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The magnetless Clarion® cochlear implant in a patient with neurofibromatosis 2

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Abstract
We present our experience using the Clarion® magnetless multichannel cochlear implant with a woman profoundly deafened following bilateral acoustic neuromata as a consequence of neurofibromatosis 2 (NF2). The right neuroma had been previously removed without an attempt at neural preservation. On the left, however, a posterior fossa approach had been taken with the aim of preserving hearing. Although the left cochlear nerve appeared to be undamaged at the end of the operation, no hearing thresholds could be elicited on post-operative audiometry, because of damage either to the cochlear nerve or to the blood supply to the cochlea. Round window electrical stimulation subsequently produced a perception of sound, confirming that the cochlear nerve was capable of functioning and that a cochlear implant would be effective. Because she would need regular magnetic resonance imaging (MRI) to monitor existing and future NF2 lesions, it was decided to use a magnetless Clarion® implant, which has been shown to be MRI compatible. We report our experience of using the device in this case and discuss some of the issues related to the provision of cochlear implants to patients with NF2.

Key words: Cochlear implant; Neuroma, acoustic; Neurofibromatosis

Introduction

Neurofibromatosis type 2 (NF2) is a dominant inherited condition associated with the development of bilateral vestibular schwannomas and other peripheral and central neural tumours. Surgery for bilateral vestibular schwannomas carries a high risk of complete loss of functional hearing. In cases where both the left and right cochlear nerves are divided at the time of surgery, auditory brainstem implants (ABI) have been placed in the lateral recess of the fourth ventricle to stimulate the cochlear nucleus, with some success (Luetje *et al.*, 1992; Shannon *et al.*, 1993). However, the benefits of ABI have been variable and not, so far, comparable to those provided by conventional multichannel cochlear implants for patients with profound deafness and intact cochlear nerves.

Using a middle or posterior fossa approach, it is possible to attempt preservation of the cochlear nerve in surgery for NF2 vestibular schwannomas. Although preservation of hearing has been reported, with useful levels of post-operative speech discrimination, this is only possible in a relatively small percentage of cases, and usually for tumours less than 2 cm in diameter. Recently there have been reports (Pensak *et al.*, 1991; Hoffman *et al.*, 1992; Doyle and Shelton, 1993; Arriaga and Marks, 1995; Hulka *et al.*, 1995; Tono *et al.*, 1996) suggesting that in some cases where hearing preservation has failed, the intact cochlear nerve may be capable of being stimulated by a multichannel cochlear implant. Auditory perception with the implant (including for speech) has been reported with these patients as being substantially better than the best results so far achieved by ABI.

Many patients with NF2 will continue to need regular MRI examination of the brain to check for or to monitor the growth of residual vestibular schwannoma and the other tumours (such as meningiomas) that may be present in the brain and spinal cord. Conventional transcutaneous multichannel cochlear implants contain magnets to maintain the connection between transmitter and receiver. The magnetic fields generated by MRI may cause demagnetization of these magnets, and there is a risk that the implant may physically move, heat up or cause undesirable output during the scanning process. Additionally the field itself is disrupted by the presence of the magnet giving rise to extensive artefacts or shadows in the images obtained.

Implant manufacturers have recognized this issue, and several solutions have been proposed to overcome the problems arising from the use of magnets in transcutaneous systems. One solution is to incorporate a removable magnet, however the frequent surgical interference with the skin flap required for regular scanning in this case of NF2 would have been undesirable. Advanced Bionics, in conjunction with Medizinisch Hochschule (Hannover) and Bruckhoff and Partners have developed a magnetless transcutaneous multichannel implant. A research version of this device was made available for our patient.

The magnetless Clarion® 1.2 cochlear implant

The internal components of the Clarion® magnetless implant are identical to the existing Clarion® 1.2 system except there is no magnet. The difference externally is the use of the Hannover headpiece, which allows effective

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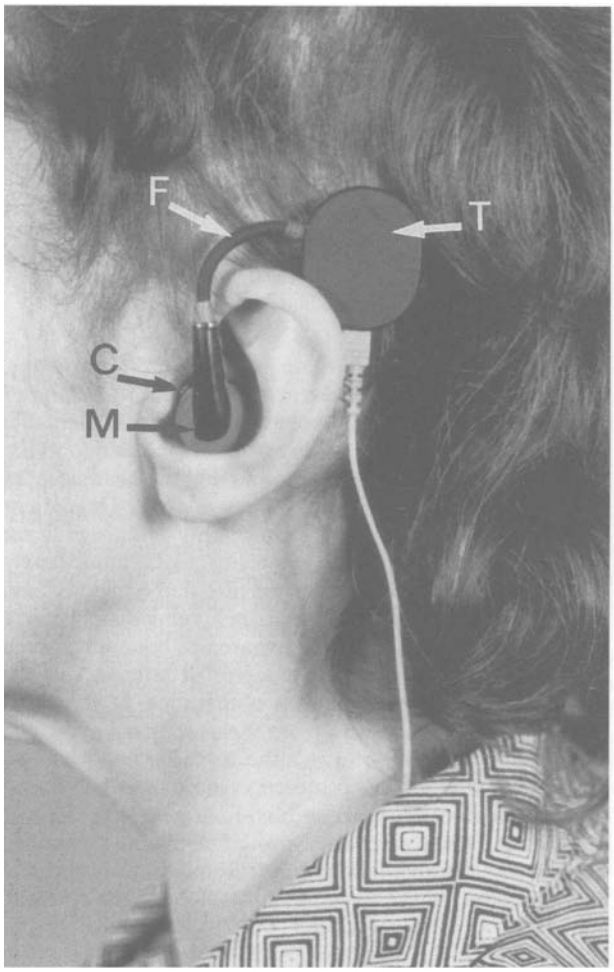

FIG. 1
Hannover headpiece. M: microphone inlet; C: customized earmould; F: flexible padded wire; T: transmitter.

alignment of the transmitter and receiver. The headpiece comprises a stiff padded wire, linking the transmitter to a customized earmould, and an integrated microphone. The earmould is designed to support the padded wire with the correct alignment and pressure against the scalp needed for successful transmission. When correctly fitted, this ensures that the transmission link remains stable during all normal orientations and movements of the head. The microphone is located at the earmould end of the headpiece (Figure 1). Early clinical results with this device are to be presented by Weber *et al.* (in press).

Magnetic resonance imaging with magnetless cochlear implant

The magnetless device produces substantially less distortion in the MRI image than its magnet-containing counterpart (Figures 2 and 3). Severe image degradation in the region of the ipsi-lateral hemisphere has been demonstrated in conventional devices (Weber *et al.*, 1998). Studies have shown that the Clarion® magnetless device may be considered MRI compatible, causing relatively minor image distortion. In a 1.5 T field, the distortion was found to be in the region of 0–30 mm radius around the device. Using a field of 0.3 T the distortion had been measured to a maximum radius of 45 mm. The unexpected finding that the weaker magnetic field produced more distortion has been attributed to the less

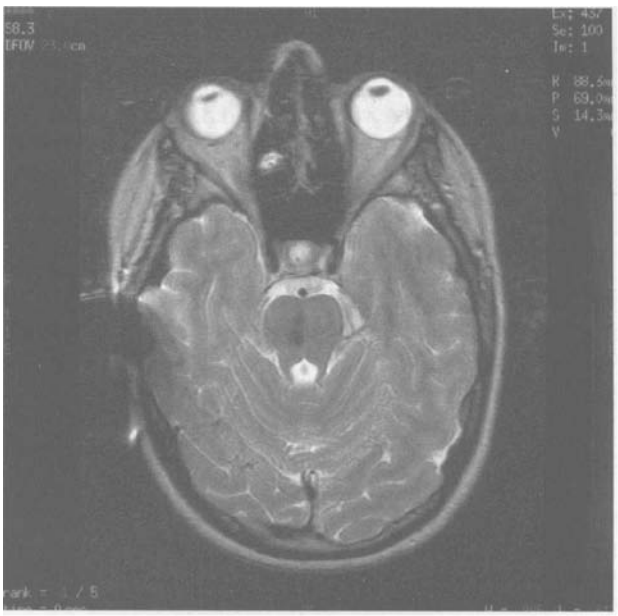

FIG. 2
MRI of Clarion® magnetless implant. MRI scan of volunteer wearing the magnetless implant on side of head. This illustrates the approximate degree of distortion in a 1.5 Tesla field. *(By kind permission of J. Goldring).*

sophisticated imaging technique employed by the older 0.3 T scanner. Figures 2 and 3 compare the extent of distortion produced by the 1.5 T field between a magnetless and a conventional Clarion® implant (note: the implant is strapped to a volunteer's head rather than in situ). In both cases a minor torque of less than 0.01N was produced, and no evidence of significant heating or unintentional output was found.

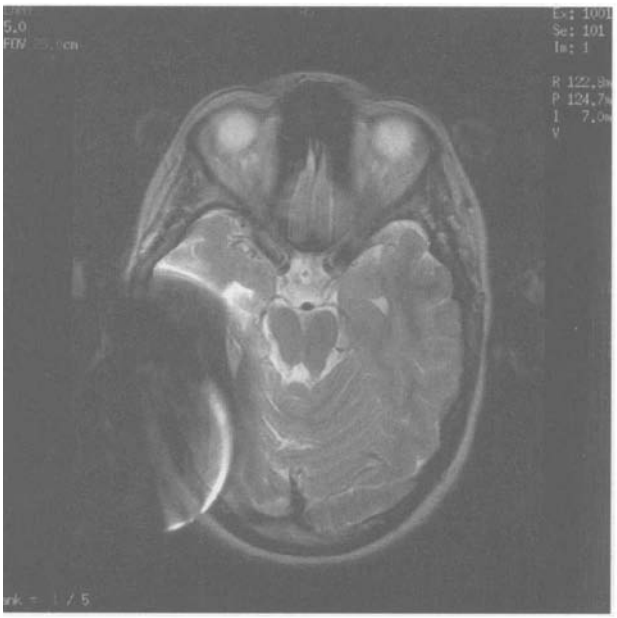

FIG. 3
MRI of conventional Clarion® implant (containing magnet). MRI scan of volunteer wearing conventional implant (containing magnet) on side of head. This illustrates the approximate degree of distortion in a 1.5 Tesla field. *(By kind permission of J. Goldring).*

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Case report

A woman presented in 1987 aged 34, with a 10-year history of progressive right hearing impairment, which had become total after five years. She had developed left-sided tinnitus two years previously, with an initial slight hearing impairment on this side. More recently there had been several episodes of sudden severe left hearing loss, which had all recovered satisfactorily with steroid treatment. She also reported intermittent infrequent vertigo. Later neurogenetic review of the family established the diagosis of neurofibromatosis 2, due to a new mutation.

Pure tone audiometry at this time showed a total right hearing loss, and a mean left hearing threshold of 23 dBHL over the speech range (0.5, 1, 1 and 4 kHz). Air computed tomography (CT) meatography and MRI showed bilateral vestibular nerve tumours. The right extended extracanalicularly, while the left was confined to the internal auditory canal. In addition, three asymptomatic meningiomas were present with an intra-medullary cord lesion at C2/3, a neurofibroma at the base of her right index finger and a cafe-au-lait skin lesion on the left leg. Subsequently, she developed further neurofibromas of the right greater auricular nerve, left trigeminal nerve and left jugular fossa, with a further neuroma in the lumbar spinal cord.

Translabyrinthine excision of her right superior vestibular nerve tumour was carried out on 27 September 1988. She had normal facial function post-operatively but the cochlear nerve was not preserved.

By September 1990, she had experienced further episodes of left sensorineural hearing loss with incomplete recovery, demonstrating a mean PTA across the speech range of 45 dBHL. Surgery to excise her left vestibular nerve tumour, which was 8 mm in size and still intracannalicular, was therefore recommended with a view to attempting hearing preservation. Posterior fossa surgery to remove the left neuroma was carried out on 21 October 1990. The posterior internal auditory canal wall was removed and the tumour excised completely, preserving the cochlear facial nerves and adjacent blood vessels. Unfortunately she had no hearing post-operatively, presumably due to cochlear ischaemia. She was therefore referred to the UCL Cochlear Implant Programme for consideration of cochlear implantation.

Following referral to the UCL Cochlear Implant Programme in 1990, transtympanic promontory stimulation of the left ear was performed two weeks after the excision of her second acoustic neuroma. She did not report any auditory percept during this procedure. At that stage she decided to defer undergoing the round window stimulation which would have provided a more definitive functional test of the nerve's capacity to respond to electrical stimulation. In February 1991, a craniotomy with excision of multiple meningiomas was carried out. She made a complete recovery without further neurological deficit. By August 1991 she had developed a neurofibroma in the cervical region, and a recurrence of her intramedullary tumour was apparent. Serial scanning for the next five years showed no significant change in the size of these various lesions and there was clearly a continuing need for regular scanning. Six years after the initial promontory stimulation she was again referred to the UCL/RNTNE Cochlear Implant Programme and round window stimulation was performed in May 1997. Under local anaesthetic and using a ball electrode placed in the round window niche through a small flap raised in the tympanic membrane, encouraging results were obtained (Table I).

Her left-sided tinnitus was suppressed during electrical stimulation, and for a 50 Hz stimulus, she was able to reliably detect a gap of 50 ms.

TABLE I
ROUND WINDOW STIMULATION RESULTS

Freq. Hz	Threshold microamp	Uncomfortable loudness level microamp	Percept
50	28.2	>99.9	'Phone ring'
100	30.6	>99.9	'Crackle'
200	40.1	>99.9	'Crackle/buzz'
400	190.0	350.0	'Crackle/buzz'
800	200.0	400.0	'Scratch/buzz'

Cochlear implant surgery

Our patient received a research version of the Clarion® magnetless implant on 5 August 1997. The implantable device needed to be placed very precisely in relation to the external auditory canal and pinna so that the headpiece, as described above, could be held firmly in place. The patient had had a posterior fossa craniectomy with significant absence of bone over the posterior fossa immediately behind the mastoid bone. She had also had a middle fossa craniotomy performed with a burr hole immediately above the ear. This left a rather narrow area of bone into which to sink the implant. A further factor of interest was the presence of unexpected partial obliteration of the basal turn of the cochlea 0.5 cm from the cochleostomy, which was placed just anterior to the round window. Two possible reasons for this obstruction are either that during posterior fossa surgery some blood entered the posterior semicircular canal or that a protein-containing exudate, similar to that which is found in CSF in cases of posterior fossa tumours, had entered the perilymph via the cochlear aqueduct.

Post-operative progress

The implant was activated in September 1997. Using all available eight channels, the CIS strategy was employed. Some early problems were encountered with the stability of the padded wire designed to hold the external transmitter against the scalp. A remake of the customized earmould was required. Eventually a stable connection was achieved, which was maintained even during vigorous head movement. After three months of implant use, an irritating ticklish sensation occurred at the back of her mouth. This was associated with activation of the highest frequency channel. There was no visible palatal or pharyngeal movement associated with this sensation, which was therefore likely to be of sensory nerve origin, possibly associated with existing neoplasms in the region. With manipulation of pulse-width, the sensation was eliminated without functional loss of this channel. Six months post-operatively, the implant is being worn for all waking hours with considerable benefit (see below).

Results

In the six months following initial activation of the device the results shown in Table II were obtained.

Ten months after initial stimulation, the patient reported that her auditory perception is continuing to improve. She currently attends for regular re-tuning and rehabilitation sessions, and has a home practice programme. She reports a high degree of satisfaction with the device. Further improvements are anticipated.

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TABLE II
IMPLANT-AIDED SPEECH PERCEPTION SCORES

Test administered via POCIA* System	Pre-implant	6 weeks	3 months	6 months
CUNY/UCL Sentences (with lip-reading)	30%	88%	87%	97%
BKB Sentences in quiet (no lip-reading)	0%	23%	17%	34%
Environmental sound recognition	not tested	note tested	63%	not tested

*Prediction of outcome of cochlear implantation in adults
POCIA tests were carried out using the system devised by the MRC Institute of Hearing Research, Nottingham and we are grateful for their permission to quote results

Post-operative MRI scan

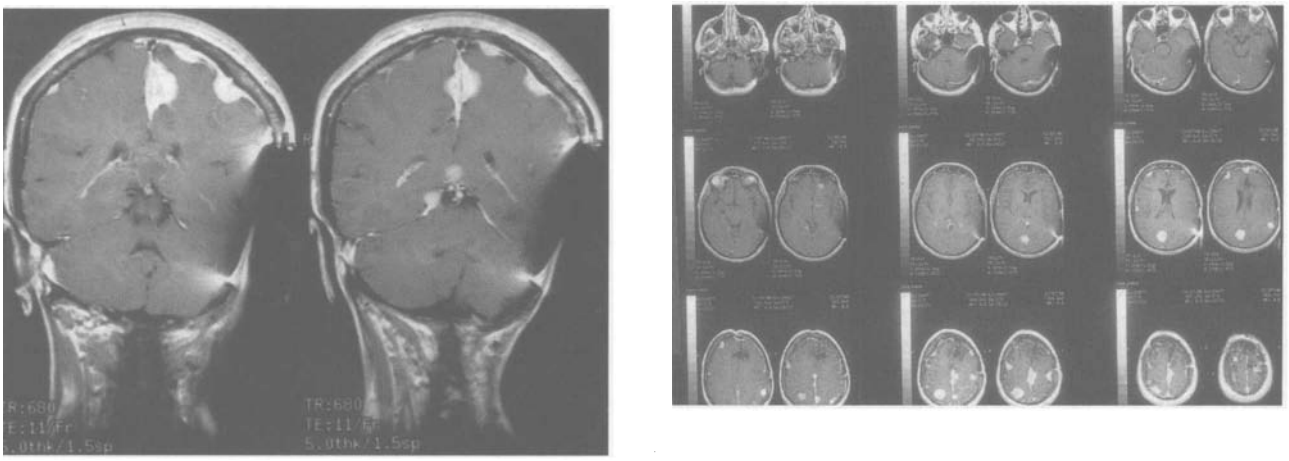
As part of the on-going neurological monitoring, a post-operative MRI scan was performed in August 1998 (see Figure 4 below). This shows a small amount of artefact but allows good visualization of the residual tumours.

Discussion

For patients with NF2, preservation of hearing on at least one side after removal of the vestibular schwannoma is an important goal in trying to avoid the appalling handicap of total acquired deafness. With present techniques, hearing preservation is unlikely for tumours greater than 2 cm in diameter but Black *et al.* (1995) point out that advances in genetic identification of NF2 patients and the availability and increasing sophistication of MRI scanning have increased the rate of diagnosis of earlier, and so smaller tumours. As these smaller tumours are more reliably identified, so the amount and quality of residual hearing is likely to be greater at the time of diagnosis. The dilemma is that while earlier excision can reduce post-operative morbidity, there is a risk that some patients with a degree of useful hearing pre-operatively may be made profoundly deaf by bilateral tumour removal. Cohen *et al.* (1993) recommend early surgery in smaller tumours because they are associated with a better chance of hearing preservation; in larger tumours they recommend a delay in surgery until the hearing is lost or other complications appear. Black *et al.* (1995) summarized techniques for predicting hearing preservation in tumours smaller than 2.0 cm, and the surgical techniques designed to preserve hearing using a middle fossa or suboccipital approach; compared with the alternative of total deafness, they point out that any residual hearing after surgery is of benefit to these patients. Pensak *et al.* (1991) recommend attempting to preserve the cochlear nerve in tumours less than 2 cm, partial tumour removal in larger tumours and watchful waiting in those patients with tumours less than 1 cm. Glasscock *et al.* (1993) reported a 44 per cent rate of hearing preservation in 25 NF2 patients, falling to a 24 per cent rate of useful hearing during a follow-up period of one to 12 years.

The reported quality of hearing preservation in terms of pure tone threshold and speech discrimination (using standard single-syllable speech testing material) is between normal and zero. Sanna *et al.* (1995) reviewed 57 papers in the English language literature dealing with hearing preservation after acoustic neuroma surgery, and concluded that there is wide range of results and an even wider range of criteria for evaluating what constitutes useful preservation. Ten separate classification systems (including Sanna's own) were described, with a plea for an accepted international classification of the results of hearing conservation surgery.

When hearing preservation fails, leaving the patient with total bilateral deafness, the only two techniques currently available for restoring some perception of sound are by an auditory brainstem implant (ABI) and by a conventional cochlear implant. In cases where the cochlear nerve has been divided on both sides, ABI is the only option. The insertion of the ABI electrode into the lateral recess of the fourth ventricle carries a significant risk, that is likely to be less when the electrode is placed at the time of tumour removal rather than at a later separate operation. Current results using ABI appear comparable to those found with a single channel cochlear implant, with limited discrimination of sounds and useful assistance to lip-reading (Shannon *et al.*, 1993; Laszig *et al.*, 1995). There will continue to be an important place for ABI, particularly in patients with large tumours where preservation of the nerve is not possible.



(a) (b)

FIG. 4a and b
Post-operative MRI scan of patient. MRI scan (T_1 weighted sequence with gadolinium enhancement and 1.5 Tesla field) showing minor degradation of the image around implant site. Multiple intracranial tumours are visible.

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In contrast, the potential benefit from a multichannel intracochlear implant is greater. Hoffman *et al.* (1992) report the case of a man whose second side 1.5 cm acoustic neuroma was totally removed by a retrosigmoid approach, leaving the cochlear nerve intact but with no preservation of hearing. Round window stimulation was initially negative (perhaps due to neuropraxia) but positive after eight weeks, and the patient received a Nucleus® 22 multichannel cochlear implant. One year after implantation, he scored 72 per cent correct on the CID sentence test and 'was able to use the telephone for limited conversations'. Hulka *et al.* (1995) reported a similar case where a 2 mm intracanalicular tumour was completely removed by a retrosigmoid approach, leaving the patient profoundly deaf, although the cochlear nerve was anatomically intact. Promontory stimulation was positive seven weeks later, and a Nucleus® CI22 device implanted. Three months later, pure tone thresholds of 30–40 dBA (250 Hz to 4 kHz) were recorded on sound-field testing and the patient was reported to be 'using the implant successfully'. A third NF2 case was reported by Tono *et al.* (1996). An 8 mm intracanalicular tumour was removed by the middle fossa approach, leaving the cochlear nerve intact, but no hearing. Promontory stimulation was positive at 50 Hz and 100 Hz one month later; MRI confirmed that the cochlear duct was patent and a Nucleus® 22 device was inserted. Facial nerve stimulation was produced by the five electrodes nearest to the intralabyrinthine portion of the facial nerve and these electrodes were switched off. A year after implantation the patient scored 43 per cent in an open-set test of sentence recognition without lip-reading (rising to 80 per cent with lip-reading) and was able to use the telephone for 'limited conversation'. A fourth case was reported by Arriaga and Marks (1995) of a patient with a solitary acoustic neuroma in an only hearing ear. The tumour was removed using a translabyrinthine approach and a Nucleus® CI22 device inserted through a cochleostomy anterior to the round window. The Eustachian tube was sealed and the contents of the middle ear stripped and the external ear canal everted and closed laterally. Sound field thresholds between 25 and 35 dBA were recorded using the implant, with 50 per cent enhancement of lip-reading reported. Results in these four cases are comparable to those reported for average multichannel cochlear implant users.

Our own case had a negative result on promontory stimulation using a transtympanic needle two weeks after tumour removal; round window stimulation was subsequently positive, but had been deferred for six years at the patient's own request and because further intracranial surgery was necessary for her other tumours. It is not clear whether the initial negative result was because of high impedance of the transtympanic needle or because there was a period of neuropraxia (as may have occurred in Hoffman *et al.*'s case, 1992).

Arriaga and Marks (1995) did not delay the insertion of the implant in their case because of the risk of fibrous tissue infiltration of the cochlea after translabyrinthine surgery. Our own case is the only one of the five so far reported where some obstruction of the basal turn of the cochlea was encountered. The delay between tumour surgery and implantation however was much longer (at six years) than the other cases, suggesting that normally implantation should take place with the minimum practical delay to avoid the risk of cochlear duct obstruction.

In all previously reported cases the implant contained a magnet. Tono *et al.* (1996) stated that the contralateral residual tumour that was present in their case would be monitored in future by CT rather than by MRI. A magnetless transcutaneous implant is clearly an advantage in such cases, particularly for our patient who still has a number of residual intracerebral tumours requiring regular scanning with good resolution relatively close to the implant site.

The purpose of the magnet is to hold the external transmitter coil in close alignment with the buried receiver and this remains the best strategy for most cases. However our experience between 1982 and 1990 with the UCH/RNID single channel implant (which does not contain a magnet) suggested that simple mechanical fixation of the transmitter coil was feasible, and we have found the Hannover headpiece to provide a stable and effective coupling.

A further problem that could arise in cases of NF2 is that surgery, perhaps for a larger tumour, could leave an intact nerve and profound deafness but with the added complication of residual tumour attached to the nerve. Residual tumours of this kind seem to be slow growing, and it would be reasonable to consider offering such a patient an implant, perhaps after a period of observation of the growth, or absence of growth of the residual tumour.

Conclusion

We report a further case of hearing restoration in a patient with NF2, profound bilateral deafness and an intact cochlear nerve. The benefit provided by the implant is comparable to that found in more standard multichannel cochlear implant recipients, and greater than would have been possible with any currently available auditory brainstem implant. Some obstruction of the cochlear duct was encountered at surgery and this might have been avoided by performing the implant within a few weeks, rather than six years after tumour surgery. A magnetless implant can still allow stable and effective coupling of the external and internal transmitter and receiver, and the absence of a magnet permits use of MRI to monitor existing intracerebral and spinal tumours, and any new tumours that might develop.

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