Neurofibromatosis Type 2 (NF2) is characterized by bilateral vestibular schwannomas (VSs) in more than 90% of affected patients. These VSs are mostly accompanied by further tumor manifestation in the head, spine, and peripheral nerves. The condition is genetically defined by a mutation of a tumor suppressor gene on chromosome 22 coding for merlin, a Schwann cell inhibitor. The lack or dysfunction of merlin leads to an excessive Schwann cell proliferation, with multiple nerve sheath tumors.

Auditory brainstem implants in neurofibromatosis Type 2: is open speech perception feasible?

Clinical article

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Object. Patients with bilateral auditory nerve destruction may perceive some auditory input with auditory brainstem implants (ABIs). Despite technological developments and trials in new stimulation sites, hearing is very variable and of limited quality. The goal of this study was to identify advantageous and critical factors that influence the quality of auditory function, especially speech perception.

Methods. The authors conducted a prospective study on ABI operations performed with the aid of multimodality neuromonitoring between 2005 and 2009 in 18 patients with neurofibromatosis Type 2. Outcome was evaluated by testing word recognition (monotrochee-polysyllabic word test at auditory-only mode [MTPa]) and open speech perception (Hochmair-Schulz-Moser [HSM] sentence test), both in pure auditory mode. The primary outcome was the HSM score at 24 months. The predictive meaning of general clinical data, tumor volume, number of active electrodes, duration of deafness, and early hearing data was examined.

Results. In 16 successful ABI activations the average score for MTPa was 89% (SD 13%), and for HSM it was 41% (SD 32%) at 24 months. There were 2 nonresponders, 1 after radiosurgery and the other in an anatomical variant. Direct facial nerve reconstruction during the same surgery was followed by successful nerve recovery in 2 patients, with a simultaneous very good HSM result. Patients’ age, tumor extension, and tumor volume were not negative predictors. There was an inverse relationship between HSM scores and deafness duration; 50% or higher HSM scores were found only in patients with ipsilateral deafness duration up to 24 months. The higher the deafness sum of both sides, the less likely that any HSM score will be achieved (p = 0.034). In patients with total deafness duration of less than 240 months, higher numbers of active electrodes were significantly associated with better outcomes. The strongest cross-correlation was identified between early MTPa score at 3 months and 24-month HSM outcome.

Conclusions. This study documents that open-set speech recognition in pure auditory mode is feasible in patients with ABIs. Large tumor volumes do not prevent good outcome. Positive preconditions are short ipsilateral and short bilateral deafness periods and high number of auditory electrodes. Early ability in pure auditory word recognition tests indicates long-term capability of open speech perception.

Key Words • acoustic neuroma • auditory brainstem implant • auditory midbrain implant • neurofibromatosis • open-set sentence recognition • speech perception • vestibular schwannoma • oncology • skull base

Abbreviations used in this paper: ABI = auditory brainstem implant; ABR = auditory brainstem response (acoustic stimulation); CN = cranial nerve; E-ABR = ABR (electric stimulation); HSM = Hochmair-Schulz-Moser; MTP, MTPa, MTPav = monotrochee-polysyllabic word test, MTP at auditory-only mode, MTP at audio-visual mode; NF2 = neurofibromatosis Type 2; VS = vestibular schwannoma.
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cochlear nerve dysfunction, and the natural course results in deafness within periods of 9–14 years. In NF2, the phase of tumor manifestation happens much earlier in life, frequently in adolescence, and hearing loss occurs often within a few years or weeks. Patients are faced with a double burden: life-threatening multiple tumors and impending loss of auditory communication—a devastating experience. Early diagnostic tests, refinement of microsurgical tumor resection, and monitoring control have enabled and improved hearing preservation in up to 36% of patients with NF2. Still, many patients will retain this preserved function for a limited period, until it is destroyed by tumor recurrence.

In bilateral retrocochlear deafness, rehabilitation of some auditory sensation can only be achieved by auditory prosthesis of the brainstem or midbrain. Functional results show great variability; from noise perception only to useful speech discrimination in audiovisual mode in a few patients so far. Since the early trials by Hitselberger and House in 1979, as reported by Edgerton et al. and Hitselberger et al., profound improvements in implant array, technology, and speech processing strategies have brought about useful improvement in functional outcome. As a consequence, some colleagues advocate tumor surgery and prosthesis implantation when tumors are at very small stages to achieve good auditory results. Still, ABIs after resection of small tumors give only modest results far inferior to cochlear implants.

Furthermore, patients with NF2 who have unilateral deafness and a tumor on the remaining hearing side wish to postpone surgery and to keep their natural hearing in the second ear as long as possible. This attitude must be accepted because there is no treatment equivalent to natural hearing. It is the interdisciplinary treatment philosophy of our team to opt for long-lasting conservation of natural auditory function; this goal is accomplished by careful microsurgical tumor resection with primary hearing preservation. Preserved hearing in patients with NF2 is frequently limited to periods of some years, and it is endangered by tumor recurrence or regrowth. As a consequence of this tendency to postpone surgery, ABIs for auditory rehabilitation will be indicated at tumor stages in which the lesions are large, recurrent, life-threatening, and presenting with further deficits such as ataxia and caudal cranial nerve (CN IX–XII) lesions. But is good rehabilitation of hearing possible at such advanced tumor stages, and by which means?

In the present study we focused on 2 major aspects: first, to outline the current microsurgical optimization of the ABI procedure with the interdisciplinary team; and second, to identify some advantageous prognostic factors. This study was approved by the Würzburg University Ethics Committee.

**Methods**

**Patient Selection, Preparation, and Treatment**

Of a consecutive series of 104 patients with NF2 who were treated between 2005 and 2009, 18 patients with NF2 were selected for ABI surgery. All patients were seen regularly in a specialized NF2 outpatient clinic. Before inclusion in the hearing implant program, counseling and interviews with the patient were performed by the interdisciplinary team in at least 2 separate presentations, on the pros and cons of hearing implants in general, on the individual disease manifestation, on the treatment steps, and on the realistic expectations of functional outcome. The patient’s ability in lipreading or his/her individual improvement was evaluated at different presentations, and furthermore his/her social context and likely support from family, relatives, and friends were assessed.

Indications for and timing of surgery were based on the following clinical grounds: 1) the patient’s interest in hearing rehabilitation; 2) severe brainstem compression from VS; 3) bilateral deafness or diminishing hearing on the remaining side with an interest in conservation; or 4) if conservation is shown as impossible during surgery, an interest in rehabilitation by ABI.

Preparation for surgery included the following: 1) audiometry and auditory brainstem response (ABR) testing, if residual hearing was present; 2) promontory stimulation test and promontory ABR testing; 3) MRI studies of the head (Fig. 1) and whole spine, and bone window cranial CT; 4) high-resolution bone window CT scans of the skull base; 5) cervical radiograph in ante- and retro-
flexion; 6) cervical positioning test for preparation of the semisitting position; and 7) intensive counseling sessions by the surgeons and engineers of the neurosurgical and otological team.

Surgery of the tumor and implantation of the auditory device were performed after induction of general total intravenous anesthesia with the patient in the semisitting position, controlled by precordial Doppler sonography and transesophageal sonography. Perioperative and intraoperative monitoring included somatosensory evoked potentials for control of long tracts, electromyography of motor CNs, motor evoked potentials, and ABRs by acoustic and/or electric stimulation (ABR and E-ABR). Via the lateral suboccipital approach, microsurgical tumor resection was performed with special consideration for preservation of microvascular structures, brainstem, and nerves, and by avoidance of any bipolar cauterization (Fig. 2A and B). Utmost care was taken with motor CNs VII–XII, with application of minimal electrical stimulation.

For identification of the ideal site of ABI placement, CNs VII and IX were followed (Fig. 2C) up to the brainstem and, by slight retraction of the flocculus and the choroid plexus, the region of the dorsal cochlear nucleus was exposed (Fig. 2C). Mapping of the cochlear nucleus was performed with a quadripolar test electrode (Fig. 2D) until a clear waveform complex of E-ABR components III–V was obtained (Fig. 2E); attention was paid to any side effects observed on neurophysiological and cardiovascular monitoring. If adequate activation of the cochlear nucleus was evident, the test electrode was removed and the permanent 12-channel ABI electrode (Med-El Pulsar CI 100) was placed (Fig. 2F). Adequate positioning was again tested by E-ABR and by resistance control of each electrode contact, and then the final position was secured by superficial fibrin plaster at the cable site laterally at the brainstem. Throughout the procedure cauterization was avoided and only applied at the end during jugular vein compression before dura mater closure, if there were any accidentally opened vessels that needed sealing.

Postoperative treatment included 1 night of treatment and observation in the neurosurgical ICU, neuroanesthesiology control of caudal CNs' function, facial nerve documentation, and cranial CT scan. In case of caudal CN dysfunction, specific ergotherapeutic training was applied, and, in case of delayed recovery, temporary tracheotomy was indicated and carried out.

The ABI activation and programming was scheduled at 6–8 weeks after surgery, when the patients were readmitted for 3 days. The first ABI activation session was performed in the ICU under electrocardiographic

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**Fig. 2.** Case 2. Intraoperative photographs and E-ABR readout illustrating some steps of surgery in this 22-year-old man. He had been deaf on the right side since tumor resection in 2004 and developed deafness in his other (left) ear in 2006, along with tumor progression on the left side, with ataxia and CN V deficit. At surgery of the large tumor, caudal CNs IX and X were found at the lower part (A); the facial nerve fibers were spread apart and elongated, but could all be preserved under continuous control by motor CN monitoring from the brainstem to the meatus (B). Using CN IX as a guide, the lateral recess was identified and opened and the region of the cochlear nucleus was thereby exposed (C). After successful stimulation by a quadripolar test electrode (D) and registration of reproducible E-ABR complexes (E; ms = millisecond), the final ABI electrode was placed and fixed on the cochlear nucleus (F). The patient had immediate useful auditory function, but experienced a significant improvement toward better voice and speech recognition after 12 months, with a score of 60%, and with a score of 84.9% after 4 years on the HSM test.
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control with preparations for emergency resuscitation. The ABI testing consisted of control of each electrode for resistance, sensation perception as auditory or non-auditory (or silent), by sensitivity level, pitch perception, pitch ranking, and intensity level. After identification of auditory electrodes, the next programming sessions were performed at the hearing implant center outside the ICU.

Stimulation strategy was continuous interleaved stimulation technique (CIS+) as a routine,\textsuperscript{2,65} at a maximum rate of 50,760 pulses/second.

Subsequent ABI control and programming sessions were scheduled at 1 month, then at 3-month intervals in the 1st year, and thereafter at 6- to 12-month intervals depending on the needs of the individual patient. At 3–6 months, an in-clinic auditory rehabilitation program was organized. In patients with advanced NF2 and multiple neurological deficits, intensive neurological rehabilitation was performed.

\textbf{Data Analysis}

The following parameters were considered for each patient: 1) age; 2) sex; 3) side of implantation; 4) previous surgery or radiosurgery; 5) duration of ipsilateral deafness; 6) duration of contralateral deafness; 7) number of active acoustic electrodes; 8) word recognition tests at closed set (a: monotonche-polyphonic word test [MTP] at auditory-only mode [MTPa], and b: MTP at audiovisual mode [MTPav]); 9) speech recognition test at open set by Hochmair-Schulz-Moser (HSM) sentence test at auditory-only mode;\textsuperscript{22} 10) duration of follow-up; 11) tumor extension toward the brainstem according to Hannover classification\textsuperscript{32} (this classification was extended by Grade T4c for most severe brainstem compression with tumor growth over the midline to the contralateral side [for example see Fig. 1]); and 12) tumor volume—the tumor was delineated in each slice of continuous contrast-enhanced CT or T1-weighted MRI by using the application MIPAV (Medical Image Processing, Analysis, and Visualization) provided by CIT (Center for Information Technology) at the NIH (http://mipav.cit.nih.gov/index.php). Summing the obtained areas and multiplication with the slice thickness resulted in the tumor volume. Because continuous data are not always available, volume could be calculated as an ellipsoid (4:3 × \Pi × abc (abc is the radius in 3 dimensions: a = transverse, b = sagittal, c = craniocaudal; Fig. 1).

Clinical outcome classes were applied according to pure auditory mode results in speech recognition tests and, if not applicable, in word recognition tests: Class 1, excellent (100%–76% HSM score for open-set speech recognition); Class 2, good (75%–51% HSM score); Class 3, medium (50%–25% HSM score); Class 4, fair (24%–6% HSM score); Class 5, useful (0% HSM score, no open-set speech recognition, but closed word recognition [≥ 40% MTPa score]).

\textbf{Statistical Analysis}

The primary outcome was the HSM score at 24 months. In 2 patients who had no measurement at the 24-month visit but had a score of 0 on all preceding visits, the outcome was set to 0. The numbers of patients who achieved a 24-month HSM score of 25% or greater, and of 10% or greater were evaluated. Exact 95% CIs for these percentages were computed using the binomial distribution.

The predictive meaning of several pre- and postoperative variables and of short-term results for the long-term outcome after 24 months was examined. In particular, we considered tumor volume, ipsilateral deafness, total duration of deafness (the sum of the durations of left- and right-sided deafness as a measure of the cumulative burden of disease), the number of active electrodes as a measure of procedural success, and MTPa and HSM scores after 3 months. The relationship between quantitative variables was assessed using the Kendall tau coefficient, a rank correlation coefficient that is independent of the normal distribution of the data. Corresponding p values refer to the test of the null hypothesis of no correlation. SPSS 20 (IBM Corp.) was used as statistical software; p values < 0.05 were considered significant.

\textbf{Results}

\textbf{Data Acquisition and Follow-Up}

In 2 patients ABI activation was not successful. One patient remained deaf after previous radiosurgery; she had developed deafness and facial palsy 4 months after radiosurgery at another clinic. Rapid further tumor progression with severe brainstem distortion necessitated tumor resection, and ABI placement was possible with reliable responses. After unsuccessful ABI activation, a surgical reextraction and proof of correct stimulation site were performed; still she did not experience any hearing sensation. The other patient remained deaf (probably because of an anatomical variant) and a repeat operation could not be done due to another neoplastic disease, a gynecological tumor.

In 16 of 18 implantations ABI activation was successful, with reproducible auditory sensation (89%). Long-term follow-up is available in 15 patients, ranging from 1 to 5 years. For medical reasons, in some patients not all parameters could be obtained; in 2 patients their general status did not allow long investigations such as the HSM sentence test, but in all patients repeated word recognition tests could be performed. The patient in Case 5 had only 4 months of follow-up; she died of a recurrent tumor of the pharynx and mediastinal region. In 6 patients volumetric tumor calculations were not directly possible by the radiographic program, but by mathematical calculation.

\textbf{Surgical Data and Sequelae Related to Tumor Surgery}

Patients’ ages ranged from 19 to 66 years (mean 37.6 years, SD 12.6 years; see Table 1). Tumor recurrences were present in 9 (56%) of the 16 patients (Cases 1, 4–7, 10, 12, 14, and 16) after previous surgery for hearing preservation, and after a previous ABI procedure in 2 of them (Cases 16 and 17). In 3 patients (Cases 14, 16, and 18), tumor surgery had been undertaken some years before; the goal for surgery now was hearing rehabilitation. To judge the possible impact of tumor compression, the extension and volume of those previous tumors were considered.
TABLE 1: Auditory function in 18 patients with VSs and deafness treated with ABIs within the 1st year after activation*  

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Side</th>
<th>Previous Op/RS, Yrs Before</th>
<th>Deafness (mos)</th>
<th>3-Mo FU Scores</th>
<th>6-Mo FU Scores</th>
<th>12-Mo FU Scores</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Ipsilat Contralat</td>
<td>MTPa MTPav HSM</td>
<td>MTPa MTPav HSM</td>
<td>MTPa MTPav HSM</td>
</tr>
<tr>
<td>1</td>
<td>36, M</td>
<td>lt</td>
<td>12 yrs ago, rec</td>
<td>2 120</td>
<td>100.0 100.0 64.2</td>
<td>100.0 100.0 87.7</td>
<td>100.0 95.8 86.8</td>
</tr>
<tr>
<td>2</td>
<td>22, M</td>
<td>lt</td>
<td>none</td>
<td>12 24</td>
<td>95.8 100.0 12.3</td>
<td>87.5 100.0 28.3</td>
<td>100.0 100.0 60.4</td>
</tr>
<tr>
<td>3</td>
<td>47, F</td>
<td>rt</td>
<td>1 yr ago, RS</td>
<td>12 36</td>
<td>ND ND ND</td>
<td>ND ND ND</td>
<td>ND ND ND</td>
</tr>
<tr>
<td>4</td>
<td>48, F</td>
<td>rt</td>
<td>20 yrs ago, rec</td>
<td>2 48</td>
<td>87.5 100.0 2.8</td>
<td>100.0 100.0 65.1</td>
<td>100.0 95.8 81.1</td>
</tr>
<tr>
<td>5</td>
<td>25, F</td>
<td>rt</td>
<td>10 yrs ago, 13 yrs ago, rec</td>
<td>180 108</td>
<td>41.7 100.0 0.0</td>
<td>ND ND ND</td>
<td>ND ND ND</td>
</tr>
<tr>
<td>6</td>
<td>54, F</td>
<td>lt</td>
<td>12 yrs ago, rec</td>
<td>48 252</td>
<td>16.7 100.0 0.0</td>
<td>66.7 100.0 0.0</td>
<td>50.0 100.0 0.0</td>
</tr>
<tr>
<td>7</td>
<td>27, F</td>
<td>lt</td>
<td>14 yrs ago, rec</td>
<td>2 24</td>
<td>75.0 100.0 12.2</td>
<td>95.8 100.0 12.3</td>
<td>83.3 100.0 28.3</td>
</tr>
<tr>
<td>8</td>
<td>19, F</td>
<td>rt</td>
<td>none</td>
<td>12 0</td>
<td>54.2 95.8 0.0</td>
<td>70.8 100.0 0.0</td>
<td>87.5 87.5 10.4</td>
</tr>
<tr>
<td>9</td>
<td>40, F</td>
<td>rt</td>
<td>none</td>
<td>48 60</td>
<td>ND ND ND</td>
<td>ND ND ND</td>
<td>ND ND ND</td>
</tr>
<tr>
<td>10</td>
<td>51, F</td>
<td>lt</td>
<td>19 yrs ago, rec</td>
<td>48 228</td>
<td>41.7 100.0 0.0</td>
<td>62.5 100.0 0.0</td>
<td>83.3 100.0 0.0</td>
</tr>
<tr>
<td>11</td>
<td>66, M</td>
<td>rt</td>
<td>none</td>
<td>2 144</td>
<td>100.0 100.0 99.1</td>
<td>100.0 100.0 99.1</td>
<td>100.0 100.0 97.2</td>
</tr>
<tr>
<td>12</td>
<td>43, F</td>
<td>lt</td>
<td>10 yrs ago, rec</td>
<td>6 144</td>
<td>87.5 100.0 39.6</td>
<td>87.5 100.0 9.4</td>
<td>95.8 100.0 12.3</td>
</tr>
<tr>
<td>13</td>
<td>42, M</td>
<td>rt</td>
<td>10 yrs ago; unsuccessful</td>
<td>144</td>
<td>75.0 100.0 3.8</td>
<td>91.7 100.0 7.5</td>
<td>91.7 100.0 14.2</td>
</tr>
<tr>
<td>14</td>
<td>31, M</td>
<td>lt</td>
<td>8 yrs ago, no rec</td>
<td>96 3</td>
<td>83.3 100.0 0.0</td>
<td>75.0 100.0 1.9</td>
<td>87.5 100.0 16.0</td>
</tr>
<tr>
<td>15</td>
<td>42, M</td>
<td>rt</td>
<td>2 yrs ago, regrowth</td>
<td>0 184</td>
<td>75.0 95.8 0.0</td>
<td>83.3 100.0 0.0</td>
<td>95.8 100.0 5.7</td>
</tr>
<tr>
<td>16</td>
<td>33, M</td>
<td>rt</td>
<td>ABI 12 yrs ago, no rec</td>
<td>156 168</td>
<td>66.7 100.0 0.0</td>
<td>66.7 100.0 0.0</td>
<td>50.0 100.0 0.0</td>
</tr>
<tr>
<td>17</td>
<td>27, M</td>
<td>lt</td>
<td>15 yrs ago, ABI 10 yrs ago</td>
<td>3 120</td>
<td>79.2 100.0 22.6</td>
<td>62.5 100.0 11.5</td>
<td>95.8 100.0 38.7</td>
</tr>
<tr>
<td>18</td>
<td>25, F</td>
<td>rt</td>
<td>10 yrs ago, 4 yrs ago</td>
<td>24 12</td>
<td>66.7 100.0 0.0</td>
<td>87.5 100.0 11.3</td>
<td>100.0 100.0 36.8</td>
</tr>
<tr>
<td>mean</td>
<td>37.6</td>
<td></td>
<td></td>
<td>36.3 101.1</td>
<td>71.6 99.5 16.0</td>
<td>82.5 100.0 22.3</td>
<td>88.0 98.6 32.5</td>
</tr>
<tr>
<td>SD</td>
<td>12.6</td>
<td></td>
<td></td>
<td>54.4 78.6</td>
<td>23.2 1.4 28.6</td>
<td>14.0 0.0 33.5</td>
<td>16.6 3.4 33.5</td>
</tr>
</tbody>
</table>

* Auditory scores are expressed as percentages. CI = cochlear implant; FU = follow-up; ND = not done (patients unable to perform test); rec = recurrence; RS = radiosurgery.
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Postoperative sequelae included 2 cases of hydrocephalus treated by ventriculoperitoneal shunt implantation (Cases 11 and 12), and 2 surgical revisions for local CSF effusion. Caudal CN palsies were partially present in 9 patients before surgery and deteriorated in 4 of them; in 1 patient a temporary tracheotomy and in 3 temporal parenteral endogastric tube implantation were necessary.

Facial nerve reconstruction by sural nerve graft was necessary in 2 patients (Cases 1 and 4) at the same surgery, with successful reinnervation and with complete eye closure within the 1st postoperative year.

**Surgical Data and Sequelae Related to ABI Procedures**

Two patients had had ABI trials performed by other surgeons without success, and asked for a second attempt. The patient in Case 13 had received an ABI contralaterally 10 years ago with only minimal, dull noise perception, which was why he had kept it switched off; when tumor growth on the second side made surgery necessary, ABI insertion was planned and completed successfully (Case 13). The patient in Case 16 also had undergone a previous ABI surgery 12 years ago with no useful perception, and therefore desired to undergo another attempt.

In 2 patients a reexploration surgery was undertaken because of suspicion of electrode dislocation; after previous radiotherapy, the patient in Case 3 did not experience any sound perception, either after implantation or after revision, and despite intraoperative E-ABR. In the patient in Case 10 some electrode dislocation was presumed due to side effects at first implant activation; after surgical revision this patient attained useful auditory perception.

**Preoperative Auditory Function**

Some useful preoperative auditory function was present in 3 of 16 cases. In Cases 13 and 15, large tumors with brainstem compression made surgery absolutely necessary. In Case 8, the patient had bilateral large tumors with unilateral deafness and useful hearing on the contralateral side; furthermore, she suffered from orbital tumor infiltration in her only functioning eye; with the prospect of impending deafness on the second side, unilateral tumor surgery on the deaf side and simultaneous ABI insertion appeared reasonable. All the other patients suffered from bilateral deafness (which is calculated from the date when the second side becomes deaf) for 0–180 months, 3 years on average (mean 36.3 months, SD 54.4 months), on the ipsilateral side. Duration of deafness was longer on the contralateral side, 9 years on average (101.1 months, SD 78.6 months) (Table 1).

**Postoperative Auditory Function With ABI**

Of 18 implanted devices 16 could be activated, with successful hearing rehabilitation documented by audiometric laboratory tests (Tables 1 and 2). Those 16 patients have been using their devices regularly every day for an average of 8 hours. Nine patients are active in their profession or in an education and/or training program.

**Hearing Function During the 1st Year of Follow-Up**

Word recognition results as documented by MTP tests at 3-month, 6-month, and 12-month follow-up are given in Table 1. Pure auditory-only (MTPa) word understanding ranged from 16.7% to 100% (mean 71.6%, SD 23.2%) at 3-month follow-up; it increased to an average of 82.5% (SD 14%) at 6 months and to 88% (SD 16.6%) at 12 months. Combined audiovisual testing (MTPav) resulted in 95.8%–100% understanding (mean 99.5%, SD 1.4%) at 3 months and increased to 100% at 6 months. At 12 months, 12 patients had achieved 100% and 3 had 87%–96% MTPav.

Open-set speech understanding in pure auditory mode by HSM test could be identified in 8 patients at 3 months, at scores of 2.8%–99.1%. At 6 months, 10 patients had an average of 22.3% speech recognition (SD 33.5%). At 12 months the average HSM score was 32.5% (SD 33.5%) in 12 (75%) of 16 patients.

**Hearing Function at 24-Month Follow-Up**

As shown in Table 2, at 24 months the overall results in word recognition were similar to the 1st year, with an average of 88.7% (SD 12.7%) correct MTPa, and speech recognition improved further in 7 patients, resulting in an average HSM score of 41.4% (SD 32.4%). Eight of 16 patients (50%, 95% CI 25%–75%) had HSM scores of 25% or higher after 24 months. Twelve patients (75%, 95% CI 48%–93%) achieved HSM scores of at least 10%.

**Peak Performance at 1–5 Years of Follow-Up**

Further increases in word and sentence recognition could be identified in some patients after the first 2 years, as is shown in Table 2. Meanwhile, 13 patients experienced some open-set speech perception at scores of 6% to 99% (mean 45.6%, SD 35.2%); of those, 9 patients have attained 25% or higher HSM scores (outcome Classes 1, 2, or 3).

Excellent- and good-quality hearing (Classes 1 and 2) was present in 6 patients (Cases 1, 2, 4, 7, 11, and 18). Medium-quality understanding (Class 3) was found in 3 patients (Cases 12, 14, and 17), and fair quality (Class 4) was found in 4 patients (Cases 8, 13, 15, and 16). Class 5 outcomes with no open, but closed word understanding (≥ 40%) were found in 3 patients (Cases 5, 6, and 10).

**Characteristics in the Different Outcome Groups**

Classes 1, 2, and 3 (25%–100% HSM score) contained 5 men and 4 women, mean age 35.9 (SD 13.9) years, with large tumor volumes (an average of 12.7 cm$^3$) and extensions (7 severe cases [T4; Hannover classification]), no preference for side (3 right and 6 left ABI), a short period of ipsilateral deafness (average 16.6 months, SD 30.7 months), and a high number of active electrodes (7.8, SD 1.3). The duration of contralateral deafness was 71 months (SD 59.7 months).

In Classes 4 or 5 the average age was 41.2 years (SD 10.9 years), tumor extensions were T4 according to the Hannover classification, tumor volume was 20.9 cm$^3$, duration of deafness was 72 months (SD 77.8 months), and the number of electrodes was 7.5 (SD 3.5). The duration of contralateral deafness was 180.7 months (SD 53.2 months).
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Deafness (mos)</th>
<th>Tumor</th>
<th>24-Mo FU Scores</th>
<th>Peak Performance Scores</th>
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<tbody>
<tr>
<td></td>
<td>Ipsilat</td>
<td>Contralat</td>
<td>Extension†</td>
<td>Radio Vol (cm³)</td>
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<tr>
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<tr>
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</tr>
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<td>SD</td>
<td>54.4</td>
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* Act Els = active electrodes; radio = radiological.
† Tumor extension was categorized by extended Hannover classification.
Factors Related to High-Quality Outcome in Patients With ABI Procedures

No significant correlation was found between tumor volume and HSM score after 24 months (Fig. 3). Note, in particular, that the patient with the third-largest tumor had the second-best outcome. On the other hand, a relatively small tumor can be associated with clinical failure in speech recognition as well.

We found an inverse relationship between duration of deafness and 24-month HSM scores (Fig. 4). Short duration of ipsilateral deafness is no guarantee, but an HSM score of 50% or better is not found at more than 24 months’ duration (Fig. 4 upper). The correlation becomes obvious when considering the sum of ipsilateral and contralateral deafness periods: at less than 200 months’ “deafness sum,” some HSM score is likely to be obtained. At up to 100 months’ deafness sum, an HSM score ≥ 40% is achieved. At periods longer than 200 months an HSM score is unlikely, despite adequate electrode placement with 6–12 auditory electrodes. The higher the cumulative deafness load, the less likely that any HSM score will be achieved (the Kendall tau correlation coefficient = –0.39, p = 0.034) (Fig. 4 lower).

The overall correlation of the number of active electrodes and 24-month HSM scores was not significant (Fig. 5). The significance of this relationship was negated by 2 outlier patients who had poor outcomes, despite a maximum of active electrodes. The latter are probably attributable to long-lasting deafness (see Fig. 4). In patients whose total deafness duration was less than 240 months, higher numbers of active electrodes were significantly associated with better outcomes.

Good short-term results in terms of the MTPa score are strongly correlated with favorable long-term outcomes as measured by the HSM score (Fig. 6). Remarkably, this cross-correlation was stronger than the longitudinal relationships between 3-month and 24-month scores for MTPa (τ = 0.69) and HSM (τ = 0.72), and stronger than the cross-sectional correlations between MTPa and HSM at 3-month (τ = 0.63) and 24-month follow-up assessments (τ = 0.55, p < 0.001 for all correlations).

Illustrative Cases

Case 1

This 36-year-old man had a compressive left-sided...
The tumor grew over the midline, with severe compression at all levels (midbrain, brainstem, medulla) and planes (anterior, at meatus, and dorsal to meatus). The patient had undergone hearing conservation surgery 12 years before and had now become functionally deaf in his other ear and developed dramatic ataxia with frequent falls. He underwent complete tumor resection and ABI insertion, and his facial nerve was reconstructed by direct nerve suture during the same surgery and recovered well within the 1st year. Immediately after ABI activation he was able to lead a conversation, and obtained open-set speech perception of 87.7% within the first 6 months.

Meanwhile he has undergone further surgery of the contralateral schwannoma recurrence, of peripheral schwannomas, and of a spinal meningioma.

**Case 2**

This 22-year-old man had been deaf on the right side since tumor resection in 2004, and developed deafness in his other (left) ear in 2006, along with tumor progression on the left side, with ataxia and CN V deficit. Major tumor compression was exerted at all levels, at the plane of the meatus, the brainstem, and the cerebellum.

At surgery (Fig. 2A) of the large tumor, motor CNs were found to be spread apart and elongated, and especially caudal CNs were adherent to the tumor, but were all preserved, as was the facial nerve (Fig. 2B), under continuous control by motor CN monitoring. By opening the lateral recess above the caudal CNs and using CN IX as a guide to the region of the cochlear nucleus, this was exposed (Fig. 2C) and investigated by stimulation testing using a quadripolar test electrode (Fig. 2D). At all electrode combinations E-ABR could be registered and reproduced, and consisted of the characteristic peaks III, IV, and V (Fig. 2E). Therefore, a permanent ABI electrode was placed and fixed (Fig. 2F). At first fitting the patient had immediate useful auditory function, but experienced a significant improvement toward better voice and speech recognition after 6 months, with a score of 60% on the HSM test. Over 4 years he showed further improvement, to 84.9% on the HSM test.

**Discussion**

Auditory brainstem implants offer a realistic option for hearing rehabilitation in patients with bilateral retrocochlear deafness. In the early stages of this treatment modality results were scarce, and recent reports in the last decade show more satisfying hearing quality. In part this may be attributed to improved speech processor technology. Most centers encounter a slow long-term improvement in auditory perception in the individual patient far beyond 1 year after activation. The most frequently considered factors related to results are discussed below.

**Test Modalities**

Single- to multiple-syllable–containing word series and sentence tests were originally developed for patients with cochlear implants to evaluate their speech comprehension. The HSM sentence test has become an established method for evaluation of speech recognition; it identifies patients’ abilities at quiet and at noisy conditions and is mainly applied in cochlear implants. The test battery contains an extensive number of sentence series in all languages to guarantee several completely unknown tests at each setting with the patient.

**Nonresponder Status**

In reports on ABI surgery, some patients do not experience any hearing sensations. There are some negative predictors such as large tumors with distorted brainstem...
Speech perception in patients with auditory brainstem implants

anatomy, previous radiation or radiosurgery, long-standing deafness, hemorrhage, and meningitis that have been encountered so far. Nonetheless, none of these factors means an absolute exclusion from ABI surgery because there are single positive results despite those negative predictors. Gray et al. reported on 5 (22%) of 23 patients with neurofibromatosis who had no auditory sensations; 3 of them had large tumors. In the series by Schwartz and coworkers there were 13 (15%) of 86 nonresponders and 12 (15%) of 82 lost to activation and follow-up. Kanowitz and colleagues found 2 nonresponders (11%) of 18 implants. The failure rate in our series was 2 (11%) of 18 activated implants, with 2 specific causes identified—previous radiation and anatomical variation. Although radiotherapy and radiosurgery are no longer factors for primary exclusion from ABI trial, overall results are less favorable after previous radiation. After radiosurgery, patients with previously implanted devices obtained some environmental sound recognition, but never any open speech recognition, either with cochlear implants or with ABIs.

Tumor Size, Brainstem Compression, and Deformation

As the first investigators to analyze the influence of tumor presence or absence on ABI outcome, Colletti and colleagues compared ABI outcome in patients with tumors to those without tumors, and identified better results if no tumor had ever been present: patients without tumors achieved an average of 59% and patients with tumors had a maximum of 11% of open-set speech understanding with ABI. Those were the only ABI-treated patients reported to have obtained some open-set speech recognition.

It is not clear whether molecular changes due to a previous tumor or the deformation of brain structures is responsible for less favorable results in tumor cases. As a consequence, some authors advocate ABI surgery at stages in which tumor size is small or medium to avoid brain deformation, difficult surgery, and unfavorable results. Despite early surgery in tumors that were at the small stage, open speech recognition was not observed in their series.

In our series the most favorable results were achieved in 4 patients with very large tumors and in 1 patient with a medium-sized tumor. As is evident in Fig. 3, there were small tumors with poor or moderate outcome and several large tumors with the best HSM results. Our results correspond to those reported by Otto et al., who could exclude any relevant influence of tumor sizes on ABI outcome. Beyond tumor size, the induced changes on the brainstem in the present series were quite severe, with deformation of the brainstem by tumors growing up to the midline or even across it (Fig. 1). This might have led to structural and functional changes within the cochlear nucleus, the olivary complex, and the lemniscus of the auditory pathway by chronic compression. If several essential relay stations of the pathway are deteriorated, a less favorable outcome with ABI could be understood. Furthermore, the whole surgery is more difficult and more risky, because the relationship between the brainstem and the tumor is quite adhesive, and the brainstem is deformed and softer and more vulnerable than usual. The identification and opening of the lateral recess may be more difficult and harmful as well.

Electrodes and Stimulation

Discussion on electrodes has to include the number, type, and site of implanted electrodes.

Electrode Number. A certain number of electrodes is necessary to represent different frequencies and pitch, especially high pitch, as a precondition to speech understanding. The Med-El implant used in this series contains 12 channels. An active channel count of 8–9 channels was mostly associated with open-set auditory understanding; still, with only 6 channels activated, some hearing of this quality was achieved. Regarding experiences with other devices, electrode numbers cannot be directly compared. Kuchta et al. investigated the nucleus 8-electrode device and found no improvement in speech discrimination at more than 5 auditory electrodes. Most authors found good results if 60%–75% of available electrodes or channels gave auditory perception.

Electrode side effects are a common finding in ABI programming. Up to 42% of activated electrodes have been reported to cause unintended side effects. There are 2 groups of such electrodes; those causing only nonauditory side effects and those with a combination of auditory and nonauditory effects. The first case is simple to solve by programming out. In the second situation this programming out may have disadvantageous consequences. Different sites of stimulation may lead to different pitch percepts and code for different frequencies. This diversity is reduced or lost by programming out the auditory electrodes with side effects.

Electrode Types: Superficial or Penetrating. As an alternative to surface electrodes, an array with combined penetrating and superficial electrodes has been studied by Otto and colleagues. The idea was to obtain a better coupling with neurons representing different pitch. In practice, the advantages were a low electrical charge and a wide range of pitch perception as anticipated, but the clinical results with those penetrating electrodes were disappointing. In comparison with simultaneous superficial stimulation, the results yielded by penetrating electrodes were less positive, with fewer electrodes being ready to use and worse performance. As a consequence, the increased risk from placing penetrating electrodes at the cochlear nucleus is not worthwhile, and this type of ABI at the cochlear nucleus has been abandoned.

Electrode Site: Cochlear Nucleus or Inferior Colliculus. As an alternative to the cochlear nucleus, Colletti and colleagues investigated the inferior colliculus by using a superficial electrode array. Recently, a penetrating electrode model for the inferior colliculus was designed and tried out experimentally and clinically by Lenarz and coworkers. In view of the tonotopic gradient within the inferior colliculus, it was presumed to be an ideal target for such electrodes. Again the penetrating electrode showed the advantages of low electrical charge, and some improvement was observed regarding loudness, pitch, and temporal and directional cues, but open-set speech perception was not obtained until now. As of this writing, this target has not yielded any superior outcome, and altogether the inferior colliculus has not led to any signifi-
cant improvement, either by superficial or by penetrating electrodes. The lack of responsiveness of the colliculus is speculated to be caused by cochlear nucleus compression. Experimental data by acute cochlear nucleus compression suggest immediate decrease of function in the inferior colliculus and recovery after nucleus release. Whether reversibility of this phenomenon is maintained in chronic compression is unknown.

A corresponding investigation by Sekiya et al. was undertaken by performing experimental acoustic nerve compression, resulting in degeneration of 50% of neurons within parts of the cochlear nucleus, specifically the dorsal and the posteroventral. One may speculate that a similar degeneration occurs at the onset of deafness due to schwannomas, which in turn could be stopped or attenuated by early stimulation of the cochlear nucleus by an implant.

To date the functional results in trials with inferior colliculus implants are less favorable than previously expected; this may be attributed to proximal degeneration, due in part to the complexity of the structure and to difficulties in exact electrode placement.

Chronic degeneration may be a cause of limited success in both nucleus and colliculus implants. Colletti et al. speculate that the cochlear nucleus contains a group of neurons for “a separate pathway of auditory processing that is specialized for modulated sounds,” which they presume to be critical for speech understanding and which might be damaged in NF2. In his analysis of the physiology of the auditory pathway Möller outlines the functional anatomy of the cochlear nucleus, with 3 divisions of each auditory neuron performing some kind of parallel processing of incoming information. Whether a VS or its removal causes the degeneration of 1 group of these neurons is open to speculation. On the other hand, we have to assume that a significant number of neurons will have been lost if implantation is performed at a late stage, and this will result in limited hearing quality.

Duration of Preimplant Deafness

A short duration of deafness is identified as a positive factor in many previous reports; however, there is a large variation in absolute data. Most authors discuss periods of a few years to more than 10 years, and advocate ABI surgery before 10 years of deafness.

Corresponding to experiences with cochlear implants, durations of a few years must be considered and might decide whether there will be any hearing sensation at all. High-quality hearing, however, is dependent on much shorter periods. In our series the best auditory outcome (open-set speech understanding greater than 50%) was related to short periods of deafness and was never identified in deafness periods of more than 24 months. Long-standing deafness did not prevent recovery of some useful hearing, but a long deafness duration was invariably associated with rather moderate results in speech perception. On the other hand, a short period of deafness on the implanted side was no guarantee of excellent results. In these patients the duration of contralateral deafness could also be of some importance. The cumulative deafness load by the sum of ipsi- and contralateral deafness periods is of significant importance.

Studies on cochlear implants in postlingual deaf patients also show an influence of the duration of deafness on functional outcome, especially speech perception: Oh et al. report 5 years of deafness as critical, whereas Durinsin et al. found significant differences when comparing scores from before and after 6 months of deafness; also, over long training periods patients could not catch up in speech recognition. Peasgood and colleagues found no speech recognition in adults with late cochlear implants, and much less favorable results than are associated with cochlear implants in general.

Study Limitations

Due to the small size of our cohort it is impossible to compute reliable multivariable prediction models. Therefore, it cannot be decided at this stage whether, for example, duration of deafness or the number of active electrodes (or both) independently contribute to the clinical outcome.

Patients’ Judgment on ABI Outcome

Patients’ satisfaction with their ABI is only partially dependent on laboratory hearing results. Independent of their individual result, all of these patients with functioning ABIs are very satisfied with their reparticipation in normal life, and their increased awareness of the movements of others and their surroundings; they do hear and can control their own voice, and they are aware of continuous improvement over at least 3 years or longer. A patient’s personal attitude, with an interest in learning and training, plays an important role in progress and in satisfaction; on the other hand, a patient’s individual constitution and the presence of useful eyesight is helpful in relearning speech comprehension, and the absence of frequent further tumor surgeries is important in this respect as well.

Still, patients with severe NF2 manifestation and frequent tumor surgery are those who most often will need auditory rehabilitation by ABI and will appreciate also a small improvement in communication.

Conclusions

So far, no other study has reported on such short intervals of deafness and high rates of speech perception. Our study documents that open-set speech perception is feasible on a regular basis in patients with NF2, and also in brainstem compressive tumors. The presented results are similar and in part better than in the nontumor situation, and they are approaching the result level of cochlear implants.

If high-quality hearing with speech perception is the goal, there are 2 factors lying in the surgeon’s hands: first, adequate timing of ABI surgery may help to avoid time-dependent degeneration; and second, utmost microsurgical care at the cochlear nucleus will avoid any manipulation or compression of this structure and its microvascular supply.

These data should not be misunderstood as a de-recommendation of ABI in patients with long-standing deafness: to achieve medium-quality hearing with good audiovisual speech perception conveys a major advantage for the individual patient in the quality of his or her everyday life.
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The chance to regain some auditory communication facilitates the whole management of this serious disease.

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Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Matthies, Helms, Shehata-Dieler, Müller. Acquisition of data: Matthies, Brill, Varallyay, Mlynski, Shehata-Dieler, Müller. Analysis and interpretation of data: Matthies, Varallyay, Gelbrich, Hagen, Mlynski, Shehata-Dieler. Drafting the article: Matthies. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Matthies. Statistical analysis: Matthies, Gelbrich. Administrative/technical/material support: Brill. Study supervision: Matthies.

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