Auditory brainstem implants in neurofibromatosis Type 2


Those of us involved in the care of patients with neurofibromatosis Type 2 (NF2) and placement of auditory brainstem implants (ABIs) have been aware for some time of excellent results being obtained in a few European centers. Prior to this report, most of this information had been anecdotal or delivered in small meetings and conferences. There was one previous report in the otolaryngology literature. In fact, awareness of these results in NF2 patients led to a small meeting to discuss the possible reasons for differences in results, held in Munich in 2012, and attended by Professors Matthies and Behr, who have been the primary surgeons for these cases.

Due to the rarity of the disease, there are no truly large centers for ABI placement and only a few with substantial numbers of cases. Given the improved results that have now been reported, those of us continuing to place ABIs are obliged to critically evaluate our own methods to determine whether there is room for improvement. For instance, as a surgeon with a greater than 50 case experience in ABI placement for NF2, I have a number of patients with excellent open-set speech discrimination but not nearly the 44% (8 of 18) achieving greater than 25% open-set discrimination in this series.

It is also possible, however, that part of the difference in results lies in methodological aspects that cannot or need not be addressed. In addition, there may be both a numerator issue and a denominator issue.

In terms of issues that can clearly be addressed, I would agree with the authors that an extremely important one is the technical care taken in tumor resection and ABI placement. The authors report that they do not use bipolar cautery in the process of tumor resection, and we minimize cautery use as well. In my opinion, the important concept is to understand that there is a higher degree of delicacy needed akin to that needed for hearing preservation surgery.

The use of the semisitting position may also facilitate brain relaxation and bloodless dissection. We do not utilize the semisitting position, and I am not aware of any center in the US that does (for vestibular schwannoma resection). Adoption of this technique may pose significant challenges in this country due to medicolegal concerns regarding the perceived increased risk of air embolism, as well as surgeon comfort and familiarity. It is possible that the benefits of the semisitting position are required to obtain improved ABI results. These benefits would have to be weighed against any additional risks, either real or perceived.

The next issue is that of the ABI device. The authors use the Med-EI ABI, while the only device available in the US is the one manufactured by Cochlear Ltd. The editorial and response preceding the article itself debate this issue in some detail in terms of electronics and processor technology. I would posit, however, that differences in outcome could as well be due to the physical characteristics of the electrode arrays or even the cables. For instance, the Cochlear Nucleus ABI cable is not ideally flexible, which sometimes interferes with exact array positioning.

Yet another issue is that of postimplantation ABI follow-up. The authors stress the importance of continuous intervention in ABI programming by experienced audiologists. In the experience of our center, follow-up is often problematic due mostly to vast geographic distances in North America, an issue that may pose less of a problem in Europe. Our patients typically travel long distances for surgery and initial ABI activation. They must then choose to continue contact with our audiologist on a regular basis, and not all can do this due to either practical or financial concerns. I am not sure there is an easy solution to this problem, since the presence of more centers necessarily dilutes experience at each center.

In terms of “denominator” issues, it is unclear to me 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 of vestibular schwannomas, observation of vestibular schwannomas, or investigation of other NF2-related pathology? While our center treats patients for a variety of NF2-related issues, a higher proportion presents for vestibular schwannoma surgery and ABI placement. Perhaps we are less selective in whom we implant the devices.

This selectivity could be on the basis of preoperative evaluation but also could be on the basis of intraoperative findings. The Med-El device includes a 4-channel test electrode via which electric auditory brainstem responses (E-ABRs) can be confirmed prior to ABI implantation. The authors report that the ABI was implanted “if adequate activation of the cochlear nucleus was evident,” suggesting that the ABI was not implanted in all cases. In contrast, the Cochlear ABI allows E-ABR testing only with the ABI device itself. The device must first be implanted to even test, and testing is carried out only to optimize positioning. We do perform electrophysiological mapping in all cases, but only after the ABI has already been implanted.

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 lessons for all readers are the possibility of achieving better than previously thought au-
diological 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 reject 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. 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 1 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.

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