Clinical UM Guideline

Subject:	Cochlear Implants and Auditory Brainstem Implants		
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Description

This document addresses cochlear implants, auditory brainstem implants, and replacement or upgrade of speech processor and controller components. This document does not address replacement parts other than as specifically described below.

A single- or multi-channel unilateral or bilateral cochlear implant is intended to restore a level of auditory sensation to an individual with bilateral severe to profound sensorineural hearing loss by means of electrical stimulation of the auditory nerve. A unilateral hybrid cochlear implant is intended to restore a level of auditory sensation to an individual with residual low-frequency hearing sensitivity and bilateral severe to profound sensorineural hearing loss.

An auditory brainstem implant is a device designed to restore some hearing in an individual with neurofibromatosis type 2 (NF-2) who has been rendered deaf by the surgical removal of neurofibromas involving both auditory nerves.

Clinical Indications

I. Cochlear Implants

Medically Necessary:

A. Unilateral or bilateral implantation of a single- or multi-channel cochlear implant is considered **medically necessary** in an individual with bilateral severe-to-profound pre- or postlingual hearing loss (sensorineural deafness), defined as a hearing threshold of pure tone average of 70 decibels or greater, when **all** of the following criteria are met:
   1. The individual, including those with hearing loss due to meningitis, has obtained limited benefit from conventional hearing aids; **and**
   2. The individual is free from lesions in the auditory nerve and acoustic areas of the central auditory pathway (nervous system); **and**
   3. The individual is free from otitis media or other active middle ear infections; **and**
   4. The individual* is able to participate in a post-cochlear implant rehabilitation program in order to achieve benefit from the cochlear implant device.
      *Note: For a young child, a parent or guardian may act as the surrogate for participation.

B. Unilateral implantation of a single- or multi-channel cochlear implant is considered **medically necessary** for subsequent bilateral implantation (that is, sequential implantation) without retesting of hearing when the above criteria are met at the time of the initial (first) cochlear implantation.

C. Upgrade to or replacement of an existing external speech processor, controller or speech processor and controller (integrated system) is considered **medically necessary** for an individual whose response to existing components is inadequate to the point of interfering with the activities of daily living or when components are no longer functional.

D. Cochlear implantation with a hybrid cochlear implant device (for example, Nucleus® Hybrid™ L24 Cochlear Implant System) is considered **medically necessary** in an individual 18 years of age and older with bilateral severe-to-profound high-frequency sensorineural hearing loss with residual low-frequency hearing sensitivity (that is, able to hear low-frequency sounds) when **all** of the following preimplantation criteria are met:
   1. Limited benefit from appropriately fit bilateral hearing aids; **and**
   2. Normal to moderate hearing loss in the low-frequencies (that is, hearing thresholds no poorer than 60 decibels hearing level up to and including 500 hertz [averaged over 125, 250, and 500 hertz]) in the ear selected for implantation; **and**
   3. Severe to profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 hertz greater than or equal to 75 decibels hearing level) in the ear to be implanted; **and**
   4. Moderately severe-to-profound mid- to high-frequency hearing loss (threshold average of 2000, 3000, and 4000 hertz greater than or equal to 60 decibels hearing level) in the contralateral ear; **and**
   5. Preoperative speech perception scores as follows:
      a. Consonant-Nucleus-Consonant word recognition score from 10% to 60% in the ear to be implanted; **and**
      b. Consonant-Nucleus-Consonant word recognition score in the contralateral ear equal to or better than in the ear to be implanted, but not more than 80% in the best-aided condition; **and**
   6. Individual is free from lesions in the auditory nerve and acoustic areas of the central auditory pathway (nervous system); **and**
   7. Individual is free from otitis media or other active middle ear infections; **and**
   8. Individual is able to participate in a post-hybrid cochlear implant rehabilitation program in order to achieve benefit from the hybrid cochlear implant device.

Not Medically Necessary:

A. Upgrade to or replacement of an existing external speech processor, controller or speech processor and controller (integrated system) is considered **not medically necessary** when the criteria specified above are not met or when requested for convenience or to upgrade to a newer technology when the current components remain functional.

B. A cochlear implant is considered **not medically necessary** for all other indications when the above criteria are not met, including but not limited to treatment of **any** of the following:
   1. Auditory neuropathy spectrum disorders; **or**

2. Tinnitus in individuals who do not also have bilateral severe-to-profound hearing loss (sensorineural deafness); **or**
3. Unilateral deafness.

II. Auditory Brainstem Implants

Medically Necessary:

A. An auditory brainstem implant is considered **medically necessary** in an individual when **all** of the following criteria are met:
   1. Is 12 years of age or older; **and**
   2. Diagnosed with neurofibromatosis type 2; **and**
   3. Is completely deaf as a result of bilateral neurofibromas of the auditory nerve.

B. Upgrade to or replacement of an existing external sound processor, remote assistant or both components is considered **medically necessary** for an individual whose response to existing components is inadequate to the point of interfering with the activities of daily living or when components are no longer functional.

Not Medically Necessary:

A. Upgrade to or replacement of an existing external sound processor, remote assistant, or both components is considered **not medically necessary** when the criteria specified above are not met or when requested for convenience or to upgrade to a newer technology when the current components remain functional.

B. An auditory brainstem implant is considered **not medically necessary** for all other indications when the above criteria are not met.

Coding

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

Cochlear Implants
When services may be Medically Necessary when criteria are met:

CPT

69930	Cochlear device implantation, with or without mastoidectomy
69949	Unlisted procedure, inner ear [when specified as implantation of hybrid cochlear device]

HCPCS

L8614	Cochlear device, includes all internal and external components
L8619	Cochlear implant external speech processor and controller, integrated system, replacement
L8627	Cochlear implant, external speech processor, component, replacement
L8628	Cochlear implant, external controller component, replacement
L8699	Prosthetic implant, not otherwise specified [when specified as hybrid cochlear device, including all internal and external components]

ICD-10 Procedure

09HD05Z-09HD45Z	Insertion of single channel cochlear prosthesis into right inner ear [by approach; includes codes 09HD05Z, 09HD35Z, 09HD45Z]
09HD06Z-09HD46Z	Insertion of multiple channel cochlear prosthesis into right inner ear [by approach; includes codes 09HD06Z, 09HD36Z, 09HD46Z]
09HE05Z-09HE45Z	Insertion of single channel cochlear prosthesis into left inner ear [by approach; includes codes 09HE05Z, 09HE35Z, 09HE45Z]
09HE06Z-09HE46Z	Insertion of multiple channel cochlear prosthesis into left inner ear [by approach; includes codes 09HE06Z, 09HE36Z, 09HE46Z]

ICD-10 Diagnosis

H90.3	Sensorineural hearing loss, bilateral
H90.5	Unspecified sensorineural hearing loss
H90.6	Mixed conductive and sensorineural hearing loss, bilateral
H90.8	Mixed conductive and sensorineural hearing loss, unspecified
H91.93	Unspecified hearing loss, bilateral

When services are Not Medically Necessary:
For the procedure codes listed above when criteria are not met or for all other diagnoses not listed; or when the code describes a procedure or situation designated in the Clinical Indications section as not medically necessary.

Auditory Brainstem Implants
When services may be Medically Necessary when criteria are met:

HCPCS

L8699	Prosthetic implant, not otherwise specified [when describing replacement components of an auditory brain stem implant]
S2235	Implantation of auditory brain stem implant

ICD-10 Diagnosis

D33.3	Benign neoplasm of cranial nerves [when specified as neurofibromatosis type 2]
Q85.00	Neurofibromatosis, unspecified
Q85.02	Neurofibromatosis, type 2

When services are Not Medically Necessary:

For the procedure codes listed above when criteria are not met or for all other diagnoses not listed; or when the code describes a procedure or situation designated in the Clinical Indications section as not medically necessary.

Discussion/General Information

Single- or Multi-Channel Cochlear Implantation

Background Information and Description of the Technology

A single- or multi-channel 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 headset, an external sound processor, and an external transmitter/audio input selector. 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. The microphone picks up sounds that are carried to the external sound processor and transformed into coded signals that are then transmitted transcutaneously to the implanted internal receiver. The receiver converts the incoming signals to electrical impulses, which are conveyed to the electrode array, ultimately resulting in stimulation of the auditory nerve.

Once an individual is referred to a cochlear implant center, the implant team will perform additional testing to determine whether or not the individual is a suitable candidate for cochlear implantation. This testing includes audiologic testing, psychological testing, medical examination, and additional tests performed by the surgeon. An otolaryngologist examines the ear canal and middle ear to ensure that no active infection or other abnormality precludes the implant surgery; a physical examination identifies any potential problems with the use of general anesthesia needed for the implant procedure. The process often involves evaluation using radiographic tests such as computerized tomography scan or magnetic resonance imaging to evaluate the inner ear anatomy.

A speech-language pathologist and audiologist are involved in the cochlear implant evaluation process, performing extensive hearing tests that include pure tone and speech audiometric tests to determine how much the individual can hear with or without a hearing aid. An audiologist may perform objective measures of auditory function in infants and the youngest children as an alternative to standard testing methods. These testing methods define hearing loss is rated on a scale based on the threshold of hearing. Severe hearing loss is defined as a bilateral hearing threshold of 70 to 90 decibels, and profound hearing loss is defined as a bilateral hearing threshold of 90 decibels and above. Electrophysiological and objective measures have a valuable role in the management of individuals receiving cochlear implants; and in particular, young children, complex cases, and difficult-to-test persons. A challenge of performing these studies in children < age 1 year is the lack of available, effective tools for measuring speech perception abilities, including concern regarding the reliability of audiometric results for this age group. Behavioral audiometric testing, the standard for measuring hearing sensitivity, is performed in infants using visual reinforcement audiometry but is not appropriate for infants < age 5.5 months because they do not respond to sound with directed head turns (Holt and Svirsky, 2008). Therefore, audiologists use objective measures of auditory function as an alternative, including evoked otoacoustic emissions (OAE) testing, auditory brainstem response testing (ABR), and auditory steady-state response testing (ASSR) to assess various elements of the auditory system.

Electrically evoked auditory brainstem responses (EABR), middle latency responses (MLR), or acoustic reflexes (EART) may be used intraoperatively with stimuli delivered to the cochlear implant prior to leaving the operating room or postoperatively on an outpatient basis to facilitate the implant fitting process. These objective measures are also useful in a small subset of children who are unable to respond consistently to the electrical stimuli used to program the speech processor after implantation. The promontory/round window EABR (prom-EABR) stimulation test (or PST) may be indicated in a small subset of infants and children with conditions such as congenital malformations, cochlear nerve dysplasia or selected aplasia, or narrow internal auditory canal, to confirm the integrity of the auditory nerve (Nikolopoulos, 2000). For cochlear implantation performed in centers in the United States, the PST is no longer considered to be a prognostic factor, as the majority of the test results are recorded as "positive;" however, some centers continue to use the PST results during the implant evaluation process as an exclusion for candidates with complete absence of preoperative ability to stimulate the auditory nerve (Kuo, 2002).

One of the most commonly used speech recognition tests performed in the evaluation of adolescent and adult cochlear implant candidates is the Hearing In Noise Test (HINT), which measures an individual's ability to hear speech in quiet and in noise in the context of sentences. Psychological counseling of candidates is also performed. These tests are completed to ensure that the candidate will benefit from implantation and are motivated to participate in the process. It is important that the candidate understands what the implant will and will not do, and also understands the commitment required for care and follow-up services (National Institute on Deafness and other Communication Disorders [NIDCD], 2017).

Several FDA-approved cochlear implant systems are commercially available in the United States.

Cochlear Implant Manufacturer	Advanced Bionics® Corporation (Valencia, CA)	Cochlear Americas (Englewood, CO)	MED-EL Corporation, USA (Durham, NC)
Currently Marketed Cochlear Implant Systems	HiResolution® Bionic Ear System (HiRes 90K)	Cochlear Nucleus® System includes Nucleus 5 and 6 series of CI devices	Maestro (Sonata or Pulsar) Synchrony CI (FDA approved for 3.0T MRI without magnet removal)
Predecessor Cochlear Implants	Clarion Multi-Strategy or HiFocus CII Bionic Ear (P940022)	Nucleus 22, 24, Freedom with Contour (P840024)	Combi 40+ (P000025)

Subsequent generations of the various components of these devices have focused on improved electrode design and speech processing capabilities. Furthermore, smaller devices and the accumulating experience with use of cochlear implants in children have resulted in broadening of the candidate selection criteria to include children as young as 12 months of age. Specific criteria vary with the type of device. The FDA-labeled indications for each device are available on the FDA Premarket Approval (PMA) website (FDA, 2018).

While the benefits of unilateral cochlear implants are well accepted, the implantation of a single device does not provide normal (binaural) hearing to an individual with severe bilateral hearing loss. Binaural hearing provides certain auditory effects that assist in localizing sound and understanding speech in a noise environment. The auditory benefits enabled by binaural hearing include addressing "head shadow," "binaural summation," and "binaural squelch" (Gantz, 2002). Head shadow is the barrier the head creates between sounds emitted from one

direction and the contralateral ear. Head shadow dampens noise reaching the contralateral ear and delivers an intelligible signal to noise (SNR) or "speech-to-noise" ratio. Head shadow is believed to permit a bilateral cochlear implant user the flexibility of attenuating to the ear with the better SNR. This shadowing or attenuating effect works best for high frequency sounds.

With binaural hearing, each ear receives both unique and redundant information (acoustical representation) that is processed in the brain. The processing of this redundant information, "binaural summation," improves the hearing threshold and increases sensitivity to small differences in sound frequency and intensity. Binaural summation can lead to improved speech perception in both quiet and noise. The third effect of binaural hearing is "binaural squelch." With two ears, the brain uses cues to separate sounds coming from different locations. Optimal sound localization requires the ability to detect differences in time and amplitude between signals reaching both ears (Tyler, 2003).

Centers for Disease Control and Prevention (CDC) and Other Recommendations

Cochlear implant recipients should be up-to-date on age-appropriate pneumococcal conjugate and Hemophilus influenza type b conjugate vaccinations in accordance with current CDC and the Advisory Committee on Immunization Practices (ACIP) recommendations (CDC, 2018).

In 2010, the American Academy of Pediatrics (AAP) issued a policy statement on cochlear implants in children addressing surgical site infections and prevention and treatment of acute otitis media and meningitis. The policy statement includes the following recommendations:

> Children with profound deafness who are candidates for cochlear implantation should receive all age-appropriate doses of pneumococcal conjugate and Hemophilus influenzae type b conjugate vaccines and appropriate annual immunization against influenza. In addition, starting at 24 months of age, a single dose of 23-valent pneumococcal polysaccharide vaccine should be administered. Before implant surgery, primary care providers and cochlear implant teams should ensure that immunizations are up-to-date, preferably with completion of indicated vaccines at least 2 weeks before implant surgery. Imaging of the temporal bone/inner ear should be performed before cochlear implantation in all children with congenital deafness and all patients with profound hearing impairment and a history of bacterial meningitis to identify those with inner-ear malformations/cerebrospinal fluid fistulas or ossification of the cochlea (Rubin, 2010).

Efficacy and Safety of Single or Multi-Channel Cochlear Implantation

Cochlear implants are recognized as an effective treatment of sensorineural deafness. While use of a unilateral cochlear implant in an individual with severe to profound hearing loss has become a standard clinical practice, bilateral implantation is less common. Evolution of cochlear implant devices has focused on minimizing the internally implanted electrodes, such that one device, the Nucleus® 24 (Cochlear Americas, Englewood, CO), received FDA approval (2000) for use in children 12 months of age and older. A review of the early peer-reviewed literature includes several reports on individuals with bilateral cochlear implants (Long, 2003; Muller, 2002; Schoen, 2005; Tyler, 2002; van Hoesel, 2002; van Hoesel, 2003). These early reports evaluated small numbers of

individuals and provided limited outcome information. In these reports, most, but not all, individuals reported very slight to modest improvements in sound localization and speech intelligibility with bilateral cochlear implants, especially with noisy backgrounds, but not necessarily in quiet environments. When reported, the combined use of binaural stimulation improved hearing by only a few decibels or percentage points. This improvement appeared marginal, and 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 (Morera, 2005).

Subsequently, a number of prospective case series designed to assess whether bilateral cochlear implants could provide some of the benefits of binaural hearing were published in the peer-reviewed literature. These studies assessed the benefits of bilateral cochlear implants on sound localization and speech perception in both adults (Gantz, 2002; Laszig, 2004; Litovsky, 2004; Schoen, 2005; Verschuur, 2005), and, to a lesser extent, in children (Kuhn-Inacker, 2004; Litovsky, 2006a). The largest and most complete of these early case studies included a case series of 30 U.S. children with sequentially placed bilateral cochlear implants (Peters, 2007).

Lammers and colleagues (2014) summarized the evidence on the effectiveness of bilateral cochlear implantation compared with unilateral implantation among children with sensorineural hearing loss. A total of 21 studies evaluated bilateral implantation in children, with no randomized controlled trials identified. The authors were unable to perform pooled statistical analysis due to a limited number of studies, heterogeneity in outcomes and comparison groups and a high risk for bias in the studies. A "best-evidence synthesis" was performed, with results "…indicating the positive effect of the second implant for especially sound localization and possibly for preverbal communication and language development." One study demonstrated improvements in language development, although other studies found no significant improvements. The currently available evidence consists solely of cohort studies that compare a bilaterally implanted group with a unilaterally implanted control group, with only one study providing a clear description of matching techniques to reduce bias.

Cochlear Implantation in Adults with Bilateral Severe-to-Profound Hearing Loss

Over the past several decades, many studies have investigated the use of bilateral cochlear implantation in adults with severe postlingual hearing loss (Gantz 2002, Litovsky 2004, Laszig 2004, Schoen 2005, Verschuur 2005, Litovsky 2006b). When viewed together, these small retrospective case studies show that bilateral cochlear implantation can improve sound localization in most adults and may improve speech perception in others when compared with unilateral implantation.

Kraaijenga and colleagues (2017) conducted a multicenter, randomized clinical trial at five tertiary referral centers evaluating objective and subjective measures of simultaneous versus sequential bilateral cochlear implantation in 40 adults with postlingual severe to profound hearing loss and a maximum duration of 10 years without hearing aid use in both ears. The simultaneous bilaterally implanted group received two cochlear implants during one surgical procedure. The sequential bilaterally implanted group received two cochlear implants with an interval of 2 years between implants. Results 1 year after receiving simultaneous bilateral cochlear implants were compared with the results 1 year after receiving sequential bilateral cochlear implants, and results of 3 years of follow-up for both groups were compared separately. The primary outcome measure was speech intelligibility in noise from straight ahead. Secondary outcome measures included speech intelligibility in noise from spatially separated sources,

speech intelligibility in silence, localization capabilities, and self-reported benefits assessed with various hearing and quality of life questionnaires. Nineteen participants were randomized to receive simultaneous bilateral cochlear implants (11 women and 8 men; median age, 52 years [interquartile range, 36-63 years]), and another 19 participants were randomized to undergo sequential bilateral cochlear implants (8 women and 11 men; median age, 54 years [interquartile range, 43-64 years]). Three participants did not receive a second cochlear implant and were unavailable for follow-up. Comparable results were found 1 year after simultaneous or sequential bilateral cochlear implants for speech intelligibility in noise from straight ahead (difference, 0.9 dB; 95% confidence interval [CI], -3.1 to 4.4 dB) and all secondary outcome measures except for localization with a 30° angle between loudspeakers (difference, -10%; 95% CI, -20.1% to 0.0%). In the sequential bilateral cochlear implant group, all participants performed significantly better after the bilateral cochlear implants on speech intelligibility in noise from spatially separated sources and on all localization tests, which was consistent with most of the participants' self-reported hearing capabilities. Speech intelligibility-in-noise results improved in the simultaneous bilateral cochlear implant group up to 3 years following implantation. This study shows comparable objective and subjective hearing results 1 year after receiving simultaneous bilateral cochlear implants and sequential bilateral cochlear implants with an interval of 2 years between implants. Also reported was a significant benefit of sequential bilateral cochlear implants over a unilateral cochlear implant.

A technology assessment on the effectiveness of cochlear implants in adults completed by the Tufts Evidence-based Practice Center for the Agency for Health Care Research and Quality (AHRQ, 2011) examined 22 studies with 30 or more participants, concluding 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. The available evidence was insufficient to draw conclusions on improvements in open-set sentence test scores and any relationship between pre-implantation characteristics of individuals and outcomes (for example, age, duration of hearing impairment, HINT scores, and pre- or post-linguistic deafness). Gaylor and colleagues (2013) published an update to the 2011 AHRQ technology assessment evaluating the communication-related outcomes and health-related quality of life outcomes after unilateral or bilateral cochlear implantation in adults with sensorineural hearing loss. A total of 16 of 42 studies published through May 2012 evaluated use of unilateral cochlear implants. Most unilateral implant studies showed a statistically significant improvement in mean speech scores as measured by open-set sentence or multisyllable word tests; meta-analysis of 4 studies revealed a significant improvement in cochlear-implant relevant quality of life after unilateral implantation (standard mean difference: 1.71; 95% CI, 1.15 to 2.27). However, these studies varied in design, and there was considerable heterogeneity observed across studies.

In summary, studies of cochlear implants in adults with severe-to-profound hearing loss who only receive limited or no benefit from amplification with conventional hearing aids show consistent clinical effectiveness in speech reception (especially in noise) and in sound localization with bilateral devices.

Cochlear Implantation in Unilateral Deafness (with or without Tinnitus)

The evidence in the peer-reviewed literature is insufficient to permit conclusions concerning the effect of cochlear implantation for the treatment of unilateral hearing loss (single-sided deafness), including cochlear implants for

tinnitus relief in individuals with unilateral deafness. The available evidence consists of observational studies and case series with small sample sizes and heterogeneity in evaluation protocols and outcome measurements (Arndt, 2017; Arts, 2012; Dillon, 2017a; Dillon, 2017b; Mertens, 2017; van de Heyning, 2008).

Blasco and Redleaf (2014) published a systematic review and meta-analysis of studies evaluating cochlear implantation for improvement of tinnitus and speech comprehension in unilateral sudden deafness. In the pooled analysis of nine studies with 36 participants, subjective improvement in tinnitus occurred in 96% of 27 participants assessed, subjective improvement in speech understanding occurred in 100% of 16 participants assessed, and subjective improvement in sound localization occurred in 87% of 16 participants assessed. The meta-analyses demonstrated a high-degree of inter-study heterogeneity among a small study population. The authors recommend additional study with follow-up of longer duration (> 24 months) to compare symptoms and performance between unilateral cochlear implantation and more traditional strategies for hearing rehabilitation.

Van Zon and colleagues (2015) published a systematic review of studies evaluating cochlear implantation for single-sided deafness or asymmetric hearing loss. A total of nine studies (n=112 subjects) were considered in the data review, but included no high-quality studies of cochlear implantation in this population. The authors were unable to pool the data for meta-analysis due to high between-study heterogeneity in participants (classification of hearing loss, duration of deafness, and the indication for cochlear implantation [hearing loss vs. tinnitus]), test conditions (pre-implant vs. post-implant), follow-up duration, and outcome measurements (that is, different test configurations, word tests, and/or questionnaires). The authors concluded the "current literature suggests important benefits of cochlear implantation in this population regarding sound localization, quality of life, and tinnitus. Although results for speech perception in noise are promising as well, varying results between studies were reported for this outcome."

Sladen and colleagues (2016) conducted a prospective repeated-measures study examining speech recognition and self-perceived health-related quality of life in a cohort of 20 adults and children who received a cochlear implant for unilateral hearing loss (> 6 months, but < 2 years). The etiology of the hearing loss was attributed to idiopathic sudden sensorineural hearing loss (n=15), otosclerosis (n=2), vestibular schwannoma (n=1), cholesteatoma (n=1), or bacterial sepsis (n=1). A total of 15 (75%) participants reached the 6-month post-activation point and were included in the final data analysis. A significant improvement in speech recognition in both quiet and noise was reported at the 6-month post-activation point. Consonant-Nucleus-Consonant (CNC) scores in quiet improved from 4.8% in the preoperative period to 42.3% a 6-months post-activation. However, limitations of this study exist, including the small sample size, comprised of predominantly younger adults with a short duration of reported hearing loss prior to implantation, and the short follow-up period.

Sladen and colleagues (2017) retrospectively reviewed data (prospectively collected) of short-term outcomes for 23 adults and children with single-sided deafness from multiple etiologies (most frequently, sudden sensorineural hearing loss [n=12] and congenital conditions [n=3]) who received a cochlear implant. Participants had mild to severe sensorineural hearing loss with ≤ 40% CNC word recognition on the affected side and normal hearing in the nonimplanted ear. CNC word recognition improved significantly from pre-implantation to 3 months post-activation (p=0.001), but not between the 3-month and 6-month intervals in 13 of the 20 evaluable participants. There was no significant improvement in 8 participants from pre-implantation to 6 months post-activation for AzBio sentence understanding in noise (+ 5 decibels signal-to-noise). Twelve of the 13 participants who reported tinnitus prior to

surgery reported subjective improvement after implantation. Limitations of this study include the small sample size with retrospectively collected data from a variable number of participants at the 3- and 6-month intervals; additionally, study outcomes were evaluated using separate and unique testing protocols at the two participating treatment centers. Finally, the heterogeneous study population comprised of children and adults with various hearing loss etiologies and duration of deafness limits drawing conclusions as to the net health benefit of cochlear implantation in individuals with unilateral sensorineural hearing loss.

In summary, the evidence for cochlear implantation for unilateral deafness, with or without tinnitus, consists of non-randomized, retrospective studies and case series reporting varying results and a high-degree of interstudy heterogeneity among small study populations. Additional study is needed measuring long-term outcomes, in order to establish the clinical utility of cochlear implantation as a treatment for unilateral deafness with or without tinnitus.

Cochlear Implantation in Children 12 Months of Age and Older

The early peer-reviewed literature concerning the use of bilateral cochlear implants in children is limited. Kuhn-Inacker and colleagues (2004) observed significant improvement in speech discrimination in noise with bilateral implants compared to unilateral implants. Litovsky and colleagues (2006a) reported that 9 of 13 (70%) children with bilateral cochlear implants discriminated source separations of $\leq 20^{o}$ and 7 of 9 children performed better when using bilateral (vs. unilateral) devices. Sharma and Dorman (2006) studied congenitally deaf children to establish the existence and time limits of a sensitive period for the development of central auditory pathways in humans. The authors reported that central auditory pathways are "maximally plastic" for a period of about 3½ years in children with cochlear implants. Stimulation delivered within this timeframe results in auditory evoked potentials that reach normal values in 3 to 6 months. However, when stimulation occurs after 7 years, changes occur within 1 month, but then have little to no subsequent change.

Peters and colleagues (2007) reported experience with 30 children between 3 and 13 years old who were bilaterally implanted with sequential surgeries at least 6 months apart. All children received their first cochlear implant prior to 5 years of age. Children acquired speech perception in the second ear within 6 months. However, children under 8 years of age acquired speech perception in the second ear more rapidly and ultimately gained a higher level of speech understanding than older children. The authors reported that sequentially implanted children in all age groups showed better mean speech perception scores in background noise with bilateral implants than with a single implant. Speech performance in quiet was improved to a lesser degree, but this difference did not meet a level of statistical significance. In younger children, speech perception scores improved for 12 months following the second implant while scores for older children plateaued at 6 months. The rate of improvement in speech perception scores in the second ear was inversely related to the child's age at the time of the second implant.

Other studies have reported benefits for children (and adults) with a unilateral cochlear implant with hearing aid in the opposite ear. Ching and colleagues (2006) reported on 29 children (and 21 adults) with unilateral cochlear implants and a contralateral hearing aid. The authors noted that both children and adults derived binaural advantages related to sentence perception in noise and localized sound that was better with bilateral cochlear implants. In another report, Holt and colleagues (2005) concluded that children with severe hearing loss who used a

unilateral cochlear implant benefited from wearing an appropriately-fitted hearing aid in the nonimplanted ear to maximally benefit from bilateral stimulation.

Cochlear Implantation in Infants Younger than 12 Months of Age

On November 10, 2020 the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) revised its position statement on cochlear implants to, "Consider unilateral and bilateral cochlear implantation as appropriate treatment for adults and children over 9 months of age with moderate to profound hearing loss who have failed a trial with appropriately fit hearing aids." In April 2020, the AAO-HNS issued a position statement on pediatric cochlear implants stating:

> Cochlear implantation should occur as soon as practicable, including in infants between six and 12 months of age. The Academy states that implantation below 12 months of age is associated with better language outcome and as such, implantation should not be delayed by a hearing aid trial.

Other studies and developments in clinical practice have occurred with the advent of universal neonatal hearing screening in some countries and the availability of screening programs for at-risk infants in other countries, including earlier referral, diagnosis, and intervention for infants with hearing loss. Improvements in device technology, over 2 decades of pediatric clinical experience, and a growing recognition of the efficacy of cochlear implants have led to increasing numbers of young children receiving cochlear implants. There are multiple small retrospective studies and small and large case series which suggest improved health outcomes in several objective measures in children implanted before 12 months of age. There is some data to suggest that earlier implantation leads to improved language acquisition during this critical period of development of spoken and aural language skills; it has been shown that the age at the time of auditory stimulation is a strong predictor of auditory and language outcomes (Harrison, 2005; Miyamoto, 2005; Nicholas, 2006; Sharma, 2005; Tomblin, 2005). Cumulatively, these reports and an increase in recent evidence suggest that cochlear implantation can be performed safely and successfully without serious complications in select children younger than 12 months of age with profound bilateral sensorineural deafness (Colletti, 2005b; Colletti, 2009; Dettman, 2007; Holman, 2013; Holt and Svirsky, 2008; Loundon, 2010; Roland, 2009; Tait, 2007; Valencia, 2008; Vlastarakos, 2010b; Waltzman and Roland, 2005).

In addition to information about children with congenital deafness, there are several small prospective and retrospective case reviews and a matched-pair analysis suggesting that children with profound bilateral sensorineural deafness secondary to bacterial meningitis may benefit from early cochlear implantation (El-Kashlan, 2003; Hehar, 2002; Johnson, 1995; Young, 2000). Philippon and colleagues (2010) performed a descriptive analysis of data, including the cause of meningitis, preoperative imaging evaluation, age at implantation, time lapse between meningitis and implantation, and relevant surgical findings, in an attempt to propose guidelines for the management of profound bilateral sensorineural hearing loss after bacterial meningitis. The mean age at implantation was 3 years 8 months; the mean time delay between meningitis and surgery was 2 years 1 month for the children. A total of 18 children (67%) were implanted within a year. Labyrinthitis ossificans was evidenced at the time of surgery in 62% of the total study subjects (children and adults). Open-set speech discrimination was achieved by 37% of the children (10 of 27). The authors recommended early cochlear implantation for individuals

with bilateral profound deafness secondary to bacterial meningitis as an interventionist approach to avoid complications presented by labyrinthitis ossificans and to optimize hearing outcomes.

Vlastarakos and colleagues (2010a) conducted a meta-analysis of prospective controlled studies, prospective and retrospective cohort studies, clinical guidelines, and review articles reporting the diagnostic challenges and safety considerations in cochlear implantation in 125 children younger than 12 months of age. Overall, no major anesthetic complication was reported; the rate of surgical complications was reported at 8.8% (3.2% major complications), similar to the respective complication rates in older implanted children (major complications ranging from 2.3 to 4.1%). The authors summarized that cochlear implantation can be performed in otherwise healthy infants, provided that the attending pediatric anesthesiologist is considerably experienced and appropriate facilities of pediatric perioperative care are readily available. A number of concerns, with regard to anatomic constraints, existing co-morbidities or additional disorders, tuning difficulties, and special phases of the developing child should be also taken into account. The meta-analysis did not find an increased rate of anesthetic or surgical complications in implanted infants, although it identified a lack of studies with long-term follow-up reporting improved health outcomes.

Colletti and colleagues (2011) reported on the 10-year results of 19 children who received cochlear implants between the ages of 2 to 11 months, comparing them to 21 children implanted between 12-23 months and 33 children implanted between 24-35 months. Within the first 6 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.

In summary, given the strong evidence of benefit in children with profound hearing loss when cochlear implantation is performed after the age of 12 months, the additional evidence suggesting further benefit at ages under 12 months, and the lack of evidence that performing implantation at an earlier age is harmful, it is reasonable to perform cochlear implantation in children under 12 months of age.

Cochlear Implantation in Special Populations or Clinical Conditions

Several systematic reviews have evaluated outcomes after cochlear implantation for specific causes of deafness and in subgroups of the pediatric population. Eze and colleagues (2013) published a systematic review comparing outcomes of cochlear implantation for children with developmental disability with those without developmental disability. The authors stated that while approximately 30% to 40% of children who receive cochlear implants have developmental disability, evidence about outcomes in this group is limited. A total of 13 studies met the inclusion criteria that compared receptive or expressive language outcomes in children with cochlear implants with and without developmental disability. A meta-analysis of pooled data was not performed as the studies were heterogeneous in terms of comparator groups and outcome measures. Seven of the eligible studies in the structured systematic review demonstrated a significantly poor cochlear implant outcome in children with developmental disability, while the remaining 6 studies reported no significant difference in outcomes between the groups.

Humphriss and colleagues (2013) published a systematic review evaluating outcomes after cochlear implantation in a pediatric population 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. A total of 27 observational studies were selected for review, including case studies, cohort studies, and comparison studies between children with ANSD and severe sensorineural hearing loss. Eleven of the 15 noncomparative studies 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 children with ANSD; a single small study that used this design found no significant differences between the groups. The quality of the available evidence was limited by methodological concerns, including risk of bias and confounding.

Fernandes and colleagues (2015) evaluated 18 published studies and 2 dissertations that reported hearing performance outcomes for children with ANSD and cochlear implants. Five studies were nonrandomized controlled trials considered high quality, 5 randomized controlled trials considered low quality, and 10 were clinical outcome studies. A total of 14 studies compared speech perception in children with ANSD and cochlear implants with speech perception in children with sensorineural hearing loss and cochlear implants. Most of the studies (n=13) concluded that children with ANSD and cochlear implants developed hearing skills similar to children with sensorineural hearing loss and cochlear implants; however, these types of studies do not allow comparisons of outcomes between children with ANSD treated with cochlear implants and those treated with usual care.

In a subsequent small retrospective case series, Kontorinis and colleagues (2014) retrospectively evaluated pre- and post-cochlear implantation hearing outcomes for 27 implanted children with ANSD. Cognitive disorders were found in 4 subjects: 3 children were diagnosed with autism and another child with dyspraxia. Another child had severe additional co-morbidities (hemiplegia). One of the autistic children became a non-user of her cochlear implant. The average age at implantation was 35.4 months (range, 19-68 months) with follow-up for an average of 63.1 months (range, 6-140 months). Nine children were implanted bilaterally and 18 unilaterally. The mean preoperative categories of auditory performance (CAP) and Manchester spoken language development scale (MSLDS) scores with hearing aids were 2.5 (range, 0-5) and 2.5 (range, 0-6), respectively. The mean post-cochlear implant CAP and MSLDS scores were 5.8 (range, 2-9) and 7.7 (range 3-10), respectively. The difference between the preoperative and post-cochlear implant scores was statistically significant for both CAP ($p<0.001$) and MSLDS ($p<0.001$). In 3 children the post-cochlear implant outcomes were fluctuating. The authors identified cognitive disorders as a significantly negative prognostic factor for cochlear implant outcome. Limitations of this study include the retrospective design and lack of information on how many children would have otherwise qualified for a cochlear implant without ANSD.

In summary, additional well designed studies are needed to draw any conclusions around the effectiveness of cochlear implantation in improving speech recognition in children with ANSD.

Hybrid Cochlear Implantation

Background Information and Description of the Technology

On March 20, 2014, the FDA granted premarket approval (PMA) status to the Nucleus Hybrid™ L24 Cochlear Implant System (also referred to as "Hybrid L24") (Cochlear Americas, Englewood, CO) as a unilateral cochlear implant system for individuals with residual low frequency hearing sensitivity who have obtained limited benefit from bilateral conventional hearing aids. The Hybrid L24 is an electric-acoustic stimulation (EAS) cochlear implant

system that consists of both internal and external components. The Hybrid L24 is designed to allow individuals to hear in two ways: electrically (similar to approved cochlear implants) for severe to profound hearing loss at mid and high frequencies, and acoustically (similar to hearing aids) for normal to moderate hearing loss at low frequencies. The Hybrid L24 includes an implant consisting of a receiver/stimulator and an intracochlear electrode array using short implant electrodes placed in the cochlea through a small cochleostomy or round window insertion to preserve low-frequency hearing. An externally worn sound processor can be fit with an acoustic component, programming software/instruments, and various remote control options. Using a small microphone, the sound processor picks up sound from the person's surroundings and separates it into different groups of sounds by frequency (that is, the low or high "pitch" of a sound). The higher frequency information about the sound is sent to the receiver/stimulator and electrode array of the implanted part of the device. Since the electrode array is located inside the person's cochlea, this sound-related information is relayed to the brain, allowing the person to hear. For person's who have enough of their own acoustic low-frequency hearing after implantation, the sound processor also provides amplified low-frequency sound to the ear through the acoustic component. After implantation, some persons do not have enough low-frequency hearing to use the acoustic component so these individuals hear only electrically using the implant for all (lower and higher) sound frequencies (FDA, 2014).

The Hybrid L24 implant is intended for use in one ear by persons aged 18 years or older who obtain limited benefit from appropriately fitted conventional hearing aids in both ears and meet the following criteria (FDA, 2014):

- Typical preoperative hearing of candidates ranges from normal to moderate hearing loss in the low frequencies, with severe to profound loss in the mid to high frequencies; and
- The CNC word recognition score will be between 10% and 60%, inclusively, in the ear to be implanted in the preoperative aided condition and in the other ear will be equal to or better than that of the ear to be implanted but not more than 80% correct; and
- Previously have undergone a suitable hearing aid trial, unless already appropriately fit with conventional hearing aids.

The Hybrid L24 should not be used for individuals who have any of the following conditions:

- Deafness due to lesions of the acoustic nerve or central auditory pathway; or
- Active middle ear disease, with or without tympanic membrane perforation; or
- Absence of cochlear development; or
- Duration of severe to profound hearing loss of 30 years or greater.

Other hybrid hearing devices have been developed but do not have FDA 510(k) clearance or premarket approval for use in the U.S., including the Synchrony Electric Acoustic Stimulation (EAS) Hearing Implant System (MED-EL, USA, Durham, NC). The hybrid implant's manufacturer states it is designed for use in individuals with "partial deafness," that is, mild to moderate low-frequency hearing loss combined with profound hearing loss in the higher frequencies.

Efficacy and Safety of Hybrid Cochlear Implantation

The FDA approval of the Hybrid L24 was based on a multicenter, nonrandomized, unblinded single-subject study where participants served as their own control (so that post-implant performance was compared to each participant's baseline or pre-implant performance) (Roland, 2016). According to the FDA PMA (P130016), criteria for study inclusion required that candidates be aged 18 years and older with preoperative hearing ranges from normal to moderate hearing loss in the low frequencies (thresholds no poorer than 60 decibels HL up to and including 500 hertz), with severe to profound mid-to-high frequency hearing loss (threshold average of 2000, 3000, and 4000 hertz ≥ 75 decibels HL) in the ear to be implanted, and moderately severe-to-profound mid-to-high frequency hearing loss (threshold average of 2000, 3000, and 4000 hertz ≥ 60 decibels HL) in the contralateral ear. A CNC word recognition test score was required to be between 10% and 60%, inclusively, in the ear to be implanted in the preoperative aided condition and in the contralateral ear, equal to or better than that of the ear to be implanted, but not more than 80% correct. Individuals with severe to profound high-frequency hearing loss for > 30 year's duration or congenital hearing loss (onset prior to 2 years of age) were excluded from participation. Prospective candidates were required to go through a suitable 2 week hearing aid trial unless already appropriately fitted with conventional hearing aids.

The study was conducted at 10 United States sites, enrolled 50 participants (age at implantation, range: 23 to 86.2 years), and involved up to 9 visits before and after implantation for approximately a 1-year period. The Hybrid L24 was implanted in one ear and activated following a healing period of 2 to 4 weeks. Postoperative measurements included verification of hearing device functioning, unaided hearing thresholds and tympanometry, aided audiometric thresholds, aided CNC test in quiet, aided AzBio sentences-in-noise test, and adverse event reporting at 3-, 6-, and 12-month intervals. Co-primary effectiveness endpoints included CNC word-recognition scores and AzBio sentence-in-noise scores compared across 2 conditions: the baseline "Acoustic Alone" condition and the 6-month "postactivation Hybrid" condition. Success was defined as statistically significant improvement in both co-primary endpoint measures. A total of 49 of the 50 enrolled participants (98%) completed all effectiveness outcome assessments at the 6-month interval, while 48 participants completed the audiometric testing for hearing sensitivity; 46 subjects were evaluable at the 12-month interval. Based on data from 50 participants (including worst-case imputed scores for data missing for 1 participant), both co-primary effectiveness endpoints were met. Statistically significant improvements in mean CNC word score (35.7%: 27.8% to 43.6%; 95% CI, $p<0.0001$) and mean AzBio sentence-in-noise score (32.0%: 23.6% to 40.4%; 95% CI, $p<0.0001$) occurred from the baseline (Acoustic alone, hearing-aided) to the 6-month interval postactivation (Hybrid condition), respectively. Secondary effectiveness endpoints compared 6-month postoperative performance in the Hybrid condition to preoperative (ipsilateral) Acoustic Alone performance. All secondary endpoints were met as more than 75% of the participants performed similar to or better on each of the 3 specified measures: CNC words (96%), CNC phonemes (96%), and AzBio sentences (88%).

The primary safety endpoint was the number and proportion of participants experiencing an adverse event, defined as any surgical and/or device events. A total of 71 adverse events were reported to have occurred during the study. Of the 50 implanted participants, 34 (68%) experienced at least 1 adverse event, with multiple (2 to 4) adverse events experienced by 20 to 50 subjects; 24 of 71 adverse events in 23 participants were unresolved during the study. The two most frequently observed adverse events that were reported as resolved included tinnitus-related issues and device-related open shorts experienced by 28% and 22% of participants, respectively. By May 31, 2013, 30 of 50 (60%) participants exhibited more than a 30 decibels loss in their residual low-frequency hearing. Unresolved adverse events observed included profound/total loss of residual low-frequency hearing occurring in 22

of 50 (44%) participants. As of February 10, 2014, 6 of 50 (12%) participants were subsequently explanted and reimplanted with a standard cochlear implant. Based on these results, the FDA stated "It is yet to be determined over the long-term how many additional subjects who experience profound loss will be explanted and re-implanted with a traditional cochlear implant array."

Despite an improvement in speech recognition in terms of CNC words and AzBio sentences reported for the majority of the study population, the FDA concluded the potential risks of profound and possibly total loss of low-frequency hearing that occurred in 44% of the participants:

> …is a known risk and renders the device usage to electrical (cochlear implant) stimulation only since the acoustic amplification is ineffective for these levels of hearing loss. For subjects who lost low-frequency residual hearing to the profound/total level(s), the device showed benefit for only about half (9 of 17) or 53% of these subjects. Furthermore, 6/50 subjects who lost residual low-frequency hearing chose to undergo explantation of the Hybrid L24 and be reimplanted with an approved standard cochlear implant.

Considering the risks and benefits of the device, the FDA approved the hybrid implant only for unilateral use after a sufficient trial of conventional hearing aids, with an expiration date at 1 year. Continued approval of the PMA is contingent upon submission of periodic reports, which include reporting of device distribution in order to determine the frequency and prevalence of adverse events.

In 2016, Roland and colleagues published the results of the multicenter clinical trial presented to the FDA (2014) as part of the Nucleus Hybrid L24 implant system PMA review process. The study inclusion criteria (discussed previously in this document), evaluated 50 individuals aged 18 years or older with low-frequency hearing and severe-to-profound high-frequency sensorineural hearing loss who were implanted with the hybrid device at 10 clinical sites in the United States. Acoustic thresholds were measured for each ear preoperatively and postoperatively, at device activation, and 3, 6, and 12 months post-activation. The outcomes reported were consistent with the clinical trial data submitted to the FDA, that is, participants experienced a significant mean improvement ($p<0.001$) in both co-primary endpoints with the hybrid device over a preoperative hearing aid: CNC words (35.8 ± 27.9 percentage point change; 95% CI 27.9, 43.7) and AzBio sentences in noise (32.0 ± 29.4 percentage point change; 95% CI, 23.7, 40.4); secondary endpoints were met with the hybrid implant relative to performance with a hearing aid. Of the 17 participants that did not maintain functional acoustic hearing, 5 participants chose to have the hybrid implant explanted and replaced with a standard cochlear implant. A total of 65 adverse events were reported involving 34 of 50 participants. The most frequently occurring adverse events were profound/total hearing loss (22 events [33.8%] in 22 participants [40%]), open/short-circuited electrodes (11 events [16.9%] in 11 participants [22%]), and increased tinnitus or tinnitus not present preoperatively (6 events each [9.2%] in 6 participants [12%]). A total of 50 adverse events including tinnitus, vertigo, and other symptoms, were considered to be medical or surgically associated with a mastoidectomy with the facial recess approach used in cochlear implantation. Limitations of this clinical trial include the nonrandomized design, small number of participants, and short duration of follow-up.

The results of a prospective study conducted at 16 European cochlear implant centers evaluated the performance benefits up to 1 year post-Hybrid L24 implantation in terms of speech recognition, sound quality, and quality of life

(Lenarz, 2013). Postoperative performance using a Freedom Hybrid sound processor was compared with that of preoperative hearing aids in 66 individuals with bilateral severe-to-profound high-frequency hearing loss. The group median increase in air conduction thresholds in the implanted ear for test frequencies 125 to1000 hertz was < 15 decibels across the population, both immediately and 1 year postoperatively. A total of 88% of subjects used the Hybrid L24 processor at 1 year post-implantation. A total of 65% of subjects had significant gain in speech recognition in quiet, and 73% in noise (≥ 20 percentage points per 2 decibel signal-to-noise ratio). The mean Speech Spatial Qualities (SSQ) of Hearing Questionnaire subscale scores were significantly improved (+1.2, +1.3, +1.8 points, $p<0.001$), as was the mean health utility-13 (HUI3) score (+0.117, $p<0.01$). Combining residual hearing with the hybrid device implantation gave 22 to 26 percentage points mean benefit in speech recognition scores over hybrid device implantation alone ($p<0.01$). Partially preserved residual hearing was maintained in 89% of subjects within 1 month follow-up, however, some degradation was reported over time with 74% of subjects retaining residual hearing within 30 decibels or preoperative levels at 1 year, and approximately 50% retained hearing within 15 decibels. There were 16 subjects whose 500 hertz thresholds increased by > 30 decibels, with 4 of 16 participants obtaining a large gain in speech recognition score for the implanted ear (55 to 65 percentage points), although the remainder of subjects had limited benefit as either no change (8 of 16 participants) or a reduced score was reported. One subject withdrew from the study after 1 month follow-up due to non-device related health problems and 4 subjects withdrew after 6 months follow-up. The authors reported missing data and inconsistent data collection throughout the study, in particular, at the 1-year primary endpoint for a total of 5 subjects, so only 6-month speech recognition scores were used if available for the Hybrid L24 implanted ear and best aided pre-to postoperative comparisons. Three adverse events were reported during the study which may have been related to the device or surgery, including prickling pain below the eye/sinusitis, middle-ear infection, and dizziness. Two individuals required re-positioning of the implant device due to poor fixation, and confirmed partial extrusion of the electrode array.

Gantz and colleagues (2016) reported on the final outcomes of a multicenter, longitudinal, single-subject (three-stage) study conducted between 2002 and 2011 evaluating 87 individuals implanted with a Nucleus Hybrid S8 implant for high-frequency hearing loss. Speech perception in quiet (CNC words) and in noise (Bamford-Kowal-Bench Sentences-In-Noise [BKB-SIN]) were collected pre- and postoperatively at 3, 6, and 12 months. Subjective questionnaire data were also collected using the Abbreviated Profile for Hearing Aid Benefit (APHAB). The surgical implantation of the 10-mm electrode in the scala tympani resulted in some level of hearing preservation in 98% of participants, with 90% maintaining a functional low-frequency pure tone average (PTA) at initial activation. Two participants experienced immediate postoperative total hearing loss. An additional 6 participants lost enough hearing in the low frequencies to be considered nonfunctional. At 3 months post-activation, a total of 14 participants experienced a total loss of hearing in the implanted ear and nonfunctional hearing. An additional 2 participants experienced total hearing loss at 12 months post-activation, resulting in 16 (18%) participants with nonfunctional hearing loss at 12 months. Data was available for 75 participants at the 12-month evaluation. A significant percentage of participants demonstrated improvement in speech perception as measured by CNC words: 82.5% of participants in the hybrid ear (that is, combined acoustic and electric in the same ear) and 87.5% of participants in the hybrid and combined condition (that is, acoustic plus electric combination when listening with both ears plus the Hybrid S8 device). In addition, all participants reported positive improvements in hearing in three of the four subscales of the APHAB. A total of 14 participants requested explantation of the Hybrid S8 implant for various reasons of dissatisfaction with the device and subsequently had placement of a standard length Nucleus Freedom cochlear implant. The authors concluded that combining acoustic plus electric speech processing has

significant advantages for hearing-impaired individuals with residual low-frequency hearing. Limitations of this study include a lack of data to confirm the etiology of the decline in acoustic hearing sensitivity that resulted in the 19% rate of individuals with nonfunctional hearing loss at 12 months.

Kelsall and colleagues (2017) conducted a multicenter, prospective, nonrandomized, single-arm repeated measures, single-subject study of 50 adults (mean age 64.1 years) with severe to profound sensorineural hearing loss implanted with the Cochlear Nucleus Hybrid implant system. Subjects had residual low-frequency hearing with aided word recognition scores between 10% and 60% in the ear to be implanted and ≥ 80% in the contralateral ear. Patient-reported outcomes were assessed using the SSQ of Hearing Questionnaire, a device use questionnaire (DUQ), and the University of Washington Clinical Assessment of Music (UW-CAMP) questionnaire at preoperative intervals and after 6 and 12 months (SSQ and DUQ only) of hybrid cochlear implant use. Overall, significant improvements were reported at 6 and 12 months postactivation in mean SSQ ratings ($p<0.0001$) and in speech hearing, spatial hearing, and sound quality when compared with the bilateral hearing aid condition preoperatively. Significant improvement was also reported in overall satisfaction on the DUQ in the following measures: 1) listening in speech in various 1:1 and group conversation and listening at a distance; 2) listening in a number of non-speech environments; 3) listening to live and recorded music with and without signing; 4) locating sounds; 5) sound quality of one's own voice, naturalness and clarity of speech, and clarity of environmental sounds; and, 6) across specific listening situations including social engagement. UW-CAMP pitch discrimination and music perception abilities were retained postoperatively. Although some limitations of this study exist, including the non-randomized design, limited sample size, short duration of follow-up, and data collected from subjective measures, the authors stated the benefits reported by study participants were consistent with the objective benefits reported by Roland and colleagues (2016) in the Hybrid L24 clinical trial.

Summary

For individuals who have high-frequency sensorineural hearing loss with preserved low-frequency hearing who receive a hybrid cochlear implant, there are inherent risks of surgical implantation, frequency of adverse events post-implantation, and the potential for profound and possibly total loss of low-frequency hearing resulting in explantation of the device and reimplantation with a standard cochlear implant. The available evidence reporting relevant outcomes includes prospective and retrospective studies using single-arm, within-subject comparison pre- and post-intervention (Gantz, 2016; Lenarz, 2013; Roland, 2016). These trials, along with the views of relevant medical specialists practicing in otolaryngology and neurotology, suggest that for individuals with high-frequency sensorineural hearing loss with preserved low-frequency hearing, hybrid cochlear implantation is associated with improvements in hearing of speech in quiet and noise. In addition, the available evidence also suggests that a hybrid cochlear implant improves speech recognition better than a conventional hearing aid alone in select individuals.

Auditory Brainstem Implantation

Background Information and Description of the Technology

An auditory brainstem implant (ABI) is a device designed to restore some hearing in individuals with neurofibromatosis type 2 rendered deaf by bilateral surgical removal of neurofibromas involving the auditory nerve. The device consists of an externally worn speech processor that provides auditory information to an electrical

signal that is transferred to a receiver/stimulator implanted in the temporal bone. The receiver stimulator is, in turn, attached to an electrode array implanted on the surface of the cochlear nerve in the brainstem, thus bypassing the inner ear and auditory nerve. The electrode stimulates multiple sites on the cochlear nucleus, which is then processed normally by the brain. One device has received FDA approval for auditory brainstem implantation, the Nucleus® 24 Auditory Brain Stem Implant System (Cochlear Americas, Englewood, CO). The speech processor and receiver are similar to the devices used in cochlear implants; the electrode array placed on the brainstem is the novel component of the device.

Efficacy and Safety of Auditory Brainstem Implantation

The FDA approval of the Nucleus 24 Auditory Brain Stem Implant System was based on results of a case series of 90 individuals in which 28 complications occurred in 26 individuals; 26 of these complications resolved without surgical or extensive medical intervention (Donaldson, 2001). Two individuals had infections of the postoperative flap requiring explantation of the device. Effectiveness outcomes were evaluated in 60 individuals with a minimum experience of 3 to 6 months with the device. Device benefit was defined as a significant enhancement of lip-reading or an above-chance improvement on sound-alone tests. Based on this definition, a total of 95% (57 of 60) derived benefit from the device. While the use of an ABI is associated with a modest improvement in hearing, this level of improvement is considered significant in this group of individuals with no other treatment options. Among the 90 individuals receiving the implant, 16 did not receive auditory stimulation from the device postoperatively, either due to migration of the implanted electrodes or surgical misplacement. To place the electrode array on the surface of the cochlear nucleus, the surgeon must be able to visualize specific anatomical landmarks. Because large neurofibromas compress the brainstem and distort the underlying anatomy, it may be difficult or impossible for the surgeon to correctly place the electrode array. For this reason, individuals with large, longstanding tumors may not benefit from the device.

Studies reported by Colletti (2005a, 2006, 2007) showed improvements in word and sentence recognition over a 1 year follow-up.

Merkus and colleagues (2014) reported on a systematic review of ABIs for non-NF2 indications which included 144 non-NF2 ABI cases from 31 publications. Non-NF2 indications for which ABIs were evaluated included auditory neuropathy, autoimmune inner ear disease, bilateral traumatic cochlear nerve disruption, cochlear nerve aplasia, cochlear otosclerosis, temporal bone fractures, and vestibular schwannoma (in the only hearing ear). Cochlear implants generally resulted in better hearing than ABIs when the cochlea and cochlear nerve were intact. Complete bilateral disruption of the cochlear nerve from trauma did not exist in the literature and cochlear malformation did not preclude cochlear implantation. While comparative evidence is limited, cochlear implants appeared to demonstrate greater hearing benefits than ABI in individuals with non-NF2 indications. It was suggested that ABI may only have potential for improved outcomes in bilateral complete cochlear and inner ear aplasia, when imaging and electrophysiologic testing demonstrate that the cochlear nerve is absent. At this time, however, the available evidence in the peer-reviewed published literature is limited in drawing conclusions on the benefits of ABI for these individuals.

Sennaroglu and colleagues (2016) reported on the long-term outcomes of 35 of 60 prelingually deaf children in Turkey who received one of three different ABI models implanted for severe inner ear malformations. A total of 19

children in the analysis were deaf due to cochlear hypoplasia. At regular follow-up, children were evaluated with the CAP, Speech Intelligibility Rate (SIR), Functional Auditory Performance of Cochlea Implantation (FAPCI), and Manchester scores. Approximately one-half of the children were in the CAP category 5 and could understand common phrases without lip reading. In the subgroup with better hearing thresholds (25-40 decibels), some children (17.6%) were able to understand conversation without lip reading, use the telephone with known speaker (11.8%), and follow group conversation in a noisy room (5.9%). For children with higher hearing thresholds (> 50 decibels), none exceeded CAP category 5. SIR and Manchester scores were also better with greater hearing thresholds. Auditory performance measured with the FAPCI was in the 10th percentile for all groups and was worse compared to cochlear implantation. Children with additional nonauditory disabilities (for example, intellectual challenges) had worse outcomes. The authors reported that hearing progress was faster in the initial 2 years when compared with longer use of the ABI. Limitations of this study include the small number of participants in each hearing anomaly group which resulted in lack of certain statistical comparisons of outcomes. Additionally, as most children will have device failure at least two or three times over their lifespan, complications of revision surgery may result because of fibrotic changes in the lateral recess of the ear channel.

Definitions

Auditory brainstem response (ABR): A neurologic test of auditory brainstem function in response to auditory (click) stimuli; the most common application of auditory evoked responses.

Auditory evoked potential: Evaluates the nerve pathways from the ear to the brain; consists of a very small electrical voltage originating from the brain recorded from the scalp in response to an auditory stimulus (for example, different tones, speech sounds).

Asymmetric hearing loss (AHL): A condition in which hearing in the better ear is not normal, but can be restored using a conventional hearing aid (pure-tone average [PTA] between 30 dB HL and 55–60 dB HL).

AzBio Sentences-in-Noise Test: A test to assess a cochlear implant recipient's ability to understand sentences in the presence of background noise.

Binaural hearing: Normal hearing in both ears.

Cochlea: Part of the inner ear that processes sound.

Conductive hearing loss (CHL): Hearing loss that occurs when sound is conducted inefficiently through the outer ear canal to the eardrum and the small bones (ossicles) of the middle ear; this disorder involves a reduction in sound level or the ability to hear faint sounds.

Consonant-Nucleus-Consonant Test (CNC): An open set word recognition test (administered in quiet) consisting of 10 recorded lists of 50 monosyllabic words used to determine speech intelligibility in listeners with hearing impairments.

Decibel (dB): A unit of measurement indicating the loudness of sound. The intensity relates to how loud or soft a sound is. Sound scales are based on either sound pressure level (dB SPL) or hearing level (dB HL).

Electrically evoked auditory brainstem response (EABR): A measurement of auditory brainstem (ABR) integrity using an electrical stimulus with the purpose of determining if the auditory nerve responds as expected to electrical stimulation. The EABR test may be used presurgically in selected individuals to determine if cochlear implantation should be attempted and postsurgically to determine if the implant is working properly.

Evoked otoacoustic emissions (OAE): Sounds measured in the external ear canal that are a reflection of the working of the cochlea. OAE is used in the screening as well as the diagnosis of hearing impairments in neonates and young children. While the test is considered part of the standard battery of tests in infants, it is considered a specialized test in children and adults.

Hearing in Noise Test (HINT): A commonly used speech recognition test consisting of 250 sentences (25 lists of 10 sentences per list) performed in the evaluation of an individual's ability to hear speech in quiet and in noise in the context of sentences.

Meningitis: Inflammation of the meninges, the membranes that surround the brain and the spinal cord. This condition may result in hearing loss or deafness.

Mixed hearing loss: Hearing loss that is both conductive and sensorineural, occurring in one or both ears. This term refers to a condition where conductive hearing loss coexists with sensorineural hearing loss.

Neural plasticity: Ability of the brain, certain parts of the nervous system, or both to adapt to new conditions, such as an injury.

Neurofibromatosis Type 2 (NF-2): A group of inherited disorders in which noncancerous tumors grow on several nerves that usually include the nerve involved with hearing.

Otitis externa: Inflammation or infection of the ear canal.

Otitis media: Inflammation of the middle ear caused by infection.

Postlingual deafness: Hearing loss that occurs after acquiring language or speech.

Prelingual deafness: Hearing loss that occurs before the development of language or speech.

Promontory/round window stimulation test (PST): The application of controlled electrical current to the promontory or round window niche of the middle ear. The current is delivered via an electrode that is inserted either through the tympanic membrane by myringotomy or puncture by the otolaryngologist. The test has been used to predict the electrical response of surviving spiral ganglion nerve fibers and felt to verify a functioning cochlear nerve.

Pure-tone average (PTA): The average of hearing sensitivity (that is, the minimum volume that the person hears) calculated at multiple frequencies (perceived by pitch), typically within the range of 0.25 to 8 kHz (kilohertz).

Scala tympani: The lower tube of the cochlear canal extending from the opening in the medial wall of the middle ear leading into the cochlea.

Sensorineural hearing loss: A permanent hearing loss related to the sensory or neural structures responsible for hearing that involves a reduction in sound level or ability to hear faint sounds; this disorder affects speech understanding or the ability to hear clearly; the involved structures include, but are not limited to, the cochlea and the acoustic nerve.

Severity of hearing loss: Severity of hearing loss is defined in terms of decibels (dB); graded as Mild (21-40 dB), Moderate (41-55 dB), Moderately Severe (56-70 dB), Severe (71-90 dB), Profound (91 dB or more).

Single-sided deafness (SSD): Significant or total hearing loss in one ear; this disorder is sometimes referred to as unilateral sensorineural hearing loss. SSD is defined as a unilateral severe-to-profound deafness (PTA ≥ 70 dB HL), with a contralateral ear that has better, normal or near-normal hearing (PTA ≤ 30 dB HL). SSD may be a result of a congenital unilateral hearing loss, a sudden sensorineural hearing loss, significant head trauma affecting the ear(s), and surgery to treat acoustic neuroma or other tumors of the eighth cranial nerve.

Speech recognition test: A test frequently used to determine cochlear implant candidacy; currently used tests include the following:

- Central Institute for the Deaf (CID) Test
- City of New York (CUNY) Sentence Test
- Hearing in Noise Test (HINT)
- Lexical Neighborhood Test (LNT)
- Multi-syllabic Lexical Neighborhood Test (MLNT)
- Phonetically Balanced-Kindergarten (PBK) Test

Tinnitus: A sensation of ringing or other noises heard in one or both ears which is not caused by an external sound, and other people usually can't hear it. Tinnitus is usually caused by an underlying condition, such as age-related hearing loss, an ear injury or a problem with the circulatory system.

Unilateral hearing loss (UHL): Is generally defined as a condition in which an individual has non-functioning hearing in one ear, receives little or no clinical benefit from amplification in that ear, and has normal or near normal audiometric function in the contralateral ear. UHL includes single-sided deafness (SSD) and asymmetric hearing loss (AHL).

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Index

Auditory brainstem implants
Cochlear implants
Med-El Cochlear Implant System
MED-EL COMBI 40+ Cochlear Implant System
Nucleus hybrid L24 implant system

The use of specific product names is illustrative only. It is not intended to be a recommendation of one product over another, and is not intended to represent a complete listing of all products available.

History

Status	Date	Action
Reviewed	05/12/2022	Medical Policy & Technology Assessment Committee (MPTAC) review. The Definitions and References sections were updated.
Reviewed	05/13/2021	MPTAC review. The Discussion, Definitions, References and Index sections were updated. Reformatted Coding section.
Reviewed	05/14/2020	MPTAC review. References were updated.
Revised	06/06/2019	MPTAC review. The medically necessary statements were revised to remove reference to FDA approved devices. References were updated.

New	07/26/2018	MPTAC review. Initial document development. Moved content of SURG.00014 Cochlear Implants and Auditory Brainstem Implants to a new clinical utilization management guideline document with the same title.