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Auditory brainstem implants: current neurosurgical experiences and perspective

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Abstract

The objective of this study was to present aspects of the current treatment protocol, such as patient evaluation and selection for therapy, multimodality monitoring for optimal auditory brainstem implant (ABI) positioning and radiological evaluation, that might have an impact on the functional results of ABI.

Out of a series of 145 patients with bilateral vestibular schwannomas 10 patients received an ABI, eight of which are reported here. Patient selection was based on disease course, clinical and radiological criteria (according to the Hannover evaluation and prognosis scaling of neurofibromatosis type 2 (NF2)), extensive otological test battery and psycho-social factors. ABI placement was controlled by multimodality electrophysiological monitoring in order to activate the auditory pathway and to prevent false stimulation of the cranial nerve nuclei or long sensory or motor tracts. Results of hearing function were correlated with patients' ages, duration of deafness, tumour extension, tumour-induced compression or deformation of the brainstem, and numbers of activated electrodes without any side-effects.

Out of 59 patients with pre-operative deafness eight patients received an ABI of the Nucleus 22 type. All these patients became continuous users without any side effects and experienced improved quality of life. Speech reception in combination with lip-reading was markedly improved, with further improvement over a long period. A short duration of deafness may be favourable for achieving good results, while age was not a relevant factor. Lateral recess obstruction may necessitate a more meticulous dissection, but did not prevent good placement of the ABI in the lateral recess. Pre-existing brainstem compression did not prevent good results, but brainstem deformation and ipsi- and contralateral distortion were followed by a less favourable outcome.

Among the factors that can be influenced by the therapy management are the selection of patients with a slow progressing NF2 disease, a short duration of deafness, a careful analysis of brainstem deformation and consideration of either side for implantation. Long-standing brainstem deformation might not lead to recovery, but instead lead to a low number of active electrodes and possibly only moderate results.

ABI treatment is a safe procedure that can increase a patient's quality of life considerably. ABI placement along with neurophysiological control helps to prevent side effects and to improve acoustic activation. Further studies on structural and functional changes of the brainstem after previous tumour compression and distortion should increase our understanding and facilitate a decision on the best side for ABI implantation.

Key words: Brain Stem; Prosthesis Implantation; Neurosurgical Procedures

Introduction

Auditory brainstem implants (ABI) were designed and first applied 20 years ago in an attempt to restore some auditory function for patients with bilateral retrocochlear deafness.[1,2] Except for the improvements in the prevention of bilateral deafness by function-conserving microsurgery of schwannomas[3–5] and even bilateral schwannomas,[6,7] there is no alternative therapy in case of lost nerve function. While there are promising trials to restore the auditory pathway by biological means,[8] at present the major concerns for improvement of the technological concept of restitution lie in the optimization of the shape of the implant,[9–13] the site of implantation, the mode of positioning and the pattern of stimulation.[14]

The present study focused on criteria for patient selection, improvement in ABI positioning and on clinical parameters affecting the functional outcome.

Patients and methods

Out of a series of 145 patients with neurofibromatosis type 2 (NF2) treated microsurgically from 1978

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to 1998, 10 patients were selected for implantation of an ABI. Patient evaluation and selection for ABI were based on the Hannover NF2 manifestation and prognosis scaling,[15] absence of actual signs of serious disease progression, an extensive audiology test battery,[16,17] successful training in lip-reading, stability of social and professional life conditions, and participation in three patient counselling interviews. Neurophysiological monitoring included recording of median and tibial nerve somatosensory evoked potentials (MSEP, TSEP), auditory brainstem responses (ABR) and electrical ABR (EABR), motor cranial nerve electromyography (EMG) of the cranial nerves VIIth, IXth, Xth, XIth and XIIth by bipolar registration with monopolar needle electrodes. Besides the control of patient position and tumour resection, monitoring was applied during ABI placement and test stimulation to visualize activation of the auditory pathway and exclude false activation of other tracts or nuclei. Outcome was evaluated by an audiology test battery and correlated with clinical and radiological parameters such as patients' ages, duration of deafness, tumour extension, brainstem compression, dislocation or distortion ipsi- and contralateral, tumour extension to and/or tumour obstruction of the lateral recess.

Results

Incidence of neurofibromatosis type 2

In a series of 1 750 patients treated from 1978 to 1998 for vestibular schwannomas (VS) 145 patients presented with bilateral tumours (8.3 per cent); of those 145 patients 195 VS have been resected so far. Pre-operative auditory function was present in 126 of 195 tumour-endangered ears (65 per cent), while post-operative auditory function was preserved in 36 of 126 ears (29 per cent). Of 59 patients with pre-operative functional deafness, 10 patients were, so far, selected for implantation of an ABI, the results of eight are to be presented. In all these eight patients the Nucleus 22 device was used, and in seven of these patients within the European Study Group.[18]

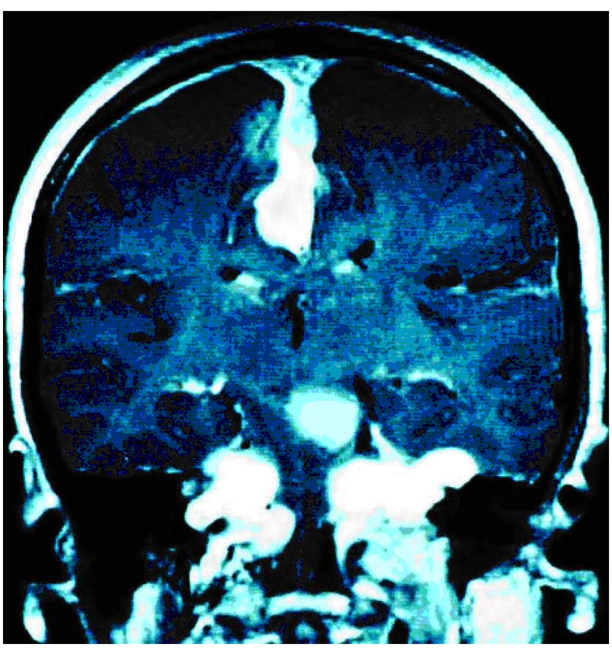

FIG. 1

Multiple cranial nerve origin of cerebellopontine angle schwannomas and generalized meningiomatosis are indicators of severe neurofibromatosis type 2. Its most severe type involves the sinuses, especially the superior sagittal sinus.

Indication for auditory brainstem implant surgery

All the patients had been known to the NF2 out-patient clinic for a minimum of six months to 10 years, on average 3.5 years. All the eight patients received a low or moderate scaling factor in the NF2 manifestation classification and a good rating regarding life prognosis and presented with a stable life phase of their NF2 disease. None of the patients had a grape-like tumour formation suggesting a tumorous change of the cranial nerves (Figure 1), none had a VS recurrence before a five-year-interval after previous tumour surgery according to the Nordstadt protocol, none had signs of meningiomatosis or of sinus infiltration (Figure 1), and none had progressive multiple cervical tumours. All patients' social (and, in six, professional) situations were stable and supportive of the planned measures. All patients had undergone lessons and succeeded in improving lip-reading.

Neurophysiological monitoring

During monitoring control of ABI placement all the eight patients had reproducible electrically-evoked auditory responses EABR.[17] At test stimulation SEP were stable and the majority of activated electrodes were free of any signs of activation of motor cranial nerves, especially of nerves VII, IX and X.

Clinical parameters and outcome

(Tables I and II). Patients' ages varied from 24 to 52 years, with a mean of 37 years. They had been deaf for two to 120 months, on average 26 months. Most of the tumours were large and brainstem compressive (Table I). Except for one patient (*Case 2*) who had been operated on for his tumour 1.5 years before and was then operated on without any signs of recurrence for implantation of the ABI, all the other patients needed tumour resection and received testing and implantation of the ABI at the same setting. One patient (*Case 5*) needed a revision to

TABLE I

CLINICAL FACTORS AND SURGERY

Patient	Age (years)	Deaf (months)	Tumour mm,	Ext.	Surgery tumour - ABI
1 TL	29	5	15	T2	05.96
2 AA	52	19	40	T4a	12.94–07.96
3 BC	35	120	50	T4b	07.96
4 MP	24	2	37	T4a	11.96
5 TK	31	36	50	T4b	11.96–03.97
6 JP	34	12	34	T4b	05.98
7 HP	49	4	27	T3b	10.98
8 CW	43	12	30	T4a	11.98

ABI = auditory brainstem implant.

TABLE II
TUMOUR EVOLUTION

Patient	Course
1	Bilateral hearing preserved for 7 and 9 years, recurrences with deafness, VII–XII palsies
2	No recurrence, stable
3	Bilateral tumour progression
4	Small recurrence, but visual deterioration of last eye
5	Large recurrence after 9 years
6	Bilateral recurrences and progression
7	Unilateral recurrence
8	Unilateral recurrence and progression

correct the positioning of the ABI and stop false activation of long tracts (Table II).

Functional outcome is regarded as useful by all the patients and all are using the implant every day and would not live without it any more. Among the various audiology tests speech tracking is reported here as a mode that reflects the individual patient's coping with listening to speech. Results vary from 9 words/minute to 41 words/minute. Most patients showed an improvement over time.[19] This test and the data correspond very well to the patient's general performance in conversations with different doctors.

Correlations of clinical and radiological factors and outcome

Age. (Table III). The youngest of the implanted patients achieved the best results (24 years, 41words/minute) and the second best result was obtained by the eldest patient (52 years, 36 words/minute). Obviously, youth is not a pre-condition for a good result.

Duration of deafness. (Table III). The best result was obtained by the patient with the shortest period of deafness (two months, 41 words/minute). But also patients experiencing 12 to 19 months of deafness presented with very good results. A fairly long duration of deafness of 36 to 120 months was followed by medium hearing quality.

Relation of brainstem and tumours. (Tables IV and V). In six of the eight patients the schwannoma showed extension towards the lateral recess. In four of these patients this tumour extension led to radiologically evident obstruction of the lateral recess, and at surgery special dissection was necessary to remove the adherent tumour and re-open the exit of the lateral recess. In two patients, although the tumour was growing towards the lateral recess, there was CSF visible and CSF circulation was found preserved at surgery. In relation to the outcome, this was *not* a factor of importance. Obviously, the adhesions and the need for more meticulous dissection did not mean a disadvantage in placing the ABI into the lateral recess.

TABLE III
CLINICAL FACTORS AND OUTCOME

Patient	Age (years)	Deafness (months)	Speech tracking (w/min)
1 TL	29	5	20
2 AA	52	19	36
3 BC	35	120	12
4 MP	24	2	41
5 TK	31	36	9
6 JP	34	12	17
7 HP	49	4	14
8 WP	43	12	30 (early)

w/min = words per minute.

TABLE IV
LATERAL RECESS OBSTRUCTION

Patient	Lateral recess	Speech tracking (w/min)
1 TL	Tumour	20
2 AA	(Tumour)	36
3 BC	Tumour, CSF	12
4 MP	Tumour	41
5 TK	CSF	9
6 JP	Tumour	17
7 HP	–	14
8 WP	Tumour, CSF	30

( ) = previous tumour surgery more than 1 year ago.
w/min = words per minute.

Most patients had schwannomas of considerable size with evident brainstem compression, and in most cases the tumours on either side were compressive (Figure 2). Careful analysis of the change in brainstem delineation reveals two different modes of deformation, an even compression of the whole laterality of the side of the brainstem or a mode of indentation and displacement of a part of the brainstem, leading to a torsion or distortion of the less displaced parts (Figure 3). In two patients brainstem deformation showed this kind of distortion before tumour removal and ABI implantation; both these patients showed the weakest functional outcome of the whole group. These same two patients and one further patient (three of the eight patients) had also had a distortion on the contralateral side before the previous surgery of the contralateral tumour. This third patient had medium quality of function.

Correlation of activated electrodes and outcome

(Table VI). Although a high number of active electrodes may be advantageous for good hearing quality, it is no guarantee. The three most successful patients had nine, 10 and 15 activated electrodes. The patient with the lowest number of activated electrodes, five electrodes, ranks at the lower range of functional outcome, but, a high number of active electrodes is no guarantee for good hearing, as the

TABLE V
BRAINSTEM COMPRESSION

Patient	Ipsilateral brainstem	Contralateral brainstem	Speech tracking (w/min)
1 TL	–	Distorted	20
2 AA	Displaced	–	36
3 BC	Displ., distort.	(Distorted)	12
4 MP	Compressed	Compressed	41
5 TK	Displ., distort.	(Distorted)	9
6 JP	Compressed	Compressed	17
7 HP	Compressed	–	14
8 WP	Compressed	–	30

( ) = previous tumour surgery more than 1 year ago.
w/min = words per minute

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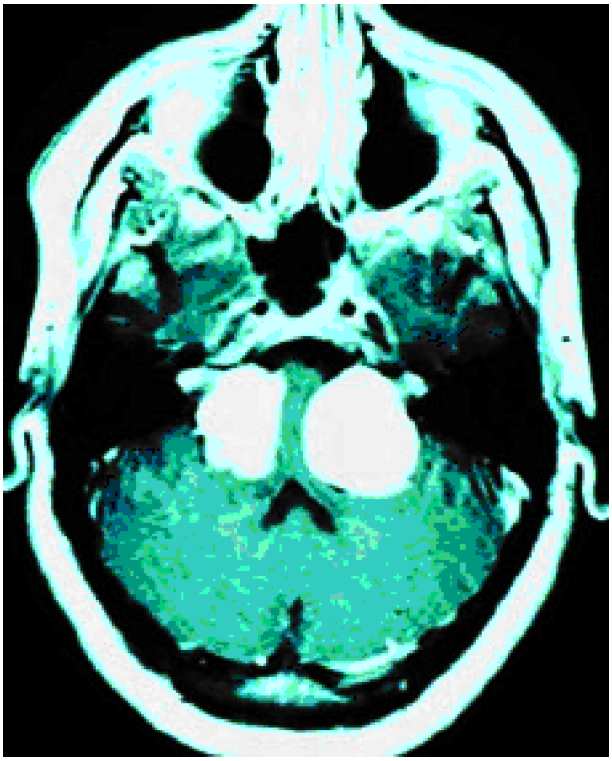

FIG. 2

The younger the neurofibromatosis type 2 patients are, the more pronounced the bilateral brainstem compression by cerebellopontine angle schwannomas may be. The compression in this patient is caused by an even mass which affects the whole of the lateral aspect of the brainstem. After complete tumour removal, the potential for brainstem re-expansion and normal brainstem function is good.

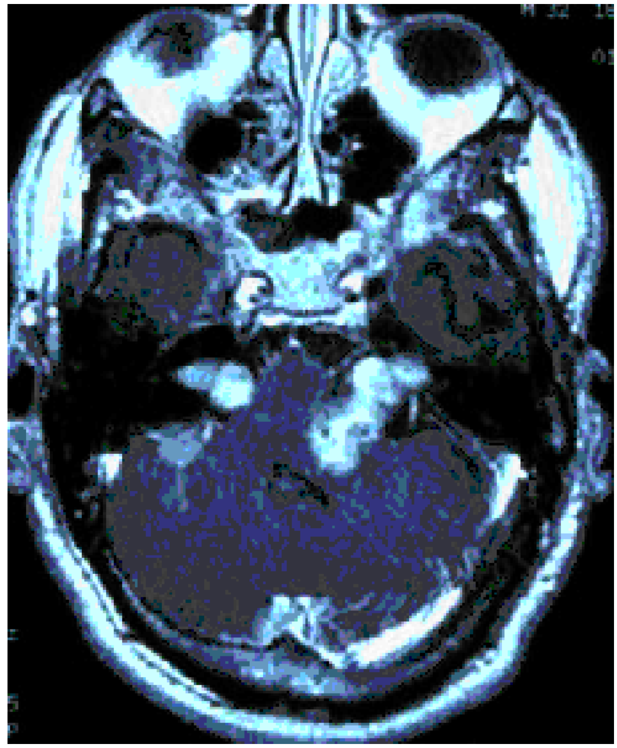

FIG. 3

Instead of homogenous tumour formation and brainstem compression as in Fig. 2, uneven tumour borders and multiple nerve origin of schwannomas may lead to indentation and deformation of the brainstem. The structural changes within the brainstem may be more pronounced and less reversible (than those of Fig. 2), i.e., brainstem expansion after tumour resection remains partial and some nuclei and tracts do not regain their functions.

weakest result was obtained by one patient with 12 active electrodes. This is one of the patients who had had the severe brainstem distortion (*Case 5*).

Discussion

A multitude of factors influence the functional outcome in ABI therapy. Some of them may be influenced by the treatment strategy, some possibly not. A stable general condition is indispensable to enable the whole procedure and to add quality of life to patients who have to undergo numerous operations. The recently developed method of evaluation of individual disease courses of NF2 provides a chance to differentiate between slow and fast progressing types of NF2 or at least of life phases respectively. All the 8 patients of this study are continuous ABI users, they judge the device as a considerable help in every day life and they do not suffer any side effects from it. Moreover, as seven of them needed tumour surgery anyway, this was no additional burden to them.

The quality in hearing performance shows a wide range however. The question arises as to whether certain parameters in some patients would have been accessible to management and to induction of a superior auditory activation. Among the factors amenable to management are the site and the side of implantation, and hereby, possibly, the number of activated electrodes.

Regarding the number of activated electrodes causing useful acoustic sensations, there were two patients with only five and seven activated electrodes respectively; all the others hear with nine or more activated electrodes. Information on the adequate position of the ABI is gained by EABR recording. A considerable number of EABR positive electrodes at surgery is judged as a positive finding; and their presence was a pre-condition to leaving an ABI in place. However, in some patients despite very good EABR a correction of placement was necessary mainly because of facial or glossopharyngeal side-effects. In some patients obviously the individual spaces between nuclei limit the region of acoustic activation. It is unknown to what extent the pre-

TABLE VI
ACTIVATED ELECTRODES AND OUTCOME

Patient	Active electrodes	Speech tracking (w/min)
1 TL	12	20
2 AA	15	36
3 BC	5	12
4 MP	9	41
5 TK	12	9
6 JP	7	17
7 HP	9 (17)	14
8 WP	10	30

w/min = words per minute.

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existing brainstem compression or deformation plays a role in this relation among the nuclei. Moreover, this is not necessarily a static situation, but after tumour removal and with brainstem re-expansion most likely a different spatial relation of nuclei might develop over weeks or months. On the other hand, in some patients with very pronounced changes and real distortion of brainstem structures, no recovery might occur and some nuclei or tracts may stay mute and cannot be activated. Only a moderate or scarce result might be the consequence. The data from this series indicate the importance of these structural parameters. The type and possibly the duration of brainstem deformation should undergo careful analysis in a larger series and might yield valuable information. It is well known that the major part of the auditory pathway crosses to the contralateral side and passes upwards via the contralateral medial lemniscus to the contralateral inferior colliculus. Hence, the ipsilateral as well as the contralateral brainstem changes need to be analysed.

The duration of deafness is a further factor open to management. Short duration of deafness may be favourable for the functional outcome, perhaps it is even a pre-condition to excellent results, but it is no guarantee to this.

In summary, the application of ABI to date is a safe procedure. But by adequate indication and controlling the site of implantation by multimodality monitoring satisfying results can be obtained, while side-effects can be excluded. On the whole, the individual patient's quality of life is definitely improved as is the likelihood that he or she will maintain a stable social and often professional life for significantly longer phases than bilaterally deaf patients.

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