INTRODUCTION
The auditory brainstem implant (ABI) for patients was originally developed for patients with neurofibromatosis type 2 (NF2) in order to electrically stimulate the cochlear nucleus complex. Drs. William House and William Hettleberger first used the ABI in such a patient in 1979, who continues to use it daily. Patients with NF2 usually have bilateral vestibular schwannomas (VS) necessitating tumor removal, which often results in profound deafness, and cochlear implants (CIs), which electrically activate peripheral neural processes within the cochlea, are usually not an option for patients with NF2 because of their loss of integrity of the auditory nerve. The ABI is introduced into the lateral recess of the fourth ventricle and placed over the area of the ventral and dorsal cochlear nuclei after tumor removal. The ABI is similar in design and function to multichannel CIs, except for differences in the design of the stimulating electrode arrays. Multichannel CIs and ABIs were developed to capitalize on the frequency tuning of neurons in the human cochlea and cochlear nucleus complex, respectively. The programming of ABI devices, however, differs in several important aspects from CI programming.

In multichannel CIs, the electrode is placed into the cochlea. Consistent placement of the electrode carrier and its depth of insertion are assured in normal cochleas. However, in ABI recipients, anatomical landmarks that are used in electrode array placement may be altered or obscured due to the presence of tumors making electrode array placement more challenging. This chapter describes the surgical anatomy of the cochlear nucleus complex, our experience, and results with ABI placement in individuals with and without NF2.

NEUROFIBROMATOSIS TYPE 2
NF2 is an autosomal dominant condition. Bilateral acoustic neuromas (VS) are the hallmark of this disease and are pathognomonic for NF2. The diagnostic criteria for NF2, redefined by the U.S. National Institutes of Health Consensus Development Conference, are met if a person has (1) bilateral eighth-nerve masses seen with computed tomographic scanning or magnetic resonance imaging (MRI) or (2) a first-degree relative (parent or sibling) with NF2 and either unilateral eighth-nerve masses or one of the following: neurofibroma, meningioma, glioma, schwannoma, or juvenile posterior subcapsular lenticular opacity. The incidence of NF2 is one in 33,000–40,000 live births. Patients with NF2 present complex and challenging management problems. Previously, delayed diagnosis, growth of tumor, or surgical removal of the acoustic tumors usually resulted in total hearing loss. Early diagnosis using gadolinium-enhanced MRI and refinements in hearing preservation surgery has improved our ability to prevent total hearing loss while achieving complete tumor removal. Appropriate family screening and DNA analysis have helped early diagnosis. For patients with larger tumors or no useful hearing, the ABI allows restoration of some auditory function when the tumor is removed.
**PATIENT SELECTION**

The nucleus ABI (Cochlear Corporation, Centennial, CO) (Fig. 34.1) is approved by the US Food and Drug Administration (FDA) for implantation at the time of VS removal. Suitable candidates are patients undergoing translabyrinthine VS removal who have (1) nonaidable hearing or an only-hearing ear with a symptomatic tumor or (2) serviceable hearing in the contralateral ear but a contralateral tumor of sufficient size to indicate that hearing will likely be lost in a short period. Criteria for patient selection for receiving an ABI are as follows: evidence of bilateral eighth-nerve tumors, competency in the English language, age 12 years or older, psychological suitability, willingness to comply with the follow-up protocol, and realistic expectations.

While there are some exceptions, the large majority of patients to have received the ABI at the House Clinic have NF2 and bilateral acoustic neuromas. In these patients, the goal is to restore some auditory function in order for these individuals to continue to be a part of the hearing world and to improve their quality of life. The ABI may be implanted during removal of either the first- or second-side tumor, even if some hearing remains on the other side, which is often the case. This approach allows patients to become familiar with the use of the device and prepares them for when all hearing is lost.\(^9\)\(^{-11}\)

More recently, other potential indications for ABI placement have developed. Postlingually deafened adults with cochlear ossification following meningitis or with cranial trauma resulting in bilateral cochlear nerve transection or avulsion have been implanted. In Europe, congenitally deaf young children who are deemed not to be candidates for CI due to severe cochlear malformation or cochlear nerve aplasia have been implanted with very good results. FDA-approved trials of patients with these diagnoses are now beginning in the United States.\(^12\)\(^{-14}\)

**SURGICAL TECHNIQUE AND ANATOMY OF THE COCHLEAR NUCLEUS**

The cochlear nucleus complex (dorsal and ventral cochlear nuclei) lies in the lateral recess of the fourth ventricle. It is partially obscured by the cerebellar peduncles. A surface electrode array introduced in the lateral recess crossing the taenia chooroidea will stimulate viable cochlear nucleus structures.

At the House Clinic, we have almost exclusively used the translabyrinthine approach for placement of the ABI. Typically, we use a C-shaped incision that starts behind the pinna and is approximately 2 cm away from the postauricular fold at the level of the mastoid tip, as shown in Figure 34.2. It allows the placement of the internal receiver and magnet under the scalp. It is important that the incision not directly cross the area of the receiver/stimulator.

The translabyrinthine approach provides direct access to the cochlear nuclei. The jugular bulb is skeletonized to provide the widest access to this area (Fig. 34.3). Anatomical landmarks used for placement include the stump of the eighth nerve, the glossopharyngeal nerve, the facial nerve, and the taenia chooroidea as well as the mouth of the later-

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**Fig. 34.1:** Nucleus auditory brainstem implant.

**Fig. 34.2:** C-shaped incision currently used for the placement of an auditory brainstem implant (ABI). The incision starts behind the pinna and is approximately 2 cm away from the postauricular fold at the level of the mastoid tip.
Auditory Brainstem Implants

al recess where all of these structures converge (Fig. 34.4). Two features frequently used to identify the lateral recess are its relationship to the ninth nerve and the position of the jugular bulb. In the surgical setting where there is almost always distortion of the brain stem from the tumor, the lateral recess is superior to the ninth nerve. The ninth nerve is generally in a fixed anatomic position, and going from there, the lateral recess may be identified in almost every case. The jugular bulb is important because its position may vary. Indeed, with a contracted mastoid and a high jugular bulb, the exposure may be more difficult although it should not be an impediment to placement of the ABI electrode.

Location of the ventral cochlear nucleus, the main target for placement of the ABI, can be problematic. After clearly identifying the basic anatomic landmarks of the lateral recess, including the choroid plexus and the more reflective ependymal surface, dissection is temporarily stopped and the posterior fossa is occluded with Gelfoam. At this point, a subgaleal pocket is created superior and posterior to the mastoidectomy site. A seat is then drilled in the outer table of the skull to secure the position of the ABI receiver. Next, dissection continues in the posterior fossa with the ABI on the operative field. After placing the ground electrode under the temporalis muscle, the ABI electrode array is carefully inserted into the lateral recess. Correct anatomic placement is confirmed using electrophysiological monitoring. Electrically evoked auditory brainstem responses are elicited by stimulation of the nucleus, and the position of the ABI electrode array is optimized using information derived from electrophysiological monitoring, as determined by an experienced auditory physiologist. In addition to facial nerve monitoring, the lower cranial nerves are also monitored to avoid side effects and non-auditory sensations.

Once the optimal position is determined, Teflon felt or muscle is used to secure the electrode in the lateral recess of the fourth ventricle. In patients with NF2 who will require frequent MRIs, the magnetic disc in the receiver/stimulator is replaced with a nonmagnetic plug. The receiver is then placed in the subgaleal pocket prior to filling the mastoid cavity with abdominal fat, carefully inserting a titanium mesh cranioplasty, and closing the scalp (Fig. 34.5).

In other centers, the retrosigmoid approach has been used to implant ABIs with similar success. Both supine/lateral and semi-sitting positions have been used. The latter may offer some benefit in terms of brain relaxation and ease of access to the lateral recess. In young children, the retrosigmoid approach is used universally.

In the case of the previously investigational penetrating ABI (PABI), after establishing landmarks and identifying the cochlear nuclei, the penetrating electrode was placed first into the ventral portion of the nucleus, which is then followed by placement of the surface electrode as in a regular ABI surgery (Fig. 34.6).
Implantation is facilitated by preservation of landmarks during tumor resection. Great care is also taken during exposure of the lateral recess, which may be obstructed by arteries, veins or a thin membrane, and during device placement. Very gentle manipulation of brainstem, tumor, and surrounding neurovascular structures may lead to improved audiologic results. With the largest tumors, this may be very difficult or impossible.

**DEVICE**

We use the current-generation nucleus ABI manufactured by Cochlear Corporation. This device, which is the only one approved for commercial use by the FDA, consists of 21 electrodes embedded in a silicone carrier that is fixed to a fabric mesh, connected to an implantable internal receiver/stimulator. A competing device manufactured by the MedEl Corporation is of similar construction and has an array of 12 electrodes. This device has not been approved by the FDA but is in use in Europe and elsewhere. The external equipment for both devices consists of an external transmitter coil held in place by a tape and metal disk placed on the scalp over the receiver/stimulator coil and connected to a microphone and sound processor, which contains the battery power source. All of this is similar to a CI. As long as the magnet is removed from the implanted receiver/stimulator, follow-up serial MR imaging scans (1.5 Tesla maximum) can be obtained with minimal artifact. The MedEl implant does not have a removable magnet, and therefore further MR imaging requires externally securing the implant in place. Presence of the magnet may further affect ability to assess ipsilateral disease, and coronal MR imaging may be more able to assess the cerebellopontine angle in such situations.\(^\text{16}\)

The sound processing unit (speech processor) requires appropriate programming and must be fitted to individual users. Programming speech processors involves psychophysical assessment of electrically induced auditory (and nonauditory) percepts including threshold, comfort level, and pitch. The outcomes of these measures are used to program the sound processor appropriately and control the amplitude and the sequential patterns of stimulation.

In multichannel auditory implants, different sites of stimulation can often generate different pitch percepts for the listener. Changes in the frequency spectrum of sound can therefore be coded by appropriate changes in the patterns of electrode activation. CIs can usually employ a relatively standard pattern of neural stimulation because of the homogeneous tuning of neurons in the cochlea. ABI recipients, however, have variations in brainstem anatomy, electrode array placement, and tumor effects that require the use of more individualized stimulus patterns to code frequency cues and manage any nonauditory sensations. Therefore, special techniques and additional time are usually required to program ABI sound processors.

Initial testing and activation of the ABI is typically carried out 1–2 months after the surgery. Any nonauditory sensations are reduced or, if possible, eliminated by altering the electrical parameters of stimulation (particularly pulse duration and reference ground electrode). Nonauditory sensations have included dizziness, sensation of vibration in the eye, throat sensations, and ipsilateral...
tingling sensations in the head or body. Generally, non-auditory sensations have decreased in magnitude when longer pulse duration stimuli are used.

## RESULTS

Auditory outcomes and speech perception performance have generally improved significantly since initial development of the ABI. The first 25 patients implanted prior to 1992 at the House Research Institute (HRI) received a single-channel system. Since 1992, we have used the nucleus multichannel ABI device that has resulted in improved performance. The nucleus multichannel ABI completed clinical trials and received approval from the FDA for commercial release on October 20, 2000.

A number of articles have been published detailing results obtained with the ABI. To date, more than 280 patients have been implanted at HRI with more than 1250 total recipients worldwide. The safety of this device has been comparable to the safety of CIs. At our institution, only two patients (early users of the “single-channel” ABI) were explanted due to infection, and this was likely related to the percutaneous connector in use at the time rather than the ABI electrode array.

Presently, 80% of the patients are device users, and 92% have received auditory sensations from their ABIs. Approximately 25% of ABI users have achieved open-set speech discrimination (at least 20% correct without lipreading cues on the CUNY Sentence Test). Ten of our patients have scored 65% or better, and three patients have scored 82% or better on this test. The majority of patients recognize a high percentage of environmental sounds, and speech understanding ability has enhanced an average of 35% when ABI sound is combined with lipreading. This enhancement has reached as high as 75% in some individuals.

Initially, most of our patients with NF2 were implanted at the time of surgery to remove second side VS, but in the mid-1990s application was made to FDA to begin implanting when first side acoustic tumors were removed. This can assist in easing the transition of patients to hearing exclusively with the ABI after becoming completely deaf. Now, about a third of our patients have received ABIs on their first tumor sides, and their ABIs have been activated soon after surgery. Even though useable hearing may remain on the second-tumor side, this experience has resulted in as much as a 25% “headstart” in speech perception when patients ultimately become completely reliant on ABI sound. Most first-side recipients have commented on practical advantages of this approach, including avoiding any substantial period of complete deafness, being able to remain on the job, and maintaining some ability to hear and monitor their children’s activities.

Since about 9% of our NF2 cases do not ultimately obtain hearing sensations from their ABIs, another advantage of first-side implantation is that it can provide a valuable second opportunity to achieve a hearing result when needed. Usually, the second opportunity has resulted in beneficial hearing outcomes; however, in about 20% of these patients, the second ABI did not provide substantially better outcomes than the first. This suggests that whatever affected outcomes on the first side can also influence outcomes on the second side.

In our NF2 cases, we have found that very large acoustic tumors can sometimes distort or damage brainstem anatomy, complicate device implantation, and impact ABI outcomes. Some treatments of acoustic tumors, including surgery, also may affect the stimulability of brainstem auditory structures. For example, we have noted that patients with a history of “gamma knife” radiation therapy for acoustic tumors have had a higher rate of not getting hearing from their ABIs than patients without this history (about 30% vs. 9%). However, there are many examples of patients with large acoustic tumors, or a history of gamma knife, who have experienced great benefit from their ABIs.

## EXPANDED APPLICATIONS

### FOR THE ABI

The PABI, developed at HRI and Huntington Medical Research Institute (Pasadena, CA) in collaboration with Cochlear Corporation (Centennial, CO), was an effort to improve the efficiency of the ABI through microstimulation with needle electrodes, and to increase access to the subsurface tonotopic organization of the ventral cochlear nucleus. We hoped that this would contribute to higher levels of speech understanding. The PABI was studied in clinical trials under auspices of the FDA. Patients had both needle electrode arrays and conventional surface arrays.

Ten patients were implanted, and we found that microstimulation resulted in hearing sensations at much lower electrical levels than conventional surface electrodes, that a wide range of pitch percepts could be elicited depending on the location and depth of the penetrating electrodes, and that sound quality and speech perception performance were better with sound processor maps that used a combination of penetrating and surface electrodes. In two PABI recipients, we also found that stimulation on longer
electrodes (2 mm in length) could result in uncomfortable facial sensations, possibly related to activation of trigeminal tracts. Unfortunately, microstimulation with the PABI alone did not substantially improve speech recognition over the conventional surface electrodes. This may have been due to difficulty in identifying the precise location of the subsurface cochlear nucleus structures in NF2 patients with distorted brainstem anatomy, and thus obtaining a high percentage of useable penetrating electrodes. Once placed, the position of the penetrating array could not be adjusted if needed. Clear EABR waveforms also could not be observed when recording from scalp electrodes to assist with accurate placement of the penetrating array. Therefore, the PABI is not in current clinical use. Given their demonstrated advantages, however, newer penetrating electrode array designs are continuing to be studied.

While the original purpose of the ABI was to treat deafness in adults with NF2, we anticipated that it could be useful in treating deafness in non-NF2 cases without viable auditory nerves or implantable cochleas. Recently, many of these cases have been implanted, primarily in Europe. Positive outcomes have led to renewed interest in the ABI as an effective treatment of deafness resulting from traumatic transection or avulsion of the auditory nerve, cochlear ossification after meningitis, and congenital cochlear aplasia/agenesis.

Such non-NF2 cases, both adult and pediatric, were first implanted by Vittorio Colletti in Verona, Italy. While outcomes have been variable, in general results have exceeded those in the historic NF2 population. Many of these cases also were reported to have substantial “sound-only” word recognition ability. Such patients are implanted without concurrent tumor resection and generally have more normal brainstem anatomy, which helps preserve neural function and facilitates the identification of important anatomical landmarks used in device implantation. This may be particularly important in maximizing ABI outcomes in pediatric cases.

ABIs are now being used in the treatment of deafness in pediatric cases with cochlear malformations or cochlear nerve aplasia. In 2004, we reported on ABI outcomes in 21 children with NF2 implanted as young as age 12.\(^{19}\) We found that these individuals could experience substantial communication benefit. A key factor was appropriate family and other support. Several years ago, we also conducted device programming and comprehensive evaluation of a 3-year-old non-NF2 case implanted with an ABI in Verona,\(^\) and then subsequently four other very young children implanted at the age of 2–3 years (also in Verona). Our findings have demonstrated that considerable, and in some cases remarkable, benefit is possible. Some of these children have developed a substantial level of auditory function, including discrimination of environmental sounds, some open set recognition of speech, development of useful speech and language, and in some cases have been able to attend a normal hearing classroom with appropriate support services.

Generally, pediatric cases without concurrent developmental delay or syndromic problems have been reported to perform best.\(^{21}\) Of course, benefits of ABI implantation must be weighed against the risk of surgery in this group of patients in whom craniotomy is performed solely for ABI placement. In the pediatric population, a retrosigmoid surgical approach is typically used, as the mastoid is less developed. As with pediatric CIs, early implantation seems to provide the best outcomes with generally diminishing returns as duration of deafness increases.

In a recent retrospective cohort study,\(^{22}\) a group of children were studied who initially underwent CI and failed to progress with auditory perception. Before CI, all of the children had severe-to-profound sensorineural hearing loss and a diagnosis of cochlear nerve deficiency. Exploration of the CI and simultaneous ABI on the same side was performed, and all patients demonstrated an absent cochlear nerve at surgery.

Performance as measured by the categories of auditory performance scale was significantly improved after ABI. Patients with cognitive deficits did gain performance benefits from the ABI, but the difference in performance compared to children without cognitive impairment was significant. Additionally, children implanted before age 3 also performed better than children implanted at an older age.

Cases of a small cochlear nerve on imaging may in reality represent an absent cochlear nerve. The decision on whether to place a CI in a patient with cochlear nerve deficiency is never an easy one, and currently both imaging and electrophysiology are not fully predictive of outcome with CI. Although this cohort was selected from patients with failed CI, ABI is a potential alternative to CI in select cases. In patients who fail to progress with intensive rehabilitation with CI, ABI may be considered as an alternative. While in this study, the CI was explanted, we may see more situations where an ABI is considered on the contralateral side, if any CI benefit has been obtained, to allow for continued use of both a CI and an ABI. After more than 33 years of experience with ABIs in adults, we have initiated our own pediatric ABI clinical trials under
Auspices of the FDA. Ten children aged 2–5 years will be implanted and their auditory performance tracked on an extensive battery of tests.

Since the beginning of our ABI program in 1979, we have utilized a team approach to maximize patient benefit, satisfaction, and safety. We feel that this is particularly necessary in pediatric ABI implantation given the possibility for eliciting nonauditory sensations, and the much more limited ability of young children to provide detailed feedback about the effects of electrical stimulation. They also are not typically capable of making precise judgments regarding the psychophysical parameters (e.g. electrical threshold, comfortable loudness, and electrode-specific pitch) that are normally considered necessary to properly program ABI sound processors. Initially, best estimates of these perceptual effects and parameters must be determined by skilled clinicians using age-appropriate behavioral testing. The efficacy of the ABI depends on the accuracy of the processor settings that must be objectively verified by further sound-field testing and close observation of the child at home and at school. Regular follow-ups for further behavioral testing and device reprogramming are critical in achieving maximum benefit. Since hearing with the ABI can change over time, some electrodes may become useable that could not be used initially.

Electrophysiological measures in sedated young children have been used, mostly in Europe, to program ABI sound processors, however we have found that these tests can significantly underestimate required stimulation levels. Therefore, we do not consider electrophysiological measures as a substitute for reliable behavioral responses in programming sound processors in pediatric ABI recipients. One area where both electrophysiological and behavioral measures fall short in pediatric cases is in the assessment of electrode-specific pitch sensations. In very young deaf children, the concept of perceptual pitch may not exist, or at least may not be reliably testable. In adults, such testing is used to sensibly assign electrodes to processor frequency analysis bands so that lower and higher frequency spectral cues are sent to electrodes that generate appropriate-sounding pitch percepts. Of course, each adult ABI user is somewhat different in this respect. In pediatric cases, the lack of this data has not been a serious obstacle, and both a generally increasing and a generally decreasing assignment of frequency information across electrodes have resulted in good performance. With little if any prior auditory experience, pre- or perilingually deafened young children appear to have adapted to either electrode-assignment strategy. We feel, however, that a reasonable practice for very young children is to apply the predominant pitch pattern seen in adults, which is a tendency for pitch percepts to rise in a lateral to medial direction across the electrode array. Despite these psychophysical complexities, it is clear that experienced pediatric ABI teams can safely implant and program the devices with very beneficial outcomes.

Recently, there has been encouraging data regarding the long-term safety of ABI use. Histological sections through the cochlear nuclei on both sides were obtained from an adult patient who used his ABI daily for 15 years. This individual demonstrated very high levels of speech recognition performance up until the time of his death from complications of NF2. The analysis showed good and comparable populations of auditory neurons on both implanted and unimplanted sides with no deleterious effects from long-term electrical stimulation. Also in this regard, the very first recipient of an ABI initially implanted in 1979 continues in 2013 to use her ABI daily with benefit. This is a very encouraging finding for pediatric cases potentially facing a lifetime of ABI use.

Recent ABI results in patients with NF2 also have been encouraging. Performance improvements were first seen in patients operated via the retrosigmoid approach in the semisitting position with the 12-electrode MedEl ABI device. In this group, reports indicated that about 30% of patients were experiencing some degree of “sound-only” word recognition ability. Due to concerns of other possible intraoperative complications, such as air embolism, the sitting position is not typically used for VS resection in the United States. When taking care to achieve meticulous tumor resection and brainstem handling via the translabyrinthine approach, our group is also achieving about a 25% incidence of “sound-only” speech understanding ability.

CONCLUSION

The ABI has been utilized to provide auditory benefit to deaf patients who are not candidates for cochlear implantation due to loss or absence of the cochlear nerves or abnormalities of the cochlea itself. By far, the most common indication for ABI is NF2. The safety of the stimulation of the cochlear nuclei using this device has been established. Most patients perceive beneficial auditory sensations and improve their communication abilities over lipreading only. A smaller number achieve substantial speech discrimination using only ABI sound, and
nontumor patients may achieve greater average benefit. A key factor is regular device use, and with that improvements can occur for 10 years or more. More recent ABI results demonstrate the possibility of achieving improved auditory results even in patients with NF2. Additionally, ABI implantation in prelingually deafened children who are not CI candidates holds much promise for providing auditory stimulation and allowing for language and speech development.

REFERENCES