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First Croatian auditory brainstem implantation

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Abstract

A deaf female patient was diagnosed with bilateral acoustic neurinomas. Diagnosis incorporated the standard audiological battery for sensorineural hearing loss, computed tomography and magnetic resonance imaging. The left side had been operated on four years previously in another clinic using the suboccipital approach. The auditory brainstem implant surgery was performed on the 'second side' using the same approach. It was an uneventful operation with good anatomy and no serious post-operative complications. Post-operatively, the patient performed exceptionally well, with up to 50 per cent of words recognized in the opened set and 85 per cent in the closed set, both without lip-reading.

Key words: Brain Stem; Prosthesis Implantation; Neuroma, Acoustic; Neurofibromatosis 2; Hearing Loss, Sensorineural; Croatia

Introduction

Neurofibromatosis type 2 (NF2) is a rare disease, formerly called 'central neurofibromatosis', that most often involves the VIIIth cranial nerve, sometimes bilaterally. In such cases hearing damage or even deafness is no exception. In recent years advances in technology led us to cochlear implants that tend to replace the non-functioning cochlea. Now we are going a step further; auditory brainstem implant (ABI) where cochlear nuclei are stimulated directly. Although it enables us to bypass the VIIIth nerve and the possible pathology, it confronts us with many new issues. The safety of the device is one of the issues. Acute and chronic studies on influence of electrical stimulation of cochlear nuclei were performed showing no measurable changes.[1,2] Biocompatibility of the implanted materials has been inherited from cochlear implant technology. Several approaches to the fourth ventricle are in present use, translabyrinthine and suboccipital being the most common. The midline approach is also interesting.[3] Intra-operative orientation is often reduced due to the tumour size or position, so identification of reliable landmarks such as the root of the VIIIth nerve, choroid plexus of the fourth ventricle and the IXth nerve, is necessary.[4] Acoustic nerve tumour surgery and brainstem implantation are demanding surgical procedures. A multidisciplinary approach is also required, starting from diagnostics, anaesthesiology issues, surgery, intra-operative monitoring, post-operative care, fitting and rehabilitation procedures.[5]

Auditory brainstem implantation is considered to be a useful procedure for patients with bilateral neural deafness. They can now regain their acoustic contact with the environment and add a new quality to their communication abilities.[6,7]

Patient and methods

Our patient is a 28-year-old female who first came to our Department in 1994. The first clinical sign of the disease was a sudden, bilateral hearing loss in 1992. The right ear improved a little, and she was able to use a hearing aid. The left ear remained deaf. From the very beginning she has been suffering tinnitus on the left side as if she had water in her ear canal. Before this incident, her hearing was subjectively very good on the right and slightly less so on the left. Since the moment hearing deteriorated, she suffered from slight vertigo and nausea. From time to time she had a moderate bitemporal headache. In the early stage of the disease she suffered pain in both ears. There is no traceable sign of neurofibromatosis in her family.

A tonal audiogram showed a sensory hearing loss on the right side with a hearing threshold between 50 and 60 dB in the speech frequencies, falling to 25 dB at 12 kHz. There was complete deafness on the left, without any response.

Tympanometry was normal, and the cochleostapedial reflex showed a response on the right side upon ipsilateral stimulation. Even with contralateral stimulation there was some response to stimulus intensities of 120 to 125 dB. On the left side there was no response to the ipsilateral stimulation.

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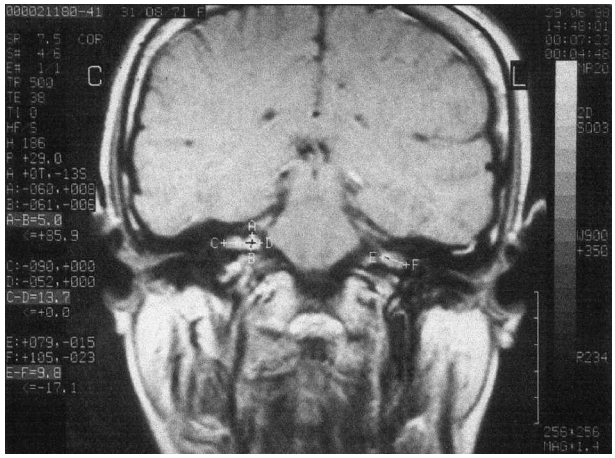

FIG. 1

Pre-operative magnetic resonance imaging showing the tumour on the right side, and an apparent scar on the left.

Contralateral stimulation elicited response on all characteristic frequencies. Threshold levels on the right were reduced.

Prolonged stapedial reflex testing showed signs of fatigue.

Brainstem evoked response audiometry (BERA) showed no response on the left side. On the right all characteristic waves were present. After comparing the expected latency of the wave V according to the hearing threshold at 2 and 4 kHz with the actually measured latency, the hearing damage was suggested to be cochlear.[8]

Electronystagmography showed a left-sided, partially compensated severe peripheral vestibular damage. Retrocochlear hearing damage was suspected, and MRI showed a bilateral tumour of the internal hearing canal, small on the right, only 2–3 mm, and bigger, but still intracanalicular, on the left side, approximately 14 mm in diameter.

The patient was first operated on in another clinic in January 1995. The larger, left-sided tumour was removed using a suboccipital approach. Facial paresis persisted for several months afterwards.

The hearing level on the right started to vary, and the patient reported of different types of tinnitus. In the later course of the disease the hearing level on the right changed for the worse and finally in August 1998 she became deaf. The promontory test showed no response on both sides. Repeated MRI showed a slow but consistent growth of the tumour on the right. No changes of the shadow on the left side were noticed (Figure 1).

FIG. 2

Tumour of the VIIIth nerve, intra-operative finding.

The right side was operated on 25 January, 1999, at our Department. A retrosigmoidal, suboccipital right-sided craniotomy was performed. After visualization of all relevant anatomical structures, the cranial nerves were checked electrophysiologically. The tumour was about 15 mm at its largest diameter, located mostly intracanalicularly. To expose the lateral part of the tumour the posterior wall of the inner hearing canal was drilled away. The tumour was removed, and the plate brainstem implant electrode inserted into the lateral recesses of the fourth brain chamber. Intra-operative stimulation was performed and placement of the electrode adjusted accordingly. The implant body was placed into a drilled-out bed and fixed with non-resorbable sutures (Figure 2).

Post-operatively, the patient complained of severe vertigo for the first few days, and headache in the occipital region mostly at daytime. During the night there was no headache. After a while, vertigo gave way to slight, occasional dizziness. The headaches were controllable with ordinary analgesics. A post-operative CT scan showed the electrode to be in a good position and no sign of ventricular enlargement.

An auditory brainstem implant Cochlear CI21+1M was implanted. All speech understanding results regard listening to a known speaker.

Results

The first fitting was performed in our operating theatre, six weeks after the operation for removal of the tumour. Fifteen active channels were formed and sorted according to a subjective pitch. On the first session the patient was able to recognize a spectrum of intensities, from threshold to discomfort. There were a few unwanted, non-auditory effects of the stimulation. Some channels produced a feeling of pressure in the throat, and involuntary twitching of the chin or facial muscles. She also experienced an unpleasant sound in the opposite ear. All such channels were disconnected.

In the next sessions the threshold levels did not change much, nor did the pitch recognition. Channels are divided into three groups according to a reliable pitch recognition. Today she has 11 active

TABLE I

PITCH AND INTENSITY RESOLUTION THRESHOLD

	Frequency (Hz)		
	500	1000	2000
Δf (%)	1	2	>5
Δl (dB)	1.25	3	4

Δf = pitch discrimination threshold; Δl = intensity discrimination threshold.

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TABLE II
SPEECH UNDERSTANDING WITH AUDITORY BRAINSTEM IMPLANT AND LIP-READING

	Months after surgery			
	1	2	4	6
Consonants (%)	46	53	57	67
Numbers (%)	60	71	95	100
Words (%)	68	96	96	96
Sentences (%)	68	96	96	100

TABLE III
SPEECH UNDERSTANDING, WITH AUDITORY BRAINSTEM IMPLANT WITHOUT LIP-READING

	Months after surgery			
	1	2	4	6
Words CS (%)	60	80	85	85
Words OS (%)	0	10	35	50
Sentences CS (%)	68	70	80	96
Sentences OS (%)	0	0	40	52

CS = closed set; OS = opened set.

channels, pulse width is 100 us, frequency range is between 200 and 3573 Hz. SPEAK coding strategy is used.

The tinnitus she suffered before the surgery has now gone.

The tone duration resolution threshold tested with a 1000 Hz tone lasting 500 ms, was more than ± 35 per cent. Recognition of consonants in Table II was tested in the median position. In the initial position of the consonant in the word, the results are better (60 per cent after two months, 76 per cent after four and 78 per cent after six months). The most repeated error was mistaking plosives 'p' for 'b' or 't' for 'd' and switch of africates and fricates.

Discussion

The patient had bilateral neurofibromas of the VIIIth nerve, although we could not actually prove a generalized or central (type 2) neurofibromatosis. The tumours were suspected after a thorough audiological procedure, and finally diagnosed using MRI. CT failed to reveal the tumour, but it should be mentioned that the inner hearing canal was not targeted at all (simple CT of the brain). Even if it was targeted, it is questionable if the tumour on the right side would have been diagnosed. ABR and CT are no more a screening/diagnostic standard for acoustic nerve tumours. Nowadays it is MRI.[9]

We performed the surgery on the other side, since one side had been operated on already in another clinic in 1995. The decision to use an ABI was made after the promontory test results. A cochlear implantation was considered, but that would leave the patient and us with problems if it failed. Secondary ABI surgery is of a questionable efficacy and an open medico-legal issue.

Surgery was performed using a suboccipital approach. We considered that it would provide better anatomy and broader view, both, for the tumour removal and electrode placement. The tumour was middle-sized, smaller than 2 cm. Anatomy was well preserved, as expected in such cases. The position of the electrode was verified by intra-operative monitoring as suggested by most authors.[5,10]

The first fitting was done in the operating theatre with the patient monitored and the anaesthetist by her side. This first switch on was very promising, and from the very beginning the patient was doing well. Cessation of tinnitus can be found in other ABI patients, and stands along the fact that listening is the best therapy for tinnitus.[11]

The results are very encouraging, and after review of the literature, our patient belongs to the upper class of the ABI users.[5,12,13]

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