



BlueCross BlueShield
of Alabama

Name of Blue Advantage Policy:
Auditory Brain Stem Implant

Policy #: 146

Latest Review Date: March 2024

Category: Surgery

BACKGROUND:

Blue Advantage medical policy does not conflict with Local Coverage Determinations (LCDs), Local Medical Review Policies (LMRPs) or National Coverage Determinations (NCDs) or with coverage provisions in Medicare manuals, instructions or operational policy letters. In order to be covered by Blue Advantage the service shall be reasonable and necessary under Title XVIII of the Social Security Act, Section 1862(a)(1)(A). The service is considered reasonable and necessary if it is determined that the service is:

1. *Safe and effective;*
2. *Not experimental or investigational*;*
3. *Appropriate, including duration and frequency that is considered appropriate for the service, in terms of whether it is:*
 - *Furnished in accordance with accepted standards of medical practice for the diagnosis or treatment of the patient's condition or to improve the function of a malformed body member;*
 - *Furnished in a setting appropriate to the patient's medical needs and condition;*
 - *Ordered and furnished by qualified personnel;*
 - *One that meets, but does not exceed, the patient's medical need; and*
 - *At least as beneficial as an existing and available medically appropriate alternative.*

Routine costs of qualifying clinical trial services with dates of service on or after September 19, 2000, which meet the requirements of the Clinical Trials NCD are considered reasonable and necessary by Medicare. Providers should bill **Original Medicare for covered services that are related to **clinical trials** that meet Medicare requirements (Refer to Medicare National Coverage Determinations Manual, Chapter 1, Section 310 and Medicare Claims Processing Manual Chapter 32, Sections 69.0-69.11).*

POLICY:

Blue Advantage will treat **unilateral auditory brain stem implants** that are FDA-approved as a **covered** benefit in carefully selected patients with **neurofibromatosis type 2** who are at least 12 years of age or older and who are rendered deaf due to bilateral resection of neurofibromas of the auditory nerve.

Blue Advantage will treat **bilateral** use of an auditory brainstem implant as a **non-covered benefit** and as **investigational**.

Blue Advantage will treat **upgrades of existing components for next-generation devices** as a **covered** benefit when the patient's existing components are inadequate to the point of interfering with the activities of daily living or the components are no longer functional.

Blue Advantage will treat **upgrades of an existing, functioning external system to achieve aesthetic improvement** such as smaller profile components, or a switch from a body-worn, external sound processor to a behind-the-ear (BTE) model, as a **non-covered** benefit.

Blue Advantage will treat **an auditory brainstem implant** as a **non-covered** benefit and as **investigational** for all other conditions, **including non-neurofibromatosis type 2 indications**.

Blue Advantage will treat **penetrating electrode auditory brainstem implant (PABI)** as a **non-covered** benefit and **investigational**

Blue Advantage does not approve or deny procedures, services, testing, or equipment for our members. Our decisions concern coverage only. The decision of whether or not to have a certain test, treatment or procedure is one made between the physician and his/her patient. Blue Advantage administers benefits based on the members' contract and medical policies. Physicians should always exercise their best medical judgment in providing the care they feel is most appropriate for their patients. Needed care should not be delayed or refused because of a coverage determination.

DESCRIPTION OF PROCEDURE OR SERVICE:

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.

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 the 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.

KEY POINTS:

The most recent literature update was performed through December 18, 2023.

Summary of Evidence

For individuals who are deaf due to bilateral resection of neurofibromas of the auditory nerve who receive an auditory brainstem implant (ABI), the evidence includes a large, prospective case series and a technology assessment that included observational studies. Relevant outcomes are functional outcomes, quality of life, and treatment-related morbidity. The technology assessment found the highest quality evidence for improvement in hearing function, but evidence on other outcomes was lacking. The U.S. Food and Drug Administration 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. A systematic review of 16 studies found that ABI was associated with improved sound recognition and speech perception. Based on these results, ABIs are considered appropriate for the patient population age ≥ 12 years with neurofibromatosis type 2 and deafness following tumor removal. The evidence is sufficient to determine that the technology results in an improvement in the net health outcome.

For individuals who are deaf due to nontumor etiologies who receive an auditory brainstem implant (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, and some older (now obsolete) ABI models have been associated with high rates of device failure and adverse events in this population. In addition, ABI studies have shown inferior outcomes in children with other disabilities. However, ABIs hold promise for select patients when the cochlea or cochlear nerve is absent. Evaluation is currently ongoing with the recently available Nucleus ABI541 to determine its efficacy and durability in children. Thus, further study is needed to define populations that would benefit from

these devices. The evidence is insufficient to determine that the technology results in an improvement in the net health outcome.

Practice Guidelines and Position Statements

National Institute for 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."

U.S. Preventive Services Task Force Recommendations

Not applicable.

KEY WORDS:

Auditory brain stem implant (ABI), neurofibromatosis II, neurofibromas, auditory nerve, penetrating electrode brainstem implant, PABI, Nucleus 24® Auditory Brainstem Implant System, Nucleus ABI541

APPROVED BY GOVERNING BODIES:

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. Nucleus® 24 Auditory Brainstem Implant System (Cochlear Corp.) labeling states: "The efficacy of bilateral implantation with the ABI has not been studied." The Nucleus® 24 is currently obsolete.

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

BENEFIT APPLICATION:

Coverage is subject to member's specific benefits. Group-specific policy will supersede this policy when applicable.

CURRENT CODING:

CPT codes:

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

HCPCS:

S2235	Implantation of auditory brain stem implant
-------	---

REFERENCES:

1. Colletti L, Shannon R, Colletti V. Auditory brainstem implants for neurofibromatosis type 2. *Curr Opin Otolaryngol Head Neck Surg*. Oct 2012; 20(5):353-357.
2. Colletti L, Wilkinson EP, Colletti V. Auditory brainstem implantation after unsuccessful cochlear implantation of children with clinical diagnosis of cochlear nerve deficiency. *Ann Otol Rhinol Laryngol*. Oct 2013; 122(10):605-612.
3. Colletti L. Beneficial auditory and cognitive effects of auditory brainstem implantation in children. *Acta Otolaryngol*, September 2007; 127(9): 943-946.
4. Colletti V, Carner M, Miorelli V, et al. Auditory brainstem implant (ABI): New frontiers in adults and children. *Otolaryngol Head Neck Surg*, July 2005; 133(1): 126-138.
5. Colletti V, Fiorino FG, Carner M, et al. Auditory brainstem implant as a salvage treatment after unsuccessful cochlear implantation. *Otol Neurotol*. Jul 2004; 25(4):485-496; discussion 496.
6. Colletti V, Shannon RV, Carner M et al. Complications in auditory brainstem implant surgery in adults and children. *Otol Neurotol* 2010; 31(4):558-64.
7. Colletti V. Auditory outcomes in tumor vs. non-tumor patients fitted with auditory brainstem implants. *Adv Otorhinolaryngol* 2006; 64: 167-185.
8. Ebinger K, Otto S, Arcaroli J, et al. Multichannel auditory brainstem implant: U.S. clinical trial results. *Journal of Laryngology Otology Supplement*, 2000; 27: 50-3.
9. Food and Drug Administration. Nucleus 24 Auditory Brainstem Implant System. FDA Summary of Safety and Effectiveness. 2000; www.accessdata.fda.gov/cdrh_docs/pdf/P000015B.pdf. Accessed January 25, 2021.
10. Food and Drug Administration. Premarket Approval (PMA). Nucleus ABI541 Auditory Brainstem Implant. 2016. www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfpma/pma.cfm?id=P000015S012. Accessed January 25, 2021.
11. Goffi-Gomez MV, Magalhaes AT, Brito Neto R, et al. Auditory brainstem implant outcomes and MAP parameters: report of experiences in adults and children. *Int J Pediatr Otorhinolaryngol*. Feb 2012; 76(2):257-264.
12. IOM (Institute of Medicine). 2011. *Clinical Practice Guidelines We Can Trust*. Washington, DC: The National Academies Press.

13. Jackson KB, Mark G, Helms J, et al. An auditory brainstem implant system. *American Journal of Audiology*, 2002; 11(2): 128-33. (Abstract)
14. Kaplan AB, Kozin ED, Puram SV, et al. Auditory brainstem implant candidacy in the United States in children 0-17 years old. *Int J Pediatr Otorhinolaryngol*. Mar 2015; 79(3):310-315.
15. Lenarz M, Matthes C, et al. Auditory brainstem implant, part II: Subjective assessment of functional outcome, 2002; 23(5): 694-7. (Abstract)
16. Lenarz T, Moshrefi M, Matthies C, et al. Auditory brainstem implant, part i: auditory performance and its evolution over time. *Otology and Neurotology*. 2001; 22(6): 823-33. (Abstract)
17. Matthies C, Brill S, Kaga K, et al. Auditory brainstem implantation improves speech recognition in neurofibromatosis type II patients. *ORL J Otorhinolaryngol Relat Spec*. 2013; 75(5):282-295.
18. Medina M, Di Lella F, Di Trapani G, et al. Cochlear implantation versus auditory brainstem implantation in bilateral total deafness after head trauma: personal experience and review of the literature. *Otol Neurotol*. Feb 2014; 35(2):260-270.
19. Merkus P, Di Lella F, Di Trapani G, et al. Indications and contraindications of auditory brainstem implants: systematic review and illustrative cases. *Eur Arch Otorhinolaryngol*. Jan 2014; 271(1):3-13.
20. National Institute for Clinical Excellence (NICE). *Interventional Procedure Guidance 108. Auditory brain stem implants*. 2005; //guidance.nice.org.uk/IPG108.
21. Nevison B, Laszig R, Sollmann WP, et al. Results from a European clinical investigation of the nucleus multichannel auditory brainstem implant. *Ear and Hearing*, 2002; 23(3): 170-83. (Abstract) Accessed January 25, 2021.
22. Noij KS, Kozin ED, Sethi R, et al. Systematic review of nontumor pediatric auditory brainstem implant outcomes. *Otolaryngol Head Neck Surg*. Nov 2015; 153(5):739-750.
23. Nucleus ® 24 Auditory Brainstem Implant System. FDA Summary of Safety and Effectiveness. www.accessdata.fda.gov/scripts/cdrh/cfdocs/cftopic/pma/pma.cfm?num=p000015.
24. Ontario Health (Quality). Auditory brainstem implantation for adults with neurofibromatosis 2 or severe inner ear abnormalities: a health technology assessment. *Ont Health Technol Assess Ser [Internet]*. 2020 Mar;20(4): 185. www.hqontario.ca/evidence-to-improve-care/health-technology-assessment/reviews-and-recommendations/auditory-brainstem-implantation-for-adults-with-neurofibromatosis-2-or-severe-inner-ear-abnormalities. Accessed January 14, 2021.
25. Otto S, Brachmann D, Hitselberger W, et al. Brainstem electronic implants for bilateral anacusis following surgical removal of cerebello pontine angle lesions. *Otolaryngologic Clinics of North America*, 2001; 34(2).
26. Otto S, Brachmann D, Hitselberger W, et al. Multichannel auditory brainstem implant: Update on performance in 61 patients. *Journal of Neurosurgery*, 2002; 96(6): 1063-71. (Abstract)

27. Otto SR, Shannon RV, Wilkinson EP, et al. Audiologic outcomes with the penetrating electrode auditory brainstem implant. *Otol Neurotol*, December 2008; 29(8): 1147-1154.
28. Puram SV, Barber SR, Kozin ED, et al. Outcomes following Pediatric Auditory Brainstem Implant Surgery: Early Experiences in a North American Center. *Otolaryngol Head Neck Surg*. Jul 2016; 155(1):133-138.
29. Sanna M, Di Lella F, Guida M et al. Auditory brainstem implants in NF2 patients: results and review of the literature. *Otol Neurotol* 2012; 33(2):154-64.
30. Schwartz MS, Otto SR, Shannon RV, et al. Auditory brainstem implants. *Neurotherapeutics*, January 2008; 5(1): 128-136.
31. Sennaroglu L, Sennaroglu G, Yucel E, et al. Long-term Results of ABI in Children With Severe Inner Ear Malformations. *Otol Neurotol*. Aug 2016; 37(7):865-872.
32. Sennaroglu L, Ziyal I, Atas A, et al. Preliminary results of auditory brainstem implantation in prelingually deaf children with inner ear malformations including severe stenosis of the cochlear aperture and aplasia of the cochlear nerve. *Otol Neurotol*. Sep 2009; 30(6):708-715.
33. Toh E, Luxford W. Cochlear and brainstem implantation. *Otolaryngologic Clinics of North America*, 2002; 35(2).

POLICY HISTORY:

Adopted for Blue Advantage, March 2005

Available for comment May 1-June 14, 2005

Medical Policy Group, August 2006

Available for comment September 1-October 16, 2006

Medical Policy Group, August 2008

Medical Policy Group, August 2010

Available for comment July 23-September 6, 2010

Medical Policy Group, September 2012

Medical Policy Group, December 2013

Medical Policy Group, November 2014

Medical Policy Group, March 2015

Medical Policy Group, September 2016

Medical Policy Group, February 2017

Available for comment March 1 through April 14, 2017

Medical Policy Group, February 2018

Medical Policy Group, February 2019

Medical Policy Group, February 2020

Medical Policy Group, February 2021

Medical Policy Group, February 2022

Medical Policy Group, February 2023

UM Committee, December 2023: Policy approved by UM Committee for use for Blue Advantage business.

Medical Policy Group, March 2024

UM Committee, March 2024: Annual review of policy approved by UM Committee for use for Blue Advantage business.

This medical policy is not an authorization, certification, explanation of benefits, or a contract. Eligibility and benefits are determined on a case-by-case basis according to the terms of the member's plan in effect as of the date services are rendered. All medical policies are based on (i) research of current medical literature and (ii) review of common medical practices in the treatment and diagnosis of disease as of the date hereof. Physicians and other providers are solely responsible for all aspects of medical care and treatment, including the type, quality, and levels of care and treatment.

This policy is intended to be used for adjudication of claims (including pre-admission certification, pre-determinations, and pre-procedure review) in Blue Cross and Blue Shield's administration of plan contracts.