Retrosigmoid Craniotomy for Auditory Brainstem Implantation in Adult Patients with Neurofibromatosis Type 2

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Abstract

Objective To report our technique and experience using a retrosigmoid craniotomy approach for auditory brainstem implantation (ABI) placement in adult neurofibromatosis type 2 (NF2) patients.

Design Retrospective case series.

Setting Single-center study, Boston, Massachusetts, United States.

Participants All NF2 patients who underwent evaluation at Massachusetts Eye and Ear Infirmary and surgery at Massachusetts General Hospital from 2009 to 2013 were reviewed. Six cases of retrosigmoid craniotomy for ABI surgery in five adult NF2 patients were identified. The clinical history, operative course, and outcomes in these patients were reviewed.

Main Outcome Measures Postoperative complications and audiological outcomes.

Results Indications for ABI surgery were profound hearing loss associated with growth or treatment of bilateral vestibular schwannomas. In all cases, a retrosigmoid craniotomy was performed for tumor resection and ABI placement without complication. Electrode placement was confirmed intraoperatively using electrical-evoked auditory brainstem responses. The ABI was activated in the awake patient 4 to 6 weeks postoperatively. Audiological testing was used to evaluate sound detection and speech perception with the ABI. There were no cases of cerebrospinal fluid leak.

Conclusion Retrosigmoid craniotomy is a safe and effective means to provide access to the cochlear nucleus for ABI placement following tumor resection in the adult NF2 patient. Preliminary data indicate that this approach has few complications while offering benefits for hearing. The retrosigmoid craniotomy should be considered a reasonable alternative to the traditional translabyrinthine approach for placement of the ABI in deaf patients who are not candidates for the cochlear implant.

Keywords

- ▶ auditory brainstem implant (ABI)
- ▶ neurofibromatosis type 2 (NF2)
- ▶ retrosigmoid craniotomy

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Introduction

The cochlear implant (CI) is considered to be the most successful neural prosthesis, providing meaningful sound and speech perception in most deaf pediatric and adult users. However, a subset of patients are not candidates for the CI due to anatomical considerations such as auditory nerve injury, cochlear ossification, or congenital hypoplasia/aplasia of the auditory nerve or cochlea. Neurofibromatosis type 2 (NF2) patients represent one challenging group for which CI does not typically work.¹⁻³ NF2 patients carry an autosomal dominant mutation that predisposes them to gliomas, meningiomas, juvenile cortical cataracts, and bilateral vestibular schwannomas (VS). Growth or removal of these schwannomas is often associated with injury or sacrifice of the auditory nerve, rendering these NF2 patients ineligible for the CI to rehabilitate retrocochlear deafness.¹⁻³

The auditory brainstem implant (ABI) was developed in 1979 by Hitselberger and House and first tested and validated in NF2 patients.^{4,5} The ABI electrode bypasses the damaged or absent auditory nerve and directly stimulates the second-order auditory neurons found in the cochlear nucleus (CN).⁶⁻⁹ Most ABI users have improved sound detection that aids in lipreading, with overall outcomes that lag behind CI patients. More recently, the use of the ABI has expanded to include non-NF2 patients who are deaf from cochlear ossification, cochlear nerve avulsion, transection, or injury, as well as children with congenital cochlear or auditory nerve aplasia.^{4,5} Data from Colletti and colleagues suggest that audiological performance among "non-tumor" ABI users (who underwent retrosigmoid craniotomy and electrode placement) is better than NF2 ABI recipients.^{6,9} The reasons for these differences in performance are not known.

ABI surgery is classically performed using a translabyrinthine craniotomy that provides direct access to the posterior fossa for tumor resection and to the lateral recess of the fourth ventricle for ABI electrode placement on or near the dorsal cochlear nucleus. This approach offers a wide view of the internal auditory canal, the cerebellopontine angle, the lower cranial nerves (CNs), and foramen of Luschka as well as access to the entire course of the facial nerve should grafting be needed.⁴ Insertion of the ABI electrode requires the use of landmarks such as the choroid plexus and the glossopharyngeal nerve to provide indirect access to the CN via the lateral recess of the fourth ventricle. Because final placement of the ABI electrode is generally blind due to the relative inaccessibility of the CN, electrode position is guided using intraoperative electrophysiology. Specifically, electrical-evoked auditory brainstem responses (EABRs) are recorded from different electrode pairs in the array to help determine if the array will stimulate the central auditory pathway.

Advantages of the translabyrinthine craniotomy include low rates of postoperative cerebrospinal fluid (CSF) leak and headache,^{10,11} and based on our experience, shorter hospital stays compared with the retrosigmoid approach. The main disadvantage of the translabyrinthine approach is that any residual hearing and the vestibular labyrinth is destroyed. In

the case of deaf non-NF2 ABI recipients, a translabyrinthine craniotomy would eliminate any residual vestibular function.

More recently, there has been substantial interest in retrosigmoid (suboccipital) craniotomy for ABI surgery. Although this approach has been frequently used during ABI surgery abroad for both tumor (NF2) and nontumor patients, it has only recently gained widespread acceptance in the United States for this indication. Retrosigmoid craniotomy offers several advantages including the possibility of hearing and vestibular preservation because the cochlea and vestibular labyrinth are left intact.¹² Thus if a tumor is completely removed in an NF2 patient using the retrosigmoid approach with preservation of acoustically evoked auditory potentials consistent with preservation of hearing, an ABI is not needed. Other benefits of this approach include a panoramic view of larger tumors, direct access to the lateral recess, and improved visualization of lower CNs. Additional operative advantages over the translabyrinthine craniotomy include reduced operative time and a decreased risk of contaminating the wound with microorganisms from the mastoid cavity.⁴

Although the retrosigmoid approach for ABI has been briefly described and reviewed,¹²⁻¹⁴ there are few detailed descriptions of retrosigmoid craniotomy for ABI. In addition, because many of our NF2 patients at Massachusetts Eye and Ear Infirmary and Massachusetts General Hospital undergo an initial trial of medical management, our description captures the unique experiences of NF2 patients who have received biologics such as inhibitors to vascular endothelial growth factor (VEGF) or endothelial growth factor receptor (EGFR) to control tumor growth and stabilize or improve hearing before surgery.¹⁵ In this article, we first describe the retrosigmoid technique for ABI surgery in detail, and then we briefly review our institutional experience with six cases in five adult patients who were anatomically amenable to this approach due to tumor size and location. Together, our experience suggests that retrosigmoid craniotomy for ABI is safe, with few complications and benefits for hearing. Moving forward, retrosigmoid craniotomy may be a valuable surgical technique for placement of ABIs in both NF2 and non-NF2 patients who need hearing rehabilitation.

Methods

Patient Selection

A retrospective review of ABI patients at Massachusetts Eye and Ear Infirmary over the period from 2009 to 2013 was conducted to identify patients who had undergone ABI surgery using a retrosigmoid approach. All patients were evaluated postoperatively with a mean follow-up time of 2.5 years for the patients in this series.

Audiological Testing

Pure-tone audiometry was done according to American National Standards Institute standards for testing and calibration.¹⁶⁻¹⁹ Speech perception was assessed using the Early Speech Perception Test²⁰ and/or the Consonant-Nucleus-Consonant (CNC) monosyllable word test (House Ear Institute). All speech testing used recorded materials. Sound detection thresholds and speech

perception using the ABI were measured in the sound field with the patient sitting at 0 degrees azimuth to the loudspeaker. Warble tones were used for sound field thresholds at 250, 500, 1000, 2000, and 4000 Hz.

Imaging

Imaging protocols have been previously described.^{21,22} In brief, if high-resolution scans were available from an outside institution, patients were not subjected to repeat scanning. However, in most cases, patients underwent magnetic resonance imaging (MRI) using a dedicated protocol including axial and sagittal unenhanced T1-weighted images, axial T2-weighted images, and axial fluid attenuated inversion recovery images through the entire brain as well as high-resolution three-dimensional constructive interference in steady state (CISS) images through the temporal bone. Parameters for the CISS sequence vary by scanner. Total scanning time was ~20 minutes, and conscious sedation was used when needed. High-resolution temporal bone computed tomography (CT) scans were also performed to generate contiguous axial and coronal images through the temporal bones at a thickness of 0.6 or 0.75 mm.

Data Analysis and Institutional Review

All aspects of our study were approved by the Human Studies Committee of the Massachusetts Eye and Ear Infirmary (13-029H).

Results

We begin by describing the surgical technique of retrosigmoid craniotomy for ABI placement and then present our institutional experience with this approach. Because the procedure for vestibular schwannomas removal has been extensively described, we focus our description primarily on placement of the ABI device.

Surgical Technique and Operative Considerations

Before beginning the procedure, CN monitors (typically CN V and VII) and evoked potential equipment needed for intraoperative testing of ABI placement are established. Patients are typically positioned supine with the head (placed in a Mayfield headholder) and ipsilateral shoulder tilted away from the side of surgery. The expected site of the ABI receiver

stimulator superior to the auricle is marked using methylene blue (►Fig. 1A), and the area for nonmagnetic markers is tattooed on the skin (►Fig. 1B). An incision is then made extending posteriorly in the suboccipital area, dividing the musculature. The retrosigmoid craniotomy is completed and then enlarged to reveal the inferior aspect of the transverse sinus and the posterior edge of the sigmoid sinus. We typically do not harvest a pericranial graft to retain full-thickness skin over the area where the internal antenna coil of the ABI will eventually be deposited. The dura is then opened, and microdissection under an operating microscope is used to dissect down to the tumor capsule with minimal cerebellar retraction. We stimulate the capsule to ensure that there is no involvement of the facial nerve. The capsule is then incised with debulking of tumor and dissection of the mass away from the cerebellum and peduncle. After locating the origin of the facial nerve, a combination of capsular dissection and internal debulking is used to dissect along the brainstem. The internal auditory canal is decompressed with a combination of cutting and diamond burrs. The facial nerve is identified in the fundus anterosuperiorly, and the tumor is dissected from the facial nerve and remnant cochlear nerve from lateral to medial toward to the porus acusticus. The remaining tumor fragment in the cistern is then debulked as extensively as possible with facial nerve preservation, and hemostasis is then achieved.

The microscope is then oriented to visualize the lower CNs. The choroid plexus and the glossopharyngeal nerve are used as landmarks to reach the foramen of Luschka. Blunt dissection in this region results in egress of CSF from the fourth ventricle.

To begin placement of the ABI device, the periosteum is elevated off the temporal calvarium to create a subperiosteal pocket for the implant (►Fig. 2A). The silicone implant dummy is then used to mark out the location on the calvarium for the inset well (►Fig. 2B). Cutting and diamond burrs are then used to create a well for the implant, located just superior to the craniotomy cavity (►Fig. 2C). We take great care to avoid getting any bone dust into the intracranial cavity by keeping this area covered with moist gauze during drilling. A small tract for the electrode from the well toward the craniotomy cavity is also drilled (►Fig. 2D).

The ABI receiver stimulator is then anchored by placing a self-drilling 1.5 × 4-mm titanium screw on either side of the

Fig. 1 Marking and planning for auditory brainstem implantation (ABI) device placement using the retrosigmoid approach. (A) The anticipated site of the ABI receiver-stimulator superior to the auricle is marked using methylene blue. (B) The area for nonmagnetic markers is tattooed on the skin as shown.

Fig. 2 Preparation of the bony well for the auditory brainstem implantation (ABI) device. (A) The periosteum is elevated off the temporal calvarium revealing a subperiosteal pocket for the ABI device. (B) The silicone dummy is utilized to mark out a presumptive location on the calvarium for device insertion. (C) A bony well is drilled just superior to the craniotomy site. (D) A small tract is drilled for the electrode, running from the bony well toward the craniotomy site.

implant for tie-down sutures (►Fig. 3A).²³ A 3-0 nylon is tied around each of these screws, and the area is thoroughly irrigated (►Fig. 3B). The magnet is first removed from the receiver stimulator before placement and replaced with the nonmagnetic metallic spacer (►Fig. 3C). This allows for MRI surveillance up to 1.5 T in these NF2 patients who often require follow-up of significant tumor burden. The implant is then placed into the subperiosteal pocket and bony well and seated securely (►Fig. 3D). The approximate location of the center of the telecoil is then tattooed using methylene blue. This allows the clinician and patient to easily find the ABI for

placement of the externally worn speech processor and headpiece that is attached using a magnetic adhesive disk placed on the scalp.

Using an operating microscope, the landmarks of the foramen of Luschka are identified again. The electrode array of the ABI is placed into the lateral recess of the fourth ventricle (►Fig. 4A and B and ►Video 1). In all cases, a Cochlear Nucleus 24 ABI (Englewood, Colorado, United States) was used (the only ABI approved by the Food and Drug Administration [FDA]); this system has 21 stimulating contacts and a ground electrode. Once it is in the presumptive

Fig. 3 Placement of the auditory brainstem implantation (ABI) receiver stimulator. (A) Screws are used to provide an anchor point for tie-down sutures. (B) Nylon sutures are tied to the screws; the area is irrigated. (C) The receiver stimulator is opened, and the magnet is replaced with the nonmagnetic spacer. (D) The implant is then carefully placed into the bony well so that it fits securely within the subperiosteal pocket.

Fig. 4 Placement of the auditory brainstem implantation (ABI) electrode array in the cochlear nucleus. (A) The electrode array is placed into craniotomy site and gently advanced into the lateral recess of the fourth ventricle. (B) The cochlear nucleus is located, and the electrode array is carefully and securely positioned. (C) Proper positioning is confirmed based on electrical-evoked auditory brainstem responses tracings, impedances, and good coverage of the electrodes within the array. (D) A ground electrode is placed in the subperiosteal pocket, and the receiver stimulator is tied down and anchored.

location of the CN, electrical evoked potentials are recorded from selected electrode pairs sampling different areas of the electrode array. The electrode position is often shifted to optimize the number of electrode pairs that elicit responses consistent with stimulation of the auditory pathway and consequently increase the potential number of electrodes eliciting auditory sensations (►Fig. 4C).

Video 1

Video demonstrating placement of the auditory brainstem implantation electrode array into the lateral recess of the fourth ventricle and presumptive location of the cochlear nucleus. Array positioning is confirmed with evoked auditory brainstem responses. 34 seconds. 21 MB. Online content including video sequence is viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0034-154412>

At this point, Dacron mesh associated with the electrode paddle is tucked into the lateral recess of the fourth ventricle followed by Teflon felt to provide stability to the array. The ground electrode is then tucked anterior to the receiver-stimulator in a subperiosteal pocket (►Fig. 4D). Tie-down sutures are tied across the implant to secure its position (►Fig. 4D). A portion of the dura is closed primarily with a graft of DuraGen (Plainsboro, New Jersey, United States) used to fill any remaining dural defect anterior to the electrode. The mastoid is then carefully waxed, and a small titanium mesh cranioplasty is made to occlude the superior portion of the craniectomy. After irrigation and hemostasis, the incision is closed and a sterile dressing applied. EABRs are monitored for

stability of response during closing. Positioning of the electrode and receiver-stimulator is confirmed by CT and X-ray (►Fig. 5A, 5B). The ABI is activated 4 to 6 weeks postoperatively following recovery and healing.

Institutional Experience with Retrosigmoid Craniotomy for ABI Surgery

From 2009 to 2013, we completed six ABI surgeries using a retrosigmoid craniotomy (►Table 1). These patients were all NF2 patients, most of whom had failed medical management with biologics such as VEGF inhibitors and therefore required surgical resection of their progressive vestibular schwannomas. Here we briefly describe their history, operative course, and outcomes.

Case 1

Our first retrosigmoid ABI case was a 66-year-old man with NF2 and bilateral vestibular schwannomas. His history was notable for an ependymoma of the cervical cord and an L3 nerve sheath tumor. Before any surgery, his audiogram indicated a severe sensorineural loss in the left ear and a severe-to-profound sensorineural loss in the right ear. Word recognition scores (CNC, open set monosyllables, House Ear Institute) were 2% in the right ear and 64% in the left ear. In 1999, he had partial resection of his right-sided vestibular schwannomas with preservation of facial nerve function but profound ipsilateral hearing loss across all frequencies tested. He was placed on a trial of medical management with Erlotinib, a biological inhibitor of the EGFR receptor. On this regimen, tumor growth stabilized and his residual hearing on the left stabilized as well; however, after 33 months, treatment was stopped due to growth of both tumors and loss of residual hearing. He underwent retrosigmoid craniotomy on the left with resection of a 4-cm vestibular schwannoma and placement of a Nucleus 24 ABI. Facial nerve

Fig. 5 Confirmation of electrode and receiver stimulator positioning postoperatively. (A) Computed tomography is used to confirm positioning of the electrode array within the anatomical location of the cochlear nucleus. (B) X-ray is used to confirm the secure placement of the receiver stimulator within the subperiosteal pocket.

function was preserved, and he was discharged uneventfully on postoperative day (POD) 6. At activation, the patient had nine electrodes that elicited auditory stimulation without any non-auditory sensations using monopolar stimulation. After 10 months of use, the patient continued to use all nine electrodes and had sound detection thresholds between 30 and 45 dB HL across a wide range of frequencies (**Fig. 6B**). The patient also achieved category 1 on the Early Speech Perception test, that is, pattern perception of speech. Routine follow-up 2 years later revealed a recurrence of his right-sided vestibular schwannoma. He therefore underwent resection of this mass with placement of a second ABI on the right side. Although he had no postoperative complications, he did not return for activation of his right ABI and was lost to follow-up.

Case 2

The patient was a 23-year-old woman with NF2 and bilateral vestibular schwannomas, with a past history notable for a left-

sided cataract surgery and right-sided ptosis due to a right cavernous sinus meningioma. She also had a history of cervical ependymoma, pharyngeal schwannomas, and cutaneous schwannomas. Patient had normal hearing in each ear with good word recognition until 2000 when at age 13, she underwent resection of her left vestibular schwannoma resulting in complete loss of hearing in that ear. Hearing in the right ear was stable until 2009. She was briefly treated with PTC299, an oral VEGF inhibitor, for management of her right-sided vestibular schwannoma, but this was stopped due to tumor growth. An audiogram in 2010 indicated a mild sensorineural hearing loss in the right ear with a CNC word recognition score of 62% (**Fig. 7A**). A trial of intravenous bevacizumab was begun every other week but stopped after a few months due to tumor growth. She therefore underwent retrosigmoid craniotomy with complete resection of a 5.5-cm tumor resection on the right and subsequent placement of a Nucleus 24 ABI. Facial nerve function was preserved, and the patient was discharged

Fig. 6 Case 1 audiogram. (A) Preoperative. (B) Postoperative (17 months) after left-sided auditory brainstem implantation placement (C). ANSI, American National Standards Institute; PTA, pure tone acuity; SDT, speech detection threshold; SRT, speech recognition threshold.

Table 1 Relevant clinical characteristics of five auditory brainstem implantation patients (six total cases) implanted using a retrosigmoid craniotomy

Patient no.	Age, y	M/F	Diagnosis	Relevant history	Prior biological therapy?	Side of surgery/ Tumor size	Device implanted	Postoperative course	Facial nerve function	Audiological performance with ABI
1 (left)	66 (case 1)	M	NF2	Bilateral vestibular schwannomas, cervical ependymoma, L3 nerve sheath tumor	Erlotinib	Left/4 cm	Nucleus 24 ABI	Uneventful; discharged POD 6	Intact	Environmental sound awareness
1 (right)	68 (case 2)	M	NF2	Bilateral vestibular schwannomas, cervical ependymoma, L3 nerve sheath tumor	Erlotinib	Right/3.5 cm	Nucleus 24 ABI	Uneventful; discharged POD 6	Intact	Lost to follow-up
2	23	F	NF2	Bilateral vestibular schwannomas, OS cataract, OD ptosis due to cavernous sinus meningioma	PTC299; Bevacizumab	Right/5.5 cm	Nucleus 24 ABI	Uneventful; discharged POD 5	Intact	Environmental sound awareness
3	20	M	NF2	Bilateral vestibular schwannomas	Bevacizumab	Left/4 cm	None (intra-operative EABRs preserved)	Uneventful; discharged POD 4	Intact	NA
4	53	M	NF2	Bilateral vestibular schwannomas, trochlear nerve schwannoma, multiple falx and frontal meningiomas, schwannomas of cauda equina, right facial palsy with gracilis transfer	None	Right/2.7 cm	Nucleus 24 ABI	Discharged POD 4; returned with seizure, PNA, and PE	N/A	Threshold 30–50 dB across all frequencies; ESP of 62% at 70 dB
5	30	M	NF2	Bilateral vestibular schwannomas, facial nerve schwannoma, peripheral schwannomas including multiple kidney masses	Bevacizumab	Left/2.7 cm	Nucleus 24 ABI	Uneventful; discharged POD 6	Stable House-Brackmann IV/VI	Threshold 15–30 dB across all frequencies; ESP of 88% at 60 dB

Abbreviations: ABI, auditory brainstem implantation; EABR, electrical-evoked auditory brainstem responses; ESP, Early Speech Perception (test); F, female; M, male; NA, not applicable; NF2, neurofibromatosis type 2; OD, oculus dexter (right eye); OS, oculus sinister (left eye); PE, pulmonary embolism; PNA, pneumonia; POD, postoperative day.

Fig. 7 Case 2 audiogram. (A) Preoperative. (B) Postoperative (15 months). ANSI, American National Standards Institute; PTA, pure tone acuity; SDT, speech detection threshold; SRT, speech recognition threshold.

on POD 5. The patient had eight electrodes that elicited auditory sensations using monopolar stimulation at activation but did not wear the device. Even without daily use of the device, she had sound detection thresholds between 50 and 65 dB HL when tested with the ABI (►Fig. 7B). She has subsequently refused to use her device, even though she recognizes that it provides auditory input and, at present, prefers to use text messaging for communication.

Case 3

The patient was an otherwise healthy 20-year-old man with NF2, initially diagnosed in 2005 presenting with hearing loss and subsequently found to have bilateral vestibular schwannomas. In 2007, he underwent a right-sided retrosigmoid craniotomy with 3-cm tumor resection. The tumor filled the internal auditory canal and extended into the cerebellopontine angle. His left vestibular schwannoma was managed conservatively with bevacizumab until 2009, but eventually the tumor grew to 3 × 4 cm, and he developed grade 2 facial weakness. Audiograms indicated no measurable hearing on the right side and a mild to moderate sensorineural hearing loss in the left ear with a word recognition of 92% (CNC). In 2010, he underwent left retrosigmoid craniotomy for tumor resection with planned ABI placement. After most of the tumor was debulked, intraoperative auditory evoked potential testing (auditory brainstem response [ABR]) indicated preservation of the patient's preoperative ABR waveform consistent with hearing preservation. As a result, the ABI was not placed. The patient was discharged on POD 4 with preservation of facial nerve function and reported preservation of hearing on his left side (►Fig. 8). A postoperative audiogram was not done.

Case 4

This patient was a 53-year-old man with NF2 and bilateral vestibular schwannomas. His past medical history was notable for bilateral posterior falcine meningiomas and a left frontal meningioma, as well as multiple schwannomas of the cauda equina. His left vestibular schwannoma was resected in 1989 and 1991 with placement of a cochlear

implant at the time of the first surgery. Due to tumor regrowth and worsening hearing on the left, he underwent radiation therapy in 2011. On the right, he had been previously operated on in 1984 and 1990 using a translabyrinthine and suboccipital craniotomy approach, respectively, which were complicated by facial nerve paralysis. Right facial palsy was managed with a gracilis free transfer graft innervated by the trigeminal in 2010. In 2012, he was found to have a new right trochlear nerve schwannoma that due to progressive growth was managed surgically. Preoperative testing revealed no hearing on the right with limited hearing on the left with his implant (►Fig. 9A). He subsequently underwent retrosigmoid craniotomy with subtotal resection of the trochlear schwannoma and Nucleus 24 ABI placement. He was discharged on POD 4 with stable baseline right-sided facial weakness and some facial movement from his gracilis

Fig. 8 Case 3 postoperative audiogram. ANSI, American National Standards Institute; PTA, pure tone acuity; SDT, speech detection threshold; SRT, speech recognition threshold.

Fig. 9 Case 4 audiogram. (A) Preoperative interrogation of left-sided cochlear implant. Of note, this patient had right-sided anacusis preoperatively. (B) Postoperative assessment of right-sided hearing using auditory brainstem implantation (8 months). ANSI, American National Standards Institute; PTA, pure tone acuity; SDT, speech detection threshold; SRT, speech recognition threshold.

transfer. His postoperative course was complicated by presentation to the emergency department 2 weeks later with a suspected seizure, bilateral pneumonia, and subsegmental pulmonary emboli. These were managed appropriately, and he was eventually discharged. Patient had 12 electrodes that elicited auditory sensation using monopolar stimulation when activated. After 5 months of use, his sound detection thresholds were between 30 and 60 dB HL (► **Fig. 9B**).

Case 5

Our final patient was a 30-year-old man with NF2 and bilateral vestibular schwannomas. His past medical history was notable for a left facial nerve tumor with baseline House-Brackmann deficit of IV/VI, cervical schwannomas, as well as peripheral schwannomas including several masses in his kidneys bilaterally. He was otherwise healthy. He had previously undergone resection of a right vestibular schwannoma in 1999 with resulting right-sided deafness (► **Fig. 10A**). Thereafter, he had experienced subsequent left ear hearing loss, with a 2.7-cm expanding left-sided tumor noted on imaging. Given his worsening symptoms, he had been started on bevacizumab beginning in 2010, but his tumor continued to grow with progression of his hearing loss. Preoperative testing revealed profound bilateral hearing loss (► **Fig. 10B**). He therefore underwent resection of his left vestibular schwannoma with simultaneous placement of a Nucleus 24 ABI using a retrosigmoid craniotomy. His postoperative course was uneventful, and he was discharged on POD 6 with stable left-sided facial weakness. All electrodes elicited auditory sensations at activation using monopolar stimulation. After 8 months of daily use, sound detection thresholds are between 15 and 40 dB across all frequencies (► **Fig. 10C**). Remarkably, this patient had an open set monosyllable score of 12% (CNC).

Discussion

We describe our surgical technique for placement of an ABI using a retrosigmoid craniotomy. This description is meant to

provide guidance to otologic surgeons and neurosurgeons who wish to perform ABI surgery in the future. Notably, the retrosigmoid craniotomy may have some advantages over the translabyrinthine craniotomy, and these differences should be kept in mind when considering what approach to use for ABI placement. We also describe our experience with ABI placement using a retrosigmoid craniotomy. In particular, we detail six cases of ABI surgery using this approach and describe the patients' history, surgical course, and outcomes. In our experience, all implanted patients were able to achieve environmental sound awareness, with most achieving reasonable sound detection across all frequencies tested. It is notable that nearly all the patients described here underwent initial medical management with biological therapy such as VEGF or EGFR inhibitors prior to surgical resection of their tumor and ABI surgery. Thus a trial of biological therapy did not complicate the procedure by increasing bleeding or adversely influence the ability to carry out ABI surgery successfully using a retrosigmoid craniotomy approach.

Although the translabyrinthine approach has traditionally been used for ABI surgery, this approach obliterates the inner ear and does not allow for the possibility of hearing (or vestibular) preservation in patients with reasonable preoperative hearing.¹²⁻¹⁴ Our third case is a good example of how the retrosigmoid craniotomy approach leaves open the possibility of forgoing ABI surgery in a patient with preserved intraoperative ABRs. From a technical standpoint, the retrosigmoid approach provides a panoramic view of larger tumors, good access to the lateral recess, and improved visualization of lower CNs.^{4,24,25} Other benefits may include reduced bleeding, decreased operative time, and decreased risk of wound contamination with tympanomastoid flora.¹²⁻¹⁴ Based on these advantages, several centers now prefer the retrosigmoid craniotomy approach for ABI insertion in the vast majority of patients.^{7,9,26}

Surgeons outside the United States, particularly those in Europe, have been performing ABI surgery in non-NF2 patients, such as those with profound deafness who have

Fig. 10 Case 5 audiogram. (A) Postoperative assessment of hearing after resection of right vestibular schwannoma with resulting ipsilateral deafness. (B) Assessment before auditory brainstem implantation (ABI) after bevacizumab treatment for expanding left-sided schwannoma with progressive bilateral hearing loss. (C) Postoperative assessment of left-sided hearing using ABI (4 months). ANSI, American National Standards Institute; PTA, pure tone acuity; SDT, speech detection threshold; SRT, speech recognition threshold.

not or cannot benefit from a cochlear implant due to cochlear or cochlear nerve pathology such as cochlear nerve aplasia, traumatic injury of the cochlear nerve, cochlear ossification, or cochlear nerve hypoplasia/aplasia. Interestingly, the European experience suggests that surgical and audiological outcomes are more favorable in nontumor NF2 patients compared with NF2 patients.⁶⁻⁹ We are now using the retrosigmoid craniotomy approach for nontumor pediatric and adult patients as part of an FDA-approved study to determine the safety and efficacy of ABI surgery using either the translabyrinthine or retrosigmoid approaches. Future analyses of these cases will be critical to determine if the benefits and disadvantages of the two approaches are similar in nontumor patients and NF2 patients.

Previous studies in the ABI literature are variable in terms of their inclusion of patients with prior medical management using biological agents. To our knowledge, there is no study dedicated to reviewing outcomes following ABI surgery in patients previously treated with agents such as bevacizumab or erlotinib. At our institution, nearly all patients are initially managed medically with VEGF or EGFR inhibitors to determine whether disease progression can be slowed and the

duration of hearing preservation extended. Most of the cases described here were managed with these agents until treatment failure dictated a need for surgery. We found that retrosigmoid craniotomy in these patients was feasible with few complications. Pathologic analyses of these tumors did not reveal any discernible changes in the gross appearance of histology of the tumors compared with those not exposed to biological therapy. In addition, prior biological therapy did not appear to affect the surgical approach adversely or the ability technically to remove the tumor. In the future, it will be critical to perform a more comprehensive retrospective or even prospective analysis of patients who have undergone treatment with biologics before surgery to clarify the impact of such treatment on surgical factors related to ABI surgery.

Our institutional experience with retrosigmoid craniotomy for ABI surgery has been favorable, with reasonable postoperative safety and efficacy outcomes. In all cases, patients were discharged home within 4 to 6 days after surgery, and baseline facial nerve function was preserved. Although our study does not formally compare the translabyrinthine approach with a retrosigmoid craniotomy,

patients in this series achieved reasonable audiological outcomes with no significant postoperative complications. Thus, based on our the cases, retrosigmoid craniotomy for ABI surgery appears safe and effective with no added complications compared with the translabyrinthine approach. In the future, it will be informative to compare surgical outcomes in large patient cohorts to determine definitively the advantages and disadvantages of a retrosigmoid craniotomy compared with a translabyrinthine approach.

Importantly, all of our implanted patients achieved sound awareness upon activation of their ABI with most patients obtaining reasonable thresholds of sound detection across the frequencies tested if the device is worn daily. These audiological outcomes are in line with previously published studies from other institutions that demonstrate auditory sensations in all implanted patients.^{9,11,26} Notably, in two of our five patients, some degree of speech perception was achieved ranging from perception of speech patterns to some open set word recognition, thus conveying the range of audiological outcomes after ABI surgery. Together, the findings from our case series validate the efficacy of retrosigmoid craniotomy for ABI surgery.

Conclusions

- A retrosigmoid craniotomy can be used for placement of ABI devices, with specific procedural steps and surgical considerations that must be kept in mind.
- The retrosigmoid craniotomy approach offers distinct advantages over the more traditional translabyrinthine approach.
- In our institutional experience, ABI surgery using a retrosigmoid craniotomy is safe with few postoperative complications.
- A preoperative trial of biological therapy does not seem to adversely influence the ability to carry out ABI surgery successfully.
- Patients at our institution who have undergone ABI surgery using the retrosigmoid approach have all obtained at minimum sound awareness, with most patients achieving reasonable hearing thresholds across a wide range of frequencies if the device is worn daily.

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