Auditory brainstem implant in auditory rehabilitation of patients with neurofibromatosis type 2: Hannover programme


Abstract
An auditory brainstem implant (ABI) is indicated for patients suffering from bilateral neural deafness. The most affected patients are those with neurofibromatosis type 2 (NF2). An implantation is possible either at the same time as, or after, surgical removal of an acoustic neuroma. This paper demonstrates the results of eight out of 11 patients with NF2, seven of whom received an ABI after tumour removal. Pre-operatively, all of them were deaf. Post-operatively, the first fitting served to determine the individual stimulation parameters for each electrode. The stimulation-dependent side-effects were eliminated by reducing the stimulus intensity without causing negative effects on the hearing with the ABI. Only in one case was an open set understanding achieved within the first year. However, all patients had a better speech understanding when they combined their hearing with the ABI and their lip-reading abilities. There is no correlation between the performance with ABI and the tumour size or the duration of deafness.

Key words: Brain Stem; Prosthesis Implantation; Neurofibromatosis 2; Rehabilitation of Hearing Impaired

Introduction
An auditory brainstem implant (ABI) is indicated for patients suffering from bilateral neural deafness. Patients with neurofibromatosis type 2 (NF2) are particularly affected because of the high incidence of bilateral acoustic neurina. The ABI differs as compared to the cochlear implant (CI) only by the electrode design itself and its anatomical location where the electrode has to be placed. The electrode consists of 22 (Nucleus) or 16 (Clarion) small platinum plates which are located in three rows on a Dacron mesh. The electrode must be placed in the lateral ventricle on the ventral cochlear nucleus on one side only. So far 12 patients have been implanted (eight Nucleus 21+1 ABI, two Nucleus 24M ABI, two Clarion ABI). In this paper the results of the eight patients implanted with the Nucleus 21+1 ABI will be demonstrated.

Materials and method
After a standardized audiological and neurosurgical examination the bilateral neural deafness and the individual status of the NF2 were evaluated. Depending on the tumour size and the compression of the auditory brainstem the indication for tumour removal and, at the same time, an implantation is determined in an interdisciplinary approach by the neurosurgeon and the ENT surgeon. During implantation extensive multimodal monitoring is used to optimize the location of the electrode. Only in the case of a reliable response from the auditory central pathway while using a test electrode is an implantation successful. The first fitting takes place six to eight weeks post-operatively while determining the individual map of the electrode configuration. Before determining the individual threshold (T-level) and uncomfortable level (C-level) the side-effects are eliminated while reducing the energy or changing the stimulation configuration. This is followed by a pitch ranking and the fitting of the the speech processing strategy. Initial basic hearing and speech training for 14 days, comparable to the rehabilitation with CI, then takes place. During the regular follow-up visits refitting and correction of the pitch ranking is performed. The results of the individual performance is tested by means of a standard test battery for CI patients. In this study all patients were tested with live voice vowel and consonant confusion test and speech tracking.
Results of hearing and lip-reading are displayed. The horizontal line indicates the lip-reading ability alone. Apart from two patients, no patient had any residual hearing before surgery. In seven cases a tumour removal was necessary before implantation (Table I).

## Discussion

Auditory rehabilitation with cochlear implants has become an established procedure. Although the indication for a CI has been extended, there is still a contra-indication for retrocochlear disorders. But with the introduction of the ABI, especially in patients suffering from NF2 and bilateral acoustic neuroma, an alternative is provided. Nevertheless the primary aim of the treatment of those patients is the recovery of hearing after reducing or extirpating the tumour. Slattery et al. found that the recovery of hearing is not dependent on the location but on the tumour size and the radicality of the tumour removal. The risk of total hearing loss is estimated to be up to 35–45 per cent. Bance and Ramsden see the challenge of the NF2 treatment as being in the improvement of the quality of life, which means that before the decision is taken to implant an ABI, the hearing should be saved by tumour reduction instead of total tumour removal, if possible.

The first patients with NF2 and implanted with an ABI achieved sound discrimination without causing pathologic side-effects to the neighbouring neural structures in the brainstem. The sound discrimination is comparable to that of a single channel CI. New electrode designs improved the performance with the ABI. In animal experiments with penetrating electrodes, better electrophysiological results could be obtained with lower thresholds and increased dynamic ranges. However, histological analysis showed tissue damage that has not allowed the use of these electrodes in humans until now.

### Table I

<table>
<thead>
<tr>
<th>Patient</th>
<th>Onset of deafness</th>
<th>Tumour size (cm)</th>
<th>Age at implantation (years)</th>
<th>Number of electrodes used</th>
<th>Side effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ac</td>
<td>1994</td>
<td>No</td>
<td>52</td>
<td>15</td>
<td>Vertigo, sensation in the leg</td>
</tr>
<tr>
<td>Co</td>
<td>1985</td>
<td>4×5</td>
<td>36</td>
<td>5</td>
<td>Vertigo, tickling in the tongue</td>
</tr>
<tr>
<td>Ku</td>
<td>1993</td>
<td>4</td>
<td>31</td>
<td>12</td>
<td>Sensation in the extremities</td>
</tr>
<tr>
<td>Lü</td>
<td>1995</td>
<td>1</td>
<td>29</td>
<td>12</td>
<td>Vibration in the hand</td>
</tr>
<tr>
<td>Pa</td>
<td>1998</td>
<td>2</td>
<td>49</td>
<td>9</td>
<td>Vertigo</td>
</tr>
<tr>
<td>Pe</td>
<td>1983</td>
<td>3</td>
<td>34</td>
<td>7 (17)</td>
<td>Vertigo, sensation in the body</td>
</tr>
<tr>
<td>Pr</td>
<td>1996</td>
<td>3</td>
<td>24</td>
<td>9</td>
<td>Pounding in the ear</td>
</tr>
<tr>
<td>W-P</td>
<td>1998</td>
<td>1.5</td>
<td>43</td>
<td>10</td>
<td>Vertigo</td>
</tr>
</tbody>
</table>

**Fig. 1**
Vowel confusion test - scores for each patient over time.

**Fig. 2**
Consonant confusion test - scores for each patient over time.
Comparing the results of CI patients and ABI patients, lower thresholds for the low frequencies with a comparable dynamic range are found in ABI patients. The difference in temporal resolution and integration led to improved speech strategies. All patients involved in this study improved their communication ability. At first all patients achieved a better sound discrimination with an overall improvement in communication combined with lip-reading. But already after six months one patient has achieved an open-set speech understanding. Otto et al. also observed an open-set speech understanding in three patients within the first year, the 20 other patients achieved a significantly improved communication in combination with lip-reading. In the literature and in our experience there is no correlation found between the tumour size, the pre-operative hearing loss and the psychophysical data of the speech strategies. However, all studies confirm that there is, compared to CI patients, often a change in the individual map within the first year.

ABI is an established treatment in the rehabilitation of bilaterally deaf patients suffering from neural disorders such as NF2. If there is no auditory recovery expected after tumour removal, an implantation should take place in the same session. The post-operative fitting depends on the detection of side-effects, that can be eliminated by reducing the stimulus intensity and changing the electrode configuration. The communication skills improve in all cases, but an open-set speech understanding is still difficult to achieve. In the long-run better speech processing strategies should improve the stimulation modalities as well as the performance.

References

7 Bance M, Ramsden RT. Management of neurofibromatosis Type 2. Ear Nose Throat J 1999;78:91–6

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