Cochlear Implant

Policy Number: 7.01.05  Last Review: 9/2017
Origination: 10/1988  Next Review: 9/2018

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.

Replacement of internal and/or external components is considered medically necessary only in a small subset of members who have inadequate response to existing component(s) to the point of interfering with the individual’s activities of daily living, or the component(s) is/are no longer functional and cannot be repaired. Copies of original medical records must be submitted either hard copy or electronically to support medical necessity.

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 (eg, the Nucleus® Hybrid™ L24 Cochlear Implant System) may be considered medically necessary for patients ages 18 years and older who meet all of the following criteria:

- Bilateral severe-to-profound high-frequency sensorineural hearing loss with residual low-frequency hearing sensitivity; AND
- Receive limited benefit from appropriately fit bilateral hearing aids; AND
- Have the following hearing thresholds:
  - Low-frequency hearing thresholds no poorer than 60 dB hearing level up to and including 500 Hz (averaged over 125, 250, and 500 Hz) in the ear selected for implantation; AND
  - Severe to profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz ≥75 dB hearing level) in the ear to be implanted; AND
*Moderately severe to profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz ≥60 dB hearing level) in the contralateral ear; AND*

*aided consonant-nucleus-consonant word recognition score from 10% to 60% in the ear to be implanted in the preoperative aided condition and in the contralateral ear will be equal to or better than that of the ear to be implanted but not more than 80% correct.*

**When Policy Topic is not covered**

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

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*.

Replacement of internal and/or external components solely for the purpose of upgrading to a system with advanced technology or to a next-generation device is 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, ≥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. Cochlear ossification may prevent electrode insertion, and the absence of cochlear development as demonstrated on computed tomography scans remains an absolute contraindication.

CPT has a range of codes (92601-92609) 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.

**Description of Procedure or Service**

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<th>Populations</th>
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<td>With bilateral sensorineural hearing loss</td>
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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.

For individuals who have bilateral sensorineural hearing loss who receive cochlear implant(s), the evidence includes randomized controlled trials (RCTs) and multiple systematic reviews and technology assessments. Relevant outcomes are
symptoms, functional outcomes, and treatment-related mortality and morbidity. The available studies have reported improvements in speech reception and quality-of-life measures. And, although the available RCTs and other studies measured heterogeneous outcomes and included varying patient populations, the findings are consistent across multiple studies and settings. In addition to consistent improvement in speech reception (especially in noise), studies showed improvements in sound localization with bilateral devices. Studies have also suggested that earlier implantation may be preferred. The evidence is sufficient to determine qualitatively that the technology results in a meaningful improvement in the net health outcome.

For individuals who have unilateral sensorineural hearing loss who receive cochlear implant(s), the evidence includes prospective and retrospective studies reporting within-subjects comparisons and systematic reviews of these studies. Relevant outcomes are symptoms, functional outcomes, and treatment-related mortality and morbidity. Given the natural history of hearing loss, pre- and postimplantation comparisons may be appropriate for objectively measured outcomes. However, 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. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have high-frequency sensorineural hearing loss with preserved low-frequency hearing who receive a hybrid cochlear implant that includes a hearing aid integrated into the external sound processor, the evidence includes prospective and retrospective studies using single-arm, within-subjects comparison pre- and postintervention and systematic reviews. Relevant outcomes are symptoms, functional outcomes, and treatment-related mortality and morbidity. The available evidence has suggested that a hybrid cochlear implant system is associated with improvements in hearing of speech in quiet and noise. The available evidence has also suggested that a hybrid cochlear implant improves speech recognition better than a hearing aid alone. Some studies have suggested that a shorter cochlear implant insertion depth may be associated with preserved residual low-frequency hearing, although there is uncertainty about the potential need for reoperation after a hybrid cochlear implantation if there is loss of residual hearing. The evidence is insufficient to determine the effects of the technology on health outcomes.

Clinical input strongly supported the use of a hybrid cochlear implant for patients with high-frequency hearing loss but preserved low-frequency hearing.

**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.

<table>
<thead>
<tr>
<th>Variables</th>
<th>Advanced Bionics® HiResolution Bionic Ear System (HiRes 90K)</th>
<th>Cochlear® Nucleus 5</th>
<th>Med El® Maestro (Sonata or Pulsar)</th>
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<td>Predicate devices</td>
<td>Clarion Multi-Strategy or HiFocus CII Bionic Ear (P940022)</td>
<td>Nucleus 22, 24, Freedom with Contour (P840024)</td>
<td>Combi 40+ (P000025)</td>
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| Adults      | ● ≥18 y  
● Postlingual onset of severe to profound bilateral sensorineural HL (≥70 dB)  
● Limited benefit from appropriately fitted hearing aids, defined as scoring ≤50% on a test of open-set HINT sentence recognition | ● ≥18 y  
● Pre-, peri-, or postlingual onset of bilateral sensorineural HL, usually characterized by:  
  ○ Moderate-to-profound hearing loss in low frequencies; and  
  ○ Profound (≥90 dB HL) HL in mid-to-high speech frequencies  
● Limited benefit from binaural hearing aids (≤50% sentence recognition in ear to be implanted) | ● ≥18 y  
● Severe to profound bilateral sensorineural HL (≥70 dB)  
● ≤40% correct HINT sentences with best-sided listening condition |
| Children   | ● 12 mo to 17 y of age  
● Profound bilateral sensorineural deafness (≥90 dB)  
● Use of appropriately fitted hearing aids for at least 6 mo in children 2-17 | 25 mo to 17 y 11 mo  
● Severe to profound bilateral sensorineural HL  
● MLNT scores ≤30% in best-aided condition in children 25 mo to 4 y 11 mo | ● 12 mo to 18 y with profound sensori-neural HL (≥90 dB)  
● In younger children, little or no benefit is |
### Variables

- y or at least 3 mo in children 12-23 mo
- Lack of benefit in children <4 y defined as a failure to reach developmentally appropriate auditory milestones (eg, spontaneous response to name in quiet or to environmental sounds) measured using IT-MAIS or MAIS or <20% correct on a simple open-set word recognition test (MLNT) administered using monitored live voice (70 dB SPL)
- Lack of hearing aid benefit in children >4 y defined as scoring <12% on a difficult open-set word recognition test (PBK test) or <30% on an open-set sentence test (HINT for Children) administered using recorded materials in the soundfield (70 dB SPL)
- LNT scores ≤30% in best-aided condition in children 5 y to 17 y and 11 mo-12-24 mo
- Profound sensorineural HL bilaterally
- Limited benefit from appropriate binaural hearing aids
- Defined by lack of progress in the development of simple auditory skills with hearing aids over a 3- to 6-mo period
- In older children, lack of aided benefit is defined as <20% correct on the MLNT or LNT, depending on child’s cognitive ability and linguistic skills
- A 3- to 6-mo trial with hearing aids is required if not previously experienced

### HINT: Hearing in Noise Test; HL: hearing loss; 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.

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In March 2014, FDA approved the Nucleus® Hybrid™ L24 Cochlear Implant System (Cochlear Corporation, Centennial, CO) through the premarket approval process. 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.

Although 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 evidence review was originally created in December 1995 and has been updated regularly with searches of the MEDLINE database. The most recent literature search was performed through December 20, 2016. The following is a summary of the key literature to date. Unless otherwise noted, this evidence review refers to traditional cochlear implants (ie, not hybrid cochlear implant/hearing aid systems [eg, the Nucleus Hybrid L24 Cochlear Implant System]).

**Cochlear Implantation for Bilateral Sensorineural 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 (NIH) Consensus Development conference, which offered the following conclusions²:
“Cochlear implantation improves communication ability in most adults with severe to profound deafness and frequently leads to positive psychological and social benefits as well.” “Prelingually deafened adults may also be suitable for implantation, although these candidates must be counseled regarding realistic expectations. Existing data indicate that these individuals achieve minimal improvement in speech recognition skills.

“However, other basic benefits, such as improved sound awareness, may provide psychological satisfaction 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 United States and abroad. In 2009, Bond et al 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 articles, 2 of which were randomized controlled trials (RCTs) (1513 deaf children; 1379 adults). They defined 62 different outcome measures and overall evidence was of moderate-to-poor quality. (Reviewers’ summaries of the effectiveness of bilateral cochlear implants and cochlear implants in children are presented in the following appropriate sections.) Reviewers 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 published 2 other systematic reviews on the clinical and cost-effectiveness of unilateral cochlear implants, first focusing on children in 2009 and then adults in 2010. Both were conducted with a literature review that identified 1580 articles and abstracts on cochlear implants. In the 2010 review, they identified 9 studies that met inclusion criteria addressing implantation in adults; all were methodologically weak and too heterogeneous to perform a meta-analysis. However, reviewers concluded that 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 2009, the National Institute for Health and Care Excellence (NICE) released a technology guidance on cochlear implants for children and adults with severe-to-profound deafness. This guidance was based on Bond’s 2009 technology assessment.

The NICE guidance included the following recommendations:
1.1 “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....
1.5 For the 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. Adequate benefit from acoustic hearing aids is defined for this guidance as:

- for adults, a score of 50% or greater on Bamford-Kowal-Bench (BKB) sentence testing at a sound intensity of 70 dB SPL [sound pressure level].

1.7 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 2011, the Agency for Healthcare Research and Quality (AHRQ) published a technology assessment on the effectiveness of cochlear implants in adults. 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 were effective in adults with sensorineural hearing loss. Pre- and post-cochlear implant scores on multisyllable tests and open-set sentence tests demonstrated significant gains in speech perception whether or not a contralateral hearing aid was used 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%), or any relation 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 updated the AHRQ assessment. Sixteen (of 42) studies published through May 2012 evaluated 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 considerable heterogeneity was observed across studies. 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.

In 2011, Berrettini et al published results of a systematic review on the effectiveness of cochlear implantation in adults. Included in the review were 8 articles on unilateral cochlear implantation in patients of advanced age. All studies reported benefits with cochlear implantation, despite age at time of implant (ages ≥70 years). In 6 studies, results did not differ significantly between younger and older patients. However, 2 studies reported statistically significant inferior perceptive results (eg, HINT and Consonant-Nucleus-Consonant [CNC] test scores) in older patients. This systematic review also examined 3 studies assessing prelingual deafness in 56 adults who received unilateral cochlear implants.
Reviewers concluded that unilateral cochlear implantation 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, 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 technology assessment (described above) made the following conclusions on the evidence related to bilateral cochlear implants in adults. The strongest evidence for an advantage from bilateral over unilateral implantation related to for understanding speech in noisy conditions. The comparison of bilateral to unilateral cochlear implants plus an acoustic hearing aid was limited by small sample sizes and poor reporting. Reviewers 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 2009, the NICE guidance (noted above) indicated:

1.2 “Simultaneous bilateral cochlear implantation [in adults] is recommended as an option for ... people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids ... [and] ... who are blind or who have other disabilities that increase their reliance on auditory stimuli as a primary sensory mechanism for spatial awareness....

1.3 Sequential bilateral cochlear implantation is not recommended as an option for people with severe to profound deafness.

1.5 For the 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. Adequate benefit from acoustic hearing aids is defined for this guidance as:

- for adults, a score of 50% or greater on Bamford-Kowal-Bench (BKB) sentence testing at a sound intensity of 70 dB SPL.

1.6 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. The objective was to evaluate 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 through January 2012. Nineteen studies conducted in the United States and Europe were included in this update; 6 had been 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. All studies compared bilateral with unilateral implantation, and 2 compared bilateral implants with a unilateral implant plus acoustic hearing aid. The studies selected were of moderate-to-poor quality, including the 2 RCTs. Meta-analyses could not be performed due to heterogeneity between studies in outcome measures and study designs. 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%, p<0.001) and when noise was ipsilateral to the first ear (21%, p<0.001); another RCT found a significant benefit for spatial hearing at 3 months postimplantation compared with preimplantation (mean difference, 1.46; p<0.01). QOL results varied, showing bilateral implantation might improve QOL in the absence of worsening tinnitus.

Van Schoonhoven et al (2013) independently published a systematic review as an update to the original NICE review. As with the Crathorne review, all 19 studies (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 selected studies was of moderate-to-poor quality.

The 2011 AHRQ technology assessment (noted earlier) examined the effectiveness of cochlear implants in adults by reviewing 16 studies on bilateral cochlear implantation of fair-to-moderate quality published since 2004. The assessment concluded that bilateral cochlear implants provide greater benefits in speech perception test scores, especially in noise, 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 follow-up. 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 were needed to understand the benefits associated with bilateral cochlear implantation and to identify criteria for appropriate candidates, given the risks of a second surgery and the irreversible changes to the cochlea, which could preclude future medical intervention.

The Gaylor update to the AHRQ 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. 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 designs between studies.

In the 2011 Berrettini review of cochlear implant effectiveness in adults (noted earlier), 13 articles on bilateral cochlear implants were reviewed. Sound localization improved with bilateral cochlear implants compared with monaural hearing in 6 studies. Significant improvements were reported in hearing in noise and in quiet environments with bilateral implants compared with unilateral implants in 10 studies and 7 studies, respectively. Five studies reviewed addressed simultaneous implantation, 5 studies assessed sequential implantation, and 3 studies included a mix of simultaneous and sequential implantation. However, no studies compared simultaneous with sequential bilateral implantation results, and no conclusions could be made on the timing of bilateral cochlear implantation. Smulders et al (2011) 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). One study of adults showed that 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 high risks of bias.

Since the publication of the systematic reviews described above, additional comparative studies (eg, Blamey et al [2015]) and case series (eg, Harkonen et al [2015]) have reported on outcomes after bilateral cochlear implantation. For example, in a 2016 prospective observational study including 113 patients with postlingual hearing loss, of whom 50 were treated with cochlear implants and 63 with hearing aids, cochlear implant recipients’ depression scores improved from preimplantation to 12 months posttreatment (Geriatric Depression Scale score improvement, 31%; 95% CI, 10% to 47%).

**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 technology assessment on cochlear implants made the following observations regarding cochlear implantation in children: All studies in children that compared 1 cochlear implant with nontechnologic support or an acoustic hearing aid reported gains on all outcome measures. Weak evidence showed greater gain from earlier implantation (before starting school). The 2009 NICE guidance noted earlier, included the following recommendations for children:

1.1 “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....

1.2 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....
1.3 Sequential bilateral cochlear implantation is not recommended as an option for people with severe to profound deafness.

1.5 For the 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, a]dequate benefit from acoustic hearing aids is defined for this guidance as:

- speech, language and listening skills appropriate to age, developmental stage, and cognitive ability.

1.6 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 and then adults. Both 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; all were methodologically weak and too heterogeneous to perform a meta-analysis. However, the authors concluded that there is sufficient, consistent evidence demonstrating positive benefits with unilateral cochlear implants in severely to profoundly hearing impaired children compared with acoustic hearing aids or no hearing support.

Cochlear Implant Timing in Pediatrics

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 (2006), central auditory pathways are “maximally plastic” for about 3.5 years, making a case for earlier cochlear implantation of children with hearing impairment. 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 effect. Sharma et al (2005) observed this result when they reported on auditory development in 23 children with unilateral or bilateral implants. 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. Heterogeneity of studies precluded 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 suggested improvements in hearing and communicative outcomes in children receiving implants before 1 year of age, although it is uncertain whether these improvements were related to duration of cochlear implant usage rather than age of implantation. However, 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 noted, the 1995 NIH Consensus Development conference concluded cochlear implants are recognized as an effective treatment of sensorineural deafness. 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 manifest 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.

Studies published since the systematic reviews above have suggested that cochlear implant removal and reimplantation (due to device malfunction or medical/surgical complications) in children is not associated with worsened hearing outcomes.

**Specific Indications for Cochlear Implantation in Pediatrics**

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. A quantitative meta-analysis was not 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 2012. Anomalies included inner ear dysplasia
such as large vestibular aqueduct and anomalous facial nerve anatomy. Twenty-two studies were reviewed (total N=311 patients). Reviewers found implantation surgery was more difficult and speech perception was poorer in patients with severe inner ear dysplasia. Heterogeneity across studies limited interpretation of these findings.

In 2013, Eze et al published a systematic review comparing outcomes for cochlear implantation among children with and without developmental disability. Reviewers noted that while 30% to 40% of children who receive cochlear implants have a 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 in both groups. The selected studies were heterogeneous in terms of comparator groups and outcome measures, precluding data pooling and meta-analysis. In a structured systematic review, reviewers reported that 7 of the eligible studies demonstrated significantly poor outcomes with cochlear implantation in children with developmental disability, while the remaining studies reported no significant differences in outcomes between the 2 groups.

Auditory Neuropathy Spectrum Disorder
Humphriss et al published a systematic review in 2013 that evaluated 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. Reviewers 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 to children with ANSD with hearing aids, and 12 that compared children with ANSD who received a cochlear implant to 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, provided the most information about the effectiveness of cochlear implantation among patients with ANSD; the 1 small study that used this design found no significant differences between the groups. Overall, reviewers suggested that further RCT evidence is needed.

In a 2015 systematic review, Fernandes et al evaluated 18 published studies and 2 dissertations that reported hearing performance outcomes for children with ANSD and cochlear implants. Studies included 4 nonrandomized controlled studies considered high quality, 5 RCTs considered low quality, and 10 clinical outcome studies. Most studies (n=14) compared the speech perception in children with ANSD and cochlear implants to the speech perception in children with sensorineural hearing loss and cochlear implants. Most of these studies concluded that children with ANSD and cochlear implants developed hearing skills similar to those with sensorineural hearing loss and cochlear implants; however, these types
of studies do not permit comparisons across outcomes between ANSD patients treated with cochlear implants and those treated with usual care.

**Cochlear Implantation in Infants Younger Than 12 Months**

While currently available cochlear implants are labeled by the Food and Drug Administration for use in children older than 12 months of age, earlier diagnosis of congenital hearing loss with universal hearing screening has prompted interest in cochlear implantation in children younger than 12 months old.

In 2010, Vlastarakos et al conducted a systematic review of studies on bilateral cochlear implantation in a 125 children implanted before age 1. For this off-label indication, reviewers 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 was limited and of poor quality.

A number of small studies from outside the United States have reported results on cochlear implantation in infants younger than 12 months old. For example, in a study from Australia, Ching et al (2009) published an interim report on early language outcomes among 16 children implanted prior to 12 months of age, compared with 23 who were implanted after 12 months of age (specific timing implantation was not provided). The 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 implanted later performed at 2 standard deviations below normal. Reviewers noted that these results were preliminary, because 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 (2009) reported on findings from 13 infants who had implants placed before 12 months of age. The procedures were performed between 1998 and 2004. In this small study, the rate of receptive language growth for these early-implant infants overlapped with scores of normal-hearing children. This overlap was not detected for those implanted at 12 to 23 or 24 to 36 months of age. Subsequently, Colletti et al (2011) reported on 10-year results among 19 infants with cochlear implants received between the ages of 2 and 11 months (early implantation group) compared with 21 children implanted between the ages of 12 and 23 months and 33 children implanted between the ages of 24 and 35 months. Within the first 6 months postimplantation, there were no significant differences among groups in Category of Auditory Performance testing, but patients in the infant group had greater improvements than older children at the 12- and 36-month testing.

A more recent (2016) prospective study of 28 children with profound sensorineural hearing loss who were implanted early with cochlear implants (mean age at device activation, 13.3 months) reported that these children had social and conversational skills in the range of normal-hearing peers 1 year after device activation.
Cochlear Implantation in Children: Bilateral Stimulation

In a 2014 systematic review, Lammers et al compared the evidence on the effectiveness of bilateral cochlear implantation to unilateral implantation among children with sensorineural hearing loss. Reviewers identified 21 studies that evaluated bilateral cochlear implantation in children, with no RCTs identified. Due to the limited number of studies, heterogeneity in outcomes and comparison groups, and high risk for bias in the studies, reviewers 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. Reviewers noted that the currently available evidence consisted solely of cohort studies that compared a bilaterally implanted group with a unilaterally implanted control group, with only 1 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. Due to the heterogeneity of identified studies, reviewers 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 than 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. Reviewers concluded that, although bilateral cochlear implants provided benefits of bilateral hearing in children, further research would be needed.

As noted earlier, Smulders et al (2011) examined the timing of sequential bilateral cochlear implantation in a systematic review of 11 studies, of which 7 addressed prelingually deafened children. Sound localization was not affected by second implantation delay in any study of children, but delays in second implantation resulted in poorer outcomes in quiet environments in 1 study and poorer outcomes in noise in 2 studies. 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. These studies, ranging in size from 91 to 961 patients, have generally reported improved speech outcomes with bilateral implantation, compared with unilateral implantation. In another retrospective case series (2013) of 73 children and adolescents who underwent sequential bilateral cochlear implantation with a long (>5 year) interval between implants, performance on the second implanted side was worse than the primary implanted side, with outcomes significantly associated with the interimplant interval.
Section Summary: Cochlear Implantation for Bilateral Sensorineural Hearing Loss
Multiple trials of cochlear implantation in patients with bilateral sensorineural hearing loss, although in varying patient populations, have consistently demonstrated improvements in speech recognition in noise and improved sound localization.

Cochlear Implantation for Unilateral Sensorineural Hearing Loss
As noted, a number of potential benefits to binaural hearing exist, including binaural summation, which permits 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.

Systematic Reviews
In 2014, Vlastarakos et al published a systematic review of the evidence related to cochlear implantation for single-sided deafness and/or unilateral tinnitus. Reviewers selected 17 studies (total N=108 patients), including prospective and retrospective comparative studies, case series, and case reports. Reviewers reported that sound localization improved after cochlear implantation, although statistical analysis was not included in some of the relevant studies. In most patients (95%), unilateral tinnitus improved. Reviewers noted that most studies selected had short follow-up times, and evaluation protocols and outcome measurements were heterogeneous.

In 2015, van Zon et al published another systematic review of studies evaluating cochlear implantation for single-sided deafness or asymmetric hearing loss. Reviewers reviewed 15 studies, 9 of which (n=112 patients) were considered of sufficient quality to be included in data review. Reviewers identified no high-quality studies of cochlear implantation in this population. Data were not be pooled for meta-analysis due to high between-study heterogeneity, but reviewers concluded that studies generally report improvements in sound localization, QOL scores, and tinnitus after cochlear implantation, with varying results for speech perception in noise.

In 2014, Blasco and Redleaf published a systematic review of studies evaluating cochlear implantation for unilateral sudden deafness. Reviewers included 9 studies (total N=36 patients). In pooled analysis, subjective improvement in tinnitus occurred in 96% of patients (of 27 assessed), subjective improvement in speech understanding occurred in 100% of patients (of 16 assessed), and subjective improvement in sound localization occurred in 87% of patients (of 16 assessed). The small number of patients assessed for each outcome limited conclusions that may be drawn.

Case Series
Several individual studies have reported on outcomes for cochlear implantation for single-sided deafness since publication of the systematic reviews described above.
The longest follow-up was reported by Mertens et al (2015) in a case series with structured interviews, which included 23 individuals who received cochlear implants for single-sided deafness with tinnitus. Eligible patients had either single-sided deafness or asymmetric hearing loss and ipsilateral tinnitus. Subjects had a mean 8 years of experience with their cochlear implant (range, 3-10 years). Tinnitus symptoms were assessed by structured interview, visual analog scale (VAS), and the Tinnitus Questionnaire (TQ; a validated scale). Patients demonstrated improvements in VAS scores from baseline (mean score, 8) to 1 month (mean score: 4; \( p<0.01 \) vs baseline) and to 3 months (mean score, 3; \( p<0.01 \) vs baseline) after the first fitting. TQ scores improved from baseline to 3 months post fitting (55 vs 31, \( p<0.05 \)) and were stable for the remainder of follow-up.

In 2015, Arndt et al reported outcomes for 20 children who underwent cochlear implantation for single-sided deafness, which represented a portion of their center’s cohort of 32 pediatric patients with single-sided deafness who qualified for cochlear implants. Repeated-measure analyses of hearing data sets were available for 13 implanted children, excluding 5 who had undergone surgery too recently to be evaluated and 2 children who were too young to be evaluated for binaural hearing benefit. There was variability in the change in localization ability across the tested children. Self- or child-reported hearing benefit was measured with the Speech, Spatial and Qualities of Hearing Scale. Significant improvements were reported on the child and parent evaluations for the scale’s total hearing score and 3 subcategories (speech hearing, spatial hearing, hearing quality).

In 2017, Sladen et al retrospectively reviewed prospectively collected data of short-term (6-month) follow-up for 23 adults and children with single-sided deafness from a variety of mechanisms who received a cochlear implant. In the implanted ear, CNC word recognition improved significantly from preimplantation to 3 months postactivation (\( p=0.001 \)). However, for AzBio sentence understanding in noise (+5 dB signal-to-noise), there was no significant improvement from preimplantation to 6 months postactivation.

In a prospective repeated-measures cohort study that included 20 subjects with single-sided deafness implanted with cochlear implants (15 of whom had reached 6-month follow-up), Sladen et al (2016) reported on speech recognition and QOL. Pure-tone audiometry improved with air conduction in the implanted ear. CNC scores in quiet improved from 4.8% in the preoperative period to 42.3% at the 6-month postactivation check in patients who reached that follow-up.

Also in 2016, Rahne et al reported on a retrospective review of 4 children and 17 adults with single-sided deafness treated with cochlear implants and followed for 12 months. Sound localization with aided hearing improved from preimplantation for all individuals. The speech recognition threshold in noise (signal-to-noise) ratio improved from -1.95 dB (CI off, SD=2.7 dB) to -4.0 dB after 3 months (SD=1.3 dB; \( p<0.05 \)), with continued improvements through 6 months.
Earlier studies, such as those reported by Arndt et al (2011), with 11 adult patients, and by Hansen et al (2013), with 29 patients, reported improvements in hearing ability compared with baseline for implanted subjects.

**Cochlear Implant for Tinnitus Relief in Patients With Unilateral Deafness**

The application of cochlear implants for tinnitus relief in patients with unilateral deafness has also been described. Studies of cochlear implants in patients with bilateral hearing loss, which is often associated with tinnitus, have reported improvements in tinnitus. For example, van Zon et al (2016) reported on a prospective study focusing on tinnitus perception conducted as a part of a multicenter RCT comparing unilateral with bilateral cochlear implantation in patients with severe bilateral sensorineural hearing loss. This analysis included 38 adults enrolled from 2010 to 2012 and randomized to simultaneous bilateral or unilateral cochlear implants. At 1 year postimplantation, both unilaterally and bilaterally implanted patients had significant decreases in score on the Tinnitus Handicap Inventory (THI; a validated scale): change in score of 8 to 2 (p=0.03) and from 22 to 12 (p=0.04) for unilaterally and bilaterally implanted patients, respectively. Bilaterally implanted patients had a significant decrease in TQ score: change in score of 20 to 9 (p=0.04).

Based on observations about tinnitus improvement with cochlear implants, several studies have reported on improvements in tinnitus after cochlear implantation in individuals with unilateral hearing loss. For example, in the meta-analysis by Vlastarakos et al (2014) described above, tinnitus improved in most patients (95%).

Ramos Macias et al (2015) reported results of a prospective multicenter study with repeated measures related to tinnitus, hearing, and QOL, among 16 individuals with unilateral hearing loss and severe tinnitus who underwent cochlear implantation. All patients had a severe tinnitus handicap (THI score ≥58%). Eight (62%) of the 13 patients who completed the 6-month follow-up visit reported a lower tinnitus handicap on the THI score. Perceived loudness/annoyingness of the tinnitus was evaluated with a 10-point VAS. When the cochlear implant was on, tinnitus loudness decreased from 8.4 preoperatively to 2.6 at the 6-month follow-up; 11 of 13 patients reported a change in score of 3 or more.

In 2008, Van de Heyning et al published a study 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. Three patients showed complete tinnitus relief, whereas most demonstrated a significant reduction in tinnitus loudness based on VAS scores (2 years after implantation, 2.5; before implantation, 8.5).

Tavora-Vieira et al (2013) 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. 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.

**Section Summary: Cochlear Implantation for Unilateral Sensorineural Hearing Loss**

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.

**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 preserve 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, approval was based on 2 clinical studies conducted outside of the United States and 1 pivotal study of the Hybrid L24 device conducted under investigational device exemption.

The pivotal trial was a prospective, multicenter, 1-arm, nonrandomized, nonblinded, repeated-measures clinical study among 50 subjects at 10 U.S. sites. Results were reported in FDA documentation and in peer-reviewed form by Roland et al (2016). Eligible patients were selected on the basis of having severe high-frequency sensorineural hearing loss (≥70 dB hearing level averaged over 2000, 3000, and 4000 Hz) with relatively good low-frequency hearing (≤60 dB hearing level averaged over 125, 250, and 500 Hz) in the ear selected for implantation.

Performance was compared pre- and postimplant within each subject; outcomes were measured at 3, 6, and 12 months postoperatively. The trial tested 2 coprimary efficacy hypotheses: (1) that outcomes on CNC, a measure of word recognition, and (2) AzBio sentences in noise presented through the hybrid implant system would be better at 6 months postimplantation than preoperative performance using a hearing aid.

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, AzBio sentence-in-noise score), there were significant within-subject improvements from baseline to 6-month follow-up. Mean improvement in CNC word score was 35.8% (95% CI, 27.8% to 43.6%); for AzBio score, mean improvement was 32.0% (95% CI, 23.6% to 40.4%). For safety outcomes, 65 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 (2013) reported results of a prospective multicenter European study evaluating the Nucleus Hybrid L24 system. 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.

**Hearing Benefit With Shorter Cochlear Array**

The Nucleus Hybrid L24 system was designed with a shorter cochlear implant with the intent of preserving low-frequency hearing. A relevant question is whether a shorter implant is associated with differences in outcomes, although studies addressing this question do not directly provide evidence about hybrid implants themselves. Gifford et al (2013) 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 ear (bimodal condition) and cochlear implant plus low-frequency hearing in both ears (best-aided condition). They reported 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.

In 2014, Santa Maria et al published a meta-analysis of hearing outcomes after various types of hearing-preservation cochlear implantation, which included implantation of hybrid devices, cochlear implantation with surgical techniques designed to preserve hearing, and the use of postoperative systemic steroids. Reviewers included 24 studies, but only 2 focused specifically on a hybrid cochlear implant system, and no specific benefit from a hybrid system was reported.

Causon et al (2015) evaluated factors associated with cochlear implant outcomes in a meta-analysis of articles published from 2003 to 2013, which reported on pure-tone audiometry measurements pre- and post-cochlear implantation. Twelve studies with available audiometric data (total N=200 patients) were included. Reviewers standardized degree of hearing preservation after cochlear implant using the HEARRING consensus statement formula. This formula calculates a percentage of hearing preservation at a specific frequency band, which is scaled to the preoperative audiogram by dividing the change in hearing by the difference between the maximum measurable threshold and the preoperative hearing threshold. The association of a variety of patient- and surgery-related factors, including insertion depth, and improvement in low-frequency hearing were evaluated. In this analysis, insertion depth was not significantly associated with low-frequency residual hearing.

In a 2015 retrospective review that included 10 subjects implanted with a cochlear implant with either a standard electrode (n=12) or the Nucleus Hybrid L24
electrode (n=10), loss of acoustic hearing to the severe-profound level occurred in a higher proportion of patients who received a standard electrode (58% vs 30%).

Since the publication of the Santa Maria and Causon studies, which evaluated factors associated with cochlear implant outcomes, additional studies have attempted to evaluate whether shorter cochlear arrays are more likely to preserve hearing.

**Section Summary: Hybrid Cochlear Implantation**

Prospective and retrospective studies using a single-arm, within-subjects comparison pre- and postintervention have suggested that a hybrid cochlear implant system is associated with improvements in hearing of speech in quiet and noise. For patients who have high-frequency hearing loss but preserved low-frequency hearing, the available evidence has suggested that a hybrid cochlear implant improves speech recognition better than a hearing aid alone. Some studies have suggested that a shorter cochlear implant insertion depth may be associated with preserved residual low-frequency hearing, although there is uncertainty about the potential need for reoperation following a hybrid cochlear implantation if there is loss of residual hearing.

**Summary of Evidence**

For individuals who have bilateral sensorineural hearing loss who receive cochlear implant(s), the evidence includes randomized controlled trials (RCTs) and multiple systematic reviews and technology assessments. Relevant outcomes are symptoms, functional outcomes, and treatment-related mortality and morbidity. The available studies have reported improvements in speech reception and quality-of-life measures. Although the available RCTs and other studies measured heterogeneous outcomes and included varying patient populations, the findings are consistent across multiple studies and settings. In addition to consistent improvement in speech reception (especially in noise), studies showed improvements in sound localization with bilateral devices. Studies have also suggested that earlier implantation may be preferred. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have unilateral sensorineural hearing loss who receive cochlear implant(s), the evidence includes prospective and retrospective studies reporting within-subjects comparisons and systematic reviews of these studies. Relevant outcomes are symptoms, functional outcomes, and treatment-related mortality and morbidity. Given the natural history of hearing loss, pre- and postimplantation comparisons may be appropriate for objectively measured outcomes. However, 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. The evidence is insufficient to determine the effects of the technology on health outcomes.
For individuals who have high-frequency sensorineural hearing loss with preserved low-frequency hearing who receive a hybrid cochlear implant that includes a hearing aid integrated into the external sound processor, the evidence includes prospective and retrospective studies using single-arm, within-subjects comparison pre- and postintervention and systematic reviews. Relevant outcomes are symptoms, functional outcomes, and treatment-related mortality and morbidity. The available evidence has suggested that a hybrid cochlear implant system is associated with improvements in hearing of speech in quiet and noise. The available evidence has also suggested that a hybrid cochlear implant improves speech recognition better than a hearing aid alone. Some studies have suggested that a shorter cochlear implant insertion depth may be associated with preserved residual low-frequency hearing, although there is uncertainty about the potential need for reoperation after a hybrid cochlear implantation if there is loss of residual hearing. The evidence is insufficient to determine the effects of the technology on health outcomes.

Supplemental Information

Clinical Input From Physician Specialty Societies and Academic Medical Centers
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.

2016 Input
In response to requests, input was received from 2 specialty societies, one of which provided 4 responses and one of which provided 3 responses, and 3 academic medical centers while this policy was under review in 2016. Clinical input focused on the use of hybrid cochlear implants. Input was consistent that the use of a hybrid cochlear implant/hearing aid device that includes the hearing aid integrated into the external sound processor of the cochlear implant improves outcomes for patients with high-frequency hearing loss but preserved low-frequency hearing.

2010 Input
In response to requests, input was received from 2 physician specialty societies and 4 academic medical centers while this policy was under review in 2010. In addition, unsolicited input was received from a specialty society. Most providing input supported the use of cochlear implants in infants younger than 12 months of age; many supporting this use noted that there are major issues when determining hearing level in infants of this age group, and others commented that use could be considered in these young infants only in certain situations. Those providing input were divided on the medical necessity of upgrading functioning external systems—some agreed and others did not.
Practice Guidelines and Position Statements

American Academy of Otolaryngology – Head and Neck Surgery Foundation
The American Academy of Otolaryngology – Head and Neck Surgery Foundation has a position statement on cochlear implants that was revised in 2014. The Foundation “...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 perform significantly better with two cochlear implants [rather] than one, bilateral cochlear implantation is accepted medical practice.”

Agency for Health Care Research and Quality
In 2011, a technology assessment for the Agency for Health Care Research and Quality assessed the effectiveness of cochlear implants in adults. The assessment conclusions are noted within the body of this evidence review.

National Institute for Health and Care Excellence
In 2009, the National Institute for Health and Care Excellence released technology appraisal guidance on cochlear implants for children and adults with severe-to-profound deafness. This guidance included recommendations for use of unilateral and bilateral cochlear implants in children and adults, as noted within the body of this evidence review.

National Institutes of Health
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:

- Cochlear implantation has a profound impact on hearing and speech perception 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.

**U.S. Preventive Services Task Force Recommendations**
Not applicable.

**Medicare National Coverage**
Existing national coverage states:

“...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.”

**Ongoing and Unpublished Clinical Trials**
Some currently unpublished trials that might influence this review are listed in Table 2.

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NCT: national clinical trial.

References
### Billing Coding/Physician Documentation Information

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>69930</td>
<td>Cochlear device implantation, with or without mastoidectomy</td>
</tr>
<tr>
<td>92507</td>
<td>Treatment of speech, language, voice, communication, and/or auditory processing disorder; individual</td>
</tr>
<tr>
<td>92601</td>
<td>Diagnostic analysis of cochlear implant, patient under 7 years of age; with programming</td>
</tr>
<tr>
<td>92602</td>
<td>Diagnostic analysis of cochlear implant, patient under 7 years of age; subsequent reprogramming</td>
</tr>
<tr>
<td>92603</td>
<td>Diagnostic analysis of cochlear implant, age 7 years or older; with programming</td>
</tr>
<tr>
<td>92604</td>
<td>Diagnostic analysis of cochlear implant, age 7 years or older; subsequent reprogramming</td>
</tr>
<tr>
<td>92605</td>
<td>Evaluation for prescription of non-speech-generating augmentative and alternative communication device</td>
</tr>
<tr>
<td>92606</td>
<td>Therapeutic service(s) for the use of non-speech-generating device, including programming and modification</td>
</tr>
<tr>
<td>92607</td>
<td>Evaluation for prescription for speech-generating augmentative and alternative communication device, face-to-face with the patient; first hour</td>
</tr>
<tr>
<td>92608</td>
<td>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)</td>
</tr>
<tr>
<td>92609</td>
<td>Therapeutic services for the use of speech-generating device, including programming and modification</td>
</tr>
<tr>
<td>92618</td>
<td>Evaluation for prescription of non-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)</td>
</tr>
<tr>
<td>L8614</td>
<td>Cochlear device, includes all internal and external components</td>
</tr>
<tr>
<td>L8615</td>
<td>Headset/headpiece for use with cochlear implant device, replacement</td>
</tr>
<tr>
<td>L8616</td>
<td>Microphone for use with cochlear implant device, replacement</td>
</tr>
<tr>
<td>L8617</td>
<td>Transmitting coil for use with cochlear implant device, replacement</td>
</tr>
<tr>
<td>L8618</td>
<td>Transmitter cable for use with cochlear implant device, replacement</td>
</tr>
<tr>
<td>L8619</td>
<td>Cochlear implant, external speech processor and controller, integrated system, replacement</td>
</tr>
<tr>
<td>L8621</td>
<td>Zinc air battery for use with cochlear implant device, replacement, each</td>
</tr>
<tr>
<td>L8622</td>
<td>Alkaline battery for use with cochlear implant device, any size, replacement, each</td>
</tr>
<tr>
<td>L8623</td>
<td>Lithium ion battery for use with cochlear implant device speech processor, other than ear level, replacement, each</td>
</tr>
<tr>
<td>L8624</td>
<td>Lithium ion battery for use with cochlear implant device speech processor, ear level, replacement, each</td>
</tr>
<tr>
<td>L8627</td>
<td>Cochlear implant, external speech processor, component, replacement</td>
</tr>
<tr>
<td>L8628</td>
<td>Cochlear implant, external controller component, replacement</td>
</tr>
<tr>
<td>L8629</td>
<td>Transmitting coil and cable, integrated, for use with cochlear implant device, replacement</td>
</tr>
</tbody>
</table>
ICD-10 Codes

H90.3- H90.8
Sensorineural hearing loss code range (H90.3 is bilateral)

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.
10/1/15  No policy statement changes.
9/1/16  Policy statement changed to indicate that cochlear implantation with a hybrid cochlear implant/hearing aid system is considered medically necessary for patients meeting criteria.
5/1/17  Statements added to indicate that replacement of cochlear implant components only to upgrade to a system with advanced technology or to a next-generation device is considered not medically necessary and that replacement of components when individuals have inadequate response to existing components or components are no longer functional is medically necessary.
9/1/17  No policy statement changes.
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