Medical Policy Manual Surgery, Policy No. 08

Cochlear Implant

Effective: June 1, 2022

Next Review: March 2023
Last Review: April 2022

IMPORTANT REMINDER

Medical Policies are developed to provide guidance for members and providers regarding coverage in accordance with contract terms. Benefit determinations are based in all cases on the applicable contract language. To the extent there may be any conflict between the Medical Policy and contract language, the contract language takes precedence.

PLEASE NOTE: Contracts exclude from coverage, among other things, services or procedures that are considered investigational or cosmetic. Providers may bill members for services or procedures that are considered investigational or cosmetic. Providers are encouraged to inform members before rendering such services that the members are likely to be financially responsible for the cost of these services.

DESCRIPTION

A cochlear implant is a device for the 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.

MEDICAL POLICY CRITERIA

Notes:

- This policy does not apply to surgically anchored bone-conduction hearing aids or externally worn air-conduction hearing aids. Cochlear implants are not hearing aids. While hearing aids function by amplifying sound, cochlear implants replace the functions of an absent or nonfunctioning cochlea.
- This policy does not address the use of the Nucleus® 24 Auditory Brain Stem Implant, which is designed to restore hearing in patients with neurofibromatosis who are deaf secondary to removal of bilateral acoustic neuromas.
- Hybrid cochlear implant/hearing aid systems are devices that include a hearing aid integrated into the external sound processor of the cochlear implant. If hearing aid components of such systems are billed separately, there may be specific member

benefit language addressing coverage of hearing aids that would be applicable. Contract language takes precedence over medical policy.

- Repeat hearing tests or trials of hearing aids are not necessary for patients who have previously met Criteria I. and II. as it is unlikely that natural hearing or the benefit from hearing aids will improve significantly over time.

I. **Unilateral or bilateral implantation of cochlear implants**, ***other than*** cochlear implant/hearing aid ***hybrid*** devices, and associated aural rehabilitation may be considered **medically necessary** when all of the following criteria (A. – D.) are met:

A. Meets one of the following age requirements:

1. Age 9 months or older for the Nucleus 24 cochlear implant system (with any of the Cochlear® sound processors); or
2. Age 12 months or older.

B. Meets one or more of the following:

1. Patients diagnosed with enlarged vestibular aqueduct (EVA) (greater than 1mm at the midpoint), as evidenced by MRI or CT imaging; or
2. Patients with both of the following (a. and b.):
   a. Bilateral severe to profound pre- or postlingual (sensorineural) hearing loss, defined as a pure-tone average of 70 decibels (dB) hearing threshold or greater at 500 Hz (hertz), 1000 Hz and 2000 Hz; and
   b. Limited or no benefit from hearing aids (defined below) unless hearing aids are unreasonable.
      i. Adults: Scores less than or equal to 50 percent correct on tape recorded sets of open-set sentence recognition in the ear to be implanted.
      ii. Children: Failure to develop basic auditory skills, and in older children, less than or equal to 30 percent correct on open-set tests.

C. Implanted device is FDA approved (PMA or 510k only).

D. Patients do not have any of the following contraindications:

1. Deafness due to lesions of the acoustic nerve (eighth cranial nerve), central auditory pathways, or brain stem in the implanted ear.
2. Active or chronic infections of the external or middle ear and mastoid cavity in the implanted ear, including but not limited to otitis media.
3. Tympanic membrane perforation.
4. Radiographic evidence of absent cochlear development in the implanted ear.
5. Inability or lack of willingness to participate in post-implantation aural rehabilitation.

II. **Unilateral implantation of *hybrid* cochlear implant/hearing aid systems** that include the hearing aid integrated into the external sound processor of the cochlear

implant may be considered **medically necessary** when all of the following criteria are met (A. – F.):

A. Age 18 years or older.

B. Bilateral severe to profound pre- or postlingual (sensorineural) hearing loss, defined as a pure-tone average of 70 decibels (dB) hearing threshold or greater at 500 Hz (hertz), 1000 Hz and 2000 Hz.

C. Limited or no benefit from hearing aids unless hearing aids are unreasonable, defined as scores less than 50 percent correct on tape recorded sets of open-set sentence recognition in the ear selected for implantation.

D. Meets all of the following (1. and 2.):

1. All of the following in the ear selected for implantation (a. – c.):

   a. Low frequency hearing thresholds no poorer than 60 dB hearing level up to and including 500 Hz (averaged over 125, 250, and 500 Hz; i.e., threshold average of 125, 250, and 500 Hz less than or equal to 60 dB hearing level); and

   b. Severe to profound mid-to-high frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz greater than or equal to 75 dB hearing level); and

   c. Aided consonant-nucleus-consonant word recognition score from 10 percent to 60 percent in the preoperative aided condition.

2. All of the following for the contralateral ear (a and b):

   a. Moderately severe to profound mid-to-high frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz greater than or equal to 60 dB hearing level); and

   b. Aided consonant-nucleus-consonant word recognition score equal to or better than that of the ear selected for implantation but not more than 80 percent correct.

E. Implanted device is FDA approved (PMA or 510k only).

F. Does not have any of the following contraindications:

1. Deafness due to lesions of the acoustic nerve (eighth cranial nerve), central auditory pathways, or brain stem in the implanted ear

2. Active or chronic infections of the external or middle ear and mastoid cavity in the implanted ear, including but not limited to otitis media

3. Tympanic membrane perforation

4. Radiographic evidence of absent cochlear development in the implanted ear

5. Inability or lack of willingness to participate in post-implantation aural rehabilitation

6. A duration of severe to profound hearing loss of 30 years or greater.

III. Implantation of cochlear implants is considered **not medically necessary** when Criterion I. or II. above is not met.

IV. **Implant replacement**, including **replacement parts or upgrades** to existing cochlear implants and/or components, may be considered **medically necessary** when components are no longer functional, or for functional devices only in the small subset of patients whose response to existing components is inadequate to the point of interfering with activities of daily living, which would include school and work.

V. **Implant replacement, including replacement parts or upgrades** to existing cochlear implants and/or components, are considered **not medically necessary** when Criterion IV. is not met, including but not limited to upgrades of existing, functioning external systems to achieve aesthetic improvement, such as smaller profile components, or a switch from a body-worn external sound processor to a behind-the-ear (BTE) model.

NOTE: A summary of the supporting rationale for the policy criteria is at the end of the policy.

POLICY GUIDELINES

A Pure Tone Average (PTA) is determined by averaging the hearing threshold levels at a set of specified frequencies: for example, 500, 1000, and 2000 Hz (PTA = 500 Hz (T)+ 1000 Hz (T) + 2000Hz (T) ÷ 3).

LIST OF INFORMATION NEEDED FOR REVIEW

REQUIRED DOCUMENTATION:

It is critical that the list of information below is submitted for review to determine if the policy criteria are met. If any of these items are not submitted, it could impact our review and decision outcome.

- History and Physical/Chart Notes
- Manufacturer and Model Name of Cochlear Implant being requested
- Audiology test results

CROSS REFERENCES

1. Transcutaneous Bone-Conduction and Bone-Anchored Hearing Aids, Surgery, Policy No. 121

BACKGROUND

A cochlear implant provides direct electrical stimulation to the auditory nerve, bypassing the usual transducer cells that are absent or nonfunctional in deaf cochlea. The basic components of a cochlear implant include both external and internal components. The external components include a microphone, an external sound processor, and an external transmitter. The internal components are implanted surgically and include an internal receiver implanted within the temporal bone, and an electrode array that extends from the receiver into the cochlea through a surgically created opening in the round window of the middle ear.

Sounds that are picked up by the microphone are carried to the external signal 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.

Hearing loss is rated on a scale based on the threshold of hearing. Severe hearing loss is defined as a bilateral hearing threshold of 70-90 decibels (dB) and profound hearing loss is defined as a hearing threshold of 90 dB and above.

A post-cochlear implant rehabilitation program is necessary to achieve benefit from the cochlear implant. The rehabilitation program includes development of skills in understanding running speech, recognition of consonants and vowels, and tests of speech perception ability.

REGULATORY STATUS

Note: Full FDA approval includes only Premarket Approval (PMA) and 510k approval. Devices with Investigational Device Exemption (IDE) or Humanitarian Device Exemption (HDE) are not considered fully FDA approved.

Several cochlear implants are commercially available in the United States. The FDA-labeled indications for currently marketed electrode arrays are summarized in the table below. Over the years, subsequent generations of the various components of the devices have been FDA approved, 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.

Manufacturer and FDA approved Cochlear Implants	Indications for Adults or Children
CONVENTIONAL COCHLEAR IMPLANTS	
Advanced Bionics® • HiRes™ Ultra implant • HiResolution Bionic Ear System (HiRes 90K*) Sound Processors: • ClearVoice • HiRes Fidelity 120 • HiRes Optima Predecessors: • Clarion Multi-Strategy • HiFocus CII Bionic Ear	<u>Adults</u>: • ≥ 18 years of age • Post-lingual onset of severe to profound bilateral sensorineural hearing loss [≥70 decibels (dBs)] • Limited benefit from appropriately fitted hearing aids, defined as scoring ≤ 50% on a test of open-set Hearing in Noise Test (HINT) sentence recognition <u>Children:</u> • 12 months to 17 years of age • Profound bilateral sensorineural deafness (>90dB) • Use of appropriately fitted hearing aids for at least 6 months in children 2 to 17 years of age or at least 3 months in children 12 to 23 months of age. • Lack of benefit in children <4 years of age is defined as a failure to reach developmentally-appropriate auditory milestones (e.g., spontaneous response to name in quiet or to environmental sounds) measured using the Infant-Toddler Meaningful Auditory Integration Scale or Meaningful Auditory Integration Scale or < 20% correct on a simple open-set word recognition test (Multisyllabic Lexical Neighborhood Test) administered using monitored live voice [70 dB SPL (sound pressure level)] • Lack of hearing aid benefit in children >4 years of age is defined as scoring < 12% on a difficult open-set word recognition test (Phonetically Balanced-Kindergarten Test) or < 30% on an open-set sentence test (HINT for Children) administered using recorded materials in the soundfield (70 dB SPL)

Manufacturer and FDA approved Cochlear Implants	Indications for Adults or Children
Cochlear® • Nucleus CI600 series • Nucleus CI500 series • Nucleus CI24RE series • Nucleus 24 series Sound Processors: • Kanso® 2 • Kanso® • Nucleus® 7 • Nucleus® 6 • Nucleus® 5* • Nucleus Freedom Predecessors: • Nucleus 22, 24	<u>Adults</u>: • ≥ 18 years old • Pre- or post-lingual onset of moderate to profound bilateral sensorineural hearing loss • ≤50% sentence recognition in the ear to be implanted • ≤60% sentence recognition in the opposite ear or binaurally <u>Children 9 months to 24 months:</u> • Profound sensorineural hearing loss bilaterally • Limited benefit from appropriate binaural hearing aids • Lack of progress in the development of auditory skills <u>Children 25 months to 17 years 11 months</u>: • Severe to profound bilateral sensorineural hearing loss • Multi-syllabic Lexical Neighborhood Test (MLNT) scores of ≤30% in best-aided condition in children 25 months to 4 years 11 months • Lexical Neighborhood Test (LNT) scores of ≤30% in best-aided condition in children 5 years to 17 years and 11 months • Lack of progress in the development of auditory skills

Med El®
 	
• Maestro system
• Synchrony Implant
• Synchrony 2 Implant
• Concerto Implant
 	
Sound Processors:
• Sonnet
• Sonnet 2
• Concerto implant
• Opus
• Opus 2
• Rondo 2
 	
Predecessors:
• Combi 40+
• Sonata
• Pulsar
	<u>Bilateral Hearing Loss</u>
	<u>Adults</u>:
	• ≥ 18 years old
	• Severe to profound bilateral sensorineural hearing loss (≥70dB)
	• ≤40% correct Hearing in Noise test (HINT) sentences with best-sided listening condition
	
	<u>Children:</u>
	• 12 months to 18 years with profound sensorineural hearing loss (≥90dB)
	• In younger children, little or no benefit is defined by lack of progress in the development of simple auditory skills with hearing aids over a 3-6 month period
	• In older children, lack of aided benefit is defined as <20% correct on the MLNT or LNT depending upon the child's cognitive ability and linguistic skills
	• A 3-6 month trial with hearing aids is required if not previously experienced
	
	<u>Single-Sided Deafness and Asymmetric Hearing Loss</u>
	• ≥ 5 years old
	• Single-sided deafness (SSD) or asymmetric hearing loss (AHL), where:
	  o SSD is defined as profound sensorineural hearing loss in one ear and normal hearing or mild sensorineural hearing loss in the other ear.
	  o 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.

Manufacturer and FDA approved Cochlear Implants	Indications for Adults or Children
	• Limited benefit from an appropriately fitted unilateral hearing aid in the ear to be implanted. • For ages 18 years-old and above, limited benefit from unilateral amplification is defined by test scores of 5% correct or less on monosyllabic consonant-nucleus-consonant (CNC) words in quiet when tested in the ear to be implanted alone. • For ages between 5 and 18 years-old, 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 alone • At least 1 month experience wearing a Contra Lateral Routing of Signal (CROS) hearing aid or other relevant device and not show any subjective benefit
Oticon Medical Neuro Cochlear Implant System (Neuro 2 sound processor and Neuro Zti implant)	Adults: • Severe-to-profound bilateral SNHL (≥70 dB at 500, 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
HYBRID COCHLEAR IMPLANTS	
Cochlear® • Nucleus® Hybrid™ L24 Cochlear Implant (Nucleus 6)	Adults: • ≥ 18 years old • Residual low-frequency hearing sensitivity • Severe to profound high-frequency sensorineural hearing loss • Limited benefit from appropriately fit bilateral hearing aids
Med El® • Med EL EAS™	Adults: • ≥ 18 years old • Residual low-frequency hearing sensitivity • Severe to profound high-frequency sensorineural hearing loss • Candidates should go through a suitable hearing aid trial, unless already appropriately fit with hearing aids
RECENTLY FDA-APPROVED DEVICES	
• New devices that come onto the market are added to the policy at policy updates. In the interim, new devices may be approved for coverage for FDA-approved indications when applicable criteria are met.**	

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

** FDA-approved indications can be found by searching by device name in the FDA 510(k) Premarket Notification Database or the De Novo Database and viewing the Summary.

While cochlear implants have typically been used mono laterally, in recent years, interest in bilateral cochlear implantation has arisen. The proposed benefits of bilateral cochlear implants are to improve understanding of speech in noise and localization of sounds. Improvements in

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

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

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

In September 2016, FDA approved the Med EL EAS™ (Electric Acoustic Stimulation) Hearing Implant System (Med EL Corp.).[2] 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 the combination of the SYNCHRONY cochlear implant and the SONNET EAS audio processor. According to the FDA's premarket approval notification:[3]

> The MED-EL EAS System is indicated for partially deaf individuals aged 18 years and older who have residual hearing sensitivity in the low frequencies sloping to a severe/profound sensorineural hearing loss in the mid to high frequencies, and who obtain minimal benefit from conventional acoustic amplification. Typical preoperative hearing of candidates ranges from normal hearing to moderate sensorineural hearing loss in the low frequencies (thresholds no poorer than 65 dB HL up to and including 500 Hz) with severe to profound mid- to high-frequency hearing loss (no better than 70 dB HL at 2000 Hz and above) in the ear to be implanted. For the non-implanted ear, thresholds may be worse than the criteria for the implanted ear, but may not be better. The CNC word recognition score in quiet in the best-aided condition will be 60% or less,

in the ear to be implanted and in the contralateral ear. Prospective candidates should go through a suitable hearing aid trial, unless already appropriately fit with hearing aids.

EVIDENCE SUMMARY

Cochlear implants (CI) are recognized effective treatment of sensorineural deafness in select patient, as noted in a 1995 National Institutes of Health Consensus Development conference, which offered the following conclusions:[4]

- Cochlear implantation has a profound impact on hearing and speech reception in postlingually deafened adults with positive impacts on psychological and social functioning.
- The results are more variable in children. Benefits are not realized immediately but rather are manifested over time, with some children continuing to show improvement over several years.
- Prelingually deafened adults may also benefit, although to a lesser extent than postlingually deafened adults. These individuals achieve minimal improvement in speech recognition skills. However, other basic benefits, such as improved sound awareness, may meet safety needs.
- Training and educational intervention are fundamental for optimal post implant benefit.
- 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 have been implanted under the age of two years due to the risk of new bone formation associated with meningitis, which may preclude a cochlear implant at a later date.

ENLARGED VESTIBULAR AQUEDUCTS (EVA)

Enlarged vestibular aqueduct (also known as enlarged vestibular aqueduct syndrome (EVAS), large vestibular aqueduct, large vestibular aqueduct syndrome (LVAS), or dilated vestibular aqueduct) is a condition which is associated with childhood hearing loss. According to the NIH National Institute on Deafness and other Communication Disorders (NIDCD):[5] most children with enlarged vestibular aqueducts (EVA) will develop some amount of hearing loss, and approximately 5 to 15% of children with sensorineural hearing loss (hearing loss caused by damage to sensory cells inside the cochlea) have EVA.

Systematic Reviews

Pan (2022) reported a systematic review and meta-analysis of the safety and effectiveness of cochlear implantation for patients with large vestibular aqueduct deformity.[6] A total of five randomized controlled trials met inclusion criteria. There was low to high risk of bias for blinding of participants and personnel and low or unclear risk of bias for the other evaluated biases. Meta-analysis evaluated postoperative hearing ability and speech intelligibility rate between EVA patients and those with normal inner ear structure. No significant differences between groups were identified.

In 2014, Xu conducted a systematic review in Chinese to assess the efficacy and safety of cochlear implantation in deaf patients with inner ear malformations compared to deaf patients with normal inner ear structure, including 11 RTCs (n=655 patients).[7] In terms of postoperative complications, electrode impedance, behavior T-level, hearing abilities and

speech discrimination; patients with mixed inner ear malformations, Mondini syndrome or EVA were not significantly different than controls. However, the reviewers concluded that additional larger controlled studies with longer follow-up may help to evaluate the efficacy of cochlear implantation for deaf patients with inner ear malformation more reliably.

In 2012, Pakdaman conducted a systematic review to determine if abnormal cochleovestibular anatomy influences surgical and audiologic outcomes following cochlear implant (CI) surgery in children, including 22 studies.[8] Out of the 311 children included, 89 (29%) were diagnosed with EVA, considered to be a mild/moderate anomaly. Outcomes of CI surgery were analyzed based on the severity of the ear malformation (mild/moderate anomaly versus severe), and subgroup analyses were not performed based on the different malformations observed. The reviewers reported that severe inner ear dysplasia was associated with increased surgical difficulty and lower speech perception.

Nonrandomized Studies

There have been a number of case series and retrospective analyses published on the efficacy of cochlear implants in patients with EVA, all generally reporting an improvement of outcomes including various clinical scores for hearing improvement and scores measuring quality of life. These studies range in size from three to 47 cases.[9-19] Some of these studies have focused on pediatric patients, while others have included mixed patient populations and have not analyzed pediatric patients from adults in terms of outcomes. Overall, these studies report that outcomes in EVA patients are comparable to cochlear implant patients with no malformations, including similar risk of cerebrospinal fluid (CSF) gusher during cochlear implantation.

There is research indicating that the age of cochlear implantation for patients with EVA affects health outcomes. In 2013, Ko conducted a study (1) to assess health outcomes of Mandarin-speaking patients with EVA after cochlear implantation (CI); (2) to compare their performance with a group of CI users without EVA; (3) to understand the effects of age at implantation and duration of implant use on the CI outcomes.[20] Forty-two patients with EVA participating in this study were divided into two groups: the early group received CI before five years of age and the late group after five years of age. The patients with EVA with more than five years of implant use (18 cases) achieved a mean score higher than 80% on the most recent speech perception tests and reached the highest level on the CAP/SIR scales. The early group developed speech perception and intelligibility steadily over time, while the late group had a rapid improvement during the first year after implantation. The two groups, regardless of their age at implantation, reached a similar performance level. These patients do not necessarily need to wait until their hearing thresholds are higher than 90 dB HL or PB word score lower than 40% to receive CI. Similar results have been reported in small pediatric case series, indicating that if patients receive cochlear implants prior to becoming severely to profoundly deaf, that residual hearing is preserved.[9, 21]

In contrast to studies reporting favorable outcomes, one small retrospective study performed by Bichy in 2002 that reported better hearing outcomes in patients with EVA using hearing aid than those who had undergone cochlear implantation.[22] The analysis in this study included 16 children and adults with EVA that had undergone cochlear implantation and 10 children and adults undergoing treatment of progressive or fluctuant sensorineural hearing loss with the use of a hearing aid alone. Although the hearing aid group had a better mean pure-tone average (70.8 dB; SD 24.4) versus (107.0 dB; SD 21.7) for the cochlear implant group, the use of

health utility indexes determined that greater net health benefit (including quality of life) was derived from cochlear implantation over hearing aids.

INFANTS UNDER AGE 12 MONTHS

The literature review focused on studies comparing the impact on hearing, speech development and recognition, and complication rates of implantation in infants younger than 12 months with those of older age groups. This includes the question of whether any early benefits that may occur in these very young patients later converge with those in older patients.

Systematic Reviews

Sbeih (2022) reported a systematic review that assessed the safety of cochlear implantation in children 12 months and younger.[23] A total of 18 studies met inclusion criteria. Major and minor complications were reported in 3.1% and 2.4% of patients, respectively. The authors noted that this is similar to rates of complications in older cohorts.

Two older systematic reviews were identified that addressed CI in children under 12 months of age. The reviews, summarized below, reported few studies of CI in this age group compared with CI in children over one year of age. Both systematic reviews ranked the available studies as poor to fair due to heterogeneity in study participants and study designs, and high risk for potential bias. In addition, differences in outcomes between the age groups did not reach statistical significance.

In 2011 Forli reported similar findings in seven studies comparing CI implanted prior to one year of age with implantations performed after one year of age.[24] The studies precluded meta-analysis due to heterogeneity of age ranges analyzed and outcomes evaluated. While studies suggested improvements in hearing and communicative outcomes in children receiving implants prior to one year of age, between-group differences did not reach statistical significance. In addition, it is not certain whether any improvements were related to duration of cochlear implant usage rather than age of implantation. Nor is it clear whether any advantages of early implantation are retained over time.

In 2010, Vlastarakos conducted a systematic review of studies on bilateral cochlear implants in a total of 125 children implanted before one year of age.[25] The authors noted that follow-up times ranged from a median duration of 6 to 12 months and, while results seemed to indicate accelerated rates of improvement in implanted infants, the evidence available was limited and of lower quality. Additionally, the lack of reliable outcome measures for infants demonstrated the need for further research before cochlear implantation prior to one year of age becomes widespread.

Nonrandomized Studies

In March 2020, the FDA approved an expansion of the indications for Cochlear Americas' Nucleus 24 cochlear implant system for infants aged 9 to 12 months of age with bilateral profound sensorineural deafness who demonstrate limited benefit from appropriate binaural hearing aids. Previously, this device was approved for ages 12 months and older. According to the FDA's summary of safety and effectiveness data, approval was based on supporting evidence from a comprehensive literature review and a clinical feasibility study. The clinical feasibility study was a retrospective clinical analysis of 84 subjects implanted with cochlear implants between the ages of 9 and 12 months. Descriptive statistics were reported for time

under anesthesia (unilateral: 2hrs 34min, bilateral: 4hrs 15min), estimated blood loss (unilateral: 10.75 cc, bilateral: 19.88 cc), time in recovery (unilateral: 2hr 18min, bilateral: 1hr 59min), and adverse events (Percent of subjects: 2.4% cerebral spinal fluid leak; 2.4% facial weakness; 2.4% infection; 7.1% minor post-op complication; 3.6% minor skin irritation; 3.6% otitis media; 2.4% seroma; 7.1% temperature regulation during procedure).

The supporting literature review identified 49 articles including 750 total (not necessarily unique) patients implanted with cochlear implants prior to 12 months of age. Safety results were reported on a per-study basis with no meta-analysis. Complication rates were reported between 1.5% and 10% except for two studies. One reported a rate of 29%, and the other reported on two techniques, one of which had a rate of 20.6% and the other 61.5%. Two studies compared complications across different age ranges. One reported similar complication rates across ages and the other reported higher rates for younger ages. The summary section states that the study findings support that the safety profile for cochlear implantation in pediatric patients who are implanted between 9 and 12 months of age is comparable to that of the currently approved population of age 12 months and older. Effectiveness results were reported on a per-study basis with no meta-analysis. No study reported worse hearing outcomes for the early-implanted group and many reported significantly better outcomes for this group.

A 2017 retrospective study by Kalejaiye assessed surgical complications, operative times, and reoperation rates in 73 patients under one year of age.[27] They compared these patients, identified from the American College of Surgeons National Surgical Quality Improvement Program Pediatric database (2012-2013), with pediatric patients in the database above the age of one. They found that the patients under one year had higher readmission rates (6.9% vs. 2.7%) and longer mean operative times (191 minutes vs. 160 minutes), but no significant differences were noted in complication rate, postoperative length of stay, or reoperation rate.

In 2015, Guerzoni conducted a 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).[28] The investigators reported that at one-year follow-up, assertiveness and responsiveness scores were within the normal range of normal-hearing age-matched peers. Age at cochlear implant activation exerted a significant impact, with the highest scores associated to the youngest patients.

In 2011, Colletti reported on the 10-year results comparing 19 children with cochlear implants received between the ages of 2 to 11 months to 21 children implanted between 12-23 months and 33 children implanted between 24 to 35 months.[29] Within the first six months post-implantation, there was no significant difference among groups in Category of Auditory Performance testing but differences became significantly better in the infant group (early implantation) at the 12 and 36 month testing. Previously, Colletti reported on findings from 13 infants who had implants placed before 12 months.[30] The procedures were performed between 1998 and 2004. In this small study, the rate of receptive language growth for these early implant infants overlapped scores of normal-hearing children. This overlap was not detected for those implanted at 12 to 23 or 24 to 36 months.

In 2009 Ching published an interim report on early language outcomes of children with cochlear implants.[31] This study evaluated 16 children who had implants before 12 months of age compared to 23 who had implants after 12 months (specific time of implantation was not provided). The preliminary results demonstrated that children who received an implant before

12 months of age developed normal language skills at a rate comparable to normal-hearing children, while those with later implants performed at two standard deviations below normal. The authors noted that these results are preliminary, as there is a need to examine the effect of multiple factors on language outcomes and the rate of language development.

Johr (2008) highlighted the surgical and anesthetic considerations when performing cochlear implant surgery in very young infants.[32] This was an observational study and literature review by pediatricians at a tertiary children's hospital in Switzerland. Surgical techniques and anesthetic management aspects of elective surgeries in small infants were analyzed in patients younger than one year of age undergoing cochlear implant surgeries. The results demonstrated that the age of the patient and the pediatric experience of the anesthesiologist, but not the duration of the surgery, are relevant risk factors. The authors concluded, "Further research is needed to provide more conclusive evidence that the performance outcome for children implanted before 12 months of age does not converge with the results of children implanted between 12 and 18 months."

ADULTS AND CHILDREN OVER AGE 12 MONTHS

Since there is sufficient evidence that bilateral and unilateral cochlear implants are safe and lead to improvements in health outcomes in adults and children over the age of twelve months with bilateral severe to profound pre- or postlingual (sensorineural) hearing loss, the evidence reviewed below will be focused on systematic reviews and randomized studies. Nonrandomized studies will not be described in detail.

Systematic Reviews

The following is a summary of the most recent systematic reviews related to CI. These reviews included a critical analysis of the quality of the included studies. While noting the heterogeneity of the studies, and the potential for bias, these reviews found that the studies consistently reported beneficial outcomes for both bilateral and unilateral CI in select children and adults compared with no hearing devices or with conventional hearing aids.

Adults

A technology assessment published by Health Quality Ontario in 2018 evaluated bilateral cochlear implantation in adults and children in separate analyses.[33] The literature search conducted through March 2017 identified 10 studies on bilateral cochlear implantation in adults: three RCTs and seven prospective observational studies. Two of the three RCTs included data from a single RCT and compared simultaneous bilateral with unilateral cochlear implantation for severe bilateral sensorineural hearing loss. The third RCT randomized 24 adult patients with severe bilateral sensorineural hearing loss to receive bilateral implantation immediately or after a six-month waiting period. The observational studies performed within- or between-patient comparisons of bilateral cochlear implantation with unilateral cochlear implantation with or without hearing aids in the nonimplanted ear. Study quality was evaluated using the GRADE system. The quality of the RCTs was high, medium, and low and the quality of the prospective observational studies ranged from very low to low. The GRADE of evidence for adults overall was rated moderate to high. Overall, the authors concluded that bilateral cochlear implantation improved sound localization, speech perception in noise, and subjective benefits of hearing and that the safety profile was acceptable.

In a meta-analysis, McRackan (2018) examined the impact of cochlear implantation on quality of life (QOL).[34] 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 (standardized mean difference [SMD]=51.77). 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). A 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 (2018) aimed to determine the change in general health-related quality of life (HRQOL) after cochlear implantation and association with speech recognition.[35] Twenty-two articles met criteria for meta-analysis of HRQOL improvement, but 15 (65%) were excluded due to incomplete statistical reporting. From the seven 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.

In 2013, the authors of the 2011 AHRQ technology assessment reported the following findings of an updated systematic review of studies published through May 2012:[36]

- Unilateral cochlear implants

  Sixteen (of 42) studies were of unilateral cochlear implants. Most unilateral implant studies showed a statistically significant improvement in mean speech scores as measured by open-set sentence or multi-syllable word tests. A meta-analysis of four studies revealed a significant improvement in cochlear-implant relevant quality of life (QOL) after unilateral implantation. However, these studies varied in design and there was considerable heterogeneity observed across studies, making it difficult to compare outcomes across studies.

- Bilateral cochlear implants

  Thirteen studies reported improvement in communication-related outcomes with bilateral implantation compared with unilateral implantation and additional improvements in sound localization compared with unilateral device use or implantation only. The risk of bias varied from medium to high across studies. Based on results from at least two studies, the QOL

outcomes varied across tests after bilateral implantation. A meta-analysis was not performed because of heterogeneity in design between the studies.

In 2012 and 2013 Crathorne and van Schoonhoven, respectively, published updated systematic reviews for the National Institute for Health and Care Excellence (NICE). Included studies were from the U.S. and Europe and compared bilateral with unilateral cochlear implants. In two studies the unilateral implant group also had an acoustic hearing aid for the contralateral ear. Neither systematic review was able to conduct a meta-analysis due to the heterogeneity of the studies and the level of evidence of the studies which was rated as moderate-to-poor.

In October 2011, Berrettini published results of a systematic review of unilateral and bilateral cochlear implant effectiveness in adults.[37]

- Unilateral cochlear implants

  Eight articles on unilateral cochlear implants in advanced age patients were included. All of the studies reported benefits with cochlear implantation despite advanced age at time of implant (age 70 years or older). In six studies, results were not significantly different between younger and older patients. However, two studies reported statistically significant inferior perceptive results (e.g., hearing in noise test and consonant nucleus consonant test) in older patients. This systematic review also examined three studies totaling 56 adults with pre-lingual deafness who received unilateral cochlear implants. The authors concluded unilateral cochlear implants provided hearing and quality-of-life benefits in prelingually deaf patients, but results were variable.

- Bilateral cochlear implants

  Thirteen articles on bilateral cochlear implants were reviewed. Sound localization improved with bilateral cochlear implants compared with monaural hearing in six studies. Significant improvements in hearing in noise and in quiet environments with bilateral implants compared with unilateral implants were reported in ten studies and seven studies, respectively. Five of the studies reviewed addressed simultaneous implantation, five studies reviewed sequential implantation, and three studies included a mix of simultaneous and sequential implantation. However, no studies compared simultaneous to sequential bilateral implantation results, and no conclusions could be made on the timing of bilateral cochlear implantation.

In June 2011 the most recent technology assessment, by the Tufts Evidence-based Practice Center for the Agency for Health Care Research and Quality (AHRQ), reported the following findings on the effectiveness of unilateral and bilateral cochlear implants (CIs) in adults:[38]

- Unilateral cochlear implants

  The assessment examined 22 studies with 30 or more patients and concluded that, while the studies reviewed were rated as poor to fair quality, unilateral cochlear implants are effective in adults with sensorineural hearing loss. Pre- and post-cochlear implant scores on multi-syllable tests and open-set sentence tests demonstrated significant gains in speech perception regardless of whether a contralateral hearing aid was used along with the cochlear implant. Additionally, the assessment found generic and disease-specific health-related quality of life improved with unilateral cochlear implants. However, the

available evidence was insufficient to draw conclusions on improvements in open-set sentence test scores (i.e., >40% and ≤50% or >50% and ≤60%), and any relationship between pre-implantation patient characteristics and outcomes [e.g., age, duration of hearing impairment, Hearing in Noise Test (HINT) scores and pre- or post-linguistic deafness.]

- Bilateral cochlear implants

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

Children

The technology assessment published by Health Quality Ontario in 2018 discussed above regarding its findings on adult implantation identified 14 studies (all prospective observational studies) on bilateral cochlear implantation in children.[33] Two studies included both sequential and simultaneous bilateral implantation while the rest evaluated sequential only. As for adults, overall, the authors concluded that bilateral cochlear implantation improved sound localization, speech perception in noise, and subjective benefits of hearing and that the safety profile was acceptable (GRADE of evidence: moderate to high). The authors additionally concluded that bilateral cochlear implantation allowed for better language development and more vocalization in preverbal communication in children (GRADE of evidence: moderate).

In a 2015 systematic review, Fernandes evaluated 18 published studies and two dissertations that reported hearing performance outcomes for children with ANSD and cochlear implants.[39] 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 with ANSD and cochlear implants with the speech perception in children with sensorineural hearing loss and cochlear implants. Most of these studies concluded that children with ANSD and cochlear implants developed hearing skills similar to those with sensorineural hearing loss and cochlear implants; however, these types of studies do not allow comparisons of outcomes between ANSD patients treated with cochlear implants and those treated with usual care.

In a 2014 systematic review, Lammers summarized the evidence on the effectiveness of bilateral cochlear implantation compared with unilateral implantation among children with

sensorineural hearing loss.[40] The authors identified 21 studies that evaluated bilateral cochlear implantation in children, with no RCTs identified. Due to the limited number of studies, heterogeneity in outcomes and comparison groups, and high risk for bias in the studies, the authors were unable to perform pooled statistical analyses, so a best-evidence synthesis was performed. The best-evidence synthesis demonstrated that there was consistent evidence indicating the benefit of bilateral implantation for sound localization. One study demonstrated improvements in language development, although other studies found no significant improvements. The authors noted that the currently available evidence 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.

In 2013, Eze published a systematic review comparing outcomes for cochlear implantation for children with developmental disability with those without developmental disability.[41] The authors noted that while approximately 30% to 40% of children who receive cochlear implants have developmental disability and that evidence about outcomes in this group was limited. Their review included 13 studies that compared receptive or expressive language outcomes in children with cochlear implants with and without developmental disability. The included studies were heterogeneous in terms of comparator groups and outcome measures, precluding data pooling and meta-analysis. In a structured systematic review, the authors reported that seven of the eligible studies demonstrated a significantly poor cochlear implant outcome in children with developmental disability, while the remaining studies reported no significant difference in outcomes between the groups.

Humphriss (2013) published a systematic review evaluating outcomes after cochlear implantation among pediatric patients with auditory neuropathy spectrum disorder (ANSD), a sensorineural hearing disorder characterized by abnormal auditory brainstem response with preserved cochlear hair cell function as measured by otoacoustic emissions testing.[42] The authors identified 27 studies that included an evaluation of cochlear implantation in patients with ANSD, including 15 noncomparative studies, one that compared children with ANSD who received a cochlear implant with children with ANSD with hearing aids, and 12 that compared children with ANSD who received a cochlear implant with children with severe sensorineural hearing loss who received a cochlear implant. Noncomparative studies were limited in that most (11/15) did not include a measure of speech recognition before cochlear implantation. Among the comparative studies, those comparing cochlear implantation to “usual care”, typically a hearing aid, provided the most information about effectiveness of cochlear implantation among patients with ANSD; the one small study that used this design found no significant differences between the groups. Overall, the authors suggested that further RCT evidence is needed.

Randomized Trials

In 2016, Smulder conducted a small prospective multi-center randomized trial to evaluate the benefits of bilateral implants compared to unilateral implants in adults with postlingual deafness, including 38 patients.[43] At one-year follow-up, there were no significant differences between groups on the speech-in-noise or the consonant-vowel-consonant test. The bilaterally implanted group performed significantly better when noise came from different directions ($p < 0.001$) and was better able to localize sounds ($p < 0.001$) compared to the unilaterally implanted group. These results were consistent with the patients' self-reported hearing capabilities. The results were consistent at a two year follow up, reported in 2017.[44]

Nonrandomized Studies

Adults

Numerous case series have been published on adult patients with bilateral cochlear implants.[45-53] Most but not all studies report slight to modest improvements in sound localization and speech intelligibility with bilateral cochlear implants especially with noisy backgrounds but not necessarily in quiet environments. In addition, depression scores improved in cochlear implant patients from pre-implantation to 12 months post-treatment (geriatric depression scale improvement: 31%, 95% CI 10% to 47%) in a prospective observational study including 113 patients with postlingual hearing loss, of whom 50 were treated with cochlear implants and 63 with hearing aids.[54]

When reported, the combined use of binaural stimulation improved hearing in the range of one to four decibels or 1 to 2%. While this improvement seems slight, any improvement in hearing can be considered beneficial in the deaf. However, this improvement may not outweigh the significant risks of a second implantation. In addition, similar binaural results can be achieved with a contralateral hearing aid, assuming the contralateral ear has speech recognition ability. A number of studies have reported benefits for patients with a unilateral cochlear implant with hearing aid (HA) in the opposite ear.

Children

Several recent publications have evaluated bilateral cochlear implants in children.[55-57] These studies, ranging in size from 91 to 961 patients, generally report improved speech outcomes with bilateral implantation, compared with unilateral implantation. In a retrospective case series of 73 children and adolescents who underwent sequential bilateral cochlear implantation with a long (>five year) interval between implants, performance on the second implanted side was worse than the primary implanted side, with outcomes significantly associated with the interimplant interval.[48, 52, 58-64]

Adults and Children

Ching (2006) subsequently reported on 29 children and 21 adults with unilateral cochlear implant and a contralateral hearing aid.[46] They noted that both children and adults localized sound better with bilateral inputs.

UNILATERAL HEARING LOSS WITH OR WITHOUT TINNITUS

The use of cochlear implants in patients with unilateral hearing loss is an off-label use of these devices. As noted in the 2011 AHRQ technology assessment, a number of narrative literature reviews[65-67] and small ($n \leq 30$) observational studies (described below) conducted primarily in adult patients have been published. However, these studies have included small numbers of patients ($n \leq 30$) and had risk of reporting bias.

Systematic Reviews

Oh (2022) reported on a systematic review and meta-analysis of cochlear implantation in adults with single-sided deafness.[68] A total of 50 studies with 674 patients (3 to 45 patients meeting inclusion criteria per study) were included. Of these, 41 were prospective cohort studies, seven were retrospective cohort studies, and two were case series. A meta-analysis of speech perception outcomes, which included five studies, found a standardized mean

difference (SMD) post- versus pre-implantation of 2.8 (95% CI 2.16 to 3.43), with some evidence of publication bias. A meta-analysis of QoL, which included eight studies, found a significant improvement, with an SMD of 0.68 (95% CI 0.45 to 0.91), and no evidence of publication bias. Meta-analysis of sound localization (seven studies; SMD, -1.13 [95% CI -1.68 to -0.57]), and tinnitus score reduction (seven studies; SMD -1.32 [95% CI -1.85 to -0.80]) also reported significant improvements. Limitations include the small sample sizes of included studies, imprecise definitions of single-sided deafness used across studies, and heterogeneity in outcomes measured, follow-up time frames, and etiology of single-sided deafness.

Assouly (2021) published a systematic review of cochlear implantation for tinnitus.[69] A total of seven prospective cohort studies, with 105 total subjects (range 10 to 26) met inclusion criteria. Two studies had a moderate risk of bias and five had serious risk of bias. Due to considerable methodological and statistical heterogeneity (I^2>75%), no meta-analysis was performed. Each included study reported a statistically significant improvement in tinnitus distress (measured via questionnaire). The only reported adverse event was worsening of tinnitus loudness following implantation in one participant.

Benchetrit (2021) published a systematic review and meta-analysis evaluating audiological and patient-reported outcomes in children <18 years with single-sided deafness (SSD).[70] 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 to 9.58).

Levy (2020) published a systematic review of cochlear implantation for tinnitus in SSD.[71] A total of 17 studies including 247 patients met inclusion criteria. The mean age was 50.2 years (range 23 to 71). Tinnitus outcomes were measured using the Tinnitus Handicap Inventory (THI). Based on six studies, an improvement of 35.4 points (95% CI -55.8 to -15.0, p<0.001) was reported. Based on 13 studies reporting on subjective improvement, with proportions weighted based on patients per study, 14.9% (CI 6.4 to 26.1) of patients reported complete resolution of tinnitus, 74.5% (CI 63.1 to 84.5) reported partial improvement; 7.6% (CI 4.1 to 12.6) of patients had no change in severity, and 3.0% (CI 1.0 to 6.7) reported worsening of their tinnitus.

A 2019 SR published by Peter identified 13 studies that met inclusion criteria and evaluated the influence of cochlear implantation on tinnitus in patients with single-sided deafness.[72] All identified studies were cohort studies. They mainly reported tinnitus questionnaire scores using the THI. Overall, of the 153 included patients, 34.2% demonstrated complete suppression, 53.7% demonstrated an improvement, 7.3% demonstrated a stable value, and 4.9% showed an increase of tinnitus. No patients reported an induction of tinnitus.

In 2015, van Zon published a systematic review of studies evaluating cochlear implantation for single-sided deafness or asymmetric hearing loss.[73] The authors reviewed 15 studies, nine of which (n=112 patients) were considered high enough quality to be included in data review. The authors identified no high-quality studies of cochlear implantation in this population. Data were not able to be pooled for meta-analysis due to high between-study heterogeneity, but the

authors conclude that studies generally report improvements in sound localization, quality of life scores, and tinnitus after cochlear implantation, with varying results for speech perception in noise.

In 2014, Vlastarakos published a systematic review of the evidence related to cochlear implantation for single-sided deafness.[74] The authors included 17 studies, including prospective and retrospective comparative studies, case series and case reports that included 108 patients. The authors report that sound localization is improved after cochlear implantation, although statistical analysis was not included in some of the relevant studies. In most patients (95%), unilateral tinnitus improved. The authors note that most of the studies included had short follow-up times, and evaluation protocols and outcome measurements were heterogeneous.

In 2014, Blasco and Redleaf published a systematic review and meta-analysis of studies evaluating cochlear implantation for unilateral sudden deafness.[75] The review included nine studies with a total of 36 patients. In pooled analysis, subjective improvement in tinnitus occurred in 96% of patients (of 27 assessed), subjective improvement in speech understanding occurred in 100% of patients (of 16 assessed), and subjective improvement in sound localization occurred in 87% of patients (of 16 assessed). However, the small number of patients in which each outcome was assessed limits any conclusions that may be drawn.

Randomized Trials

Marx (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.[76] Primary outcomes included HRQOL, auditory-specific quality of life, and tinnitus severity as assessed after six 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 overall EQ-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 six months, indicating no significant effect on binaural hearing within this timeframe.

Peters (2021) randomized 120 adults with single-sided deafness (median duration, 1.8 years) into three 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).[77] 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 three and six months compared to baseline. At three months follow-up, the cochlear implant group performed significantly better than all other groups. At six 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 five years is ongoing.

Nonrandomized Studies

Dillon (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.[78] 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 one month of implantation. At the 12-month interval, spatial hearing localization was significantly improved ($p<0.001$). Masked sentence recognition was found to improve at the 12-month interval in the SoNcontra configuration ($p<0.001$), but there was no significant difference in the SoNo or SoNci spatial configurations. Subjects demonstrated a significant improvement in CNC word recognition between one and six months ($p=0.002$) and 6 and 12 months ($p=0.010$). Findings were compared with previously published data for patients in the unilateral hearing loss cohort of this study.[79] 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 AHL.[78]

In 2019, Dillon 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.[80] Unaided hearing thresholds at 125 Hz were compared between the preoperative and initial activation intervals in 24 participants to assess the change in low-frequency hearing. At activation, a significant elevation in the unaided hearing thresholds at 125 Hz was noted ($p<0.001$), with the majority of subjects (n=16) demonstrating no response to stimulus. The remaining nine 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 EAS vs standard cochlear implants.

Galvin III (2019) reported data from on FDA-approved study of cochlear implantation in 10 patients with SSD.[81] 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 one, three, and six months postactivation. Performance was assessed with both ears (binaural), with the implanted ear alone, and the normal hearing alone. No patient had previous experience with a contralateral routing of signal (CROS) or bone conduction device (BCD) system. Mean improvement for consonant-nucleus-consonant (CNC) word recognition vs baseline was 66.8%, 76.0%, and 84.0% at one, three, and six months postactivation, respectively. The normal hearing ear performed significantly better compared to the implanted ear for all outcome measures at all intervals ($p<0.05$). Audiological performance of the implanted ear at one, three, and six months postactivation was significantly better compared to baseline ($p<0.05$), with no significant difference across postactivation intervals ($p>0.05$). The change in root mean square error (RMSE) in localization with binaural listening

postactivation reduced by 6.7, 7.6, and 11.5 degrees at one, three, and six months postactivation. Binaural performance was significantly improved compared to the normal hearing ear alone at all postactivation time intervals ($p<0.05$). Tinnitus visual analog scale (VAS) scores significantly decreased with the implant on at all postactivation time intervals ($p<0.05$). Significant improvements on SSQ scores were reported for the Speech ($p=0.003$), Spatial ($p<0.001$), and Quality ($p=0.034$) 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 (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.[82] A total of 10 patients were evaluated. Two years post-implantation, 90% of patients used their implant regularly for an average of more than 11 hours per day. Twelve months postactivation, speech from the front and noise at the healthy ear achieved a 2.7 dB improvement ($p=0.0029$). Speech to the implanted ear and noise from the front achieved a 1.5 dB improvement ($p=0.018$). The mean sound localization error of all participants was improved by 10.2 degrees ($p=0.030$) at 12 months postactivation. 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 decreased significantly 12 months postactivation from 41.2 points (SD 26.5) preoperatively to 23.0 points (SD 17.5; $p=0.004$) on the Tinnitus Handicap Inventory (THI). Quality of life measures showed a significant improvement on the global subscale of the WHO Quality of Life questionnaire ($p=0.007$). The Speech, Spatial, and Qualities of Hearing Scale questionnaire (SSQ) indicated a significant improvement from 4.2 to 6 ($p=0.004$) in speech comprehension and from 3 to 5.3 ($p=0.009$) in spatial hearing. No significant difference was noted in the subscale qualities of hearing (6.2 to 6.9; $p=0.13$). The scores of the patients on the three subscales were significantly lower than for the normal hearing control group, with an average speech comprehension score of 8.7 ($p=0.001$), an average spatial hearing of 8.6 ($p<0.001$), and an average qualities of hearing score of 9.1 ($p=0.005$). Adverse events were not reported.

In July 2019, the FDA approved to expand the indication for the MED-EL Cochlear Implant System to include individuals aged five years and older with single-sided deafness (SSD) or asymmetric hearing loss (AHL). According to the FDA's summary of safety and effectiveness data, 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 CROS 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, soundfield 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, preoperative, unaided/best-aided condition and the bilateral, 12-month post-operative cochlear implant (CI) + normal hearing (NH) or hearing aid (HA) condition. Study results are summarized in Table 1. 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.

Table 1. Feasibility Study Results for MED-EL Cochlear Implant System for SSD and AHL

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 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	NR

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; BKB-SIN: Bench-Kowal-Banford Speech in Noise Test; 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.

The FDA decision was further supported by a literature search yielding six publications comprising a total of 58 adults with SSD (n=50 of which implanted with MED-EL devices) and a total of 52 adults with AHL (n=37 of which implanted with MED-EL devices). The candidacy criterion of ages five and older was based on a literature search yielding five publications comprising a total of 26 children with SSD (n=5 of which implanted with a MED-EL device) and a total of nine children with AHL. While the overall benefits of CI 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 in methodology and outcome assessment, and at 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.

Buss (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.[79] 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 ≤ 80dB 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.

A 2016 study from Sladen reported on a retrospective review of prospectively-collected data of short-term (six-month) follow-up for 23 adults and children with single-sided deafness from a variety of mechanisms who received a cochlear implant.[85] In the implanted ear, CNC word recognition improved significantly from pre-implantation to three months post-activation (P=0.001). However, for AzBio sentence understanding in noise (+5 dB signal-to-noise [SNR]), there was no significant improvement from pre-implantation to six months post-activation.

Also in 2016, Rahne reported on a retrospective review of four children and 17 adults with single-sided deafness treated with cochlear implants and followed for 12 months.[86] Sound localization with aided hearing improved from pre-implantation to aided hearing for all individuals. The Speech recognition threshold in noise (signal-to-noise) ratio improved from -1.95 dB (CI off, SD: 2.7 dB) to -4.0 dB after three months (SD 1.3 dB, P<0.05), with continued improvements through six months.

In 2016, Mertens reported a case series including 23 individuals who received cochlear implants for single-sided deafness with tinnitus.[87] Eligible patients had either single-sided deafness or asymmetric hearing loss and ipsilateral tinnitus. Subjects had a mean eight years of experience with their cochlear implant (range, 3 to 10 years). Patients demonstrated improvements in VAS from baseline (mean score, 8) to one month (mean score: 4; p<0.01 vs baseline) and three months (mean score: 3; p<0.01 vs baseline) after the first fitting. Tinnitus scores improved from baseline to three months post fitting (55 vs 31, p<0.05) and were stable for the remainder of follow-up.

In 2015, Ramos Macias reported 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.[88] All patients had a severe tinnitus handicap (THI score ≥ 58%). Eight (62%) of the 13 patients who completed the six-month follow-up visit reported a lower tinnitus handicap on the THI score. Perceived loudness/annoyingness of the tinnitus was evaluated with a 10-point VAS. When the CI was on, tinnitus loudness decreased from 8.4 preoperatively to 2.6 at the six-month follow-up; 11 of 13 patients reported a change in score of three or more.

In 2015, Arndt reported outcomes for 20 children who underwent cochlear implantation for single-sided deafness, which represented a portion of their center’s cohort of 32 pediatric

patients with single-sided deafness who qualified for cochlear implants.[89] Repeated-measure analyses of hearing data sets were available for 13 implanted children, excluding five who had undergone surgery too recently to be evaluated and two children who were too young to be evaluated for binaural hearing benefit. There was variability in the change in localization ability across the tested children. Self- (or child-) reported hearing benefit was measured with the Speech, Spatial and Qualities of Hearing Scale (SSQ). Significant improvements were reported on the child and parent evaluations for the scale's three subcategories: speech hearing, spatial hearing, hearing quality, and total hearing.

In 2013, Hansen reported results of a prospective study of cochlear implantation for severe-to-profound single-sided sensorineural hearing loss in 29 patients, 10 of whom had single-sided deafness due to Meniere's disease.[90] Performance was compared pre- to post-implant within each subject; outcomes were measured at three-, six-, and 12-months postoperatively. Patients showed significant improvements in CNC word and AzBio sentence scores showed improvement in the implanted ear pre-and post-implant. For the 19 patients with pre- and post-operative data available, the average improvement on CNC word score was 28% (range: -26% to 64%). The average AzBio score improvement was 40% (range: -57% to 92%).

Tavora-Vieira (2013) reported results of a prospective case series that included nine 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.[91] Speech perception was improved for all subjects in the "cochlear implant on" state compared with the "cochlear implant off" state, and subjects with tinnitus generally reported improvement.

Section Summary

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. 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. However, studies assessing outcomes compared to best-aided hearing controls beyond six months are lacking.

COCHLEAR RESTORATION

The optimal timing of cochlear implantation in children is of particular interest given the strong associations between hearing and language development. While there is current research investigating the ability to restore hearing by stimulating cochlear hair cell regrowth, cochlear implantation damages the cochlea and eliminates the possibility of cochlear restoration. However, the potential to restore cochlear function is not foreseeable in the near future; therefore, if implantation of cochlear implants is felt to be most beneficial at a younger age when the nervous system is "plastic", this potential development seems too far in the future to benefit young children who are current candidates for a cochlear implant.

HYBRID COCHLEAR IMPLANTATION

Systematic Review

Santa Maria (2014) conducted a systematic review and meta-analysis of hearing outcomes after various types of hearing-preservation cochlear implantation, including implantation hybrid devices, cochlear implantation with surgical techniques designed to preserve hearing, and the use of post-operative systemic steroids.[92] The study included 24 studies, but only two studies

focused specifically on a hybrid cochlear implant system, and no specific benefit from a hybrid system was reported.

Nonrandomized Studies

The pivotal trial for the Med-EL EAS system was a prospective, multi-center, non-randomized, non-blinded, repeated measures clinical study of 73 subjects at 14 U.S. sites, implanted with either SONATA FLEX24 or a PULSAR FLEX24.[3] Final outcomes were reported in 2018 by Pillsbury.[93] Sixty-seven of 73 subjects (92%) completed outcome measures at 3, 6, and 12 months postactivation. A 30 dB or less low-frequency pure-tone average shift was experience by 79% and 97% were able to use the acoustic unit at 12 months postactivation. 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 subject performed similarly or improved (85%) on CNC words in quiet. Improvements in speech perception scores were statistically significant ($p<0.001$). The Abbreviated Profiled of Hearing Aid Benefit (APHAB) was administered preoperatively and at 12 months postactivation; 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 ($p<0.001$). 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%).

The pivotal trial for the Nucleus® Hybrid™ L24 Cochlear Implant System, published by Roland in 2016, was a prospective, multi-center, one-arm, non-randomized, non-blinded, repeated-measures clinical study of 50 subjects at 10 U.S. sites.[94] Performance was compared pre- to post-implant within each subject; outcomes were measured at three-, six-, and 12-months postoperatively. Post-operatively, patients' hearing was evaluated in three states: Hybrid (simultaneous electric and acoustic stimulation in the implanted ear via the Hybrid L24 including the acoustic component), Bimodal (electric stimulation only using the Hybrid L24 minus the acoustic component with contralateral acoustic stimulation), and Combined (electric and acoustic stimulation via the Hybrid L24 and contralateral acoustic stimulation). Results from the Bimodal and Combined conditions were grouped into an "Everyday Listening" category, which was not prospectively defined by the manufacturer. All 50 subjects enrolled underwent device implantation and activation. One subject had the device explanted and replaced with a standard cochlear implant between the three- and six- 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 additional subjects were explanted after 12 months. For the two primary effectiveness endpoints, CNC word-recognition score and AzBio sentence-in-noise score, a measure of sentence understanding in noisy environments, there were significant within-subject improvements from baseline to six-month follow up. The mean improvement in CNC word score was 35.7% (95% confidence interval [CI] 27.8% to 43.6%); for AzBio score, the mean improvement was 32.0% (95% CI 23.6% to 40.4%) For safety outcomes, 71 adverse events were reported, most commonly profound/total loss of hearing (occurring in 44% of subjects) with at least one adverse event occurring in 34 subjects (68%).

Five-year outcomes for the pivotal trial were reported by Roland in 2018.[95] Thirty-two out of 50 subjects (64%) enrolled in the postapproval study. Out of the 18 subjects who did not participate, six had been explanted and reimplanted 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 postapproval study. At

five years postactivation, 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 six months were statistically significant ($p<0.001$), but changes six months through five years postactivation were not statistically different ($p>0.05$). Acoustic component amplification was utilized by 84% and 81% of patients at 12 and three years postactivation, respectively. Mean CNC word recognition in quiet scores were significantly improved over the preoperative condition at each postactivation interval ($p<0.001$). However, mean scores did not significantly differ after 12 months postactivation. At five years postactivation, 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 one 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 postactivation intervals ($p<0.001$).

In 2016, Gantz published outcomes from a multicenter, longitudinal study evaluating outcomes with the Nucleus Hybrid S8 featuring a shorter cochlear array.[96] Eighty-seven subjects received an implant. At 12 months postactivation, five subjects had total hearing loss, whereas functional hearing was maintained by 80%. CNC word scores demonstrated 82.5% of subjects had experience 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 of 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 five of the six remaining subjects.

In 2015, Friedmann conducted a retrospective review that included 22 subjects implanted with a cochlear implant with either a standard electrode (n=12) or the Nucleus Hybrid L24 electrode (n=10).[97] At one year post-implant, 30% patients with the Hybrid-L and 58% patients with the standard electrode lost residual acoustic hearing resulting in a profound hearing loss in the implanted ear. The authors reported that while hearing preservation rates with the hybrid electrode tended to be better, among recipients who lost residual hearing, speech perception was better in those with the longer standard electrode.

Lenarz (2013) reported results of a prospective multi-center European study evaluating the Nucleus Hybrid™ L24 system.[98] The study enrolled 66 adults with bilateral severe-to-profound high frequency hearing loss. At one year post-operatively, 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.

Gifford (2013) compared hearing outcomes pre- and post-implantation for 44 adult cochlear implant recipients with preserved low-frequency hearing in two test conditions: cochlear implant plus low-frequency hearing in the contralateral plus low-frequency hearing in the contralateral ear (bimodal condition) and cochlear implant plus low-frequency hearing in both ears (best-aided condition).[99] The authors reported that there were small but statistically significant differences in improvements in adaptive sentence recognition and speech recognition in a noisy “restaurant” environment, suggesting that the presence of residual hearing is beneficial.

A small number of studies in a small number of patients suggest that a hybrid cochlear implant system is associated with improvements in hearing of speech in quiet and noise. However, there are currently no available studies that compare the use of a standard hearing aid with a hybrid cochlear implant, which would be an appropriate comparison to determine if a hybrid device improves outcomes for patients who currently have hearing loss, but might not be candidate for a cochlear implant. In addition, there is only limited data to suggest that the preservation of residual hearing associated with a hybrid device is associated with improved outcomes compared with a standard cochlear implant.

Section Summary

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

PRACTICE GUIDELINE SUMMARY

AMERICAN ACADEMY OF OTOLARYNGOLOGY- HEAD AND NECK SURGERY

In 2020, the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) published a revised position statement on cochlear implants. The Academy "considers unilateral and bilateral cochlear implantation as appropriate treatment for adults and children over 9 months of age with severe to profound hearing loss who have failed a trial with appropriately fit hearing aids."[100]

In 2020, the AAO-HNS published a position statement on pediatric cochlear implants.[101] The Academy states that "there is ample evidence that early cochlear implantation of children with sensorineural hearing loss (SNHL) for whom hearing aids provide inadequate access to sound is advantageous." The statement goes on to say that "Children with bilateral severe to profound SNHL (4-frequency PTA > 80 dB HL or 2-frequency PTA > 85) will not receive adequate benefit from amplification and are candidates for bilateral cochlear implantation. Children with this degree of SNHL, including infants between 6 and 12 months, should receive cochlear implants as soon as practicable."

AMERICAN ACADEMY OF AUDIOLOGY

In July 2019, the American Academy of Audiology published clinical practice guidelines on cochlear implants.[102] These guidelines include recommendations regarding cochlear implant evaluation. They recommend determining unaided air conduction and bone conduction thresholds using developmentally appropriate assessment measures. They additionally recommend determining auditory speech perception using appropriately fit amplification using developmentally appropriate assessment measures. Other recommendations are included regarding non-audiologic evaluation prior to implantation, and surgical and post-surgical roles for the audiologist.

SUMMARY

There is enough research to show that cochlear implants improve health outcomes, specifically, speech reception (especially in noise) and sound localization, for some patients who have severe to profound bilateral sensorineural hearing loss. Therefore, cochlear implants may be considered medically necessary in specific patients with bilateral hearing loss who meet the policy criteria.

The current research on cochlear implantation in patients diagnosed with enlarged vestibular aqueducts (EVA) has limitations. Despite these limitations, there is enough research to show that cochlear implants improve health outcomes, specifically, speech recognition, for patients for patients with EVA. In addition, early placement of cochlear implants avoids atrophy and preserves hearing patients with EVA with moderate hearing loss. Therefore, cochlear implants may be considered medically necessary in patients with EVA when policy criteria are met.

The current research on hybrid cochlear implant/hearing aid systems has limitations. Despite these limitations, there is enough research to show that hybrid cochlear implant/hearing aid systems improve health outcomes, specifically, speech recognition, for patients aged 18 years or older who have high frequency sensorineural hearing loss with preserved low frequency hearing. Therefore, hybrid cochlear implant/hearing aid systems may be considered medically necessary in specific patients with high frequency sensorineural hearing loss with preserved low frequency hearing who meet the policy criteria.

There are currently no cochlear implants that have approval from the U.S. Food and Drug Administration (FDA) for use in patients who are younger than 9 months of age. There is not enough research to show that cochlear implants improve health outcomes in patients younger than 9 months of age and it is unclear that the benefits of early cochlear implantation outweigh the risk of surgery and anesthesia in these very young patients. In addition, there are no clinical practice guidelines from U.S. professional societies that recommend cochlear implantation in these very young patients. Therefore, cochlear implantation in patients younger than 9 months of age is considered not medically necessary

In all other situations, cochlear implants and hybrid cochlear implant/hearing aid systems do not improve health outcomes. Therefore, cochlear implants and hybrid cochlear implant/hearing aid systems are considered not medically necessary when the policy criteria are not met, including but not limited to unilateral hearing loss with or without tinnitus.

Implant replacement, including replacement parts or upgrades to existing cochlear implants and/or components may be considered medically necessary only in those patients whose response to the existing device is inadequate to the point of interfering with activities of daily living, including school or work. Replacement of an existing cochlear implant device is considered not medically necessary when the policy criteria are not met.

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CODES

Codes	Number	Description
CPT	69930	Cochlear device implantation, with or without mastoidectomy
	92601	Diagnostic analysis of cochlear implant, patient younger than 7 years of age; with programming
	92602	;subsequent reprogramming
	92603	Diagnostic analysis of cochlear implant, age 7 years or older; with programming
	92604	;subsequent reprogramming
	92630	Auditory rehabilitation; pre-lingual hearing loss
	92633	Auditory rehabilitation; post-lingual hearing loss
HCPCS	L8614	Cochlear device, includes all internal and external components
	L8615	Headset/headpiece for use with cochlear implant device, replacement
	L8616	Microphone for use with cochlear implant device, replacement
	L8617	Transmitting coil for use with cochlear implant device, replacement
	L8618	Transmitter cable for use with cochlear implant device or auditory osseointegrated device, replacement

	L8619	Cochlear implant external speech processor and controller, integrated system, replacement
	L8621	Zinc air battery for use with cochlear implant device and auditory osseointegrated sound processors, replacement, each
	L8622	Alkaline battery for use with cochlear implant device, any size, replacement, each
	L8623	Lithium ion battery for use with cochlear implant device speech processor
	L8624	Lithium ion battery for use with cochlear implant or auditory osseointegrated device speech processor, ear level, replacement, each
	L8625	External recharging system for battery for use with cochlear implant or auditory osseointegrated device, replacement only, each
	L8627	Cochlear implant, external speech processor, component, replacement
	L8628	Cochlear implant, external controller component, replacement
	L8629	Transmitting coil and cable, integrated, for use with cochlear implant device, replacement

Date of Origin: *January 1996*