# **Original Research Article**

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# Management of advanced otosclerosis: experience in a tertiary care centre

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## **ABSTRACT**

**Background:** Advanced otosclerosis affects approximately 10% of patients with otosclerosis. Ossification of the cochlea increases with the course of the disease and may cause sensori-neural or mixed hearing loss. Hearing aids, stapedotomy and cochlear implants are management options for hearing loss associated with advanced otosclerosis.

**Methods:** A retrospective study of 153 patients with advanced otosclerosis was done in a tertiary ENT centre. 110 patients with advanced otosclerosis underwent stapedotomy and 43 patients with advanced otosclerosis underwent cochlear implantation (CI) from 1997 till date. Exclusion criteria included patients with profound hearing loss from causes other than otosclerosis. The aim was to study the indications, selection criteria, as well as surgical issues of stapedotomy and cochlear implantation in patients with advanced otosclerosis.

**Results:** Stapedotomy followed by use of hearing aid was found to give good outcomes in advanced otosclerosis. Patients with advanced otosclerosis who had poor pre-operative speech discrimination underwent CI. Complete insertion was possible in 39 patients and partial insertion was done in 4 patients. The outcomes of cochlear implantation were found to be satisfactory. Facial nerve stimulation was seen in 5 patients who underwent CI; however this was successfully managed by reprogramming or switching off the concerned electrodes.

**Conclusions:** In our experience, stapedotomy or CI in advanced otosclerosis has proven successful, with a low complication rate. The selection criteria for stapedotomy versus CI have to be stringent for optimal outcomes.

Keywords: Advanced otosclerosis, Stapedotomy, Cochlear implantation

#### INTRODUCTION

Advanced otosclerosis is known to affect approximately 10% of patients with otosclerosis. Ossification of the cochlea may increase with the course of the disease resulting in sensori-neural or mixed hearing loss. Hearing aids, stapedotomy and cochlear implants are management options in these patients. In advanced otosclerosis with mixed hearing loss, stapedotomy can help in correction of

the conductive component with acceptable hearing levels. In patients with advanced otosclerosis, effective and safe hearing rehabilitation can be accomplished with cochlear implantation (CI). However, extensive cochlear ossification can impede complete electrode insertion. Facial nerve stimulation can pose programming challenges. Selection criteria should be individualized and based on a thorough evaluation.

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#### **METHODS**

The study included 153 adult patients with advanced otosclerosis who were managed in Madras ENT Research Foundation, Chennai a tertiary care ENT center over a 20 year period (1997-2017). The aim was to study the indications, selection criteria, as well as surgical issues of stapedotomy and cochlear implantation in these patients. There were seventy one men and eighty two women in this study and the age ranged from 25-62 years (average age was 43 years). All patients were evaluated by audiometry, speech tests and imaging. Exclusion criteria included patients with profound hearing loss due to causes other than otosclerosis. One hundred and ten patients with advanced otosclerosis underwent stapedotomy. Forty three patients underwent cochlear implantation (Figure 1-4).

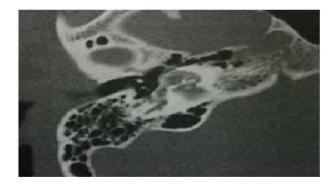


Figure 1: Pre-op scan showing cochlear otosclerosis.



Figure 2: Cochlear ossification being removed.



Figure 3: Complete insertion of electrodes into the scala tympani.



Figure 4: CI insertion into the second turn of the cochlea.

#### **RESULTS**

One hundred and ten patients with advanced otosclerosis underwent stapedotomy. In 104 patients the post-operative hearing reached an aidable level (95%) (Figure 5). The remaining 6 patients who did not benefit from stapedotomy and hearing aid usage were counseled for cochlear implantation. Of these, four patients underwent cochlear implantation and achieved significant hearing improvement.

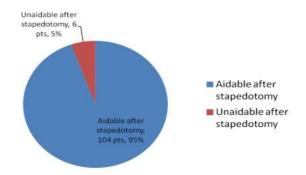


Figure 5: Post-operative outcome after stapedotomy.

Forty three patients underwent primary cochlear implantation from 1997 till date. In 32 patients, there was no cochlear ossification noted intra-operatively (74%) and the electrode insertion was uneventful. In 11 patients varying degrees of ossification of the cochlea was encountered (26%) (Figure 6).

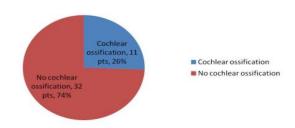


Figure 6: Incidence of intra-operative cochlear ossification in otosclerosis.

Of the 43 patients who underwent CI, complete electrode insertion was possible in thirty nine patients (91%) and in 4 patients, partial insertion was possible (9%). In one patient the electrode was inserted in the second turn (2%). None of the patients had a scala vestibuli insertion. Occlusion of the round window niche by otospongiotic foci was not seen in any of the patients. Misplacement of the electrode array did not occur in any patient and this was confirmed by intra-operative telemetry and a post-operative X-ray. CSF gusher was not encountered intra-operatively in any patient.

The average pre-operative pure tone audiometry level in our patients was 90 dBHL. The categories of auditory perception (CAP) score was used to assess the benefit obtained from cochlear implantation. The average pre-operative CAP score was 1. The mean post-operative CAP score value was 2 (range 0-3) at 6 months and improved to 4 (range 2-6) at 12 months (p $\leq$ 0.05) which was statistically significant and confirmed that CI is beneficial in the management of advanced otosclerosis.

Of the 43 patients who underwent CI, post-operative facial nerve stimulation was seen in 5 patients (12%). In two patients, the concerned electrodes were switched-off. In two patients, programming was done with a lower charge unit & pulse width to avoid excess current dissipation onto the facial nerve. In one patient, one electrode was switched-off & adjacent electrodes were set with a lower charge unit to avoid undue facial nerve stimulation from that region of the cochlea (Figure 7-9).

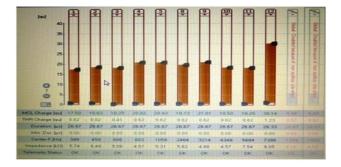


Figure 7: Electrodes 6 & 7 have been switched-off due to facial nerve stimulation.

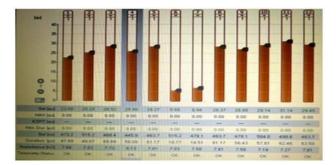


Figure 8: Electrodes 6 and 7 are programmed with a lower charge unit and pulse width to avoid excess current dissipation onto the Facial Nerve.



Figure 9: Electrode 7 has been switched-off and adjacent electrodes set with lower charge Unit to avoid facial nerve stimulation.

In our series the average follow up period of cochlear implantees was 3 years and 4 months. The outcomes after cochlear implantation were found to be satisfactory as 15 patients (35%) achieved a CAP score of 4 one year post CI, 8 patients (19%) achieved a CAP score of 5 and 8 patients (19%) achieved a CAP score of 6 one year after implantation.

## **DISCUSSION**

The indications for cochlear implantation have expanded in recent years. Patients with profound hearing loss due to advanced otosclerosis are increasingly being considered as CI candidates. Sensori-neural hearing loss in advanced otosclerosis occurs due to proteolytic enzymes from otospongiotic foci leaking into the perilymph. Subsequent diffusion into the endolymph causes damage to hair cells. Another factor possibly contributing to SNHL is hyalinization of the spiral ligament. Evaluation of these patients includes a thorough clinical examination, including tuning fork tests. A negative Rinne test result is useful to differentiate advanced otosclerosis from sensorineural hearing loss of other causes. Pure tone and impedance audiometry, Oto Acoustic Emissions, Brain Stem Evoked Response Audiometry and imaging (CT / MRI scans) are essential.

Radiology is essential in the pre-surgical evaluation of otosclerosis to evaluate the extent of the lesion and cochlear patency. HRCT is considered to be the imaging technique of choice for the diagnosis of otosclerosis. HRCT can detect subtle otosclerotic foci in and around the cochlea, and may predict the risk of complications during surgery. The CT grading system of Rotteveel is partially based on location and on the type of lesion: solely fenestral (grade 1), retrofenestral: double ring or halo effect (grade 2A), narrowed basal turn (grade 2B) or both (grade 2C), and diffuse confluent retrofenestral involvement (grade 3). Symons and Fanning proposed a classification similar to Rotteveel, except grade 2 is based on anatomic location instead of the type of lesion: basal turn (2A), middle/apical turns (2B), both basal and middle/apical turns (2C).<sup>3</sup>

Mercus et al described an algorithm for management of advanced otosclerosis based on CT grading, speech discrimination and air-bone gap. In patients with speech discrimination (SD) scores of <30%, the most effective intervention is CI. Patients with an SD between 30% and 50% may be treated with either CI or stapedotomy. In cases of Rotteveel grade 2C or 3 otosclerosis on HRCT, CI is the better option. If the CT scan shows less cochlear involvement (Rotteveel grade 1, 2A, or 2B), the air-bone gap (ABG) will guide the surgeon to either stapedotomy or CI. If the ABG is 30 dB or more, a stapedotomy can result in improvement of hearing. If the ABG is 30 dB or less, patients should be treated with CI. Patients with an SD of 50% to 70% are candidates for stapedotomy, rehabilitation with hearing aids, or CI. Patients with limited cochlear involvement on HRCT (Rotteveel grade 1, 2A, or 2B) and an ABG of 30 dB or more should be treated with stapedotomy. When the ABG is 30 dB or less, and HRCT shows limited cochlear involvement, patients will generally benefit from hearing aids and follow-up. If, HRCT shows extensive retrofenestral otosclerosis (Rotteveel grade 2C or 3), CI should be recommended.3

Stapedotomy with subsequent hearing aid use is a useful treatment option for patients with good speech discrimination. 46%-100% hearing improvement has been reported after stapedotomy. The effectiveness with regard to the improvement of the level of speech discrimination varies around 38%–75%. <sup>4</sup> The advantages of stapedotomy for advanced otosclerosis include simple and safe procedure, reduced cost, good sound quality, and lack of any adverse effect on outcomes of future cochlear implantation. In 1996, Glasscock et al. showed that stapedectomy was effective in 60% of patients with advanced and far-advanced otosclerosis.<sup>5</sup> In our study, in 95% of patients who underwent stapedotomy, the hearing improved to an aidable level. The disadvantage with a stapedotomy is that it has no influence on the sensorineural component of the hearing loss in patients with severe mixed hearing loss. An increase of sensorineural hearing loss, after stapedotomy could result in a functionally deaf ear.

Cochlear implantation has been increasingly used in the management of severe to profound sensori-neural hearing loss in advanced otosclerosis. Ruckenstein et al. studied cochlear implanted patients with advanced otosclerosis with an excellent hearing benefit. Hearing results were found to be comparable with those for other indications.<sup>6</sup> Mean speech recognition scores of 54% to 75% have been reported after CI.<sup>6</sup> When stapedotomy does not yield a satisfactory result, the option of CI should be considered because a previous stapedotomy will not affect the outcome of CI. The improvement in hearing and the improvement in speech perception have been reported to be better after CI than after stapedotomy. However, cochlear implantation is an expensive that requires an experienced surgeon, procedure especially because otosclerosis can cause problems

during implantation. The surgery may be complicated by ossification of the cochlea which has been reported in 10 to 37% of patients.<sup>7</sup> In patients with ossification, more drilling may be required to access the lumen. Eleven patients in our study had ossification of the cochlea. Occasionally insertion in the second turn or the scala vestibuli may be required. In our series, one patient had CI insertion in the second turn. A scala vestibuli electrode insertion may be required in 2% to 25% of the cases. Several other issues may complicate CI in advanced otosclerosis. Intracochlear osteoneogenesis might result in a partial electrode insertion. This was seen in 9% of our patients. The electrode may enter a false lumen or the internal auditory canal. Furthermore, programming of the CI can be challenging because the progression of otosclerosis can cause postoperative failure of the CI. Cochlear ossification has been postulated as a cause of facial nerve stimulation (FNS) in approximately 20% of patients with advanced otosclerosis due to increase of conductivity due to bone spongiosis.4 Facial nerve stimulation is found most commonly due to electrodes in close proximity with the geniculate and labyrinthine segment of the facial nerve. Patients with a higher CT classification are significantly more likely to develop FNS. Use of modiolar hugging contour electrodes has been reported to be associated with a reduced incidence of post-operative FNS.8 In 12% of our patients, FNS was observed and was successfully managed. The management of facial nerve stimulation consists of a reduction in stimulus levels of the cranially located electrodes, totally deactivating the causative electrodes or reimplantation. Variable mode programming can also solve FNS; while the nonoffending electrodes receive normal pulses, the offending electrodes receive wider pulses. Decreased performance of CI occurs if too many electrodes must be inactivated due to FNS. Rotteveel et al described the factors contributing to the success of a CI, namely little spongiosis of the cochlea on CT scans, full insertion of the electrode array, and little or no facial electrical stimulation.4

The management of advanced otosclerosis can pose challenges to the otologist. The pre-operative air-bone gap, speech discrimination and extent of involvement on CT scans are the main criteria in deciding on the choice of treatment in these patients. Stapedotomy has shown to be a simple, cost-effective procedure for restoring hearing in patients with an air-bone gap on pure tone audiometry and good speech discrimination. Cochlear implantation has proved to be effective in hearing restoration in advanced otosclerosis, either as a primary procedure or following failure of stapedotomy. It is vital that these patients are managed by a team experienced in CI surgeries. Awareness regarding the intra-operative challenges and post-operative programming issues is essential for optimal outcomes in patients with advanced otosclerosis. In this series of patients, clear cut selection criteria were used to decide about the choice of the surgical procedure. Both stapedotomy and CI have proven successful in patients with advanced otosclerosis, with good outcomes and a low complication rate.

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Ethical approval: The study was approved by the

Institutional Ethics Committee

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