

ORIGINAL ARTICLE

Cochlear Implantation in far Advanced Otosclerosis: A Surgical, Audiological and Quality of Life Review of 35 Cases in a Single Unit

Edward P. Flook, Stephen J Broomfield, Shahad Saeed, Iain A. Bruce, Kevin M. Green

ENT Registrar, Manchester (EPF, SJB)

Pre-registration Clinical Scientist (audiology), Manchester Cochlear Implantation Programme (SS)

ENT Consultant, Manchester (IA, KMG)

Objective: This is a review of our experience of implanting patients with far advanced otosclerosis, specifically reviewing pre-operative predictors of differences in audiological outcome, complications (including facial nerve stimulation (FNS)) and patient satisfaction.

Materials and Methods: A retrospective case review including audiology assessments was undertaken.

Results: Thirty-five patients were identified as having far advanced otosclerosis from the Manchester Cochlear Implant programme database. Twenty-seven patients had good outcomes (PTA threshold mean of 39dB, AB phonemes mean of 67%, BKB mean of 80%, CUNY mean of 93%). The remaining 8 had reasonable PTA thresholds but poorer audiological test scores. Analysis of these 8 cases showed that incomplete electrode insertion and electrode "switch off", to reduce FNS, are likely to have had the most effect. Six patients had FNS, all of which resolved with altered electrode mapping. Fourteen patients had implant devices with a straight, lateral wall lying electrode, and 6 (43%) of these had FNS. None of the 21 patients implanted with a perimodiolar electrode had any FNS.

Conclusion: Results for cochlear implantation of patients with far advanced otosclerosis generally have good surgical success, good audiological outcomes and high satisfaction rates. Tinnitus is improved in most patients. The main factor leading to poorer outcome was in the use of only a limited number of electrode channels due to FNS or partial electrode insertion. Electrode design has a key role in facial nerve stimulation.

Submitted : 23 August 2010

Revised: 22 December 2010

Accepted : 22 December 2010

Introduction

Cochlear implantation (CI) is a highly successful technique of improving severe and profound hearing loss. It is performed in adults and children with many different aetiologies of hearing loss.

Otosclerosis is caused by abnormal bone growth in the middle or inner ear. This otospongiotic bone can vary in site and severity with resultant stapes fixation, fenestral otosclerotic plaques and retrofenestral disease in cochlear otosclerosis. The site of otosclerotic bone growth can reliably be seen on high resolution CT scanning [1]. Initially hearing loss is conductive due to the ossicular fixation and later sensorineural hearing loss if the cochlea is involved. Mixed hearing loss is not uncommon. When surgical intervention is initiated the majority of patients undergo a stapedotomy or stapedectomy. Far advanced otosclerosis has been defined as when bone conduction pure tone thresholds are not measurable using a

standard audiometer and air conduction thresholds are no better than 85dB but other authors have suggested alternate definitions [2]. Even in far advanced otosclerosis, stapedectomy can improve the conduction mechanism to allow the hearing to be aidable [2-6]. CI may play a role in two scenarios: In dead ears implicating unsuccessful stapedectomy or if stapedectomy is deemed unlikely to help.

Facial nerve stimulation (FNS) is when the current from the CI electrode unintentionally stimulates the facial nerve causing facial twitching or paraesthesia [7]. Eleven previous international case series have shown varying rates of FNS in otosclerotic patients. A total of 227 otosclerosis patients received cochlear implantation and had generally good outcomes, with 54 (24%) having some FNS [8-18]. Most papers describe that altering the electrode mapping will reduce or stop the FNS and only very rarely does it result in implant failure.

Corresponding address:

Edward P. Flook
17 Willow Way, Manchester, M20 6JT, UK
Phone: +44 (0) 7932081662
E-mail: edflook@yahoo.com

The Glasgow Benefit Index (GBI) is a measure of patient benefit, and not of health status per se. It measures the changes in health status resulting from otorhinolaryngological intervention, but does not provide a measure of the status of the patient either prior to or after the intervention. The benefit scale ranges from -100 (maximal negative benefit), through 0 (no benefit), to +100 (maximal benefit) was recorded.

This is a review of our experience of implanting far advanced otosclerotic patients, specifically reviewing pre-operative predictors of differences in audiological outcome, complications (including FNS) and patient satisfaction.

Materials and Methods

Thirty-five patients with otosclerosis were identified from the Manchester Cochlear Implant programme database. A retrospective case note review, including audiology assessments was undertaken. Audiological outcomes (including quality of life questionnaire scores) were reviewed 3 months post switch-on and again between 9 months and 2 years. The departmental standard audiological assessment included pure tone audiogram (PTA), Arthur Boothroyd (AB) speech recognition test (using phonemes and words), Bamford-Kowal-Bench (BKB) sentences in quiet and noise and City University of New York sentence test (CUNY). Glasgow Benefit Index and tinnitus severity were also reviewed.

The patients identified received their implants from 1989 to 2009. A few of these patients have received a sequential second side CI, but these patients were only included in this study for their first CI. With cases that had CI in the late Eighties and early Nineties, audiological testing, quality of life questionnaires and pre-operative scanning were not performed or stored reliably in the medical notes resulting in a number of patients with incomplete datasets (these cases were included). No other cases were excluded.

Patients had been identified as having otosclerosis prospectively either clinically and/or surgically and/or radiologically.

Results

Thirty-five patients underwent cochlear implantation to rehabilitate their severe to profound hearing loss resulting from otosclerosis. Various devices were implanted: 13 had Cochlear CI24K-CON (Cochlear Ltd, NSW, Australia), 10 patients had a Cochlear CI22M, 8 had Cochlear Freedom, 2 had Med-El C40+ (Med-El, Innsbruck, Austria), 1 had Med-El Sonata Flex and 1 had Ineraid (Symbion Inc, Utah, USA).

Nine cases were diagnosed clinically, all others had stapes footplate fixation confirmed on middle ear exploration or had CT evidence of otosclerosis. Review of the 23 CT scan reports showed that 8 had fenestral otosclerosis, 12 had retrofenestral (cochlear) otosclerosis while 3 had no radiological evidence of otosclerosis. In these 3 patients a clinical diagnosis of otosclerosis was made due to the gradual nature of hearing loss, family history of otosclerosis, and a significant deterioration during pregnancy.

Patients were implanted aged 35-79, median 64 yrs old. Duration of profound deafness ranged from 18 months to 40 years, median 12.5 years. Surgical implantation was mostly uneventful. Difficulty was noted in 4 cases where 3 had partial insertion of electrode, and the fourth noted difficulty in finding the correct cochlear lumen due to new bone growth. Post implantation aided hearing thresholds and speech testing improved in 95% of cases between 3 month testing and 9 month testing and audiology outcome scores were therefore taken from the 9 month assessment. Thresholds after 9 months (recorded in 33 patients, averaged 500Hz, 1, 2, 4kHz) had a mean of 41 db (range 28-65). Audiological testing showed mean scores of 58% (3-90) for AB phonemes (recorded in 31 patients), 64% (2-100) for BKB in quiet (33 patients), and 83% (3-100) for CUNY (32 patients).

After 9 months 8 patients had worse audiology outcome compared to the rest of the patients (recording at least one from either AB phonemes <30%, BKB in quiet <40%, CUNY scores <50% or PTA mean >55dB). Excluding these poorer outcome patients, the rest of the implantees (25) had very good outcomes with a PTA mean threshold of 39 dB (range 28-53), AB phonemes mean 67% (50-90), BKB mean 80% (44-100) and CUNY mean score 93% (59-100). Figures 1-4 highlight that although 8 worse outcome patients had poor AB and BKB scores, their PTA thresholds and CUNY scores ranged from poor to good. These 8 patients,

although worse than the rest of the patients, still had mean hearing thresholds of 48dB and mean CUNY scores of 50. Six of the 8 patients could not use the full complement of electrode channels (3 patients required some electrode channels to be “switched off” due to FNS, 2 patients had only partial insertion of the electrode, 1 patient the basal turn lumen was difficult to find due to new bone formation).

Cochlear otosclerosis was seen on CT in 12 CI recipients, 4 of these were in the worse outcome group as discussed above while the other 8 had good outcomes, full electrode insertion and good electrode channel usage. Audiological scores of these 8 showed a mean PTA threshold of 35dB (range 28-51), AB phonemes mean score of 71% (56-90), BKB mean score of 90% (71-100) and CUNY scores of 93% (60-100) in keeping with the rest of the good outcome patients. Two patients with cochlear otosclerosis seen on CT scanning had FNS.

FNS was seen in 6 patients during implantation “switch on” and mapping. In all of these cases it was not possible to use a full complement of electrodes for the channel mapping, and in 2 patients less than half could be used. None of these patients had to have the implant switched off or removed. Three of these patients had worse audiological outcomes than the cohort average, 2 had good audiological outcomes while 1 patient had no audiological data available. 5 patients with FNS had

Cochlear CI22M implants whilst 1 had Med-El C40+. Only one patient had other extra-cochlear stimulation in which they had FNS and pain from many electrodes (NB partial electrode array insertion only), this settled with altered mapping but resulted in only 5 electrode channels being used.

Previous stapedectomies that were initially successful then failed, were not successful at all or resulted in dead ear had occurred in 10 patients. Their outcome from CI was good and unaffected by the previous surgery.

Twenty patients had the GBI recorded; the mean GBI score was 53 suggesting that the patients felt it was of good benefit. The range of scores was 31 to 81 demonstrating that no patients considered CI to have had little or no benefit. 4 of the 8 worse outcome patients had GBI scores taken with a mean of 54 (range 36-75).

Tinnitus severity scoring, using the Tinnitus Handicap Inventory, was taken in 12 patients pre-operatively and again post implantation. 10 patients had improvement with 1 showing no change and 1 showing an increase at the 3 month assessment then improving (but still slightly worse than pre-operatively) at the 9 month assessment. An incomplete data set made it difficult to correlate tinnitus scores to implant audiological outcomes or FNS; there is no correlation from our results.

Discussion

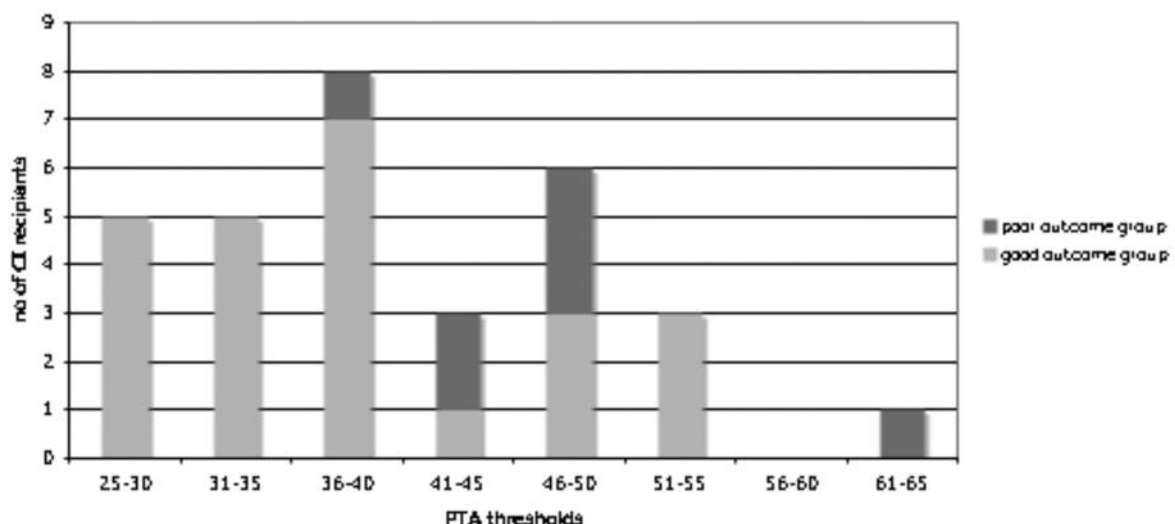


Figure 1. Mean hearing threshold scores

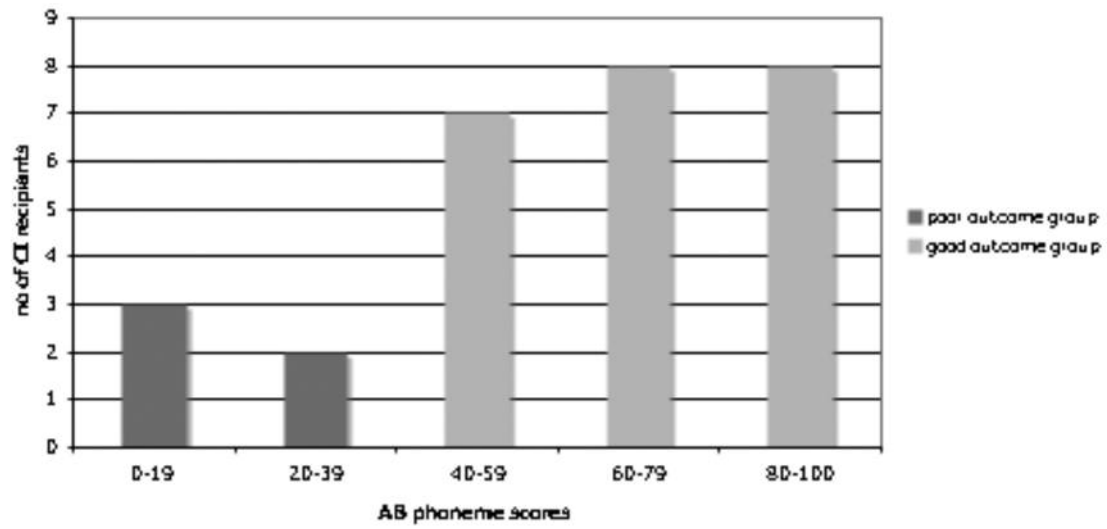


Figure 2. Mean AB phoneme scores

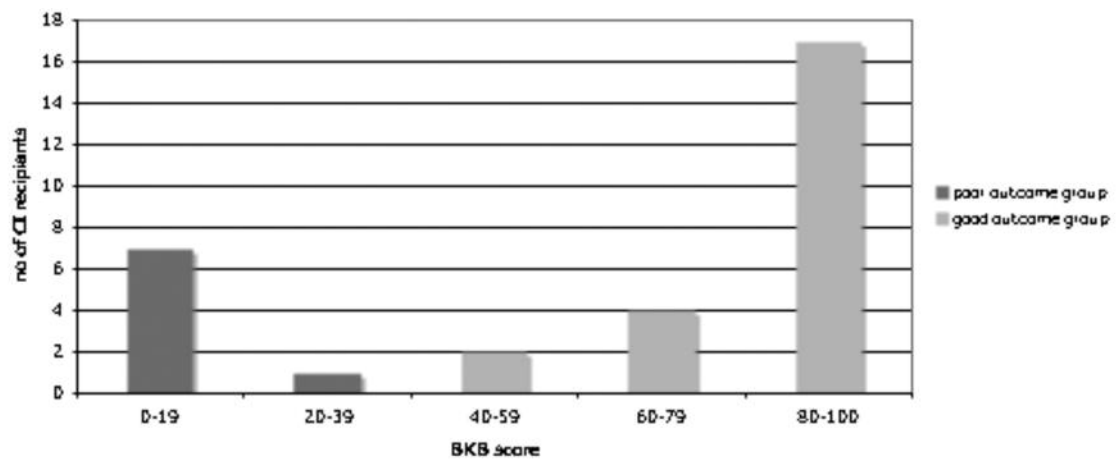


Figure 3. Mean BKB in quiet scores

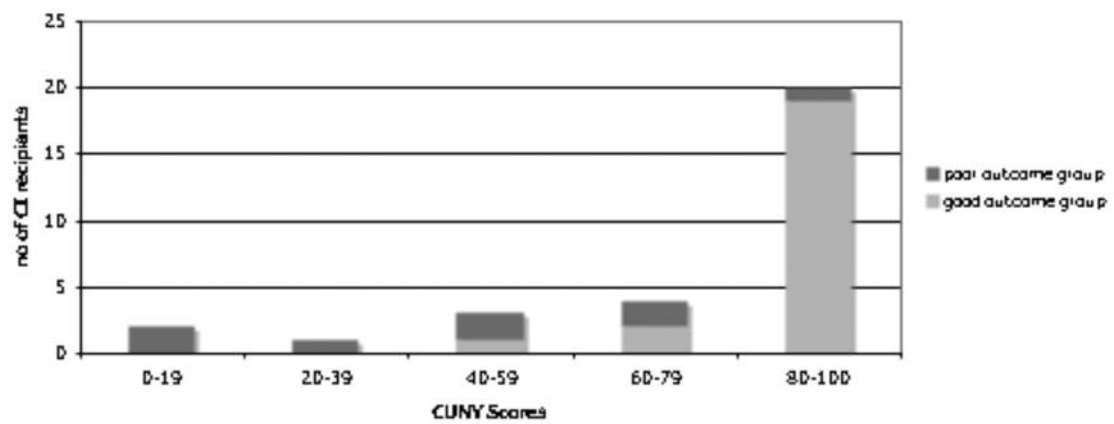


Figure 4. Mean CUNY scores.

Although in far-advanced otosclerosis stapedectomy and hearing aid use may be a suitable treatment option, this review has shown that cochlear implantation may play a role. Our results and those of other workers in this area demonstrate that previous stapedectomy does not influence outcomes from cochlear implantation [9,11,12].

In the present study we identified 8 cases where there were generally worse audiological results than the rest, although still gained reasonable PTA thresholds. Possible factors influencing the outcome in these 8 patients were further analysed. The inability to use the full complement of electrode channels is most likely to have had most effect with poorer sound quality as a result. Ages and duration of profound deafness prior to implantation were similar to the whole group averages.

Pre-operative findings that influence outcome are not clear. 4 out of the 12 patients with cochlear otosclerosis seen on CT had a worse outcome, related to partial electrode insertion and FNS, but the other 8 had good outcomes. This shows that it is not a clear predictor of outcome. No other factors could be distinguished from our case review and results to predict the poor outcome patients pre-operatively.

Twenty-one patients had cochlear implant devices with perimodiolar electrodes and 14 had devices with straight electrodes which rest on the lateral wall in the cochlea. Of the 6 patients who had some FNS, all 6 had devices with a straight electrode (Cochlear CI22M and Med-El 40+), and none of the perimodiolar electrode devices caused FNS. It has been shown on computer modelling that there is a requirement for increased stimulation levels due to the increased conductivity in surrounding otosclerotic bone that reduces current density in the scala tympani as current leaks more easily out of the cochlea [18]. Facial nerve stimulation can occur due to current leak from the electrode, especially when there is partial electrode insertion. No patient factors pre-operatively (i.e. far advanced disease, cochlear otosclerosis or age) appear to influence outcome. The type of implant and electrode design may affect the level of current threshold required for stimulation. Perimodiolar designs with more focused electrical stimulation (and therefore

lower thresholds for stimulation and less current leak) reduces the likelihood of FNS [19]. Other studies have had similar findings [8,14]. All FNS could be controlled by altering mapping but resulting in less electrode channels being used and potentially worse audiological results. Two reports (6 CI cases in total) of FNS affecting electrode channel use resultant poor audiological outcome had the straight electrode device exchanged for a perimodiolar device and showed great improvement of audiological tests and no FNS [20,21]. The current study has shown 6 of 14 (43%) straight electrode implants resulted in some FNS while 0 of 21 perimodiolar electrodes had FNS.

Conclusion

Results for cochlear implantation of patients with far advanced otosclerosis generally have good surgical success, good audiological outcomes and high satisfaction rates. Tinnitus is improved in most patients. The main factor leading to poorer outcome was in the use of only a limited number of electrode channels due to FNS or partial electrode insertion. Perimodiolar electrode devices produced no facial nerve stimulation compared with 43% of recipients of straight electrode devices, strongly suggesting that electrode design has a key role in facial nerve stimulation.

References

1. Lee TC, Aviv RI, Chen JM, Medzelski JM, Fox AJ, Symons SP. CT Grading of Otosclerosis. *American Journal of Neuroradiology* 2009;30:1435-9.
2. Frattali MA, Sataloff RT. Far-advanced otosclerosis. *Ann Otol Rhinol Laryngol* 1993;102:433-7.
3. Glasscock ME, Storper IS, Haynes DS, Bohrer PS. Stapedectomy in profound cochlear loss. *Laryngoscope* 1996;106:831-3.
4. Khalifa A, el-Guindy A, Erfan F. Stapedectomy for far-advanced otosclerosis. *J Laryngol Otol* 1998;112:158-60.
5. Lippy WH, Burkey JM, Arkis PN. Word Recognition Score Changes After Stapedectomy for Far Advanced Otosclerosis. *Am J Otol* 1998; 19: 56-8.
6. Shea PF, Ge X, Shea JJ Jr. Stapedectomy for Far-Advanced Otosclerosis. *Am J Otol* 1999;20: 425-9.
7. Broomfield S, Mawman D, Woolford TJ, O'driscoll

- M, Luff D, Ramsden RT. Non-auditory stimulation in adult cochlear implant users. *Cochlear Implants Int* 2000;1:55-66.
8. Ruckenstein MJ, Rafter KO, Montes M, Bigelow DC. Management of far advanced otosclerosis in the era of cochlear implantation. *Otol Neurotol* 2001;22:471-4.
9. Rayner MG, King T, Djalilian HR, Smith S, Levine SC. Resolution of facial stimulation in otosclerotic cochlear implants. *Otolaryngol Head Neck Surg* 2003;129:475-80.
10. Marshall AH, Fanning N, Symons S, Shipp D, Chen JM, Nedzelski JM. Cochlear implantation in cochlear otosclerosis. *Laryngoscope* 2005;115:1728-33.
11. Quaranta N, Bartoli R, Lopriore A, Fernandez-Vega S, Giagnotti F, Quaranta A. Cochlear implantation in otosclerosis. *Otol Neurotol* 2005;26:983-7.
12. Rama-López J, Cervera-Paz FJ, Manrique M. Cochlear implantation of patients with far-advanced otosclerosis. *Otol Neurotol* 2006;27:153-8.
13. Psillas G, Kyriafinis G, Constantinidis J, Vital V. Far-advanced otosclerosis and cochlear implantation. *B-ENT* 2007;3:67-71.
14. Calmels MN, Viana C, Wanna G, et al. Very far-advanced otosclerosis: stapedotomy or cochlear implantation. *Acta Otolaryngol* 2007;127:574-8.
15. Matterson AG, O'Leary S, Pinder D, Freidman L, Dowell R, Briggs R. Otosclerosis: selection of ear for cochlear implantation. *Otol Neurotol* 2007;28:438-46.
16. Ramsden R, Rotteveel L, Proops D, Saeed S, van Olphen A, Mylanus E. Cochlear implantation in otosclerotic deafness. *Adv Otorhinolaryngol* 2007;65:328-34.
17. Mosnier I, Bouccara D, Ambert-Dahan E, Ferrary E, Sterkers O. Cochlear implantation and far-advanced otosclerosis. *Adv Otorhinolaryngol* 2007;65:323-7.
18. Sainz M, Garcia-Valdecasas J, Ballesteros JM. Complications and pitfalls of cochlear implantation in otosclerosis: a 6-year follow-up cohort study. *Otol Neurotol*. 2009;30:1044-8.
19. Frijns JH, Kalkman RK, Briaire JJ. Stimulation of the facial nerve by intracochlear electrodes in otosclerosis: a computer modelling study. *Otol Neurotol*. 2009;30:1168-74.
20. Battmer R, Pesch J, Stöver T, Lesinski-Schiedat A, Lenarz M, Lenarz T. Elimination of facial nerve stimulation by reimplantation in cochlear implant subjects. *Otol Neurotol* 2006;27:918-22.
21. Polak M, Ulubil SA, Hodges AV, Balkany TJ. Revision cochlear implantation for the facial nerve stimulation in otosclerosis. *Arch Otolaryngol Head Neck Surg*. 2006;132:398-404.