Auditory brainstem implants

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We read with great relish the clinical report by Matthies et al.² describing their experience with auditory brainstem implants (ABIs) in patients suffering from neurofibromatosis Type 2. To put it simply, these are the best results in the literature in terms of auditory perception in this patient population. What is most interesting about this report, though, is the fact that these results are so vastly better than anything else recorded in the literature. In this series, at 24 months after implantation, 13 (72%) of 18 patients had open-set speech recognition present at least to some degree on a sentence-reading test. This means that without any visual cues at all, using sound input only from the ABI, these patients were able to understand sentences. In the literature to date, the best reported results worldwide have been closer to 11% of patients attaining open-set speech recognition. The easy way out here would be just to attribute these results to the excellence of the surgeons and center involved and call it a day, but that wouldn’t help us understand what other factors might potentially explain these results. It bears repeating that these results are so much better than what has been achieved before that we think it behooves us all, including the authors, to understand what factors might be critical and potentially translatable to other centers in order to improve results in all programs implanting these devices.

One important factor emerges from the data in this report that has been seen in other reports and in multicenter trials. Dedicated and continuous revision of the programming has an important effect on the ultimate outcome. The best results in hearing perception were only achieved at 24 months. At 6 months, by contrast, only 3 patients had greater than 30% accuracy on the open-set sentence test. In our own series of patients and in other trials, the dedication of the patient in returning for revision of programming and in continued use and training was critical in achieving better outcomes.¹ The strength of the current series is in the stringent follow-up and continued training of the users. We should also note that the authors implanted the Med-El device exclusively. We could speculate that the improved results might be due to alternative processing strategies and stimulation paradigms. We would be curious to hear the authors’ views on the importance of the device type. One thing is clear; this type of surgery cannot be undertaken except in centers with experienced audiologists who are well versed in advanced processing for cochlear implants and ABIs.

Finally, the authors show for the first time that implantation can be successful despite the presence of a large tumor, provided that landmarks can be retained. As they mention, one of their best results was in a patient with a large tumor. They also identified the duration of ipsilateral deafness as an important factor. This has long been suspected, because loss of neurons in the dorsal cochlear nucleus has been previously demonstrated in animal models involving destruction of the cochlear nerve. The data in the current report seem to show a clear trend toward improved results in patients who still have residual hearing on the side with the implanted device or who are only recently deaf. This report, like others, has a limited number of “star” patients whose results are so much better than the other patients. Only 4 of the original 18 patients had more than 75% accuracy on the sentence test at 24 months (bearing in mind, nonetheless, that a total of 13 patients had some open-set speech recognition). Three of these 4 outperformers had experienced only 2 months of ipsilateral deafness. For us, this is compelling evidence that ABI surgery should be undertaken earlier rather than later in a patient’s course.

Matthies et al. also mention trials of the midbrain implant in the inferior colliculus. We don’t share their enthusiasm for this strategy. In the history of neural prostheses, the farther one goes along the pathway from the primary sensory receptor, the poorer the results of the prosthesis. This was true in the visual pathway, so that current prostheses are retinal rather than cortical. Similarly, too much signal processing may occur in the dorsal cochlear nucleus, or even in the cochlear nerve itself, so that implantation at the inferior colliculus may not recreate auditory perceptions.

We congratulate the authors for these outstanding results. We are still left wondering what the critical factors are in obtaining such high levels of open-set speech perception. Most likely it is a combination of the factors identified in this series: dedicated programming over 2 full years, getting patients scheduled for implantation within 2–3 months of deafness, and meticulous surgical technique. We also note that the average use of the ABI was still only 8 hours a day in this patient cohort, which is in line with what has been reported in other series, despite the high rate of open-set speech recognition. It is also a valuable lesson to take
away that large tumor size does not preclude implantation. We would welcome more study on different processing strategies and stimulation protocols to see if the results can become even more reliable.

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**Disclosure**

The authors report no conflict of interest.

**References**


**Response**

CORDULA MATTHIES, M.D., PH.D.,1 STEFAN BRILL, DIP. ING. (M.SC.E.E.),2 AND WAFAA SHEHATA-DIELER, M.D., PH.D.2

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We thank Drs. Golfinos et al. for their sincere interest in our study and their encouraging remarks and discussion regarding speech perception in patients with ABIs. We gratefully accept the opportunity to respond to their editorial. They suggest some further discussion of “what factors might be critical and potentially translatable to other centers in order to improve results.” Some of those are addressed below.

**Clinical Factors**

Golfinos et al. ascribe the quality of results to the stringent long-term patient follow-up and revision of programming.

In fact, the patients in our series participated very reliably in the postsurgical control and rehabilitation program, with repeated clinical and functional controls and adaptation of the stimulation parameters. Although this continuity in clinical treatment and follow-up is probably a positive factor, the converse applies as well: because our patients noticed very early on the usefulness of their device and experienced fast and ongoing improvement, they were very motivated to stick to this schedule.

For the ABI fitting procedure a pragmatic and individualized approach is mandatory, with the main focus on improving speech and environmental sound perception and taking into account the underlying disease and tendency to fatigue and concentration difficulties. Loudness perception, compound action potentials, telemetric impedance values of individual electrodes, and side effects are considered, but to a lesser extent than in cochlear implant programming, as long as a benefit is achieved for the overall speech and sound perception.

We also believe that the repeated open-set testing with sentences at each of the patients’ visits somehow motivated them to use their devices and to involve their families and friends in training their hearing, both with and without lipreading. To state the obvious, we never used the same sentence lists twice and we never handed out any of the sentence material to the patients, for example for training purposes.

As pointed out by the reviewers, not all patients experience open-set speech perception early on (at 3-month follow-up there were 3 patients with better than 30% perception in the open-set sentence test), but most do after 2 years (11 of 16 have some open-set speech perception). So the reviewers speculate that later improvement might have resulted from changes in programming. Although adaptation of programming is important, our clinical data point to another interpretation: most of the patients who needed longer to obtain very good results had gone through longer periods of deafness and needed longer periods for complex hearing recovery. Already at 3 months, however, in many patients, results on syllable and word tests at closed set (monotrochee-polysyllabic [MTP] test) are very good, namely much better than in previous series, and statistically this early MTP test is a strong predictor of the development of open-set speech perception (see Fig. 6 in our article). Therefore, very early on—at the 3-month follow-up—the individual potential for a good outcome can be predicted by MTP test results. A good MTP test result reflects a good level of perception early on. At this early phase the majority of patients were already much better than those reported on in previous studies. Advanced processing of auditory input needs some revival after a long duration of deafness, and some reorganization of the interplay between the cochlear nucleus and auditory cortex.

The short duration of deafness is identified as a very powerful factor according to our data and our experience. What is new in this series is the finding that critical periods are much shorter than previously suspected. So far, discussion has concentrated on deafness periods of 5 or 10 years that might decide on hearing with the ABI, regarding sound and tone perception, or words with lipreading.1,2

Now we have learned that very short periods of a few months may cause a difference in outcome and that open-set speech perception is possible and becomes realistic if implantation is performed early enough. This is very valuable knowledge because the surgeon may be able to influence the duration of deafness to some extent; communicating this knowledge and perspective to the patient will support him or her in the decision for or against an ABI. Golfinos and colleagues mention that there is time-dependent degeneration in the dorsal cochlear nucleus. As in other nuclear lesions, structural changes at higher (cortical) functions with loss of diversity and complexity are the consequence.

**Surgical Factors**

The basis for a good MTP test result early on (the predictor of open-set speech perception) consists of 2 factors: the delicate handling of the brainstem and the ex-