and cost-effectiveness of bilateral multichannel cochlear implants compared with unilateral cochlear implantation alone or in conjunction with an acoustic hearing aid in adults with severe to- profound hearing loss. A literature search was updated in July 2011 and January 2012. Nineteen studies were included in this update; 6 of these studies were included in the original NICE review. Two studies were RCTs with waiting list controls, 10 were prospective pre-/post repeated measure or cohort designs, 6 were cross-sectional in design, and 1 was an economic evaluation. The studies were conducted in the U.S. and Europe; all compared bilateral with unilateral implantation, and 2 compared bilateral implants with a unilateral implant plus acoustic hearing aid.(11)

The included studies in the Crathorne review were of moderate-to-poor quality, including 2 RCTs.(11) Meta-analyses could not be performed due to heterogeneity between studies in outcome measures and study design. However, all studies reported that bilateral cochlear implants improved hearing and speech perception. One RCT found a significant binaural benefit over the first ear alone for speech and noise from the front ($12.6\% \pm 5.4\%$, $p < 0.001$) and when noise was ipsilateral to the first ear ($21\% \pm 6\%$, $p < 0.001$), and another RCT found a significant benefit for spatial hearing at 3 months postimplantation compared with preimplantation (mean difference [SD] scores, 1.46 [0.83-2.09], $p < 0.01$). QOL results varied, showing bilateral implantation may improve QOL in the absence of worsening tinnitus.(11)

Van Schoonhoven et al independently published a systematic review in 2013 as an update to the original NICE review.(12) As with the Crathorne review, all studies ($n=19$, published through March 2011) showed a significant bilateral benefit in localization over unilateral cochlear implantation. Similarly, meta-analyses could not be performed due to the heterogeneity of the studies and the level of evidence of the included studies, which was of moderate-to-poor quality.(12)

The April 2011 AHRQ technology assessment, noted earlier, completed by the Tufts Evidence-based Practice Center on the effectiveness of cochlear implants in adults examined 16 studies on bilateral cochlear implantation of fair-to-moderate quality published since 2004.(7) The assessment concluded bilateral cochlear implants provide greater benefits in speech perception test scores, especially in noise, when compared with unilateral cochlear implants (with or without contralateral hearing aids). Significant binaural head shadow benefits were noted along with some benefit in binaural summation, binaural squelch effects, and sound localization with bilateral cochlear implants. However, it was unclear if these benefits were experienced under quiet conditions, although benefits increased with longer bilateral cochlear implant usage indicating a need for longer term studies. Hearing-specific QOL could not be assessed because only 1 study evaluated this outcome. Additionally, the evidence available on simultaneous bilateral implantation was found to be insufficient, although gains were experienced in speech perception using open-set sentences or multisyllable tests compared with unilateral cochlear implants or unilateral listening conditions. The assessment noted longer term studies are needed to further understand the benefits with bilateral cochlear implantation and identify candidacy criteria given the risks of a second surgery and the destruction of the cochlea preventing future medical intervention.

The update by Gaylor et al to the assessment previously reported showed improvement across 13 studies in communication-related outcomes with bilateral implantation compared with unilateral implantation and additional improvements in sound localization compared with unilateral device use or implantation only.(8) The risk of bias varied from medium to high across studies. Based on results from at least 2 studies, QOL outcomes varied across tests after bilateral implantation; meta-analysis was not performed because of heterogeneity in design between the studies.(8)

In the 2011 Berrettini et al review of cochlear implant effectiveness in adults (noted earlier), 13 articles on bilateral cochlear implants were reviewed.(10) Sound localization improved with bilateral cochlear implants compared with monaural hearing in 6 studies. Significant improvements in hearing in noise and in quiet environments with bilateral implants compared with unilateral implants were reported in 10 studies and 7 studies, respectively. Five of the studies reviewed addressed simultaneous implantation, 5 studies reviewed sequential implantation, and 3 studies included a mix of simultaneous and sequential implantation. However, no studies compared simultaneous to sequential bilateral implantation results, and no conclusions could be made on the timing of bilateral cochlear implantation. Smulders et al also examined the timing of bilateral cochlear implantation in a systematic review of 11 studies; 5 studies addressed postlingually deafened adults and 7 studies addressed prelingually deafened children (discussed next).(13) One study on adults showed a delay in the timing of the second implantation resulted in poorer outcomes in quiet environments. Nevertheless, all studies reported benefits with bilateral implants, but all studies were considered to be of poor quality and with a high risk of bias.

Cochlear Implantation in Pediatrics

Similar to the adult population, the evidence related to the use of cochlear implants in children has been evaluated in several systematic reviews and technology assessments.

The 2009 Bond et al technology assessment on cochlear implants made the following observations regarding cochlear implantation in children: All studies in children that compared one cochlear implant with nontechnologic support or an acoustic hearing aid reported gains on all outcome measures. Weak evidence shows greater gain from earlier implantation (before starting school).(3)

The 2009 NICE technology appraisal guidance 166, *Cochlear Implants for children and adults with severe to profound deafness*,(6) noted earlier includes the following recommendations for children:

“Unilateral cochlear implantation is recommended as an option for people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids.

1. Simultaneous bilateral cochlear implantation is recommended as an option for children with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids:
2. Sequential bilateral cochlear implantation is not recommended as an option for people with severe to profound deafness.
3. For purposes of this guidance, severe to profound deafness is defined as hearing only sounds that are louder than 90 dB HL at frequencies of 2 and 4 kHz without acoustic hearing aids. For children, adequate benefit from acoustic hearing aids is defined for this guidance as, speech, language and listening skills appropriate to age, developmental stage, and cognitive ability.
4. Cochlear implantation should be considered for children only after an assessment by a multidisciplinary team. As part of the assessment, children should also have had a valid trial of an acoustic hearing aid for at least 3 months (unless contraindicated or inappropriate)."

As also noted earlier, Bond et al published 2 systematic reviews on the clinical and cost-effectiveness of unilateral cochlear implants, first focusing on children in 2009(4) and subsequently focusing on adults in 2010.(5) Both reviews were conducted with a literature search that identified 1580 titles and abstracts on cochlear implants. In the 2009 review, the authors identified 15 studies that met their inclusion criteria addressing cochlear implantation in children.(4) The authors found the studies available were methodologically weak and too heterogeneous to perform a meta-analysis. However, they concluded there is sufficient, consistent evidence demonstrating positive benefits with unilateral cochlear implants in severely to profoundly hearing impaired children when compared with acoustic hearing aids or no hearing support.

The optimal timing of cochlear implantation in children is of particular interest given the strong associations between hearing and language development. While there is current research investigating the ability to restore hearing by stimulating cochlear hair cell regrowth, cochlear implantation damages the cochlea and eliminates this possibility. However, the potential to restore cochlear function is not foreseeable in the near future. If cochlear implantation is believed to be most beneficial at a younger age, when the nervous system is "plastic," the potential for cochlear hair cell regrowth seems too far in the future to benefit young children and should not be a deterrent to current candidates for a cochlear implant. A number of studies have evaluated the effect of age of implantation on hearing outcomes after cochlear implantation in children.

As reported by Sharma and Dorman, central auditory pathways are "maximally plastic" for a period of about 3.5 years, making a case for earlier cochlear implantation of children with hearing impairment.(14) Stimulation delivered before about 3.5 years of age results in auditory evoked potentials that reach normal values in 3 to 6 months. However, when stimulation occurs after 7 years of age, changes occur within 1 month, but then have little to no subsequent change. Sharma et al observed this result when they reported on auditory development in 23 children with unilateral or bilateral implants.(15) In 1 child who received a bilateral device with implantation of the second ear after age 7 years, the auditory responses in the second ear were similar to that seen in "late-implanted" children.

In 2011, Forli et al conducted a systematic review of 49 studies on cochlear implant effectiveness in children that addressed the impact of age of implantation on outcomes.(16) Heterogeneity of studies precluded performance of a meta-analysis. Early implantation was examined in 22 studies, but few studies compared outcomes of implantations performed before 1 year of age to implantations performed after 1 year of age. Studies suggest improvements in hearing and communicative outcomes in children receiving implants before 1 year of age, although it is not certain whether these improvements are related to duration of cochlear implant usage rather than age of implantation. However, the reviewers noted hearing outcomes have been shown to be significantly inferior in patients implanted after 24 to 36 months. Finally, 7 studies were reviewed that examined cochlear implant outcomes in children with associated disabilities. In this population, cochlear implant outcomes were inferior and occurred more slowly but were considered to be beneficial.

As previously noted, the 1995 National Institutes of Health Consensus Development conference concluded cochlear implants are recognized as an effective treatment of sensorineural deafness.(2) This conference offered the following conclusions regarding cochlear implantation in children:

- Cochlear implantation has variable results in children. Benefits are not realized immediately but rather are manifested over time, with some children continuing to show improvement over several years.
- Cochlear implants in children under 2-years-old are complicated by the inability to perform detailed assessment of hearing and functional communication. However, a younger age of implantation may limit the negative consequences of auditory deprivation and may allow more efficient acquisition of speech and language. Some children with postmeningitis hearing loss under the age of 2 years have received an implant due to the risk of new bone formation associated with meningitis, which may preclude a cochlear implant at a later date.

Several systematic reviews have evaluated outcomes after cochlear implantation for specific causes of deafness and in subgroups of pediatric patients. In a 2011 systematic review of 38 studies, Black et al sought to identify prognostic factors for cochlear implantation in pediatric patients.(17) A quantitative meta-analysis was not able to be performed due to study heterogeneity. However, 4 prognostic factors: age at implantation, inner ear malformations, meningitis, and Connexin 26 (a genetic cause of hearing loss), consistently influenced hearing outcomes.

Pakdaman et al conducted a systematic review of cochlear implants in children with cochleovestibular anomalies in 2011.(18) Anomalies included inner ear dysplasia such as large vestibular aqueduct and anomalous facial nerve anatomy. Twenty-two studies were reviewed totaling 311 patients. The authors found implantation surgery was more difficult and speech perception was lower in patients with severe inner ear dysplasia. However, heterogeneity in the studies limited interpretation of these findings.

In another 2011 systematic review, Roush et al examined the audiologic management of children with auditory neuropathy spectrum disorder.(19) The review included 15 studies that addressed cochlear implantation in these patients. All of the studies reported auditory benefit with cochlear implantation in children with auditory neuropathy spectrum disorder. However, the studies were noted to be limited methodologically and further research is needed in this population.

In 2014, Eze et al published a systematic review comparing outcomes for cochlear implantation for children with developmental disability with those without developmental disability.(20) The authors note that while approximately 30% to 40% of children who receive cochlear implants have developmental disability, evidence about outcomes in this group is limited. Their review included 13 studies that compared receptive or expressive language outcomes in children with cochlear implants with and without developmental disability. The included studies were heterogeneous in terms of comparator groups and outcome measures, precluding data pooling and meta-analysis. In a structured systematic review, the authors report that 7 of the eligible studies demonstrate a significantly poor cochlear implant outcome in children with developmental disability, while the remaining studies reported no significant difference in outcomes between the groups.

Humphriss et al published a systematic review evaluating outcomes after cochlear implantation among pediatric patients with auditory neuropathy spectrum disorder (ANSD), a sensorineural hearing disorder characterized by abnormal auditory brainstem response with preserved cochlear hair cell function as measured by otoacoustic emissions testing.(21) The authors identified 27 studies that included an evaluation of cochlear implantation in patients with ANSD, including 15 noncomparative studies, 1 that compared children with ANSD who received a cochlear implant with children with ANSD with hearing aids, and 12 that compared children with ANSD who received a cochlear implant with children with severe sensorineural hearing loss who received a cochlear implant. Noncomparative studies were limited in that most (11/15) did not include a measure of speech recognition before cochlear implantation. Among the comparative studies, those comparing cochlear implantation to “usual care”, typically a hearing aid, provide the most information about effectiveness of cochlear implantation

among patients with ANSD; the 1 small study that used this design found no significant differences between the groups. Overall, the authors suggest that further RCT evidence is needed.

Cochlear Implantation in Infants Younger Than 12 Months

While currently-available cochlear implants have FDA labeling for only children older than 12 months, earlier diagnosis of congenital hearing loss with universal hearing screening has prompted interest in cochlear implantation in children younger than 12 months.

A number of small studies from outside the U.S. have reported results on cochlear implantation in infants younger than 12 months, which would be an off-label indication. For example, in a study from Australia, Ching et al(22) published an interim report on early language outcomes of children with cochlear implants. This study evaluated 16 children who had implants before 12 months of age compared with 23 who had implants after 12 months (specific time of implantation was not provided). The preliminary results demonstrated that children who received an implant before 12 months of age developed normal language skills at a rate comparable with normal-hearing children, while those with later implants performed at 2 standard deviations below normal. The authors noted that these results are preliminary, as there is a need to examine the effect of multiple factors on language outcomes and the rate of language development. Similarly, in a study from Italy, Colletti(23) reported on findings from 13 infants who had implants placed before 12 months. The procedures were performed between 1998 and 2004. In this small study, the rate of receptive language growth for these early implant infants overlapped scores of normal-hearing children. This overlap was not detected for those implanted at 12 to 23 or 24 to 36 months. Data from these small studies are viewed as preliminary and not conclusive. Subsequently, Colletti et al reported on the 10-year results comparing 19 children with cochlear implants received between the ages of 2 and 11 months with 21 children implanted between 12 and 23 months and 33 children implanted between 24 and 35 months.(24) Within the first 6 months postimplantation, there was no significant difference among groups in Category of Auditory Performance testing, but differences became significantly better in the infant group (early implantation) at the 12- and 36-month testing.

Johr et al(25) highlighted the surgical and anesthetic considerations when performing cochlear implant surgery in very young infants (<1 year of age). This is an observational and literature review by pediatricians at a tertiary children's hospital in Switzerland. Patients younger than 1 year of age undergoing cochlear implant surgeries were analyzed concerning surgical techniques and anesthetic management aspects of elective surgeries in small infants. The results demonstrated that the age of the patient and the pediatric experience of the anesthesiologist, but not the duration of the surgery, are relevant risk factors. The authors concluded, "Further research is needed to provide more conclusive evidence that the performance outcome for children implanted before 12 months of age does not converge with the results of children implanted between 12 and 18 months." Currently, there is no conclusive published evidence to support performing cochlear implant surgery on an infant younger than 12 months of age.

In 2010, Vlastarakos et al, conducted a systematic review of studies on bilateral cochlear implants in a total of 125 children implanted before age 1.(26) The authors noted follow-up times ranged from a median duration of 6 to 12 months and, while results seemed to indicate accelerated rates of improvement in implanted infants, the evidence available is limited and of lower quality. Additionally, the lack of reliable outcome measures for infants demonstrates the need for further research before widespread cochlear implantation before 1 year of age.

Cochlear Implantation in Children: Bilateral Stimulation

In a 2014 systematic review, Lammers et al summarized the evidence on the effectiveness of bilateral cochlear implantation compared with unilateral implantation among children with sensorineural hearing loss.(27) The authors identified 21 studies that evaluated bilateral cochlear implantation in children, with no RCTs identified. Due to a limited number of studies, heterogeneity in outcomes and comparison

groups and high risk for bias in the studies, the authors were unable to perform pooled statistical analyses, so a best-evidence synthesis was performed. The best-evidence synthesis demonstrated that there is consistent evidence indicating the benefit of bilateral implantation for sound localization. One study demonstrated improvements in language development, although other studies found no significant improvements. The authors noted that the currently available evidence consists solely of cohort studies that compare a bilaterally implanted group with a unilaterally implanted control group, with only one study providing a clear description of matching techniques to reduce bias.

In 2010, Sparreboom et al conducted a systematic review of bilateral cochlear implants in children with severe-to-profound deafness.(28) Due to the heterogeneity of the studies identified, the authors were unable to perform a meta-analysis. A qualitative review of the studies found binaural ability takes time to develop; bilateral cochlear implants seem to provide better speech perception over unilateral implants; and delays in implanting the second cochlear implant seem to decrease speech perception in quiet and decrease or eliminate the potential for binaural summation. The authors concluded while bilateral cochlear implants provide benefits of bilateral hearing in children, further research is needed.

As noted, Smulders et al examined the timing of sequential bilateral cochlear implantation in a systematic review of 11 studies; 5 studies addressed postlingually deafened adults (previously discussed), and 7 studies addressed prelingually deafened children.(13) Sound localization was not affected by second implantation delay in any study of the studies on children, but delays in second implantation resulted in poorer outcomes in quiet environments in 1 study and poorer outcomes in noise in 2 studies. However, all studies were considered to be of poor quality and with a high risk of bias.

Several publications not included in the Lammers and Sparreboom systematic reviews have evaluated bilateral cochlear implants in children. Broomfield et al reported surgical safety results from bilateral cochlear implants from a prospective registry of pediatric patients who received cochlear implants starting in 2009.(29) The study included 1397 cochlear implant procedures in 961 patients who were implanted at one of 14 centers in the United Kingdom from January 2010 to December 2011. The procedures included 436 bilateral simultaneous, 394 bilateral sequential, and 131 unilateral implants. Overall, 15 major complications (defined as events that necessitated further major surgical intervention, intensive care unit admission, exposure to invasive intervention, permanent disability, or device failure), including 6 device failures. Overall, this study suggests that bilateral cochlear implantation has a reasonable safety profile in children.

Sarant et al conducted a cohort study comparing language development among 91 Australian children aged 5 to 8 years implanted with either unilateral or bilateral cochlear implants.(30) Subjects had been implanted early, with a first implant by 3.5 years of age and a second implant (if bilaterally implanted) by 6 years of age. Children with bilateral implants had significantly better vocabulary outcomes and significantly better scores on overall and expressive language subscales of the Clinical Evaluation of Language fundamentals.

Illg et al reported results from a retrospective case series of 73 children and adolescents who underwent sequential bilateral cochlear implantation with a long (>5 year) interval between implants.(31) The primary outcome measures were speech perception testing with the German Freiburger Monosyllabic Word test in quiet and the German Hochmair-Desoyer, Schutlz, Moser Sentence Test in quiet and in noise. Performance on the second implanted side was worse than the primary implanted side, with the interimplant interval significantly associated with outcomes.

Cochlear Implantation for Unilateral Hearing Loss

As noted, a number of potential benefits to binaural hearing exist, including binaural summation, which allows improved signal detection threshold, and sound localization. The potential benefits from binaural hearing have prompted interest in cochlear implantation for patients with unilateral hearing loss.

In 2013, Vlastarakos et al published a systematic review of the evidence related to cochlear implantation for single-sided deafness.(32) The authors included 17 studies, including prospective and retrospective comparative studies, case series, and case reports, that included 108 patients. The authors report that sound localization is improved after cochlear implantation, although statistical analysis was not included in some of the relevant studies. In most patients (95%), unilateral tinnitus improved. The authors note that most of the studies included had short follow-up times, and evaluation protocols and outcome measurements were heterogeneous.

Some of the representative studies on cochlear implantation for unilateral hearing loss follow.

Arndt et al, for example, published a German pilot study in 2010 of 11 adult patients with unilateral hearing loss of various causes.(33) The aim was to evaluate the use of unilateral electrical stimulation with normal hearing on the contralateral side and after a period of 6 months compared with the preoperative unaided situation, conventional contralateral routing of signal or bone-anchored hearing aid hearing aids. Ten (of 11) patients also suffered from tinnitus. Two tests were used to assess speech comprehension, localization was assessed using an array of multiple speakers, and QOL was evaluated using 3 questionnaires. The study results were presented as p values without adjustment for multiple testing. The authors reported that cochlear implantation improved hearing abilities in these study patients and was superior to the above alternative treatment options. The use of the cochlear implant did not interfere with speech understanding in the normal-hearing ear.

The application of cochlear implants for tinnitus relief in patients with unilateral deafness has also been described in previous studies. Van de Heyning et al, for example, published a study in 2008 of 21 patients with unilateral hearing loss accompanied by severe tinnitus for at least 2 years who underwent cochlear implants at a university center in Belgium.(34) Three (of 21) patients showed complete tinnitus relief, whereas most demonstrated a significant reduction in tinnitus loudness based on a visual analogue scale (2 years after implantation, 2.5 ± 1.9 ; before implantation, 8.5 ± 1.3). Based on the data and narrative reviews, the evidence-base to date on unilateral hearing loss is based on a few observational studies with a small number of patients ($n \leq 30$), with a tendency toward reporting bias across these studies.(35)

In 2013, Hansen et al reported results of a prospective study of cochlear implantation for severe-to-profound single-sided sensorineural hearing loss in 29 patients, 10 of whom had single-sided deafness due to Meniere disease.(36) Performance was compared pre- with postimplant within each subject; outcomes were measured at 3, 6, and 12 months postoperatively. Patients showed significant improvements in CNC word and AzBio sentence scores in the implanted ear pre- and postimplant. For the 19 patients with pre- and postoperative data available, the average improvement on CNC word score was 28% (range, -26%-64%). The average AzBio score improvement was 40% (range, -57%-92%).

Tavora-Vieira et al reported results of a prospective case series that included 9 postlingually deaf subjects with unilateral hearing loss, with or without tinnitus in the ipsilateral ear, with functional hearing in the contralateral ear, who underwent cochlear implantation.(37) Speech perception was improved for all subjects in the “cochlear implant on” state compared with the “cochlear implant off” state, and subjects with tinnitus generally reported improvement.

Hybrid Cochlear Implantation

A concern about traditional cochlear implants is that the implantation process typically destroys any residual hearing, particularly for hearing in the low-frequency ranges. Newer devices have used a shorter cochlear electrode in combination with a hearing aid-like amplification device to attempt to mitigate the damage to the cochlea and permit residual hearing.

In March 2014, FDA approved Nucleus® Hybrid™ L24 Cochlear Implant System for use through the premarket approval process. According to FDA’s Summary of Safety and Effectiveness Data, the

approval was based on 2 clinical studies conducted outside of the U.S. and 1 pivotal study of the Hybrid L24 device conducted under investigational device exemption.(1) The pivotal trial was a prospective, multicenter, 1-arm, nonrandomized, nonblinded, repeated-measures clinical study among 50 subjects at 10 U.S. sites. Performance was compared pre- with postimplant within each subject; outcomes were measured at 3, 6, and 12 months postoperatively. Postoperatively, patients' hearing was evaluating in 3 states: hybrid (simultaneous electric and acoustic stimulation in the implanted ear via the Hybrid L24 including the acoustic component), bimodal (electric stimulation only using the Hybrid L24 minus the acoustic component with contralateral acoustic stimulation), and combined (electric and acoustic stimulation via the Hybrid L24 and contralateral acoustic stimulation). Results from the bimodal and combined conditions were grouped into an "Everyday Listening" category, which was not prospectively defined by the manufacturer. All 50 subjects enrolled underwent device implantation and activation. One subject had the device explanted and replaced with a standard cochlear implant between the 3- and 6-month follow-up visit due to profound loss of low-frequency hearing; an additional subject was explanted before the 12-month follow-up visit and 2 additional subjects were explanted after 12 months. For the 2 primary effectiveness end points, CNC word-recognition score and AzBio sentence-in-noise score, a measure of sentence understanding in noisy environments, there were significant within-subject improvements from baseline to 6-month follow-up. The mean improvement in CNC word score was 35.7% (95% CI, 27.8% to 43.6%); for AzBio score, the mean improvement was 32.0% (95% CI 23.6% to 40.4%) For safety outcomes, 71 adverse events were reported, most commonly profound/total loss of hearing (occurring in 44% of subjects) with at least 1 adverse event occurring in 34 subjects (68%).

Lenarz et al reported results of a prospective multicenter European study evaluating the Nucleus Hybrid™ L24 system.(38) The study enrolled 66 adults with bilateral severe-to-profound high-frequency hearing loss. At 1 year postoperatively, 65% of subjects had significant gains in speech recognition in quiet, and 73% had significant gains in noisy environments. Compared with the cochlear implant hearing alone, residual hearing significantly increased speech recognition scores.

The Nucleus Hybrid L24 system was designed with a shorter cochlear implant with the intent of preserving low-frequency hearing. Gifford et al compared hearing outcomes pre- and postimplantation for 44 adult cochlear implant recipients with preserved low-frequency hearing in 2 test conditions: cochlear implant plus low-frequency hearing in the contralateral plus low-frequency hearing in the contralateral ear (bimodal condition) and cochlear implant plus low-frequency hearing in both ears (best-aided condition).(39) The authors report that there were small but statistically significant differences in improvements in adaptive sentence recognition and speech recognition in a noisy "restaurant" environment, suggesting that the presence of residual hearing is beneficial.

Ongoing Clinical Trials

A search of the database ClinicalTrials.gov on April 4, 2013, identified the following prospective, comparative studies evaluating cochlear implants:

- Children's Bilateral Cochlear Implantation in Finland (FinBiCI) (NCT00960102) – This is a prospective, nonrandomized comparative trial to compare bilateral cochlear implants with unilateral cochlear implant and hearing aid among patients up to age 24 months with congenital severe or profound bilateral hearing impairment. Primary outcomes include assessment of auditory performance skills. Enrollment is planned for 40 subjects; the planned study completion date is December 2014.
- Evaluation of Revised Indications (ERID) for Cochlear Implant Candidacy for the Adult CMS Population (NCT02075229) – This is a prospective case-control study designed to evaluate the safety and efficacy of multichannel cochlear implant systems for newly implanted adults with an indication that expands current Center for Medicare and Medicaid Services. The primary outcome includes the Hearing in Noise Test at baseline and postimplantation. Enrollment is planned for 90 subjects; the planned study completion date is January 2016.

- Outcomes In Children With Developmental Delay And Deafness (NCT01256229) – This is a prospective, randomized, open label trial that includes an experimental group, children with developmental delay who are randomized hearing aids or cochlear implant, and an active control group, children without developmental delay who receive a cochlear implant. Enrollment is planned for 252 subjects; the planned study completion date is August 2014.

Clinical Input Received Through Physician Specialty Societies and Academic Medical Centers

In response to requests, input was received through 2 physician specialty societies and 4 academic medical centers while this policy was under review for February 2010. In addition, unsolicited input was received from a specialty society. While the various physician specialty societies and academic medical centers may collaborate with and make recommendations during this process, through the provision of appropriate reviewers, input received does not represent an endorsement or position statement by the physician specialty societies or academic medical centers, unless otherwise noted. Most of those providing input supported use of cochlear implants in infants younger than 12 months of age; many of those supporting this use noted that there are major issues determining hearing level in infants of this age group, and others commented that use could be considered in these young infants in certain situations only. Those providing input were divided in their comments regarding the medical necessity of upgrading functioning external systems; some agreed with this and others did not.

Summary

A cochlear implant is a device for people with severe-to-profound hearing loss who only receive limited benefit from amplification with hearing aids. A cochlear implant provides direct electrical stimulation to the auditory nerve, bypassing the usual transducer cells that are absent or nonfunctional in deaf cochlea.

The available evidence, summarized in multiple systematic reviews and technology assessments, is sufficient to conclude that cochlear implants improve hearing outcomes for both adults and children. Studies show consistent improvement in speech reception (especially in noise) and in sound localization with bilateral devices. Studies also suggest that earlier implantation may be preferred. Based on these studies, and several systematic reviews that have provided additional evidence in support of unilateral and bilateral cochlear implantation, cochlear implants have been shown to provide benefits sufficient to improve net health outcomes in patients with bilateral hearing loss. Therefore, unilateral and bilateral cochlear implants are considered medically necessary for individuals with bilateral hearing loss in individuals aged 12 months and older.

The available evidence for the use of cochlear implants in improving outcomes for patients with unilateral hearing loss, with or without tinnitus, is limited by small sample sizes, short follow-up times, and heterogeneity in evaluation protocols and outcome measurements.

Future controlled studies with appropriate patient selection comparing cochlear implants with alternative treatment options are needed. Therefore, cochlear implantation as a treatment for patients with unilateral hearing loss is considered to be investigational.

Hybrid cochlear implant/hearing aid systems have not been demonstrated to improve outcomes compared with either a cochlear implant alone; therefore, they are considered to be investigational.

Practice Guidelines and Position Statements

The American Academy of Otolaryngology-Head and Neck Surgery has a position statement on cochlear implants that was revised in 2014. The Academy “considers unilateral and bilateral cochlear implantation as appropriate treatment for adults and children with severe to profound hearing loss. Based on extensive literature demonstrating that clinically selected adults and children can significantly perform better with two cochlear implants rather than one, bilateral cochlear implantation is accepted medical practice.”(40)

In 2006, the American Academy of Otolaryngology-Head and Neck Surgery Foundation (41) released criteria for cochlear implants for adults and pediatric patients.

Adult Criteria

1. Be 18 years or older, with bilateral, severe to profound sensorineural hearing loss, i.e., 70dB or greater PTA (pure-tone air-conduction average) at 500, 1000, and 2000 Hz;
2. Have tried but have limited benefit from adequately fitted binaural hearing aid; or
3. Have sentence recognition score of 50 percent or less in the ear to be implanted and 60 percent or less in the contralateral ear in best aided conditions using Hearing in Noise Test (HINT) or City University of New York (CUNY) tests.

Pediatric Criteria

1. Be 12 months to 17 years of age.
2. Infants age 12-24 months should have bilateral, profound hearing loss with thresholds of 90dB or greater at 1000 Hz.
3. Children 24 months to 17 years should have bilateral severe to profound (greater than 70dB) hearing loss.
4. Infants and older children should demonstrate lack of progress in simple auditory skills in conjunction with appropriate auditory amplification and participation in intensive aural habilitation for three to six months. Less than 0.14520 percent correct on the Multi-syllabic Lexical Neighborhood Test (MLNT) or Lexical Neighborhood Test (LNT), depending on the child's cognitive and linguistic abilities.
5. A three-to six-month trial of appropriate hearing aids is required. If meningitis is the cause of hearing loss or if there is radiologic evidence of cochlear ossification a shorter hearing aid trial and earlier implantation may be reasonable."

In April 2011, a technology assessment was completed by the Tufts Evidence-based Practice Center for the AHRQ on the effectiveness of cochlear implants in adults.(7) The assessment conclusions are noted above.

In January 2009, NICE released technology appraisal guidance 166,(6) "Cochlear Implants for children and adults with severe to profound deafness," which includes recommendations for use of unilateral and bilateral cochlear implants in children and adults as noted above.

In May 2008, the British Cochlear Implant Group released a position paper on bilateral cochlear implants.(42) The position paper includes the following regarding indications for use of bilateral implantation:

- "For all profoundly deaf children in order to stimulate both auditory pathways and optimize speech, language and auditory development and maximize potential academic achievement.
- For all profoundly deaf adults, unable to benefit from bimodal hearing;
- For patients following meningitis or other risk of ossification, where failure to implant may result in obliteration of the cochlea, preventing future stimulation;
- For patients with additional sensory handicap, where there is greater reliance on binaural hearing;
- For patients who experience a loss of performance in the first implanted ear or loss of device function in the first ear but re-implantation in the same ear is contra-indicated;
- For patients who agree to participate in research studies into bilateral implantation."

Cochlear implants are recognized as an effective treatment of sensorineural deafness, as noted in a 1995 National Institutes of Health Consensus Development conference, which offered the following conclusions(2):

- Cochlear implantation has a profound impact on hearing and speech reception in postlingually deafened adults with positive impacts on psychological and social functioning.
- Prelingually deafened adults may also benefit, although to a lesser extent than postlingually deafened adults. These individuals achieve minimal improvement in speech recognition skills. However, other basic benefits, such as improved sound awareness, may meet safety needs.
- Training and educational intervention are fundamental for optimal postimplant benefit.

The conference offered the following conclusions regarding cochlear implantation in children:

- Cochlear implantation has variable results in children. Benefits are not realized immediately but rather are manifested over time, with some children continuing to show improvement over several years.
- Cochlear implants in children under 2-years-old are complicated by the inability to perform detailed assessment of hearing and functional communication. However, a younger age of implantation may limit the negative consequences of auditory deprivation and may allow more efficient acquisition of speech and language. Some children with postmeningitis hearing loss under the age of 2 years have received an implant due to the risk of new bone formation associated with meningitis, which may preclude a cochlear implant at a later date.

Medicare National Coverage

Existing national coverage states(43), "...cochlear implantation may be covered for treatment of bilateral pre- or-post-linguistic, sensorineural, moderate-to-profound hearing loss in individuals who demonstrate limited benefit from amplification" which "is defined by test scores of less than or equal to 40% correct in the best-aided listening condition on tape-recorded tests of open-set sentence cognition." Coverage for cochlear implants may also be provided when the patient has "hearing test scores of greater than 40% and less than or equal to 60% only when the provider is participating in, and patients are enrolled in, either an FDA-approved category B investigational device exemption clinical trial as defined at 42 CFR 405.201, a trial under the Centers for Medicare & Medicaid (CMS) Clinical Trial Policy as defined at section 310.1 of the National Coverage Determinations Manual, or a prospective, controlled comparative trial approved by CMS as consistent with the evidentiary requirements for National Coverage Analyses and meeting specific quality standards."

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Billing Coding/Physician Documentation Information

69930	Cochlear device implantation, with or without mastoidectomy
92507	Treatment of speech, language, voice, communication, and/or auditory processing disorder; individual
92601	Diagnostic analysis of cochlear implant, patient under 7 years of age; with programming
92602	Diagnostic analysis of cochlear implant, patient under 7 years of age; subsequent reprogramming
92603	Diagnostic analysis of cochlear implant, age 7 years or older; with programming
92604	Diagnostic analysis of cochlear implant, age 7 years or older; subsequent reprogramming
92605	Evaluation for prescription of non-speech-generating augmentative and alternative communication device
92606	Therapeutic service(s) for the use of non-speech-generating device, including programming and modification
92607	Evaluation for prescription for speech-generating augmentative and alternative communication device, face-to-face with the patient; first hour
92608	Evaluation for prescription for speech-generating augmentative and alternative communication device, face-to-face with the patient; each additional 30 minutes (List separately in addition to code for primary procedure)
92609	Therapeutic services for the use of speech-generating device, including programming and modification
L8614	Cochlear device, includes all internal and external components
L8615	Headset/headpiece for use with cochlear implant device, replacement
L8616	Microphone for use with cochlear implant device, replacement
L8617	Transmitting coil for use with cochlear implant device, replacement
L8618	Transmitter cable for use with cochlear implant device, replacement
L8619	Cochlear implant, external speech processor and controller, integrated system, replacement

L8621	Zinc air battery for use with cochlear implant device, replacement, each
L8622	Alkaline battery for use with cochlear implant device, any size, replacement, each
L8623	Lithium ion battery for use with cochlear implant device speech processor, other than ear level, replacement, each
L8624	Lithium ion battery for use with cochlear implant device speech processor, ear level, replacement, each
L8627	Cochlear implant, external speech processor, component, replacement
L8628	Cochlear implant, external controller component, replacement
L8629	Transmitting coil and cable, integrated, for use with cochlear implant device, replacement

Additional Policy Key Words

N/A

Policy Implementation/Update Information

10/1/88	New policy, covered with criteria.
3/1/00	No policy statement changes.
3/1/01	Prior Authorization requirement is added. Additional criteria added.
3/1/02	Added to Prosthetics section. Policy statement revised to indicate the use of bilateral cochlear implants is investigational.
5/1/03	Policy statement revised to change minimum age from 18 months to 1 year old.
3/1/04	New CPT codes added. No policy statement changes.
3/1/05	Policy statement revised to include next generation devices as not medically necessary.
3/1/06	No policy statement changes.
3/1/07	No policy statement changes.
6/1/07	Interim change with policy statement revised to indicate bilateral cochlear implantation may be considered medically necessary.
3/1/08	No policy statement changes.
3/1/09	No policy statement changes.
10/1/09	No policy statement changes.
10/1/10	Policy statements modified for clarity; intent of policy statements unchanged
10/1/11	No policy statement changes.
10/1/12	No policy statement changes.
10/1/13	Policy statement added that cochlear implantation as a treatment for patients with unilateral hearing loss with or without tinnitus is considered investigational.
10/1/14	Policy statement added that cochlear implantation with a hybrid cochlear implant/ hearing aid system is considered investigational. Added HCPCS code L8619.

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