© Med Sci Monit, 2009; 15(6): CS100-104
PMID: 19478705

WWW.MEDSCIMONIT.COM
Case Study

Received: 2008.12.30
Accepted: 2009.03.17
Published: 2009.06.01

Bilateral electric stimulation from auditory brainstem implants in a patient with neurofibromatosis type 2

Henryk Skarżyński[1ADFG], **Robert Behr**[2AD], **Artur Lorens**[1ABCDEF], **Robert Podskarbi-Fayette**[1EF], **Krzysztof Kochanek**[1DE]

[1] Institute of Physiology and Pathology of Hearing, Warsaw, Poland
[2] Department of Neurosurgery, Klinikum Fulda gAG, Academic Hospital of the University of Marburg, Fulda, Germany

Authors' Contribution:
A Study Design
B Data Collection
C Statistical Analysis
D Data Interpretation
E Manuscript Preparation
F Literature Search
G Funds Collection

Source of support: Departmental sources

Summary

Background: Recent developments in the field of electronic hearing prostheses have allowed for the introduction of auditory brainstem implants in patients with neurofibromatosis type 2.

Case Report: Bilateral electric stimulation from 2 sequentially placed auditory brainstem implants was applied in a 27-year-old man with neurofibromatosis type 2.

Conclusions: Results of the present case support further application of bilateral electric stimulation from auditory brainstem implants for patients with neurofibromatosis type 2.

key words: **neurofibromatosis type 2 • auditory brainstem implants • bilateral stimulation**

Full-text PDF: http://www.medscimonit.com/fulltxt.php?ICID=869668

Word count: 1307
Tables: 2
Figures: 6
References: 8

Author's address: Henryk Skarżyński, Institute of Physiology and Pathology of Hearing, Warsaw, Poland e-mail: sekretariat@ifps.org.pl

BACKGROUND

An auditory brainstem implant (ABI) is the primary choice of auditory prosthesis to treat deafness in patients with neurofibromatosis type 2 (NF2), a disease that affects 1 in 40 000 individuals. This autosomal-dominant disorder was first described by Wishart in 1822 and is characterized by a defect on the long arm of chromosome 22 (22q), resulting in the development of tumors of the VIIIth cranial nerve. For many years, treatment of this disease was limited to eradication of the tumor by surgical removal or irradiation (gamma knife). After successful removal of the tumor, the cochlear nuclei can be localized, and a multichannel stimulating electrode can be implanted. Optimal placement of the electrode, good neurosurgical outcome, and adequate auditory skills enable restoration of auditory perception (sensation) in patients who have lost the vestibulocochlear nerve as a result of this disease [1].

In January 1998, we initiated the Auditory Brainstem Implant Program at the Institute of Physiology and Pathology of Hearing in Warsaw, Poland, in close cooperation with the Ear, Nose, and Throat and Neurosurgery Clinics of the University of Wurzburg and Klinikum Fulda, Germany [2]. The aim of this report was to present the results of bilateral stimulation from 2 sequentially implanted ABIs in a patient with NF2. To our knowledge, report of bilateral ABI stimulation has not been described in the literature.

CASE REPORT

We present the case of a 27-year-old man with NF2. The selection of this patient was based on the course of NF2, especially in terms of tumor growth, as assessed by means of clinical and radiologic prognoses. The diagnosis was confirmed, and monitoring of the growth rates of both tumors was carried out initially by means of gadolinium-enhanced magnetic resonance imaging (Figure 1). After the first ABI placement, the growth rate of the contralateral tumor was monitored by high-resolution computed tomography (Figure 2). The progression of sensorineural hearing loss as a result of tumor growth was the main criterion for primary excision surgery and implant placement. The patient's speech discrimination had deteriorated, and other symptoms, such as bilateral tinnitus and disequilibrium, were having an increasingly negative impact on his quality of life.

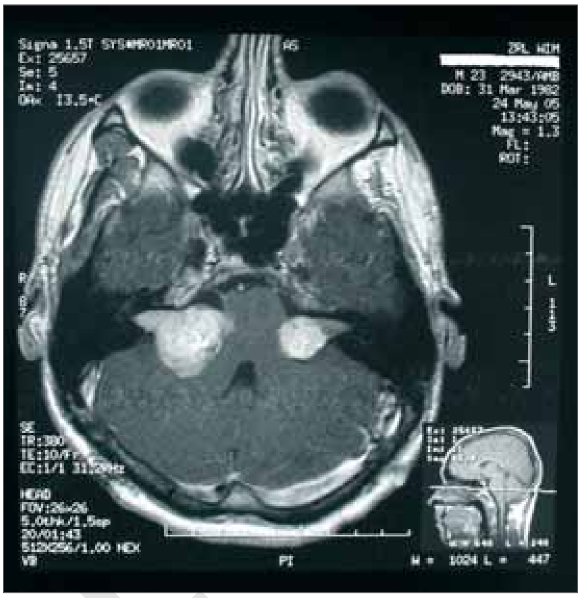

Figure 1. Magnetic resonance image before tumor removal.

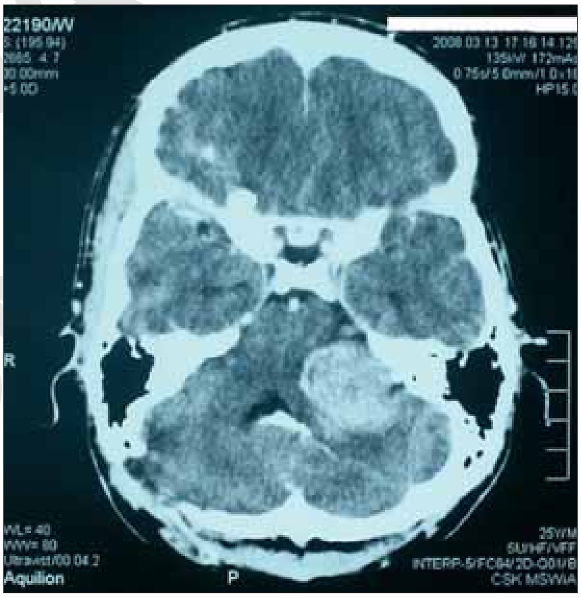

Figure 2. Computed tomography scan of the patient before the second auditory brainstem implant was placed, showing progression of the tumor on the left side.

Surgical removal of the tumor on the right side, with subsequent implantation of the ABI on the same side, was conducted on February 9, 2006. Activation of the right-side ABI system was performed on April 4, 2006. The vestibular schwannoma on the left side was removed and the second ABI was implanted on March 28, 2008 (Figure 3). Activation of the left-side ABI system was performed on June 26, 2008. Both surgeries were led by prof. Robert Behr.

We used the C40+ ABI system manufactured by Med-El. This system consists of an implantable portion, which includes the ABI stimulator, the active electrode array, and the reference electrode. The internal stimulator portion is similar in structure to that of the class C40+ ceramic body cochlear implant and is implanted in the bony bed behind the ear. The active electrode array consists of 12 active platinum surface-to-surface contacts partially embedded in a silicone paddle to stimulate the cochlear nuclei when placed directly on the brainstem. There is also a polyester mesh embedded in the silicone to allow for tissue ingrowth for stabilization of the electrode paddle, as shown in Figure 4. The external portion, coupled to the receiver by a magnet, is a digital speech processor that uses technology similar to that used in cochlear implants. This system converts acoustic signals to an electrical signal, which is processed into a pattern of electrical pulses according to the speech-coding strategy used.

The patient was evaluated postoperatively for auditory sensations, adverse effects, most comfortable loudness (MCL), threshold, and tonotopy organization by means of electric

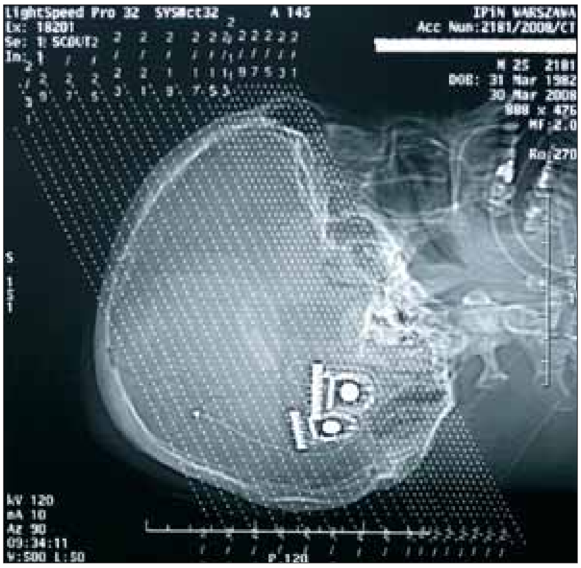

Figure 3. Computed tomography prescan of the patient after the second auditory brainstem implant was placed.

stimulation of each of the 12 electrodes in the array. The loudness scaling was performed several times for each active electrode. From the loudness scaling results, the MCL levels and threshold levels were derived in current units and corresponded to 25 points on a subjective loudness scale. To determine the tonotopy organization of the electrode array, a pitch-ranking procedure was performed according to the method described by Lorens and associates [3]. Perceptual performance was assessed by free-field audiometric testing and the Sound Effects Recognition Test (SERT).

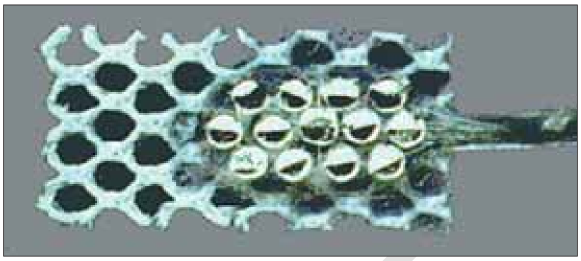

Figure 4. Med-El Combi 40+ auditory brainstem implant electrode array.

Speech comprehension was tested under the "sound only" condition using the Pruszewicz monosyllabic Polish word test (20 words per list, 20 lists) [4]. Lists of each test were randomized between test conditions. Results are presented as the mean of 3 test lists. Monosyllable tests were also conducted in noisy surroundings at a signal-to-noise ratio of 10 dB. For subjective assessment of sound quality, a visual analog scale (VAS) was used, where 0 corresponded to bad quality, and 10 corresponded to good quality. The SERT, Pruszewicz test, and VAS test were administered 1 month after activation of the second ABI under 3 conditions: sound on right side only, sound on left side 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 side and left side, respectively. Electrodes eliciting adverse effects were switched off. A tonotopic pattern was obtained by stimulating various electrodes

Table 1. Psychophysical evaluation of auditory and nonauditory sensations; right side.

Electrode No./Dur (µs)		Programmed Levels (Current Units/ Duration; [ms])		Adverse Effects (may be with or without auditory stimulus)			Electrode status
No.	**Dur**	**THR**	**MCL**	**Location of sensation**		**Description**	
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

No. – number; Dur – duration; THR,; MCL – most comfortable loudness.

Table 2. Psychophysical evaluation of auditory and nonauditory sensations; left side.

Electrode No./Dur (μs)		Programmed Levels (Current Units/ Duration; [ms])		Adverse Effects (may be with or without auditory stimulus)			Electrode Status
No.	**Dur**	**THR**	**MCL**	**Location of Sensation**		**Description**	
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 Twitching of left ear	Off
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

No. – number; Dur – duration; THR,; MCL – most comfortable loudness.

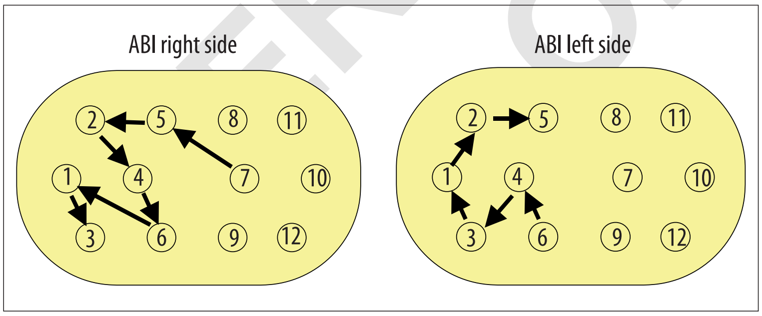

Figure 5. Tonotopy orientation of the electrode array.

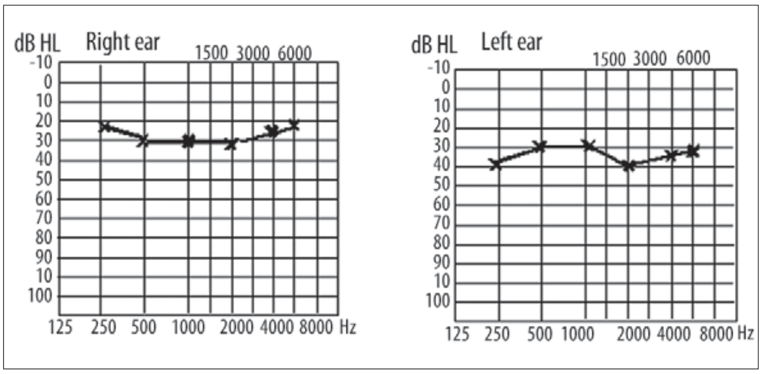

Figure 6. Results of audiometric evaluation.

of the right side and left side, as shown in Figure 5. The free-field audiogram is shown in Figure 6. The SERT score for the right side was 90%, and that for the left side was 20%. The left side score was at the chance performance level; the SERT requires the patient to choose from among 5 foils. The bilateral SERT score was 90%. Word recognition scores obtained for the right and left sides separately were 70% and 0%, respectively at a 60-dB hearing level (dBHL) presentation level in quiet surroundings and 50% and 0%, respectively at 10 dB SNR in noisy surroundings. Bilaterally, the score was 70% in quiet surroundings, with 50% in noisy surroundings being equal to the score of the right side. Subjective sound-quality assessment rankings were 6 points, 1 point, and 9 points for the right side, left side, and bilaterally, respectively.

DISCUSSION

Much data from several leading neuro-otosurgical centers worldwide has shown that unilateral stimulation of the cochlear nucleus with a multichannel ABI provides benefit to patients with hearing loss of retrocochlear origin, especially those with NF2. The level of satisfaction, measured by patients and professionals, varies considerably, from providing environmental sound awareness and assisting in lip-reading to open-set speech understanding comparable to that provided by a cochlear implant [5,6]. Our positive experiences with unilateral ABI in patients with NF2 [7] triggered discussion as to whether patients with bilateral sacrifice of the vestibulocochlear nerves would benefit more from bilateral than from single-sided stimulation of the auditory pathway with an auditory brainstem prosthesis. Much data has shown that bilateral stimulation of the cochlear nerves by means of cochlear implants provides more benefit to patients than single-sided stimulation [8], so we made the assumption that the same effect could be achieved with bilateral stimulation from ABIs. Results from the present study, obtained shortly after activation of the second ABI, support this assumption. The free-field audiograms for the left and right sides were comparable, demonstrating that both sides showed the same sensitivity to sounds across a wide frequency range. Lack of sound recognition and lack of open speech recognition in response to electric stimulation of the left ABI alone were most probably due to the early stage of rehabilitation after left ABI activation. The subjective benefit from bilateral stimulation, as shown by VAS testing, was encouraging.

CONCLUSIONS

Results achieved with bilateral ABI stimulation in the present patient with NF2 demonstrated that bilateral electrical stimulation from ABIs provide at least the same or better sound perception benefit as that provided by unilateral stimulation. Results from the present case support the further application of bilateral electric stimulation from ABIs for patients with NF2.

REFERENCES:

1. Shannon RV, Fayad J, Moore JK et al: Auditory brainstem implant. II. Post surgical issues and performance. Otolaryngol Head Neck Surg, 1993; 108: 635–43
2. Skarżyński H, Szuchnik J, Lorens A, Zawadzki R: First auditory brainstem implantation in Poland: auditory perception results over 12 months. J Laryngol Otol, 2000; 114 (27): 44–45
3. Lorens A, Piotrowska A, Skarżyński H: Pitch perception and the number of electrodes vs. long term development of speech perception ability in auditory brainstem implant. International Congress Series: Elsevier, 2004; 1273: 429–32
4. Pruszewicz A, Demenko G, Richter L, Wika T: New articulation lists for speech audiometry. Part II. Otolaryngol Pol, 1994; 48(1): 56–62
5. Otto SR, Brackmann DE, Hitselberger W: Auditory Brainstem Implantation in 12- to 18-Year-Olds. Arch Otolaryngol Head Neck Surg, 2004; 130: 656–59
6. Vincenti V, Pasanisi E, Guida M et al: Hearing Rehabilitation in Neurofibromatosis Type 2 Patients: Cochlear versus Auditory Brainstem Implantation. Audiol Neurotol, 2008; 13: 273–80
7. Skarżyński H, Szuchnik J, Miszka K, Lorens A et al: Rehabilitation Results of the Auditory Brainstem Implant User Based on Tests in Polish and German Language. 4th European Congress of Oto-Rhino-Laryngology Head and Neck: Klaus Jahnke, Marcus Fisher. Monduzzi Editore, Berlin, 2000; 155–58
8. Zeitler DM, Kessler MA, Terushkin V et al: Speech perception benefits of sequential bilateral cochlear implantation in children and adults: a retrospective analysis. Otol Neurotol, 2008; 29(3): 314–25