The auditory brain stem implant (ABI) is a device designed to restore some hearing in people with neurofibromatosis type II who are rendered deaf by bilateral removal of the characteristic neurofibromas involving the auditory nerve.

The auditory brainstem implant (ABI) consists of an externally worn speech processor that provides auditory information to an electrical signal that is transferred to a receiver/stimulator that is implanted in the temporal bone. The receiver stimulator is, in turn, attached to an electrode array that is 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.

Regulatory Status
One device has received approval by the U.S. Food and Drug Administration (FDA) for auditory brainstem implantation: the Nucleus 24® Auditory Brainstem Implant System (Cochlear Corporation). 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 (NF2).
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 (PABI) 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 64573 (incision for implantation of neurostimulator electrodes; cranial nerve).

In 2007, a CPT code for diagnostic analysis with programming of this device became effective:

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

Rationale

Unilateral Auditory Brainstem Implant In Patients With Neurofibromatosis Type 2

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. (1, 2) 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. A total of 60 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, a total of 95% patients (57 of 60) derived benefit from the device. While the use of an auditory brainstem implant (ABI) is associated with a very modest improvement in hearing, this level of improvement is considered significant in this group of patients who have no other treatment options. 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. To place the electrode array on the surface of the cochlear nucleus, the surgeon must be able to visualize specific anatomical landmarks. Because large neurofibromas compress the brainstem and distort the underlying anatomy, it may be difficult or impossible for the surgeon to correctly place the electrode array. For this reason, patients with large, long-standing tumors may not benefit from the device.

In 2012, Sanna and colleagues reported on 25 ABIs placed in 24 patients with NF2. (3) In this retrospective case study, patients were followed up for a range of 2–53 months. One patient died due to NF2 progression. 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 and 75-100% speech discrimination and telephone use in 4 patients). A multivariate analysis did not identify any factors that were statistically significant in predicting ABI performance outcomes. The authors also conducted a review of the literature on ABIs and found it difficult to compare outcomes as reporting methods and outcomes measured were inconsistent and imprecise.

Unilateral Auditory Brainstem Implant In Nontumor Patients

In a 2004 study in Italy, V. Colletti and colleagues reported on the use of ABIs in patients with deafness unrelated to neurofibromatosis and who had a poor response to cochlear implants. (4) However, there are inadequate data from this study to permit scientific conclusions regarding this additional indication.

V. Colletti and colleagues, in 2005, presented data from ABIs in 16 children and adults who had non-tumor diseases of the cochlear nerve or cochlea and 13 patients with NF2. (5) Ages ranged from 14 months to 70 years; the non-tumor group included patients with head trauma, complete cochlear ossification, 1 child with auditory neuropathy, and 5 children with bilateral cochlear nerve aplasia. Following implantation, the adult non-tumor group scored substantially higher than the patients with NF2 in open set speech perception tests. Some of the children showed dramatic improvements in word and sentence recognition over a 1-year follow-up. Short-term adverse effects included dizziness or tingling sensations in the leg, arm, and throat (20 of 29 patients).

Additional studies report improvement in hearing with ABIs in “non-tumor” patients; V. Colletti has reported results on 54 non-tumor patients, (6) and L. Colletti has reported results on 22 non-neurofibromatosis patients. (7)

In a 2010 retrospective review, the authors previously cited, V. Colletti and colleagues, reported on the complications from ABI surgery in 83 adults and 31 children, 78 of whom had nontumor cochlear or cochlear nerve disorders. (8) The authors found complication rates were similar to cochlear implant surgery. Additionally, major and minor complications were significantly fewer in
nontumor patients than in NF2 patients. These authors concluded ABIs can be used in a wider population of patients than only those with NF2. However, this review did not evaluate hearing outcomes.

Sennaroglu et al., in 2009, reported on the use of ABIs in 11 prelingually deaf children ages 30-56 months. (9) Results showed mean performance on the Meaningful Auditory Integration Scale improved in all patients. However, the results of this small study are described as only preliminary.

**Bilateral Auditory Brainstem Implants**

Nucleus 24® Auditory Brainstem Implant System (Cochlear Corporation) labeling states (1) “The efficacy of bilateral implantation with the ABI has not been studied.” No evidence was identified to support bilateral auditory brainstem stimulation. The studies included to date only included patients with unilateral auditory brainstem implantation.

**Penetrating Electrode Auditory Brainstem Implant**

In 2008, Otto et al. (10) conducted a prospective clinical trial (n=10) with patients with NF2 who received a penetrating electrode auditory brainstem implant (PABI) after vestibular schwannoma removal. The PABI is an extension of the ABI technology that uses surface electrodes on cochlear nuclei. 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 result in improved speech recognition. These data are inadequate to draw conclusions regarding the effectiveness of PABI as compared to conventional ABI.

Schwartz et al. (11) discuss in a 2008 review article the future directions in central implants for hearing, including PABI, the use of ABI in non-tumor patients, and the auditory midbrain implant.

**Ongoing Clinical Trials**

A search of online site ClinicalTrials.gov identified no ongoing studies on auditory brainstem implant.

**Summary**

The Nucleus 24 Auditory Brainstem (ABI) Implant received FDA approval only for patients with neurofibromatosis type 2 (NF2) following tumor removal. The available evidence for unilateral use of ABI devices in patients with NF2 is sufficient to demonstrate improvements in net health outcomes. Therefore, the policy statement indicates an auditory brainstem implant may be considered medically necessary in this condition.

ABIs hold promise for patients with cochlear and cochlear nerve abnormalities when cochlear implants are not indicated. However, studies on ABIs for non-NF2 conditions are limited with small numbers of patients and insufficient data to make scientific conclusions. Given the lack of both high-quality evidence and FDA approval, ABI for non-NF2 conditions and bilateral ABI are considered investigational. Penetrating electrode auditory brainstem implant is also considered investigational since the very limited evidence available is insufficient to draw conclusions on health outcomes.

**Practice Guidelines and Position Statements**
In January 2005, National Institute for Clinical Excellence (NICE) issued Interventional Procedure Guidance 108, *Auditory Brain Stem Implants*. (12) The guidance states the following: “…evidence on safety and efficacy of auditory brain stem implants appears adequate to support the use of this procedure by surgical teams experienced in this technique.”

**Medicare National Coverage**

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. (13) However, devices which 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 utilized. Along with cochlear and auditory brainstem implants, the benefit manual specifically refers to osseointegrated implants as prosthetic devices.

**References:**


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