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# Audiometric Outcomes of Auditory Brainstem Implantation during Vestibular Schwannoma Resection in NF2 Patients

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## Abstract

**Background** Many patients with neurofibromatosis type 2 (NF2) suffer from sensorineural hearing loss, and associated cochlear nerve compromise in NF2 patients makes auditory brainstem implant (ABI) an attractive treatment option. The long-term outcomes and benefits of the device are still being explored.

**Methods** A retrospective review was conducted for 11 ABI recipients at a single-institution tertiary center between November 2017 and August 2022. Patients diagnosed with NF2 undergoing resection for concurrent vestibular schwannoma (VS) were included. Pre- and postaudiometric assessments were reviewed. Evaluation included pure-tone audiometry and speech testing.

**Results** Our cohort included 11 patients with a median age of 34 years. All patients underwent a translabyrinthine approach for implant placement with concurrent VS resection. Average tumor size of VS was 2.87 cm. Preoperatively, 8 patients had pure-tone averages with no response at 110 dB, 2 were within mild–moderate hearing loss (25–45 dB), and one patient had a PTA of profound loss (92 dB). Postoperatively, 9 (81%) patients had improvement in PTA. In total, seven patients reported mild side effects upon ABI activation which included dizziness ( $n=2$ ), tinnitus ( $n=1$ ), and abdominal and lower extremity tingling sensation ( $n=3$ ). Of the seven patients with early speech perception (ESP) scores, five had a score >75%, indicating the auditory ability to detect pattern perception upon auditory stimulation through the ABI alone.

**Conclusion** Nine of 11 patients derived benefits from ABI placement. These findings demonstrate that ABI placement during concurrent VS resection can provide a significant hearing benefit for NF2 patients.

## Keywords

- ▶ auditory brainstem implant
- ▶ vestibular schwannoma
- ▶ NF2
- ▶ neurofibromatosis 2
- ▶ audiometric outcomes

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## Background

Neurofibromatosis type 2 (NF2) results from autosomal dominant mutations in the NF2 tumor suppressor gene.<sup>1</sup> This tumor predisposition syndrome is estimated to have a lifetime prevalence of 1 in 25,000 individuals and exhibits a penetrance of 90%.<sup>2</sup> NF2 patients account for 5% of incident vestibular schwannomas (VS), benign growths that can impose a variety of effects including progressive hearing loss (90%) and tinnitus (60%).<sup>3</sup> Auditory brainstem implantation (ABI) was developed in 1979 to address the sensorineural hearing loss associated with NF2 patients.<sup>4</sup> The ABI device consists of a multielectrode surface array surgically implanted along the brainstem to stimulate the cochlear nucleus complex, allowing it to bypass the peripheral auditory system making it a possible option when the cochlear nerve is sacrificed—thus, ABI is often the option over cochlear implantation in NF2 patients, especially as they are less likely to experience audiometric decline with tumor growth and radiation therapy.<sup>5–7</sup> Currently, ABI is primarily used to restore hearing to NF2 patients with VS who experience complete hearing loss because of tumor growth, radiation, and/or surgical resection.<sup>8</sup> ABIs in NF2 patients typically provide sound awareness and may also improve lip reading, with only a portion of patients exhibiting open-set speech recognition.<sup>9</sup>

Long-term outcomes and benefits of ABI are still being explored due to the rarity of NF2, the relative novelty of the ABI surgical procedure, and the evolving technology of ABI devices across recent decades. Current literature regarding ABI procedures consists of a variety of case series, many of which describe procedures that took place when older models of the ABI device existed.<sup>6,7</sup> Audiologically, both aural rehabilitation and postactivation monitoring of these patients are critical to understand the risks and benefits of this complex procedure.

In this single-institution epidemiology cross-sectional study, our objective is to investigate the medical and audiological risks and benefits of this procedure in recent years with newer device models. We hypothesize that ABI demonstrates strong audiological outcomes with minimal negative effects for patients living with NF2, which we define as improvement of pattern perception, auditory awareness, audition, closed-set, and open-set speech testing without deterioration of these abilities. By closely and comprehensively examining the variables involved in this intervention, we hope to better understand the potential risks and benefits of ABI and to inform decision-making about the use of this intervention in patients with NF2.

## Methods

In this study, a retrospective review was performed for adult patients (>18 years old) who received an ABI at a single tertiary care institution activated between June of 2018 and March of 2022. Only patients with a diagnosis of NF2 undergoing resection for concurrent VS were included.

Analyzed patient data included demographic, surgical/clinical, and device characteristics. Surgical/clinical characteristics included surgical approach (retrosigmoid, translabyrinthine, etc.), tumor size, site of ABI placement, dates of ABI placement and activation, extent of follow-up period, as well as any side effects that may have been present at the time of ABI activation. Device characteristics included type of ABI, processor used, and number of electrodes placed.

Additionally, a comprehensive array of audiological outcomes was assessed. Preoperative and postactivation audiology assessments were reviewed and analyzed, with preoperative assessments performed at the time of their initial consultation and postactivation assessment performed at their final follow-up appointment; all audiology assessments involved effective masking of the contralateral ear to prevent confounding results. We did not include data on leads resulting in auditory response at the time of surgery. In the case of the Cochlear Nucleus ABI541 implant used in all these cases, the electrode lead terminates in 21, 0.7-mm platinum disk electrodes arranged in three rows of seven. During surgery, placement of the ABI leads is performed at the anterosuperior surface of the recess of the fourth ventricle. Lead placement resulting in auditory response is evaluated intraoperatively by an audiologist. In our practice, we reposition leads that result in responses from less than 10 electrodes. Such cases typically occur in giant tumors with significant distortion of the brainstem. We do this for three reasons: (1) electrode response typically decreases between surgical implantation and postoperative programming. We believe that (2) pitch perception and (3) long-term auditory perception is best when there is response from greater than 10 electrodes at the time of surgery.

Audiologic evaluations performed pre- and postoperatively include pure-tone audiometry, pure-tone averages (PTAs), early speech perception (ESP), and auditory spondee identification. Open-set and closed-set word lists were used to assess auditory abilities with the use of the ABI. Open-set testing included AzBio Sentences, Hearing in Noise Test, and Consonant-Nucleus-Consonant (CNC) words. Pure-tone behavioral audiometry was used for sound awareness at a variety of different frequencies. Speech testing was performed sequentially from most basic (ESP) to most complex (AzBio) repetition tasks depending on the patient's progress. Auditory, auditory-visual, and visual conditions for CNC were administered to assess advantage of speechreading in comprehension.

## Results

In our cohort of 11 patients, 6 (55%) were male, 9 (82%) were white, and median age was 34 years (see ► **Table 1**). Among this cohort, Patient 2 underwent unilateral VS resection with concurrent ABI placement via two separate operations (denoted by Patient 2a and Patient 2b within the tables). All patients underwent a translabyrinthine approach for implant placement with concurrent VS resection. Average tumor size was 2.81 cm (1.1–4.6 cm) at the time of resection, and 6 of the 12 (50%) were right-sided ABI placements.

**Table 1** Auditory brainstem implant patient characteristics

| Patient | Age (at the time of procedure) | Reason for ABI | Tumor size (cm) | Side of auditory brainstem implant | Auditory brainstem implant | Processor | Follow-up (months) |
|---------|--------------------------------|----------------|-----------------|------------------------------------|----------------------------|-----------|--------------------|
| 1       | 21                             | NF2, AN        | 3.2             | Right                              | ABI 541                    | N7        | 21                 |
| 2a      | 20                             | NF2, AN        | 4.6             | Right                              | ABI 541                    | N7        | 24                 |
| 2b      | 33                             | NF2, AN        | 3.8             | Left                               | ABI 541                    | N7        | 6                  |
| 3       | 34                             | NF2, AN        | 2.5             | Left                               | ABI 541                    | N7        | 5                  |
| 4       | 67                             | NF2, AN        | 2.2             | Left                               | ABI 541                    | N7        | 7                  |
| 5       | 22                             | NF2, AN        | 3.7             | Left                               | ABI 541                    | N7        | 29                 |
| 6       | 56                             | NF2, AN        | 1.9             | Left                               | ABI 541                    | N7        | 13                 |
| 7       | 10                             | NF2, AN        | 2.3             | Right                              | ABI 541                    | N7        | 22                 |
| 8       | 19                             | NF2, AN        | 4.2             | Right                              | ABI 541                    | N7        | 36                 |
| 9       | 69                             | NF2, AN        | 1.1             | Left                               | ABI 541                    | N7        | 8                  |
| 10      | 49                             | NF2, AN        | 2.8             | Right                              | ABI 541                    | N7        | 14                 |
| 11      | 42                             | NF2, AN        | 1.5             | Right                              | ABI 541                    | N7        | 22                 |

Abbreviations: ABI, auditory brainstem implantation; NF2, neurofibromatosis type 2.

**Table 2** Electrodes, side effects, and auditory outcomes

| Number | Hearing perioperatively               | Follow-up (months) | Electrode activation | Side effects at activation                 | Electrode used | Sound                        | Early speech perception |
|--------|---------------------------------------|--------------------|----------------------|--|----------------|------------------------------|-------------------------|
| 1      | Contralaterally normal                | 21                 | 13                   | Dizzy and tinnitus                         | 11             | 25–45 dB HL for 250–8,000 Hz |                         |
| 2a     | Contralaterally ABI, bilaterally deaf | 24                 | 10                   | –  | 10             | 30–40 dB HL for 250–4,000 Hz |                         |
| 2b     | Contralaterally ABI, bilaterally deaf | 6                  | 18                   | Spasm, wave down legs, paralyzing wave     | 14             | 20–40 dB HL for 250–6,000 Hz |                         |
| 3      | Contralaterally severe hearing loss   | 5                  | 8                    | Tingling on left                           | 7              | 45–60 dB HL for 125–1,000 Hz | 70                      |
| 4      | Bilaterally deaf                      | 7                  | 9                    | None                                       | 9              | 25–45 dB HL for 250–8,000 Hz | CNT                     |
| 5      | Contralaterally severe hearing loss   | 29                 | 17                   | None                                       | 17             | 25–40 dB HL at 250–8,000 Hz  |                         |
| 6      | Contralaterally normal                | 13                 | 10                   | None                                       | 10             | 20–30 dB HL for 250–8,000 Hz |                         |
| 7      | Contralaterally normal                | 22                 | 19                   | None                                       | 19             | 25–30 dB HL for 250–8,000 Hz |                         |
| 8      | Contralaterally normal                | 36                 | 7                    | Yes, unspecified                           | 7              | 25–45 dB HL for 250–3,000 Hz |                         |
| 9      | Contralaterally severe hearing loss   | 8                  | 19                   | Dizziness                                  | 19             | 15–30 dB HL for 250–4,000 Hz |                         |
| 10     | Contralaterally moderate hearing loss | 14                 | 11                   | None                                       | 11             | 20–45 dB HL for 250–8,000 Hz |                         |
| 11     | Contralaterally moderate hearing loss | 22                 | 9                    | Nonauditory sensation down abdomen and leg | 9              | 55–75 dB HL at 250–2,000 Hz  |                         |

Abbreviation: ABI, auditory brainstem implantation.

Placement dates ranged from March 2018 to August 2021, and activation dates ranged from June 2018 to March 2022, with the average time between ABI placement and activation set at 5.4 months. Extent of follow-up ranged from 5 to 36 months with a mean of 18 months. Upon activation of ABI, 7 of 11 (64%) patients reported mild side effects which included tinnitus ( $n=1$ ), dizziness ( $n=2$ ), and abdominal and lower extremity tingling sensation ( $n=3$ ). All patients were implanted with the ABI 541 model and N7 processor, and the number of electrodes returning a response at postoperative audiologic follow-up ranged from 7 to 19 out of 21, with a mean of 12 (see ►Table 2). As for preoperative audiologic assessments, 8 of the 11 (73%) patients had PTAs with no response at 110 dB. Two of the 11 patients had a PTA within mild–moderate hearing loss (25–45 dB) and one patient had a PTA of profound loss (92 dB). Postoperatively, 9 of 11 (81%) had improvement in PTAs from profound and no response levels to mild–moderate hearing loss in response to auditory stimuli providing access to sound. One patient with a mild PTA remained unchanged. One patient had a worsening of PTAs from a moderate level to a severe PTAs. Of the 7 patients with ESP scores, 5 (71%) had a score greater than 75%, indicating the auditory ability to detect pattern perception upon auditory stimulation through the ABI alone (see ►Table 3).

In our study, ABI patients tested with CNC words demonstrated improvement in all conditions between first appointment and follow-up. Improvement for the auditory-alone condition was between 7 and 29% whereas visual-alone condition revealed an improvement of 1 to 24%. The greatest improvement was seen in the auditory-and-visual condition with an improvement of 20 to 30%. The lowest score at the first appointment was 20%. As for AzBio sentences, the visual-alone condition revealed a low score of 4% but improved to 41% when tested in the auditory–visual condition.

The one patient with sufficient open-set quiet performance to warrant testing in noise scored 84% in the auditory conditions and 100% in the auditory visual condition (see ►Table 4).

## Discussion

This study reports the largest cohort of adult NF2 patients fitted with ABI in 6 years at a single institution, with the goal of contributing to a growing body of literature on the potential benefits and risks of ABI for NF2 patients.<sup>8,10–12</sup> At this time, NF2 is in an age of medical trials and ABI continues to be an exceedingly rare procedure; thus, the importance of volume cannot be understated in achieving dual goals of outcome reporting among the scientific community and, more importantly, optimized patient outcomes. Our results demonstrate that NF2 patients can experience significant improvements in hearing after ABI placement during concurrent VS resection. Postoperatively, 9 of 11 patients derived benefits from their ABI placement, demonstrating improvements in PTAs from preoperative levels. Additionally, five of seven patients with ESP scores were able to detect pattern perception upon auditory stimulation through their ABI alone. Still, two patients had variable outcomes: one patient whose PTAs remained unchanged and one whose worsened. There are several underlying factors contributing to the variability in ABI outcomes, including patient-related factors such as patient age, social support, tumor size, duration of deafness, history of radiation, or the presence of surgical landmarks.<sup>13</sup> Improvement in ABI outcomes have been shown to occur over time, so it should be acknowledged that, because the time interval for the postoperative audiologic assessment varied by patient in this study, its findings may underestimate the audiologic improvement post-ABI placement for some individuals.<sup>10</sup> It

**Table 3** Additional auditory outcomes

| Number | Auditory pattern perception | Auditory spondee identification | Auditory monosyllabic | Auditory visual pattern perception | Auditory visual spondee identification | Auditory visual monosyllabic identification | Visual pattern perception |
|--------|-----------------------------|---------------------------------|-----------------------|------------------------------------|--|---|---------------------------|
| 1      | 75%                         | 13%                             | –                     | 92%                                | 96%                                    | 63%   | 92%                       |
| 2a     | 88%                         | 70%                             | –                     | –                                  | –                                      | –   | 100%                      |
| 2b     | –                           | –                               | –                     | –                                  | –                                      | –   | –                         |
| 3      | –                           | –                               | –                     | –                                  | –                                      | –   | –                         |
| 4      | –                           | –                               | –                     | –                                  | –                                      | –   | –                         |
| 5      | 71%                         | 17%                             | –                     | –                                  | –                                      | –   | –                         |
| 6      | 100%                        | 58%                             | 46%                   | –                                  | –                                      | –   | –                         |
| 7      | 100%                        | 92%                             | 83%                   | –                                  | –                                      | –   | –                         |
| 8      | 54%                         | –                               | –                     | –                                  | –                                      | –   | 70%                       |
| 9      | 63%                         | –                               | –                     | 100%                               | 83%                                    | 21%   | –                         |
| 10     | 88%                         | 29%                             | –                     | –                                  | –                                      | –   | 88%                       |
| 11     | –                           | –                               | –                     | –                                  | –                                      | –   | –                         |

**Table 4** Additional auditory outcomes

| Number | Visual monosyllabic identification | CNC Auditory (recorded) | CNC Auditory visual (monitored live voice) | CNC visual | AzBio Sentences MLV, auditory visual | AzBio Sentences MLV, visual | HINT audio | HINT audiovisual |
|--------|------------------------------------|-------------------------|--|------------|--------------------------------------|-----------------------------|------------|------------------|
| 1      | 63%                                | –                       | –  | –          | –                                    | –                           | –          | –                |
| 2a     | 93%                                | 0%, 13%                 | 58%, 77%                                   | –          | 42%                                  | –                           | –          | –                |
| 2b     | –                                  | –                       | –  | –          | –                                    | –                           | 84%        | 100%             |
| 3      | –                                  | –                       | –  | –          | –                                    | –                           | –          | –                |
| 4      | –                                  | –                       | –  | –          | –                                    | –                           | –          | –                |
| 5      | –                                  | –                       | –  | –          | –                                    | –                           | –          | –                |
| 6      | –                                  | 8%, 28%                 | 40%, 63%                                   | 4%, 29%    | 41%                                  | 4%                          | –          | –                |
| 7      | –                                  | 0%, 7%                  | 20%, 51%                                   | 18%, 41%   | –                                    | –                           | –          | –                |
| 8      | 33%                                | –                       | –  | –          | –                                    | –                           | –          | –                |
| 9      | –                                  | –                       | –  | –          | –                                    | –                           | –          | –                |
| 10     | –                                  | 4%, 29%                 | 20%, 42%                                   | 0%, 1%     | –                                    | –                           | –          | –                |
| 11     | –                                  | –                       | –  | –          | –                                    | –                           | –          | –                |

Abbreviation: CNC, consonant-nucleus-consonant; HINT, Hearing in Noise Test; MLV, monitored live voice.

is unclear why Patient 2 displayed such successful outcomes; two possible explanations are that this individual was highly motivated to maximize their recovery and thus underwent an extensive aural rehabilitation lasting several hours per day, and that this patient received bilateral ABI, either or both of which may have contributed to the audiologic success. Approaches to and outcomes of ABI placement in NF2 patients represent a growing point of interest among stakeholders within the past few decades, and the exact precautions and indications of the procedure for NF2 patients and nontumor patients are continuing to be elucidated. Studies affirm that cochlear implant (CI) is a favorable option for hearing rehabilitation for a subset of NF2 patients with an intact cochlear nerve and may provide the NF2 patient with improved communication skills and potential enhanced open-set speech communication.<sup>4</sup>

There are additional factors related to the device and device placement that may contribute to variation in outcomes such as differences in surgical technique, level of surgeon experience, postimplant programming, signal-coding strategies, and postactivation follow-up treatment.<sup>14</sup> Additionally, device migration may occur postoperatively, contributing to further unpredictable variability in the performance of the device. Overall, while the potential benefits for patients undergoing ABI can be significant, the results can also be unpredictable, speech recognition may be poor, and nonstimulation may occur. Due to the unpredictable nature of ABI outcomes, it is not only critical to determine ABI candidacy on a case-by-case basis but also to carefully track auditory responses after ABI placement and activation.

To this effect, tracking auditory development after ABI activation begins with sound awareness which is commonly assessed using questionnaires and pure-tone audiometry. Pure-tone audiometry is used to indicate the average

pure-tone threshold over a specific frequency range and clinically, this measure is used to describe the degree of hearing loss an individual has. For patients with CIs, these thresholds should be between 30 and 45 dB due to the limited dynamic range. Previous literature assigns 45 dB as the postoperative soundfield PTAs of CI recipients.<sup>15</sup> The results from our study show that 8 of the 11 patients had no responses at 110 dB pre-ABI placement. Of these 11 patients, 9 had postoperative aided soundfield thresholds measured to be between 15 and 45 dB and thus demonstrated improvement from preoperative PTAs. The ABI was seen to provide auditory stimulation in patients that were otherwise unable to perceive sound preoperatively.

Furthermore, speech comprehension for both normal hearing and hearing-impaired individuals is a multifaceted process that relies on both visual and auditory information. These visual cues in speech are a major contributing factor for speech recognition in patients with hearing loss, and previous literature has found patients with ABIs had significant improvement in speech reading as compared with their baseline, though outcomes may vary.<sup>16,17</sup> Our results are consistent with prior studies that have found improvements in comprehension when speech reading and additional visual cues are provided, and especially so in the case of ABI recipients.<sup>17–19</sup> Furthermore, they provide evidence that ABI is successful in aiding aural and verbal communication when both auditory and visual modalities are utilized.

Audiometrically, ABI recipients had variable outcomes with 9 of 11 experiencing an improvement in pattern perception, auditory awareness, audition, closed-set, and open-set speech testing. These findings, along with Patient 2 scoring 100% in the auditory visual condition, provide promising results of advancements in ABI, as previous literature has found that open-set speech perception was limited

among ABI patients as compared with CI patients.<sup>18</sup> It is important to note that of the patients tested in auditory-alone, visual-alone, and auditory-visual conditions, the auditory-visual group showed a great improvement in speech recognition scores compared with the visual-alone condition. Lip reading or, the use of these auditory visual cues has been seen to improve with ABI use.<sup>9,20–22</sup> Improvement of auditory responses is seen up to 8 years postactivation.<sup>21</sup> This is important when looking at an ABI recipient's communication potential. Consideration of the patient's sound-field thresholds is crucial because they provide valuable information regarding the access to sound the patient may have and thus influences their ability to respond appropriately and effectively to auditory stimuli. Pattern perception, auditory awareness, audition, closed-set, and open-set speech testing give insight into a patient's auditory abilities and provide insight to what they can achieve. This testing also allows for patient progress to be monitored with ABI use.

There is a growing body of knowledge of the indications and contraindications of ABI placement for patients with NF2. While many of our patients derived benefits from ABI placement, further investigation with larger patient volumes must be dedicated toward understanding ABI outcomes especially as influenced by factors such as patient etiology, surgical methods, electrode placement, or device specifications. Additionally, the safety and effectiveness of the device is being explored for patients with conditions other than NF2, such as postmeningitis hearing loss and cochlear nerve aplasia, or in the cases of unsuccessful CI implantation.<sup>23,24</sup> In our study, tumor size did not demonstrate an association with ABI performance postactivation, and previous studies have arrived at the same conclusion among both NF2 and non-NF2 patients.<sup>13,14</sup> Despite there not being an observable impact on tumor size among our population, the presence of tumor or lack thereof has interestingly been linked to a difference in outcomes in prior studies, with nontumor patients observed to have better outcomes than some NF2 patients<sup>9</sup> and thus, this may serve as a starting point for further uses for ABI among a broader population of patients in need of hearing restoration and rehabilitation.

This study should be considered within the context of its limitations. One limitation is the inconsistent methodology utilized to assess audiologic capabilities of patients pre- and postoperatively. Additionally, postoperative audiologic monitoring with the above-described testing is limited by the patient's audition—if a patient is unable to perform pattern perception, they will be unable to proceed with further, increasingly complex audiological testing which leads to inconsistent monitoring of progress and success.

## Conclusion

In total, 9 of the 11 NF2 patients benefited from ABI placement. These findings demonstrate that the placement of an ABI during concurrent VS resection can provide various significant hearing benefits for an NF2 patient.

## Conflict of Interest

None declared.

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