Cochlear Implantation in Ménière’s Disease

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Introduction
Ménière’s disease (MD) is characterized by cochleovestibular dysfunction that manifests as fluctuating sensorineural hearing loss (SNHL), episodic vertigo, aural fullness, and tinnitus.1 The SNHL is most often initially unilateral and low-frequency in nature, but bilateral involvement may develop at some point in 30 to 50%.2 The incidence of severe to profound SNHL in MD is low, estimated to progress in 1 to 6% of those diagnosed with MD.1 Cochlear Implantation (CI) has been approved by the FDA for rehabilitation of bilateral severe to profound SNHL since 1984, and over 60,000 patients in the US alone have received a CI.3 However, given that the prevalence of MD in the US is estimated at 0.2%,8 the number of CI recipients whose SNHL is secondary to MD is extremely small. Consequently, there is a paucity of data describing the effects of CI on hearing performance and vestibular function in MD.

Patients with severe to profound bilateral SNHL due to MD stand to experience significant benefit from CI. The objectives of this study are to: (1) Determine whether CI in MD patients improves hearing performance as measured by standard CI audiological testing and (2) Describe the impact of CI on subjective auditory and vestibular quality of life measures.

Methods
This study is a retrospective review and was approved by the Institutional Review Board at Wake Forest University (IRB #03022050). Subjects were identified by querying the Otolaryngology and Audiology patient databases for all patients who have undergone CI since 1997. All CI patients who also carried the diagnosis of MD were included in this group. Patient charts were then reviewed for pertinent information, including demographics, past medical and surgical history, audiological data, etc. Only MD patients aged 18 or older who met AAO-HNS diagnostic criteria1 for definite MD were included. Eligible patients were invited to answer a series of questions regarding their current hearing ability via the Hearing Handicap Index (HHI), subjective perception of MD symptoms, and functional status related to their MD before and after implantation via the MD Functional Level Scale (MD-FLS).3 Sentence testing scores, frequency and severity of MD vestibular and auditory symptoms, and hearing quality of life were compared pre- and post-CI. pertinent results were then analyzed for statistical significance.

Results
Thirty CIIs were performed in 12 patients who met study inclusion criteria. The male to female ratio was 2:1. The mean age at first implantation was 62.6 years (range 40-84). 25% of patients underwent pre-CI ablative procedures for their vestibular symptoms (surgical or chemical labyrinthectomy) and 33% had pre-CI endolymphatic sac decompressive surgery. Of those who had ablative procedures, one had the procedure (intratympanic gentamicin injections) performed on the eventual CI ear. Pre- and post-operative audiograms and sentence testing (HINT and A2-Bloc) were available for 10 of the 12 patients and compared (Fig. 1). Mean long-term follow-up was 28.4 months. Mean sentence testing in quiet improved from 26.8% pre-CI to 81.6% post-CI (p<0.0001) at the most recent follow-up.

Patient questionnaires were sent to all 12 patients and all but one were returned. Results from the post-operative Hearing Handicap Inventory (HHI) demonstrated a mean total score of 53.36 ± 26.3. Mean values for the subscales12 were also calculated: Emotional distress 1.96 ± 1.1, Difficulty in hearing 2.96 ± 1.1, and Social restriction 2.18 ± 1.8 (Fig. 2). Despite the fact that mean pre- and post-CI MD-FLS vestibular scores were similar: 3.9 ± 1.9 and 3.4 ± 1.8, respectively (p=0.50), patients reported nearly a ten-fold reduction in the number of annual vertigo episodes post-CI (p=0.033, Fig. 3). Four patients reported persistent functional hearing loss in their implanted ear. 80% of the respondents reported persistent tinnitus (implanted ear) in the non-implanted ear, three in both). All but one patient with tinnitus rated their tinnitus as the "same" or "better" after implantation (Fig. 4).

Discussion
This study further demonstrates that MD patients who undergo CI may obtain hearing performance outcomes similar to CI patients who have other more common acquired forms of severe to profound SNHL.6 A review of the available literature reveals that only five English-language papers investigating CI in the setting of MD have been published.1,4 Of these, only one looked at hearing performance pre- and post-implantation.5 This study concluded there was a significant improvement in hearing performance in these patients.5 The remainder were either single case studies or expert opinion papers. Only Lustig, et al.4 reported on vestibular symptoms post-CI, and their findings were similar to this study’s results.

Hearing fluctuation persists even with electrical stimulation of the cochlea for a certain segment of MD CI recipients, as one third of patients subjectively noted hearing performance fluctuation.5 This fluctuation was also revealed on audiological testing. However, these changes were not severe, and the fluctuations were easily overcome by minor CI programming modifications, resulting in excellent long-term overall stability in sentence testing scores.

Regarding vestibular symptoms, it is encouraging that the introduction of a current emitting electrode within the cochlear microenvironment does not induce vertiginous symptoms in patients with a tendency towards endolymphatic hydrops (EH). This finding is most likely a result of the natural history of vertigo symptoms in longstanding end-stage MD (one of relative resolution). The known histologic changes of intracochlear fibrosis after standard implantation could conceivably affect the endolymphatic space associated with the vestibular end-organs, since the cochlear endolymphatic space may be constricted secondary to adjacent fibrotic changes, making the vestibular endolymphatic space perhaps more vulnerable to EH. However, this theoretic pathophysiology is not supported by this data, and CI did not seem to significantly degrade vestibular function in this sample population.

With current CI efforts aimed at preserving the intracochlear anatomy as much as possible in order to preserve residual hearing by using more forgiving electrode arrays and insertion techniques, future investigations may also look at how these techniques may impact the overall auditory and vestibular symptoms in MD CI recipients.

Conclusion
Patients who undergo CI for severe to profound SNHL attributed to MD are capable of achieving a significant improvement in their hearing performance. This improvement is comparable to the gains experienced by non-MD patients who undergo CI. The introduction of the implant electrode into the cochlea adjacent to the endolymphatic space neither seems to adversely alter the natural history of vestibular dysfunction, nor significantly exacerbate auditory symptoms in CI recipients who have MD. Cochlear implantation remains an excellent option for eligible patients who sustain severe to profound SNHL secondary to MD.

References