

Medical Policy



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Populations	Interventions	Comparators	Outcomes
Individuals: • With bilateral sensorineural hearing loss	Interventions of interest are: • Cochlear implant(s)	Comparators of interest are: • Best-aided hearing	Relevant outcomes include: • Symptoms • Functional outcomes • Treatment-related mortality • Treatment-related morbidity
Individuals: • With unilateral sensorineural hearing loss	Interventions of interest are: • Cochlear implant(s)	Comparators of interest are: • Best-aided hearing	Relevant outcomes include: • Symptoms • Functional outcomes • Treatment-related mortality • Treatment-related morbidity

Populations	Interventions	Comparators	Outcomes
Individuals: <ul style="list-style-type: none"> • With high-frequency sensorineural hearing loss with preserved low-frequency hearing 	Interventions of interest are: <ul style="list-style-type: none"> • Hybrid cochlear implant that includes a hearing aid integrated into the external sound processor of the cochlear implant 	Comparators of interest are: <ul style="list-style-type: none"> • Best-aided hearing 	Relevant outcomes include: <ul style="list-style-type: none"> • Symptoms • Functional outcomes • Treatment-related mortality • Treatment-related morbidity

DESCRIPTION

A cochlear implant is a device for treatment of severe-to-profound hearing loss in individuals 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.

OBJECTIVE

The objective of this policy is to determine whether use of a cochlear implant improves the net health outcome for patients with unilateral or bilateral hearing loss.

BACKGROUND

The basic structure of a cochlear implant includes 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 Americas, 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 Product Code: MCM.

Table 1. FDA-Approved Cochlear Implant Systems^a Approved by the FDA

Variables	Manufacturer and Currently Marketed Cochlear Implants		
	Advanced Bionics® HiResolution® Bionic Ear System (HiRes 90K)	Cochlear® Nucleus 22 and 24	Med El® Maestro Combi 40+
PMA	P960058	P840024, P970051	P000025
Predicate devices	Clarion Multi-Strategy or HiFocus CII Bionic Ear (P940022)	Freedom with Contour	
Indications			
Adults ≥18 y	<ul style="list-style-type: none"> • Postlingual onset of severe-to-profound bilateral SNHL (≥70 dB) • Limited benefit from appropriately fitted hearing aids, defined as scoring ≤50% on a test of open-set HINT sentence recognition 	<ul style="list-style-type: none"> • Pre-, peri-, or postlingual onset of bilateral SNHL, usually characterized by: <ul style="list-style-type: none"> ○ Moderate-to-profound HL in low frequencies; and ○ Profound (≥90 dB) HL in mid-to-high speech frequencies • Limited benefit from binaural hearing aids (≤50% sentence recognition in ear to be implanted) 	<ul style="list-style-type: none"> • Severe-to-profound bilateral SNHL (≥70 dB) • ≤40% correct HINT sentences with best-sided listening condition
Children	<ul style="list-style-type: none"> • 12 mo to 17 y of age • Profound bilateral SNHL (>90 dB) • Use of appropriately fitted hearing aids for at least 6 mo in children 2-17 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 sound field (70 dB SPL) 	<ul style="list-style-type: none"> • 25 mo to 17 y 11 mo • Severe-to-profound bilateral SNHL • MLNT scores ≤30% in best-aided condition in children 25 mo to 4 y 11 mo • LNT scores ≤30% in best-aided condition in children 5 y to 17 y and 11 mo 12-24 mo • Profound SNHL bilaterally • Limited benefit from appropriate binaural hearing aids 	<ul style="list-style-type: none"> • 12 mo to 18 y • Profound sensorineural HL (≥90 dB) • In younger children, little or no benefit is defined by lack of progress in the development of simple auditory skills with hearing aids over 3 to 6 mo • 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.

^a 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. Cochlear Ltd. voluntarily recalled the Nucleus CI500 range in September 2011 for device malfunction in the CI512 implant.

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 aids. 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, interest in bilateral cochlear implantation has arisen in recent years. 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; ie, 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, ie, 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 may be used. 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.

POLICY

- A. 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 ages 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, 1000, and 2000 Hz (hertz), who have shown limited or no benefit from hearing aids.
- B. 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 model, are considered **not medically necessary**.
- C. Cochlear implantation as a treatment for patients with unilateral hearing loss with or without tinnitus is considered **experimental / investigational**.
- D. 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:
1. Bilateral severe to profound high-frequency sensorineural hearing loss with residual low-frequency hearing sensitivity; **AND**
 2. Receive limited benefit from appropriately fit bilateral hearing aids; **AND**
 3. Have the following hearing thresholds:
 - a) 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**
 - b) Severe to profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz \geq 75 dB hearing level) in the ear to be implanted; **AND**
 - c) Moderately severe to profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz \geq 60 dB hearing level) in the contralateral ear; **AND**
 - d) 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.
- E. 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.

- F. 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**.

Policy Guidelines

1. 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; ie, in those patients with hearing loss of a magnitude where a hearing aid will not produce the required amplification.
2. In certain situations, implantation may be considered before 12 months of age. One scenario is postmeningitis when cochlear ossification may preclude implantation. Another is in cases with a strong family history, because establishing a precise diagnosis is less uncertain.
3. 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 to 90 dB at frequencies of 1, 2, and 3 kHz, and profound hearing loss is defined as a bilateral hearing threshold of 90 dB and above at frequencies of 1, 2, and 3 kHz.
4. 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% or less correct on open-set tests.
5. 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.
6. Contraindications to cochlear implantation may include deafness due to lesions of the eighth cranial (acoustic) nerve, central auditory pathway, or brain stem; active or chronic infections of the external or 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.

RATIONALE

The most recent literature search was performed through January 11, 2019. The following is a summary of the key literature to date.

Evidence reviews assess the clinical evidence to determine whether the use of a technology improves the net health outcome. Broadly defined, health outcomes are length of life, quality of life, and ability to function³/₄including benefits and harms. Every clinical condition has specific outcomes that are important to patients and to managing the course of that condition. Validated outcome measures are necessary to ascertain whether a condition improves or worsens; and whether the magnitude of that change is clinically significant. The net health outcome is a balance of benefits and harms.

To assess whether the evidence is sufficient to draw conclusions about the net health outcome of a technology, 2 domains are examined: the relevance and the quality and credibility. To be relevant, studies must represent one or more intended clinical use of the technology in the intended population and compare an effective and appropriate alternative at a comparable intensity. For some conditions, the alternative will be supportive care or surveillance. The quality and credibility of the evidence depend on study design and conduct, minimizing bias and confounding that can generate incorrect findings. The randomized controlled trial (RCT) is preferred to assess efficacy; however, in some circumstances, nonrandomized studies may be adequate. RCTs are rarely large enough or long enough to capture less common adverse events and long-term effects. Other types of studies can be used for these purposes and to assess generalizability to broader clinical populations and settings of clinical practice.

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

Clinical Context and Test Purpose

The purpose of cochlear implants is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as best-aided hearing, in patients with bilateral sensorineural hearing loss.

Contraindications to cochlear implantation may include deafness due to lesions of the eighth cranial (acoustic) nerve, central auditory pathway, or brainstem; active or chronic infections of the external or 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.

The question addressed in this evidence review is: does the use of a cochlear implant improve the net health outcome for patients with bilateral hearing loss?

The following PICOTS were used to select literature to inform this review.

Patients

The relevant population of interest are individuals with bilateral sensorineural hearing loss.

Interventions

The therapy being considered is the cochlear implant, which has 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.

Comparators

Comparators of interest include best-aided hearing.

Outcomes

The general outcomes of interest are symptoms, functional outcomes, treatment-related mortality, and treatment-related morbidity.

Timing

The existing literature evaluating cochlear implant(s) as a treatment for bilateral sensorineural hearing loss has varying lengths of follow up, ranging from 6 months. While studies described below all reported at least one outcome of interest, longer follow-up was necessary to fully observe outcomes. Therefore, 1-year of follow-up is considered necessary to demonstrate efficacy.

Setting

Patients with bilateral sensorineural hearing loss are actively managed by otolaryngologists, audiologists, and primary care providers in an outpatient clinical setting.

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

- a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
- c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

Studies with duplicative or overlapping populations were excluded.

Cochlear Implantation: 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 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. Bond et al (2009) authored a

technology assessment 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.³ The clinical effectiveness review included 33 articles (1513 deaf children; 1379 adults), 2 of which were RCTs. They defined 62 different outcome measures, and overall evidence was of moderate-to-poor quality. 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."

Gaylor et al (2013) published an updated technology assessment for the Agency for Healthcare Research and Quality.⁴ Sixteen (of 42) studies published through May 2012 evaluated unilateral cochlear implants. Most unilateral implant studies showed statistically significant improvement in mean speech scores, as measured by open-set sentence or multisyllable word tests; meta-analysis of 4 studies revealed significant improvements in cochlear implant relevant quality of life (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.

Cochlear Implantation: Bilateral Stimulation

While the 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.

In a meta-analysis, McRackan et al (2018) determined the impact of cochlear implantation on quality of life (QOL) and determined the correlation. From 14 articles with 679 CI patients who met the inclusion criteria, pooled analyses of all hearing-specific QOL measures revealed a very strong improvement in QOL after cochlear implantation (SMD=51.77).⁵ Subset analysis of CI-specific QOL measures also showed very strong improvement (SMD=51.69). Thirteen articles with 715 patients met the criteria to evaluate associations between QOL and speech recognition. Pooled analyses showed a low positive correlation between hearing-specific QOL and word recognition in quiet ($r=50.213$), sentence recognition in quiet ($r=50.241$), and sentence recognition in noise ($r=50.238$). Subset analysis of CI-specific QOL showed similarly low positive correlations with word recognition in quiet ($r=50.213$), word recognition in noise ($r=50.241$), and sentence recognition in noise ($r=50.255$) between QOL and speech recognition ability. Using hearing-specific and CI-specific measures of QOL, patients report significantly improved QOL after cochlear implantation. This study is limited in that widely used clinical measures of speech recognition are poor predictors of patient-reported QOL with CIs.

In another meta-analysis, McRackan et al (2018) aimed to determine the change in general health-related quality of life (HRQOL) after cochlear implantation and association with speech recognition.⁶ Twenty-two articles met criteria for meta-analysis of HRQOL improvement, but 15 (65%) were excluded due to incomplete statistical reporting. From the 7 articles with 274 CI patients that met inclusion criteria, pooled analyses showed a medium positive effect of cochlear implantation on HRQOL (SMD=0.79). Subset analysis of the HUI-3 measure showed a large effect (SMD=0.84). Nine articles with 550 CI patients met inclusion criteria for meta-analysis of

correlations between non-disease specific PROMs and speech recognition after cochlear implantation (word recognition in quiet [$r=0.35$], sentence recognition in quiet [$r=0.40$], and sentence recognition in noise [$r=0.32$]). Some limitations are, though regularly used, HRQOL measures are not intended to measure nor do they accurately reflect the complex difficulties facing CI patients. Only a medium positive effect of cochlear implantation on HRQOL was observed along with a low correlation between non-disease specific PROMs and speech recognition. The use of such instruments in this population may underestimate the benefit of cochlear implantation.

Crathorne et al (2012) published a systematic review.⁷ 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. The review included 2 RCTs with waiting-list controls, 10 studies with prospective pre/post repeated-measure or cohort designs, 6 cross-sectional studies, and 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 both RCTs. Meta-analyses could not be performed due to heterogeneity among 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.

The Gaylor Agency for Healthcare Research and Quality 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.⁴ 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 across studies.

Since the publication of the systematic reviews described above, additional comparative studies and case series 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%).⁸

The van Zon et al (2016) prospective study focused on tinnitus perception conducted as a part of a multicenter RCT comparing unilateral with bilateral cochlear implantation in patients who had severe bilateral sensorineural hearing loss.⁹ 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 (a validated scale), with a change in score from 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 Tinnitus Questionnaire score (change in score, 20 to 9; $p=0.04$).

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 Bond technology assessment (2009) 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.³ Weak evidence showed greater gain from earlier implantation (before starting school).

In a review, Bond et al (2009) 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.¹⁰ However, reviewers concluded that there was 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.

Buss et al (2018) published the results of a single-center, retrospective review of 109 children and adolescents who received a second, sequential cochlear implant (CI) between 2008 and 2016.¹¹ Inclusion criteria included <20 years at first CI (CI-1), and minimum 12 years follow-up after second CI (CI-2). Subjects were evaluated at baseline using tests for speech intelligibility and performance, auditory performance, and word and sentence recognition in silence and in noise. Patients were divided into two groups according to inter-CI interval: <3 years (Early Group), versus ≥ 3 years (Late Group); and into 2 groups according to initial performance with the first CI: word recognition <85% (Weak Group), versus $\geq 85\%$ (Strong Group). On the Categories of Auditory Performance (CAP) scale, 28.1% of patients showed improvement at 3 months post-CI-2, 47% at 12 months, and 51.9% at 24 months. Progression in CAP score between CI-1 and M3, M12 and M24 post-CI-2 was significant ($P < 0.05$). On the SIR scale, 33.7% of patients showed improvement at 3 months, 45.4% at 12 months, and 52.6% at 24 months ($P < 0.05$). On word recognition, 47.4% of patients showed improvement at 3 months, 50.8% at 12 months, and 55% at 24 months ($P < 0.05$). On sentence recognition in silence, 66.6% of patients showed improvement at 3 months, 61.2% at 12 months, and 60.6% at 24 months ($P < 0.05$). Progression on sentence recognition in noise, on the other hand, was not significant ($P=0.55$). In the Early group, CAP score improved in 44.4% of patients at M3, 72.4% at M12 and 76.1% at M24 ($P < 0.05$). In the Late group, progression was not significant at M3 ($P = 1$) or M12 ($P = 0.06$) but was significant at M24 ($P < 0.05$). In the Early group, SIR score improved in 49.1% of patients at M3, 63.0% at M12 and 72.1% at M24. In the Late group, SIR score improved in 14.3% of patients at M3, 23.3% at M12 and 27.3% at M24. Improvement was significant in both groups at M3, M12 and M24 ($P < 0.05$). The following are some biases and limitations: (1) subjects' ages advance over the study period. Audiometric and speech-therapy tests are age-adapted, and were not necessarily the same at the various assessment time points; tests for older subjects are correspondingly more "difficult", so that speech therapy scores at 1-year post-CI-2 might be better than at 2 years, due to the nature of the respective tests. This biases assessment of individual progression over time. Patients were implanted between 1.2 and 24 years of age. Speech therapy tests at M3, M12 and M24 thus differed between younger and older patients, introducing an inter-individual bias. (2) certain factors were not taken into

account, like socioeconomic level, parental investment in the project, or associated behavioral, cognitive, psychomotor or sensory disorders, although these strongly impact CI results. They are, however, difficult to quantify, being subjective.

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. 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.¹² Stimulation delivered before about 3.5 years of age results in auditory evoked potentials that reach normal values in 3 to 6 months.

Forli et al (2011) conducted a systematic review of 49 studies on cochlear implant effectiveness in children that addressed the impact of age of implantation on outcomes.¹³ 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 with 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 the duration of cochlear implant usage or 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 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 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.¹⁴

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 systematic review of 38 studies, Black et al (2011) sought to identify prognostic factors for cochlear implantation in pediatric patients.¹⁵ A quantitative meta-analysis was not performed due to study heterogeneity.

However, 4 prognostic factors³⁴age at implantation, inner ear malformations, meningitis, and connexin 26 (a genetic cause of hearing loss)³⁴consistently influenced hearing outcomes.

Pakdaman et al (2012) conducted a systematic review of cochlear implants in children with cochleovestibular anomalies.¹⁶ 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.

Auditory Neuropathy Spectrum Disorder

In a systematic review, Fernandes et al (2015) evaluated 18 published studies and 2 dissertations that reported hearing performance outcomes for children with auditory neuropathy spectrum disorder (ANSD) and cochlear implants.¹⁷ 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 who had ANSD and cochlear implants to the speech perception in children who had 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 (FDA) 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.

Vlastarakos et al (2010) conducted a systematic review of studies on bilateral cochlear implantation in a 125 children implanted before age 1.¹⁸ 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 on cochlear implants 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 before 12 months of age, compared with 23 who were implanted after 12 months of age (specific timing implantation was not provided).¹⁹ 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 of the need to examine the effect of multiple factors on language outcomes and the rate of language development.

Similarly, in a study from Italy, 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.²⁰ 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.^{21,}

Cochlear Implantation in Children: Bilateral Stimulation

In a systematic review, Lammers et al (2014) compared the evidence on the effectiveness of bilateral cochlear implantation with that for unilateral implantation among children with sensorineural hearing loss.^{22,} 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 could not 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.

Several publications not included in the Lammers systematic review 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.^{23,24,25,26,} 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.^{27,}

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

Clinical Context and Test Purpose

The purpose of cochlear implant(s) is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as best-aided hearing, in patients with unilateral sensorineural hearing loss.

Contraindications to cochlear implantation may include deafness due to lesions of the eighth cranial (acoustic) nerve, central auditory pathway, or brainstem; active or chronic infections of the external or 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.

The question addressed in this evidence review is: does the use of a cochlear implant improve the net health outcome for patients with unilateral hearing loss?

The following PICOTS were used to select literature to inform this review.

Patients

The relevant population of interest are individuals with unilateral sensorineural hearing loss.

Interventions

The therapy being considered is cochlear implant(s).

Comparators

Comparators of interest include best-aided hearing.

Outcomes

The general outcomes of interest are symptoms, functional outcomes, treatment-related mortality, and treatment-related morbidity.

Timing

The existing literature evaluating cochlear implant(s) as a treatment for unilateral sensorineural hearing loss has varying lengths of follow up, ranging from 3-months to 6-months. While studies described below all reported at least one outcome of interest, longer follow-up was necessary to fully observe outcomes. Therefore, 6-months of follow-up is considered necessary to demonstrate efficacy.

Setting

Patients with unilateral sensorineural hearing loss are actively managed by otolaryngologists, audiologists, and primary care providers in an outpatient clinical setting.

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

- a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
- c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

Studies with duplicative or overlapping populations were excluded.

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

Van Zon et al (2015) published a systematic review of studies evaluating cochlear implantation for single-sided deafness or asymmetric hearing loss.²⁸ Reviewers assessed 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 pooled for meta-analysis due to high between-study heterogeneity, but reviewers concluded that studies generally reported improvements in sound localization, QOL scores, and tinnitus after cochlear implantation, with varying results for speech perception in noise.

Nonrandomized Trials

Baron et al (2018) published the results of an FDA clinical trial that investigated the potential benefit of cochlear implant (CI) for use in adult patients with moderate-to-profound unilateral sensorineural hearing loss and normal to near-normal hearing on the other side.²⁹ The study population was 20 CI recipients with one normal or near-normal ear (NH) and the other met criterion for implantation (CI). All subjects received a MED-EL standard electrode array, with a full insertion based on surgeon report. They were fitted with an OPUS 2 speech processor. This group was compared to 20 normal hearing persons (control group) that were age-matched. Outcome measures included: sound localization on the horizontal plane; word recognition in quiet with the CI alone, and masked sentence recognition when the masker was presented to the front or the side of normal or near-normal hearing. The follow-up period was 12-months. While the majority of CI recipients had at least one threshold ≤ 80 dB prior to implantation, only three subjects had these thresholds after surgery. For CI recipients, scores on consonant-nucleus-consonant (CNC) words in quiet in the impaired ear rose an average of 4% (0-24%) at the postoperative test to a mean of 55% correct (10%-84%) with the CI alone at the 12-month test interval.

Case Series

Several individual studies have reported on longer-term outcomes for cochlear implantation for single-sided deafness since the publication of the van Zon systematic review.

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.³⁰ 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, and the Tinnitus Questionnaire (a validated scale). Patients demonstrated improvements in visual analog scale 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. Tinnitus Questionnaire scores improved from baseline to 3 months after fitting (55 vs 31, $p < 0.05$) and were stable for the remainder of follow-up.

Rahne et al (2016) reported on a retrospective review of 4 children and 17 adults with single-sided deafness treated with cochlear implants and followed for 12 months.³¹ 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, standard deviation, 2.7 dB) to -4.0 dB after 3 months (standard deviation, 1.3 dB; $p < 0.05$), with continued improvements through 6 months.

Cochlear Implant for Tinnitus Relief in Patients with Unilateral Deafness

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), tinnitus improved in most patients (95%).^{32,}

Ramos Macias et al (2015) reported on 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.^{33,} All patients had a severe tinnitus handicap (Tinnitus Handicap Inventory score $\geq 58\%$). Eight (62%) of the 13 patients who completed the 6-month follow-up visit reported a lower tinnitus handicap on the Tinnitus Handicap Inventory score. Perceived loudness/annoyingness of the tinnitus was evaluated with a 10-point visual analog scale. Tinnitus loudness decreased from 8.4 preoperatively to 2.6 at the 6-month follow-up.

Tavora-Vieira et al (2013) reported on 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.^{34,} 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 for Individuals with High-Frequency Sensorineural Hearing Loss with Preserved Low-Frequency Hearing Loss

Clinical Context and Test Purpose

The purpose of a hybrid cochlear implant that includes a hearing aid integrated into the external sound processor of the cochlear implant is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as best-aided hearing, in patients with high-frequency sensorineural hearing loss with preserved low-frequency hearing.

The question addressed in this evidence review is: does the use of a cochlear implant improve the net health outcome for patients with unilateral or bilateral hearing loss?

The following PICOTS were used to select literature to inform this review.

Patients

The relevant population of interest are individuals with high-frequency sensorineural hearing loss with preserved low-frequency hearing.

Interventions

The therapy being considered is a hybrid cochlear implant that includes a hearing aid integrated into the external sound processor of the cochlear implant.

Comparators

Comparators of interest include best-aided hearing.

Outcomes

The general outcomes of interest are symptoms, functional outcomes, treatment-related mortality, and treatment-related morbidity.

Timing

The existing literature evaluating a hybrid cochlear implant that includes a hearing aid integrated into the external sound processor of the cochlear implant as a treatment for high-frequency sensorineural hearing loss with preserved low-frequency hearing has varying lengths of follow up, including 3-, 6-, and 12-months. While studies described below all reported at least one outcome of interest, longer follow-up was necessary to fully observe outcomes. Therefore, 1-year of follow-up is considered necessary to demonstrate efficacy.

Setting

Patients with high-frequency sensorineural hearing loss with preserved low-frequency hearing are actively managed by otolaryngologists, audiologists, and primary care providers in an outpatient clinical setting.

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

- a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
- c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

Studies with duplicative or overlapping populations were excluded.

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 mitigate the damage to the cochlea and preserve residual hearing.

In March 2014, the FDA approved the Nucleus Hybrid L24 Cochlear Implant System for use through the premarket approval process. According to the FDA's summary of safety and effectiveness data, approval was based on 2 clinical studies conducted outside of the United States and a pivotal study of the Hybrid L24 device conducted under investigational device exemption.¹

The pivotal trial was a prospective, multicenter, single-arm, nonrandomized, nonblinded, repeated measures clinical study among 50 subjects at 10 U.S. sites. Results were reported in FDA documentation and peer-reviewed form by Roland et al (2016).³⁵ 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. The 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 consonant-nucleus-consonant, 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 other subjects were explanted after 12 months. For the 2 primary effectiveness end points (consonant-nucleus-consonant word recognition score, AzBio sentence-in-noise score), there were significant within-subject improvements from baseline to 6-month follow-up. Mean improvement in consonant-nucleus-consonant 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 on results of a prospective multicenter European study evaluating the Nucleus Hybrid L24 system.³⁶ 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.

Santa Maria et al (2014) 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.³⁷ Reviewers included 24 studies, but only two 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.³⁸ 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.

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 hybrid cochlear implantation if there is a loss of residual hearing.

SUMMARY OF EVIDENCE

For individuals who have bilateral sensorineural hearing loss who receive the cochlear implant(s), the evidence includes 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 randomized controlled trials 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 the 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 a 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 of the cochlear implant, the evidence includes prospective and retrospective studies using single-arm, within-subject 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 hybrid cochlear implantation if there

is a loss of residual hearing. The evidence is insufficient to determine the effects of the technology on health outcomes.

CLINICAL INPUT RECEIVED THROUGH 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 of those providing input supported the 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.

PRACTICE GUIDELINES AND POSITION STATEMENTS

American Academy of Otolaryngology – Head and Neck Surgery

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."³⁹

Agency for Health Care Research and Quality

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.⁴⁰ The assessment conclusions are noted within the body of this policy.

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.⁴¹ This guidance was originally based on Bond's (2009) technology assessment,³ and no changes to guidance were made following an updated review of the evidence in 2011.

The 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, as defined in 1.5.
- 1.2 Simultaneous bilateral cochlear implantation is recommended as an option for the following groups of people with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids.
 - a. Children
 - b. Adults 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 [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:
 - a. for adults, a score of 50% or greater on Bamford-Kowal-Bench (BKB) sentence testing at a sound intensity of 70 dB SPL
 - b. for children speech, language and listening skills appropriate to age, developmental stage, and cognitive ability.
- 1.4 Cochlear implantation should be considered for children and adults only after an assessment by a multidisciplinary team. As part of the assessment, children and adults should also have had a valid trial of an acoustic hearing aid for at least 3 months (unless contraindicated or inappropriate)."
- 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)."

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 conclusions²:

- 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.

ONGOING AND UNPUBLISHED CLINICAL TRIALS

Some currently unpublished trials that might influence this policy are listed in Table 2.

Table 2. Summary of Key Trials

NCT No.	Trial Name	Planned Enrollment	Completion Date
Ongoing			
NCT02204618	Cochlear Implantation in Single Sided Deafness and Asymmetrical Hearing Loss: a Cost/Utility Study	160	Aug 2018
NCT02941627 ^a	The Neuro Zti Cochlear Implant System Efficacy and Safety in Adults	55	Jul 2018
NCT03007472 ^a	Clinical Evaluation of the Cochlear Nucleus(R) CI532 Cochlear Implant in Adults	100	Jul 2019
NCT02075229	A Proposal to Evaluate Revised Indications for Cochlear Implant Candidacy for the Adult CMS Population	90	Jun 2019
NCT02203305 ^a	Cochlear Implantation in Cases of Single-Sided Deafness	50	Dec 2018

NCT: national clinical trial.

^a Industry-sponsored or partially sponsored.

CODING

The following codes for treatment and procedures applicable to this policy are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

CPT/HCPCS

69930	Cochlear device implantation, with or without mastoidectomy
92601	Diagnostic analysis of cochlear implant, patient younger than 7 years of age; with programming
92602	Diagnostic analysis of cochlear implant, patient younger than 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
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 or auditory osseointegrated 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 or auditory osseointegrated device speech processor, ear level, replacement, each
- L8625 External recharging system for battery for use with cochlear implant or auditory osseointegrated device, replacement only, 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

ICD-10 Diagnoses

- H90.3 Sensorineural hearing loss, bilateral
- H90.41 Sensorineural hearing loss, unilateral, right ear, with unrestricted hearing on the contralateral side
- H90.42 Sensorineural hearing loss, unilateral, left ear, with unrestricted hearing on the contralateral side

REVISIONS

03-21-2006	In Policy section added, "Bilateral cochlear implantation is considered experimental/investigational" per Medical Director interim guide.
Effective 10-31-2006 Posted 03-01-2007	In Policy section deleted "A FDA-approved cochlear implant and associated rehabilitation may be considered medically necessary in patients with bilateral deafness one year and older with severe to profound pre-or postlingual hearing loss, defined as a hearing threshold of 70 decibels (dB) or worse (adult) and 90dB (child), and have shown no response from hearing aids. Children 18 and under will be reviewed by a consultant" per Medical Director.
	In Policy section added, "Unilateral cochlear implantation is considered medically necessary in patients one year and older with bilateral severe to profound pre-or postlingual hearing loss, defined as a hearing threshold of 70 decibels (dB) or worse (adult) and 90dB (child), and have shown no response from hearing aids" per Medical Director.
	In Policy section added "Bilateral cochlear implantation in children one through 18 years of age with bilateral severe to profound pre-or postlingual hearing loss, defined as a hearing threshold of 90dB (child), who have shown no response from hearing aids will be reviewed by a consultant for medical necessity" per Medical Director.
	In Policy section added, "in adults over 18 years of age" to Bilateral cochlear implantation is considered experimental/investigational, per Medical Director.
	In References Government Agency; Medical Society; and Other Authoritative Publications section, added #3 and #4 per Medical Director.
01-01-2010	In coding section: Updated Wording for: L8614 Added HCPCS Codes: L8327, L8628, L8329

03-19-2010	<p>In Policy section: Replaced prior policy section stating, "Unilateral cochlear implantation is considered medically necessary in patients one year and older with bilateral severe to profound pre-or postlingual hearing loss, defined as a hearing threshold of 70 decibels (dB) or worse (adult) and 90dB (child), and have shown no response from hearing aids. Bilateral cochlear implantation in children one through 18 years of age with bilateral severe to profound pre-or postlingual hearing loss, defined as a hearing threshold of 90dB (child), who have shown no response from hearing aids will be reviewed by a consultant for medical necessity. Bilateral cochlear implantation is considered experimental/investigational in adults over 18 years of age." With policy language currently shown.</p> <p>In Coding section: Added CPT Codes: 92601, 92602, 92603, 92604 Added HCPCS Codes: L8615, L8616, L8617, L8618, L8619, L8621, L8622, L8623, L8624, L8627, L8628, L8629, Added Diagnosis Codes: 389.13, 389.15, 389.16, 389.17</p>
10-04-2013	<p>Description section updated.</p> <p>In Policy section:</p> <ul style="list-style-type: none"> ▪ Revised the following language: "Unilateral or Bilateral cochlear implantation, for adult and children, (simultaneous or sequential) will be reviewed for medical necessity. Documentation for the second implanted ear should parallel the candidacy for unilateral implantation and provide information regarding the anticipated benefits from the second implant. <u>Adults: (18 years of age or older)</u> <ul style="list-style-type: none"> • Post-lingual onset of profound sensorineural hearing loss bilaterally (≥ 90 dB HL); or • Pre- or Post-lingual onset of severe to profound hearing loss bilaterally (≥ 70 dB HL) and; limited or no benefit from appropriately fitted hearing aids. Limited benefit is defined as aided scoring $\leq 50\%$ in the ear to be implanted with $\leq 60\%$ in the better ear on an open set sentence test (HINT). • The minimum duration of hearing aid use is waived if x-rays or clinical information indicate ossification of the cochlea. <u>Children: (1 year to 17 years of age)</u> <ul style="list-style-type: none"> • Pre-or Post-lingual onset of profound hearing loss bilaterally (≥ 90dB HL); and • Limited or not benefit from appropriately fitted hearing aids. For children 12-23 months of age a 3 to 6 month trial of hearing aids is required. Limited use as defined on age appropriate testing. • For children ≤ 4 years of age, lack the benefit is defined as failure to reach developmentally appropriate auditory milestones. • The minimum duration of hearing aid use is waived if x-rays or clinical information indicate ossification of the cochlea." <p>Rationale section added.</p> <p>In Coding section: <ul style="list-style-type: none"> ▪ Added ICD-10 Diagnosis (<i>Effective October 1, 2014</i>) </p> <p>Reference section updated.</p>
09-18-2014	<p>Description section updated</p> <p>In Policy section:</p> <ul style="list-style-type: none"> ▪ Added Item D "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 experimental / investigational (see Policy Guidelines section)."

	<p>In Policy Guidelines</p> <ul style="list-style-type: none"> ▪ Added the following items: <p>"1. 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; ie, in those patients with hearing loss of a magnitude where a hearing aid will not produce the required amplification.</p> <p>2. In certain situations, implantation may be considered before 12 months of age. One scenario is postmeningitis when cochlear ossification may preclude implantation. Another is in cases with a strong family history, because establishing a precise diagnosis is less uncertain.</p> <p>3. 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 to 90 dB at frequencies of 1, 2, and 3 kHz, and profound hearing loss is defined as a bilateral hearing threshold of 90 dB and above at frequencies of 1, 2, and 3 kHz.</p> <p>4. 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% or less correct on open-set tests.</p> <p>6. Contraindications to cochlear implantation may include deafness due to lesions of the eighth cranial (acoustic) nerve, central auditory pathway or brain stem, active or chronic infections of the external or 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.</p> <p>7. 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.</p> <p>8. Hybrid cochlear implant devices that include a hearing aid integrated into the external sound processor of the cochlear implant are considered investigational.</p> <ul style="list-style-type: none"> ▪ Revised Item 5 from, "A post-cochlear implant rehabilitation program is medically necessary for adults and children to achieve benefit from the cochlear implant. The rehabilitation program usually consists of 6 to 12 sessions that last approximately 2.5 hours each. The rehabilitation program is to develop skills in understanding running speech, recognition of consonants and vowels, and tests of speech perception ability. Variability in the number of rehabilitation sessions needed for each patient exists. Rehabilitation should continue until there is a plateau in the patient's language ability or a target endpoint has been reached. There may be need for reprogramming of the device as time goes on due to cochlea-implant interface changes." to "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. Variability in the number of rehabilitation sessions needed for each patient exists."
	Rationale section updated
	Updated References
07-08-2015	Updated Description section.
	Updated Rationale section.
	Updated References section.
10-27-2015	Updated Rationale section.

08-17-2016	<p>Updated Description section.</p> <p>In Policy section:</p> <ul style="list-style-type: none"> ▪ In Item D, added "(eg, ", ")", "may be", and "medically necessary for patients ages 18 years and older who meet all of the following criteria:", and removed ", including but not limited to", "is", and "experimental / investigational (see Policy Guidelines section)." to read, "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:" ▪ Added Item D 1, "Bilateral severe to profound high-frequency sensorineural hearing loss with residual low-frequency hearing sensitivity; AND" ▪ Added Item D 2, "Receive limited benefit from appropriately fit bilateral hearing aids; AND" ▪ Added Item D 3, "Have the following hearing thresholds: <ul style="list-style-type: none"> a) 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 b) Severe to profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz \geq75 dB hearing level) in the ear to be implanted; AND c) Moderately severe to profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz \geq60 dB hearing level) in the contralateral ear; AND d) 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." ▪ In Policy Guidelines, removed Items 7 and 8. <p>Updated Rationale section.</p> <p>Updated References section.</p>
03-15-2017	<p>Updated Description section.</p> <p>In Policy section:</p> <ul style="list-style-type: none"> ▪ Added new Item E, "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." ▪ Added new Item F, "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." <p>Updated Rationale section.</p> <p>Updated References section.</p>
01-01-2018	<p>In Coding section:</p> <ul style="list-style-type: none"> ▪ Added HCPCS codes: L8625, L8694. ▪ Revised nomenclature to HCPCS codes: L8618, L8624. ▪ Removed ICD-9 codes.
03-28-2018	<p>Updated Description section.</p> <p>In Policy section:</p> <ul style="list-style-type: none"> ▪ In Item A, removed "and" and added "who" to read, "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 ages 12 months and older

	<p>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, 1000, and 2000 Hz (hertz), who have shown limited or no benefit from hearing aids."</p> <ul style="list-style-type: none"> ▪ In Item B, removed "(BTE)" to read, "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 model, are considered not medically necessary."
	Updated Rationale section.
	In Coding section: <ul style="list-style-type: none"> ▪ Removed HCPCS code: L8694.
	Updated References section.
03-27-2019	Updated Description section.
	Updated Rationale section.
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