Evolution of Otosclerosis to Cochlear Implantation


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KEYWORDS
Otosclerosis; Cochlear implant; Computerised tomography; Facial nerve

Abstract
Introduction: Otosclerosis is an osteodystrophy of the labyrinthine capsule producing conductive hearing loss. If the process invades the cochlea, a sensorineural hearing loss usually takes place. The cochlear implant is a good alternative in these patients.
Objective: To ascertain the behaviour of cochlear implantation (CI) in otosclerosis.
Material and methods: We reviewed a database of 250 patients who underwent CI, performing a retrospective study of 13 patients with clinical, audiological and/or imaging findings of bilateral otosclerosis. The 26 ears were studied as to their natural history, previous surgeries, evolution to profound hearing loss, computed tomography (CT) images, complications and functional results.
Results: Of the cases studied, 46% were female and 54% were men, with a mean age of 26 years at the onset of conductive hearing loss. Stapes surgery was performed in 19 ears (73%), with a mean patient age of 29 years, and 53% of them underwent CI. CT results showed that there were signs of different degrees of radiological affectation in 54% of the ears. A total of 3 complications took place (23%): implant failure, overstimulation of the facial nerve (FN) and bilateral tinnitus were found. One year after implantation, the average percentages of correct 2-syllable words were 80% and 85% in open sentences.
Conclusions: Patients having profound bilateral sensorineural hearing loss secondary to otosclerosis obtain great benefit from CI.
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Palabras clave
Otosclerosis; Implante coclear; Tomografía computarizada; Nervio facial

Evolución de la otosclerosis hacia la implantación coclear

Resumen

Introducción: La otosclerosis es una osteodistrofia de la cápsula laberíntica que produce hipoacusia de conducción. Si el proceso invade la cóclea, aparece una hipoacusia neurosensorial, siendo el implante coclear una buena alternativa en estos pacientes.

Objetivo: Conocer el comportamiento de la otosclerosis en la implantación coclear.

Material y métodos: Se revisó una base de datos de 250 pacientes intervenidos de implante coclear realizando un estudio retrospectivo sobre 13 pacientes con criterios clínicos, audiológicos y/o de imagen de otosclerosis bilateral. Se estudian los 26 oídos analizando la historia natural, cirugías previas, evolución hacia hipoacusia profunda, hallazgos en imagen mediante tomografía computarizada, complicaciones y resultados funcionales.

Resultados: El 46% fueron mujeres y el 54% hombres con una edad media de 26 años al inicio de la hipoacusia conductiva. La cirugía del estribo fue llevada a cabo en 19 oídos (73%) a una edad media de 29 años y de estos en el 53% se realizó posteriormente un implante coclear. Los resultados de la tomografía computarizada muestran que en el 54% de los oídos existen signos de afectación radiológica en diferente grado. Se registraron un total de 3 complicaciones (23%): un fallo de implante coclear, una estimulación del nervio facial y un acúfenos bilateral. Al año de la implantación obtuemos un porcentaje promedio de aciertos en bisiilabos del 80% y del 85% en frases en contexto abierto.

Conclusiones: Pacientes que presentan una hipoacusia neurosensorial profunda bilateral secundaria a una otosclerosis se benefician extraordinariamente de la implantación coclear.

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Introduction

Otosclerosis is an aberrant process of the labyrinthine capsule which appears as conductive or mixed hearing loss. The bone increases in size as it undergoes a maturation process (otospongiosis–otosclerosis phase)^1^ and may invade the round window or go deeper into the labyrinth, resulting in atrophy of the spiral ligament and stria vascularis. With the evolution of the disease, detritus caused by the unusual osteoclastic/osteoblastic activity may alter the normal composition of perilymphatic fluids and cause perceptual hearing loss due to the appearance of otosclerotic foci in the cochlea or inner ear canal, by inflammatory reactions mediated by immune complexes in the perilymph or by the sum of several of these factors.\(^2\,^3\) Therefore, sometimes the disease progresses gradually with worsening hearing loss, starting at low frequencies and subsequently increasing to high frequencies of the bone curve (mixed hearing loss). Eventually, low frequencies will be affected in more advanced stages,\(^4\) culminating in deep hearing loss. Long-term monitoring studies estimate that approximately 10% of ears with otosclerosis and conductive hearing loss develop sensorineural hearing loss.\(^5\) Shea et al.\(^6\) estimated that 1.6% of patients with otosclerosis will evolve to deep hearing loss. Despite this loss, there are no obstacles for the use of prostheses and, except in cases where good results are obtained, the possibility of including patients within CI programmes should be considered.

Implantation in these patients involves surgical and programming challenges that must be taken into account. There is a possibility that electrode insertion within the cochlea results partial or incomplete hearing and overstimulates the FN. The first problem is mainly due to ossification and cochlear hypodensity, which lead to incorrect placement of the electrode guide.\(^7\,^8\) The current design of electrodes facilitates the development of the technique and the use of all channels. The second problem is due to the change in bone electrical impedance and the reduction in the distance from the electrode to the FN caused by the possible formation of cavities.\(^9\)

Imaging tests are of great help in order to know the status of the ear prior to CI, because they help to confirm it as the disease evolves.\(^10\) High-resolution CT scans can analyse dysplastic foci in the labyrinth and plaque\(^2\,^11\) based on studies of the pre-stapedial region, where the fissura ante fenestram is located.\(^12\) This is the best method available for evaluating the windows and otic capsule, confirming the diagnosis in approximately 95% of cases.\(^13\,^15\)

In order to present our experience with this group of patients, we have conducted a study describing the clinical features, CT results, surgical findings, complications and functional results obtained.

Material and Method

A retrospective, descriptive and comparative study was conducted by the Otolaryngology service, with the aim of analysing a sample of patients with a prior diagnosis of otosclerosis included in the CI programme. The database was extracted only from our centre and we selected patients with clinical and/or radiological criteria of bilateral otosclerosis who had been implanted by the same surgeon. Therefore, we included patients with no prior history of stapedial surgery, but with otosclerotic foci described in the CT images requested during our preimplantation protocol, as well as patients without CT findings, but with a history of stapedial surgery. We excluded those who presented unilateral criteria in the course of their disease, with a natural evolution of 1 of the 2 ears, without conductive hearing loss or images suggestive of otosclerosis on CT scans.
All patients in this study were comprehensively evaluated through a complete medical record which reflected their audiological history and the existence of events therein. All data concerning the natural history of each ear are presented in Table 1. As shown, we have attempted to analyse the evolution of each ear and quantify the years during which sensorineural hearing loss developed until patients became candidates for ear implantation. Although many of these measurements are subjective, they help to calculate the duration of hearing loss progression and the duration of profound hearing loss.

Imaging techniques used were part of the standard preimplantation protocol employed at most CI centres. Temporal bone CT scan and magnetic resonance imaging (MRI) were requested for all candidate patients. The results were examined by the head and neck specialist radiologist, who described the CT findings of the otosclerotic process, both fenestral (narrow or elongated window, dense plaque) and retrofenestral (double cochlear ring effect, narrow basal turn). Active foci appeared as a low-density mesh of trabecular bone due to the numerous cells and vessels present, whereas mature foci appeared as areas of increased density due to their being practically avascular and acellular. The description was based on the histological classification of Lindsay,16 although we agree with the subsequent proposal by Rotteveel et al.17 and have used it in the analysis, since we believe that it reflects CT findings more accurately (Table 2).

Regarding the surgical technique, we used the same as in routine CIs, with the exception of performing a wider than usual posterior tympanotomy, in order to obtain maximum visibility, especially of the round window. Previously, we thinned the posterior wall of the duct until we obtained an involuntary extraction of the electrode guide after reaching the posterior wall of the ear canal.

Following CI surgery, we studied the short-term and long-term complications of each patient at the same time as we started programming. Regarding functional results, we reflected those obtained after 1, 3, 6, 9, and 12 months, using the percentage of correct responses in syllables and open-context sentences. These results were compared with those obtained in the rest of the implanted adult population.

We used measures of central tendency for the descriptive statistical analysis, which was performed with the software package SPSS version 15.0, using non-parametric tests in comparative studies (Mann–Whitney U test and Chi-Square test).

### Results

Of the 250 cochlear implants performed since 1998, 13 patients presented clinical and/or audiological and/or imaging criteria of bilateral otosclerosis and were included as candidates for the study. Therefore, the total number of analysed ears was 26.

As for the descriptive analysis, we observed that 46% (n=6) of patients were female and 54% (n=7) were male, with a mean age of 64 years (range: 47–76 years) at the time of study, and a mean age of 26 years (range: 10–49 years) at the beginning of conductive hearing loss. A total of 5 patients (38%) presented a positive family history (family members with early onset of hearing loss and/or history of stapedial surgery). Within this group, positive CT findings were identified in 47%. The CT results of all 13 patients are shown in Table 2. Of the 26 ears studied, 46% (12 of 26) showed no involvement, while 54% (14 of 26) presented positive findings. Only 2 (8%) had fenestral otosclerotic lesions (type 1) (Fig. 1A), 7 (27%) had retrofenestral lesions with or without fenestral involvement (type 2) (Fig. 1B and C). Of these 7 ears, the double ring sign was identified in 5 (type 2a), a narrow basal turn was observed in only 1 ear (type 2b), and the presence of both signs was also observed in only 1 ear (type 2c). Finally, 5 ears (19%) presented an abnormal structure of the otic capsule, which was unrecognisable due to advanced otosclerosis (type 3) (Fig. 1D). In some cases it is very difficult to assess involvement due to extensive cochlear destructuring. This situation was not found in our sample. The severity of radiographic involvement was asymmetrical in only 1 patient: 1 ear showed signs of retrofenestral otosclerosis and in the other the lesions were very advanced and affected the entire cochlea. In this patient

### Data Collected in the Clinical History.

| Age of onset of conductive hearing loss: | Years |
| Stapedial surgery: | No | Yes: | Years |
| Labyrinthisation: | No | Yes: | Years |
| Time without requiring prosthetic aid after diagnosis: | No | Yes: | Years |
| Prosthetic aid: | No | Yes: | Years |
| Age of profound sensorineural hearing loss (inefficacy of aids): | Years |
| Years with total deafness until implantation: | No | Yes: | Date |
| Cochlear implant: | No | Yes: | Date |

### Classification of Otic Capsule Lesions Described in Computerised Tomography.

| Type 1 | No involvement | 12 (46%) |
|        | Only fenestral involvement | 2 (8%) |
|        | (spongiotic or sclerotic lesions) | |
| Type 2 | Retrofenestral lesions (double ring effect, narrow basal turn or both) with or without mechanical lesions | |
| Type 2a | Double ring effect | 5 (19%) |
| Type 2b | Narrow basal turn | 1 (4%) |
| Type 2c | 2a+2b | 1 (4%) |
| Type 3 | Severe retrofenestral lesions with loss of the normal architecture of the cochlea | 5 (19%) |

Source: Güneri et al.1

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**Table 1** Data Collected in the Clinical History.

**Table 2** Classification of Otic Capsule Lesions Described in Computerised Tomography.
with radiological asymmetry we decided to implant the least affected ear.

Stapedial surgery was performed in 19 ears (73%) at a mean age of 29 years (range: 18–50 years). Within these cases, cochlear implant was performed subsequently in 53%. In 6 operated ears there was an immediate process of labyrinthisation or development of postoperative endocochlear perceptive hearing loss after stapedial surgery and none of these underwent subsequent surgery for CI. Therefore, we considered these patients as having a cochlear otosclerosis, either due to the CT result or to the evolution of hearing by bone conduction. Only 1 implanted ear (Table 3) showed only fenestral radiological signs, although it did not show bone conduction hearing.

Of the 13 implanted patients, none was suggested for double CI. Neither did any of them undergo subsequent review surgery; only 1 ear had to undergo extraction and implantation of a new device due to component failure. We used conventional multichannel implant systems (2 CI, Advanced Bionics; 11 Combi40+, Medel) in all patients, although some new compressed or forked devices were available in the operating room in case of need.

We reviewed the 13 reports and surgical notes elaborated by the same otological team, in order to study the surgical findings and possible intraoperative complications. In some of these cases the inspection of the middle ear revealed abnormalities, but they did not complicate the placement of cochlear implants. However, no ossified round windows were found. The stapes prostheses were present and without displacement in most ears, except for 1 case in which it was dislocated. Once cochleostomy was performed it was possible to insert all the electrodes in the scala tympani, with no obliteration being found in any of them, including the 2 implanted ears classified as suffering cochlear disintegration according to CT. In these cases, the MRI reported good cochlear permeability. Neither were there any cases of partial insertion.

Few complications occurred during the surgical procedure and after surgery. There were a total of 3 complications (23%): 1 cochlear implant failure, 1 overstimulation of the FN and 1 bilateral tinnitus. The case of cochlear implant failure was caused by involuntary extraction of the electrode array after removal of an ear wax plug. This guide had been in contact with the posterior wall of the ear canal. It was reimplanted without further problems. During programming, 1 patient developed an overstimulation of the FN (7.69%). This occurred during the third month after surgery and led to the cancellation of channels 5, 6, 7, 1 and 2 (12-electrode guide), respectively. However, despite the cancellation of 5 channels, the functional outcome of this patient was acceptable (70% correct disyllables and open-context phrases). In addition, the CT results showed that she was classified within the group of patients who presented no pathological findings. Although it could be thought that those ears classified as type 3 should be more related to FN stimulations, this was not so. The management of overstimulation consists in reducing the stimuli levels or eventually disconnecting the causative electrode.

Regarding the comparative analysis, if we consider the implanted and non-implanted ears separately we can observe that there were no statistically significant differences regarding the start of conductive hearing loss. The start in implanted ears ranged from 10 to 49 years, with a mean age of 27 years. This value was similar in the nonimplanted ears but with a greater age range, from 12 to 49 years. Greater variation between implanted and nonimplanted ears was found in the progression of hearing loss towards total deafness. This progression included the years of need for prosthetic support and the years where this

![Figure 1](http://www.elsevier.es) Pathological findings obtained in axial sections of temporal bone in patients with otosclerosis candidates for cochlear implantation. (A) Fenestral focus: type 1. (B) Double ring: type 2a. (C) Narrow basal turn: type 2b. (D) Severe retrofenestral cochlear involvement: type 3.

<table>
<thead>
<tr>
<th>Type</th>
<th>Ear With Cochlear Implant</th>
<th>Ear Without Cochlear Implant</th>
</tr>
</thead>
<tbody>
<tr>
<td>No Involvement</td>
<td>6 (46%)</td>
<td>6 (46%)</td>
</tr>
<tr>
<td>Type 1</td>
<td>1 (7%)</td>
<td>1 (7%)</td>
</tr>
<tr>
<td>Type 2</td>
<td>4 (31%)</td>
<td>3 (23%)</td>
</tr>
<tr>
<td>Type 2a</td>
<td>3 (23%)</td>
<td>2 (15%)</td>
</tr>
<tr>
<td>Type 2b</td>
<td>0</td>
<td>1 (7%)</td>
</tr>
<tr>
<td>Type 2c</td>
<td>1 (7%)</td>
<td>0</td>
</tr>
<tr>
<td>Type 3</td>
<td>2 (15%)</td>
<td>3 (31%)</td>
</tr>
</tbody>
</table>

Source: Güneri et al.¹
no longer obtained any benefits until the confirmation of total deafness. Thus, the duration of this progression had a mean value of 13 years in the implanted ears and 8 years in the non-implanted, with this difference being statistically significant.

We found no statistically significant differences when comparing the different types of radiological findings (types 1, 2 and 3) according to implantation or not. This could be due to the small number of patients (Table 3). If we look at those patients with asymmetrical radiographic involvement, we can see that the least affected ear was implanted, hence we find less type 3 cases among implanted patients. If we compare those ears with greater radiographic extension of otosclerosis towards the retrocochlear area (type 3) with ears showing no radiographic involvement, we see that 40% of ears in the first group developed profound sensorineural hearing loss immediately after stapedial surgery, whereas this only occurred in 25% of the unaffected ears. Therefore, we can say that, in our study, greater radiographic involvement took place among those who developed a rapidly progressive sensorineural hearing loss.

The mean functional results obtained at 1, 3, 6, 9, and 12 months are reflected in Table 4. The mean percentage of correct disyllables at 12 months was 80±10 (60–90), whereas for open phrases it was 85±12 (60–98). As we can see, the results were very satisfactory for this population; the percentages of correct disyllables and open sentences were similar to those obtained in the rest of the implanted adult population. We found no statistically significant differences when we analysed the hearing results by comparing the percentage of correct disyllables and open phrases in terms of the presence or absence of positive radiographic findings on CT. Neither did we find significant differences (P>.05) when we compared only those ears classified as type 3 with the rest of the ears.

**Table 4** Functional Results.

<table>
<thead>
<tr>
<th>Time</th>
<th>Percentage of Correct Disyllables</th>
<th>Percentage of Correct Open Phrases</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 month</td>
<td>11%</td>
<td>6%</td>
</tr>
<tr>
<td>3 months</td>
<td>25%</td>
<td>20%</td>
</tr>
<tr>
<td>6 months</td>
<td>40%</td>
<td>40%</td>
</tr>
<tr>
<td>9 months</td>
<td>60%</td>
<td>65%</td>
</tr>
<tr>
<td>12 months</td>
<td>80%</td>
<td>85%</td>
</tr>
</tbody>
</table>

Discussion

We know that the classical, clinical, stapedial-vestibular form of otosclerosis has a nonlinear progressive conductive, mixed evolution with severe cochlear impairment towards the end. One of the most feared complications of otosclerosis surgery is the onset of endocochlear hearing loss due to various causes (granuloma, penetration of the prosthesis into the inner ear, perilymph fistula). These patients present signs such as vertigo and hearing loss in the immediate postoperative period or after some time. Once this happens, they depend on the evolution of the other ear towards the stabilisation of hearing, with or without a hearing aid, or towards the development of profound hearing loss. Regardless of whether this complication appears or not, the aberrant maturation process of the otic capsule continues its evolution. Thus, out of the 6 ears which developed postoperative perceptive endocochlear hearing loss, 3 showed radiographic evidence of cochlear involvement in the preimplantation CT scan. In 2 of these 3 cases, involvement was very severe (type 3). In these cases, we attempted to implant the ear with the least auditory deprivation time, that is, the one which had retained hearing for the longest time, prior to the development of total deafness.

Despite our study being carried out in a single hospital, we were able to collect data on a significant number of patients with bilateral otosclerosis and requiring a cochlear implant. Out of the adults who were implanted at our institution until the end of 2010 (n=250), 5.2% presented bilateral otosclerosis as the cause of bilateral, profound, sensorineural hearing loss. Thus, we should consider otosclerosis not as a frequent but probable cause of indication for CI, despite being initially considered as a contraindication for this surgery.

Regarding the inclusion of patients in our study, all were selected by their previous history of stapedial surgery and the findings described in CT scans. However, other studies also included patients who presented findings suggestive of plaque fixation during implantation surgery.

As explained in the introduction section, the imaging diagnosis of otosclerosis was mainly based on findings observed through high-resolution CT. The study was based on the analysis of the pre-stapedial region where the *fissula ante fenestram* is located. Occasionally, we discovered foci on the asymptomatic side, showing that not all radiological locations were always accompanied by clinical expression. Cochlear otosclerosis is less common and it is rare for it to be observed without fenestral involvement.

In our study, not all ears with clinical disease expression presented radiological findings, even after the disease reached severe audiometric involvement.

The incidence of FN stimulation among the implanted population is around 14.6%, although this varies in the literature. According to different studies, this stimulation is much more common in patients with otosclerosis. However, we only observed this complication in 7.69% of cases. This observation could be explained by the low numbers of patients in our study and also by the fact that most of their cochleae had not suffered complete destructuring of their architecture. Some authors report that a high percentage of patients experienced FN stimulation mainly at the expense of the distal electrodes. Other studies, such as that by Rama-López et al. on 30 patients, did not detect any abnormalities in FN stimulation. Another possible complication is the presence of tinnitus. In our study, 1 patient developed a transient episode of tinnitus after programming which did not require any action. However, the study by Sainz et al. describes 2 patients (13%) in whom the onset of sudden and progressive intense tinnitus after 2 years required the performance of a contralateral cochlear implant and removal of the previous device.

Occasionally, CI surgery can find windows which are apparently longer due to otospongiosis lesions located in the margins, or windows which are narrower and/or closed due to mature foci of otosclerosis. A typical sign of retrocochlear otosclerosis is the presence of the ‘‘double ring’’ or ‘‘halo’’
effect. This ring represents the confluence of pericochlear foci around the cochlear lumen. These were observed in many of our patients.

In some studies, ears which suffer an immediate labyrinthisation process, that is, those developing sensorineural hearing loss after stapedial surgery, are not implanted. Our study did not include implanted ears with immediate labyrinthisation after stapedial surgery.

While there is diversity in the choice of electrode guide, particularly in cases of partially ossified cochlea, this has not been discussed or compared in our study. CT and MRI scans and the studies of ears prior to implantation did not make us foresee the need for an electrode guide designed for that purpose. We are aware of the existence of electrodes that shorten the gap between active plaques (compressed) and others which double it (forked). Our current design facilitates the technique and enables the insertion and use of all channels. None of the histories and evolutions of hearing loss in our patients made us anticipate the existence of patent cochlea, so we used the same devices as in the adult population candidates for implantation. Therefore, despite the recommendations for the use of special electrode guides to prevent partial insertions, we have not had any problems using conventional devices, perhaps due to the small sample size. However, we did find some series, like that of Cohen and Waltzman, in which partial insertion of the implant took place in 7% of cases (about 110), with the number of active electrodes varying between 10 and 18 due to semilibrated cochlea with apparently normal radiological images. The only exception is that it included patients with ossified cochlea due to meningitis. The study by Marshall et al. identified the scala tympani in all 30 patients studied, requiring greater milling in 3 of them. There was only 1 case in which the surgeon was unsure of having inserted the electrode into the scala tympani, otherwise there were no problems with insertion. We can consider that the excellent results obtained in these cases are linked to the number of nerve fibres covered by the bony tissue. In other words, those that remained undamaged by the pathology were made use of. Furthermore, the different electrical impedance offered by bone also contributed. The prognosis regarding speech discrimination depends on the increase in electrical resistance and the functional status of the spiral ganglion.

As for the dilemma of which ear to implant, the one with longer or shorter duration of deafness, the study by Matterson et al. concluded that there were no long-term advantages or disadvantages in speech perception if the ear with longer duration of sensorineural hearing loss was implanted, but concluded that it should have preference.

Although some studies on ossified cochlea, such as that by Balkany et al., have presented poor results regarding normal cochlea with respect to ours, this could be because in our case patients presented cochlear ossification characteristics, but also good permeability. Also, our study only included patients diagnosed with otosclerosis and excluded patients with ossified cochlea due to meningitis.

The functional results were similar to those of implanted adults, without significant differences. We used the percentage of success in disyllables and open-context phrases, which is the trend observed in most of the literature.

Conclusions

Thanks to technological advances, the experience accumulated by otological surgeons and progress in imaging techniques that help to plan surgery, indications for cochlear implants have expanded their range to more complex pathologies, with cochlear otosclerosis being one of them. Therefore, patients with bilateral, profound, sensorineural hearing loss secondary to otosclerosis benefit extraordinarily from CI, despite possible cochlear ossification.

Conflict of Interests

The authors have no conflicts of interest to declare.

References

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