Cochlear Implantation in Advanced Otosclerosis: Pitfalls and Successes

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Abstract

Purpose of Review This review will highlight recent outcome-based evidence guiding decision making for cochlear implantation in advanced otosclerosis, related complications, and technical surgical considerations in otosclerosis and the obstructed cochlea.

Recent Findings Cochlear implantation in advanced otosclerosis results in consistent, excellent auditory outcomes with improvement in both objective speech recognition scores and subjective quality of life measures. Facial nerve stimulation may occur at higher rates in otosclerosis cochlear implant recipients. Cochlear implantation in the setting of luminal obstruction in osteosclerotic patients may be managed with altered surgical technique to achieve successful auditory improvements. Pre-operative imaging with high resolution CT or MRI may help anticipate intraoperative challenges and post-operative complications in cochlear implantation.

Summary Cochlear implantation is an established, successful treatment for profound hearing loss in advanced otosclerosis. Surgeon knowledge of outcomes, complications, and potential surgical challenges is important to appropriately counsel patients regarding auditory rehabilitation options in advanced otosclerosis.

Keywords Cochlear implantation · Cochlear ossification · Cochlear obstruction · Facial nerve stimulation · Hearing loss · Otosclerosis

Introduction

Otosclerosis is a relatively uncommon disease within the general population, but a common cause of hearing loss encountered in otologic practice. Clinically relevant otosclerosis is estimated to affect 0.3% of the population with a greater predilection for females and Caucasian backgrounds, whereas histologic evidence of otosclerosis is higher, ranging 3.4–12.75% across temporal

bone series [1]. The most common clinical presentation of this disease is conductive hearing loss, which is commonly treated with stapes surgery, hearing aids or a combination. A smaller proportion of those affected by otosclerosis, reported between 8.9 and 34%, will eventually progress to a mixed or sensorineural hearing loss (SNHL), for which one treatment option includes cochlear implantation [2, 3]. This article reviews outcomes and specific complications related to cochlear implantation in otosclerosis, highlighting important decision-making factors and changing trends in management over past decades. Additionally, histologic and radiologic findings in otosclerosis are covered in the context of complex surgical presentations, including cochlear implantation in the ossified or obstructed cochlea.

Histologic Findings Related to SNHL in Otosclerosis

Otosclerosis has been described as a bony dysplasia confined to the otic capsule that is likely multifactorial in etiology [4]. Histologic hallmarks of this disease include bony

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otosclerotic foci involving adjacent areas of bone resorption and deposition, characterized by both loose, spongiosus (otospongiosis) and dense, sclerotic lesions [5]. The most common site for this pathology, in up to 95.9% of temporal bones, is the otic capsule bone anterior to the oval window (fissula ante fenestrum), resulting in eventual stapes footplate fixation and the most common symptom of otosclerosis, conductive hearing loss [6]. The next most common sites of otosclerotic lesions include the round window and cochlear wall, two sites which are relevant to both the development of SNHL and surgical approach to cochlear implantation [6].

The development of SNHL in otosclerosis is not completely understood and has been attributed to the degree of cochlear wall endosteal involvement and resulting disruptions in the endocochlear potential and other local inflammatory processes [4, 7, 8]. Although endosteal otosclerosis has shown variable correlation with sensory hair cell loss, the spiral ganglion neuron population appears unaffected by the disease process [7, 9]. This finding suggests that it is possible for a sufficient population of spiral ganglion neurons to remain in the setting of cochlear otosclerosis to allow for successful cochlear implantation.

Macroscopic histologic changes of relevance to cochlear implantation in otosclerosis include structural changes within the otic capsule that may impede usual surgical technique, including round window obliteration, cochlear ossification, or fibrous obstruction and peri-cochlear cavitation. Round window involvement of otosclerosis occurred in 30% of temporal bones in a series by Schuknecht and Barber, with 7.6% of temporal bones showing complete ossification and obliteration of the round window [6]. Ossification within the cochlea is less common in histologic studies of otosclerotic temporal bones and is usually confined to the lower basal turn of the scala tympani [6, 10–12]. Otospongiotic cavitation lesions are also described, often in continuity with the cerebrospinal fluid (CSF) space, creating a potential false tract for cochlear implant (CI) insertion [6].

Management of Profound Hearing Loss in Otosclerosis

House and Sheehy first introduced the term of “far advanced otosclerosis” in 1961 to describe otosclerosis patients with absent bone conduction thresholds and air conduction thresholds over 85 dB HL [13]. Iurato et al. added the “very far advanced otosclerosis” terminology to include those with a “blank audiogram” or immeasurable air and bone conduction thresholds [14]. In this article, we use the term advanced otosclerosis (AO) to refer to otosclerosis patients with profound hearing loss and

speech recognition scores sufficient to meet CI candidacy. Both reports describe that a subset of these patients will experience closure of an obscured air bone gap (obscured by bone conduction audiometer limits) with stapedectomy and achieve serviceable hearing levels despite the severity of pre-operative hearing loss. Several case series have supported this finding, achieving serviceable hearing with stapedectomy in a majority (up to 81.2%) of ears with far and very far advanced otosclerosis [15–17]. Based upon these rates of successful stapedectomy in AO, several authors historically advocated for middle ear exploration with possible stapedectomy and against upfront cochlear implantation in those with profound hearing loss with history or exam findings suggesting possible otosclerosis [15, 18]. Despite the success of stapedectomy in AO, up to 20% of cochlear implantations before 1983 involved otosclerosis as the cause of hearing loss, presumably including some cases of unsuccessful stapedectomy in “normal” or advanced otosclerosis or progressive hearing loss after initial successful stapedectomy [19]. Ample retrospective case series over the past 2 decades show excellent outcomes for cochlear implantation in AO and indicate that upfront cochlear implantation, without prior stapedectomy, is also offered by some practitioners [20•, 21•, 22]. Recent reports also show good results with direct acoustic cochlear implants in AO [23]. This is an experimental device that amplifies sound directly, like a bone conduction system, to a stapes prosthesis that is placed in the oval window. Thus, three interventions for profound hearing loss in AO include stapedectomy with hearing aids, cochlear implantation, and direct acoustic cochlear implants.

Cochlear Implantation Auditory Outcomes in Otosclerosis

Nearly all retrospective case series and prospective cohort studies examining cochlear implantation in AO have shown high rates of success as measured by several objective and subjective metrics [20•, 21•, 22, 24, 25•, 26, 27•, 28, 29, 30•, 31–36]. In contemporary series where numerical speech recognition values are provided, otosclerotic CI recipients show post-operative mean disyllabic word and sentence recognition scores between 68–74.2% and 75–92.5%, respectively [21•, 25•, 27•, 29, 35, 36]. Mean absolute improvement from pre-operative baseline scores for disyllabic word and sentence recognition scores was 54.6–60.4% and 57.9–74.6%, respectively [25•, 27•]. These excellent outcomes are not surprising considering most otosclerosis CI recipients are implanted in the setting of post-lingual deafness [37]. Further, Matterson and colleagues demonstrated that duration of deafness prior to CI in otosclerosis did not affect post-operative speech recognition at 6 months or 1 year post-implantation; however, those with shorter duration of deafness

did show greater initial improvement at 3 months, suggesting this group improved speech recognition faster [34]. AO CI recipients also re-acquired the ability to use the telephone at high rates between 56 and 100% [27•, 33, 35]. When compared to CI recipients with non-otosclerotic etiologies of deafness, AO patients do at least as well on objective measures of speech recognition improvement [20•, 25•, 26, 31, 35, 36].

Cochlear implantation for AO has also demonstrated significant quality of life improvements, similar to CI recipients without otosclerosis. In the most recent and extensive quality of life study, Calvino et al. examined quality of life improvements across three different surveys examining physical, psychological, and social health status (Nijmegen Cochlear Implant Questionnaire, Glasgow Benefit Inventory) as well as sound quality in everyday life (Hearing Implant Sound Quality Index) [20•]. In this study, otosclerotic CI recipients showed significant improvements from pre-operative values across all domains, and further, there were no differences when compared to CI recipients without otosclerosis [20•].

Dumas and colleagues compared outcomes between AO CI recipients who underwent prior stapedectomy or not, finding that prior surgery was correlated with poorer monosyllabic word recognition scores, pure tone average and speech reception thresholds [21•]. This finding would support the utility of cochlear implantation as a primary treatment for AO instead of upfront stapedectomy and hearing aid usage recommended by several authors [18, 38]. However, other authors have not found any difference in auditory outcomes according to prior stapedectomy status among CI recipients in AO [25•, 36, 39•].

Cochlear luminal obstruction involving round window obliteration or scala tympani ossification are common findings at the time of cochlear implantation in AO patients [27•]. These cases may necessitate an altered surgical approach including drilling of the cochlear lumen, scala vestibuli insertions, or use of split arrays and may result in incomplete electrode insertions or increased rates of facial nerve stimulation [19, 40, 41]. Despite these problems, long-term post-implantation auditory outcomes in the setting of cochlear obstruction in AO are similar to those without an obstructed cochlear lumen [19, 25•, 27•, 34]. Notably, Semaan and colleagues did observe worse speech recognition scores in those with cochlear obstruction during short-term follow-up (less than 1 year), but differences between obstructed and non-obstructed cochleae equalized at long-term follow-up [25•].

Burmeister and colleagues reported one case of attempted hearing preservation hybrid cochlear implantation in an AO patient with pre-operative HINT and CNC scores of 36% and 25% [42]. However, this patient experienced loss of residual acoustic hearing at the time of surgery and was not able to utilize an electro-acoustic stimulation paradigm. At current, it is not known what percentage of AO patients would

potentially fulfill Hybrid CI candidacy, nor what effect AO would have on the pre-operative cochlear health and potential for hearing preservation cochlear implantation.

Decision Making for Auditory Rehabilitation in AO: Stapedectomy and Cochlear Implantation

It has been well documented that some patients with advanced otosclerosis can greatly benefit from stapedectomy and subsequent hearing aids, achieving excellent speech recognition levels, even in the setting of pre-operative profound hearing loss with undetectable bone conduction thresholds [13, 14]. Van Loon and colleagues performed a systematic review of auditory outcomes following stapedectomy in AO, finding the mean pre-operative speech recognition increased from 11 to 59% [38]. Further, 72% of patients were no longer CI candidates following stapedectomy (speech recognition >50%) and 35% of patients obtained excellent post-operative aided speech recognition scores (>80%) [38]. Consistent with other meta-analysis, no prognosticators of success were found in this study, including degree of otosclerosis on HRCT or pre-operative speech recognition scores, suggesting that it is not possible to predict which AO patients will benefit from stapedectomy or would be better served by a cochlear implant [38, 39•, 43].

Several recent studies examining auditory outcomes in AO between stapedectomy and cochlear implantation highlight the more consistent improvement in speech recognition with cochlear implantation [38, 39•, 43, 44]. Kabbara et al. found a greater mean word recognition score at 1-year follow-up after cochlear implantation (75%) compared to stapedectomy (50.6%) with word recognition scores greater than 50% occurring in 85% and 60% of the cochlear implantation and stapedectomy groups, respectively [43]. Similarly, Calmels et al. reported increased speech recognition scores and rates of subjective satisfaction and telephone use after cochlear implantation for AO compared to stapedectomy [33]. A 2016 meta-analysis by Abdurehim et al. incorporating the aforementioned studies found significantly greater improvement in speech recognition scores from cochlear implantation compared to stapedectomy in AO [39•].

The decision making for auditory rehabilitation in AO can be nuanced. Cochlear implantation may result in more consistent and long-term improvement in speech recognition scores. However, a subset of patients will receive similar benefit from stapedectomy, a less costly and complex procedure than cochlear implantation. Further, there may be binaural and music appreciation benefits associated with auditory stimulation via stapedectomy and hearing aid versus cochlear implantation [38]. Considering these factors and that it is not currently possible to predict who will achieve good results with stapedectomy, several authors

have recommended initial stapedectomy for all cases of AO prior to cochlear implantation [15, 16, 18, 38]. Inherent in this recommendation is that patients with lack of improvement following stapedectomy in AO are still able to undergo cochlear implantation; thus, no opportunities are lost with upfront stapedectomy. Other authors have suggested an algorithmic approach to decision making, weighting preoperative speech recognition scores and HRCT disease grade (under the hypothesis that high grade disease indicates higher chance of long-term failure with stapedectomy from cochlear otosclerosis progression) [43–45]. Merkus et al. proposed an approach utilizing a speech recognition threshold of 30%, with values below this prompting recommendation for cochlear implantation, and values between 30 and 50% being recommended stapedectomy or cochlear implantation depending on HRCT grade and the air bone gap [44]. Thus, no universal approach is utilized and no definitive guidelines exist for auditory rehabilitation in AO. The decision between stapedectomy and cochlear implantation in otosclerosis auditory rehabilitation should be made through appropriate patient counseling and consideration of case specifics to guide decision making. In our practice, for patients presenting with no preoperative word understanding, a cochlear implant is recommended. If there is some preoperative word recognition on monosyllabic word testing, a stapedectomy could be considered.

Facial Nerve Stimulation in Otosclerosis CI Recipients

Facial nerve stimulation (FNS) is a well-documented complication of cochlear implantation in the broad CI patient population, occurring in 0.9–14.9% of patients amongst large case series [46–51]. Otosclerosis is associated with higher rates of FNS after cochlear implantation, occurring in up to 75% of patients in some series [47, 49, 50], although lower rates between 2.7% and 53% are presented in other series [22, 27•, 52•, 53, 54]. The onset of FNS can be immediate or delayed, with many cases starting over 1 year after implantation [54]. FNS has a range of symptomatic presentations from a subjective sensation of movement to gross total facial musculature stimulation; Kelsall et al. created a grading scale with 6 levels to describe the degree of FNS [48].

FNS in otosclerosis is speculated to occur at higher rates secondary to current leakage through otospongiotic bone, which may have lower impedance compared to normal otic capsule bone [55, 56]. Seyyedi et al. examined temporal bones of AO CI patients, finding that FNS only occurred in those with otospongiotic lesions in the upper basal turn of the cochlea, near the labyrinthine segment of the facial nerve [57]. An anatomic study by Kruschinski et al. demonstrated

that the most common site of electrode deactivation for FNS in their series of AO CI recipients occurred near the thinnest (average 0.33 mm) portions of bone separating the labyrinthine segment of facial nerve from the scala tympani (in the upper basal turn) [52•]. Fig. 1 demonstrates this anatomic proximity in histologic sections from an otosclerotic temporal bone. In alignment with this hypothesis, several studies have observed lower rates of FNS with perimodiolar arrays, which would have a more medial position within the scala tympani, further away from the otic capsule bone and facial nerve compared to lateral wall arrays [22, 34, 36, 49]. Additionally, as may occur in the general CI population, current leakage through the oval and round windows may produce FNS distal from the labyrinthine segment [55].

FNS may be successfully mitigated through altered programming strategies (i.e., increased pulse widths) or deactivation of select electrodes, often without significant deleterious effect on speech recognition [54, 55]. However, extensive electrode deactivation has been associated with poorer auditory outcomes in several studies [30•, 49, 50, 58]. Diminished auditory benefit from FNS requiring electrode deactivation has been successfully managed with CI removal and implantation of a new device [59]. Notably, other non-auditory stimuli which may occur, including tinnitus, headache and vertigo, may be successfully managed with similar programming strategies [60].

Incomplete and False Tract CI Insertion

Incomplete or false tract electrode insertions have been reported to occur secondary to the unique otic capsule structural changes of retrofenestral disease. Round window obliteration has been commonly associated with false tract insertions; the lack of a round window niche as an anatomic reference can result in misplaced cochleostomy drilling trajectories. Several cases of CI placement in the superior semi-circular canal have been reported in the setting of round window ossification [21•, 22, 41]. These instances are often detected on peri-operative imaging [27•] and in one case, post-operative imaging prompted by persistent vertigo [22]. Similar descriptions of false insertion or entry into the vestibule or lateral canal are also reported [22, 61]. CSF gusher has been reported following cochleostomy in cases of severe retrofenestral disease, successfully managed with periosteal plugging [41, 62]. False tract insertions may also traverse cavitary plaques into the middle ear or carotid canal in severe retrofenestral disease [41, 63]. Fortunately, these complications are rare.

Incomplete insertions, in which 1 or several electrodes lie outside the cochlea, are reported to occur in up to 10% of AO CI recipients and are not always associated with cochlear ossification or intraluminal drilling [20•, 30•, 40, 61].

Fig. 1 Histologic temporal bone sections from a 79-year-old female with pre-mortem diagnosis of otosclerosis without prior cochlear implantation. The proximity of the labyrinthine segment of the facial nerve to the upper basal turn of the scala tympani is shown in **A**, separated by only a thin layer of otic capsule bone. **B** shows an adjacent histologic section with an otospongiotic focus near the basal turn of the scala tympani and labyrinthine segment of the facial nerve. These sections demonstrate the anatomic proximities leading to facial nerve stimulation through cochlear implant current leakage through thin otic capsule bone and otospongiosis to the facial nerve. “ST” marks scala tympani; “SV” marks scala vestibuli; “FN” marks facial nerve; “*” marks otospongiosis. (Slides accessed from portions of the Harold F Schuknecht temporal bone library residing at the University of California San Diego with permissions granted by Dr. Jeffrey Harris, MD, PhD.)

The effect of incomplete insertions on auditory outcomes is variable across studies and is generally associated with lower maximum speech recognition scores, especially with an increased number of extracochlear electrodes [30•, 40].

Pre-operative HRCT imaging should be performed in AO. Pre-operative knowledge of high grade retrofenestral disease, cochlear obstruction, or cavitary plaques may be useful for anticipating potential false tract or incomplete electrode insertions. Peri-operative imaging following CI insertion is

suggested in AO to detect false tract insertions and should prompt intra-operative adjustments to correct electrode trajectory. Recently, a small case series of 5 patients suggested the pre-operative utility of OTOPLAN software in guiding electrode selection to prevent incomplete insertions (through choice of shorter, compressed implant lengths); however, this evidence is preliminary and further studies are needed to assess the efficacy of this strategy [64].

Surgical, Radiographic, and Histologic Observations of the Obstructed Cochlea in Otosclerosis and Other Conditions

Intra-operative surgical findings from cochlear implantation in AO suggest that cochlear obstruction and ossification may be more common than suggested by post-mortem histologic studies of otosclerosis, with reported rates between 10 and 60.5% in some surgical series [19, 20•, 22, 27•, 34, 36]. This aspect adds surgical complexity to cochlear implantation in AO, as additional drilling or alternative electrode insertion routes are often necessary for full CI insertion. When compared to other causes of cochlear ossification, including infectious, traumatic, and auto-immune, the extent of ossification in otosclerosis is more restricted, often confined to the proximal 5–6 mm or less of the basal turn of the cochlea [12, 19]. However, isolated cases of diffuse cochlear ossification in all 3 scalae have been reported in AO [10].

HRCT is often employed prior to cochlear implantation in AO as part of the diagnostic process and to assess the extent of disease. Although the sensitivity of HRCT in diagnosis of otosclerosis approaches 90% [65], it is less sensitive for detecting cochlear luminal obstruction. Quesnel et al. compared HRCT assessments of round window ossification with post-mortem histologic findings, finding only 50% of cases were detected on HRCT, although all cases of complete round window obstruction were identified [66]. Amongst all causes of cochlear ossification, including otosclerosis, the sensitivity and specificity of HRCT in detecting luminal obstruction are reported between 33–76.5% and 88–100%, respectively [67, 68]. High-resolution T2-weighted magnetic resonance imaging (MRI) has shown more promise in detecting cochlear luminal obstruction from ossification and fibrosis, with reported sensitivity and specificity rates as high as 94.1% and 87.5%, respectively [69]. However, other studies directly comparing HRCT and MRI have not found significant differences between either modality in detecting cochlear luminal obstruction [67, 70]. Thus, pre-operative use of HRCT or MRI may be useful for detecting cochlear obstruction prior to cochlear implantation in AO; however, the surgeon should be aware of the possibility for false negative scans.

Surgical Techniques for Cochlear Implantation in the Obstructed Cochlea

One goal of cochlear implantation in the obstructed cochlea is to insert as many electrodes as possible within the cochlea without disrupting the modiolus and residual neural elements necessary for successful electric stimulation. Although partial CI insertion in the ossified cochlea has shown speech recognition benefits, full insertion trends toward better long-term results [71]. Several different surgical techniques are described to address this challenge, depending on the extent of ossification [72, 73•, 74, 75]. Cochlear obstruction in otosclerosis is often confined to the round window and inferior segment of the basal turn of the cochlea, which can often be managed by removing the obstructing tissue manually or by drilling a tunnel until a patent lumen is identified. The consistency of the obstruction may be variable (i.e., sclerotic bone or fibrous tissue), and often presents a color differentiation compared to otic capsule bone, which Gantz et al. noted can serve as a guide for luminal drilling [74]. Obliteration of the round window niche by otospongiosis or ossification may obscure landmarks for the scala tympani; Balkany et al. recommend superficial drilling 1.5 mm inferior to the pyramidal process until either a patent round window or differentially colored scala tympani ossification is identified [73•]. Cochlear obstruction extending to the upper segments of the basal turn (past the 8–10 mm of inferior segment basal turn where intraluminal drilling is possible) may be managed with partial insertion of a compressed array (less favored due to potentially poorer outcomes with partial insertion [71]) or by extending the cochleostomy superiorly to perform scala vestibuli insertion [75, 76]. Fortunately, extensive obstruction involving both scalae (preventing scala vestibuli insertion) is rare in otosclerosis. If an open scala cannot be obtained by drilling away a few millimeters of obstructing bone, which will be whiter than the otic capsule, a more advanced drill-out can be accomplished by removing the posterior canal wall and closing the ear canal to get a more direct view of the descending basal turn of the cochlea until an opening into the cochlea is identified. Usually, the electrode can be advanced easily beyond the obstruction. An upper segment drill-out and use of a split-array CI can also be performed [72, 73•, 77, 78]. Several authors have described stepwise surgical decision making algorithms to guide surgical technique in cochlear obstruction in a similar order to that described here [45, 72, 73•]. Surgeon comfort and experience with each technique may help influence the approach used, as each of these techniques has been described in otosclerosis without obvious differences in auditory outcome when full insertion is achieved [27•, 34, 78].

Conclusion

Cochlear implantation achieves consistent, excellent auditory outcomes for those with profound hearing loss from otosclerosis. Patients should be appropriately counseled regarding the multiple modalities for hearing rehabilitation in advanced otosclerosis, including cochlear implantation. Pre-operative imaging may help guide diagnosis, treatment decision making, and anticipation of possible surgical challenges and complications. FNS after cochlear implantation occurs at higher rates in those with otosclerosis; in most instances, programming with elimination of a few electrodes can usually reduce this problem. Cochlear obstruction from ossification is commonly encountered in otosclerosis and can be surmounted by altered surgical approaches to achieve excellent auditory outcomes. Surgeon facility with managing cochlear obstruction is crucial as this cannot always be predicted by pre-operative imaging.

Compliance with Ethical Standards

Conflict of Interest The authors declare no competing interests.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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References

Papers of particular interest, published recently, have been highlighted as: • Of importance

1. Declau F, Van Spaendonck M, Timmermans JP, Michaels L, Liang J, Qiu JP, Van de Heyning P. Prevalence of otosclerosis in an unselected series of temporal bones. *Otol Neurotol*. 2001;22:596–602.
2. Ramsay HAW, Linthicum FH. Mixed hearing loss in otosclerosis: Indication for long-term follow-up. *Am J Otol*. 1994;15:536–9.
3. Ishai R, Halpin CF, Shin JJ, McKenna MJ, Quesnel AM. Long-term incidence and degree of sensorineural hearing loss in otosclerosis. *Otol Neurotol*. 2016;37:1489–96.

4. Rudic M, Keogh I, Wagner R, Wilkinson E, Kiros N, Ferrary E. The pathophysiology of otosclerosis: review of current research. *Hear Res.* 2015;330:51–6.
5. Quesnel AM, Ishai R, McKenna MJ. Otosclerosis: temporal bone pathology. *Otolaryngol Clin North Am.* 2018;51:291–303.
6. Schuknecht HF, Barber W. Histologic variants in otosclerosis. *Laryngoscope.* 1985;95:1307–17.
7. Kwok OT, Nadol JB. Correlation of otosclerotic foci and degenerative changes in the organ of corti and spiral ganglion. *Am J Otolaryngol Neck Med Surg.* 1989;10:1–12.
8. Doherty JK, Linthicum FH. Spiral ligament and stria vascularis changes in cochlear otosclerosis: Effect on hearing level. *Otol Neurotol.* 2004;25:457–64.
9. Nelson EG, Hinojosa R. Questioning the relationship between cochlear otosclerosis and sensorineural hearing loss: a quantitative evaluation of cochlear structures in cases of otosclerosis and review of the literature. *Laryngoscope.* 2004;114:1214–30.
10. Ibrahim RAA, Linthicum FH. Labyrinthine ossificans and cochlear implants. *Arch Otolaryngol.* 1980;106:111–3.
11. Gildener-Leapman N, Linthicum FH. Histopathology of cochlear otosclerosis: implications for cochlear implantation. *Otol Neurotol.* 2011;32:32–3.
12. Green JD, Marion MS, Hinojosa R. Labyrinthitis ossificans: histopathologic consideration for cochlear implantation. *Otolaryngol Neck Surg.* 1991;104:320–6.
13. House HP, Sheehy JL. Stapes surgery: selection of the patient. *Ann Otol Rhinol Laryngol.* 1961;70:1062–8.
14. Iurato S, Ettorre GC, Onofri M, Davidson C. Very far-advanced otosclerosis. *Am J Otol.* 1992;13:482–7.
15. Glasscock ME, Storper IS, Haynes DS, Bohrer PS. Stapedectomy in profound cochlear loss. *Laryngoscope.* 1996;106:831–3.
16. Shea PF, Ge X, Shea JJ. Stapedectomy for far-advanced otosclerosis. *Am J Otol.* 1999;20:425–9.
17. Sheehy J. Far-advanced otosclerosis: diagnostic criteria and results of treatment; Report of 67 Cases. *Arch Otolaryngol.* 1964;80:244–9.
18. Wiet RJ, Morgenstein SA, Zwolan TA, Pircon SM. Far-advanced otosclerosis: cochlear implantation vs stapedectomy. *Arch Otolaryngol Neck Surg.* 1987;113:299–302.
19. Fayad J, Moloy P, Linthicum FH. Cochlear otosclerosis: does bone formation affect cochlear implant surgery? *Am J Otol.* 1990;11:196–200.
- 20.● Calvino M, Sánchez-Cuadrado I, Gavilán J, Lassaletta L. Cochlear implant users with otosclerosis: are hearing and quality of life outcomes worse than in cochlear implant users without otosclerosis?. *Audiol Neurotol.* 2019;23:345–55. **This study evaluated quality of life outcomes between otosclerosis and no-otosclerosis CI recipients, finding equivalent quality of life benefit from the CI.**
- 21.● Dumas AR, Schwalje AT, Franco-Vidal V, Bébér JP, Darrouzet V, Bonnard D. Cochlear implantation in far-advanced otosclerosis: hearing results and complications. *Acta Otorhinolaryngol Ital.* 2019;38:445–52. **This retrospective study of 36 patients examined auditory outcomes and complication rates from cochlear implantation in otosclerosis. A trend toward worse outcomes and higher complication rate was seen with cochlear otosclerosis.**
22. Rotteveel LJC, Proops DW, Ramsden RT, Saeed SR, Van Olphen AF, Mylanus EAM. Cochlear implantation in 53 patients with otosclerosis: demographics, computed tomographic scanning, surgery, and complications. *Otol Neurotol.* 2004;25:943–52.
23. Verhaert N, Borgers C, De Voecht K, Boon E, Desloovere C. A role for acoustic stimulation in advanced otosclerosis: direct acoustic cochlear implant versus cochlear implant. *Audiol Neurotol.* 2018;23:89–97.
24. Sainz M, García-Valdecasas J, Garófano M, Ballesteros JM. Otosclerosis: mid-term results of cochlear implantation. *Audiol Neurotol.* 2007;12:401–6.
- 25.● Semaan MT, Gehani NC, Tummala N, Coughlan C, Fares SA, Hsu DP, Murray GS, Lippy WH, Megerian CA. Cochlear implantation outcomes in patients with far advanced otosclerosis. *Am J Otolaryngol - Head Neck Med Surg.* 2012;33:608–14. **This retrospective study of 30 patients found no long-term differences in auditory outcomes associated with the presence or absence of luminal obliteration or cochlear otosclerosis for cochlear implantation in AO.**
26. Castillo F, Polo R, Gutiérrez A, Reyes P, Royuela A, Alonso A. Cochlear implantation outcomes in advanced otosclerosis. *Am J Otolaryngol - Head Neck Med Surg.* 2014;35:558–64.
- 27.● Vashishth A, Fulcheri A, Rossi G, Prasad SC, Caruso A, Sanna M. Cochlear implantation in otosclerosis: surgical and auditory outcomes with a brief on facial nerve stimulation *Otol Neurotol.* 2017;38:e345–53. **Retrospective study of 36 patients, finding a non-significant trend to worse auditory outcomes in cochlear implantation for AO in the presence of cochlear ossification. They also found altered programming strategies to mitigate FNS did not affect auditory outcomes overall.**
28. Çatli T, Tokat T, Bozkurt EB, Adibelli ZH, Ergün U, Altaş E, Olgun L. Surgical considerations and audiological results of cochlear implantation in patients with otosclerosis. *Turkish J Med Sci.* 2020;50:855–9.
29. 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.
- 30.● Rotteveel LJC, Snik AFM, Cooper H, Mawman DJ, Van Olphen AF, Mylanus EAM. Speech perception after cochlear implantation in 53 patients with otosclerosis: multicentre results. *Audiol Neurotol.* 2010;15:128–36. **Large (n = 53) study showing better auditory outcomes in cochlear implantation for AO associated with full electrode insertions, less severe disease on HRCT, and no FNS or electrode deactivation.**
31. Quaranta N, Bartoli R, Lo Priore A, Fernandez-Vega S, Giagnotti F, Quaranta A. Cochlear implantation in otosclerosis. *Otol Neurotol.* 2005;26:983–7.
32. Rama-López J, Cervera-Paz FJ, Manrique M. Cochlear implantation of patients with far-advanced otosclerosis. *Otol Neurotol.* 2006;27:153–8.
33. Calmels MN, Viana C, Wanna G, Marx M, James C, Deguine O, Fraysse B. Very far-advanced otosclerosis: stapedotomy or cochlear implantation. *Acta Otolaryngol.* 2007;127:574–8.
34. 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.
35. Mosnier I, Bouccara D, Ambert-Dahan E, Ferrary E, Sterkers O. Cochlear implantation and far-advanced otosclerosis. *Adv Otorhinolaryngol.* 2007;65:323–7.
36. Marshall AH, Fanning N, Symons S, Shipp D, Chen JM, Nedzelski JM. Cochlear implantation in cochlear otosclerosis. *Laryngoscope.* 2005;115:1728–33.
37. Medina MDM, Polo R, Gutierrez A, Muriel A, Vaca M, Perez C, Cordero A, Cobeta I. Cochlear implantation in postlingual adult patients with long-term auditory deprivation. *Otol Neurotol.* 2017;38:e248–52.
38. Van Loon MC, Merkus P, Smit CF, Smits C, Witte BI, Hensen EF. Stapedotomy in cochlear implant candidates with far advanced otosclerosis: a systematic review of the literature and meta-analysis. *Otol Neurotol.* 2014;35:1707–14.
- 39.● Abdurehim Y, Lehmann A, Zeitouni AG. Stapedotomy vs cochlear implantation for advanced otosclerosis: systematic review and meta-analysis. *Otolaryngol - Head Neck Surg (United States).* 2016;155:764–70. **Meta-analysis**

- comparing auditory outcomes in stapedectomy versus cochlear implantation for AO across 4 studies. cochlear implantation yielded better auditory outcomes compared to all stapedectomy recipients, but successful stapedectomy candidates did just as well as cochlear implant recipients.**
40. Vashishth A, Fulcheri A, Guida M, Caruso A, Sanna M. Incomplete and false tract insertions in cochlear implantation: retrospective review of surgical and auditory outcomes. *Eur Arch Oto-Rhino-Laryngology*. 2018;275:1059–68.
 41. Vashishth A, Fulcheri A, Prasad SC, Bassi M, Rossi G, Caruso A, Sanna M. Cochlear implantation in cochlear ossification: retrospective review of etiologies, surgical considerations, and auditory outcomes. *Otol Neurotol*. 2018;39:17–28.
 42. Burmeister J, Rathgeb S, Herzog J. Cochlear implantation in patients with otosclerosis of the otic capsule. *Am J Otolaryngol - Head Neck Med Surg*. 2017;38:556–9.
 43. Kabbara B, Gauche C, Calmels MN, Lepage B, Escude B, Deguine O, Fraysse B, Marx M. Decisive criteria between stapedotomy and cochlear implantation in patients with far advanced otosclerosis. *Otol Neurotol*. 2015;36:e73–8.
 44. Merkus P, Van Loon MC, Smit CF, Smits C, De Cock AFC, Hensen EF. Decision making in advanced otosclerosis: an evidence-based strategy. *Laryngoscope*. 2011;121:1935–41.
 45. Eshraghi AA, Ila K, Ocak E, Telischi FF. Advanced otosclerosis: stapes surgery or cochlear implantation? *Otolaryngol Clin North Am*. 2018;51:429–40.
 46. Pires JS, Melo AS, Caiado R, Martins JH, Elói Moura J, Silva LF. Facial nerve stimulation after cochlear implantation: our experience in 448 adult patients. *Cochlear Implants Int*. 2018;19:193–7.
 47. 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.
 48. Kelsall DC, Shalloo JK, Brammeier TG, Prenger EC. Facial nerve stimulation after nucleus 22-channel cochlear implantation. *Am J Otol*. 1997;18:336–41.
 49. Bigelow DC, Kay DJ, Rafter KO, Montes M, Knox GW, Yousef DM. Facial nerve stimulation from cochlear implants. *Am J Otol*. 1998;19:163–9.
 50. Muckle RP, Levine SC. Facial nerve stimulation produced by cochlear implants in patients with cochlear otosclerosis. *Am J Otol*. 1994;15:394–8.
 51. Cohen NL, Hoffman RA, Stroschein M. Medical or surgical complications related to the Nucleus multichannel cochlear implant. *Ann Otol Rhinol Laryngol*. 1988;Suppl 135:8–13.
 52. • Kruschinski C, Weber BP, Pabst R. Clinical relevance of the distance between the cochlea and the facial nerve in cochlear implantation. *Otol Neurotol*. 2003;24:823–7. **This study examines the anatomic basis for facial nerve stimulation in otosclerosis.**
 53. Lombardo F, De Cori S, Aghakhanyan G, et al. 3D-Flair sequence at 3T in cochlear otosclerosis. *Eur Radiol*. 2016;26:3744–51.
 54. Berrettini S, Vito DA, Bruschini L, Passetti S, Forli F. Facial nerve stimulation after cochlear implantation: our experience. *Acta Otorhinolaryngol Ital*. 2011;31:11–6.
 55. Mens LHM, Oostendorp T, van den Broek P. Cochlear implant generated surface potentials: current spread and side effects. *Ear Hear*. 1994;15:339–45.
 56. Frijns JHM, Kalkman RK, Briare JJ. Stimulation of the facial nerve by intracochlear electrodes in otosclerosis: a computer modeling study. *Otol Neurotol*. 2009;30:1168–74.
 57. Seyyedi M, Herrmann BS, Eddington DK, Nadol JB. The pathologic basis of facial nerve stimulation in otosclerosis and multi-channel cochlear implantation. *Otol Neurotol*. 2013;34:1603–9.
 58. Shea JJ, Domico EH. Facial nerve stimulation after successful multichannel cochlear implantation. *Am J Otol*. 1994;15:752–6.
 59. Polak M, Ulubil SA, Hodges AV, Balkany TJ. Revision cochlear implantation for facial nerve stimulation in otosclerosis. *Arch Otolaryngol - Head Neck Surg*. 2006;132:398–404.
 60. 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.
 61. West N, Brand M, Foghsgaard S, Cayé-Thomasen P. Surgical results and complications of cochlear implantation in far-advanced otosclerosis. *J Int Adv Otol*. 2017;13:304–7.
 62. Toungh JS, Zwolan T, Spooner TR, Telian SA. Late failure of cochlear implantation resulting from advanced cochlear otosclerosis: Surgical and programming challenges. *Otol Neurotol*. 2004;25:723–6.
 63. Ramsden R, Bance M, Giles E, Mawman D. Cochlear implantation in otosclerosis: a unique positioning and programming problem. *J Laryngol Otol*. 1997;111:262–5.
 64. Lovato A, Marioni G, Gamberini L, Bonora C, Genovese E, de Filippis C. OTOPLAN in cochlear implantation for far-advanced otosclerosis. *Otol Neurotol*. 2020;41:e1024–8.
 65. Shin YJ, Fraysse B, Deguine O, Cognard C, Charlet JP, Sévely A. Sensorineural hearing loss and otosclerosis: a clinical and radiologic survey of 437 cases. *Acta Otolaryngol*. 2001;121:200–4.
 66. Quesnel AM, Moonis G, Appel J, Malley JTO, McKenna MJ, Curtin HD, Merchant SN. Correlation of computed tomography with histopathology in otosclerosis. 2012;22–8.
 67. Bettman R, Beek E, Van Olphen A, Zonneveld F, Huizing E. MRI versus CT in assessment of cochlear patency in cochlear implant candidates. *Acta Otolaryngol*. 2004;124:577–81.
 68. Frau GN, Luxford WM, Lo WW, Berliner KI, Telischi FF. High-resolution computed tomography in evaluation of cochlear patency in implant candidates: a comparison with surgical findings. *J Laryngol Otol*. 1994;108:743–8.
 69. Isaacson B, Booth T, Kutz JW, Lee KH, Roland PS. Labyrinthitis ossificans: how accurate is MRI in predicting cochlear obstruction? *Otolaryngol - Head Neck Surg*. 2009;140:692–6.
 70. Yan T, Zong F, Ma X, Xu X, Chen W, Song Z. Am J Otolaryngol Cochlear implantation in patients with ossified cochleas. *Am J Otolaryngol* 2019;40:183–6.
 71. Rotteveel LJC, Snik AFM, Vermeulen AM, Mylanus EAM. Three-year follow-up of children with postmeningitic deafness and partial cochlear implant insertion. *Clin Otolaryngol*. 2005;30:242–8.
 72. Coelho DH, Roland JT. Implanting obstructed and malformed cochleae. *Otolaryngol Clin North Am*. 2012;45:91–110.
 73. • Balkany T, Gantz BJ, Steenerson RL, Cohen NL. Systematic approach to electrode insertion in the ossified cochlea *Otolaryngol - Head Neck Surg* 1996;114:4–11. **Review of surgical approaches for cochlear implantation in the ossified cochlea with excellent surgical technical descriptions and figures.**
 74. Gantz BJ, McCabe BF, Tyler RS. Use of multichannel cochlear implants in obstructed and obliterated cochleas. *Otolaryngol Neck Surg*. 1988;98:72–81.
 75. Steenerson RL, Gary LB, Wynens MS. Scala vestibuli cochlear implantation for labyrinthine ossification. *Am J Otol*. 1990;11:360–3.
 76. Lin K, Marrinan MS, Waltzman SB, Roland JT. Multichannel cochlear implantation in the scala vestibuli. *Otol Neurotol*. 2006;27:634–8.
 77. Lenarz T, Büchner A, Tasche C, Cristofoli T, Lesinski-Schiedat A, Wallenberg EV, Battmer RD, Busby PA, Frohne C. The results in patients implanted with the nucleus double array cochlear implant: pitch discrimination and auditory performance. *Ear Hear*. 2002;23:90S–101S.

78. Dejaco D, Prejban D, Fischer N, Freysinger W, Stephan K, Seebacher J, Widmann G, Riechelmann H, Schmutzhard J. Successful cochlear implantation of a split electrode array in a patient with far-advanced otosclerosis assisted by electromagnetic navigation: a case report. *Otol Neurotol*. 2018;39:e532–7.

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