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Stimulation of the cochlear nucleus with multichannel auditory brainstem implants and long-term results: Freiburg patients

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Abstract

Since 1992 18 patients with bilateral retrocochlear deafness have been provided with a multichannel auditory brainstem implant (ABI). The surgical procedure implies tumour removal and ABI implantation in one stage. Most implantations were via the translabyrinthine approach. The long-term follow-up varied between nine and 80 months. In one case auditory perception could not be achieved and in a second case post-operative stimulation was not possible as the subject died due to lung emboli. In all the other cases auditory perception was achieved and only two subjects became non-users during the follow-up period. The presented long-term results suggest that deaf neurofibromatosis type 2 patients regain acoustic contact with the environment, enlarge their communication skills and improve their quality of life by using a multichannel auditory brainstem prosthesis.

Key words: Brain Stem; Prosthesis Implantation; Neurofibromatosis 2; Cochlear Nucleus; Electric Stimulation

Introduction

The history of electric stimulation of the auditory pathway began with Alessandro Volta in 1800 when he experienced auditory sensations after stimulation of his external auditory canal filled with saline solution.[1] In 1957 Djourno and Eyries[2] published their first experience with an extracochlear electrode stimulating the cochlear nerve and started the era of cochlear implantation. In the middle 50s Penfield[3] stimulated the auditory cortex during neurosurgical approaches under local anaesthesia, and in 1964 Simmons *et al.*[4] reported their experience with electrical stimulation of the auditory nerve and the inferior colliculus. As cochlear implants became more reliable in the late 70s, William House implanted a single-channel electrode array on the brainstem surface after removal of a neurofibroma on 24 May, 1979.[5] This was the first auditory brainstem implant (ABI).

As multichannel stimulation and transcutaneous signal transmission in cochlear implants proved to be superior in providing pitch information with maximal comfort, we developed a multichannel ABI device, based on the Mini22 cochlear implant of Nucleus®, and implanted a female patient with neurofibromatosis in Hannover on 10 September, 1992.[6,7] The results with this first multichannel ABI were so promising that, in Freiburg we continued implanting 17 more patients.[8] Meanwhile our so-called 'European ABI' has been implanted in a few patients in other European countries, whereas the so-called 'American ABI', developed parallel to the European has been implanted in several subjects in the United States. Since 1998 we have been using the new ABI based on the Nucleus® C124M cochlear implant, which provides additional telemetry facilities. This paper describes the device and reports on our long-term experience.

Materials and method

The auditory brainstem implant

The system is based on the commercial cochlear implant consisting of the same speech processor (SPECTRA) and external parts (microphon and antenna), whereas the electrode carrier of the receiver/stimulator is modified and adapted to the anatomy of the brainstem surface. Our first three patients received an array with 20 electrodes of 0.5 mm each, the carrier was round and had to be cut according to the anatomical situation. Fixation of the array on the brainstem surface was achieved by a round dacron mesh. Two further electrodes, one on the implant package and another as a separate ball electrode in the temporal muscle could be used as reference; thus, monopolar stimulation (each of the brainstem electrodes *vs* external reference) as well as bipolar stimulation (every possible combination between two brainstem electrodes) could be

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selected. The actual ABI electrode carrier has 21 electrodes of 0.7 mm on a pre-shaped silicon carrier (8.5 × 3.0 mm) and a dacron mesh between the carrier and the cable for fixation. The current density does not exceed 40 $\mu C/cm^2$ in order to protect brain tissue. The magnet is easy to remove should magnetic resonance imaging (MRI) be necessary for following up neurofibromatosis patients with multiple CNS tumours. The current CI24M hardware implements advanced speech coding strategies such as CIS or ACE in addition to the SPEAK strategy.

The 'American device' has eight electrodes of 1.0 mm placed on a similar silicon carrier (8.5 × 2.5 mm). The main advantage of our 21-electrode array is the fact that more electrodes with non-auditory side-effects can be rejected without dramatic decrease of the number of active electrodes that might reduce place pitch information.

Indications and surgery

The main indication for ABI is bilateral neural deafness due to bilateral tumours of the cerebellopontine angle (neurofibromatosis 2) and damaged auditory nerves. In cases of sensory deafness and functioning auditory nerve, a conventional cochlear implant is preferred. Bilateral injury of the auditory nerve or cochlear deformities and ossification that do not allow intracochlear electrode placement might be future indications as well. Bilateral complete ossification of the cochlears due to meningitis is not an indication for an ABI in our centre, as surgical techniques allow at least partial insertion of a conventional cochlear implant with better results.

The translabyrinthine surgical approach is recommended as it allows complete tumour removal even if the tumour is growing into the labyrinth and provides sufficient access to the lateral recess of the fourth ventricle for inserting the array. Intralabyrinthine tumour growth due to neurofibromatosis is not rare (five out of our 18 patients); in these cases complete removal cannot be achieved via the retrosigmoid approach. After tumour removal, the lateral recess and the foramen of Luschka are exposed by dissecting the choroid plexus of the fourth ventricle. Surgical landmarks are the facial nerve, cranially, and the IXth and Xth cranial nerves, caudally, as the proximal part of the VIIIth nerve cannot always be identified, especially in cases of large or recurrent tumours and previous surgery. Correct placement of the array on the surface corresponding to the cochlear nuclei can be controlled by electrically recording evoked brainstem potentials after stimulation with the implant. However, a suboccipital approach can also be used by skillful surgeons, if intralabyrinthine tumour growth is excluded by MRI.

Subjects

All our patients (Table I), 13 male and five female, had bilateral acoustic tumours of the neurofibromatosis type 2 (NF2) except one with tumours due to Bourneville-Pringle disease (*Case 3*), one with Hippel-Lindau disease (*Case 16*) and one with bilateral VIIIth nerve neuropathy and additional retrobulbary blindness (*Case 17*). The patients were aged between 17 and 58 years. Only five were referred for primary tumour removal, 12 had recurrent tumours after previous surgery. The tumour size varied between five and 70 mm. The first patient underwent gamma-knife therapy and developed facial palsy and deafness due to interstitial haemorrhage, as confirmed intra-operatively. This patient developed somatosensoric side-effects

TABLE I
SUMMARY OF PATIENTS IMPLANTED

	Subject	Aetiology	Age	Tumour	OP	Device	Electrodes	Stim. mode	Strategy	Comments
1	HK	NF2	51	15 P	Sep 92	20+2	3	BP	F0F2	LNU
2	GS	NF2	60	12 P	Nov 93	20+2	7	MP	SPEAK	
3	AL	BPD	31	70 R	Nov 93	20+2	5	MP	F0F2	x
4	SG	NF2	30	15 P	Feb 94	21+1M	6	MP	F0F1F2	
5	RF	NF2	35	20 P	Jun 94	21+1M	10	MP	F0F1F2F5	LNU
6	PF	NF2	35	30 P	Jun 94	21+1M				x
7	CH	NF2	30	5 R	Feb 95	21+1M	14	MP	SPEAK	
8	FU	NF2	42	15 R	Feb 95	21+1M	2	MP	SPEAK	LNU
9	ID	NF2	40	30 R	Feb 96	21+1M	7	MP+BP	SPEAK	Removed
10	MK	NF2	33	5 R	Feb 96	21+1M	7	MP	SPEAK	
11	AT	NF2	25	25 R	Feb 96	21+1M	11	MP	SPEAK	
12	NW	NF2	13	44 P	Apr 97	21+1M	15	BP	SPEAK	OS
13	CHe	NF2	40	None	Apr 97	21+1M	7	MP+BP	SPEAK	
14	LK	NF2	26	40 R	Nov 97	21+1M	10	CG	SPEAK	
15	EB	NF2	22	14 R	Apr 98	24M	8	MP	SPEAK	
16	JS	HLS	43	None	Apr 98	24M	0			NU
17	MM	DP	16	None	Apr 98	24M	12	MP	SPEAK	
18	StG	NF2	19	35 P	Apr 99	24M	7	MP	SPEAK	

The column 'electrodes' indicates the number of electrodes that resulted in pure auditory perception without side-effects. NF2 = neurofibromatosis type 2; BPD = Bourneville-Pringle disease; HLS = Hippel-Lindau syndrome; DP = degenerative polyneuropathy. Tumour size in mm of maximal diameter according to MRI, P for primary surgery and R for recurrent tumour. Monopolar stimulation mode (MP) was the most commonly used, bipolar mode (BP) was used only in two cases, common ground (CG) in one and a combination of monopolar and bipolar mode in two cases. In one case (*Case 16*) no auditory perception could be achieved (non-user, NU) and three cases (*Cases 1, 5 and 8*) became late non-users (LNU). In *Case 12* ABI is used occasionally (occasional stimulation, OS) due to normal hearing on the contralateral side. Subjects with (x) are no longer followed up due to death.

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during stimulation with the ABI, probably due to tissue damage after radiotherapy so that only a few electrodes could be used. One subject died due to pulmonary emboli prior to initial stimulation (*Case 6*), and one developed somatosensory side-effects eight months later so that she has not been using the device anymore (*Case 5*). Furthermore, one subject died after 18 months of continuous use (*Case 3*) and the last subject uses the ABI occasionally, as she is not deaf on the contralateral side (*Case 12*). In one case, explantation was necessary due to flap necrosis and infection after head injury two years after full time use (*Case 9*). Ten subjects are 'full time users', being followed up for four to 80 months.

Results

During the first tune-up session, all subjects except one (recurrent angioblastoma of the posterior fossa due to Hippel-Lindau syndrome) described auditory perception after stimulation of several electrodes. The electrode number varied between two (one patient) and 20 (two patients). The average electrode number without side-effects was 10. In patients with several active electrodes, pitch was projected on the array in an oblique pattern so that the most medial and caudal electrodes elicited higher and the most lateral and cranial electrodes lower pitch perception (Figure 1).

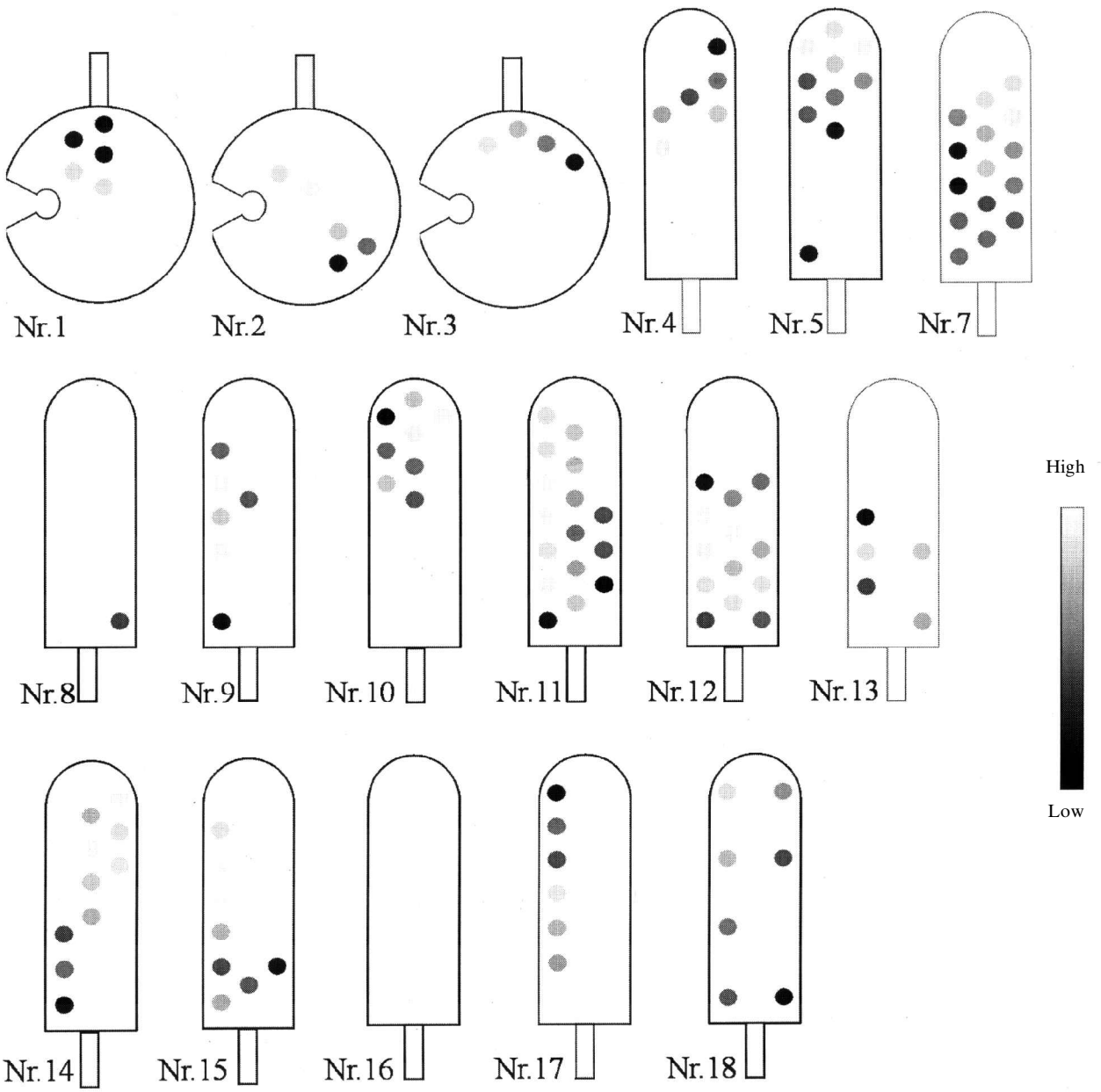

FIG. 1

Projection of different pitch perception (higher to lower pitch) on the electrodes of all subjects according to their subjective descriptions. Note the oblique projection in cases of numerous electrodes available (*Case 7, 11, 14*). In *Case 8* the device is placed too deep into the lateral recess and in *Case 10* too laterally.

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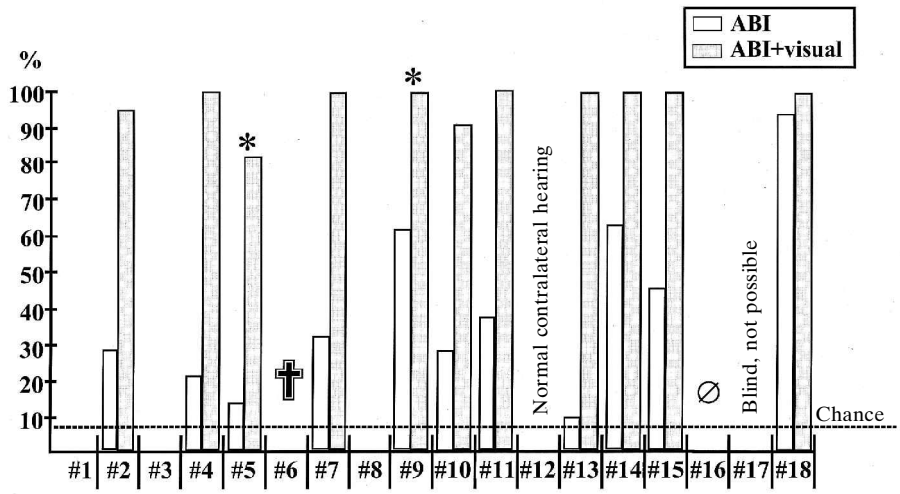

FIG. 2

Closed word discrimination (out of 24 words) of all subjects that could be tested. *Case 3* could speak French, *Case 6* died before stimulation, *Cases 1, 8,* and *16* are non-users, *Case 12* has normal contralateral hearing and subject 17 is bilaterally blind. ∗ = when auditory brainstem implant (ABI) used.

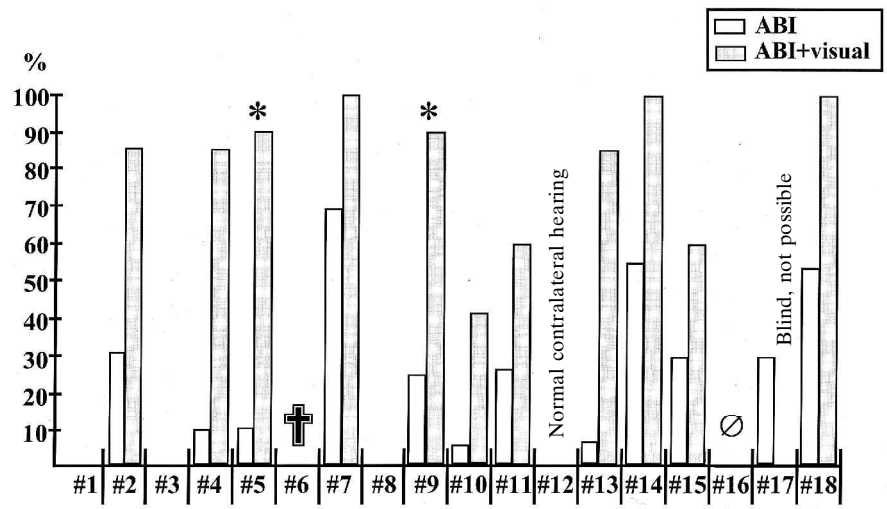

FIG. 3

Results of open-set discrimination of polysyllabic numbers (13–99) with auditory brainstem implant (ABI) alone and combined with lip-reading (except patient 17 who is bilaterally blind). ∗ = when ABI used.

Currently, the average electrode number is 7.7, as two subjects are not using the device any more. The number of electrodes without side-effects is not identical to the number of channels used for speech processing, as inversion of polarity (electrode 2 *vs* 8 and 8 *vs* 2, for example) results in different pitch perception so that these can be used as two channels. Further, combinations of monopolar and bipolar modes (as in *Cases 9* and *13*) can be used to increase the number of channels for a given number of electrodes.

All subjects can identify environmental sounds and discriminate them from speech signals. All are experiencing a dramatic increase of their communication skills by combining both ABI and lip-reading so that they are using their device permanently. Figure 2 summarizes the results of word discrimination in closed-set (out of 24 words) with the ABI alone as well as with ABI and lip-reading.

Only one patient with normal contralateral hearing (*Case 12*) is using the device occasionally in order to prepare herself for expected deafness due to a contralateral tumour.

The ABI allows limited speech understanding without lip-reading. Twelve patients were able to understand polysyllabic words (numbers) without lip-reading in a range between 10 and 60 per cent (Figure 3). All patients have been improving their skills continuously during the follow-up (Figure 4), comparable to cochlear implant patients with earlier speech coding strategies. There has been no evidence of electrode migration or technical failure so far, stimulation parameters showed only slight changes regarding pitch perception but no significant changes of current levels and thresholds. Thus, there has been no evidence of tissue damage due to chronic electrical stimulation of the brainstem surface.

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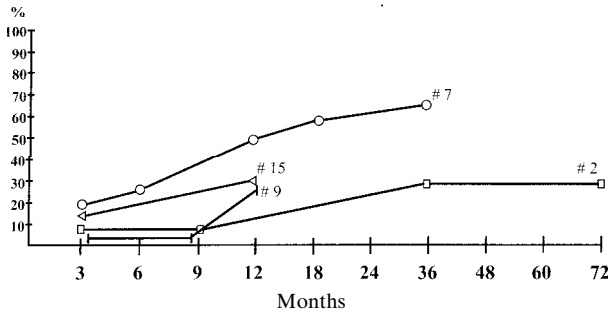

FIG. 4

Long-term number discrimination over time for the four subjects with an initially good result and sufficient follow-up period. Performance increased in all cases.

Discussion

Our long-term results with the auditory brainstem implant are very promising. Multi-electrode arrays allow different pitch perception and global use of the tonotopic organization of the cochlear nucleus complex. With a sufficient number of electrodes and correct placement on the brainstem surface, speech discrimination can be achieved. These results may be improved by further development of electrode arrays and speech processing strategies, simulating the physiological processing at the level of the cochlear nuclei.

Early and late complications related to the ABI could not be observed. Thus, possible complications can be compared to those of the surgery for tumour removal so that indications for an ABI may be extended to cochlear deformities or total ossification as well as temporal bone fractures with damage of the auditory nerve.

However, ABI already presents a reliable and important aid for completely deaf neurofibromatosis patients, and helps them regain part of their communicative skills and acoustic orientation. All our patients acknowledge this fact by using their device permanently; even our single non-user is asking for revision surgery, as she experienced the benefits of her ABI for several months before its failure.

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