American Journal of Otolaryngology–Head and Neck Medicine and Surgery xxx (2017) xxx–xxx

Contents lists available at ScienceDirect

American Journal of Otolaryngology–Head and Neck Medicine and Surgery

journal homepage: www.elsevier.com/locate/amjoto

Cochlear implantation in patients with otosclerosis of the otic capsule

Jay Burmeister [a,*,1], Susan Rathgeb [b,1], Jacques Herzog [b,c,2]

[a] Des Peres Hospital Otolaryngology Residency, 2345 Dougherty Ferry Rd., St. Louis, MO 63122, United States
[b] Center for Hearing and Balance Disorders, 226 South Woods Mill Road, Suite 58 West, Chesterfield, MO 63017, United States
[c] Department of Otolaryngology, Washington University School of Medicine, 660 S. Euclid Ave., St. Louis, MO 63110, United States

ARTICLE INFO

Article history:
Received 14 May 2017
Available online xxxx

Keywords:
Otosclerosis
Cochlear implant
Otic capsule

ABSTRACT

Objective: To evaluate outcomes of cochlear implantation of patients with otosclerosis of the otic capsule.
Study design: A retrospective case series of 6 patients (7 ears).
Patients: 6 patients (7 ears), 5 patients with severe to profound sensorineural hearing loss; 1 patient with mild to profound sensorineural hearing loss, with radiologic evidence of otosclerosis. All patients were adult males, with or without history of stapes surgery.
Intervention: Cochlear implantation of 7 ears. 5 patients with severe to profound sensorineural hearing loss received the Nucleus Contour Advance peri-modiolar electrode array with binaural implantation performed in one patient. One patient with mild to profound sensorineural hearing loss received a Cochlear® Nucleus Hybrid L24 device.
Methods: Preoperative temporal bone CT, audiometric and speech perception testing scores were reviewed, confirming presence of otosclerosis of the cochlea as well as cochlear implant candidacy. Speech perception testing included CNC words, HINT sentences and AZ Bio scores to measure hearing outcomes post implantation.
Results: All recipients of the contour advance device had a significant improvement in hearing at both 3 and 6 month follow up. The hybrid device recipient experienced loss of residual hearing in the implanted ear without improvement at 3 months and mild improvement at 6 months.
Conclusion: Cochlear implantation has proven to be effective in the treatment of patients with sensorineural hearing loss, including those with otosclerosis of the cochlea. Hybrid candidacy in the setting of otosclerosis of the cochlea may require consideration of alternative electrode devices, most likely a peri-modiolar device.

1. Introduction

Otosclerosis is a common cause of progressive hearing loss, typically resulting in a conductive hearing loss due to fixation of the stapes. Histologically, otosclerosis is described as a remodeling process of the endochondral bone of the otic capsule. The most common site of involvement is the fissula ante fenestram, although otosclerosis can involve any part of the otic capsule [1,2]. When otosclerosis involves the cochlea it may lead to progressive sensorineural hearing loss. Far advanced otosclerosis is considered when bone conduction pure tone thresholds are not measurable using a standard audiometer and air conduction thresholds are no better than 85 dB, as first described by House and Sheehy [3].

Otosclerosis presenting with progressive conductive hearing loss is most often managed with stapes surgery. When otosclerosis has progressed to include a sensorineural component, hearing aid amplification becomes necessary. In the case of far advanced otosclerosis or any patient with severe to profound sensorineural hearing loss and poor word discrimination, cochlear implantation has proven to be the most effective treatment modality [2,5–10]. Cochlear implantation of patients with far advanced otosclerosis does present the possibility of unique adverse effects, most commonly facial nerve stimulation via transotic conduction from the electrode, as well as concerns regarding patency of the scala tympani.

Audiometric criteria for cochlear implantation have expanded significantly over time. Currently, criteria for conventional cochlear implantation as approved by the Food and Drug Administration (FDA), include moderate to profound sensorineural hearing loss in both ears and ≤50% sentence recognition in the ear to be implanted and ≤60% in the opposite ear or binaurally. Cochlear implant electrodes are currently available in both straight and modiolar hugging designs [4].

In March 2014, the FDA approved hybrid cochlear implantation of patients with normal to moderate low frequency hearing loss and severe to profound mid to high frequency loss. The audiometric criteria for hybrid cochlear implantation include: Normal to moderate hearing loss in the low frequencies (thresholds better than 60 dB HL up to and including 500 Hz), severe to profound mid to high-frequency hearing

* Corresponding author.
E-mail address: jay.d.burmeister@gmail.com (J. Burmeister).
[1] No disclosures.
[2] Disclosure: consultant for cochlear and cochlear advisory board member.

http://dx.doi.org/10.1016/j.amjoto.2017.05.011
0196-0709/

loss (threshold average of 2000, 3000, and 4000 Hz ≥75 dB HL) in the ear to be implanted, with CNC word recognition score between 10% and 60% in the preoperative aided condition. The contralateral ear should have moderately severe to profound high frequency hearing loss (threshold average of 2000, 3000, and 4000 Hz >60 dB HL) with CNC score equal to or better than that of the ear to be implanted, but not better than 80%. The hybrid cochlear implant system is not a modiolar hugging electrode system [5].

2. Methods

After institutional review board approval was obtained, this single center retrospective review was conducted. Implantation of all patients in this series was performed by the senior author(JH). All cochlear implant procedures between September 2014 and November 2015 were reviewed. 106 post-lingually deafened adult patients were identified. Six of these patients (5.6%) displayed evidence of otosclerosis of the cochlea on high resolution temporal bone CT (Fig. 1), while still meeting criteria for cochlear implantation based on current recommendations.

All patients were male, ranging from 53 to 82 years of age, with mean age of onset of hearing loss approximately 33 years of age. Four patients (5 ears) were implanted with the Cochlear® Nucleus CI512 platform (#1, 2, 3, 5L, 5R). Patient #4 received the Cochlear Nucleus contoured CI24RE platform. These patients had severe to profound preoperative hearing loss as evidenced by audiogram and sentence recognition scores. Patient #5 received binaural cochlear implantation, the left ear initially, followed by right 6 months later; each ear is listed separately with designation of 5L and 5R. Three patients had at least one previous stapes surgery (#1, #2, and #6).

Patient #6 received Cochlear® Nucleus Hybrid L24 system. This patient was a 60-year-old male with mild to profound mixed hearing loss with reduced word recognition scores, as well as history of prior stapedotomy of the ipsilateral ear, meeting criteria for hybrid implantation.

All patients met current implantation criteria and were deemed to be stable for surgery. No patients required preoperative treatment of vertigo or infectious processes. All patients were given thorough instruction of risks, benefits and alternatives to implantation and provided verbal and written consent.

All patients had uncomplicated and smooth insertion of electrodes. 2 patients, (#2 and #6) including the hybrid implant recipient, had round window obliteration recognized intraoperatively. Drill curettage was used to allow full insertion into the scala tympani without difficulty in these patients.

All implants were activated 3 weeks post operatively with weekly rehabilitation visits as necessary. Patients subsequently underwent hearing evaluations at 3 and 6 months postoperatively. All speech perception testing was performed with the contralateral ear occluded.

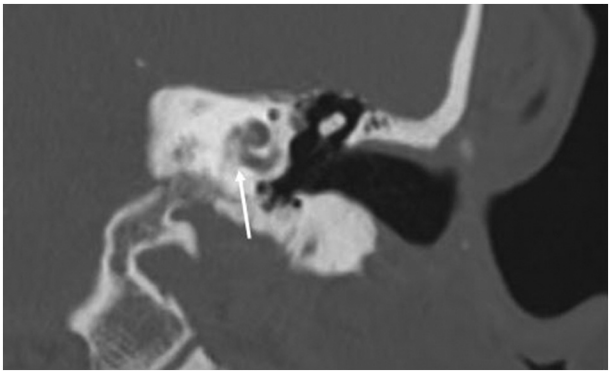

Fig. 1. Patient #6 hybrid implant recipient preoperative coronal ct of left temporal bone displaying otosclerotic focus (white arrow) involving basal turn of the cochlea.

Patients were also followed for any complications such as facial nerve stimulation.

3. Results

Preoperatively, the speech perception scores of patients 1–5 suggested profound hearing loss. These 5 patients had HINT sentence scores of 0–4% preoperatively and received a contour advance device. Patient #6 presented with HINT sentence score of 36% preoperatively.

Results are summarized in Table 1. At 3 month follow up the contour advance electrode recipients had improvement in hearing on HINT sentence scores ranging from 36 to 97%, mean = 62%. CNC scores at 3 months for these patients averaged 37%. At 6 months, improvement continued with sentence scores ranging from 38 to 100%, mean = 75%. CNC scores at 6 months ranged from 20 to 75%, mean = 42%.

The hybrid recipient had a loss of residual hearing with decrease of the preoperative HINT sentence score of 36% to 0% at 3 months postoperatively. His CNC score was also 0% at 3 months. At 6 months, there was improvement HINT sentence score to 25%, although CNC score remained at 0%. Changes in speech perception are illustrated in Fig. 2.

No patients in this series complained of facial nerve stimulation or other complications throughout the duration of the study.

4. Discussion

There have been multiple reports of cochlear implantation in patients with far advanced otosclerosis. There are however, no reported results of hybrid implantation in patients with otosclerosis. In Gantz, et al.'s report, of the 87 patients receiving a hybrid cochlear implant device, otosclerosis was not described as an etiology in any of the recipients [11]. This study did show good hearing outcomes with hybrid implantation, including a >90% rate of hearing preservation [11].

Several studies have demonstrated efficacy of cochlear implantation in patients with otosclerosis involving the otic capsule, however with the increased postoperative complication of facial nerve stimulation [2–7]. Facial nerve stimulation is found most commonly in the area around the geniculate ganglion, with facial nerve twitches being the most common manifestation of this complication [12,13]. This has been managed by removing those areas from the electrode MAP, however this may result in poorer hearing outcomes as a result of a decreased number of functioning electrodes[13]. These studies found that facial nerve stimulation was more common in patients with higher grade otosclerosis on high resolution CT and in patients who received straight rather than contoured peri-modiolar implants [13]. While none of our patients presented with any signs or symptoms of facial nerve stimulation, our hybrid implant recipient had poorer than expected hearing outcome. All patients who received peri-modiolar electrodes had improvement in hearing. This very likely explains the poor outcome of our hybrid recipient.

Seeman, et al. reported results of 30 patients who received cochlear implants with far advanced otosclerosis and found this was a safe and

Table 1
Results.

Patient #	Age	Internal	Pre-op HINT score	3 month post-op HINT score	3 month post-op CNC word score	6 month post-op HINT score	6 month post-op CNC word score
#1	82	CI512	0%	36%	16%	67%	24%
#2	67	CI512	1%	77%	32%	69%	32%
#3	71	CI512	0%	48%	32%	90%	44%
#4	65	CI24RE	0%	26%	16%	38%	20%
#5L	53	CI512	0%	97%	64%	100%	72%
#5R		CI512	4%	88%	60%	86%	60%
#6-Hybrid	60	Hybrid L24	36%	0%	0%	25%	0%
Mean	66		6%	53%	31%	68%	36%

Please cite this article as: Burmeister J, et al, Cochlear implantation in patients with otosclerosis of the otic capsule, American Journal of Otolaryngology–Head and Neck Medicine and Surgery (2017), http://dx.doi.org/10.1016/j.amjoto.2017.05.011