DISCLAIMER/INSTRUCTIONS FOR USE

Medical Policy provides general guidance for applying Blue Cross of Idaho benefit plans (for purposes of Medical Policy, the terms “benefit plan” and “member contract” are used interchangeably). Coverage decisions must reference the member specific benefit plan document. The terms of the member specific benefit plan document may be different than the standard benefit plan upon which this Medical Policy is based. If there is a conflict between a member specific benefit plan and the Blue Cross of Idaho’s standard benefit plan, the member specific benefit plan supersedes this Medical Policy. Any person applying this Medical Policy must identify member eligibility, the member specific benefit plan, and any related policies or guidelines prior to applying this Medical Policy. Blue Cross of Idaho Medical Policies are designed for informational purposes only and are not an authorization, explanation of benefits or a contract. Receipt of benefits is subject to satisfaction of all terms and conditions of the member specific benefit plan coverage. Blue Cross of Idaho reserves the sole discretionary right to modify all its Policies and Guidelines at any time. This Medical Policy does not constitute medical advice.

POLICY

Unilateral use of an auditory brainstem implant (using surface electrodes on the cochlear nuclei) may be considered medically necessary in patients with neurofibromatosis type 2, who are 12 years of age or older, and who are rendered deaf due to bilateral resection of neurofibromas of the auditory nerve.

An auditory brainstem implant is considered investigational for all other conditions including non-neurofibromatosis type 2 indications.

Bilateral use of an auditory brainstem implant is considered investigational.

Penetrating electrode auditory brainstem implant is considered investigational.

POLICY GUIDELINES

There is no specific CPT code for the implantation of this device. CPT codes that might be used include codes 61863–61868 (twist drill, burr hole, craniotomy or craniectomy code range) and code 64568 (incision for implantation of cranial nerve neurostimulator electrode array and pulse generator).

S2235: Implantation of auditory brainstem implant.

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

BENEFIT APPLICATION
BlueCard/National Account Issues

State or federal mandates (e.g., Federal Employee Program) may dictate that certain U.S. Food and Drug Administration–approved devices, drugs, or biologics may not be considered investigational, and thus these devices may be assessed only by their medical necessity.

Placement of an auditory brainstem implant is a specialized procedure that may require out-of-network referral.

Some facilities may negotiate a global fee for the implantation of the device and the associated aural rehabilitation. Charges for rehabilitation may be subject to individual contractual limitations.

BACKGROUND

The auditory brainstem implant (ABI) is intended to restore some hearing in people with neurofibromatosis type 2 who are rendered deaf by bilateral removal of the characteristic neurofibromas involving the auditory nerve. The ABI consists of an externally worn speech processor that provides auditory information by 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. To place the electrode array on the surface of the cochlear nucleus, the surgeon must be able to visualize specific anatomic landmarks. Because large neurofibromas compress the brainstem and distort the underlying anatomy, it can be difficult or impossible for the surgeon to correctly place the electrode array. For this reason, patients with large, long-standing tumors may not benefit from the device.

ABIs are also being studied to determine whether they can restore hearing for other non-neurofibromatosis causes of hearing impairment in adults and children, including absence of or trauma to the cochlea or auditory nerve. It is estimated that 1.7 per 100,000 children are affected by bilateral cochlea or cochlear nerve aplasia and 2.6 per 100,000 children are affected by bilateral cochlea or cochlear nerve hypoplasia.1

Regulatory Status

In 2000, the Nucleus® 24 Auditory Brainstem Implant System (Cochlear Corp.) was approved by the U.S. Food and Drug Administration (FDA) through the premarket approval process. 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. The device is indicated for individuals 12 years of age or older who have been diagnosed with neurofibromatosis type 2. The Nucleus® 24 Auditory Brainstem Implant System approval was based on the efficacy study of unilateral implants either at first-side or second-side tumor removal surgery.”2 The Nucleus® 24 is now obsolete.

In June 2016, the Nucleus ABI541 Auditory Brainstem Implant (Cochlear Corp.) was approved by the Food and Drug Administration through a supplement to the premarket approval for the Nucleus® 24. The new implant is indicated for individuals 12 years of age or older who have been diagnosed with neurofibromatosis type 2.

Food and Drug Administration product code: MCM.

RATIONALE

This evidence review was created in July 2002 and has been updated regularly with searches of the MEDLINE database. The most recent literature update was performed through December 10, 2018.
Evidence reviews assess the clinical evidence to determine whether the use of a technology improves the net health outcome. Broadly defined, health outcomes are length of life, quality of life, and ability to function—including benefits and harms. Every clinical condition has specific outcomes that are important to patients and to managing the course of that condition. Validated outcome measures are necessary to ascertain whether a condition improves or worsens; and whether the magnitude of that change is clinically significant. The net health outcome is a balance of benefits and harms.

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

In the case of the auditory brainstem implant (ABI), studies that compare outcomes before and after device implantation can provide useful information on health outcomes. Following is a summary of the key literature to date.

**ABI for Bilateral Resection of Neurofibromas of the Auditory Nerve**

U.S. Food and Drug Administration (FDA) approval of the Nucleus 24 Auditory Brainstem Implant System was based on results in a case series of 90 patients with neurofibromatosis type 2 (NF2), ages 12 years and older. Of the 90 subjects evaluated, 28 complications occurred in 26 patients; 26 of these complications resolved without surgical or extensive medical intervention. Two patients had infections of the postoperative flap requiring explantation of the device. Sixty patients had a minimum experience of 3 to 6 months with the device, and thus effectiveness outcomes were also evaluated. Overall device benefit was defined as a significant enhancement of lip reading or an above-chance improvement on sound-alone tests. Based on this definition, 95% (57/60) of patients derived benefit from the device. Among the 90 patients receiving the implant, 16 did not receive auditory stimulation from the device postoperatively, either due to migration of the implanted electrodes or surgical misplacement.

Similar results have been reported with other devices in European studies. Matthies et al (2013) reported on 32 patients with ABIs placed for NF2. Activation of the ABI occurred in 27 patients. Three patients experienced no auditory perception. At 12-month follow-up, significant improvements were seen on the Sound Effects Recognition Test and the Monosyllable-Trochee-Polysyllable test. Open set sentence recognition was 5% at first fitting and improved to 37% at 12 months. Performance did not significantly correlate with the number of active electrodes implanted. Sanna et al (2012) reported on 25 ABIs placed in 24 patients with NF2. In this retrospective case study, patients were followed for 2 to 53 months. Sound recognition was present in 19 patients, of whom 11 had some word recognition and 8 had good speech recognition (50% speech discrimination in 4 patients, 75%-100% speech discrimination and telephone use in 4 patients). Multivariate analysis failed to identify any statistically significant factors that predicted ABI performance outcomes. The authors also conducted a review of the literature on ABIs and found it difficult to compare outcomes because reporting methods and outcomes measured were inconsistent and imprecise.
A single small (N=10) trial from 2008 was identified on a penetrating ABI (PABI). This prospective clinical trial enrolled patients with NF2 who received a PABI after vestibular schwannoma removal. The PABI is an extension of the ABI technology that uses surface electrodes on cochlear nuclei. The PABI uses 8 or 10 penetrating microelectrodes in conjunction with a separate array of 10 to 13 surface electrodes. The PABI met the goals of lower threshold, increased pitch range, and high selectivity, but these properties did not improve speech recognition.

Section Summary: ABI for Bilateral Resection of Neurofibromas of the Auditory Nerve

The largest dataset on use of the ABI in patients with NF2 is from the FDA-regulated prospective series with the Nucleus 24 implant. This study found enhancement of lip reading and improvement in sound-alone tests in patients who had bilateral deafness and no sound perception prior to the ABI. Eighteen percent of patients did not receive auditory benefit after device implantation. Based on these results, FDA approved the Nucleus 24 implant for the indications in the trial (patients ≥12 years with NF2). Similar results have been shown in smaller retrospective series from Europe in patients with NF2. No benefit on speech recognition was found with penetrating electrodes compared with surface electrodes.

ABI for Nontumor Indications

Adults

Merkus et al (2014) reported on a systematic review of ABIs for non-NF2 indications. Included in the review were 144 non-NF2 ABI cases from 31 articles. Non-NF2 indications for which ABIs have been evaluated include cochlear otosclerosis, temporal bone fractures, bilateral traumatic cochlear nerve disruption, autoimmune inner ear disease, auditory neuropathy, cochlear nerve aplasia, and vestibular schwannoma in the only hearing ear. Cochlear implants have generally provided better hearing than ABIs when the cochlea and cochlear nerve are intact. Complete bilateral disruption of the cochlear nerve from trauma did not exist in the literature and cochlear malformation did not preclude cochlear implant. While the evidence is limited, it appears as if cochlear implants demonstrate greater hearing benefits than ABIs in patients with non-NF2 indications.

In a literature review by Medina et al (2014) assessing ABI for traumatic deafness, cochlear implant performed better than ABI. However, there was limited evidence on which to draw conclusions, because only 3 articles (total N=7 patients) were identified in the review on ABI for traumatic deafness.

Children

Systematic Reviews

A systematic review of nontumor pediatric ABI outcomes was reported by Noij et al (2015). It included 21 studies with 162 children, at a mean age of 4.3 years (range, 11 months to 17 years). Nine reports were from a single group from Italy (described below) and it could not be determined if there was patient overlap across these studies. Nearly all studies were retrospective series or cohorts; one was a case-control. Most children (63.6%) had cochlear nerve aplasia. Other conditions were cochlear aplasia, cochlear nerve hypoplasia, cochlear malformations, ossified cochlea, auditory neuropathy, trauma, and cochlear hypoplasia. Twenty-five percent of the patients had previously received a cochlear implant. Forty major and minor implant-related complications were reported, the most common being cerebrospinal fluid leak (8.5% of patients). The most common side effects associated with ABI use were discomfort of the body and/or limb, dizziness/vertigo/nystagmus, pain in the head and/or neck, and stimulation of the facial nerve or involuntary swallowing, gagging, or coughing. A variety of auditory tests were used; the most common (6 studies) was the Categories of Auditory Performance (CAP) index (range, 0-7; high score indicates better hearing). There was an improvement in CAP scores over time.
After 5 years, almost 50% of patients had CAP scores greater than 4 (5 [understanding of common phrases without lip reading] to 7 [use of telephone with known speaker]). Children who also had nonauditory disabilities never attained a CAP score greater than 4. There was no significant effect of the age of implantation.

**Case Series**

Many of the larger series on ABI in nontumor patients are from a group that includes Colletti and Colletti. In 2013, this group reported on ABIs in 21 children, ranging in age from 1.7 to 5 years, with deafness unrelated to neurofibromatosis, who had a poor response to cochlear implants. At surgery, the cochlear nerve was absent in each patient. Significant improvements in CAP index scores were seen after ABI (p<0.001).

Sennaroglu et al (2016) reported on follow-up of at least 1 year for 35 children who had received ABI. This followed a 2009 preliminary report of 11 prelingually deaf children ages 30 to 56 months who received an ABI. Sixty children had received an ABI from this center in Turkey. The children who had received the ABI in the previous year were excluded from the 2016 analysis. Over half (n=19) of the cases were due to cochlear hypoplasia. ABI models implanted were Cochlear, Med El, and Neurelec. At regular follow-up, children were evaluated with the CAP, Speech Intelligibility Rate, Functional Auditory Performance of Cochlea Implantation, and Manchester scores. About half 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 (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. Speech Intelligibility Rate and Manchester scores were also better with greater hearing thresholds. Auditory performance measured with the Functional Auditory Performance of Cochlea Implantation was in the 10th percentile for all groups and was worse compared with cochlear implantation. As was also found in the Noij systematic review (discussed above), children with additional nonauditory handicaps had worse outcomes (eg, intellectual disability).

Also, Puram et al (2016) reported on early experiences with pediatric ABI in a North American center conducting an ongoing FDA-regulated investigational device exemption trial (NCT01864291). Of 17 candidates evaluated, 5 (average age, 19.2 months) met the study selection criteria and received an implant (Nucleus AB124). Detailed inclusion and exclusion criteria are described in the report. The age at implantation ranged from 11 months to 2.5 years. After implantation, all patients had responses such as babbling and responses to sounds and speech. There were no major or minor complications such as cerebrospinal fluid leak. Two devices failed after blunt trauma (falls) at 6 and 7 months postimplantation, respectively, and 1 spontaneous device failure occurred at 15 months postsurgery. The current protocol includes use of a helmet in children who are at risk of falling.

**Mixed Populations**

Other reports from the group of Colletti and Colletti include a 2005 report on ABIs in 16 children and adults who had nontumor diseases of the cochlear nerve or cochlea and 13 patients with NF2. Ages ranged from 14 months to 70 years; the nontumor group included patients with head trauma, complete cochlear ossification, auditory neuropathy, and bilateral cochlear nerve aplasia. Following implantation, the adult nontumor group scored substantially higher than the patients with NF2 in open set speech perception tests. Some children showed dramatic improvements in word and sentence recognition over a 1-year follow-up. Short-term adverse events included dizziness or tingling sensations in the leg, arm, and throat (20/29 patients). Additional studies from this group have reported improvements in hearing.
with ABIs in “nontumor” patients, including a 2006 report on 54 nontumor patients\textsuperscript{15} and a 2007 report on 22 non-neurofibromatosis patients.\textsuperscript{16}

In a retrospective review, Colletti et al (2010) reported on complications from ABI surgery in 83 adults and 31 children, 78 of whom had nontumor cochlear or cochlear nerve disorders.\textsuperscript{17} Authors found that ABI complication rates were similar to those for cochlear implant surgery. Additionally, there were significantly fewer major and minor complications in nontumor patients than in NF2 patients.

**Section Summary: ABI in Nontumor Indications**

The evidence on ABI in nontumor patients includes case series and systematic reviews of case series. A 2014 systematic review of adults suggested that ABI might improve outcomes in bilateral complete cochlear and inner ear aplasia. Recent research includes studies of children who are deaf but would not benefit from a cochlear implant. The most common conditions in these studies are cochlear aplasia and cochlear nerve aplasia. Hearing in this age group is critical for language development, and the ABI has potential to substantially improve health outcomes for this age group. However, a 2016 U.S. study found a high rate of device failure (3/17) with the only device approved for use in the United States (now obsolete), and other studies have indicated that outcomes are inferior when children have additional disabilities. A number of studies in children are ongoing (see Table 1). Results from these studies might identify the patient populations who would benefit most from this device. Results on the recently available Nucleus AB124 are also needed to evaluate device efficacy and durability.

**Summary of Evidence**

For individuals who are deaf due to bilateral resection of neurofibromas of the auditory nerve who receive an ABI, the evidence includes a large prospective case series. Relevant outcomes are functional outcomes, quality of life, and treatment-related morbidity. The FDA approval of the Nucleus 24 device in 2000 was based on a prospective case series of 90 patients 12 years of age or older, of whom 60 had the implant for at least 3 months. From this group, 95% had a significant improvement in lip reading or improvement on sound-alone tests. While use of an ABI is associated with a very modest improvement in hearing, this level of improvement is considered significant for those patients who have no other treatment options. Based on these results, ABIs are considered appropriate for the patient population included in the trial (ie, age \( \geq 12 \) years with NF2 and deafness following tumor removal). The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who are deaf due to nontumor etiologies who receive an ABI, the evidence includes case series and systematic reviews of case series. Relevant outcomes are functional outcomes, quality of life, and treatment-related morbidity. In general, ABIs have not demonstrated hearing benefits over cochlear implants for many conditions not related to neurofibromatosis type 2. However, ABIs hold promise for select patients when the cochlea or cochlear nerve is absent. Many recent and ongoing ABI studies are being conducted in children. For children, hearing is critical for language development, and this device has potential to substantially improve health outcomes. The most common nontumor conditions in children are cochlear aplasia and cochlear nerve aplasia. There are questions about the durability of the now obsolete Nucleus 24 in active young children. Evaluation is currently ongoing with the recently available Nucleus ABI541 to determine its efficacy and durability in children. In addition, ABI studies have shown inferior outcomes in children with other disabilities. Thus, further study is also needed to define populations that would benefit from these devices. The evidence is insufficient to determine the effects of the technology on health outcomes.
SUPPLEMENTAL INFORMATION

Practice Guidelines and Position Statements

In 2005, National Institute Health and Care Excellence issued guidance on interventional procedures for auditory brainstem implants. The guidance stated: “...evidence on safety and efficacy of auditory brainstem implants appears adequate to support the use of this procedure by surgical teams experienced in this technique.”

U.S. Preventive Services Task Force Recommendations

Not applicable.

Medicare National Coverage

There is no national coverage determination. The Medicare Benefit Policy Manual references hearing aids and auditory implants, stating that hearing aids are excluded from coverage, including air-conduction and bone-conduction devices. However, devices that produce the perception of sound by replacing the function of the middle ear, cochlea, or auditory nerve are payable by Medicare as prosthetic devices. These devices are indicated only when hearing aids are medically inappropriate or cannot be used. Along with cochlear and auditory brainstem implants, the benefit manual specifically refers to osseointegrated implants as prosthetic devices.

Ongoing and Unpublished Clinical Trials

Some currently unpublished trials that might influence this review are listed in Table 1.

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<td>NCT02310399</td>
<td>Auditory Brainstem Implant (ABI) in Children With No Cochlea or Auditory Nerves</td>
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<td>NCT02102256</td>
<td>A Feasibility Study of the Placement, Use, and Safety of the Nucleus 24 Auditory Brainstem Implant in Non-Neurofibromatosis Type 2 (NF2) Pediatric Patients</td>
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Deafness

NCT: national clinical trial.

**ESSENTIAL HEALTH BENEFITS**

The Affordable Care Act (ACA) requires fully insured non-grandfathered individual and small group benefit plans to provide coverage for ten categories of Essential Health Benefits (“EHBs”), whether the benefit plans are offered through an Exchange or not. States can define EHBs for their respective state.

States vary on how they define the term small group. In Idaho, a small group employer is defined as an employer with at least two but no more than fifty eligible employees on the first day of the plan or contract year, the majority of whom are employed in Idaho. Large group employers, whether they are self-funded or fully insured, are not required to offer EHBs, but may voluntary offer them.

The Affordable Care Act requires any benefit plan offering EHBs to remove all dollar limits for EHBs.

**REFERENCES**


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literature review through December 11, 2018; no references added. Policy statements unchanged.