Simultaneous Labyrinthectomy and Cochlear Implantation in Unilateral Meniere's Disease

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Objective: In a single-institution, FDA-approved IDE study, subjects with unilateral Meniere’s disease and intractable vertigo underwent concurrent labyrinthectomy and cochlear implantation to determine speech perception, localization, and quality of life outcomes.

Methods: Three subjects with unilateral Meniere’s disease with normal or near-normal hearing in the contralateral ear underwent simultaneous labyrinthectomy and cochlear implantation. Sound localization testing demonstrated immediate benefit postimplantation with the cochlear implant (CI). RMS error with CI on was 22 degrees (±2) and with CI off was 63 (±15) at 6 months. Mean CI alone scores were 22% (±20) at 1 month and improved to 43% (±20) and 49% (±11) at the 3- and 6-month intervals, respectively. AzBio sentences in babble (0 dB SNR) scores presented in the most challenging listening condition (S0NContra) were 28% (±20) at 1 month, 38% (±18) at 3 months, and 45% (±24) at 6 months. Tinnitus Handicap Inventory (THI) significantly improved from an average preoperative score of 42 (±26) to 0 at 6 months. Quality of life measures improved overall over the postimplantation follow-up intervals.

Conclusions: Subjects with unilateral Meniere’s Disease who underwent simultaneous labyrinthectomy and cochlear implantation experienced improvements in sound localization, speech understanding, tinnitus severity, and quality of life with device use. There was a trend for better performance over the postoperative intervals.

Key Words: Meniere’s Disease, cochlear implant, labyrinthectomy, single-sided deafness.

Level of Evidence: 2b.

INTRODUCTION

Meniere’s disease is an idiopathic disease characterized by fluctuating sensorineural hearing loss (SNHL), aural fullness, tinnitus, and vertigo.1 The etiology of Meniere’s disease is unknown; however, pathological examination demonstrates endolymphatic hydrops, in addition to damage of the inner and outer hair cells with preservation of spiral ganglion cells.2,3

The management of Meniere’s disease is approached in a stepwise fashion with goals to optimize control of vertiginous symptoms and preserve hearing. Conservative medical management, followed by intratympanic steroid injections and chemical labyrinthectomy are all options for early treatment of Meniere’s disease.4 Surgical labyrinthectomy, endolymphatic sac decompression, and vestibular neurectomy may be offered for patients who fail to benefit from more conservative measures. Labyrinthectomy can provide definitive treatment of vertigo, but does so at the expense of remaining hearing.

Patients with unilateral hearing loss (UHL), as often seen in unilateral Meniere’s disease, have decreased sound localization and speech perception abilities, and overall reduced quality of life due to the loss of binaural hearing.5,6 Binaural hearing provides advantages over monaural hearing through several mechanisms, including binaural summation, binaural squelch effect, and the head-shadow effect.7–9 Sound localization is also improved with binaural hearing.10–12

Traditional rehabilitative options for UHL include contralateral routing of signal (CROS) hearing aid technology, and osseointegrated implants.13,14 Although both route sound from the affected side to the better hearing ear, hearing is not restored in the affected ear so optimization of binaural hearing is minimal.15–17 The off-label use of cochlear implantation to treat UHL has demonstrated not only improvement of auditory perception in the affected ear, but restoration of binaural hearing with resulting improvement in speech perception in noise and sound localization.18–22
Previous studies have demonstrated successful audiologic rehabilitation of Meniere’s disease patients with cochlear implantation. Simultaneous labyrinthectomy and cochlear implantation in patients with Meniere’s disease provide a unique opportunity to address intractable vertigo and restore auditory perception in a deafened ear. Previous investigation of outcomes following simultaneous labyrinthectomy and cochlear implantation in Meniere’s disease patients have either been retrospective case studies, focused on bilateral disease, or part of a larger cohort. It is therefore difficult to apply any conclusive outcomes within the current literature to this study population. In our study, we aimed to prospectively investigate outcomes of speech perception, sound localization, and quality of life measures in subjects with unilateral Meniere’s disease and normal or near-normal hearing in the contralateral ear following concurrent labyrinthectomy and cochlear implantation.

MATERIALS AND METHODS

This study was approved by the Food and Drug Administration and Institutional Review Board at the University of North Carolina at Chapel Hill as a prospective, investigational device exemption clinical trial. This was part of a larger trial investigating benefits of patients undergoing cochlear implantation at the time of either translabyrinthine vestibular schwannoma resection or labyrinthectomy for Meniere’s disease. Three patients with unilateral Meniere’s Disease scheduled for labyrinthectomy for intractable vertigo consented to undergo concurrent cochlear implantation.

Subjects met the diagnostic criteria for unilateral Meniere’s Disease based on the American Academy of Otolaryngology–Head and Neck Surgery (AAO–HNS) guidelines.

Preoperatively, subjects had a pure tone average (PTA, average unaided threshold at 500, 1000, and 2000 Hz) of less than or equal to 35 dB HL in the contralateral ear (Table I). Subjects reported a history of vertigo, hearing loss, and tinnitus average unaided threshold at 500, 1000, and 2000 Hz) of less than or equal to 35 dB HL in the contralateral ear (Table I). Subjects reported a history of vertigo, hearing loss, and tinnitus unsuccessfully controlled with either medical management or intratympanic gentamicin injections, and presented with the primary motivation to undergo labyrinthectomy due to intractable vertigo.

All subjects underwent a standard transmastoid labyrinthectomy followed by cochlear implantation with the MED-EL SYNCHRONY device with a Standard electrode array. A full electrode insertion through the round window was achieved for all three implantations and confirmed by intraoperative transorbital x-ray. Subjects were fitted with the SONNET audio processor and devices were activated 3 to 4 weeks postoperatively. Follow-up testing was completed at 1, 3, and 6 months postactivation, as described below.

Speech perception was evaluated using the consonant-nucleus-consonant (CNC) word test in quiet and AzBio sentences test. Recorded materials were presented at 60 dB SPL. CNC words in quiet were tested in the unaided condition with appropriate masking applied to the contralateral ear preoperatively and at 1, 3, and 6 months postactivation with the cochlear implant (CI) alone, and masking applied to the contralateral ear. AzBio sentence testing was administered in a 10-talker babble (0 dB SNR) at postactivation intervals in CI-on and CI-off listening conditions. AzBio sentences in babble testing were presented in three conditions: 1) speech front (0 degrees azimuth), noise front (SONC); 2) speech front, noise to the implanted ear (SONCI); and 3) the most challenging condition, with speech presented from the front and noise to the contralateral ear (SONContra).

The sound localization task presented a 200-ms bandpass filtered noise randomly across the speakers at varied intensity levels. Subjects reported the perceived sound source when listening in the CI-on and CI-off conditions. Sound localization is reported as a root mean square (RMS) error in degrees. Sound localization was evaluated post-activation at the 1-, 3-, and 6-month intervals.

Subjective questionnaires included the Speech, Spatial, and Qualities of Hearing scale (SSQ), Abbreviated Profile of Hearing Aid Benefit (APHAB), and Tinnitus Handicap Inventory (THI). The SSQ is a 50-item questionnaire used to assess the self-perception of auditory disability for speech hearing, spatial hearing, and quality of hearing. The APHAB is divided into four subscales: ease of communication, reverberation, background noise, and averseness. Both questionnaires were administered at 1, 3, and 6 months postactivation. Tinnitus severity was assessed using the THI preoperatively and at 1, 3, and 6 months postactivation. Tinnitus severity was assessed using the THI preoperatively and at 1, 3, and 6 months postactivation with subject reports based off the CI-on condition. Frequency of vertigo attacks was assessed at 1-, 3-, and 6-month intervals.

RESULTS

In this study, we wished to determine the benefits of cochlear implantation in subjects undergoing labyrinthectomy for Meniere’s Disease. Three subjects with unilateral Meniere’s disease underwent simultaneous labyrinthectomy and cochlear implantation. Subject demographics are listed in Table I. The hearing in the contralateral ear was unchanged through the postactivation intervals. Device data-logging suggested that all subjects wore their external audio processor at least 8 hours per day. Subject 2 was an exception, who wore the device only 3 logged hours per day during the first month, but then wore the processor at least 8 hours per day thereafter. Consistent device use is important because duration of daily device use has shown to be a

| Subject | Gender | Age at Onset (yr) | Age at HL (yr) | Age at Implantation (yr) | Operate Ear | Pre-op affected ear CNC | Pre-op Contra Ear CNC |
|---------|--------|------------------|---------------|------------------------|-------------|----------------------|---------------------|
| S1      | M      | 47               | 55            | 65                     | L           | 14%                  | 100%                |
| S2      | F      | 46               | 47            | 48                     | L           | 60%                  | 100%                |
| S3      | F      | 55               | 65            | 65                     | R           | 14%                  | 100%                |

CNC = consonant-nucleus-consonant; HL, hearing loss; L = left; R = right.
contributive factor in improved early postactivation sound localization performance.\textsuperscript{13}

We first wished to determine the timing and degree of improvement in sound localization following activation by reporting RMS error (Fig. 1). With a lower value indicating better performance, RMS error at the 1-month interval with CI on was on average 31 (±3) degrees compared to 58 (±5) degrees with CI off. Sound localization testing progressively improved with an average RMS error of 26 (±2) and 22 (±2) degrees with CI on compared to 67 (±18) and 63 (±15) degrees with CI off, for 3- and 6-month testing, respectively. All three study subjects individually demonstrated progressive improvement in sound localization testing immediately following implant activation.

We also aimed to investigate speech perception outcomes in both quiet and noisy environments (Figs. 2 and 3). Reported as percentage of words correct, CNC words in quiet slightly decreased at the 1-month test interval in the CI alone condition (average of 22% ± 20) compared to preoperative unaided testing (average of 29% ± 26), but then steadily improved at both 3 and 6 months postactivation to an average of 43% (±20) and 49% (±11), respectively (Fig. 2). AzBio sentences in a 10-talker babble (0 dB SNR) in the most challenging listening condition (S0Ncontra) demonstrated a similar pattern of improvement to CNC words in quiet with benefit emerging at the 3- and 6-month intervals with the CI on (Fig. 3). At the 1-month interval, AzBio scores were on average 28% with CI on (±20) compared to 30% (±20) with CI off. At 3 months, scores were averaging 38% (±18) compared to 23% (±15), and at 6 months, they were 45% (±24) compared to 33% (±24). AzBio scores in S0N0 and S0NCI noise conditions are shown in Figure 3. There is minimal differences between implant off and implant on conditions. All study subjects demonstrated an improvement in speech perception in quiet and noise in the most challenging listening condition (S0Ncontra) emerging at the 3-months postactivation with CI on compared to the CI-off listening condition.

Cochlear implantation has shown to reduce tinnitus in the affected ear in previous studies and we wished to determine if such improvement was present in our cohort by reporting THI pre- and postactivation (Fig. 4). Figure 4 presents averaged preoperative and 1-, 3-, and 6-month postactivation THI scoring, where a lower value indicates reduced severity. Preoperatively, the mean THI score was 42 (±26, range of 22–72) compared to a postactivation THI score of 1 (±2) and 3 (±5) at 1 and 3 months, respectively. All study subjects individually reported an improvement in tinnitus. At 6 months postactivation, the THI score reached zero for all three study subjects.

In order to investigate the potential improved quality of life following cochlear implantation, we report postactivation questionnaire scoring from the APHAB and SSQ (Fig. 5). APHAB subdomains improved throughout the study period with lower values indicating less perceived difficulty. Global APHAB scoring, which averages the scores from the background noise, reverberation, and ease of communication subscales, improved from an average of 38 (±25) at 1 month to 25 (±10) at 6 months postactivation. Although improvement in background noise and global APHAB scoring plateaued at 3 months, the APHAB subdomains of reverberation, aversiveness, and ease of communication showed continued improvement 1 to 6 months postactivation (Fig. 5A).

The SSQ subscale ratings scores also improved from 1 to 6 months postactivation (Fig. 5B). Quality of hearing increased from an average of 6.0 (±1.6) to 7.3 (±1.2), spatial hearing improved from 6.4 (±0.5) to 7.0 (±1.60) and speech hearing also improved from 6.1 (±1.9) to 7.1 (±1.2) at 1- and 6-month testing intervals, respectively. Improvement in subjective quality of life measures, in both the APHAB and SSQ, was seen for all three study subjects individually suggesting that
subjects perceived improved listening following cochlear implantation in their Meniere’s affected ear.

Frequency of vertigo attacks was assessed preoperatively and 1, 3, and 6 months postactivation. Subjects reported number of vertigo episodes per week. All subjects reported resolution of their vertigo at 6 months postimplantation (Fig. 6).

DISCUSSION

Simultaneous labyrinthectomy and cochlear implantation in Meniere’s disease is an attractive option for not only eliminating debilitating vertigo attacks, but also restoring hearing in a deafened ear. In this study, we aimed to report prospective outcomes of speech perception, sound localization, and quality of hearing following simultaneous labyrinthectomy and cochlear implantation in patients with unilateral Meniere’s disease. Previous studies have been limited to retrospective studies. There is also inconsistent postoperative testing. Our sample size, although small, presents consistent postoperative outcomes at 1-, 3-, and 6-month intervals for all three study subjects.

Results demonstrated immediate benefit in both sound localization and subjective tinnitus across interval testing with the CI on (Figs. 1 and 4). This continued to improve over the study period. Benefit with the CI on speech perception in quiet and noise in the most challenging condition was initially limited at the 1-month interval, but then increased at the 3- and 6-month intervals with an overall improvement at 6 months (Figs. 2 and 3). AzBio scores in conditions presented S0N0 or S0NCI showed minimal change as expected due to plateau effects from the hearing contralateral ear. A report retrospectively evaluating Meniere’s subjects implanted with shorter electrodes demonstrated slower gains in localization (12–24 months). In addition, our subjects reached a CNC word score of near 50% by 6 months postactivation, whereas their subjects did not reach a similar result until 12 months. Although methods were different, AzBio scores in the previous study did not achieve 50% until 12 months after surgery, while our subjects achieved this in the most difficult listening condition of S0Ncontra by 6 months. The subjects described in our study were implanted with the MED-EL Standard array, which has a 31 mm electrode that provides stimulation to the apical region of the cochlea, and may provide earlier improvement due to better solicitation of low-frequency cues.

Overall, self-reported quality of life and hearing measures improved with device use (Fig. 5). This was demonstrated by a reduction in global APHAB scoring from an average of 38 (±25) to 25 (±10), in addition to...
reductions in difficulty ratings on the ease of communication, reverberation, background noise, and aversiveness subscales. Each of the SSQ subscale domains, speech hearing, spatial hearing, and qualities of hearing, improved from the 1- to 6-month intervals. Although not an aim of this study, all patients were noted to have complete resolution of their vertigo by 6 months, further supporting labyrinthectomy as a successful method for eliminating attacks of vertigo (Fig. 6).

Investigations of cochlear implantation in Meniere’s disease patients have demonstrated successful auditory rehabilitation in this and other studies. Simultaneous labyrinthectomy and cochlear implantation provides many benefits, including the benefit of an ablative and restorative procedure under a single general anesthetic, restoration of binaural hearing, and the potential elimination of vertigo and tinnitus. Notably, all of our patients had complete resolution of their tinnitus. This reduction is a not a result of labyrinthectomy, rather from cochlear implantation, as previous studies have not shown a reduction in tinnitus when the labyrinth is removed and an implant is not placed. The central auditory pathway reorganization that occurs following unilateral deafness, which could theoretically exacerbate tinnitus, may be relieved by providing auditory input to the deafened ear. These results, as well as those previously published strongly advocate for cochlear implantation in this population due to its substantial benefits.

CONCLUSION

These results demonstrate that concurrent labyrinthectomy and cochlear implantation is successful in eliminating vertigo symptoms and rehabilitating hearing loss in unilateral Meniere’s disease. This study also demonstrates that patients with UHL receiving cochlear implants benefit from improved sound localization and overall subjective, self-perceived quality of hearing.

Fig. 5. Improved APHAB subscale and SSQ scoring following simultaneous labyrinthectomy and cochlear implantation in unilateral Meniere’s disease. The questionnaires were administered at 1, 3, and 6 months postactivation. Lower APHAB and higher SSQ subscale values indicate improvement. APHAB = Abbreviated Profile of Hearing Aid Benefit; SSQ = Speech, Spatial, and Qualities of Hearing scale.

Fig. 6. Vertigo attacks are eliminated following simultaneous labyrinthectomy and cochlear implantation in unilateral Meniere’s disease. Number of vertiginous episodes per week was assessed preoperatively and at 1, 3, and 6 months postactivation. All subjects reported resolution of their vertigo at 6 months.
CONFLICTS OF INTEREST
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