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Cochlear Implantation in Ménière's Disease With and Without Labyrinthectomy

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Objective: To investigate outcomes of cochlear implantation (CI) in patients with Ménière's disease (MD) with and without surgical labyrinthectomy.
Study Design: Retrospective study.
Setting: Multiple tertiary referral centers.
Subjects: Thirty one ears from 27 patients (17 men, 10 women, aged 42–84) with CI in ipsilateral MD ear.
Intervention: CI in ears with intact labyrinths (Group 1), CI with simultaneous surgical labyrinthectomy (Group 2), and CI sequential to surgical labyrinthectomy (Group 3).
Main Outcome Measure: Within-subject improvement on Bamford Kowal Bench test or City University of New York open set sentence tests.
Results: Majority of ears achieved excellent open-set speech recognition by 12 months post-CI, irrespective of intervention group. Preoperative details including patient age and sex, implant, MD and previous intervention, and audiological test results did not significantly affect outcomes. Patients with MD undergoing CI only may experience vestibular dysfunction which may cause long-term concerns. Incidental finding was noted of eight ears with fluctuating symptoms in ipsilateral ear during 12-month period post-CI, with five of eight ears showing objective fluctuating impedances and mapping.
Conclusion: CI in MD can yield good hearing outcomes in all three groups and this is possible even after a long delay after labyrinthectomy. Bilateral MD patients are complex and prospective quality of life (QoL) measures would be beneficial in being better able to manage the vestibular outcomes as well as the audiological ones. **Key Words:** Cochlear implants—Hearing outcome—Labyrinthectomy—Ménière's disease—Vestibular function.

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Ménière's disease (MD) is a disorder typically characterized by recurrent attacks of disabling vertigo, fluctuating and progressive hearing loss, and tinnitus, with an estimated prevalence of 46 to 200 per thousand patients (1). The cause and cure for MD remains unknown (2). Optimal treatment continues to be management of symptoms by minimizing vestibular symptoms while preserving hearing as possible. Unfortunately, many patients lose functional hearing in the advanced stages of the disease (3) and many interventions that aim to control vertigo risk further harm to the auditory system (4).

Cochlear implantation (CI) has become common practice in patients with MD and candidacy has changed to include patients with very poor hearing and/or uncontrolled vertigo in at least one ear (5,6) such as those who may benefit from a labyrinthectomy.

Although CI has become more common in patients with MD, our knowledge of clinical outcomes remains limited. Early histological studies predicted good outcomes in this population, as ears with end-stage MD and even labyrinthectomy had sufficient spiral ganglion cells to foresee benefit from electrical stimulation (7,8). Clinical studies thus far have been encouraging with most MD patients achieving excellent hearing outcomes with CI (8–17). However, current literature is lacking conclusive information whether previous interventions to treat MD have any effect on CI outcomes (8–11). In particular, the effect of delayed labyrinthectomy is sparsely reported (12,13) and the effect of simultaneous labyrinthectomy remains unclear, in spite of increased numbers in this latter intervention (6,13,15–18).

The aim of the current study is to investigate CI outcomes in patients with MD, including those with and without labyrinthectomy, with a focus on exploring the challenges experienced in balancing managing vestibular dysfunction and hearing optimization.

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TABLE 1. *Patient details for each intervention group*

	Group 1	Group 2	Group 3
Age (yr)			
Mean + SD	62.7 + 9.8	65.6 + 9.1	71.6 + 12.5
Range	42–76	50–75	59-84
Sex			
Male	13	4	3
Female	9	2	0
Implant			
CI24RE (CA)	11	5	3
CI24RE (ST)	3	0	0
CI512	5	0	0
CI513 (CA)	0	0	0
CI422	3	1	0
CI ear			
Unilateral	17	6	3
Bilateral	5	0	0
MD ear			
Unilateral	3	5	2
Bilateral	19	1	1
MD stage			
Burnt-out	2	0	2
Remission	14	0	1
Active	6	6	0
MD symptoms			
None	15	0	3
Cochlear only	3	0	0
Cochlear + vestibular	4	6	0
MD frequency			
0–1/yr	16	0	3
2–5/yr	2	1	0
6–12/yr	2	0	0
13–50/yr	0	1	0
>50/yr	2	4	0
Intervention			
Gentamicin	4	1	1
ELSR	5	0	0
Steroids	5	0	1
Audiology			
Hearing loss			
Duration (yr)	20.2 ± 13.3	28.5 ± 24.5	30.0 ± 21.7
Degree	Severe to profound	Moderate to profound	Dead ear
Hearing aid use	50% consistent	No or inconsistent	No (post lab.)
Speech recognition	Poor	Fair to excellent	No

Details reported as number of ears, with exception of age and duration of hearing loss (reported as mean and standard deviation in brackets) and description of audiology test results. Group 1 (N = 22) was control group and had cochlear implantation (CI) without labyrinthectomy; Group 2 (N = 6) had simultaneous CI and labyrinthectomy; Group 3 (N = 3) had CI with prolonged delayed after labyrinthectomy.

SD indicates standard deviation.

METHODS

Patients were recruited from multiple tertiary CI centers in Sydney, Wollongong, and Newcastle, Australia. Inclusion criteria were: CI with or without surgical labyrinthectomy and definite MD as per American Academy of Otolaryngology and Head and Neck Surgery (AAOHNS) criteria in the ipsilateral ear. After attaining Human Research Ethics Committee approval (St. Vincent's Hospital, Sydney), medical and audiological records were reviewed from the onset of Ménière's symptoms until 12-months post-CI switch-on. Patients were categorized into three intervention groups:

1) Group 1 (control): CI without labyrinthectomy
2) Group 2: CI with labyrinthectomy performed simultaneously
3) Group 3: CI with prolonged delay after labyrinthectomy

Patient details are listed for each intervention group in Table 1. Details include: age at implantation (mean, standard deviation in years); sex; implant; CI ear (unilateral or bilateral); MD ear (unilateral or bilateral); MD stage (based on vestibular function test results in ipsilateral ear—*burnt out* if no residual function, *remission* if residual function but no symptoms, *active* if residual function and symptoms); MD symptoms and frequency (based on patient report over 12 months before implantation); intervention (dose and frequency not always available so recorded if gentamicin, endolymphatic sac reduction, intratympanic steroids ever used in ipsilateral ear); audiology results at pre-CI assessment (using Bamford Kowal Bench and City University of New York scores), duration of hearing loss in years, hearing loss and hearing aid use.

CI Candidacy

The sample included 31 ears of 27 patients. In this study, the vast majority of patients in Group 1 had burnt out MD with very poor aided speech benefit and no vestibular symptoms (n = 20 of 22). Patients fulfilled candidacy for CI if they attained aided sentence speech scores in quiet (Bamford Kowal Bench test [BKB] or City University of New York [CUNY]) of less than 70% at 60 to 65 dB SPL. All patients had bilateral hearing loss. The cause of hearing loss in the contralateral ear was due to bilateral MD in 19 of 22 ears and prolonged hearing loss due to other causes in 3 of 22. Patients in this study did not fulfil candidacy criteria for single sided hearing loss. In, Group 2 and 3, most patients had unilateral MD (five of six in Group 2 and two of three in Group 3) and the decision for labyrinthectomy in the ipsilateral ear was based on the degree and disability from vestibular dysfunction. CI candidacy for that ear was considered secondary to the inevitable dead ear in the ipsilateral side due to labyrinthectomy. In addition, all patients in Group 3 underwent preoperative magnetic resonance imaging (MRI) scanning to confirm cochlear duct patency before being considered for candidacy.

Test methodology varied between clinics due to study being multisite research project. Therefore, there were some limitations analyzing the results. Audiological results collected were pure-tone audiogram (PTA—0.5, 1, 2, and 4 kHz) and open-set sentence test scores (repeat BKB or CUNY test within each subject in the ipsilateral ear, tested preoperatively with optimally fit hearing aids and postoperatively with CI at 3, 6, and 12-months post switch-on, test conditions auditory alone in quiet). Vestibular function test (VFT) results were included pre- and post-CI if done: caloric, cervical, and ocular vestibular evoked myogenic potentials, video head impulse test. Postoperative fluctuation was noted if patient's reported symptoms localized to ipsilateral ear and that correlated with CI impedance and MAP measurements in Custom Sound^{TM} Cochlear Ltd proprietor software (Cochlear Ltd, Sydney, Australia).

Statistical analysis was performed where relevant using IBM SPSS software (PASW for Windows, SPSS Inc., Chicago, IL).

TABLE 2. *Cochlear implant (CI) outcomes for each intervention group*

	Group 1	Group 2	Group 3
Hearing dB (pure tone average: 0.5, 1, 2, and 4 kHz)			
Pre	99 (22)	74 (22)	>120 (0)
Post	116 (21)	>120 (6)	>120 (0)
Speech (percentage aided score BKB or CUNY)			
Pre	13 (24)	40 (38)	0 (0)
Post	93 (7)	95 (10)	75 (43)

Outcomes reported as hearing (pure-tone average) and speech (repeat BKB or CUNY) test results in the ipsilateral ear pre- and 12 months post-CI, using mean and standard deviation in brackets. Group 1 (N = 22 ears) was control group and had CI without labyrinthectomy; Group 2 (N = 6 ears) had simultaneous CI and labyrinthectomy; Group 3 (N = 3 ears) had CI with prolonged delayed after labyrinthectomy.

Figures were created using combination of SPSS and CorelDRAW software (CorelDRAW X6 for Windows, Corel Corporation of Ottawa, Canada).

RESULTS

Hearing Outcomes

Hearing and speech outcomes are summarized in Table 2. The majority of ears (24 of 31) achieved speech scores above 90% by 12 months post-CI (BKB or CUNY), irrespective of group. A further 4 of 31 achieved between 80 and 90% and 2 of 31 between 70 and 80%. Statistical analysis, though conducted was viewed in the context of a sample size in Group 2 (n = 6) and especially Group 3 (n = 3) but essentially showed no difference in hearing results with variables such as age at implantation, sex, electrode choice, MD characteristics, or preoperative interventions (gentamicin, endolymphatic sac surgery, or intratympanic steroid use).

Group 1 had 13 men and 9 women patients with mean age of 62.7 (Table 1). Majority of ears were asymptomatic at time of implantation (n = 16 of 22), with two confirmed burnt-out and 14 assumed to be in remission (Table 1). The remaining ears were symptomatic (n = 6 of 22) but not appropriate for labyrinthectomy—three had cochlea symptoms only with fluctuating hearing loss and three had only occasional vertigo with two out of these patients having poor contralateral vestibular function due to MD. The majority of ears had bilateral MD (n = 19 of 22). All bilateral CIs in the total sample were only present in Group 1 which included only five patients. Of these, one patient only had unilateral MD (Patient 16, Fig. 1). His contralateral ear had prolonged hearing loss, due to mumps. The contralateral ear was implanted but data regarding that ear was noted but not analyzed in this data set as the ear was not affected with MD.

There was one patient in this group who was implanted despite having preoperative speech scores above the candidacy criteria. This patient (patient 5, Fig. 1) had bilateral MD with aided speech scores of 48% in his left ear which had burnt out MD. The right ear with best aided speech of 82% fluctuated so much due to active MD, that the patient found it very difficult to attain a functional hearing outcome overall with optimized hearing aids. Frustrated with his condition, he was being worked up for a left sided CI but preoperative caloric testing surprisingly was non-responsive in the right and normal in the left ear. In essence the left ear, the stable ear with burnt out MD and worse hearing was providing the patient his main or arguably only vestibular function. After careful and lengthy consideration and mainly due to the frequent fluctuating nature of the right ear which was rendering hearing aid use difficult in any case, the patient's right ear was chosen to be implanted weighed against the risk of bilateral vestibular failure. This was in contrast to the usual candidacy criteria for CI which considers only the audiological criteria. The patient attained stable hearing and not surprisingly attained 100% speech discrimination. He requested a CI for his second ear postoperatively but his surgeon declined this due to the small but possible risk of bilateral vestibular failure.

Group 2 had four men and two women with a mean of 65.6 years (Table 1). There was a far higher incidence of intractable vertigo and clinical indication for labyrinthectomy, with all ears experiencing vertigo on a daily or weekly basis and/or drop attacks (Table 1). One patient underwent labyrinthectomy despite good preoperative hearing as the patient was debilitated by symptoms of daily vertigo and wanted permanent relief without the risks of an intracranial operation such as vestibular nerve section. All labyrinthectomy performed were in the worse hearing ear. The majority of ears in this group had unilateral MD only (n = 5 of 6). Thus, some of the management options were less complicated than that experienced in Group 1 where there was a high proportion of patients with bilateral MD (n = 19 of 22) (Table 1).

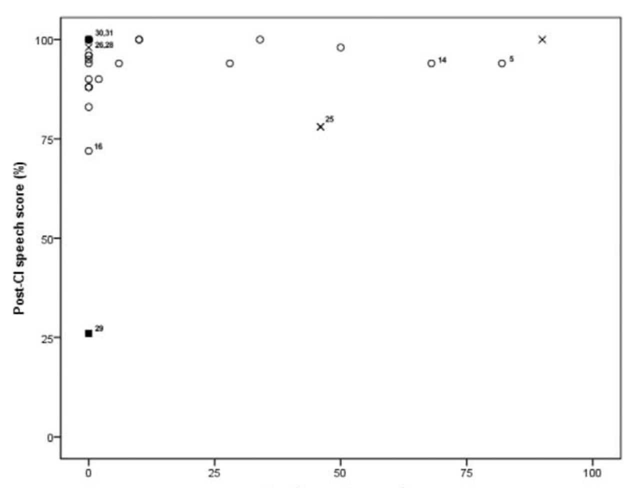

FIG. 1. Within-subject improvement of speech recognition scores for each intervention group. Group 1 results marked by circles; Group 2 results marked by crosses; and Group 3 results marked with solid squares. Pre-CI scores on *x* axis and post-CI scores on *y* axis.

In Group 3, which consisted of three ears (all men) mean age 71.6 years (Table 1), all patients had patent cochlear duct's on preoperative MRI and no intraoperative difficulties were reported with insertion though each surgeon chose modiolar hugging electrodes with stylets (CE24RECA, Cochlear Ltd, Sydney, Australia). Two patients (30 and 31, Fig. 1) attained significant improvement in speech recognition despite a 9 and 11 years delay between labyrinthectomy and CI, respectively (displayed in Fig. 1 with solid squares). One patient (29, Fig. 1) had the poorest outcome in the sample and was the only ear to require supplemental lip-reading with CI, despite only 2-year delay between labyrinthectomy and CI. The patient had undergone five applications of gentamicin before labyrinthectomy, though it was noted that in the rest of the sample there were four patients in Group 1 and one patient in Group 2 who also underwent gentamicin preoperatively and attained speech scores greater than 90%. Before labyrinthectomy, this patient's duration of hearing loss was of 20 years and she had a history of consistent hearing aid use. Furthermore, intraoperative records noted no resistance to electrode insertion suggesting fibrosis.

In addition, there were two other patients who seemed to have worse than expected hearing results compared with the rest of the cohort in Group 1 and 2, respectively. In Group 1, one patient (16, Fig. 1) had extremely difficult intraoperative anatomy and consequently underwent partial electrode insertion. His preoperative aided speech scores of zero improved to 72%. His contralateral ear which had prolonged hearing loss secondary to mumps had previously been implanted and functioned at 93% speech discrimination (though as previously noted these results have not been included in the study as the ear was not affected with MD). The second patient who was an outlier was in Group 2 (25, Fig. 1). He had a preoperative aided speech score of 46% in his ipsilateral ear experiencing daily vertigo. He had a 65-year history of hearing loss in that ear. His contralateral ear was not affected with MD but it was aided and had a PTA of 59 dB. At 3 months after CI and labyrinthectomy, his speech scores were 84%. However, due to an issue with the magnet, he did not efficiently retain the implant and with inconsistent use, his speech scores deteriorated to 49% at 6 months. He did not seek advice for it but nevertheless when the problem was noted at follow up, it was easily remedied with a magnet change. His 12 months score consequently improved to 78% and it is hoped that he may improve further. It must be noted that in this sample he has the longest duration of deafness in the ipsilateral ear and before implantation had minimal hearing aid use. This may be a confounding variable which may also contribute to his poor speech score.

Fluctuating Hearing and Speech Recognition following CI

Eight of 31 ears (26%) reported fluctuating symptoms that localized to ipsilateral ear during 12 months post switch-on period: three (10%) had cochlear only symptoms, and five (16%) had combined cochlear and vestibular symptoms. Majority (five of eight ears) had objective fluctuating CI electrode impedances and mapping (MAPs). Two ears reported worsened tinnitus in ipsilateral ear post-CI but no fluctuation.

Vestibular Outcomes

As is to be expected, no patients in Group 2 or 3 experienced any residual vestibular dysfunction in their operated ear. However, 12 of 22 ears (55%) in Group 1, reported vestibular disturbance post-CI and were able to localize symptoms to the ipsilateral ear. Four had acute vertigo or unsteadiness that resolved in less than 2 weeks, two had Benign Paroxysmal Positional Vertigo (BPPV), two had constant vestibular disturbance, two had recurrent non-positional vertigo, and two had delayed-onset non-positional vertigo many months after surgery. In this group, one patient had a particularly poor vestibular outcome. Aged 50, the patient had bilateral MD causing bilateral hearing loss. His worse hearing ear (right) with zero percent aided speech discrimination was non-responsive to caloric testing and was chosen to be implanted with a CI24R-EST (Cochlear Ltd., Sydney). He had a good outcome with postoperative speech scores of 90% and minimal vestibular dysfunction. The contralateral ear also deteriorated over time from severe to profound loss with zero percent aided speech scores and weak responses on caloric testing and he underwent a second implant with a CI24RECA (Cochlear Ltd., Sydney). However, either due to underlying MD or due to an effect of the CI, the patient experienced symptoms of bilateral vestibular failure and oscillopsia. After 3 years, he began experiencing symptoms of active MD with fluctuating impedances on his CI and also active vertigo. He reported a poor quality of life (QOL) was unable to work and lived on a disability pension. Being a retrospective study and in the absence of any preoperative QOL scores, postoperative retrospective QOL measures, though clearly desirable, would not add meaningful data and, therefore, were not measured as part of this study, though the patient symptomatically reported a poor QoL due to the vertigo.

DISCUSSION

The major finding of the current study was that most MD ears achieved overall excellent hearing benefit with CI as measured by speech recognition tests but managing their hearing loss against their vestibular dysfunction proved challenging in some patients. This finding supports early histological research that predicted good potential benefit from CI in labyrinthectomised ears (7,8). In patients with unilateral MD who required labyrinthecomy, there was no ongoing vestibular dysfunction and focus could be placed largely on auditory rehabilitation. However, labyrinthectomy cannot be considered in all patients and should be considered with caution especially in patients with bilateral MD as it is especially these patients that can prove to be challenging management dilemmas.

Hearing Results

A minority of patients who did not attain good hearing results, had other confounding variables as previously discussed which may affect the general CI population (19), except one patient in Group 3 (29, Fig. 1) whose poor outcome couldn't be adequately explained. However, since labyrinthetomy is an uncommon indication, even with a multicentre study, the sample size especially in the groups undergoing labyrinthectomy was small. This affected the power of the study. Furthermore, a ceiling effect was reached with high postoperative speech scores. As such, it is likely that in these subgroups of patients, further exploration of speech including assessment in noise would help explore subtle differences in these patient's in future prospective studies. This was not performed uniformly across different clinics and was, therefore, difficult to comment on as a group from our data. Furthermore, two patients in the total sample (one in Group 1 and one in Group 2) underwent CI with preoperative aided speech scores above 70% for reasons discussed above. As such their postoperative results introduce a bias in the data since they would be normally be expected to do well. Finally, due to a small sample size particularly in Group 2 and 3 and low power, definitive conclusions are difficult to be made about differences in results in each group, but the cohort has nevertheless been subdivided into three groups as each group poses different variables and management challenges and it is useful to focus on them separately.

Predictive Preoperative Variables Affecting Hearing Outcome

Age at implantation and sex did not affect hearing outcome in the current study. Only a limited number of studies in MD patients have controlled for age, with mixed results (12,17). In the general population, results are also mixed with some large studies showing a significant age effect (19,20) and others not, advocating for CI even in the very elderly (21). CI characteristics (unilateral or bilateral, implant type) and MD characteristics (stage, symptoms, and frequency) also did not affect hearing outcome in this study. Other trends noted were that a history of previous intervention including gentamicin, endolymphatic sac surgery, and intratympanic steroids also did not affect hearing outcome which is consistent with other studies (4,12,22).

Issues for Consideration in Balancing Vestibular Dysfunction and CI Outcome

Group 1—CI only

In Group 1, 12 patients reported vestibular disturbance that localized to ipsilateral ear post-CI. Of these, six of these were short-term and recovered but four reported less common vestibular disturbance: recurrent or non-positional vertigo not related to CI stimulation or malfunction (confirmed by scans and integrity checks) and a further two had a significant delay to onset of vertigo, which the authors cannot explain other than possible changes to underlying physiology in cochlea associated with MD.

Current candidacy criteria and clinical practice focuses heavily, if not exclusively, on hearing and auditory function when selecting which ear to implant. In our sample, one patient had bilateral MD and bilateral CI and experienced chronic oscillopsia. In contrast, after prolonged consideration, another patient was deliberately implanted in the better hearing but worse balance ear based on pre-op VFT results, which resulted in excellent hearing outcomes and no vestibular disturbance. Mick et al. (17) reported adverse vestibular outcome when the ear with better vestibular function and worse hearing was chosen for implantation and support the approach of selecting the worse balance ear in patients with bilateral hearing loss especially if both ears fulfil candidacy for CI in the MD cohort.

Mick et al. (17) also reported in a QoL assessment using a SF36 score conducted on a sample of 20 patients that patients undergoing CI in MD reported more postoperative chronic dizziness than non-Ménière's patients. They hypothesized that this could be either due to ongoing disease or that patient's with MD maybe more sensitive to even mild variation in residual dysfunction. Thus, when suffering with a vestibular destructive disease and without yet having concrete evidence whether CI can cause vestibular dysfunction in Ménière's ears, the decision to choose bilateral CI in bilateral Ménière's disease for benefits of binaural hearing needs to be weighed up against the potential loss of QoL with bilateral vestibular failure.

Another, possibly related finding from Group 1 was that of fluctuating aural symptoms post-CI. These patients experienced objective fluctuations in their CI electrode impedances and MAPs in the ipsilateral ear and the details of this phenomenon has been published recently by the authors (23). Patients with fluctuation required more frequent MAPs and audiological review over 12-month postoperative period; however, once MAPs were adjusted, patients continued to experience good hearing with CI. Patient-reported hearing fluctuation and distorted speech has been previously reported in MD population, with incidence ranging from 33 to 50% (2,4). However, further research is required to increase our understanding and awareness of this phenomenon in the MD population.

Group 2 and 3—CI and Labyrinthectomy

Patients in Group 2 achieved good hearing outcomes while receiving permanent relief of vertigo. In our cohort, candidacy for the affected ear was decided on a case-by-case basis, largely depending on the frequency of vertigo, drop attacks, and disability which made them a candidate for a labyrinthectomy. Therefore, many patients had heterogeneous audiological function in the affected ear though only one patient had aided speech function that did not also independently satisfy the criteria for CI. Clinically, caution is often exercised when considering labyrinthectomy due to the risk of developing bilateral MD in the future in a young patient or poor compensation which may be expected from deafferentation in an elderly patient, but ultimately the decision for labyrinthectomy needs to be undertaken on a case-by-case basis. It is easier to make this decision in the setting of unilateral MD and in that setting provided there is good contralateral vestibular function, good hearing, and balance outcomes are largely attained.

Patient's undergoing delayed implantation may yield good hearing outcomes with CI, though it must be noted that the sample size in this study was very small and candidacy assessment included cochlear duct patency on preoperative MRI scan as the obvious concern in this group was of intracochlear fibrosis or ossification precluding CI. Two recent studies investigated fibrosis in the cochlea following labyrinthectomy. The first, a parallel animal study showed that mild fibrosis can occur in the cochlea as early as 6 weeks in post-labyrinthectomy guinea pigs (24). The second study reported fibrosis in the cochlea of approximately one-third of human patients following labyrinthectomy, although time elapsed after labyrinthectomy did not correlate with severity of fibrosis or ossification (25). Therefore, it is conceivable that CI if delayed after labyrinthectomy may preclude a population of patients who may undergo fibrosis or ossification and, therefore, it is preferable if there are no other contraindications to consider candidacy assessment for the patient at the same time as making decisions about labyrinthectomy (6) thereby also precluding the need for a second operation (6).

A criticism of this study was that it was noted that preoperative VFT was not performed in all cases undergoing labyrinthectomy (two of six ears did not have vestibular assessment), presumably since the severity of the symptoms were such that VFT would not alter the surgical management or perhaps due to a potential delay to accessing VFT. In addition, postoperative vestibular function tests are usually not indicated unless there are symptoms. Thus, vestibular effects of CI in MD can be difficult to quantitate in a retrospective study. Patients who report subjective imbalance may not necessarily have abnormal vestibular function tests and there may still be patients who have no change to their VFT but complain of subjective symptoms and it is in those patients particularly where a QOL measure is relevant. Though these values could not be attained due to the retrospective nature of this study, ideally VFT results in all patients pre and postoperatively and QOL measures would have provided a useful insight into patient's subjective and objective reports of vertigo before and after the surgery. Therefore, authors recommend the more judicious use of preoperative VFT in all cases to quantify vestibular function in the contralateral ear, even if it doesn't alter surgical indication of the affected ear. This allows better preoperative counselling of the patient regarding the likelihood of vestibular rehabilitation and even earlier intervention by preoperative commencement of vestibular physiotherapy. Postoperative VFT should be considered if there are reports of vestibular disturbance to document any objective alteration of function. In addition, QOL measures should also be routinely considered before and after surgery to shed light on balancing the audiological and vestibular needs and goals in this complex group of patients.

CONCLUSION

Patients with MD receive overall excellent hearing benefit from CI both with and without labyrinthectomy. Good outcome is possible even with a significant delay between CI and labyrinthectomy, although there may be a risk of cochlear fibrosis and ossification precluding optimal benefit from CI.

Vestibular function tests should be routinely attained in all patients with MD undergoing CI with or without a labyrinthectomy. The effect of CI on vestibular function is unclear in MD and QOL assessments should be prospectively considered in patients with MD undergoing CI as their postoperative outcome is not only determined by audiological parameters but also colored by their own experience of vestibular function which is systemically difficult to quantify and can be multifactorial. Nevertheless, observations made from this study include that caution must be exercised in two scenarios in CI patients with bilateral MD: firstly when considering implantation of the ear with better vestibular function especially if both ears are audiological candidates for CI and secondly when considering bilateral CI in patients with bilateral MD. More research in this area is required and the benefits of binaural hearing need to be weighed against the risk of bilateral vestibular failure.

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REFERENCES

1. Minor LB, Schessel DA, Carey JP. Ménière's disease. *Curr Opin Neurol* 2004;17:9–16.
2. Lustig LR, Lalwani A. The history of Meniere's disease. *Otolaryngol Clin North Am* 1997;30:917–45.
3. Huppert D, Strupp M, Brandt T. Long-term course of Meniere's disease revisited. *Acta Otolaryngol* 2010;130:644–51.
4. Fife TD. Meniere's syndrome. *Curr Treat Options Neurol* 1999;1:57–67.
5. Wareing M, O'Connor AF. The role of labyrinthectomy and cochlear implantation. *Ear Nose Throat J* 1997;76:664–9.
6. Hansen M, Gantz B, Dunn C. Outcomes after cochlear implantation for patients with single-sided deafness, including those with recalcitrant Ménière's disease. *Otol Neurotol* 2013;34:1681–7.
7. Otte J, Schunknecht HF, Kerr AG. Ganglion cell populations in normal and pathological human cochleae. Implications for cochlear implantation. *Laryngoscope* 1978;88:1231–46.

8. Chen D, Linthicum JR, Rizer F. Cochlear histopathology in the labyrinthectomised ear: implications for cochlear implantation. *Laryngoscope* 1988;98:1170–2.
9. Lustig LR, Yeagle J, Niparko JK, et al. Cochlear implantation in patients with bilateral Ménière's Syndrome. *Otol Neurotol* 2003;24:397–403.
10. Holden LK, Neely LG, Gotter BD, et al. Sequential bilateral cochlear implantation in a patient with bilateral Meniere's disease. *J Am Acad Audiol* 2012;23:256–68.
11. Fife TA, Lewis MP, May JS, et al. Cochlear Implantation in Ménière's Disease. *JAMA Otolaryngol Head Neck Surg* 2014;140:535–9.
12. Samy RN, Houston L, Scott M, et al. Cochlear implantation in patients with Meniere's disease. *Cochlear Implants Int* 2015;16:208–12.
13. McRackan TR, Gifford RH, Kahue CN, et al. Cochlear implantation in Meniere's disease patients. *Otol Neurotol* 2014;35:421–5.
14. Osborn HA, Yeung R, Lin VY. Delayed cochlear implantation after surgical labyrinthectomy. *J Layrngol Otol* 2012;126: 63–5.
15. Zwolan TA1. Shepard NT, Niparko JK. Labyrinthectomy with cochlear implantation. *Am J Otol* 1993;14:220–3.
16. MacKeith SA, Bottrill LD, Ramsden JD. Simultaneous labyrinthectomy with cochlear implantation in patients with bilateral Ménière's disease. *Ann Otol Rhinol Laryngol* 2014;123: 485–9.
17. Mick P, Amoodi H, Arnoldner C, et al. Cochlear implantation in patients with advanced Ménière's disease. *Otol Neurotol* 2014;35: 1172–8.
18. Heywood RL, Atlas MD. Simultaneous cochlear implantation and labyrinthectomy for advanced Ménière's disease. *J Laryngol Otol* 2016;130:204–6.
19. Holden LK, Finley CC, Firszt JB, Brenner C, et al. Factors affecting open-set word recognition in adults with cochlear implants. *Ear Hear* 2013;34:342–60.
20. Blamey P, Artieres F, Başkent D, Bergeron F, et al. Factors affecting auditory performances of postlinguistically deaf adults using cochlear implants: an update with 2251 patients. *Audiol Neurotol* 2013;18:36–47.
21. Wong DJ, Moran M, O'Leary SJ. Outcomes after cochlear implantation in the very elderly. *Otol Neurotol* 2016;37:46–51.
22. Morgan M, Flood L, Hawthorne M, Raje S. Chemical labyrinthectomy and cochlear implantation for Menière's disease—an effective treatment or a last resort? *J Laryngol Otol* 1999;113:666–9.
23. McNeill C, Eykamp K. Cochlear implant impedance fluctuation in Meniere's disease: a case study. *Otol Neurotol* 2016;37:873–7.
24. Brown DJ, Mukherjee P, Pastras C, Gibson WPR, Curthoys IS. Sensitivity of the cochlear nerve to acoustic and electrical stimulation months after a vestibular labyrinthecomy in guinea pigs. *Hear Res* 2016;335:18–24.
25. Charlett SD, Biggs N. The prevalence of cochlear obliteration after labyrinthectomy using magnetic resonance imaging and the implications for cochlear implantation. *Otol Neurotol* 2015;36: 1328–30.