Auditory brainstem implants in neurofibromatosis Type 2

To The Editor: I read with great interest the article by Matthies et al. They asked whether open speech perception is feasible in patients with neurofibromatosis Type 2 (NF2) and if placement of auditory brainstem implants (ABIs) would lead to excellent results.

Before this intervention was widely accepted, methods of ABI placement were anecdotal or delivered in small meetings and conferences. However, a previous report held in Munich in February 2014, attended by Professors Matthies and Behr, illustrated the results and awareness of ABI technology.

We need to weigh these results against the benefits of ABI placement. The authors report that they do not use bipolar cautery in the process of tumor resection. While I agree with the authors that cautery may increase the risk of complications, I do not think that this is the only factor that affects outcomes.

The use of the semisitting position may also facilitate brain relaxation and bloodless dissection, which sometimes interferes with array positioning. The authors also mention the importance of continuous intervention in ABI programming.

In terms of “denominator” issues, it is unclear whether the authors’ pool of implanted patients is the same as ours or other centers in the US. The authors’ 18 implanted patients are derived from a series of 104 patients. Did these patients present for surgical treatment due to NF2-related issues, or did they present for vestibular schwannomas, observation of vestibular schwannomas, or investigation of other NF2-related pathology?

The cochlear nucleus ABI allows E-ABRs to be confirmed prior to ABI implantation. In contrast, the Cochlear Nucleus ABI cable is not ideally flexible, which sometimes interferes with exact array positioning. The authors also mention the importance of continuous intervention in ABI programming.

Despite these issues, I am extremely impressed and humbled by the authors’ results. Placement of ABIs should not be undertaken lightly, and the authors’ results certainly confirm that meticulous detail and experience are critical to achieving excellent outcomes. Perhaps the most important take-home lesson for all readers is that of achieving better than previously thought...
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diagnostic benefits with ABIs in NF2 and the importance of proceeding with ABI implantation soon after loss of hearing.

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**Disclosure**

Dr. Schwartz is a consultant for Cochlear Americas.

**References**


**RESPONSE:** I am thankful for the opportunity to respond to the letter by Dr. Marc Schwartz on our recent ABI article.

One important aspect addressed by Dr. Schwartz is acquisition and selection of patients for ABIs. In Europe, there is a great tendency for patients and colleagues to seek advice and treatment at major specialized centers and to travel long distances for this reason. Patient selection is based on a large cohort of patients with long-term follow-up who have various skull base pathologies and other neurofibromatosis-related tumors. My neurofibromatosis outpatient clinic that had been located in Hannover for 15 years is now located at Würzburg and is continuously growing. Here, patients receive complete counseling and treatment for tumors of the head, spine, and periphery from the interdisciplinary team, in addition to members of the neuro-oncology, neuro-ophthalmology, neuroradiology, and radiotherapy departments, and specialists in genetics and tissue engineering are also available to participate in patient care as needed. Currently about 250 patients with NF2 are seen for regular follow-up; of these patients 104 underwent surgery for a variety of tumors during the reported study period. For vestibular schwannomas, hearing preservation is the primary goal and is achieved in about 30% of our NF2 patients. Well-informed patients seek advice and ask for an ABI often before surgery for large tumors or are transferred after previous surgery elsewhere. As mentioned, about half of the patients in the study had undergone surgery once or several times before. In fact, in all of the patients in this continuous study, E-ABRs could be confirmed and the indication for implantation was confirmed at surgery and carried out. The quality of the E-ABRs showed specific variation and is probably an indicator for the potential of acoustic recovery.

For NF2 patients undergoing postintervention follow-up, long traveling means an additional burden in view of their lesions and physical disabilities, and also with regard to financial charges. Nonetheless, the experience of receiving complete and competent advice and care requires patients and families to keep in contact with the clinic and travel repeatedly 600 km (400 miles) or more. Since only a few centers are allowed to perform ABI surgery, health insurance often covers some of the travel expenses.

A further supportive factor is that patients, who have received the current device, experience fast hearing recovery. In contrast to previous studies, we now are seeing some useful hearing perception within the first days and weeks after ABI activation. This knowledge increases patients’ motivation to return and have the auditory frequencies tuned, loudness modulated, and program adaptations tried out.

Among the surgical aspects addressed, the semisitting position is advantageous, especially for patients with large tumors. Previous fears of transverse section syndrome can be minimized by electrophysiological control. The same applies to the risk of air embolism; this risk is now low, and, if it occurs, in my practice, related complications are rare thanks to the involvement of an experienced neuro-anesthesiologist and his close interactions with the surgical partners. In this surgical position, the option of continuous fluid irrigation helps avoid the need for cauterization. The danger of cauterization was identified and brought to light in the 1970s by Madjid Samii and Leonard Malis, before the era of monitoring. Dr. Samii started to resect giant tumors without any bipolar cauterization and obtained astonishing functional results. Without any monitoring, only the avoidance of bipolar cauterization could help protect the cranial nerves.

The inherent risks of cauterization became evident when electrophysiological monitoring was first used, though the underlying mechanisms are multiple and still debated. At the Munich ABI Meeting of Neurosurgeons (Kempinski Hotel Airport, Munich, March 23 and 24, 2012), Robert Shannon discussed the biological processes possibly induced in the nervous tissue at current application. In some ABI placement surgeries I performed in the supine position at other centers, we were able to obtain the same quality of open speech perception. Thus, I do not think that the semisitting position is mandatory in ABI surgery, but it has specific advantages.

Regarding the device, the option of using a test electrode is advantageous, especially as the paddle has the identical size as that of the final implant. If another company’s device is used, I would suggest using a bipolar stimulating probe to test the responsiveness of the cochlear nucleus. This probe may be designed for direct nerve stimulation with rounded poles. In our series with other devices (Nucleus and Clarion) implanted in Hannover, we used such a probe and connected it to our electrophysiological recording system and had it triggered by the Nucleus stimulation system.6

The cable of the Medel device is very flexible and allows gentle introduction of the implant into the recess at any angle that might be necessary according to the individual anatomy. Once the “paddle” with the electrodes is placed and moving with the brainstem, the cable ensures sufficient freedom of movement. With the exception of one of our patients who fell within a few weeks after surgery and who needed implant repositioning, I have not observed any dislocation in more than 30 implants I have placed using this model.

The current speech processor provides the engineer