Bilateral Auditory Brainstem Implantation in a Patient with Neurofibromatosis Type II

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Introduction

Rapid development of electronic hearing prostheses has allowed for the introduction of auditory brainstem implants (ABI) in patients with neurofibromatosis type II (NF2). In Poland, the Programme of Auditory Brainstem Implants commenced in January 1998 in close cooperation with the Ear, Nose, Throat and Neurosurgery Clinics of the University of Wurzburg and Klinikum Fulda, Germany (Skarzynski et al, 2000a). Since that time, there have been significant improvements in rehabilitation after implantation (Skarzynski et al, 2000b). Although the number of ABI patients world-wide has markedly increased in recent years, many of the factors associated with a successful outcome still remain unknown.

Numerous studies have shown that bilateral cochlear implantation may restore fundamental aspects of binaural hearing and provide binaural advantages experienced by normal-hearing subjects (Zeitler et al, 2008). Bilateral cochlear implantation improves sound localization and speech perception particularly in a noisy environment compared with unilateral implantation. Other benefits of bilateral cochlear implantation include more natural hearing, reduced listening effort and an improved quality of life.

Theoretically, some of these advantages of bilateral electric stimulation should also extend to auditory brainstem implants. As the auditory results in unilateral cochlear implantation are generally superior to the outcomes in ABI patients with NF2, it is unclear if the implantation of a second ABI may have the potential to improve auditory benefit.

This study documents the benefit of bilateral stimulation from sequentially implanted ABIs in a patient with NF2. To our knowledge, bilateral ABI has not been previously reported.

Case report

A 27-year-old man with NF2 presented with bilateral acoustic neuromas. Surgical removal of the right tumour and simultaneous ABI placement was performed on February 9, 2006. The right ABI was activated on April 4, 2006. The growth rate of the contralateral acoustic neuroma was monitored by high-resolution computed tomography. Tumour growth was the main criterion for primary excision of the left acoustic neuroma surgery and ABI placement. He also had clinical evidence of tumour progression. Deterioration of speech discrimination, progression of bilateral tinnitus and disequilibrium were having an increasingly negative impact on his quality of life.

The left vestibular schwannoma was removed and simultaneous placement of the second ABI was performed on March 28, 2008 (Figure 1). The left ABI was activated on June 26, 2008. Both surgeries were led by Professor Robert Behr following the established procedure (Behr et al., 2007). The Med-El C40+ ABI system was utilized. The implantable portion of this system consists of an ABI stimulator, active electrode array, and reference electrode. The structure of the internal stimulator portion is similar to the C40+ ceramic body cochlear implant and is implanted in a bony bed behind the ear. The active electrode array is placed directly on the brainstem and consists of 12 platinum surface-to-surface contacts partially embedded in a silicone paddle to stimulate the cochlear nuclei. A polyester mesh embedded in the silicone allows tissue ingrowth for stabilization of the electrode paddle (Figure 2). The patient was fitted with a Tempo+ behind-the-ear speech processor.

Auditory sensations, adverse effects, most comfortable loudness (MCL), threshold, and tonotopic organization were evaluated postoperatively by stimulation of each of the 12 electrodes. To determine the tonotopic organization of the electrode array, a pitch-ranking procedure was performed (Lorens et al., 2004). Perceptual performance was assessed by free-field audiometry and the Sound Effects Recognition Test (SERT). Speech comprehension was tested in the "sound only" condition using the Pruszewicz monosyllabic Polish word test (20 words per list, 20 lists). Test lists were randomized and the mean score of 3 lists was calculated. Monosyllabic tests were also conducted using a 10 dB signal-to-noise ratio. For subjective assessment of sound quality, a visual analogue scale (VAS) was used; 0 corresponding to poor quality, 10 corresponding to good quality. The SERT, Pruszewicz test, and VAS test were administered 1 month after activation of the second ABI. Three test conditions were utilized: sound on right only, sound on left only and bilateral sound stimulation.

Results

Tables 1 and 2 show results of the psychophysical evaluation of the auditory and nonauditory sensations elicited by electric stimulation of the right and left sides respectively. Electrodes eliciting adverse effects were switched off. A tonotopic pattern was obtained by stimulating various electrodes of the right and left sides (Figure 3). Free-field audiometric thresholds are presented in Figure 4. The SERT requires the patient to identify a sound with the correct picture from set of four. The SERT score for the right was 90% and for the left was 20% (chance performance level). The bilateral SERT score was 90%. Word recognition scores obtained for the right and left sides respectively were 70% and 0% in quiet (60 dB HL presentation level) and 50% and 0% in noise (10 dB SNR). Bilaterally, the word recognition score was 70% in quiet and 50% in noise (equal to the right side score). Subjective sound-quality assessment ratings were 6 points (right side), 1 point (left side), and 9 points (bilaterally).

Discussion

Bilateral ABI allows bilateral auditory input for patients with NF2. Free-field audiometry confirmed the same sensitivity to sounds across a wide frequency range. Lack of sound recognition and open set speech recognition with stimulation of the left ABI alone was likely due to the short time interval since activation. With further rehabilitation, it is anticipated that left ABI outcomes will improve. The subjective benefit of bilateral stimulation on VAS testing is encouraging.

Conclusion

In this patient with NF2, bilateral ABI stimulation provided at least the same or better sound perception benefit as unilateral stimulation. These results support further consideration of bilateral auditory brainstem implantation for patients with NF2.

References

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Figure 1 Computed Tomography Prescan after Placement of the second Auditory Brainstem Implant

Figure 2 Med-El Combi 40+ Auditory Brainstem Implant Electrode Array

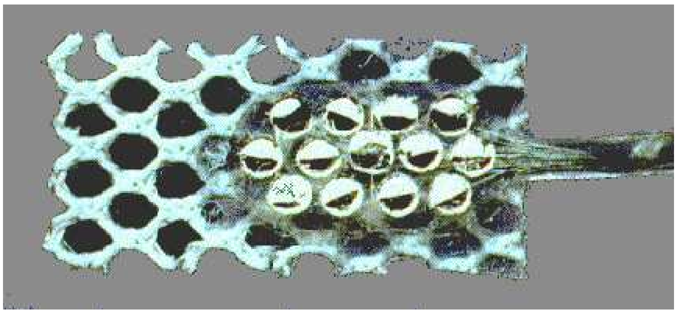

Figure 3 Tonotopic Orientation of the Electrode Array

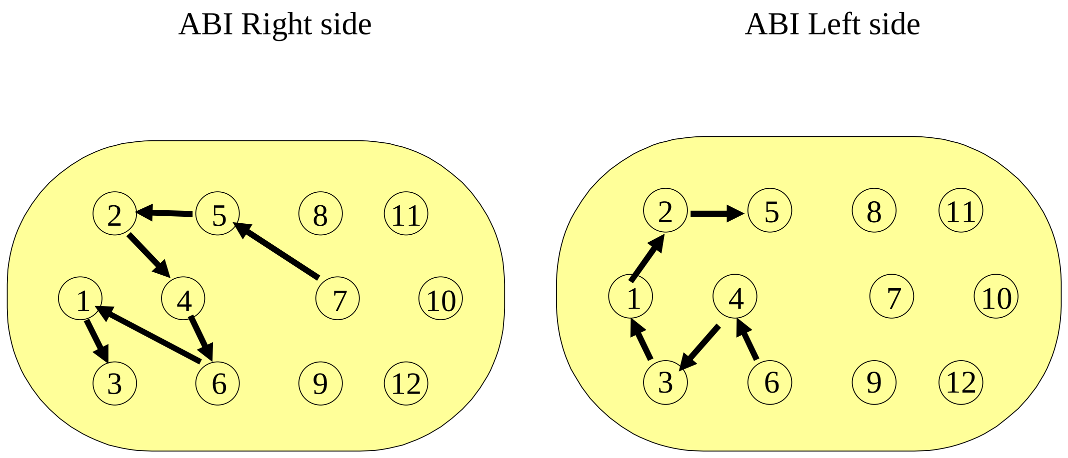

Figure 4 Audiometric thresholds: Bilateral Auditory Brainstem Implants

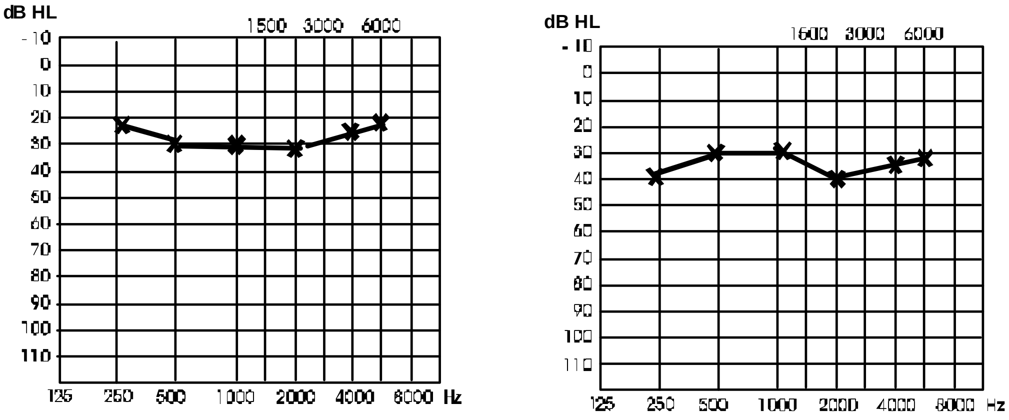

Table 1. Psychophysical Evaluation of Auditory and Nonauditory Sensations; Right Side

No.- number; Dur - duration; THR - threshold; MCL - most comfortable loudness;

electrode status: on - electrode was switched on; electrode status: off - electrode was switched off

Electrode No./Dur (μs)		**Programmed Levels (Current Units)**		**Adverse Effects (May be With or Without Auditory Stimulus)**			**Electrode Status**
				Location of Sensation	**Description**		
No.	**Dur**	**THR**	**MCL**				
1	24	10	320	No adverse effects		Auditory sensation only	On
2	24	5	350	No adverse effects		Auditory sensation only	On
3	24	10	410	No adverse effects		Auditory sensation only	On
4	24	10	410	No adverse effects		Auditory sensation only	On
5	24	15	430	No adverse effects		Auditory sensation only	On
6	24	10	515	No adverse effects		Auditory sensation only	On
7	24	10	570	No adverse effects		Auditory sensation only	On

8	24	10	614	Entire body	Mild	Trembling of entire body	Off
9	24	15	660	Entire body	Mild	Trembling of entire body	Off
10	24	15	690	No adverse effects		Auditory sensation only	On
11	24	No measurable		Right arm	Severe	Twitching of arm	Off
12	24	No measurable		Right arm	Severe	Twitching of arm	Off

Table 2. Psychophysical Evaluation of Auditory and Nonauditory Sensations; Left Side

No.- number; Dur - duration; THR - threshold; MCL - most comfortable loudness;

electrode status: on - electrode was switched on; electrode status: off - electrode was switched off

Electrode No./Dur (μsec)		**Programmed Levels (Current Units)**		**Adverse Effects (May be With or Without Auditory Stimulus)**			**Electrode Status**
				Location of Sensation	**Description**		
No.	**Dur**	**THR**	**MCL**				
1	85	400	800	No adverse effects		Auditory sensation only	On
2	85	500	Not measurable	Left arm and leg	Mild	Twitching	On
3	85	600	1000	No adverse effects		Auditory sensation only	On
4	85	600	900	Head	Mild	Tingling of throat	On
5	85	700	Not measurable	Head	Mild	Tingling of throat	On
6	85	800	1000	Head	Mild	Twitching of left ear	On
7	85	400	Not measurable	Head	Severe	Tingling of throat Twitching of left ear	Off
8	85	700	Not measurable	Head	Severe	Tingling of throat Twitching of left ear	Off
9	85	500	Not measurable	Head	Severe	Tingling of throat	Off

						Twitching of left ear	
10	85	400	Not measurable	Head	Severe	Tingling of throat Twitching of left ear	Off
11	85	No measurable		Head	Severe	Tingling of throat Twitching of left ear	Off
12	85	No measurable		Head	Severe	Tingling of throat Twitching of left ear	Off