Speech Perception after Cochlear Implantation in 53 Patients with Otosclerosis: Multicentre Results

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Key Words
Cochlear implantation · Otosclerosis · Computed tomography · Surgery · Facial nerve stimulation · Speech perception performance

Abstract
Objectives: To analyse the speech perception performance of 53 cochlear implant recipients with otosclerosis and to evaluate which factors influenced patient performance in this group. The factors included disease-related data such as demographics, pre-operative audiological characteristics, the results of CT scanning and device-related factors. Methods: Data were reviewed on 53 patients with otosclerosis from 4 cochlear implant centres in the United Kingdom and the Netherlands. Comparison of demographics, pre-operative CT scans and audiological data revealed that the patients from the 4 different centres could be considered as one group. Speech perception scores had been obtained with the English AB monosyllable tests and Dutch NVA monosyllable tests. Based on the speech perception scores, the patients were classified as poor or good performers. The characteristics of these subgroups were compared. Results: There was wide variability in the speech perception results. Similar patterns were seen in the phoneme scores and BKB sentence scores between the poor and good performers. The two groups did not differ in age at onset of hearing loss, duration of hearing loss, progression, age at onset of deafness, or duration of deafness. Conclusions: The clinical presentation of the otosclerosis (rapid or slow progression) did not influence speech perception. Better performance was related to less severe signs of otosclerosis on CT scan, full insertion of the electrode array, little or no facial nerve stimulation and little or no need to switch off electrodes.

Introduction
Nowadays, cochlear implantation is a well-accepted and effective intervention in patients with profound hearing loss. A large number of studies have shown that the majority of adults and children with a cochlear implant (CI) achieve word scores of more than 50% on speech perception tasks [Dowell et al., 1986; Valimaa et al., 2001; Oh et al., 2003]. However, performance varies widely and there are still a number of users who do not reach this level of performance.

Several attempts have been made to explain this variance in order to predict the benefit of cochlear implantation.
Otosclerosis is a heritable disease that affects the bony structure of the temporal bone. In the active phase, so-called osteospongiosis, the normal lamellar bone is resorbed and through a vascular stage is replaced by thick, irregular bone in the normal middle layer of the otic capsule [Guneri et al., 1996]. The subsequent hearing loss can be conductive, which is most commonly caused by stapes fixation due to plaque formation around the oval window, or sensorineural in the case of cochlear involvement. Sensorineural hearing loss in otosclerosis is thought to be the result of narrowing of the cochlear lumen with distortion of the basilar membrane [Linthicum et al., 1975] or it is believed to be caused by lytic enzymes that are released into the perilymph from otosclerotic foci [Mafee et al., 1985b; Linthicum, 1993; Youssef et al., 1998].

Long-term follow-up studies showed that about 10% of ears with otosclerosis and conductive hearing loss also developed sensorineural hearing loss [Browning and Gatehouse, 1984; Ramsay and Linthicum, 1994].

High-resolution computed tomography is at present the imaging modality of choice for the assessment of the osseous labyrinth, labyrinthine windows and cochlear capsule. It can detect abnormalities of the oval window area in 80–90% of patients with surgically proven otosclerosis. Sensitivity approaching 90% for fenestral otosclerosis has been demonstrated [Mafee et al., 1985a]. One cannot conclude that otosclerosis is not present when demineralization is not present in CT, but one can be virtually certain that this disease is present when it is seen. CT is therefore highly specific [Donaldson and Snyder, 1993].

In otosclerosis patients, there seems to be a trend towards fewer active electrodes and poorer scores on postoperative open-set sentences tests than in CI recipients with other causes of deafness [Blamey et al., 1992]. Histological studies have shown that otosclerosis has a relatively small effect on spiral ganglion cell survival compared to other causes of deafness [Nadol et al., 1989]. Thus the poorer scores in otosclerosis patients might be explained by the lower number of active electrodes, the altered bone properties in the otic capsule that may affect the current distributions produced by the electrodes and possibly the older average age at implantation, rather than a diminished neural response.

In this multicentre study, a group of 53 otosclerosis patients with a CI were reviewed at the CI centres in Manchester, Birmingham, Utrecht and Nijmegen. Patient characteristics, CT scans, surgical findings and the incidence of facial nerve stimulation (FNS) have been described in a previous paper [Rotteveel et al., 2004]. First, a search was made for inter-clinic differences in factors that might affect auditory performance. Second, longitudinal speech perception scores were analysed to establish relations between speech perception scores and several factors related directly or indirectly to otosclerosis.

Material and Methods

Subjects
Patients diagnosed with otosclerosis were retrieved from the databases of 4 CI centres in the Netherlands and United Kingdom that hold prospective data: University Hospital Birmingham, Manchester Royal Infirmary, Radboud University Nijmegen.
Medical Centre and University Medical Centre Utrecht. The diagnosis of ‘otosclerosis’ was based on the presence of otosclerotic lesions on the pre-operative CT scan, history of stapes surgery, or the finding of fixation of the stapes during the surgical implantation procedure. A total of 53 patients were included: 19 patients (36%) had signs of otosclerosis on the CT scan, 28 patients (53%) had a positive CT scan and a history of stapes surgery, 5 patients (9%) had a normal CT scan and a history of stapes surgery and one patient was diagnosed solely by the finding of a stapes fixation during the implantation procedure. The year of implantation of the patients ranged from 1990 to 2002. There was no difference in the mean and median year of implantation between the 4 centres. As new implant technologies are known to influence speech perception results [Krueger et al., 2008], the device types that had been used at each of the centres were evaluated (table 1). No differences were found in the distribution of the previous generation (Nucleus 22 and Clarion I) and the more recent generation devices (Nucleus 24 and Clarion II) between the 4 centres (Kruskal-Wallis test, p = 0.52).

Table 1. Number of implanted devices per CI centre

<table>
<thead>
<tr>
<th>Device type</th>
<th>Nijmegen (13 patients)</th>
<th>Utrecht (9 patients)</th>
<th>Birmingham (17 patients)</th>
<th>Manchester (14 patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Previous devices</td>
<td>9</td>
<td>5</td>
<td>11</td>
<td>10</td>
</tr>
<tr>
<td>Nucleus 22</td>
<td>4</td>
<td>3</td>
<td>11</td>
<td>9</td>
</tr>
<tr>
<td>Clarion S</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Clarion I</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Med-el 40+</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Recent devices</td>
<td>4</td>
<td>4</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td>Nucleus 24</td>
<td>3</td>
<td>4</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td>Clarion II</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Table 2. Extent of otosclerosis on the pre-operative CT scans: 3 types

<table>
<thead>
<tr>
<th>Otosclerotic lesions of the otic capsule</th>
<th>Type 1</th>
<th>Type 2</th>
<th>Type 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Solely fenestral involvement (thickened footplate and/or narrowed or enlarged windows)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Retrofenestral, with or without fenestral involvement</td>
<td>Type 2a: double ring effect</td>
<td>Type 2b: narrowed basal turn</td>
<td>Type 2c: double ring and narrowed basal turn</td>
</tr>
<tr>
<td>Severe retrofenestral (unrecognizable otic capsule) involvement, with or without fenestral involvement</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Over half of the patients had undergone stapes surgery prior to cochlear implantation. The proportion of patients with a history of stapes surgery in either ear was significantly higher in the patient group from Utrecht (100%) than in the patient groups from Birmingham (41%) and Manchester (57%); there were no differences in stapes surgery between the other groups (Fisher’s exact test). The proportions of patients who had pre-operative experience with a conventional hearing aid at the time of implantation did not differ between the 4 centres (Kruskal-Wallis test, p = 0.67).

Pre-Operative Evaluation Data

As part of the selection procedure for cochlear implantation, the patients at all 4 CI centres had undergone CT scanning of the temporal bone. When available, these CT scans were reviewed by the same experienced otologist. It appeared that the CT scans of 17 patients had been destroyed. In these cases, the diagnoses were based on the original reports by the radiologists at the CI centres. The CT scans were reviewed for fenestral involvement (narrowed or enlarged window, thickened footplate) and retrofenestral involvement (double ring effect, narrowed basal turn) of the otosclerotic process and were categorized into 3 types (table 2) [Rotteveel et al., 2004].

Postoperative Evaluation Data

First, at each CI centre, the patients’ speech processor programming notes were evaluated to gather information on the need to lower stimulation levels or switch off electrodes to eliminate non-auditory effects, such as FNS and pain or stinging sensations in the middle ear or throat.

Second, longitudinal speech perception scores were retrieved from the medical files. At all 4 clinics, speech perception measurements had been carried out in special sound-treated booths. The speech material was recorded on tape or CD and presented by a loudspeaker placed in front of the patient. Although speech perception measurements were part of the regular evaluation visits at all 4 centres, the time interval between measurements varied. The English CI centres had recorded data on the open-set Bamford-Kowal-Bench (BKB) sentences test [Bench et al., 1979] and/or phoneme scores on the open-set Arthur Boothroyd (AB) monosyllables test [Boothroyd, 1968]. Phoneme scores had also been obtained by the 2 Dutch CI groups, using the open-set NVA monosyllables tests [Bosman and Smoorenburg, 1995]. The AB and NVA are largely comparable; the 2 tests comprise a large number of lists that consist of 10 isophonemic balanced CVC (consonant-vowel-consonant) words. As the speech recognition intensity curves obtained from subjects with normal hearing were fairly comparable [Michael, 1987; Bosman and Smoorenburg, 1995] and the test scores had been obtained at a fixed level of 40 dB above the speech reception threshold of controls with normal hearing.
hearing, it was decided to pool these data for statistical analysis. This presentation level of 40 dB above speech reception threshold resembles about 65 dB SPL, the overall level of normal speech.

In this study, first the data from the 4 CI centres were compared to each other with respect to factors that might affect auditory performance such as demographics, pre-surgical audiological characteristics, CT scan results and the types of implants used, to search for interclinic differences.

For further analysis, the whole patient group was divided into poor and good performers, based on the speech perception scores of a large reference group of postlingually deaf adult CI patients using the same devices. The reference group for the phoneme scores comprised 76 Dutch CI patients implanted between 1991 and 2002, with full insertion of the electrode array, who were retrieved from the Nijmegen/Viataal CI centre’s database, excluding the patients with otosclerosis. An evaluation of the phoneme scores of the reference group showed a mean phoneme score of 55% and the 25th percentile at 40% correct. The 25th percentile was used as the criterion for inclusion in either the ‘good-performance subgroup’ (group 1) or the ‘poor-performance subgroup’ (group 2).

The reference group for the BKB sentence test scores comprised 100 English patients with full insertion of the electrode array retrieved from the database of the Birmingham CI centre; the 25th percentile of the BKB data was 47% correct. A good performer was defined as a person with a score on BKB sentences test higher than 47%; individuals with a score lower than 47% were defined as poor performers. The characteristics of the 2 subgroups of ‘good’ and ‘poor’ performers were compared to evaluate for features distinctive for good or poor outcomes.

**Results**

**Inter-Clinic Differences**

**Demographic Data**

Some demographic data from the patients at the CI centres are shown in table 3. The patients at the 4 centres did not differ in age at onset of progressive hearing loss, duration of progressive hearing loss, age at onset of deafness, duration of deafness and age at implantation (Kruskal-Wallis nonparametric test, p > 0.05). The proportions of female patients per centre ranged from 23 to 44%; the differences were not significant (Kruskal-Wallis test). On the basis of these demographics, the patients at the 4 centres were considered to be largely comparable.

Pre-Operative Evaluation Data

At all 4 centres, the patients had to be profoundly deaf to enter the cochlear implantation programme, so variations in residual hearing were limited. Mean pre-operative unaided hearing thresholds at 0.5, 1, 2 and 4 kHz exceeded 110 dB at all 4 centres.
The extent of otosclerosis on the CT scans was categorized into 3 types (table 2). Although the patient group from Utrecht seemed to have a somewhat higher proportion of patients with type 3 (i.e. severe retrofenestral otosclerosis/unrecognizable otic capsule) and fewer patients with type 2 (i.e. retrofenestral involvement) (fig. 1), $\chi^2$ tests revealed that these differences in occurrence of type 1, 2 and 3 at the 4 centres were not significant. In addition, there were no significant differences in the proportions of patients with partial insertion of the electrode array between the 4 centres (Kruskal-Wallis tests; $p = 0.87$).

### Categorization According to Patient Performance Phoneme Scores

Phoneme scores were available from 19 out of the 31 English-speaking patients on the AB monosyllable test and from all 22 Dutch-speaking patients on the NVA monosyllable tests. The English and Dutch phoneme scores were pooled. In figure 2, the phoneme scores at follow-up ‘0’ were obtained directly after the sound processor had first been fitted. The figure shows that performance varied widely. Scores improved most sharply during the first 9 months, after which they seemed to stabilize. The patients were grouped according to their performance after 9 months of implant use. As mentioned above, the 25th percentile of the reference group (phoneme score of 40%) was used as the criterion for inclusion in either the ‘good performance subgroup’ (group 1) or the ‘poor performing subgroup’ (group 2). Patients with a phoneme score of higher than 40% ($n = 24$) were categorized as good performers; patients with a score of lower than 40% ($n = 17$), which is the 25th percentile in the adult postlingually deaf CI population, were categorized as poor performers (group 2).

Group 1 and 2 did not differ in age at onset of hearing loss (Mann-Whitney $t$ test, $p = 0.32$), duration of progressive hearing loss ($p = 0.87$), age at onset of deafness ($p = 0.46$) or duration of deafness ($p = 0.65$). The distributions of recent and previous generation devices and of NVA and AB monosyllable tests in the 2 groups were not significantly different (Fisher’s exact test, $p = 0.17$). Analysis of the extent of otosclerosis on the CT scan between the good and poor performers revealed a tendency towards a lower proportion of patients with type 1 otosclerosis and a higher proportion of patients with type 3 otosclerosis in the group of poor performers (fig. 3a), although signifi-
cance was not reached (Fisher’s exact test: type 1, p = 0.26; type 2, p = 0.76; type 3, p = 0.50). Figure 3a also shows that partial insertion of the electrode array, FNS and inactive electrodes (i.e. switched off during rehabilitation) were less common in the group of good performers, but again statistical significance was not reached in these groups (Fisher’s exact test, p = 0.14, 0.10 and 0.06, respectively).

BKB Sentence Test Scores
BKB sentence test scores had been obtained from 28 English-speaking patients. Data from the 2 English centres were combined, because there were no differences in patient characteristics, pre-operative residual hearing, extent of otosclerosis on the CT scan, device-type-related factors and test procedures. Figure 4 shows the scores on the BKB sentences test: performance varied widely. The patients were grouped according to their performance after 9 months of implant use, using the 25th percentile of the BKB data of the reference group as a criterion, which was 47% correct. The criterion for inclusion in group 1 was a score of higher than 47% (n = 15); individuals with a score of lower than 47% were placed in group 2 (n = 13). The good and poor performers did not differ in age at onset of hearing loss (Mann-Whitney t test, p = 0.78), age at onset of deafness (Mann-Whitney t test, p = 0.66), duration of progressive hearing loss (Mann-Whitney t test, p = 0.55) nor duration of deafness (Mann-Whitney t test, p = 0.68).

Figure 3b shows the extent of otosclerosis (type 1, 2 or 3) on the pre-operative CT scan per group. A larger proportion of patients in the group of poor performers (group 2) had type 3 otosclerosis, i.e. severe retrofenestral otosclerosis with an unrecognizable otic capsule (Fisher’s exact test, p = 0.07). Type 1 otosclerosis, i.e. solely fenestral involvement, was more frequent in group 1 (Fisher’s exact
test, p = 0.65). However, these differences were not significant.

Figure 3b further shows the percentages of patients with full and partial insertion per performance group. There were trends towards more patients with partial insertion among the poor performers (Fisher’s exact test, p = 0.37), a lower percentage of patients with FNS in the group of good performers (Fisher’s exact test, p = 0.25) and a lower percentage of patients who had one or more inactive electrodes (i.e. that had been switched off at some point during rehabilitation to control FNS or other types of discomfort) in the group of good performers (Fisher’s exact test, p = 0.25). The poor performers contained a larger proportion of patients with relatively older-generation devices than the good performers (Fisher’s exact test, p = 0.07).

Discussion

Results were available on 53 CI users with otosclerosis as the cause of deafness at 4 different CI centres. Similarities in demographic data, pre-operative CT scans and audiological data meant that the patients from the 4 different centres could be considered as one group. The pre-operative audiological data reflected that all 4 CI teams had employed conservative inclusion criteria. Nevertheless, significantly more patients at the Utrecht centre had undergone stapes surgery than the patients at the other centres. This was not considered to have had any important influence on later performance with a CI.

The phoneme scores obtained from the English and Dutch patients were pooled although different tests had been used (AB and NVA monosyllable tests, respectively). Pooling was considered feasible, because the AB monosyllable test and the NVA monosyllable test have the same set-up, scoring procedure and level of presentation of the CVC words. Moreover, analysis of the distribution of NVA and AB monosyllable test results showed that these were equally distributed in the 2 groups (Fisher’s exact test; p = 0.54). By pooling these data, the statistical power increased significantly.

A wide variation in the speech perception scores was observed between our subgroups of good and poor performers. Categorizing patients in good and poor performers is helpful as an orientation for the further rehabilitation process [Krueger et al., 2008]. In the present study, no differences were found in demographic factors between the poor and good performers: the clinical presentation of the disease (rapid or slowly progressive) did not influence performance with a CI. Also, there were no differences in age at onset or duration of deafness be-
The differences between the poor and good performers comprised factors directly related to the disease (extent of otosclerosis on the CT scan, non-auditory sensations such as FNS) and factors indirectly related to the disease (fewer electrodes due to partial insertion or deactivation of electrodes). A reduced number of active electrodes due to partial insertion of the electrode array has already been known to negatively influence patients’ performance [Hartmann et al., 1995; Rotteveel et al., 2005]. When during rehabilitation, FNS cannot be managed by lowering stimulation levels, usually the causative electrodes are deactivated, thus reducing the number of active electrodes. FNS in otosclerosis is thought to result from lowering of the electrical impedance of the bone by the disease, or by a reduced distance from the electrode to the facial nerve by loss of bone and cavity formation [Mens et al., 1994; Weber et al., 1995; Ramsden et al., 1997; Bigelow et al., 1998]. However, Frijns et al. [2003] showed that in patients with full insertion of the electrode array the optimal number of electrodes for phoneme perception in noise had to be selected for each patient individually and was smaller than the maximum number available. In the present study, obvious trends were seen: compared to the poor performers, the good performers had less severe otosclerosis on the CT scan, the majority had full electrode array insertion, very few had FNS and very few had deactivated electrodes. Similar patterns were seen in the phoneme scores and BKB sentence scores in the poor and good performers. Although many of the differences did not reach statistical significance, the similarities between the scores on these 2 speech recognition tests indicate that these differences are of importance.

Conclusion

A previous paper showed that cochlear implant surgery in patients with otosclerosis can be challenging, with a relatively high number of partial insertions and misplacements of the electrode array demanding revision surgery. A very high percentage of patients were confronted with FNS, mainly caused by the more distal electrodes on the array.

The present study showed wide variation in speech perception scores in patients with otosclerosis. Pooling of the data for statistical analysis was found feasible after analysis of the different test procedures. Several factors were identified to influence patients’ performance. Good performance in patients with otosclerosis was related to less severe otosclerosis on the CT scan, full electrode array insertion, little or no FNS and little or no need to switch off electrodes. One indirect disease-related factor, the number of active electrodes, appeared to be the most important determinant of the outcome. Knowledge of these factors is of clinical importance during the patient selection period prior to implantation: in patients with this specific disease affecting the otic capsule, special emphasis can be put on the assessment of the cochlear structure. During counselling, the probability of a successful rehabilitation with the CI may be estimated by the CT scan obtained and by the acknowledgement of a potential partial electrode array insertion. Although exact predictions about the benefit remain uncertain and unwise, this knowledge may be of value for the patient with otosclerosis in order to develop realistic expectations.

Acknowledgements

The authors would like to thank David W. Proops and Richard T. Ramsden for putting the different CI centre databases at our disposal and for their helpful remarks on the 2 papers about this patient group with otosclerosis. We would also like to thank Frank B.M. Joosten, neuroradiologist, for reassessing a subset of CT scans to establish the test-retest reliability of the categorization into 3 types of otosclerosis.

References


