Medical Policy

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***Current Policy Effective Date: 11/1/24**
(See policy history boxes for previous effective dates)

Title: Cochlear Implant

Description/Background

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.

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 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 United States and are manufactured by Cochlear Americas, Advanced Bionics, and the MED-EL Corp. 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 9 months. The labeled indications from FDA for currently marketed implant devices are summarized in Table 1. FDA Product Code: MCM.

Table 1. Cochlear Implant Systems Approved by the Food and Drug Administration

Variables	Manufacturer and Currently Marketed Cochlear Implants			
Device	Advanced Bionics® HiResolution® Bionic Ear System (HiRes 90K)	Cochlear® Nucleus 22 and 24	Med El® Maestro Combi 40+	Neuro Cochlear Implant System (Oticon Medical)
PMA	P960058	P840024, P970051	P000025	P200021
Predicate devices	Clarion Multi-Strategy or HiFocus CII Bionic Ear (P940022)	Freedom with Contour		
Indications				
Adults ≥ 18 y	• Post lingual 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	• Pre-, peri-, or post lingual onset of bilateral SNHL, usually characterized by: ○ Moderate-to-profound HL in low frequencies; and ○ Profound (≥90 dB) HL in mid-to-high speech frequencies • Severe to profound unilateral SNHL (SSD or AHL) ○ PTA at 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz of > 80 dB HL ○ Normal or near normal hearing in the contralateral ear defined as PTA at 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz of ≤ 30 dB HL ○ Limited benefit from an appropriately fitted unilateral hearing device	• Severe-to-profound bilateral SNHL (≥70 dB) • ≤40% correct HINT sentences with best-sided listening condition • SSD (≥90 dB) or AHL (Δ15 dB PTA) ○ Limited benefit from unilateral amplification, defined by test scores of 5% or less on monosyllabic CNC words in quiet when tested in the ear to be implanted alone ○ Patients must have at least 1 month experience wearing a CROS hearing aid or other relevant device and not show any subjective benefit	• Severe-to-profound bilateral SNHL (≥70 dB at500, 1000, and 2000 Hz) • Limited benefit from appropriately fit hearing aids, defined as scoring ≤50%correct HINT sentences in quiet or noise with best-sided listening condition
Children	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	25 mo to 17 y 11 mo of age • 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	12 mo to 18 y of age • 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%	Not applicable

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 (e.g., 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)

children 5 y to 17 y and 11 mo

9 to 24 mo of age

- Profound SNHL bilaterally
- Limited benefit from appropriate binaural hearing aids**

9-12 mo (Nucleus 24 Cochlear only)

- Profound SNH bilaterally
- Limited benefit from appropriate binaural hearing aids**

** In younger children, limited benefit is defined as lack of progress in the development of simple auditory skills in conjunction with appropriate amplification and participation in intensive aural habilitation over a three to six-month period.

5 y to 18 y of age

- Severe to profound unilateral SNHL (SSD or AHL)
  - PTA at 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz of > 80 dB HL
  - Normal or near normal hearing in the contralateral ear defined as PTA at 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz of ≤ 30 dB HL
- Limited benefit from an appropriately fitted unilateral hearing device

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

5 y to 18 y of age

- SSD (≥90 dB) or AHL (Δ15 dB PTA)
  - Insufficient functional access to sound in the ear to be implanted must be determined by aided speech perception test scores of 5% or less on developmentally appropriate monosyllabic word lists when tested in the ear to be implanted
  - Patients must have at least 1 month experience wearing a CROS hearing aid or other relevant device and not show any subjective benefit

AHL: asymmetric hearing loss; CNC: consonant-nucleus-consonant; CROS: contralateral routing of signal; 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; PMA: premarket approval; PTA: pure tone average; SNHL: sensorineural hearing loss; SPL: sound pressure level. SSD: single-sided deafness.

In 2014, the Nucleus® Hybrid™ L24 Cochlear Implant System (Cochlear Americas, Centennial, CO) was approved by FDA 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 ages 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".
- "The CNC [Consonant-Nucleus-Consonant] word recognition score between 10% and 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".

In 2022, the Nucleus® Hybrid™ L24 Cochlear Implant System received expanded approval for single-sided deafness or unilateral hearing loss in adults and children age 5 or older (P970051/S205).

Other hybrid hearing devices have been developed. The Med-El® EAS System received expanded premarket approval by the FDA in 2016 (PMA P000025/S084). FDA product code: PGQ.

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 occurring in noisy environments and localization of sounds. Improvements in speech intelligibility with bilateral cochlear implants may occur with binaural summation (i.e., signal processing of sound input from two sides may provide a better representation of sound and allow the individual to separate 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 receive it 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 a single processor may be used. However, no single processor for bilateral cochlear implantation has been approved by FDA for use in the United States. 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.

In July 2019, the FDA expanded indications for the MED-EL Cochlear Implant System to include single sided deafness and asymmetric hearing loss. The indications for use are as follows:

The MED-EL Cochlear Implant System is indicated for evoking auditory sensations via electrical stimulation of the auditory pathways for individuals ages 5 years and above with single-sided deafness (SSD) or asymmetric hearing loss (AHL), where:

- SSD is defined as profound sensorineural hearing loss in one ear and normal hearing or mild sensorineural hearing loss in the other ear.
- AHL is defined as a profound sensorineural hearing loss in one ear and mild to moderately severe sensorineural hearing loss in the other ear, with a difference of at least 15 dB in pure tone averages (PTAs) between ears.
- Profound hearing loss is defined as having a PTA of 90 dB HL or greater at 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz. Normal hearing is defined as having a PTA of up to 15 dB HL at 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz. Mild hearing loss is defined as having a PTA of up to 30 dB HL at 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz. Mild to moderately severe hearing loss is defined as having a PTA ranging from 31 to up to 55 dB HL at 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz.

Medical Policy Statement

The safety and effectiveness of United States Food and Drug Administration (FDA) approved bilateral and unilateral cochlear implants and associated hybrid cochlear implant devices have been established. The implants may be considered useful therapeutic options when indicated.

Inclusionary and Exclusionary Guidelines

Inclusions

Unilateral or Bilateral cochlear implantation with an FDA approved cochlear implant is considered an established, safe, and effective therapy for individuals who are 9 months of age or older and who meet the following criteria:

- Unilateral or bilateral moderate to profound pre- or post-lingual sensorineural hearing loss **OR**
- Limited or no benefit from hearing aid(s), defined as an aided monosyllabic word score of less than or equal to 50% correct in the ear to be implanted

Replacement of internal and/or external components in a small subset of members may be considered established when ALL of the following are met:

- There is an inadequate response to existing components 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 implant with a hybrid device that includes the hearing aid integrated into the external sound processor of the cochlear implant (e.g., the Nucleus® Hybrid L24 Cochlear Implant System) may be considered established for patients 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.

In certain situations, implantation **consideration may be given before 9 months** of age. One scenario post meningitis when cochlear ossification may preclude implantation. Another is in cases with a strong family history, because establishing a precise diagnosis is less uncertain. However, **these are NOT the only examples** where consideration may be given.

Cochlear implantation outside these guidelines may also be considered medically necessary if the patient is diagnosed with auditory neuropathy spectrum disorder with limited or no benefit from hearing aid(s).

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.

Exclusions

- 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.
- 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.
- Non-FDA approved devices or indications.

CPT/HCPCS Level II Codes *(Note: The inclusion of a code in this list is not a guarantee of coverage. Please refer to the medical policy statement to determine the status of a given procedure.)*

Established codes:

69930	92601	92602	92603	92604	92605
92606	92607	92608	92609	92618	92622
92623	L7510	L8614	L8615	L8616	L8617
L8618	L8619	L8621	L8622	L8623	L8624
L8625	L8627	L8628	L8629		

Other codes (investigational, not medically necessary, etc.):

N/A

Note: The above code(s) may not be covered by all contracts or certificates. Please consult customer or provider inquiry resources at BCBSM or BCN to verify coverage.

Rationale

COCHLEAR IMPLANTATION FOR BILATERAL HEARING LOSS

Clinical Context and Therapy 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 individuals 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 following PICO was used to select literature to inform this review.

Populations

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.
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.

Study Selection Criteria
Methodologically credible studies were selected using the following principles:

- To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
- 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 conclusions:(1)

> "Cochlear implantation improves communication ability in most adults with severe to profound deafness and frequently leads to positive psychological and social benefits as well."
>
> "Pre-lingually 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 post-implant benefit."

The effectiveness of cochlear implants has been evaluated in several systematic reviews and technology assessments, both from the U.S. and abroad. Bond et al (2009) 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 to a single cochlear implant (unilateral or unilateral plus hearing aids) for severely to profoundly deaf children and adults.(2) The clinical effectiveness review included 33 papers, 2 of which were randomized controlled trials (RCTs) (1,513 deaf children; 1,379 adults). 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 pre-lingually deaf children."

Gaylor et al (2013) published an updated technology assessment for the Agency for Healthcare Research and Quality.(3) 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 multi-syllable 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]: 1.15-2.27). However, these studies varied in design and there was considerable heterogeneity observed across studies.

Cochlear Implantation: 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 (CI) 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).(4) 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.(5) 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.(6) 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 two RCTs with waiting-list controls, 10 studies with prospective pre-/post repeated measure or cohort designs, six were cross-sectional studies, and an economic evaluation. All studies compared bilateral with unilateral implantation, and two 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 design. However, all studies reported that bilateral cochlear implants improved hearing and speech perception. One RCT found a significant binaural benefit over the first ear alone for speech and noise from the front (12.6%, p<.001) and when noise was ipsilateral to the first ear (21%, p<.001), another RCT found a significant benefit for spatial hearing at three months post-implantation compared with pre-implantation (mean difference, 1.46, p<.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.(3) The risk of bias varied from medium to high across studies. Based on results from at least two studies, QOL outcomes varied across tests after bilateral implantation; meta-analysis was not performed because of heterogeneity in design between the 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 post-lingual hearing loss, of whom 50 were treated with cochlear implants and 63 with hearing aids, cochlear implant recipients' depression scores improved from pre-implantation to 12 months post-treatment (Geriatric Depression Scale score improvement, 31%; 95% CI, 10% to 47%).(7)

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.(8) This analysis included 38 adults enrolled from 2010 to 2012 and randomized to simultaneous bilateral or unilateral cochlear implants. At one year, post implantation, 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=.03) and from 22 to 12 (p=.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=.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 one cochlear implant with non-technologic support, or an acoustic hearing aid reported gains on all outcome measures.(2) 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.(9) However, reviewers 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.

Barron 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.(10) 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 ≥ 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 ($p < .05$). On the SIR scale, 33.7% of patients showed improvement at 3 months, 45.4% at 12 months, and 52.6% at 24 months ($p<.05$). On word recognition, 47.4% of patients showed improvement at 3 months, 50.8% at 12 months, and 55% at 24 months ($p<.05$). 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 ($p<.05$). Progression on sentence recognition in noise, on the other hand, was not significant ($p=.55$). In the Early group, CAP score improved in 44.4% of patients at M3, 72.4% at M12 and 76.1% at M24 ($p<.05$). In the Late group, progression was not significant at M3 ($p=1$) or M12 ($p=.06$) but was significant at M24 ($p<.05$). 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 ($p<.05$). 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 one-year post-CI-2 might be better than at two 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.

In March 2020, the U.S. Food and Drug Administration (FDA) approved to expand the indication for the Nucleus 24 Cochlear Implant System to include children aged 9 to 24 months of age who have bilateral profound sensorineural deafness and have demonstrated limited benefit from appropriate trials of binaural hearing aids.(11) Children 2 years of age and older may demonstrate severe to profound bilateral hearing loss. The approval was based on a retrospective analysis of prospective data from 5 centers in the United States in children aged between 9 and 12 months who were implanted between 2012 and 2017. Data were collected through March 2019 and included a total of 84 subjects (50% female). Average patient age was 10 months 15 days and 61 subjects received bilateral implants. Post-operative follow-up duration was 6 months. The most common adverse events observed were minor post-operative complications (7.1%) and difficulties with temperature regulation during implantation (7.1%). Twenty-four patients experienced 28 medical/surgical complications and 26 of those complications were resolved without major surgical or medical intervention. Two reimplantation surgeries were reported. The benefits of the device for the age expansion from 12 to 9 months were based on a systematic review of the literature to support premarket approval. A literature search yielded 49 peer-reviewed studies that reported data on safety and/or effectiveness of implantation in children prior to 12 months of age reflecting data on 750 subjects. Significant benefits in terms of improved speech and language development are expected through expansion of the indication in children from 12 to 9 months as reflected by significant improvements in speech intelligibility rating and categorical auditory performance scores.(12) Older implanted children (12-29 months) demonstrated more delayed and atypical language abilities over time.(13)

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.(14) 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.(15) Heterogeneity of studies precluded a meta-analysis. Early implantation was examined in 22 studies, but few studies compared outcomes of implantations performed before one year of age with implantations performed after one year of age. Studies suggest improvements in hearing and communicative outcomes in children receiving implants before one year of age, although it is uncertain whether these improvements are related to 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 24-36 months. Finally, seven 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.(1) This conference offered the following conclusions regarding cochlear implantation in children:

- Cochlear implantation has variable results in children. Benefits are not realized immediately but rather are manifested over time, with some children continuing to show improvement over several years.
- Cochlear implants in children under two-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 post-meningitis hearing loss under the age of two 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 above systematic reviews suggest that cochlear implant removal and re-implantation (due to device malfunction or medical/surgical complications) in children is not associated with worsened hearing outcomes.(16)

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.(17) A quantitative meta-analysis was not performed due to study heterogeneity. However, four 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 (2012) conducted a systematic review of cochlear implants in children with cochleovestibular anomalies.(18) 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 two dissertations that reported hearing performance outcomes for children with auditory neuropathy spectrum disorder (ANSD) and cochlear implants.(19) Studies included four nonrandomized controlled studies considered high quality, five 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.

Bo et al (2023) evaluated 15 studies to assess the effect of cochlear implantation on auditory and speech performance outcomes of children with ANSD.(55) The evidence suggested that children with ANSD who received cochlear implants appeared to achieve similar improvements in their auditory and speaking abilities as children with non-ANSD sensorineural hearing loss. According to pooled data, the categories of auditory performance, speech recognition score, speech intelligence rating score, and open-set speech perception did not significantly differ between the ANSD and sensorineural hearing loss groups.

Cochlear Implantation in Infants Younger Than 12 Months

While currently-available cochlear implants are labeled by the 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.

In 2020, the American Academy of Otolaryngology - Head and Neck Surgery Foundation released an updated position statement on cochlear implants. (55) The Foundation "...considers unilateral and bilateral cochlear implantation as appropriate treatment for adults and children over 9 months of age with moderate to profound hearing loss who have failed a trial with appropriately fit hearing aids."

A number of small studies from outside the U.S. have reported results on cochlear implantation in infants younger than 12 months. 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).(21) 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 two 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.(22) Within the first six months post-implantation, 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.(23)

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.(24) Reviewers identified 21 studies that evaluated bilateral cochlear implantation in children, with no RCTs identified. Due to a limited number of studies, heterogeneity in outcomes and comparison groups and high risk for bias in the studies, 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 one 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 report improved speech outcomes with bilateral implantation, compared with unilateral implantation.(25-28) 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 inter-implant interval.(29)

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 Therapy 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 individuals 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 following PICO was used to select literature to inform this review.

Populations
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.

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 1 outcome of interest, longer follow-up was necessary to fully observe outcomes. Therefore, 6-months of follow-up is considered necessary to demonstrate efficacy.

Study Selection Criteria
Methodologically credible studies were selected using the following principles:
- To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
- 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.

Systemic Reviews
Oh et al (2022) published a systematic review and meta-analysis of 50 studies, including prospective and retrospective observational studies and case series, evaluating cochlear implantation in adults (n=674) with single-sided deafness.(30) Pooled outcomes indicated improved scores in speech perception (SMD, 2.8; 95% CI, 2.16 to 3.43; 7 studies; I^2=73.1%), localization (SMD, -1.13; 95% CI, -1.68 to -0.57; 7 studies; I^2=71.5%). tinnitus (SMD, -1.32; 95% CI, -1.85 to -0.80; 8 studies; I^2=73.1%); and quality of life (SMD, 0.61; 95% CI, 0.45 to 0.91; 10 studies; I^2=0.0%). Study interpretation is limited by small sample sizes and heterogeneity in reported outcomes and follow-up durations.

Benchetrit et al. (2021) published a systematic review and meta-analysis evaluating audiological and patient-reported outcomes in children <18 years with single-sided deafness (SSD).(31) Twelve observational studies evaluating 119 children(mean age [standard deviation], 6.6 [4.0] years) were included. Clinically meaningful improvements in speech perception in noise (39/49 [79.6%]) and in quiet (34/42 [81.0%]) were reported. Sound localization improved significantly following implantation (mean difference [MD], -24.78°; 95% CI, -34.16° to -15.40°; I^2 = 10%). Compared to patients with congenital SSD, patients with acquired SSD and shorter duration of deafness reported greater improvements in speech and hearing quality. Patients with longer duration of deafness were also more likely to be device nonusers (MD, 6.84; 95% CI, 4.02 to9.58).

Randomized Trials
Marx et al (2021) conducted a small open-label, multicenter RCT of cochlear implantation (n=25) versus initial observation and treatment abstention (n=26) in adult patients with single-sided deafness or asymmetric hearing loss following failure of prior treatment with contralateral routing of the signal (CROS) hearing aids or bone-conduction devices.(32) Primary outcomes included HRQOL, auditory-specific quality of life, and tinnitus severity as assessed after 6 months of treatment. Both EQ-5D visual analog scale and auditory-specific quality of life

indices significantly improved in the cochlear implant arm. However, no significant difference in overallEQ-5D descriptive component scores were noted between groups. Mean improvement was most pronounced in subjects with associated severe tinnitus. A clinical rationale for the minimum clinical improvement in quality of life (0.8 SD) was not reported. No significant difference for speech recognition in noise or horizontal localization was noted between groups at 6 months, indicating no significant effect on binaural hearing within this timeframe.

Peters et al (2021) randomized 120 adults with single-sided deafness (median duration, 1.8 years) into 3 treatment groups for the "Cochlear Implantation for siNGLE-sided deafness" (CINGLE) trial: cochlear implant (n=29); first bone-conduction devices, then CROS (n=45); and first CROS, then bone-conduction devices (n=46).(33) Patients with a maximum 30 dB hearing loss in the best ear and a minimum 70 dB hearing loss in the poor ear with duration of single-sided deafness between 3 months and 10 years were eligible for inclusion. After the initial cross-over period, 25 patients were allocated to bone-conduction devices, 34 patients were allocated to CROS, and 26 patients preferred no treatment. Seven patients did not receive their allocated treatment. For the primary outcome, speech perception in noise from the front, a statistically significant improvement was noted for the cochlear implant group at 3 and 6 months compared to baseline. At 3 months follow-up, the cochlear implant group performed significantly better than all other groups. At 6 months, the cochlear implant group performed significantly better than the bone-conduction devices and no treatment groups, but no significant difference was observed between the cochlear implant group and the CROS group. Sound localization improved in the cochlear implant group only. All treatment groups improved on disease-specific quality of life compared to baseline. The study is limited by small sample size, device heterogeneity, loss to follow-up, and lack of allocation concealment. Study follow-up through 5 years is ongoing.

Nonrandomized Trials

Buss 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.(34) 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 to 24%) at the postoperative test to a mean of 55% correct (10% to 84%) with the CI alone at the 12-month test interval.

Dillon et al (2019) published a clinical update reporting on the prevalence of low-frequency hearing preservation with the use of standard long electrode arrays (MED-EL Corporation) in a subset of 25 patients (12 with unilateral hearing loss) from earlier cohorts.(35) Unaided hearing thresholds at 125 Hz were compared between the preoperative and initial activation intervals to assess the change in low-frequency hearing. At activation, a significant elevation in the unaided hearing thresholds at 125 Hz was noted among a sample of 24 patients (p<0.001), with the majority of subjects (n=16) demonstrating no response to stimulus. The remaining 9 participants maintained an unaided low-frequency hearing threshold of ≤ 95 dB, and 5/9 participants met the fitting criterion of ≤ 80 dB for electric-acoustic stimulation (EAS) at initial activation. An additional three participants demonstrated improvement in unaided low-frequency hearing thresholds at latter monitoring intervals. It is uncertain whether identifying patients with preservation of low-frequency hearing can help predict individuals that may benefit from electric-acoustic stimulation versus standard cochlear implants.

Galvin III et al (2019) reported data from on FDA approved study of cochlear implantation in 10 patients with single-sided deafness (SSD).(36) Patients were implanted with the MED-EL Concerto Flex 28 device. Speech perception in quiet and noise, localization, and tinnitus severity were measured prior to implantation at 1-, 3-, and 6-months post-activation. Performance was assessed with both ears (binaural), with the implanted ear alone, and the normal hearing alone. No patient had previous experience with contralateral routing of signal or bone conduction device system. Mean improvement for consonant-nucleus-consonant word recognition versus baseline was 66.8%, 76.0%, and 84.0% at 1-, 3-, and 6-months post-activation, respectively. The normal hearing ear performed significantly better compared to the implanted ear for all outcome measures at all intervals (p<.05). Audiological performance of the implanted ear at 1-, 3-, and 6-months post-activation was significantly better compared to baseline (p<.05), with no significant difference across post-activation intervals (p>.05). The change in root mean square error in localization with binaural listening post-activation reduced by 6.7, 7.6, and 11.5 degrees at 1-, 3-, and 6-months post-activation. Binaural performance was significantly improved compared to the normal hearing ear alone at all post-activation time intervals (p<.05). Tinnitus visual analog scale scores significantly decreased with the implant on at all post-activation time intervals (p<.05). Significant improvements in Speech, Spatial, and Qualities of Hearing Scale questionnaire (SSQ) scores were reported for the Speech (p=.003), Spatial (p<.001), and Quality (p=.034) subtests. Global scores were not reported. Adverse events were reported in 5/10 participants, including facial nerve stimulation, periorbital edema, mild postoperative balance disturbance, postauricular pain, and unresolved taste disturbance. The study is limited by small sample size.

Peter et al (2019) published the results of a Swiss multicenter study assessing cochlear implantation for use in adult patients in post-lingual single-sided deafness, defined as a hearing loss of 70 dB hearing level (HL) in the mean thresholds of 0.5, 1, 2, and 4 kHz in the affected ear, and 25 dB HL or better in the frequencies from 125 to 2 kHz and 35 dB HL or better from 4 to 8 kHz in the normally hearing contralateral ear.(37) A total of 10 patients were evaluated. Two-year post-implantation, 90% of patients used their implant regularly for an average of more than 11 hours per day. Twelve months post-activation, speech from the front and noise at the healthy ear achieved a 2.7 dB improvement (p=.0029). Speech to the implanted ear and noise from the front achieved a 1.5 dB improvement (p=.018). The mean sound localization error of all participants was improved by 10.2 degrees (p=.030) at 12 months post-activation. One participant experienced a loss in low-frequency residual hearing from surgery, resulting in poorer localization performance after surgery with an increased error of 11.3 degrees. Tinnitus severity significantly decreased 12 months post-activation from 41.2 points (SD 26.5) preoperatively to 23.0 points (SD 17.5; p=.004) on the tinnitus Handicap Inventory. Quality of life measures showed a significant improvement on the global subscale of the World Health Organization quality of life questionnaire (p=.007). The SSQ indicated a significant improvement from 4.2 to 6 (p=.004) in speech comprehension and from 3 to 5.3 (p=.009) in spatial hearing. No significant difference was noted in the subscale qualities of hearing (6.2 to 6.9; p=.13). The scores on the 3 subscales were significantly lower than for the normal hearing control group, with an average speech comprehension score of 8.7 (p=.001), an average spatial hearing of 8.6 (p<.001), and an average quality of hearing score of 9.1 (p=.005). Adverse events were not reported.

Poncet-Wallet et al (2019) reported on audiological and tinnitus outcomes of cochlear implantation in adults with SSD and tinnitus.(38) Twenty-six patients with SSD and incapacitating tinnitus (THI score > 58) underwent cochlear implantation. Masking white noise stimulation was delivered for the first month post-implantation, after which standard cochlear implant stimulation was provided. Catastrophic handicaps (grade 5, THI 78 to100) were noted for 31% of participants and severe handicaps (grade IV, THI 58 to 76) were noted for 69% of participants. The first month of white noise stimulation provided a significant improvement in THI scores (72 ± 9 to 55 ± 20; p<.05). No change was observed for the other measures at this time point. After 1 year of standard stimulation, 23 patients (92%) completed the final 13-month visit with 0% of participants reporting catastrophic handicaps, 4% reporting severe handicaps, and 26% reporting moderate handicaps (grade III, THI 38-56), 30% reporting mild handicaps (grade II, THI 18 to 36), and 39% reporting slight or no handicaps (grade I, THI 0 to16) (p<.05). All 23 patients attending the 13-month visit reported improvement of tinnitus on at least two of four tinnitus questionnaires.

Dillon et al (2020) conducted a prospective clinical trial evaluating 20 subjects with asymmetric hearing loss (AHL), defined as a hearing loss of ≥ 70 dB HL in the ear to be implanted and between 35- and 55-dB HL in the contralateral ear.(39) Patients were required to fail initial treatment with traditional or bone-conduction hearing aids. Subjects underwent cochlear implantation with the MED-EL Synchrony Standard electrode array. Significant subjective benefit was reported by patients within 1 month of implantation. At the 12-month interval, spatial hearing localization was significantly improved (p<.001). Masked sentence recognition was found to improve at the 12-month interval in the sound from 90 degrees to the contralateral ear configuration (p<.001), but there was no significant difference in the sound from the front or from 90 degrees to the cochlear implant ear spatial configurations. Subjects demonstrated a significant improvement in CNC word recognition between 1 and 6 months (*p*<.002) and 6 and 12 months (*p*=.10).Findings were compared with previously published data for patients in the unilateral hearing loss cohort of this study.(34) Significant main effects of cohort were found for localization performance and spatial configuration in masked sentence recognition, indicating that the magnitude of benefit for these outcomes was reduced for subjects with asymmetric hearing loss.(39)

In July 2019, the FDA approved to expand the indication for the MED-EL Cochlear Implant System to include individuals aged 5 years and older with SSD or asymmetric hearing loss (AHL).(40) According to the FDA's summary of safety and effectiveness data,(34) approval was based on supporting evidence from a comprehensive literature review and a clinical feasibility study conducted at the University of North Carolina at Chapel Hill under IDE# G140050 in patients treated between 2014 and 2019. In this prospective, non-blinded, repeated measures study, 40 subjects were implanted with the MED-EL CONCERT or SYNCHRONY Cochlear Implant System. Twenty patients each were enrolled into the SSD and AHL groups. All 20 patients completed testing in the SSD group. One patient withdrew from the AHL group, and one patient had not yet completed follow-up at the time of data analysis. Patients were required to have previous experience of at least one month in duration with a conventional hearing aid, bone conduction device, or contralateral routing of signal device. Exclusion criteria included Meniere's disease with intractable vertigo, tinnitus as the primary concern for cochlear implantation, and severe or catastrophic score on the THI. Aided word recognition in the ear to be implanted was required to be 60% or less as measured with a 50-word CNC word list. Speech perception and localization were evaluated at baseline and at 1, 3, 6, 9, and 12 months post-operatively utilizing CNC word recognition and AzBio sentence tests. For patients in the AHL group, sound field testing was completed with a hearing aid in the contralateral ear. Quality of life measures included the SSQ, THI, and Abbreviated Profile of Hearing Aid Benefit (APHAB) scales. Primary effectiveness measures were comparisons of speech perception and localization performance between the bilateral, pre-operative, unaided/best-aided condition and the bilateral, 12-month post-operative cochlear implant + normal hearing or hearing aid condition. Study results are summarized in Table 2. Nine device- or procedure-related adverse events were reported. Most frequently reported adverse events included vertigo/dizziness/imbalance (22.5%) and unrelated infection (7.5%). The data from the is limited by its small sample size in adult subjects only. Effectiveness endpoints were not prespecified.

The FDA decision was further supported by a literature search yielding six publications comprising a total of 58 adults with SSD (n=50 were implanted with MED-EL devices) and a total of 52 adults with AHL (n=37 of which implanted with MED-EL devices). The decision to expand the indication to pediatric patients aged 5 and older was based on a literature search yielding five publications comprising a total of 26 children with SSD (n=50 were implanted with a MED-EL device) and a total of 9 children with AHL. While the overall benefits of cochlear implants in children with SSD and AHL included improved performance in speech perception in quiet and noise, sound localization, and subjective measures of quality of life these results are limited to primarily case series with small sample sizes, heterogeneous methodology and outcome assessment, and high-risk of bias in self-reported measures. The FDA has required MED-EL to conduct a post-marketing study to continue to assess the safety and efficacy of the implant in a new enrollment cohort of adults and children.(41)

Table 2. Feasibility Study Results for MED-EL Cochlear Implant System for SSD and AHL (40)

Outcome		SSD (n=20)			AHL (n=18)	
Speech Perception in Quiet	**Baseline, Unaided**	**12-mo, Unaided**	**12-mo, CI-On**	**Baseline, Unaided**	**12-mo, Unaided**	**12-mo, CI-On**
Implant Ear CNC, Mean (SD) Range	3.5 (6.68) 0 to 22	NA	54.6 (18.15) 10 to 84	6.3 (7.98) 0 to 22	NA	56.2 (18.41) 28 to 86
Contralateral Ear CNC, Mean (SD) Range	99.3 (2.27) 90 to 100	99.8 (0.62) 98 to 100	NA	92.7 (8.68) 78 to 100	92.7 (8.68) 72 to 100	NA
Soundfield, Binaural AzBio, Mean (SD) Range	99.0 (1.56) 95 to 100	NA	99.5 (1.19) 95 to 100	87.4 (13.96) 50 to 99	NA	94.3 (8.38) 72 to 100
		SSD (N=20)			**AHL (N=17)**	
Speech Perception in Noise	**Baseline, Unaided**	**Baseline, Best-Aided (BCHA)**	**12-mo, CI-On**	**Baseline, Unaided**	**Baseline, Best-Aided (BCHA)**	**12-mo, CI-On**
Noise Front AzBio, Mean (SD) Range	37.5 (10.98) 20 to 64	31.5 (16.56) 0 to 59	47.2 (10.72) 29 to 68	22.7 (13.95) 0 to 47	20.5 (12.86) 0 to 47	33.5 (22.10) 3 to 85
Noise at CI AzBio, Mean (SD) Range	83.4 (9.51) 59 to 94	61.25 (27.92) 0 to 98	85.0 (11.04) 60 to 97	44.2 (17.70) 9 to 78	30.5 (18.23) 1 to 70	44.6 (24.74) 5 to 94
Noise at Contralateral AzBio, Mean (SD) Range	16.5 (12.78) 0 to 45	18.3 (13.50) 0 to 59	52.6 (21.43) 8 to 86	6.3 (9.49) 0 to 36	11.3 (16.69) 0 to 66	29.4 (22.59) 1 to 95
		SSD (N=20)			**AHL (N=18)**	
Localization Performance	**Baseline, Unaided**	**Baseline, Best-Aided (BCHA)**	**12-mo, CI-On**	**Baseline, Unaided**	**Baseline, Best-Aided (BCHA)**	**12-mo, CI-On**
Mean RMS Error (SD) Range	66.5 (20.47) 42.9 to 109.1	69.6 (18.71) 45.3 to 106.1	26.7 (6.32) 13.6 to 38.4	76.5 (19.23) 43.8 to 105.3	77.2 (18.89) 45.6 to 106.5	40.1 (10.65) 26.6 to 73.6
Quality of Life	SSQ (Speech)	SSQ (Spatial)	SSQ (Qualities)	APHAB (Global)	APHAB (EC, RV, BN, AV)	THI
SSD (N=20) Baseline: Mean (SD); Range 12-mo: Mean (SD); Range	3.7 (1.34); 0.6 to 7.2 7.1 (0.99); 5.4 to 8.9	2.4 (1.2); 0.5 to 4.5 6.5 (1.86); 2.8 to 8.9	5.6 (2.09); 0.5 to 9.8 7.7 (1.28); 5.6 to 9.8	49.8 (18.65); 20.3 to 86.3 17.9 (8.91); 6.1 to 36.7	EC: 31.6 (21.06); 2.8 to 81.0 8.7 (6.15); 1.0 to 24.8 BN: 70.1 (17.32); 39.3 to 95.0 25.2 (11.95); 10.2 to 56.2 RV: 47.5 (21.96); 18.7 to 87.0	NR

					19.7 (12.43); 2.8 to 41.7 AV: 43.1 (28.64); 1.0 to 93.0 26.7 (24.83); 1.0 to 91.0	
AHL (N=18) Baseline: Mean (SD); Range 12-mo: Mean (SD); Range	3.2 (1.48); 0.4 to 6.0 5.8 (1.50); 3.6 to 8.9	2.6 (1.26); 0.3 to 4.7 6.0 (1.62); 3.1 to 8.5	4.6 (1.77); 0.2 to 8.3 6.8 (1.20); 4.4 to 8.7	54.1 (16.21); 20.0 to 92.3 28.1 (10.49); 11.3 to 54.1	EC: 42.9 (24.67); 10.2 to 91.0 16.6 (13.01); 1.0 to 54.0 BN: 63.5 (16.84); 14.5 to 95.0 39.3 (17.10); 14.5 to 66.3 RV: 56.0 (18.30); 14.2 to 97.0 28.3 (11.96); 12.0 to 54.2 AV: 43.1 (35.04); 1.0 to 99.0 42.4 (29.21); 1.0 to 97.0	NR

AHL: asymmetric hearing loss; APHAB: Abbreviated Profile of Hearing Aid Benefit; AV: Aversiveness subscale; BCHA: bone conduction hearing aid; BN: Background Noise subscale; CI: cochlear implant; CNC: consonant-nucleus-consonant; EC: Ease of Communication subscale; NA: not applicable; NR: not reported; RMS: root mean square; RV: Reverberation subscale; SD: standard deviation; SSD: single-sided deafness; SSQ: Speech, Spatial, and Qualities of Hearing Scale; THI: Tinnitus Handicap Inventory.

In January 2022, the FDA approved to expand the indication for the Nucleus 24 Cochlear Implant System to individuals aged 5 years and older with single-sided deafness or asymmetrical hearing loss.(42) According to the FDA's summary of safety and effectiveness data, approval was based on unpublished data in 42 adults from a feasibility study (n=10) and real-world data from two cochlear implantation centers (n=32). Study interpretation is limited by small sample size in adult subjects only, unclear rationale for the efficacy threshold, and missing data. The FDA has required Cochlear Americas to conduct a postmarketing study to continue to assess the safety and efficacy of the implant in a new enrollment cohort of adults and children.

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%).(44)

Ramos Macias et al (2015) reported on results of a prospective multicenter study with repeated measures related to tinnitus, hearing, and quality of life, among 16 individuals with unilateral hearing loss and severe tinnitus who underwent cochlear implantation.(45) 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. Tinnitus loudness decreased from 8.4 preoperatively to 2.6 at the 6-month follow-up.

Tavora-Vieira et al (2013) reported results of a prospective case series that included 9 post-lingually 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.(46) 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.

Cochlear Implantation in Pediatric Population with Unilateral Deafness

Brown et al (2022) published results from the Childhood Unilateral Hearing Loss (CUHL) prospective, single-arm trial.(43) Twenty children aged 3-12 with moderate to profound sensorineural hearing loss and poor speech perception (word score <30%) in one ear and normal hearing in the contralateral ear were enrolled. CNC word score perception in quiet improved significantly from 1% to 50% ($p<.0001$) at 12 months after activation. Speech perception in noise by BKB-SIN score also significantly improved by 3.6 dB in head shadow ($p<.001$), 1.6 dB in summation ($p=.003$), and 2.5 dB in squelch ($p=.0001$). By 9 months, localization improved by 26°. Significant improvements were also found in SSQ speech ($p=.0012$), qualities of hearing ($p=.0056$), and spatial hearing subscales ($p<.0001$). Improvements in fatigue were not statistically significant. Study limitations include use of a single-arm study design, small sample size, and incomplete comparison to best-aided hearing at baseline, including enrollment of never aided subjects.

Section Summary: Cochlear Implantation for Unilateral Sensorineural Hearing Loss

The available evidence for the use of cochlear implants in improving outcomes for individuals with unilateral hearing loss, with or without tinnitus, is limited by small sample sizes and heterogeneity in evaluation protocols and outcome measurements. A small feasibility study in adults with SSD or AHL demonstrated improvements in sound perception, sound localization, and subjective measures of quality of life compared to baseline conditions. Multiple position statements support the use unilateral cochlear implants (see Supplemental section). Ongoing post-marketing studies in adults and children may further elucidate outcomes. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

HYBRID COCHLEAR IMPLANTATION FOR INDIVIDUALS WITH HIGH-FREQUENCY SENSORINEURAL HEARING LOSS WITH PRESERVED LOW-FREQUENCY HEARING LOSS

Clinical Context and Therapy 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 following PICO was used to select literature to inform this review.

Populations
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.

Study Selection Criteria
Methodologically credible studies were selected using the following principles:

- To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
- 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.

Review of Evidence

Nonrandomized Trials
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 September 2016, the FDA approved the MED-EL Cochlear Implant with Combined Electrical Stimulation and Acoustic Amplification System (EAS) for partially deaf individuals aged 18 years and older who have residual hearing sensitivity in the low frequencies sloping to severe/profound sensorineural hearing loss in the mid- to high-frequencies, and who receive minimal benefit from conventional acoustic amplification.(34) Final outcomes were reported in 2018 by Pillsbury et al.(47) Sixty-seven of 73 subjects (92%) completed outcome measures at 3, 6, and 12-months post activation. A 30 dB or less low-frequency pure-tone average shift was experienced by 79% and 97% were able to use the acoustic unit at 12 months post-activation. In the EAS condition, 94% of subjects performed similarly or demonstrated improvement (85%) compared to preoperative performance on City University of New York sentences in noise at 12 months. Ninety-seven percent of subjects performed similarly or improved (85%) on CNC words in quiet. Improvements in speech perception scores were statistically significant ($p<.001$). The APHAB was administered preoperatively and at 12 months post-activation; 60 subjects completed the APHAB assessment at each time point. The mean score on the APHAB Global Scale improved by 30.2%, demonstrating a significant reduction in perceived disability (p<.001). Thirty-five device related adverse events were reported for 29 of 73 subjects (39.7%). The most frequently observed adverse event was profound/total loss of residual hearing, which occurred in 8 of 73 subjects (11.0%).

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 a pivotal study of the Hybrid L24 device conducted under investigational device exemption.(48)

The pivotal trial was a prospective, multicenter, single-arm, nonrandomized, non-blinded, repeated-measures clinical study among 50 subjects ≥ 18 years of age at 10 U.S. sites. Results were reported in FDA documentation and peer-reviewed form by Roland et al (2016).(49) 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- with post implant within each subject; outcomes were measured at 3, 6, and 12 months postoperatively. The trial tested 2 co-primary 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 post-implantation 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 two other subjects were explanted after 12 months. For the two primary effectiveness end points, (consonant-nucleus-consonant word-recognition score and 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, the mean improvement was 32.0% (95% CI 23.6% to 40.4%) Ninety-six percent of subjects performed equal or better on speech in quiet and 90% performed equal or better in noise. For safety outcomes, 65 adverse events were reported, most commonly profound/total loss of hearing (occurring in 44% of subjects) with at least one adverse event occurring in 34 subjects (68%).

Five-year outcomes for the pivotal trial were reported by Roland et al. (2018).(50) Thirty-two of 50 subjects (64%) enrolled in the post-approval study. Out of the 18 subjects who did not participate, 6 had been explanted and re-implanted with a long electrode array, two discontinued for unrelated medical reasons, two withdrew for other reasons, four declined to continue follow-up evaluations, and four chose not to participate in the post-approval study. At five years post-activation, 94% of subjects had measurable hearing and 72% continued to use electric-acoustic stimulation with functional hearing in the implanted ear, and 6% had a total loss. Changes from pre-operate hearing to 6 months were statistically significant (p<.001) but changes 6 months through five years post-activation were not statistically different (p>.05). Acoustic component amplification was utilized by 84% and 81% of patients at 12- and 3-years post-activation, respectively. Mean CNC word recognition in quiet scores were significantly improved over the preoperative condition at each post-activation interval (p<.001). However, mean scores did not significantly differ after 12 months post-activation. At 5 years post-activation, 94% performed the same or better in unilateral CNC word scores, whereas 6% demonstrated a decline in performance. For bilateral CNC word scores, 97% performed the same or better, whereas 1 subject showed a decline in performance. The Speech, Spatial, and Qualities of Hearing Questionnaire (SSQ) was implemented to measure subjective implant satisfaction and benefit. Scores significantly improved and remained stable through all post-activation intervals ($p<.001$).

Lenarz et al (2013) reported on results of a prospective multicenter European study evaluating the Nucleus Hybrid™ L24 system.(51) 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 hybrid devices, cochlear implantation with surgical techniques designed to preserve hearing, and the use of postoperative systemic steroids.(52) 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.(53) 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.

Gantz et al (2016) published outcomes from a multicenter, longitudinal study evaluating outcomes with the Nucleaus Hybrid S8 featuring a shorter cochlear array.(54) Eighty-seven subjects received an implant. At 12 months post activation, five subjects had total hearing loss, whereas functional hearing was maintained by 80%. CNC word scores demonstrated that 82.5% of subjects had experienced a significant improvement in the hybrid condition.

Improvement in speech understanding in noise were demonstrated in 55% of subjects. Fourteen patients requested implant explantation due to various reasons for dissatisfaction with the device. These patients were re-implanted with a standard-length Nucleus Freedom cochlear implant. CNC scores prior to loss of residual hearing were missing for six subjects. CNC scores following re-implantation were missing for two additional subjects. Similar or better CNC scores following re-implantation were observed in 5/6 remaining subjects.

Section Summary: Hybrid Cochlear Implantation
Prospective and retrospective studies using a single-arm, within-subjects comparison pre- and post-intervention 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 suggest 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 loss of residual hearing. Studies reporting on long-term outcomes and results of re-implantation are lacking.

S**UMMARY 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 morbidity and mortality. 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 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 small open-label RCTs, a feasibility study, prospective and retrospective studies reporting within-subjects comparisons and systematic reviews of these studies. Relevant outcomes include symptoms, functional outcomes, and treatment-related morbidity and mortality. Given the natural history of hearing loss, pre- and post-implantation comparisons may be appropriate for objectively measured outcomes. Although 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 and heterogeneity in evaluation protocols and outcome measurements, devices have been approved by the U.S. Food and Drug Administration for use in single sided deafness. A small feasibility study in adults with single-sided deafness or asymmetric hearing loss demonstrated improvements in sound perception, sound localization, and subjective measures of quality of life compared to baseline conditions. Multiple position statements support the use unilateral cochlear implants (see Supplemental section). Ongoing post-marketing studies in adults and children may further elucidate outcomes. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

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 of the cochlear implant, the evidence includes prospective and retrospective studies using a single-arm, within-subjects comparison pre- and post-intervention and systematic reviews. Relevant outcomes are symptoms, functional outcomes, and treatment-related mortality and morbidity. The available evidence suggests 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 suggest 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 loss of residual hearing. Studies reporting on long-term outcomes and results of re-implantation are lacking. The evidence is insufficient to determine the effects of the technology on health outcomes.

Supplemental Information

CLINICAL INPUT RECEIVED FROM PHYSICIAN SPECIALTY SOCIETIES AND ACADEMIC MEDICAL CENTERS

2016 Input
In response to requests, Blue Cross Blue Shield Association received input 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, Blue Cross Blue Shield Association received input 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 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 Foundation
In 2020, the American Academy of Otolaryngology - Head and Neck Surgery Foundation released an updated position statement on cochlear implants.
(56) The Foundation “...considers unilateral and bilateral cochlear implantation as appropriate treatment for adults and children over 9 months of age with moderate to profound hearing loss who have failed a trial with appropriately fit hearing aids."

Agency for Health Care Research and Quality
In 2011, a technology assessment for the AHRQ assessed the effectiveness of cochlear implants in adults.(57) The assessment conclusions are noted within the body of this evidence review.

National Institute for Health and Care Excellence
In 2019, the National Institute for Health and Clinical Excellence (NICE) released a technology guidance on cochlear implants for children and adults with severe-to-profound deafness.(57)

The guidance included the following updated 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.
1.4 For the purposes of this guidance, severe to profound deafness is defined as hearing only sounds that are louder than 80 dB HL [hearing level] at 2 or more frequencies bilaterally (500 Hz, 1 kHz, 2 kHz, 3 kHz, 4 kHz) without acoustic hearing aids. Adequate benefit from acoustic hearing aids is defined for this guidance as:
   a. for adults, a phoneme score of 50% or greater on Authur Boothroyd word test presented at 70 dBa.
   b. for children speech, language and listening skills appropriate to age, developmental stage, and cognitive ability.
1.5 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)."
1.6 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)."

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:(1)

> "Cochlear implantation has a profound impact on hearing and speech reception in post-lingually deafened adults."
>
> "Pre-lingually deafened adults generally show little improvement in speech perception scores after cochlear implantation, but many of these individuals derive satisfaction from

hearing environmental sounds and continue to use their implants." However, improvements in other basic benefits, such as sound awareness, may meet safety needs.

"…training and educational intervention are fundamental for optimal post-implant benefit."

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

"Cochlear implantation outcomes are more variable in children. Nonetheless, gradual, steady improvements in speech perception, speech production, and language does occur."

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 post-meningitis hearing loss under the age of 2 years have received an implant due to the risk of new bone formation associated with meningitis, which might preclude implantation at a later date."

American Cochlear Implant Alliance

The American Cochlear Implant Alliance challenges the past assumption that "one ear is good enough" and urges carriers to provide coverage for cochlear implants. Taking advantage of early intervention in the critical period of neural plasticity, promotes the best opportunity for binaural hearing and may result in a positive impact to a child's educational and social function.

American Speech-Language-Hearing Association

Despite normal hearing in one ear, children with significant unilateral hearing loss face educational, social, cognitive, and behavioral challenges. These children also face communication challenges—including difficulties with language and understanding speech in noise—and report poorer quality of life than their peers with normal hearing in both ears. The FDA approval (MED-EL cochlear implant system for single sided deafness and asymmetric hearing loss) will benefit people who have struggled with single-sided deafness or asymmetric hearing loss who do not benefit from traditional amplification. Untreated hearing loss is known to contribute to depression and social isolation, which can directly affect a person's quality of life.

U.S. PREVENTIVE SERVICES TASK FORCE RECOMMENDATIONS

Not applicable.

ONGOING AND UNPUBLISHED CLINICAL TRIALS

Some currently unpublished trials that might influence this review are listed in Table 3.

Table 3. Summary of Key Trials

NCT No.	Trial Name	Planned Enrollment	Completion Date
Ongoing			
NCT03900897[a]	Expanded Indications in the MED-EL Pediatric Cochlear Implant Population	60	Nov 2023 (active not recruiting)

NCT04793412	Cochlear Implantation in Children With Asymmetric Hearing Loss or Single-Sided Deafness Clinical Trial	80	Dec 2025 (recruiting)
NCT04506853[a]	Single-Sided Deafness and Asymmetric Hearing Loss Post-Approval Study	65	Sep 2026 (recruiting)
NCT05154188[a]	Post Approval Study to Assure the ContInued saFety and effectIveness of Neuro Cochlear Implant System in Adult Users (PACIFIC)	60	Feb 2028 (not yet recruiting)
NCT05318417[a]	A Post-approval, Prospective, Nonrandomized, Single-arm Multicenter Investigation to Evaluate the Safety and Effectiveness of Cochlear Implantation in Children and Adults With Unilateral Hearing Loss/Single-sided Deafness	60	Jun 2027 (recruiting)
Unpublished			
NCT03236909[a]	Expanded Indications in the Adult Cochlear Implant Population	44	Mar 2023 (completed)
NCT02203305[a]	Cochlear Implantation in Cases of Single-Sided Deafness	43	Sep 2021 (completed)
NCT05052944	Single-sided Deafness and Cochlear Implantation	78	Nov 2023 (completed)
NCT02379819[a]	Nucleus Hybrid L24 Implant System: New Enrollment Study	52	Apr 2022 (completed)
NCT03052920	Cochlear Implantation in Adults With Asymmetric Hearing Loss Clinical Trial	40	Mar 2021 (completed)
NCT02105441	Cochlear Implantation Among Adults and Older Children With Unilateral or Asymmetric Hearing Loss	40	Mar 2018 (completed)

NCT: national clinical trial.
[a] Industry-sponsored or partially sponsored.

Government Regulations
National:

NCD: Cochlear Implant (50.3) Effective date: 9/26/22; Implementation date: 3/24/23
Existing national coverage states:(59)

Indications and Limitations of Coverage

B. Nationally Covered Indications

Effective for services performed on or after September 26, 2022, 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. Limited benefit from amplification is defined by test scores of less than or equal to 60% correct in the best-aided listening condition on recorded tests of open-set sentence recognition. Patients must meet all of the following criteria.

- Diagnosis of bilateral moderate-to-profound sensorineural hearing impairment with limited benefit from appropriate hearing (or vibrotactile) aids;
- Cognitive ability to use auditory clues and a willingness to undergo an extended program of rehabilitation;

- Freedom from middle ear infection, an accessible cochlear lumen that is structurally suited to implantation, and freedom from lesions in the auditory nerve and acoustic areas of the central nervous system;
- No contraindications to surgery; and
- The device must be used in accordance with Food and Drug Administration (FDA)-approved labeling.

C. Nationally Non-Covered Indications

- Medicare beneficiaries not meeting all of the coverage criteria for cochlear implantation listed *in Section B* are deemed not eligible for Medicare coverage *except as described in Section D below*.

D. Other

CMS may provide coverage of cochlear implants for beneficiaries not meeting the coverage criteria listed in Section B when performed in the context of FDA-approved category B investigational device exemption clinical trials as defined at 42 CFR 405.201 or as a routine cost in clinical trials under section 310.1 of the National Coverage Determinations Manual titled Routine Costs in Clinical Trials.

Local:

There is no local coverage determination.

(The above Medicare information is current as of the review date for this policy. However, the coverage issues and policies maintained by the Centers for Medicare & Medicare Services [CMS, formerly HCFA] are updated and/or revised periodically. Therefore, the most current CMS information may not be contained in this document. For the most current information, the reader should contact an official Medicare source.)

Related Policies

- Auditory Brain Stem Implants
- Implantable Bone-Conduction and Bone-Anchored Hearing Devices
- Prosthetic Devices
- Semi-Implantable and Fully Implantable Middle Ear Hearing Aids

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The articles reviewed in this research include those obtained in an Internet based literature search for relevant medical references through July 11, 2024, the date the research was completed.

Joint BCBSM/BCN Medical Policy History

Policy Effective Date	BCBSM Signature Date	BCN Signature Date	Comments
7/12/02	7/12/02	7/12/02	Joint policy established
2/26/03	2/26/03	3/4/03	Routine maintenance
09/07/04	9/7/04	8/27/04	Routine maintenance
11/15/05	11/15/05	11/10/05	Maintenance review: added HCPCS codes L8620, L8621, L8622 as covered; changed indications to reflect Medicare guidelines (i.e., moderate – profound hearing loss)
7/1/2007	5/10/07	6/30/07	Maintenance review: added HCPCS codes L8623, L8624; added CPT codes 69714, 69715, 69717, 69718; deleted codes 92510, L8620
11/1/08	8/19/08	10/28/08	Maintenance review: deleted HCPCS codes 69714, 69715, 69717, 69718
5/1/10	2/16/10	2/16/10	Maintenance review: added HCPCS codes L7510, L8627, L8628, L8629
1/1/12	10/11/11	11/9/11	Maintenance review
3/1/13	12/11/12	12/31/12	Routine maintenance. Updated Medicaid information; added exclusion for upgrades to functional equipment to achieve aesthetic improvement.
7/1/14	4/10/14	4/15/14	Routine maintenance
7/1/15	4/21/15	5/8/15	Routine maintenance; added hybrid cochlear hearing systems as an exclusion, also reflected in policy statement.
7/1/16	4/19/16	4/19/16	Routine maintenance
3/1/17	12/22/16	12/27/16	• Routine maintenance • BCBSA guide (added specific criteria attached to inclusion bullets r/t low and high frequency and hear loss thresholds) • Added inclusion for cochlear hybrid devices
3/1/18	12/12/17	12/12/17	Routine maintenance

11/1/18	8/21/18	8/21/18	Routine maintenance
3/1/19	12/11/18		Routine maintenance
3/1/20	1/9/20		• Routine maintenance • FDA expansion for cochlear implant in one side deafness added
11/1/20	8/18/20		• Routine maintenance • FDA expansion of age allowance for Nucleus 24 Cochlear implant to 9 months of age and older added to inclusions
11/1/21	8/17/21		• Routine maintenance • No change in policy statement
11/1/22	8/16/22		• Routine maintenance
11/1/23	8/23/23		• Routine maintenance • Added to policy under Regulatory section: o In 2022, the Nucleus® Hybrid™ L24 Cochlear Implant System received expanded approval for single-sided deafness or unilateral hearing loss in adults and children age 5 or older (P970051/S205). • Updated Table 1. Cochlear Implant Systems Approved by the FDA and the Inclusions section. • Updated Inclusions section with the below language: o Unilateral or Bilateral cochlear implantation with an FDA approved cochlear implant is considered an established, safe, and effective therapy for individuals who are 9 months of age or older and who meet the following criteria: ▪ Unilateral or bilateral moderate to profound pre- or post-lingual

			▪ sensorineural hearing loss OR ▪ Limited or no benefit from hearing aid(s), defined as an aided monosyllabic word score of less than or equal to 50% correct in the ear to be implanted Vendor: N/A Post JUMP Updated the below statement under the Inclusionary and Exclusionary section: In certain situations, implantation **consideration may be given before 9 months** of age. One scenario post meningitis when cochlear ossification may preclude implantation. Another is in cases with a strong family history, because establishing a precise diagnosis is less uncertain. However, **these are NOT the only examples** where consideration may be given. (ky)
5/1/24	2/20/24		• This policy is coming early as code update – informational to add codes 92622 and 92623 eff 1/1/24 per code update as EST. This policy will go back to its original date of August, 2024 JUMP. • Vendor: N/A (ky)
11/1/24	8/20/24		• Routine maintenance. • Vendor: N/A (ky)

Next Review Date: 3rd Qtr, 2025

BLUE CARE NETWORK BENEFIT COVERAGE
POLICY: COCHLEAR IMPLANT

I. Coverage Determination:

Commercial HMO (includes Self-Funded groups unless otherwise specified)	Covered, policy guidelines apply
BCNA (Medicare Advantage)	Refer to the Medicare information under the Government Regulations section of this policy.
BCN65 (Medicare Complementary)	Coinsurance covered if primary Medicare covers the service.

II. Administrative Guidelines:

- The member's contract must be active at the time the service is rendered.
- Coverage is based on each member's certificate and is not guaranteed. Please consult the individual member's certificate for details. Additional information regarding coverage or benefits may also be obtained through customer or provider inquiry services at BCN.
- The service must be authorized by the member's PCP except for Self-Referral Option (SRO) members seeking Tier 2 coverage.
- Services must be performed by a BCN-contracted provider, if available, except for Self-Referral Option (SRO) members seeking Tier 2 coverage.
- Payment is based on BCN payment rules, individual certificate and certificate riders.
- Appropriate copayments will apply. Refer to certificate and applicable riders for detailed information.
- CPT - HCPCS codes are used for descriptive purposes only and are not a guarantee of coverage.
- Duplicate (back-up) equipment is not a covered benefit.