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Clinical Records

Cochlear implantation in otosclerosis: a unique positioning and programming problem

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Abstract
A case is reported in which a Nucleus 22 channel cochlear implant was inserted into the basal turn of the cochlea of a patient with advanced otosclerosis. It then passed out of the anterior end of the basal turn into an otospongiotic cavity related to the cochlea. Seven electrodes were located in the basal turn and it was possible to map them sufficiently well for the patient to derive considerable benefit from the implant. The problem of implant induced facial nerve stimulation in otospongiosis is also discussed.

Key words: Cochlear implantation, otosclerosis

Introduction

Cochlear implantation (CI) using one of the several multichannel devices now available is recognized as a valuable and cost-effective means of rehabilitating many individuals born deaf, or deafened in adult life after the acquisition of spoken language (Summerfield and Marshall, 1995). In the latter category, the aetiological factors are many, and include otosclerosis. This condition when untreated may lead to total deafness, and of course the operation of stapedectomy may unfortunately on occasion hasten the progression of the hearing loss. Otosclerosis may cause quite widespread changes in the temporal bone, and these may present unique problems for the surgeon and for the rehabilitation team. Electrical stimulation of the facial nerve by the device is a well recognized hazard. This paper draws attention to a hitherto unrecorded problem of electrode positioning related to the unique pathological changes in the temporal bone which occur in otosclerosis.

Case report

A 56-year-old man was referred to the cochlear implant clinic at Manchester Royal Infirmary with a history of progressive hearing loss in both ears for many years. He had sustained a head injury with a probable skull fracture when he was 12 years old, but the details of this episode were not available. Certainly it was clear that his hearing was normal up to and after that event, and the onset of the hearing loss was in adult life. He had been totally deaf for 12 years at the time of his referral, and gained no benefit from hearing aids or from a vibrotactile device. He had had no vertigo, imbalance or tinnitus. There was a strong family history of otosclerosis. His father had been deaf, although the cause had never been established. The patient's sister and son had both had stapedectomies.

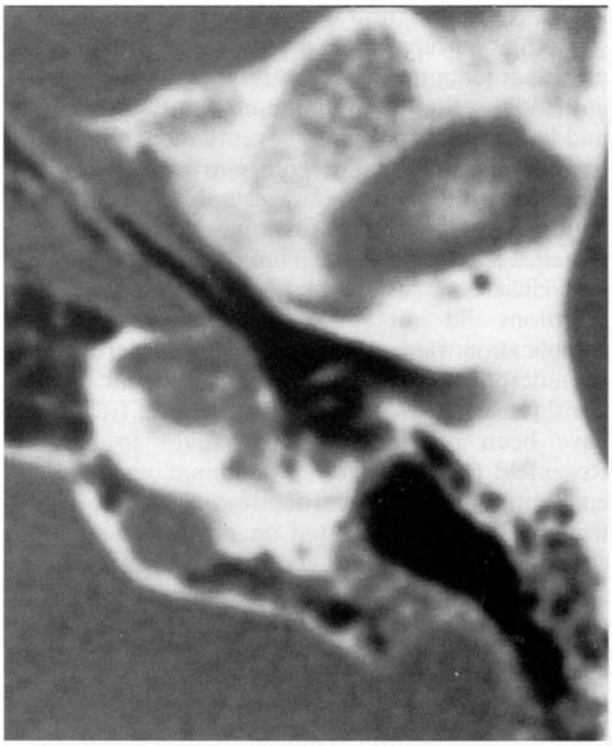

FIG. 1

High definition CT scan of the temporal bone (axial view) showing loss of the normal architecture of the cochlea with marked demineralization of the otic capsule.

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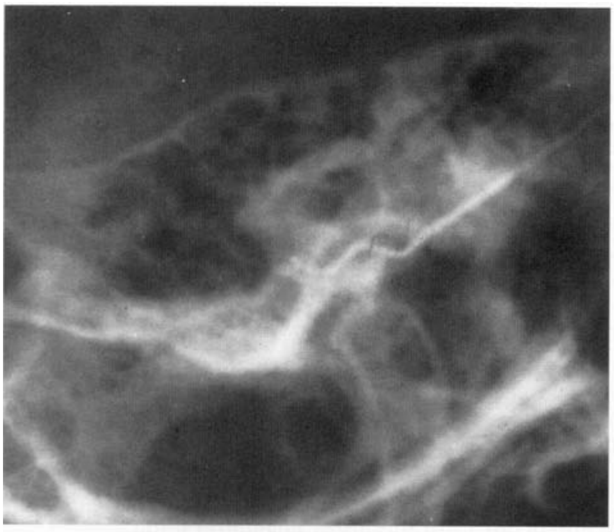

FIG. 2

Post-operative plain film of the implant showing the middle electrodes of the array (E9–E15) lying in the basal turn of the cochlea. The proximal electrodes (E1–E8) are in the middle ear and the distal electrodes (E16–E22) are seen to have passed out of the basal turn of the cochlea into a cavity in the temporal bone.

Pure tone audiometry yielded no responses to air conduction stimuli at any frequency up to 120 dB in either ear. No response was obtained on bone conduction at any frequency to 70 dB in either ear. Auditory brainstem response (ABR) using click stimuli failed to yield any response in either ear. Promontory stimulation testing was carried out using the Nucleus promontory testing device and auditory percepts were obtained in both ears at all frequencies of stimulation from 50 Hz to 800 Hz with good dynamic ranges. The Bench Kowal Bamford sentence score was 38 per cent, and the Consonant confusion test (VCV) score was 29/48 using lipreading alone. Computed tomography (CT) scanning of the temporal bones was performed and on both sides there was a loss of the normal definition of the cochlear architecture with demineralization of the surrounding otic capsule (Figure 1). The appearance was entirely typical of otosclerosis (otospongiosis). After discussion with the patient of the possible gains to be expected from cochlear implantation, the decision was made to offer him an implant and the operation was carried out on the left ear on 6/1/1994. The left ear was preferred because the CT images suggested this was the less abnormal side. A Nucleus 22 channel device was chosen. The technique employed a standard posterior tympanotomy approach retaining the incus. A separate cochleostomy was created just in front of the round window niche and a patent scala tympani was immediately entered. The electrode passed easily into the cochlea to about 7 mm at which point minimal resistance was encountered. Slight adjustment with the introducing claw combined with a little twisting movement easily overcame this resistance and the electrode passed readily into the cochlea until only two of the 22 active electrodes remained outside the scala tympani. Intra-operative stimulation of the stapedial reflex was not performed. Closure proceeded in the usual manner. Post-operatively there were no problems and on the second post-operative day a check X-ray was performed (Figure 2). This suggested that there had been some migration of the electrode. Seven electrodes (E9–E15) were seen to be in the basal turn of the cochlea; eight electrodes (E1–E8) were in the middle ear and seven electrodes (E16–E22)were seen to lie in a cavity in the temporal bone in front of the cochlea, closely related to the intrapetrous internal carotid artery. It was felt that the seven electrodes thought to be inside the cochlear might be capable of being stimulated, and that an attempt at mapping should be made rather than to remove and attempt to replace the electrode.

Programming details

Switch-on in bipolar mode (BP) one month after surgery gave auditory responses on E9–E15. Stimulation of E1–E8 which were thought to be in the middle ear gave a sensation of pressure or pain in the ear at 170 stimulus levels but no auditory sensation. Stimulation levels are arbitrary units expressed from the minimum level of stimulation 1 (approx 20μA) to a maximum level of stimulation 239 (approx 1750μA). Each arbitrary unit represents an approximate increase or decrease in current amplitude of two per cent. Typically T and C (threshold and comfort) levels are measured using stimulus levels. One of the electrodes (E6) gave rise to twitching in the orbicularis oculi muscle. Stimulation of E16–E22 gave no sensation of any type. The MAP of switch on T and C levels is shown in Table I and shows high levels for E9, E13, E14 and E15. Because of this he was initially switched on to E10, E11, and E12, using F_0 F_2 strategy. Attempts to introduce the other channels with the higher stimulation levels were unsuccessful because the patient preferred the quality of sound using the three channels alone. He used his implant in this stimulation mode for approximately 18 months during which time he underwent an intensive rehabilitation programme embracing both analytical methods, which involve the development of listening to segmental and suprasegmental elements of speech, and synthetic methods, which concentrate on real life listening skills using conversational-based training materials such as everyday sentences and connected discourse tracking. During this period he was seen for formal evaluation on three occasions (one month, nine months and 18 months) the results from which are shown in Table II. By his 18-month assessment he was scoring 60 per cent on Bench Kowal Bamford sentences in lip reading alone and 80 per cent with lip-reading + CI. His environmental sound score had risen from 6/20 at one month to 12.5/20. His own observations were very positive. 'It has made my life far more interesting and increased my confidence in meeting everyday problems. I no longer have to rely on my wife to

TABLE I

STIMULUS LEVELS AT SWITCH ON

Electrode	1	2	3	4	5	6	7	8	9	10	11
T levels	—	—	—	—	—	—	—	170	184	35	25
C levels	—	—	—	—	—	—	—	—	218	52	43
Electrode	12	13	14	15	16	17	18	19	20	21	22
T levels	40	145	160	235	235	—	—	—	—	—	—
C levels	61	164	183	239	—	—	—	—	—	—	—

TABLE II
CHANGE IN AUDITORY PERFORMANCE WITH TIME (F_0F_2 STRATEGY : THREE ACTIVE ELECTRODES

E 10, E11, E12	Pre-implant	Post-implant (1 month)	Post-implant (9 months)	Post-implant (18 months)
BKB LR	38%	—	35%	60%
LR+ES	—	—	44%	80%
VCV LR	29/40	—	20/48	20/48
LR+ES	—	—	23/48	30/48
ENV Sounds	—	6/20	11/20	12.5/20
FFA	—	48 dB	62 dB	51 dB

Key
BKB Bench Kowal Bamford Sentences.
VCV Consonant confusion test.
LR Lipreading alone.
LR+ES Lipreading plus electrical stimulation (ie with the implant).
ENV sounds Environmental sounds.
FFA Free field audiogram, with the implant; five frequency average (250Hz–4kHz).

ask all the questions. It is marvellous to hear things going on around you. I didn't realise how much I missed my hearing before. Now I don't like to switch off'. He reported using his implant from 5.30 am till 10 pm every day.

Shortly after his 18-month assessment he was upgraded to a SPECTRA speech processor for which he expressed a personal preference although there was no objective evidence of any improvement over the previous strategy. Programming was then attempted in a pseudomonopolar mode, using E1 or E7 as reference, but no improvement resulted. A further strategy was explored reverting to the BP mode of stimulation and altering the current levels rather than the stimulus levels.

Programming in this mode alters the current and the pulse width in tandem and using this strategy he was able to report pitch differences on eight channels. When

FIG. 3

Temporal bone section from a patient with otosclerosis/otospongiosis, showing a demineralized cavity in relation to the basal turn of the cochlea. (Reproduced by kind permission of Professor Bernard Fraysse, Toulouse).

assessed in this mode there was no change in free field measurements or in his environmental sound scores. However he felt that the new MAP gave him more awareness of background noise, and for the first time he was aware of voices on the radio at work and of music. (A MAP, sometimes referred to as a programme, contains patient specific parameters obtained during programming, including threshold (T) and comfort (C) levels together with the speech processing strategy being used eg SPEAK, MPEAK, $F_0F_1FD_2$ etc)

Discussion

The temporal bone changes in otosclerosis are well recognized and have been summarized by Schuknecht (1974). There is an early process of osteolysis with resorption of endchondral bone, probably as result of lysosyme activity. This may proceed to the formation of new immature basophilic bone which eventually goes on to more mature acidophilic new bone. The process of bone resorption and new bone formation go on simultaneously, and the degree of destruction of the otic capsule varies considerably from individual to individual, as does the severity of the associated cochlear deafness which probably results from toxic metabolic changes produced in the cochlear fluids. Demineralization of the temporal bone can frequently be demonstrated on high definition CT imaging, the extent varying from a small localized area of radiolucency to widespread osteolysis of the temporal bone, most extensive changes often being seen in osteogenesis imperfecta. One commonly observed pattern is that of a radiolucent band paralleling the basal turn of the cochlea and resembling a reduplication or mirror-imaging of the basal turn (Valvassori, 1984; Schwartz *et al.*, 1985). Figure 3 shows how a large osteolytic cavity may be separated from the cochlear lumen by no more than the thickness of the cochlear endosteum. It is not hard to imagine how an intracochlear implant could pass through the anterior end of the scala tympani of the best turn into such a cavity. In the case described slight resistance was felt as the electrode reached that position. However, that is a point which it is not uncommon to encounter a little resistance as the device passes from the basal to the middle turns and there was certainly nothing to suggest at the time that the electrode could have ended up where it did. Intra-operative imaging would have allowed immediate detection of this misplacement but it was not, and is still not, part of our routine.

The case also illustrates that such a misplacement need not imply a poor result. The eight electrodes in the cochlea were programmable and a MAP was created which gave

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useful information transfer and brought about a very considerable improvement in the patients communication abilities. In fact the patient actually preferred the auditory percept using only three electrodes. It is also clear that such a programming problem tests the resourcefulness of the rehabilitation team who may have to experiment with a number of different strategies in order to maximize the usefulness of the implant. The usual MPEAK strategy could not be employed as it requires a minimum of 10 active electrodes. With the F_0F_2 strategy F_0 represents the fundamental frequency and F_2 represents the mid-frequency spectral peak in the range of approximately 800 Hz-4000 Hz (ie second formant information) F_0 and F_2 are coded as rate and place of stimulation respectively.

Facial nerve stimulation in otosclerosis

Although not a prominent feature of this case, another concern associated with cochlear implantation in otosclerosis is unwanted facial nerve stimulation. Whereas facial nerve stimulation has been described in 0.9 per cent (Cohen *et al.*, 1988) to 15 per cent (Niparko *et al.*, 1991) of all cases, the incidence appears to be higher in the presence of otosclerosis. For instance, in the series described by Muckle and Levine (1994), only four out of 38 implanted patients experienced facial nerve stimulation. Of these, three had otosclerosis identified as the cause of their deafness. Similarly, Weber *et al* (1995) noted that of 17 patients they reported with facial nerve stimulation, 11 had otosclerosis. Indeed, in this series nearly 50 per cent of the implanted patients with otosclerosis suffered from facial nerve stimulation. The site of the facial nerve stimulation is unclear. Graham *et al.* (1989) had to reposition a single channel device because of facial nerve stimulation in a patient with otosclerosis. They found that the electrode caused facial twitching if placed on the promontory, attic or round window area. Placement of the electrode deep into the cochlea reduced the twitching. This suggested to them that the stimulation was in the tympanic portion of the facial nerve. Others have suggested the labyrinthine portion of the facial nerve. Electrodes that have to be inactivated to prevent facial stimulation can be basal or apical (Muckle and Levine, 1994; Weber *et al.*, 1995). Why this facial nerve stimulation occurs in otosclerosis is unclear. Current spread in temporal bones appears to follow a linear resistive model (Clopton and Spelman, 1982). Our own experience indicates that the other situation in which facial nerve stimulation from an implanted device may occur is after temporal bone fracture, and that too is presumably due to current spread through a line of decreased electrical resistance (Camilleri *et al.*, 1996; Maas *et al.*, 1996). No one has succeeded, to our knowledge, in demonstrating reduced impedance to current spread of otosclerotic bone compared to normal bone. Since the stimulus intensities needed for thresholds are not higher in otosclerotic bones, the facial nerve stimulation must result either from lowering of the bone electrical impedance by the disease, or by reducing the distance from the electrode to the facial nerve, perhaps by cavity formation. Most of the literature seems to favour the former, although the case reported here suggests that a sufficiently large otospongiotic cavity might be capable of transfer of current from the implant to the facial nerve.

Conclusion

A case is described in which a multichannel cochlear implant passed through the anterior end of the scala tympani of the basal turn of the cochlea into an otospongiotic cavity in the temporal bone. Only a small number of electrodes which could be stimulated remained in the cochlea. Useful benefits in communication skills were obtained by employing an F_0F_2 strategy.

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