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LEADING ARTICLE

HEARING RESTORATION WITH THE MULTICHANNEL AUDITORY BRAINSTEM IMPLANT

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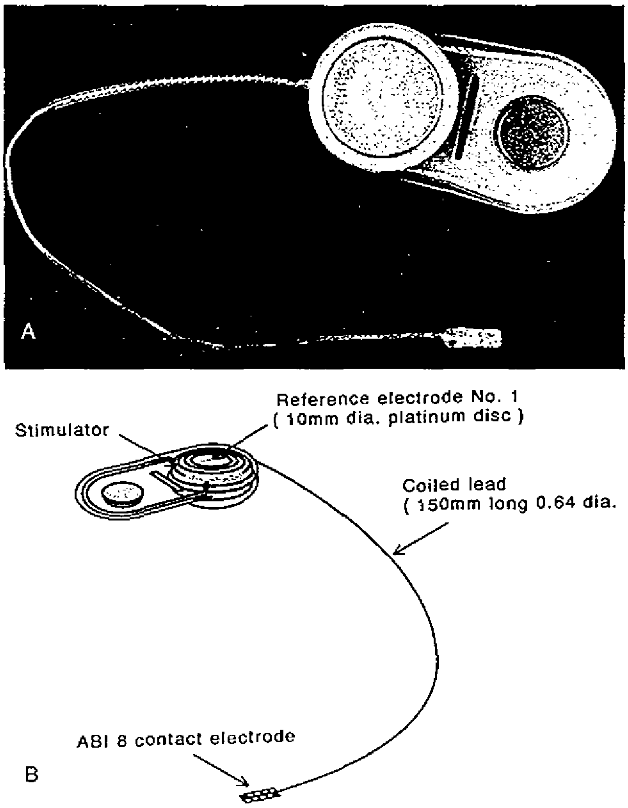
Restoration of useful hearing is now possible in patients with bilateral acoustic neuromas by direct electrical stimulation of the cochlear nucleus. Our first experience with the Multichannel Auditory Brainstem Implant is reported. A forty four year old female with bilateral acoustic neuromas and a strong family history of Neurofibromatosis Type II presented with profound bilateral hearing impairment. Translabyrinthine removal of the right tumour was performed with placement of the Nucleus eight electrode Auditory Brainstem Implant. Intraoperative electrically evoked auditory brainstem response monitoring successfully confirmed placement over the cochlear nucleus. Postoperatively, auditory responses were obtained on stimulation of all electrodes with minimal non-auditory sensations. The patient now receives useful auditory sensations using the "SPEAK" speech processing strategy. Auditory brainstem Implantation should be considered for patients with Neurofibromatosis Type II in whom hearing preservation tumour removal is not possible.

Patients with Neurofibromatosis Type II may suffer total hearing impairment due to the growth or surgical removal of their bilateral acoustic neuromas. Restoration of useful hearing is now possible in these patients by direct electrical stimulation of the cochlear nucleus using the Auditory Brainstem Implant (ABI). The first cochlear nucleus implant was performed in 1979 by William House and William Hitselberger (Edgerton et al 1982). They implanted a single channel brainstem prosthesis with a ball electrode and a percutaneous transmission system based on the 3M-cochlear implant. Following the success of this procedure, a number of patients have been implanted with a variety of prototype devices. Stimulation of the electrodes has produced auditory sensation in most patients with results similar to a single channel cochlear implant. Recently, following collaboration between Cochlear Corporation and the House Ear Institute, a fully implantable, multi-electrode prosthesis has been developed based on the Nucleus 22 channel cochlear implant (Figures 1a, 1b). The electrode array is placed over the surface of the cochlear nucleus within the lateral



FIGS. 1a, 1b. Illustration and diagram of Multichannel Auditory Brainstem Implant.

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recess of the fourth ventricle at the time of translabyrinthine acoustic neuroma removal. Stimulation is via a transcutaneous coil system with a variety of speech processing strategies available, depending on the results of electrode mapping. In most cases, multiple channels have been available for stimulation and the Speak speech processing strategy is used (Skinner et al 1996, Otto and Staller 1995).

The improved auditory perception results in patients using this multichannel ABI has led to the establishment of a multicentre international trial controlled by the U.S. Food and Drug Administration.

Case Report

Our first patient to receive an auditory brainstem implant, a 44 year old woman, presented with a progressive, bilateral, profound, sensorineural hearing impairment. Her mother had undergone multiple operations for acoustic neuromas resulting in bilateral facial paralysis and total hearing loss. The patient's sister had previously had a large acoustic neuroma removed and has a documented small contralateral tumour. Despite this strong family history of NF2, the patient herself had not previously been investigated. Magnetic resonance imaging demonstrated the presence of bilateral acoustic neuromas and a small meningioma in the left Meckel's cave region. Interestingly, the right ear in which she had had almost total hearing loss for fifteen years was the side with the smaller (2cm) acoustic neuroma, the left sided tumour measuring approximately 3cm (Figure 2). Pure tone audiometry confirmed a right profound sensorineural hearing loss with zero speech discrimination. In the left ear, the pure tone average was 100db with 25% speech discrimination at 120db. She still wore a hearing aid in the left ear but found it of limited benefit.

In view of the slight remaining hearing in the left ear, it was recommended that the smaller right sided tumour be removed first and the possibility of placement of an auditory brainstem implant was offered.

In September 1995, a translabyrinthine approach was used and total removal of the acoustic neuroma achieved. The facial nerve was preserved intact and could be stimulated at 0.05 m.a. after tumour removal. In preparation for the ABI the usual scalp incision was modified to create a large anteriorly based scalp flap so that the suture line was well behind the position of the receiver stimulator and antenna. A seat for the electronic package was drilled in the temporo parietal skull above the sinodural angle and, after tumour removal, the receiver stimulator package was secured with nylon ties. Despite the poorly pneumatised temporal bone, the translabyrinthine approach afforded adequate access to the foramen of Luschka and lateral recess. The foramen of Luschka was identified by retraction of the cerebellar flocculus, identifying where the stump of the eighth nerve entered the brainstem superior to the glossopharyngeal nerve. Positive identification of the lateral recess was confirmed by the presence of choroid plexus and egress of cerebrospinal fluid (CSF). The electrode array, with its Dacron mesh backing, was gently inserted into the lateral recess with the electrodes facing superiorly.

A sterile transmitting coil was then placed over the antenna of the receiver stimulator package, the ABI was stimulated and an electrically evoked auditory brainstem response (ABR) measured. Continuous EMG monitoring of cranial nerves 7, 9 and 10 was also performed. With the initial position of the electrode array, there was no repeatable evoked ABR with only the stimulus artefact recorded (Figure 3). The implant was then repositioned and progressively inserted into the lateral recess until a reproducible evoked ABR was obtained without activation of the cranial nerve monitors (Figure 4). The

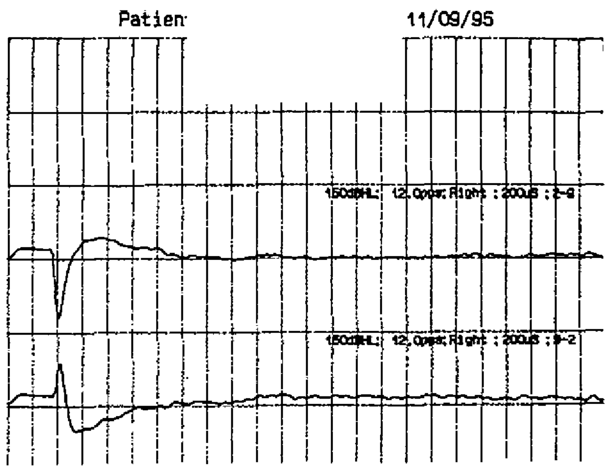

FIG. 3. Intraoperative electrically evoked ABR – initial placement.

FIG. 2. MRI Scan demonstrating bilateral acoustic neuromas.

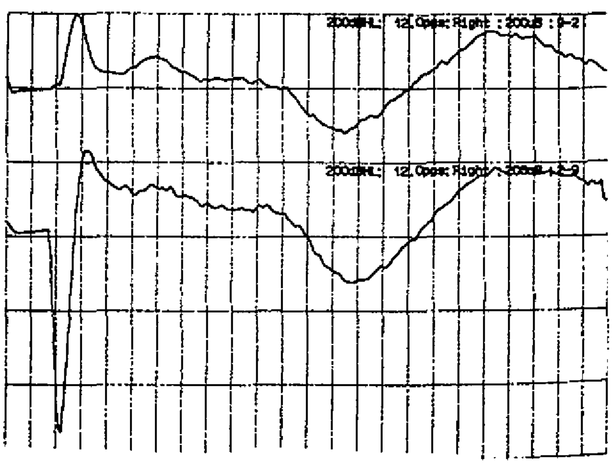

FIG. 4. Intraoperative electrically evoked ABR – final position.

FIG. 5. Postoperative CT scan demonstrating translabyrinthine defect and position of electrode array.

wave form of the evoked ABR was noted to be stable when the polarity of the electrical stimulus was reversed.

The dura of the posterior fossa was partially repaired with interrupted sutures and the translabyrinthine defect was obliterated by packing with autologous abdominal fat in the usual manner. CSF drainage at 10ml per hour via a lumbar spinal catheter was performed for 48 hours. It was felt that this may reduce the potential for CSF tracking along the electrode cable to the subcutaneous wound.

The patient tolerated the procedure well and a post operative CT scan (Figure 5) confirmed satisfactory position of the electrode array in the region of the lateral recess. Unfortunately she developed a facial nerve paralysis which progressed to a House Brackmann grade 6. Fortunately, no surgical intervention was necessary for corneal protection and by four months post operatively she had complete eye closure. Immediately following hospital discharge she developed a left deep venous thrombosis and required readmission for anti-coagulation.

Six weeks post operatively, the initial post operative stimulation of the implant was performed. In view of the potential for stimulation of non-auditory brainstem structures, the stimulation was performed in the Intensive Care Unit and pulse, blood pressure, ECG and oxygen saturation were monitored. No alteration in any parameters occurred during stimulation, however. Psychophysical testing was used to assess the stimulation thresholds and comfort levels on each electrode. Auditory sensation was noted on 5 of the 8 electrodes using a monopolar mode. In bipolar stimulation mode, there was some alteration in perceived pitch between the most medial and most lateral electrode pairs. The patient described some non-auditory perceptions, in particular facial discomfort or tingling sensation occurring with stimulation of the more medial electrodes, numbers 5, 7 and 9.

Results

The patient was fitted with a speech processor using the "SPEAK" speech processing strategy with a bipolar electrode stimulation mode. After initial fitting of the speech processor, the patient was immediately able to hear voice and environmental sound. She now wears the external device almost constantly and has given up wearing the left hearing aid. She finds the ABI extremely useful both for communication and for perception of environmental sound. Comprehensive auditory perception testing is performed regularly with tests of consonant, vowel, sentences (City University of New York, CUNY sentences) and environmental sound recognition (sound effects recognition test SERT). NU-CHIPS is the North Western University Children's Perception of speech test. The Monosyllable, Spondee, Trochee, Polysyllable (MSTP) test is a closed set test of word and stressed syllable recognition (Table 1). For sound alone testing of word discrimination she scored well above chance alone. Similarly, for vision alone her score on CUNY sentences is between 12% and 33% whereas for sound and vision her score increases to between 46% and 56%. This demonstrates a very significant aid to her speech understanding when using the ABI. This benefit is confirmed by her improved face to face communication when using the multichannel ABI.

Table 1
Auditory Perception Test results

Test	Score	(chance)
SERT	63%	(25%)
<u>Speech Perception Test Results</u>		
NU-CHIPS	56%	(25%)
MSTP	67% word	(8%)
	79% stress	(33%)
CUNY sentences	vision alone 12%	
	sound & vision 46%	

Discussion

Auditory brainstem implantation should be considered in all patients with Neurofibromatosis Type II when surgical removal of an acoustic neuroma is performed. Suitable candidates are those patients with non aidable hearing and any size tumour or patients with serviceable hearing where hearing preservation is unlikely due to tumour size. Implantation at the time of first side tumour removal is reasonable, such as in our case where there was bilateral profound hearing loss. It should also be considered in the patient with serviceable hearing in the contralateral ear, but where presence of a large contralateral tumour means that further hearing loss is likely. The selection criteria for auditory brainstem implant patients are listed in Table 2 (Brackmann et al 1993).

Table 2
Selection Criteria for Auditory Brainstem Implant

Bilateral eighth nerve tumours
Competency in English language
Age 15 years or older
Psychological suitability
Compliance with research protocol
Realistic expectations

The translabyrinthine approach is a proven method for tumour removal enabling access to the lateral recess of the fourth ventricle and dorsal cochlear nucleus complex. The cochlear nucleus is situated superior to the lateral recess of the fourth ventricle with the ventral portion of the cochlear nucleus covered by the middle cerebellar peduncle. The lateral recess is directed anterolaterally from the fourth ventricle and so the translabyrinthine approach is necessary to obtain a sufficiently anterior access to allow implant insertion. This approach also facilitates facial nerve preservation, particularly for the larger tumours which are typical of NF2 patients.

The nucleus multichannel ABI has an electrode array consisting of eight platinum disc electrodes mounted in two rows on a silastic carrier. This has a backing of Dacron mesh to prevent displacement and facilitate permanent fixation by ingrowth of fibrous tissue. The cochlear nucleus complex is surrounded by neural structures such as the vestibular nuclei, the spinothalamic and trigeminothalamic tracts, together with cranial nerves 5, 7, 9 and 10. The implant is a surface electrode array and it has been determined that the most effective position is overlying the dorsal cochlear nucleus and inferior portion of the ventral cochlear nucleus within the lateral recess (Brackmann et al 1993, Shannon et al 1993). More ventral placement is likely to produce non auditory stimulation of cranial nerves 7 or 9 or stimulation of the overlying flocculus of the cerebellum. Excessively deep insertion into the fourth ventricle is likely to cause stimulation of the spinothalamic tracts.

During surgery, the implant is initially positioned by visual identification of the lateral recess and junction of the cochlear nerve and cochlear nucleus complex. Accurate positioning of the implant is facilitated by monitoring of the facial and glossopharyngeal nerves and measurement of electrically evoked auditory brainstem potentials (Waring 1995). Intraoperative measurement of the electrically evoked auditory brainstem response (EEABR) serves the dual purpose of confirming the integrity of the implant and confirming that the electrical stimulus does activate the auditory system. After placement of the electrode array, the device is stimulated using the transduction coil. Initially, the stimulus amplitude is gradually increased and the resulting stimulus artefact monitored. Subsequently, the first part of the sweep, when the bi-phasic stimulus occurs, is electronically blanked out to eliminate the stimulus artefact and allow recognition of the EEABR wave form. When a potential EEABR wave form is identified, the polarity of the electrical stimulus is reversed, in which case the neural response will remain unchanged.

However, a stimulus current artefact will be inverted. In our case, electric stimulation of the implant in the initial position produced only stimulus artefact (Figure 3). Note the inversion of the response with reversal of electrode polarity. Stimulation of the electrodes in the final position produced a repeatable early response that did not invert with changed polarity (Figure 4). There was, however, a persistent negative peak at longer latency (7 millisecond). This was thought to be due to a myogenic response which persisted despite apparently optimal electrode position. Possibly this was due to using a 200 microsecond pulse delivered through the ABI rather than a 50 microsecond pulse.

Initial stimulation and speech processor fitting is delayed until six weeks post operatively in order to allow resolution of any brainstem oedema and recovery of any reversible neural injury. By this time, the electrode array should be stable and fixed into position. As noted above, there is considerable potential for non-auditory stimulation. Possible non-auditory effects include facial movement (cranial nerve 7), constriction in the throat (cranial nerve 9), vertigo (vestibular nuclei and nerve 8), tingling in the shoulder and arm (long tracts of the brainstem) and vibration sensation in the eye (flocculus of the cerebellum). Out patient experienced facial discomfort on stimulation of electrodes 5, 7 and 9, possibly due to stimulation of the trigeminal tract. For this reason, these electrodes were not included in the initial map. Fortunately, with time, the non-auditory sensations have diminished. Currently, stimulation produces a loud auditory sensation without side effects on six electrodes. Electrodes 8 and 9 are not used in the map because of a sensation of twitching of the corner of the eye.

A variety of electrode stimulation modes are available for the multichannel ABI. A 10mm platinum disc is present on the receiver stimulator which can be used as a reference electrode for monopolar stimulation. Alternatively, a bipolar mode can be used between individual electrodes on the electrode array. The bipolar mode requires a lower current level and has been the main mode used in our patient. Interestingly, when recently using a Monopolar stimulation mode, the patient reports the quality of sound perceived to be clearer and more normal. There is no significant improvement in results of auditory perception testing, however. A combined or "variable" mode utilising both bipolar and monopolar stimulation is also possible. This system allows flexibility in accommodating electrode specific pitch sensations and the possibility of reducing or eliminating any non-auditory sensations. Similarly, a variety of speech encoder strategies are available. The new SPEAK speech processing strategy appears to be the most effective even where as few as 2 electrodes are available for stimulation. Using the SPEAK encoder our patient receives very useful auditory sensations and her results on perceptual performance scores compare favourably with the mean scores from multichannel auditory brainstem implant patients at the House Ear Institute (Otto and Staller 1995).

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USES OF HEARING

None of the organs of sense, besides the eye, combines within itself the two faculties of perception and expression. That infinite variety of sounds which exercises so lively an influence on our passions, and by means of which so much useful knowledge is imparted to the mind, depends for its existence upon this organ. It is even capable of supplying more sublime conceptions to the imagination than that of sight. What concentration of terrific spectacles can produce on the mind of a deaf man that feeling of intolerable excitement and anxiety which a blind man experiences when he hears, for the first time, the sounds of a thunderstorm gathering above his head? Is not the idea of the trumpet, which shall wake the dead, "when the mystery of God shall be finished," one of the most awful that is associated with our anticipations of that dreadfully glorious time?

In this sense, as in that of sight, we are struck by the diminutive size of the organ, when contrasted with the magnitude of the ideas which it can receive and impart. The atmosphere is agitated by a thunder-storm – the heavens are convulsed around us from one horizon to the other – all nature is terrified by the tremendous sounds – yet the whole is received on a membrane, the dimensions of which do not exceed those of a split pea.

By appealing through the medium of this sense to our mental passions, the poet is enabled to wield them at his pleasure. By this he acquires the power of raising or of soothing our thoughts, by the grandeur or the melody of his verse. By this he can fill our minds with a pleasing terror, when he sings of the awful changes of nature – or lull them into a delicious peace and admiration, when he celebrates her gentler beauties. By this he can startle us in our chambers with the roar of angry billows – the clattering of sudden thunders – the explosion of mines – the pealing of artillery – the crash of warring elements; or refresh our spirits, amid the agitations of a worldly life, with the sounds of pastoral innocence and simplicity – the murmuring of summer streamlets – the whispering of summer winds – the singing of birds, and other peace-breathing sounds.

GERALD GRIFFIN
Dublin, 1860