Clinical Policy Bulletin: Cochlear Implants and Auditory Brainstem Implants

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Policy

I. Auditory Brainstem Implant

Aetna considers an auditory brainstem implant (ABI) medically necessary in members 12 years of age or older who have lost both auditory nerves due to disease (e.g., neurofibromatosis or von Recklinghausen's disease).

II. Cochlear Implant

Aetna considers uniaural (monaural) or binaural (bilateral) cochlear implantation a medically necessary prosthetic for adults aged 18 years and older with bilateral, pre- or post-linguistic, sensorineural, moderate-to-profound hearing impairment who meet both of the following criteria:

A. Member has bilateral severe to profound sensorineural hearing loss determined by a pure tone average of 70 dB or greater at 500 Hz, 1000 Hz, and 2000 Hz; and

B. Member has limited benefit from appropriately fitted binaural hearing aids. Limited benefit from amplification is defined by test scores of 40% correct or less in best-aided listening condition on open-set sentence cognition (e.g., Central Institute for the Deaf (CID) sentences, Hearing in Noise Test sentences (HINT), and consonant-nucleus-consonant (CNC) test.

Aetna considers uniaural (monaural) or binaural (bilateral) cochlear implantation a medically necessary prosthetic for infants and children with bilateral sensorineural hearing impairment who meet all of the following criteria:

A. Child has profound, bilateral sensorineural hearing loss determined by a pure tone average of 90 dB or greater at 500, 1000 and 2000 Hz; and

B. Child has limited benefit from appropriately fitted binaural hearing aids. For children 4 years of age or younger, limited benefit is defined as failure to reach developmentally appropriate auditory milestones measured using the
Infant-Toddler Meaningful Auditory Integration Scale, the Meaningful Auditory Integration Scale, or the Early Speech Perception test, or less than 20% correct on open-set word recognition test (Multisyllabic Lexical Neighborhood Test) in conjunction with appropriate amplification and participation in intensive aural habilitation over a 3 to 6 month period. For children older than 4 years of age, limited benefit is defined as less than 12% correct on the Phonetically Balanced-Kindergarten Test, or less than 30% correct on the Hearing in Noise Test for children, the open-set Multi-syllabic Lexical Neighborhood Test (MLNT) or Lexical Neighborhood Test (LNT), depending on the child’s cognitive ability and linguistic skills; and

C. A 3- to 6-month hearing aid trial has been undertaken by a child without previous experience with hearing aids. Note: When there is radiological evidence of cochlear ossification, this requirement may be waived at Aetna’s discretion.

The following additional medical necessity criteria must also be met for uniaural (monaural) or binaural (bilateral) cochlear implantation in adults and children:

A. The member must be enrolled in an educational program that supports listening and speaking with aided hearing; and

B. The member must have had an assessment by an audiologist and from an otolaryngologist experienced in this procedure indicating the likelihood of success with this device; and

C. The member must have no medical contraindications to cochlear implantation (e.g., cochlear aplasia, active middle ear infection); and

D. The member must have arrangements for appropriate follow-up care including the long-term speech therapy required to take full advantage of this device. (Note: Particular plans may place limits on benefits for speech therapy services. Please consult plan documents for details).

Aetna considers cochlear implantation experimental and investigational for auditory dyssynchrony, single-sided deafness, tinnitus and all other indications because its effectiveness for these indications has not been established.

III. Hybrid Cochlear Implants

Aetna considers FDA-approved hybrid cochlear implants (e.g., the Nucleus Hybrid L24 Cochlear Implant System) medically necessary for individuals 18 years of age and older with severe or profound sensori-neural hearing loss of high-frequency sounds in both ears, but who can still hear low-frequency sounds with or without a hearing aid, and the following criteria are met:

A. Severe sensorineural hearing loss with a pure-tone average (PTA) between 60-90 dB HL between 125-1500 Hz and profound loss at higher frequencies in the ear to be implanted; and

B. Speech Perception:

1. Consonant-Nucleus-Consonant (CNC) word recognition score between 0% and 35% inclusive in the ear to be implanted; and

2. CNC word recognition score in the contralateral ear equal to or better than, in the ear to be implanted but not more than 60% in the best-
C. Lack of benefit from a minimum of 30 day hearing aid trial with appropriately fit binaural hearing aids worn on a full-time basis (8 hours per day); and
D. Member has patent cochlea and normal cochlear anatomy, and no ossification or any other cochlear anomaly that might prevent complete insertion of the electrode array; and
E. The following additional medical necessity criteria must also be met:

   1. The member must be enrolled in an educational program that supports listening and speaking with aided hearing; and
   2. The member must have had an assessment by an audiologist and from an otolaryngologist experienced in this procedure indicating the likelihood of success with this device; and
   3. The member must have no medical contraindications to cochlear implantation (e.g., cochlear aplasia, active middle ear infection); and
   4. The member must have arrangements for appropriate follow-up care including the long-term speech therapy required to take full advantage of this device. **(Note: Particular plans may place limits on benefits for speech therapy services. Please consult plan documents for details).**

**Notes:**

Persons with a unilateral cochlear implant may qualify for subsequent bilateral implantation without having to be retested if medical records document that they had met criteria at the time of the initial (first) cochlear implantation.

A cochlear implant includes external components (i.e., a speech processor, a microphone headset and an audio input selector). Replacement of a cochlear implant and/or its external components is considered medically necessary when the existing device can not be repaired or when replacement is required because a change in the member's condition makes the present unit non-functional and improvement is expected with a replacement unit.

Separate assessment will be performed of the medical necessity of recommended accessories and upgrades for a cochlear implant. The member’s current condition, the member’s capabilities with his/her current cochlear implant, and the member’s capabilities of the upgrade or accessory will be considered in determining whether the upgrade or accessory offers clinically significant benefits to the member.

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 and cannot be repaired. Upgrade to or replacement of an existing external speech processor, controller or speech processor and controller (integrated system) is considered not medically necessary when such request is for convenience or to upgrade to a newer technology when the current components remain functional.

The requirement that the member be evaluated by a participating otolaryngologist and audiologist applies only to network plans; all others require documentation of hearing loss which is likely to be improved with the implant.
For adults and children, a post-cochlear implant rehabilitation program is medically necessary to achieve benefit from the cochlear implant. See CPB 0034 - Aural Rehabilitation. The rehabilitation program usually consists of 6 to 10 sessions that last approximately 2.5 hours each.

Aetna follows Medicare rules in considering cochlear implants and auditory brainstem implants as prosthetics. Medicare considers as prosthetics "[c]ochlear implants and auditory brainstem implants, i.e., devices that replace the function of cochlear structures or auditory nerve and provide electrical energy to auditory nerve fibers and other neural tissue via implanted electrode arrays".

Background

The cochlear implant is an electronic prosthesis that stimulates cells of the auditory spiral ganglion to provide a sense of sound to persons with hearing impairment. The patient selection criteria for cochlear implants described above were adapted from the Food and Drug Administration (FDA) approved indications for cochlear implants.

The Centers for Medicare and Medicaid Services (2005) has determined that the evidence is adequate to conclude that cochlear implantation is reasonable and necessary for the treatment of bilateral pre- or post-linguistic, sensorineural, moderate-to-profound hearing loss in individuals who demonstrate limited benefit from amplification. Limited benefit from amplification is defined by test scores of 40 % correct or less in the best-aided listening condition on tape recorded tests of open-set sentence cognition.

Audiologic criteria for pediatric patients follow guidelines similar to those for adults. For adults and children able to respond reliably, standard pure-tone and speech audiometry tests are used to screen likely candidates. For children, the speech reception threshold (SRT) and/or pure-tone average (PTA) should equal or exceed 90 dB; for adults, the SRT/PTA should equal or exceed 70 dB. If the patient can detect speech with best-fit hearing aids in place, a speech-recognition test in a sound field of 55 dB hearing level (HL) sound pressure level (SPL) is performed. A number of speech recognition tests are in current use.

One of the most commonly used speech recognition tests is the Hearing In Noise Test (HINT), which tests speech recognition in the context of sentences. This test uses common, simple sentences such as "How are you feeling?" or "The weather looks good today." HINT reliably and efficiently measures word recognition abilities to determine cochlear implant candidacy. HINT consists of 25 equivalent 10-sentence lists that may be presented in either condition (i.e., quiet, noise) to assess sentence understanding. The HINT test is first administered in quiet, using 2 lists of 10 sentences, scored for the number of words correctly identified. HINT in noise uses sentences administered at +10 signal to noise ratio (Sargent, 2000). For adults, the current cutoff for cochlear implant candidacy is a HINT score of less than 40 %; for children, the current cutoff is a score less than 30 %.

Alternatives to the HINT test for assessing open-set sentence recognition include the CUNY Sentence Test and Central Institute for the Deaf (CID) Test. The words and sentences used for these tests are recorded on tape and used by all cochlear implant centers. All of the tests are of a man’s voice and played at the 70 Decibel range.
Central Institute for the Deaf test consists of a list of 20 sentences. Unlike HINT sentences, CID sentences are uncommon sentences that you would not hear on a regular basis. An example of this type of sentence would be something like this: “The vacuum is in the back of the closet” or “The book is on the top shelf next to the pencil”.

The CUNY Sentence Test was developed by the City University of New York and consists of 72 lists with 12 sentences each. Each list contains 102 words and is scored for the total number of words correctly identified.

The Phonetically Balanced-Kindergarten (PBK) Test, an open-set test of word recognition is typically included in test batteries designed to assess the speech perception skills of profoundly deaf children with cochlear implants. The PBK Test has been used for almost 50 years to assess spoken word recognition performance in children with hearing impairments. The PBK contains 50 monosyllabic words that the child repeats. The PBK Test is most appropriate for children aged 5 to 7 years.

The Lexical Neighborhood Test (LNT) and the Multi-syllabic Lexical Neighborhood Test (MLNT), developed by Indiana University in 1995, are 2 new open-set tests of word recognition. These tests include words that the child repeats, and have been used to assess recognition of individual words and phonemes in children who are cochlear implant candidates. The LNT and MLNT are based on the lexical characteristics of word frequency and neighborhood density, and include words found in the vocabularies of children age 3 to 5. Results from these tests with pediatric cochlear implant users have shown that their lexicons appear to be organized into similarity neighborhoods, and these neighborhoods are accessed in open-set word recognition tests. Studies have shown that normal hearing 3- and 4-year old children are able to recognize all the words from these 2 open-set speech perception tests at very high levels of performance. Therefore, these results have been used as a benchmark for children with hearing impairments.

Children should be receptive to wearing a hearing aid before cochlear implantation because all current implants require an external processor. A period of hearing aid use to ascertain development of aided communication ability is the critical criterion for determining candidacy of young children.

For adults and children, a post-cochlear implant rehabilitation program is necessary to achieve benefit from the cochlear implant. The rehabilitation program consists of 6 to 10 sessions that last approximately 2.5 hours each. The rehabilitation program includes development of skills in understanding running speech, recognition of consonants and vowels, and tests of speech perception ability.

The auditory brainstem implant (ABI) is a modification of the cochlear implant, in which the electrode array is placed directly into the brain. The FDA has approved the Nucleus 24 Multichannel Auditory Brainstem Implant (Cochlear Corporation, Englewood, CO) for use in patients suffering from neurofibromatosis type 2, who have developed tumors on both auditory nerves. When these tumors are surgically removed it is often necessary to remove parts of the auditory nerve resulting in total deafness. Hearing aids and standard cochlear implants are not effective in these patients.

In clinical studies submitted to the FDA, 82 % of the 90 patients implanted with the Nucleus 24 Auditory Brainstem Implant System were able to detect certain familiar sounds, such as honking horns and ringing doorbells; 85 % were able to hear and understand conversation
with the aid of lip-reading; 12% were able to hear well enough to use the phone. Of the 90 patients who received this implant 18% were not able to hear any sound. The ABI System does not restore normal hearing.

The Nucleus 24 Auditory Brainstem Implant System was approved by the FDA on October 20, 2000. It is used in teenagers and adults who have a rare disease (neurofibromatosis type 2) in which tumors growing on cranial nerves need to be surgically removed. Removal of tumors on the auditory cranial nerves requires severing or cutting the nerves, which results in total loss of hearing. These patients cannot be helped by hearing aids or cochlear implants. Subjects used in the study that was submitted to the FDA were individuals aged 12 years of age or older. 

It has been estimated that the incidence of meningitis caused by *Streptococcus pneumoniae* in pediatric cochlear recipients was over 30 times that in similarly aged children in the general population. Based on the 2002 CDC recommendation, cochlear implants recipients should receive age-appropriate vaccination against pneumococcal disease. These individuals should receive the 7-valent pneumococcal conjugate (Prevnar®) or 23-valent pneumococcal polysaccharide (Pneumovax® and Pnu-Imune®) vaccine, or both, according to the Advisory Committee on Immunization Practices (ACIP) schedules for persons at high risk. See CPB 0037 - Pneumococcal Vaccine.

There is evidence of the effectiveness of binaural cochlear implants in improving audition over uniaural (monaural) cochlear implants. A recent technology appraisal prepared by the National Institute for Health and Clinical Excellence (NICE, 2007) recommended simultaneous bilateral cochlear implantation as an option for 3 groups of persons with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids: prelingual children, persons who are blind, and persons at risk for cochlear ossification.

A systematic evidence review (Murphy and O'Donoghue, 2007) concluded: "The available evidence indicates that bilateral cochlear implantation confers material benefits not achievable with unilateral implantation, specifically in terms of sound localization and understanding of speech in noise". By combining the results of available studies, the investigators estimated that adult bilateral recipients showed an increase in sentence recognition of 21% correct over their first implanted ear (p = 0.01) and mean bilateral localization errors of 24 degrees against a monaural error of 67 degrees (p = 0.05). Due to the small number and variety of studies, the investigators were not able to estimate the potential benefits of bilateral cochlear implantation in children. The investigators reported, however, that they identified no high quality evidence for bilateral cochlear implantation, and noted that available evidence has significant limitations that may have influenced these outcomes. The investigators discussed the need for more reliable evidence for bilateral cochlear implantation, and the need to design cochlear implants specifically for bilateral use. The results of these assessments are discussed in further detail below.

As explained below, however, significant product improvements or better quality evidence are unlikely to be forthcoming from industry. Thus, judgments about the effectiveness of bilateral cochlear implantation must be made without the benefit of high quality evidence.

Much of the controversy regarding bilateral cochlear implantation has stemmed from the fact that no evidence for the efficacy of bilateral cochlear implants was presented to the FDA in granting approval for cochlear implants currently on the market. The product
labeling for cochlear implants does not address bilateral versus unilateral implantation (hence, bilateral cochlear implants are not technically "off-label"), and there is no evidence that the FDA contemplated bilateral implantation in granting approval of currently marketed cochlear implants. Thus, cochlear implant manufacturers have promoted bilateral cochlear implantation without having to submit evidence of the efficacy of bilateral cochlear implants to the FDA to support specific labeling.

Although currently marketed cochlear implants were designed for unilateral use, the lack of regulatory scrutiny of bilateral placement of these implants decreases incentives for industry to invest resources to develop new cochlear implants specifically for bilateral use, as any new cochlear implants would need supporting evidence of safety and efficacy for pre-market approval (PMA) from the FDA. This lack of incentives also makes it less likely that cochlear implant manufacturers will fund a high quality study of bilateral cochlear implants to provide reliable evidence of their benefits and risks.

Although in normal listeners, binaural hearing improves sound localization and speech perception, such benefits in cochlear implant users may be limited because the implant's direct electrical stimulation of the auditory nerve does not preserve the fine frequency or fine structure of the acoustic waveforms at each ear, as is created with natural hearing. These features are "of indisputable importance" in binaural hearing (Murphy and O'Donaghe, 2007; see also Quentin-Summerfield et al, 2006). In addition, manufacturers have not developed cochlear implants specifically designed for bilateral use. Thus, bilaterally implanted patients use 2 separate signal processors, one controlling each ear, with independent automatic gain control circuitry. This may fail to preserve interaural differences in level accurately. The 2 unilateral processors are not temporally coordinated, so that they may not preserve the fine temporal differences in sound reaching each ear that facilitates sound localization.

The Swedish Council on Health Technology Assessment (SBU), a leading international technology assessment agency, conducted a comprehensive assessment of current evidence for bilateral cochlear implantation in children (SBU, 2006). The assessment concluded: "Scientific documentation on the benefits of bilateral cochlear implantation in children is insufficient. Well-designed, scientific studies are needed to determine whether the method yields positive effects that outweigh the increased risk for complications". In reviewing the best available evidence, the SBU Report found: "Only a few scientific studies (none of which included a control group) have assessed bilateral cochlear implants. Studies using children as their own controls have reported improvements in speech perception and directional hearing when children used both implants instead of only one. However, these studies provide only low-quality evidence because of their design. Results from clinical studies on complications of unilateral cochlear implantation (CI) in children showed that complication rates varied from 2 percent to 16 percent. A second cochlear implant would double the risk for complications. The SBU assessment found that no studies have specifically investigated the complications or side effects from bilateral cochlear implantation". The SBU assessment recommended prospective controlled clinical outcome studies to evaluate the potential benefits of bilateral cochlear implantation.

The SBU graded the quality of all of the evidence that was available until the time that the systematic evidence review was published. The systematic evidence review provided a structured review of all of the evidence, with explicit consideration of the quality of the evidence. This is the first of several systematic evidence reviews of bilateral cochlear

http://qawww.aetna.com/cpb/medical/data/1_99/0013_draft.html

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implants by any government agency; the fact that the review was prepared by a
government funded agency without any particular stake in the issue better assures that the
assessment is less prone to bias in its preparation and conclusions.

By contrast, industry-funded advocates have focused their arguments on the benefits of
binaural hearing, rather than address the fundamental question of whether there is any
reliable evidence that currently available cochlear implants are capable of providing the
benefits of binaural hearing. Industry-funded advocates have made reference to the
number of studies of bilateral cochlear implants without reference to the quality of that
evidence. Advocates have extensively quoted non-peer reviewed promotional literature
from cochlear implant manufacturers, and the conclusions of published studies are quoted
while omitting an information about the strength of study or the authors’ significant
qualifications to their conclusions. Advocates have also included abstracts and
unpublished articles among cited studies.

Additional literature on bilateral cochlear implants has been published since the SBU
assessment. One of these recently published studies -- a randomized controlled clinical
trial of bilateral implants in post-lingually deafened adults from the Medical Research
Council Institute for Hearing Research (Summerfield et al, 2006) -- is of stronger design
than earlier studies. (In theory, the benefits of bilateral cochlear implantation are more
likely to be manifested in post-lingually deafened persons than pre-lingually deafened
persons). This study found that any benefits of bilateral cochlear implants were modest
and offset by negative effects, such that there was no significant improvement in quality of
life. This study is important in that it is the only randomized controlled clinical study
of bilateral cochlear implants published to date; randomized controlled clinical trials are
considered more reliable than uncontrolled studies because they are significantly less
prone to bias in interpretation of results. This study demonstrates the feasibility of
conducting appropriate and ethical prospective controlled studies of bilateral cochlear
implantation. The study by Summerfield et al (2006) is also significant in that it did not only
assess intermediate outcomes of changes in audioligic parameters, but it also assessed
the clinically relevant outcome of improvement in quality of life. Even though the study
by Summerfield et al (2006) included only 24 subjects, it represented one of the largest
studies of bilateral cochlear implantation published to date.

In this randomized, controlled study (Summerfield et al, 2006), adult users of unilateral
cochlear implants were randomized either to receive a second identical implant in the
contralateral ear immediately, or to wait 12 months while they acted as controls for late-
emerging benefits of the first implant. A total of 24 subjects, 12 from each group,
completed the study. Receipt of a second implant led to improvements in self-reported
abilities in spatial hearing, quality of hearing, and hearing for speech, but to generally non-
significant changes in measures of quality of life, which were offset by decreases in quality
of life due to adverse effects. The investigators concluded: "Multi-variate analyses showed
that positive changes in quality of life were associated with improvements in hearing, but
were offset by negative changes associated with worsening tinnitus". The lack of net
improvement in quality of life precluded a calculation of the cost-effectiveness of bilateral
cochlear implantation using the actual outcomes of this study. A net improvement in
quality of life estimated only in a hypothetical a best-case scenario, in which no worsening
of tinnitus was assumed to occur. The investigators reported, however, that, even in this
hypothetical best-case scenario, the gain in quality of life was too small to achieve an
acceptable cost-effectiveness ratio". This investigator group is planning a similar
randomized controlled clinical study of bilateral cochlear implants in children.
More recently Murphy and O'Donoghue (2007) presented a systematic evaluation of the evidence for bilateral cochlear implantation, which found no high-quality evidence for bilateral cochlear implantation. The investigators found that less than 1/10 of citations retrieved met minimal criteria for consideration as evidence in the analysis, and that more than 2/3 of those citations that qualified as evidence were of poor quality. The investigators identified 387 citations with reference to bilateral cochlear implantation dating back to 1979. Of these 387 articles, 28 were studies meeting minimal criteria for consideration as evidence in this analysis. A further 9 studies were identified from an examination of references and the "gray literature". Of the 37 studies, 9 (24 %) were level 2b evidence (individual cohort study, including low quality randomized controlled trial), 2 (6 %) level 3b (individual case-control study), 16 (43 %) level 4 (case series and poor quality cohort and case-control studies), and 10 (27 %) level 5 evidence (expert opinion).

The authors stated that the results of the literature review identified studies of level 2b to 5 of the benefits of bilateral cochlear implantation (Murphy and O'Donoghue, 2007). However, the investigators found significant limitations that may have influenced their outcomes. The investigators found that most studies failed to provide details of selection criteria, and that some used the same group of cochlear implant users in multiple studies: "In general, the majority of papers failed to detail selection criteria for the participants recruited; in fact, some studies used the same group of cochlear implant users, who will undoubtedly be well-motivated and well-rehearsed in performing these experimental tasks". The investigators found that some studies did not mention the order of testing for bilateral cochlear implant users, a factor that is likely to influence outcomes. They also noted the bias introduced in studies comparing unilateral to bilateral use in persons with bilateral cochlear implants: "It is also important to know that a participant accustomed to wearing bilateral cochlear implants may well perform more poorly in the unilateral condition compared with a unilateral implant user". The investigators stated that "[t]he effect of these issues on a participant's performance could be considerable and may well have influenced the outcome of these clinical studies".

Although bilateral cochlear implantation has been promoted for infants and young children, the investigators found that more than 3/4 of available evidence focuses exclusively on adults (Murphy and O'Donoghue, 2007). Of the 37 studies, 28 (76 %) investigated adults only, 7 (19 %) investigated children only, and 2 (5 %) investigated adults and children.

The investigators concluded that, although available evidence supports the current trend toward bilateral cochlear implantation, "[c]ritical analysis of these studies has highlighted, in particular, the lack of control subjects used and the failure to report important methodologic considerations (e.g., whether sentence tests were open/closed). The low numbers of participants and the poor statistical analysis in the majority of the studies does not allow the reader to assess the true significance of the effects reported. These issues need to be addressed in future longitudinal, prospective clinical studies with sufficient numbers of early (less than 1 year old), simultaneously bilaterally implanted children". The investigators recommended that future implant systems be designed specifically for bilateral use (Murphy and O'Donoghue, 2007).

The conclusions of this assessment are similar to the conclusions of an assessment of cochlear implants prepared by the UK National Health Service (NHS, 2006), which found "no robust evidence" for bilateral cochlear implantation.
In addition, Chin et al (2007) found a lack of reliable evidence comparing the effectiveness of bilateral cochlear implants to binaural/bimodal fitting (combining a cochlear implant and a hearing aid in opposite ears). These researchers (2007) reviewed the evidence to address a question not addressed in the previously cited evidence reviews -- whether better binaural hearing can be achieved with bilateral cochlear implants or binaural/bimodal fitting. The authors found that most studies on comparing unilateral implantation to either mode of bilateral stimulation reported some binaural benefits in some test conditions on average but revealed that some individuals benefited, whereas others did not. The investigators found, however, no reliable evidence comparing bilateral cochlear implants to binaural/bimodal fitting: "There were no controlled comparisons between binaural/bimodal fitting and bilateral implantation and no evidence to support the efficacy of one mode over the other".

A technical report by the American-Speech Language Hearing Association (ASHA, 2004) on cochlear implants found: "Bilateral implantation is currently being studied in a limited number of cochlear implant recipients with mixed results. In some cases, recipients do experience enhanced speech understanding, especially in noise; in other users the improvement in speech understanding compared with unilateral performance is minimal or absent and the primary advantage of binaural implantation is sound localization. Bilateral implantation outcomes to date are encouraging but inconclusive due to the limited number of participants and the scope of the projects. There is a clear need for further exploration of the many variables that can affect the performance of people with binaural implants before widespread use is warranted". The ASHA report emphasized the need for further research on bilateral cochlear implantation: "Many of these studies are currently underway and the results will help to define prognosis and optimization of binaural implant usage. Such studies will determine the ultimate benefit and cost effectiveness of bilateral cochlear implantation".

Offeciers et al (2005) published an "international consensus" that recommended bilateral cochlear implants for all children with profound bilateral hearing loss. However, a review of this paper reveals no evidence that this statement represents anything more than the opinion of the 6 co-authors of this paper. In addition, this paper is not an evidence-based guideline because it makes no reference to the evidence that the coauthors relied upon in reaching their conclusions.

A technology appraisal prepared by the National Institute for Health and Clinical Excellence (NICE, 2007) recommended simultaneous bilateral cochlear implantation as an option for prelingual children with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids. The NICE Appraisal Committee considered the evidence for the clinical effectiveness of bilateral cochlear implants (NICE, 2007). The Committee considered that the additional benefits of bilateral cochlear implantation were less certain than the benefits of unilateral cochlear implantation. This was because of the limitations of the evidence base owing to the small number of studies and the small numbers of participants. However, the Committee considered that the studies had shown additional benefits to having a second cochlear implant in relation to speech perception in noisy situations and directional perception of sound. The Committee heard from patient experts that they considered that there were other benefits from bilateral cochlear implantation. These benefits included easier, less exhausting communication (e.g., determining the direction of the sound in group conversations without unnecessary head movement). The Committee concluded that there were additional benefits of bilateral
cochlear implants that had not been adequately evaluated in the published studies. Therefore there was potential for additional gains in quality of life, although these might vary among individuals.

The NICE technology appraisal (NICE, 2007) recommended simultaneous bilateral cochlear implantation as an option for persons with severe to profound deafness who are at risk for ossification of the cochlea (e.g., after meningitis). The Committee heard from clinical specialists that ossification of the cochlea could preclude successful re-implantation if the first implanted device failed. This would not be an issue for situations in which relatively normal cochlear anatomy is preserved and implanting a second device might be possible if the first failed.

The NICE technology appraisal also concluded that simultaneous bilateral cochlear implantation is an option for person who are blind (NICE, 2007). The Committee heard from clinical specialists that for people who are both deaf and blind, the gains in quality of life following bilateral implantation are greater than for other people. This is because of their increased reliance on auditory stimuli for spatial awareness.

Bichey and colleagues (2008) explored improvements in quality of life (QOL) and the cost-utility of bilateral cochlear implantation. A prospective case-control study was conducted on 23 bilateral cochlear implant patients with the Mark III health utility index. Results indicated a 0.48 mean gain in health utility after bilateral cochlear implantation and a discounted cost per quality adjusted life year of $24,859 in this cohort of patients. With a comparison of patient scores for unilateral and bilateral use, improvements in the domains of hearing, speech, emotion, and cognition were noted, resulting in a mean gain in health utility of 0.11. The authors concluded that this study found an improvement in QOL and a favorable cost-utility associated with bilateral cochlear implantation in patients with profound hearing loss. These patients showed additional improvements in QOL after they received their second implant. This is the first study that showed improvements in QOL and a favorable cost-utility after bilateral cochlear implantation in patients with profound hearing loss.

A statement by the Australian Association for the Deaf (2006) identified another problem with bilateral cochlear implants. They do not endorse bilateral implantation due to the fact that any residual hearing a child has will be totally destroyed by the procedure. They explain that rapid changes in related technology mean that, by leaving one ear intact, the child has the potential to benefit from future developments.

An assessment prepared for the Agency for Healthcare Research and Quality (Raman et al, 2011) found that unilateral cochlear implantation with or without additional use of hearing aids has been an effective method of hearing assistance. The reported stated that published studies show improved speech perception and health-related quality of life in adults with sensorineural hearing loss. The assessment stated that bilateral cochlear implantation provides added improvements in speech perception outcomes in noisy environments over unilateral cochlear implants. However, the report stated that further studies with longer follow-up duration are needed to assess the additional benefits in terms of improved health-related QOL and potential risks of bilateral cochlear implantation compared to unilateral implantation. The report noted that, additionally, none of the studies have been able to quantify the sensation described by patients of fusion of bilateral sound into a stereo perception within one's head. The report concluded that there is a need to develop better measures of performance and disease-specific QOL instruments that may reflect the significance of these subjective benefits.
In October 2007, the FDA reminded physicians that patients with cochlear implants for inner-ear malformations, especially implants with a positioner, are at risk for bacterial meningitis from Streptococcus pneumoniae. This warning follows the deaths of 2 children within the past year, aged 9 and 11 years, who had implants with a positioner and were not fully vaccinated. It should be noted that only 1 implant model has a positioner, and it was withdrawn from the market 5 years ago.

To decrease the risk for meningitis in this population, the FDA recommends:

- Considering prophylactic antibiotics peri-operatively;
- Educating implant recipients and their caregivers about the early signs of meningitis;
- Following the CDC's vaccination guidelines;
- Treating middle ear infections early.

There is emerging evidence for the use of cochlear implants in infants. In a meta-analysis, Vlastarakos et al (2010a) reviewed the evidence on cochlear implantation in infancy, regarding auditory perception/speech production outcomes. The number of cohort-studies comparing implanted infants with under 2-year-old children was 5; 3 represented type-III and 2 type-II evidence. No study was supported by type I evidence. Overall, 125 implanted infants were identified. Precise follow-up period was reported in 82. Median follow-up duration ranged between 6 and 12 months; only 17 children had follow-up duration equal or longer than 2 years. Reliable outcome measures were reported for 42 infants; 15 had been assessed with open/closed-set testing, 14 with developmental rating scales, and 13 with pre-lexical speech discrimination tools. Ten implanted infants assessed with open/closed-set measures had been compared with under 2-year-old implanted children; 4 had shown better performance, despite the accelerated rate of improvement after the first post-operative year. The authors concluded that neuroplasticity/neurolinguistic issues have led cochlear implant centers to implant deaf children in infancy; however, widespread policies regarding the afore-mentioned issue are still not justified. Evidence of these children's outperformance regarding auditory perception/speech production outcomes is limited. Wide-range comparisons between infant implantees and under 2-year-old implanted children are lacking. Longer-term follow-up outcomes should be also made available. They stated that there is a need to develop and validate robust measures of monitoring implanted infants. Potential factors of sub-optimal outcomes (e.g. mis-diagnosis, additional disorders, device tuning, parental expectations) should also be weighted, when considering cochlear implantation in infancy.

In a meta-analysis of diagnostic challenges and safety considerations in cochlear implantation under the age of 12 months, Vlastarakos et al (2010b) stated that the diagnosis of profound hearing loss in infancy, although challenging, can be confirmed with acceptable certainty when objective measures (auditory brainstem response, auditory steady-state response, and otoacoustic emissions) and behavioral assessments are combined in experienced centers. Reliable assessment of the pre-lexical domains of infant development is also important and feasible using appropriate evaluation techniques. Overall, 125 implanted infants were identified in the present meta-analysis; no major anesthetic complication was reported. The rate of surgical complications was found to be 8.8 % (3.2 % major complications) quite similar to the respective percentages in older implanted children (major complications ranging from 2.3 % to 4.1 %). The authors concluded that assessment of hearing in infancy is feasible with adequate reliability. If parental expectations are realistic and hearing aid trial unsuccessful, 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 present meta-analysis did not find an increased rate of anesthetic or surgical complications in infant implantees, although long-term follow-up and large numbers are lacking.

In a prospective cohort study, Colletti et al (2012) determined the long-term outcomes of cochlear implantation in children implanted younger than 6 months and evaluated auditory-based performance in very young children compared with older children, all with profound sensori-neural bilateral hearing loss. A total of 45 subjects (12 aged 2 to 6 months, 9 aged 7 to 12 months, 11 aged 13 to 18 months, and 13 aged 19 to 24 months) with profound bilateral hearing loss were fitted with cochlear implants and followed longitudinally for 4 years. Subjects were developmentally normal with no additional disabilities (visual, motor, or cognitive). Auditory-based communication outcomes included tests for speech perception, receptive language development, receptive vocabulary, and speech production. Age at cochlear implantation was a significant factor in most outcome measures, contributing significantly to speech perception, speech production, and language outcomes. There were no major complications and no significantly higher rates of minor complications in the younger children. The authors concluded that this article reported an uncontrolled observational study on a small group of infants fitted with cochlear implants following personal audiological criteria and, up to now, with limited literature support due to the innovative nature of the study. This study showed, for the first time, significantly improved auditory-based outcomes in children implanted younger than 6 months (only 12 subjects in this group) and without an increased rate of complications. They stated that the data from the present study must be considered as explorative, and a more extensive study is needed.

A retrospective chart review by Holman, et al. (2013) found that cochlear implants provides substantial benefit among infant recipients, and, when performed by an experienced cochlear implant and pediatric anesthesia team, the surgical and anesthetic risks are similar to that expected with both older pediatric and adult patients. The chart review included all children with severe-to-profound sensorineural hearing loss who underwent cochlear implantation at 12 months of age or younger and an audiometric control group implanted between 13 and 24 months of age. Twenty-six patients (41 ears) met criteria for the study. The median duration of follow-up was 58 months. The authors found that no major surgical or anesthetic complications occurred. One patient (4%) experienced device failure, which required revision surgery and implant exchange. Two other patients (8%) had individual electrode anomalies that were treated with map exclusion. At the last recorded follow-up, 73% of patients were performing at or above the level of normal-hearing age-matched peers. The authors reported that patients that were implanted at 12 months of age or younger reached age-appropriate speech and language skills by 24 months of age compared with 40 months for the older pediatric control group.

In a prospective study of patients in a manufacturer-sponsored clinical trial, Luetje et al (2007) examined the benefits of hybrid CI in patients with residual low-frequency hearing. A total of 13 patients (10 women, 3 men; mean age of 51 years) who met candidacy criteria for a hybrid CI were included in this study. Interventions included pre-operative evaluation, CI with a Nucleus Hybrid cochlear implant, subsequent programming, and diagnostic testing. Main outcome measures included benefits of high-frequency electrical
stimulation from the hybrid CI as measured by conventional audiometry, consonant-nucleus-consonant monosyllabic word and Bamford-Kowal-Bench sentence in noise testing at quarterly intervals per protocol. Follow-up ranged from 3 to 24 months. All 13 patients had preserved hearing immediately post-operative. However, 1 lost residual hearing 7 days post-operatively, and 2 patients had delayed hearing losses at 2 and 24 months, the latter apparently due to barotrauma; however, this was not conclusive.

Another had a bilateral symmetrically progressive hearing loss. Six patients showed changes in low-frequency hearing less than 10 dB; 2 showed changes in the range 11 to 20 dB; 2, 21 to 30 dB; and 3, more than 50 dB. Eleven of 13 had improved consonant-nucleus-consonant words ranging up to 83% when tested with hearing aid + CI in the operated ear. Four subjects exhibited improvement in Bamford-Kowal-Bench sentence in noise testing, although only 1 subject showed a significant decline associated with bilateral progression in hearing impairment. The authors concluded that combined electrical and acoustical hearing can result in significant improvement in speech understanding. Only 1 patient lost residual hearing as a direct result of surgery. Two others had delayed losses. There were no absolute predictive factors as to success with hybrid CI, just as there were none for conventional CI. Similarly, wide variation in results may occur. They stated that further studies may clarify factors involved in such variation.

Fitzgerald et al (2008) noted that although electroacoustic stimulation is a promising treatment for patients with residual low-frequency hearing, a small subset of them lose that residual hearing. It is unclear if these patients would be better served by leaving in the 10-mm array and providing electric stimulation through it, or by replacing it with a standard full-length array. These researchers evaluated word recognition and pitch-scaling abilities of cochlear implant users first implanted with a Nucleus 10-mm Hybrid electrode array and then re-implanted with a full length Nucleus Freedom array after loss of residual hearing. Word recognition and pitch-scaling abilities were measured in 2 users of hybrid cochlear implants who lost their residual hearing in the implanted ear after a few months. Tests were repeated over several months, first with a 10-mm array, and after, these patients were re-implanted with a full array. The word recognition task consisted of 2 50-word consonant nucleus consonant (CNC) lists. In the pitch-scaling task, 6 electrodes were stimulated in pseudorandom order, and patients assigned a pitch value to the sensation elicited by each electrode. Shortly after re-implantation with the full electrode array, speech understanding was much better than with the 10-mm array. Patients improved their ability to perform the pitch-scaling task over time with the full array, although their performance on that task was variable, and the improvements were often small. The authors concluded that (i) short electrode arrays may help preserve residual hearing but may also provide less benefit than traditional cochlear implants for some patients, and (ii) pitch percepts in response to electric stimulation may be modified by experience.

In a feasibility study, Gantz et al (2010) examined if the use of a shorter-length cochlear implant (10 mm) on 1 ear and a standard electrode (24 mm) on the contralateral ear is a viable bilateral option for children with profound bilateral sensori-neural hearing loss. A secondary purpose of this study was to determine whether the ear with the shorter-length electrode performs similarly to the standard-length electrode. The goal was to provide an option of electrical stimulation that theoretically might preserve the structures of the scala media and organ of Corti. A total of 8 pediatric patients with profound bilateral sensori-neural hearing loss between the ages of 12 and 24 months were included in this study. Interventions included Nucleus Hybrid S12 10-mm electrode and a Nucleus Freedom implant in the contralateral ear. The Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) parent questionnaire, Early Speech Perception, Glendonald Auditory Screening
Procedure word test, and Children's Vowel tests will be used to evaluate speech perception and the Minnesota Child Development Inventory and Preschool Language Scales 3 test will be used to evaluate language growth. Preliminary results for 8 children have been collected before and after the operation using the IT-MAIS. Three children showed incremental improvements in their IT-MAIS scores overtime. Early Speech Perception, Glendonald Auditory Screening Procedure word test, and Children's Vowel word perception results indicated no difference between the individual ears for the 2 children tested. Performance compared with age-matched children implanted with standard bilateral cochlear implants showed similar results to the children implanted with Nucleus Hybrid S12 10-mm electrode and a Nucleus Freedom implant in contralateral ears. The authors concluded that the use of a shorter-length cochlear implant on 1 ear and a standard-length electrode on the contralateral ear might provide a viable option for bilateral cochlear implantation in children with bilateral profound sensori-neural hearing loss. Moreover, they stated that further study of this patient population will be continued.

Reiss et al (2012) noted that because some users of a hybrid short-electrode cochlear implant lose their low-frequency residual hearing after receiving CI, these investigators tested whether increasing the cochlear implant speech processor frequency allocation range to include lower frequencies improves speech perception in these individuals. A secondary goal was to see if pitch perception changed after experience with the new cochlear implant frequency allocation. A total of 3 subjects who had lost all residual hearing in the implanted ear were recruited to use an experimental cochlear implant frequency allocation with a lower frequency cut-off than their current clinical frequency allocation. Speech and pitch perception results were collected at multiple time points throughout the study. In general, subjects showed little or no improvement for speech recognition with the experimental allocation when the cochlear implant was worn with a hearing aid in the contralateral ear. However, all 3 subjects showed changes in pitch perception that followed the changes in frequency allocations over time, consistent with previous studies showing that pitch perception changes upon provision of a cochlear implant.

Carlson et al (2012) stated that revision surgery using a newer-generation conventional length cochlear implant electrode will provide improved speech perception in patients who initially underwent hybrid electrode implantation and experienced post-operative loss of residual hearing and performance deterioration. These investigators presented 4 patients who experienced delayed post-operative hearing loss following implantation with the Nucleus Hybrid S8 device and underwent re-implantation with the Nucleus Freedom or Nucleus 5 device using the Contour Advance array. Pure-tone thresholds and speech perception data were retrospectively reviewed. Four subjects underwent re-implantation with the Nucleus Freedom or Nucleus 5 device after experiencing deteriorating performance related to delayed acoustic hearing loss. Comparison of pre-revision performance to the most recent post-revision performance demonstrated improved speech perception performance in all subjects following re-implantation. The authors concluded that a small percent of patients will experience a significant loss of residual low-frequency hearing following hybrid implantation thereby becoming completely reliant on a shorter electrode for electrical stimulation. In the current series, re-implantation with a conventional length electrode provided improved speech perception performance in such patients. Revision surgery with a conventional length electrode should be considered in "short electrode" recipients who experience performance deterioration following loss of residual hearing.
Lenarz et al (2013) examined the preservation of residual hearing in subjects who received the Nucleus Hybrid L24 cochlear implant. These researchers investigated the performance benefits up to 1 year post-implantation in terms of speech recognition, sound quality, and quality of life. Post-operative performance using a Freedom Hybrid sound processor was compared with that of pre-operative hearing aids. A total of 66 adult hearing-impaired subjects with bilateral severe-to-profound high frequency hearing loss enrolled in this study. Group median increase in air-conduction thresholds in the implanted ear for test frequencies 125-1,000 Hz was less than 15 dB across the population; both immediately and 1 year post-operatively; 88 % of subjects used the Hybrid processor at 1 year post-op. Sixty-five percent of subjects had significant gain in speech recognition in quiet, and 73 % in noise (greater than or equal to 20 % points/2 dB SNR). Mean SSQ subscale scores were significantly improved (+ 1.2, + 1.3, + 1.8 points, p < 0.001), as was mean HUI3 score (+ 0.117, p < 0.01). Combining residual hearing with CI gave 22 to 26 % age points mean benefit in speech recognition scores over CI alone (p < 0.01). The authors concluded that useful residual hearing was conserved in 88 % of subjects. Speech perception was significantly improved over pre-operative hearing aids, as was sound quality and quality of life.

An UpToDate review on “Treatment of hearing impairment in children” (Smith and Gooi, 2013) does not mention the use of hybrid cochlear implant as a therapeutic option.

Furthermore, hybrid cochlear implants (e.g., Duet EASTM Hearing System) are currently being developed to allow auditory rehabilitation of patients who are not candidates for conventional implants because their low-frequency hearing exceeds current guidelines. Short implant electrodes are placed in the cochlea through a small cochleostomy to preserve low-frequency hearing.

Karsten et al (2013) determined an optimal approach to program combined acoustic plus electric (A+E) hearing devices in the same ear to maximize speech-recognition performance. A total of 10 participants with at least 1 year of experience using Nucleus Hybrid (short electrode) A+E devices were evaluated across 3 different fitting conditions that varied in the frequency ranges assigned to the acoustically and electrically presented portions of the spectrum. Real-ear measurements were used to optimize the acoustic component for each participant, and the acoustic stimulation was then held constant across conditions. The lower boundary of the electric frequency range was systematically varied to create 3 conditions with respect to the upper boundary of the acoustic spectrum: (i) Meet, (ii) Overlap, and (iii) Gap programming. Consonant recognition in quiet and speech recognition in competing-talker babble were evaluated after participants were given the opportunity to adapt by using the experimental programs in their typical everyday listening situations. Participants provided subjective ratings and evaluations for each fitting condition. There were no significant differences in performance between conditions (Meet, Overlap, Gap) for consonant recognition in quiet. A significant decrement in performance was measured for the Overlap fitting condition for speech recognition in babble. Subjective ratings indicated a significant preference for the Meet fitting regimen. The authors concluded that participants using the Hybrid ipsilateral A+E device generally performed better when the acoustic and electric spectra were programmed to meet at a single frequency region, as opposed to a gap or overlap. Although there is no particular advantage for the Meet fitting strategy for recognition of consonants in quiet, the advantage becomes evident for speech recognition in competing-talker babble and in patient preferences.
Szyfter et al (2013) evaluated the hearing preservation rate in patients with high frequency hearing loss, treated with Cochlear Nucleus Freedom Hybrid-L implant in the Otolaryngology Department, Poznan University of Medical Sciences in Poland. A total of 21 patients were operated and implanted with Nucleus Freedom Hybrid-L implant. Pure tone thresholds were recorded prior to the surgery and at the time of speech processor switch-on. Patients were subdivided into 2 groups with respect to their PTA thresholds: (i) group A-classic indications and (ii) group B-extended indications. Average PTA for 3 frequencies (250, 500, 1,000 Hz) were calculated for each patient pre- and post-operatively. In the group of 21 implanted patients in 17 cases these investigators observed preservation of hearing (12 patients from group A, 5 patients from group B) with a mean value of 13.1 dB. In 4 out of 21 patients deafness on the implanted ear was noted. These results indicated that with standard procedure hearing preservation can be obtained in majority of patients. Hearing preservation was not achieved in 19 %, but owing to design of the electrode of the Cochlear Nucleus Hybrid-L that enables to work as CI platform alone, in patients who lost their hearing after surgery re-implantations were not required. The authors concluded that the findings of this study proved that electric acoustic stimulation is a safe and reliable method to help patients with specific type of hearing loss.

Nguyen et al (2013) noted that residual hearing could be preserved with various arrays ranging from 16 to 18 mm in insertion length and 0.25 to 0.5 mm tip diameter. Whether array insertion is performed through a cochleostomy or a round window, tip diameter is an essential criterion for the array design to improve hearing preservation results. These investigators reported the outcome of patients implanted with electric-acoustic CIs with various surgical techniques and array designs. A total of 32 implanted ears (30 patients) were included in this retrospective study. Three array models were inserted: (i) Contour Advance implant (n = 16), (ii) Nucleus Hybrid-L (n = 12), and (iii) Med-El Flex EAS (n = 4). Post-operative pure tone audiometry was performed at 3 and 12 months after implantation. Three months post-operatively, hearing preservation within 30 dB was achieved in 50 %, 50 %, and 84 % cases of patients implanted with a Contour Advance, Flex-EAS, and Hybrid-L, respectively. Two patients (Hybrid-L group) had a delayed sudden hearing loss (greater than 30 dB) 3 months post-operatively and 3 patients (Contour Advance group) had total hearing loss at 1 year. Best results were achieved using arrays with small tip diameters. Cochleostomy or round window insertion did not affect hearing preservation results.

Skarzynski et al (2014) measured benefit in terms of speech recognition in quiet and in noise, and conservation of residual hearing in 3 groups of subjects implanted with the Nucleus Straight Research Array cochlear implant. This device incorporates the Nucleus Slim Straight electrode carrier designed to be easier to insert into the cochlea via the round window while potentially minimizing insertion trauma. The study was prospective, with sequential enrolment and within-subject repeated measures; 35 subjects were 15 to 84 years of age with varying levels of bilateral high-frequency HL. Subjects were divided into 3 groups (A, B, and C) according to pre-operative air conduction hearing thresholds in the ear to implant at 500 Hz; A less than or equal to 50 (n = 11), 50 less than B greater than 80 (n = 13), and C greater than or equal to 80 (n = 11) dB HL. Speech recognition was assessed pre-operatively and at intervals up to 1 year post-implantation. Hearing thresholds were monitored over time and CT scans were used to estimate electrode positions. Pre-operative mean word recognition score was significantly greater for group A compared with group C in quiet (diff. 26.6 % pts, p < 0.05), but not so in noise (diff. 7.9 % pts, p = 0.72). However, a greater proportion of subjects in group A (81 %) achieved a
worthwhile gain in speech recognition score (greater than 20 % pts) in quiet compared with group C (63 %). More importantly, for speech recognition in noise, all subjects in groups A and B achieved a greater than 20 % pts gain compared with only 73 % in group C. Hearing in implanted ears was well conserved for low frequencies, both initially and up to 12 months post-operatively (15 dB median increase in thresholds 250 to 500 Hz). Only 3 of 35 (9 %) cases lost all residual hearing in the implanted ear by 12 months. Where characteristic frequency corresponded to a position occupied by the electrode array, threshold increase was correlated with the pre-operative hearing threshold (r = 0.7; p < 0.001) and closely approximated reported estimates of residual outer hair cell gain. For characteristic frequencies at positions apical to the electrode tip, the relation between threshold increase and residual hearing decreased in amplitude at 45 to 135 degrees (r = 0.42; p <0.05), and disappeared at greater than 135 degrees (r = 0.05; p > 0.05). The authors concluded that gains in speech recognition scores for subjects with better residual low-frequency hearing were greater or equal to those obtained by subjects with poorer residual hearing. Residual hearing after CI with the Nucleus Slim Straight electrode array was well conserved across all 3 groups. It appears that the gain provided by outer hair cell function may be completely suppressed when an electrode array is in close proximity to the organ of Corti.

On March 20, 2014, the FDA approved the Nucleus Hybrid L24 Cochlear Implant System for individuals aged 18 years and older with severe or profound sensori-neural hearing loss of high-frequency sounds in both ears, but who can still hear low-frequency sounds with or without a hearing aid. The may help those with this specific kind of hearing loss who do not benefit from conventional hearing aids.

The Nucleus Hybrid L24 Cochlear Implant System combines the functions of a CI and a hearing aid. This electronic device consists of an external microphone and speech processor that picks up sounds from the environment and converts them into electrical impulses. The impulses are transmitted to the cochlea through a small bundle of implanted electrodes, creating a sense of sound that the user learns to associate with the mid- and high-frequency sounds they remember. The hearing aid portion of the device is inserted into the outer ear canal like a conventional hearing aid, and can amplify sounds in the low-frequency range. The FDA evaluated a clinical study involving 50 individuals with severe to profound high-frequency hearing loss who still had significant levels of low-frequency hearing. The individuals were tested before and after being implanted with the device. A majority of the patients reported statistically significant improvements in word and sentence recognition at 6 months after activation of the device compared to their baseline pre-implant performance using a conventional hearing aid. The device also underwent non-clinical testing, which included the electrical components, biocompatibility and durability of the device. Of the 50 subjects participating in the study, 68 % experienced 1 or more anticipated adverse events, such as low-frequency hearing loss, tinnitus, electrode malfunction and dizziness; 22 developed profound or total low-frequency hearing loss in the implanted ear and 6 of whom underwent an additional surgery to replace the Nucleus Hybrid L24 Cochlear Implant System with a standard CI. While the risk of low-frequency hearing loss is of concern, the FDA determined that the overall benefits of the device out-weigh this risk for those who do not benefit from traditional hearing aids. The device is intended for use on 1 ear only.

Roush et al (2011) summarized current evidence related to the audiologic management of children with auditory neuropathy spectrum disorder (ANSD). These researchers performed a systematic search of the literature in 25 electronic databases (e.g., PubMed,
Cochlear Implants and Auditory Brainstem Implants

A total of 18 studies met the inclusion criteria by addressing 1 or more of 8 clinical questions. Studies were evaluated for methodological quality, and data regarding participant, intervention, and outcome variables are reported. Fifteen of the 18 studies addressed the use of CI, and 4 addressed conventional acoustic amplification. All participants demonstrated improved auditory performance; however, all 18 studies were considered exploratory, and many had methodological limitations. The authors concluded that the clinical evidence related to intervention for ANSD is at a very preliminary stage. They stated that additional research is needed to address the effectiveness of acoustic amplification and CI in children with ANSD and the impact of this disorder on developmental outcomes.

Humphris et al (2013) stated that CI is a standard treatment for severe-profound sensorineural hearing loss (SNHL). However, consensus has yet to be reached on its effectiveness for hearing loss caused by ANSD. In a systematic review, Humphris et al (2013) summarized and synthesized current evidence of the effectiveness of CI in improving speech recognition in children with ANSD. A total of 27 studies were selected for analysis from an initial selection of 237. All selected studies were observational in design, including case studies, cohort studies, and comparisons between children with ANSD and SNHL. Most children with ANSD achieved open-set speech recognition with their CI. Speech recognition ability was found to be equivalent in CI users (who previously performed poorly with hearing aids) and hearing-aid users. Outcomes following CI generally appeared similar in children with ANSD and SNHL. Assessment of study quality, however, suggested substantial methodological concerns, particularly in relation to issues of bias and confounding, limiting the robustness of any conclusions around effectiveness. The authors concluded that currently available evidence is compatible with favorable outcomes from CI in children with ANSD. However, they noted that this evidence is weak; and stronger evidence is needed to support cost-effective clinical policy and practice in this area.

The results of the systematic review by Humphris et al (2013) were similar to an earlier systematic evidence review by the American Speech Language and Hearing Association (ASHA) National Center for Evidence-Based Practice in Communication Disorder (Roush et al, 2011), which found that the studies of ANSD were exploratory and had many methodological limitations, leading them to conclude that the clinical evidence related to CI for ANSD is at a very preliminary stage.

Van de Heyning et al (2008) stated that tinnitus is a well-known, difficult-to-treat symptom of hearing loss. Users of CIs have reported a reduction in tinnitus following implantation for bilateral severe-to-profound deafness. This study assessed the effect of electrical stimulation via a CI on tinnitus in subjects with unilateral deafness and ipsilateral tinnitus who underwent implantation in an attempt to treat tinnitus with the CI. A total of 21 subjects who complained of severe intractable tinnitus that was unresponsive to treatment received a CI. Tinnitus loudness was measured with a visual analog scale (VAS); loudness percepts were recorded with the device activated and de-activated. Tinnitus distress was measured with the Tinnitus Questionnaire before and after implantation. Electrical stimulation via a CI resulted in a significant reduction in tinnitus loudness (mean +/- SD; 1 year after implantation, 2.4 +/- 1.8; 2 years after implantation, 2.5 +/- 1.9; before implantation, 8.5 +/- 1.3). With the device de-activated, tinnitus loudness was still reduced to between 6.1 and 7.0 over 24 months. The Tinnitus Questionnaire revealed a significant positive effect of CI stimulation. The authors concluded that unilateral tinnitus resulting
from single-sided deafness can be treated with electrical stimulation via a CI. The outcomes of this pilot study demonstrate a new method for treatment of tinnitus in select subjects, perhaps an important new indication for CI. The findings of this small pilot study need to be validated by well-designed studies.

Tao and Chen (2012) evaluated the effects of CI on ipsilateral tinnitus. With standard assessment table and standard testing program, 48 post-lingual hearing-impaired adults aged 18 to 62 years (mean age at implantation: 35.0) were operated at 5 clinical centers from June 2009 to March 2010. There were 23 males (47.9 %) and 25 females (52.1 %). These researchers evaluated the pre- and post-implantation degrees of tinnitus, performed free sound field audiometry and scored speech perception during different periods. Secondary analyses were conducted to examine the correlation between the effects of implantation on tinnitus and hearing or speech perception rehabilitation. Before implantation, there were 16 cases with ipsilateral tinnitus and 32 cases without tinnitus. After implantation, among 16 cases, the outcomes were recovery (n = 6), tinnitus suppression (n = 1) and no change in symptoms (n = 9). The total effective rate was 43.8 %. Among another 32 cases without pre-operative tinnitus, 2 cases developed tinnitus after implantation. The effects of CI on tinnitus were negatively correlated with the course of tinnitus. There was no more correlation with other factors. The authors concluded that CIs have significant therapeutic effects on tinnitus in 43.8 % of implant users. Better efficacies are correlated with a shorter course of tinnitus. However, they stated that tinnitus suppression using electrical stimulation via CI for deafness needs to be further evaluated.

Tavora-Vieira et al (2013) examined the effectiveness of CI in patients with unilateral deafness with and without tinnitus. A total of 9 post-lingually deafened subjects with unilateral hearing loss, with and without tinnitus ipsilaterally, and functional hearing in the contralateral ear were implanted with a standard electrode. Speech perception in noise was tested using the Bamford-Kowal-Bench presented at 65 dB SPL. The Speech, Spatial, and Qualities (SSQ) of Hearing Scale was used to evaluate the subjective perception of hearing outcomes, and the Tinnitus Reaction Questionnaire assessed the effect on tinnitus. All patients were implanted with the Med-El Flex soft electrode, Innsbruck, Austria. They were regularly wearing the speech processor and found it beneficial in improving their ability to hear, particularly in noise. Decrease of tinnitus perception and an improvement of sound localization sounds were also reported by these patients. The authors concluded that in this case series, CI was successful for all 9 patients, with improvement of speech recognition in noise, self-perceived improvement of hearing, and for tinnitus control. Moreover, they stated that several factors such as deafness duration, age of deafness onset, the presence of residual hearing, patient motivation, and the rehabilitation intensity need to be further investigated in order to understand their impact on performance after implantation.

In a review on “Cochlear implantation for single-sided deafness: The outcomes. An evidence-based approach”, Vlastarakos et al (2014) reviewed the current evidence on the effectiveness of CI as a treatment modality for SSD, and/or unilateral tinnitus. Systematic literature review in Medline and other database sources was conducted along with critical analysis of pooled data. The study selection includes prospective and retrospective comparative studies, case series and case reports. The total number of analyzed studies was 17. Overall, ten prospective and three retrospective comparative studies, two case series and two case reports which had performed cochlear implantation as a treatment for single-sided deafness and/or tinnitus in unilateral deafness were systematically analyzed.
A total of 108 patients with SSD have been implanted; 66 patients due to problems associated with SSD, and 42 primarily because of debilitating tinnitus. Among the implant recipients, four children were identified. None of those was implanted because of tinnitus. Cochlear implantation in SSD leads to improved sound localization performance and speech perception in noise from the ipsilateral side with an angle of coverage up to (but not including) 90° to the front, when noise is present in the contralateral quartile (Strength of recommendation B). Statistically significant results were reported in one Level II and one Level IV study (n = 25 patients). The remaining studies did not have any statistical analysis. Speech and spatial hearing also subjectively improve following the insertion of a CI (Strength of recommendation B); this was not the case regarding the quality of hearing. Tinnitus improvement was also reported following implant placement (Strength of recommendation B); however, patients need to be advised that the suppression is mainly successful when the implant is activated. Despite the observed improvement in the majority of cases, statistically significant results were reported in only one Level II study (n = 26 patients). Although the analysis identified a median follow-up period of 2 years for the implanted patients, the reported results in the majority of studies were based on follow-up intervals of 1 year or less. The authors stated that “There is a clear need of obtaining results based on longer follow-up periods, to delineate the indications, and further quantify outcomes and factors influencing the results of cochlear implantation in patients with single-sided deafness. This is because consistent and everyday use of the device by the implantees in the long-term is the most important outcome measure, as it verifies efficacy and, as such, justifies the intervention.” The authors also noted differences in evaluation protocols as a weakness in the literature. The authors stated that, although the overall quality of the available evidence supports a wider use of CI in SSD following appropriate selection and counseling (overall strength of recommendation B), it remains to be seen if the long-term follow-up of large number of patients in well conducted high quality studies will confirm the above mentioned results.

Farinetti et al (2014) evaluated the post-operative complications related to CI and discussed the differences observed between adult and pediatric populations. Cochlear implant complications were defined as any pathological events observed during the post-operative period, whether or not they were directly related to the surgical technique. These investigators therefore recorded all complications, in the broad sense of the term, ranging from acute otitis media to cochlear explantation. All surgical procedures (unilateral or bilateral CI, revision surgery) performed in the authors’ institution between March 1993 and January 2013 were reviewed. This population comprised 168 adults (median age at the time of implantation of 51.9 years), and 235 children (median age at the time of implantation of 4.5 years). All post-operative complications were classified as (i) major (requiring surgical revision or hospital management) or (ii) minor (requiring conservative management). The global complication rate was 19.9 % (80/403 cases), comprising 5 % of major complications (20 cases) and 14.9 % of minor complications (60 cases). This complication rate was significantly higher in the adult population (p = 0.004). The authors concluded that CI is a safe hearing rehabilitation surgical technique associated with a low complication rate. However, surgeons must be familiar with these complications in order to ensure optimal prevention. Minor complications were mainly infectious in children (acute otitis media) and cochlea-vestibular in adults (tinnitus and vertigo). Major complications were mostly re-implantation following revision surgery or device failure. Only the minor complication rate was significantly higher in the adult population.

A comparative effectiveness review prepared for the Agency for Healthcare Research and Quality (AHRQ) (Pichora-Fuller, et al., 2013) indicated that the use of cochlear
implants for tinnitus and single sided deafness is a very recent off-label indication, and indicated insufficient evidence for the use of this and other sound therapies for tinnitus.

Appendix

Table: Usual medically necessary frequency of replacement of cochlear implant parts

<table>
<thead>
<tr>
<th>Replacement Parts</th>
<th>Life Expectancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Battery charger kit</td>
<td>1 per 3 years</td>
</tr>
<tr>
<td>Cochlear auxiliary cable adapter</td>
<td>1 per 3 years</td>
</tr>
<tr>
<td>Cochlear belt clip</td>
<td>1 per 3 years</td>
</tr>
<tr>
<td>Cochlear harness extension adapter</td>
<td>1 per 3 years</td>
</tr>
<tr>
<td>Cochlear signal checker</td>
<td>1 per 3 years</td>
</tr>
<tr>
<td>Disposable batteries for ear-level processors</td>
<td>72 per 6 months</td>
</tr>
<tr>
<td>Headset (3-piece component)</td>
<td>1 per 3 years</td>
</tr>
<tr>
<td>Headset cochlear coil (individual component)</td>
<td>1 per year</td>
</tr>
<tr>
<td>Headset cochlear magnet (individual component)</td>
<td>1 per year</td>
</tr>
<tr>
<td>Headset microphone (individual component)</td>
<td>1 per year</td>
</tr>
<tr>
<td>Headset cable or cord</td>
<td>4 per 6 months</td>
</tr>
<tr>
<td>Microphone cover</td>
<td>1 per year</td>
</tr>
<tr>
<td>Pouch</td>
<td>1 per year</td>
</tr>
<tr>
<td>Rechargeable batteries (per set of 2)</td>
<td>1 per year</td>
</tr>
<tr>
<td>Transmitter cable or cord</td>
<td>4 per 6 months</td>
</tr>
</tbody>
</table>

Adapted from: Wisconsin Department of Health and Family Services, 2005.

CPT Codes / HCPCS Codes / ICD-9 Codes

Auditory Brainstem Implant:

CPT codes covered if criteria are met:

92640 Diagnostic analysis with programming of auditory brainstem implant, per hour

HCPCS codes covered if selection criteria are met:

S2235 Implantation of auditory brain stem implant
Other HCPCS codes related to the CPB:

L8699  Prosthetic implant, not otherwise specified [auditory brainstem implant]

ICD-9 codes covered if selection criteria are met:

237.70 - 237.79  Neurofibromatosis
388.5  Disorders of acoustic nerve

Cochlear Implants:

CPT codes covered if selection criteria are met:

69930  Cochlear device implantation, with or without mastoidectomy
92521  Evaluation of speech fluency (eg, stuttering, cluttering)
92522  Evaluation of speech sound production (eg, articulation, phonological process, apraxia, dysarthria)
92523  with evaluation of language comprehension and expression (eg, receptive and expressive language)
92524  Behavioral and qualitative analysis of voice and resonance
92601 - 92602  Diagnostic analysis of cochlear implant, patient younger than 7 years of age
92603 - 92604  Diagnostic analysis of cochlear implant, age 7 years or older
92626 - 92627  Evaluation of auditory rehabilitation status
92630 - 92633  Auditory rehabilitation

Other CPT codes related to the CPB:

69714 - 69715  Implantation, osseointegrated implant, temporal bone, with percutaneous attachment to external speech processor/cochlear stimulator
69717 - 69718  Replacement (including removal of existing device), osseointegrated implant, temporal bone, with percutaneous attachment to external speech processor/cochlear stimulator
90669  Pneumococcal conjugate vaccine, polyvalent, when administered to children younger than 5 years, for intramuscular use
90732  Pneumococcal polysaccharide vaccine, 23-valent, adult or immunosuppressed patient dosage, when administered to 2 years or older, for subcutaneous or intramuscular use

HCPCS codes covered if selection criteria are met:
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, replacement
L8619  Cochlear implant external speech processor, replacement
L8621  Zinc air battery for use with cochlear implant device, replacement, each
L8622  Alkaline battery for use with cochlear implant device, any size, replacement, each
L8623  Lithium ion battery for use with cochlear implant device speech processor, other than ear level, replacement, each
L8624  Lithium ion battery for use with cochlear implant device speech processor, ear level, replacement, 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

Other HCPCS codes related to the CPB:

G0009  Administration of pneumococcal vaccine
S0195  Pneumococcal conjugate vaccine, polyvalent, intramuscular, for children from five to nine years of age who have not previously received the vaccine
V5273  Assistive listening device, for use with cochlear implant

ICD-9 codes covered if selection criteria are met:

389.11  Sensory hearing loss, bilateral
389.12  Neural hearing loss, bilateral
389.18  Sensorineural hearing loss, bilateral
389.22  Mixed hearing loss, bilateral

Other ICD-9 codes related to the CPB:

381.00-382.9  Otitis media
The above policy is based on the following references:

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Immunization Practices. Pneumococcal vaccination for cochlear implant candidates
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Auditory brain stem implants. Pre-assessment No. 36. Ottawa, ON: CCHTA; June
25. van Hoesel RJ. Exploring the benefits of bilateral cochlear implants. Audiol
Institute for Health Research and Development, University of Southampton; 2003:1-12.
27. Centers for Medicare and Medicaid Services (CMS). Decision memo for cochlear
Accessed April 6, 2005.
29. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular
implants. Ear Hear. 2002;23(1 Suppl):80S-89S.
32. Swedish Council on Technology Assessment in Health Care (SBU). Bilateral
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way to optimize auditory perception abilities in deaf children? Int J Pediatr
41. Dunn CC, Tyler RS, Witt SA, Gantz BJ. Effects of converting bilateral cochlear implant subjects to a strategy with increased rate and number of channels. Ann of Oto Rhinol Laryngol. 2006;115:425-432.


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