Auditory Brainstem Implant

Introduction

Neurofibromatosis type 2 is an inherited condition. It causes noncancerous tumors to grow on nerves. The usual location of these tumors is on the auditory nerves. These nerves transmit information from the inner ear to the brain and allow us to hear. Removing neurofibromatosis type 2 tumors can damage the auditory nerve, leading to deafness. An auditory brainstem implant is a device that transmits sound directly to the brainstem. The implant has two parts: the processor, which is worn near the ear, and a surgically implanted electrode. A microphone picks up sound and the processor turns the sound waves into electrical signals. The electrical signals are sent to an electrode near the brainstem. Medical studies show that auditory brainstem implants are successful in people whose hearing was damaged by surgery to remove tumors that were specifically caused by neurofibromatosis type 2. Using auditory brainstem implants has also been proposed to treat deafness from other types of tumors or other conditions. More studies are needed to see if auditory brainstem implants work for conditions other than neurofibromatosis type 2.

Note: The Introduction section is for your general knowledge and is not to be taken as policy coverage criteria. The rest of the policy uses specific words and concepts familiar to medical professionals. It is intended for providers. A provider can be a person, such as a doctor, nurse, psychologist, or dentist. A provider also can be a place where medical care is given, like a hospital, clinic, or lab. This policy informs them about when a service may be covered.
### Policy Coverage Criteria

<table>
<thead>
<tr>
<th>Implant</th>
<th>Medical Necessity</th>
</tr>
</thead>
</table>
| **Unilateral use of an auditory brainstem implant** | **Unilateral use of an auditory brainstem implant (using surface electrodes on the cochlear nuclei) may be considered medically necessary when ALL of the following criteria are met:**  
- The patient has neurofibromatosis type 2  
- Age 12 years or older  
- The patient became deaf because of 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.** |

<table>
<thead>
<tr>
<th>Implant</th>
<th>Investigational</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Bilateral use of an auditory brainstem implant</strong></td>
<td><strong>Bilateral use of an auditory brainstem implant is considered investigational.</strong></td>
</tr>
<tr>
<td><strong>Penetrating electrode auditory brainstem implant (PABI)</strong></td>
<td><strong>Penetrating (vs. surface electrodes on the cochlear nuclei) electrode auditory brainstem implant is considered investigational.</strong></td>
</tr>
</tbody>
</table>

### Documentation Requirements

The patient’s medical records submitted for review for all conditions should document that medical necessity criteria are met. The record should include the following:

- Office visit notes that contain relevant history and physical supporting:
  - Diagnosis of neurofibromatosis type 2 (the tumors are commonly called vestibular schwannomas or acoustic neuromas)
  - That the member is completely deaf due to surgical removal of neurofibromas on the auditory nerve of both ears

### Coding
Code | Description
--- | ---
**CPT**
92640 | Diagnostic analysis with programming of auditory brainstem implant, per hour

**HCPCS**
S2235 | Implantation of auditory brain stem implant

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**Related Information**


**Definition of Terms**

**Neurofibromatosis type 2:** A rare hereditary condition commonly associated with bilateral vestibular schwannomas, which are benign tumors that occur on the nerves of the inner ear and cause loss of hearing and balance. These tumors are also known as acoustic neuromas.
Consideration of Age

The age listed in the policy statement is based on Food and Drug Administration approval of the auditory brainstem implant device for ages 12 and older.

Evidence Review

Description

An auditory brainstem implant (ABI) is designed to restore some hearing in people with neurofibromatosis type 2 who are rendered deaf by bilateral removal of neurofibromas involving the auditory nerve. ABIs have also been studied to restore hearing for other non-neurofibromatosis indications.

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
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 ≥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 the 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.

Ongoing and Unpublished Clinical Trials

Some currently unpublished trials that might influence this review are listed in Table 1.
Table 1. Summary of Key Trials

<table>
<thead>
<tr>
<th>NCT No.</th>
<th>Trial Name</th>
<th>Planned Enrollment</th>
<th>Completion Date</th>
</tr>
</thead>
<tbody>
<tr>
<td>NCT02310399</td>
<td>Auditory Brainstem Implant (ABI) in Children With No Cochlea or Auditory Nerves</td>
<td>20</td>
<td>May 2020</td>
</tr>
<tr>
<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>
<td>10</td>
<td>Feb 2019</td>
</tr>
<tr>
<td>NCT02630589</td>
<td>Implantation of an Auditory Brainstem Implant for the Treatment of Incapacitating Unilateral Tinnitus</td>
<td>10</td>
<td>Jan 2022</td>
</tr>
<tr>
<td>NCT01864291</td>
<td>Study of the Nucleus 24 and ABI541 Auditory Brainstem Implant in Pediatric Non-Neurofibromatosis Type 2</td>
<td>15</td>
<td>Nov 2022</td>
</tr>
<tr>
<td>NCT01736267</td>
<td>Study of Nucleus 24 Auditory Brainstem Implant (ABI) in Adult Non-Neurofibromatosis Type 2 Subjects</td>
<td>10</td>
<td>Nov 2022</td>
</tr>
<tr>
<td>NCT01904448</td>
<td>An Early Feasibility Study of the Safety and Efficacy of the Nucleus 24 Auditory Brainstem Implant in Children With Cochlear or Cochlear Nerve Disorders Not Resulting From Neurofibromatosis Type II</td>
<td>10</td>
<td>Apr 2023</td>
</tr>
<tr>
<td>NCT02589912</td>
<td>Compassionate Use Arm - ABI541 Auditory Brainstem Implant for Neurofibromatosis Type 2 Patients With Deafness</td>
<td>10</td>
<td>Recruitment closed</td>
</tr>
</tbody>
</table>

NCT: national clinical trial.

Practice Guidelines and Position Statements

*National Institute Health and Care Excellence*

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 brain stem implants appears adequate to support the use of this procedure by surgical teams experienced in this technique.”
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.

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. 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.

References


<table>
<thead>
<tr>
<th>Date</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>05/01/18</td>
<td>New policy, approved April 10, 2018, effective August 3, 2018. Add to Surgery section. This policy was previously archived but it is now being reinstated. Unilateral use of an auditory brainstem implant may be considered medically necessary when criteria are met, considered investigational for all other conditions. PABI and Bilateral use of an auditory brainstem implant are considered investigational. Image added. Added definition for neurofibromatosis type 2.</td>
</tr>
<tr>
<td>09/21/18</td>
<td>Minor update. Added Consideration of Age information.</td>
</tr>
<tr>
<td>05/01/19</td>
<td>Annual Review, approved April 2, 2019. Policy updated with literature review through December 2018; no references added. Policy statements unchanged.</td>
</tr>
</tbody>
</table>

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Email AppealsDepartmentInquiries@Premera.com

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U.S. Department of Health and Human Services
200 Independence Avenue SW, Room 509F, HHH Building
Washington, D.C. 20201, 1-800-368-1019, 800-537-7697 (TDD)

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037338 (07-2016)
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