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Is cochlear implantation an effective treatment for Menière's disease?

K. Vermeire[1,2], L. Van Yper[1], E. De Vel[1] and I. Dhooge[1]

[1]University Department of Otorhinolaryngology and Head and Neck Surgery, Ghent University Hospital, Ghent University, Belgium; [2]Thomas More University College, Speech-Language Therapy and Audiology, Antwerpen, België

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Abstract. *Is cochlear implantation an effective treatment for Menière's disease? Objectives*: describe the results of a retrospective study of cochlear implantation (CI) in seven subjects with Menière's disease.
Methodology: The subjects received either the Nucleus CI24RE(CA)/CI512 or the Advanced Bionics HiRes90K CI systems which use the ACE, MP3000, or HiRes S Fidelity 120 coding strategies. The audiometric measures included monosyllabic word lists (NVA) in quiet at 65 dB SPL and sentences in noise (LIST) at +10 dB SNR. The quality of life after implantation was assessed by means of the Nijmegen Cochlear Implant Questionnaire (NCIQ).
Results: After CI, the hearing of all subjects improved significantly ($p<0.001$) as did their speech recognition ($p=0.018$). Speech recognition in noise showed a mean improvement of 47%. The results were less clear for the treatment of vertigo associated with Menière's, as some patients continued to have vestibular attacks after implantation. On the NCIQ, subjects reported a mean quality of life after CI of 48.3%.
Conclusions: This study clearly demonstrates that cochlear implantation is an adequate treatment of speech perception for subjects with Menière's disease who go on to develop bilateral severe to profound sensorineural hearing loss.

Introduction

Menière's disease is an idiopathic inner-ear disorder.[1] It is characterized by episodes of vertigo, roaring tinnitus, fluctuating sensorineural hearing loss, and a sense of aural fullness in the affected ear. These symptoms, or a combination of some of these symptoms, can fluctuate over periods of months or even years.[2] A diagnosis of Menière's disease is made using the clinical guidelines of the American Academy of Otolaryngology-Head and Neck Surgery Committee on Hearing and Equilibrium (AAO-HNS CHE); these guidelines are based on a checklist of signs and symptoms.[3] The reported prevalence rate for Menière's disease ranges from 3.5 per 100,000 to 513 per 100,000.[4]

Our understanding of Menière's disease has changed in recent years. For decades, the condition was thought to be caused by excessive fluid retention (hydrops) in the endolymphatic spaces of the inner ear that led to tearing or rupture of the membranous structures that affect hearing and balance. More recent research has shown that hydrops is not always associated with Menière's disease and should thus not be considered the ultimate cause of its symptoms. Current theories are based on the notion that Menière's disease may not have a single cause but may be the common endpoint of a variety of anatomic or physiologic conditions, including ischemic or even autoimmune injuries.[5]

Providing appropriate amplification to a patient with Menière's disease presents several challenges. These challenges include the patient's fluctuating hearing loss, a rising audiometric configuration, unilateral or asymmetrical hearing loss, reduced dynamic range, and reduced word-recognition scores (i.e. patients' word-recognition scores are lower than predicted based on the magnitude and configuration of the hearing loss). The presence of any one of these characteristics can create obstacles for a successful hearing aid fit.[6]

Development of bilateral severe-to-profound hearing loss secondary to bilateral Menière's disease is relatively rare, but should it occur, cochlear implantation (CI) may be an option because of the difficulties associated with fitting the patient with a hearing aid. To date, there are limited reports of CI in Menière's patients, and these are mostly case reports.[7-9] Morgan and colleagues[7] presented a case of severe debilitating vertigo due to Ménière's disease that was treated by chemical labyrinthectomy and CI. This management strategy restored the subject's hearing and provided relief

from vertigo. Lustig *et al.*[8] examined the speech recognition abilities of a group of nine CI recipients with Menière's disease; seven of the nine patients had bilateral Menière's disease. All study patients were implanted unilaterally in the ear with poorer hearing, and all showed significant improvement in their open-set speech recognition scores six months after the initial activation of the CI compared to their preoperative scores. The only exception was one patient whose device failed. The speech recognition abilities of the Menière's group were compared to a group of post-lingual CI recipients without Menière's disease. There were no significant differences in the scores of the two groups except on the CID sentence test in quiet on which the Menière's group scored significantly higher. These results suggest that CI should be considered as an option for patients with severe-to-profound hearing loss secondary to Menière's disease.

Holden *et al.*[9] recently described a case of a 45-year-old woman with bilateral, profound sensorineural hearing loss due to Menière's disease. She received her first cochlear implant in her right ear in 2008 and a second cochlear implant in her left ear in 2010. The results of the speech recognition testing indicated a substantial improvement in the patient's ability to understand speech in noise and her ability to localize sound when using bilateral cochlear implants compared to using a unilateral implant or an implant and a hearing aid. In addition, the patient reported considerable improvement in her ability to communicate in daily life when using bilateral implants versus a unilateral implant.

Here we describe the results of a retrospective study of seven patients with Menière's disease who underwent CI at our institution. Hearing and reported vestibular outcomes, as well as quality of life issues after intervention, are discussed. The study was conducted in accordance with the ethical standards of the committee on human experimentation at Ghent University Hospital and in compliance with the tenants of the Declaration of Helsinki. All patients provided written informed consent.

Materials and methods

Seven patients who fulfilled the AAO-HNS criteria for Menière's disease were recruited at the Ghent University Hospital. Patient characteristics are shown in Table 1. The patients were implanted with the following devices: Nucleus CI512 in two patients (Nucleus Corp., Lane Cove, Australia), Nucleus CI24RE(CA) in two patients (Nucleus Corp.), and HiRES90K in three patients (Advanced Bionics, Valencia, CA, USA). Table 1 also shows the sound coding strategy the patients used and the mean electric dynamic range (DR) throughout the MAP. Audiometric measures included monosyllabic words (the NVA test[10]) in quiet at 65 dB SPL, sentences (the LIST test[11]) in quiet at 65 dB SPL, and noise at +10 dB SNR. Quality of life after

Table 1

Patient characteristics

	Age at implant (years)	**Duration of implant use (years)**	**Time from onset of Ménière symptoms to CI (years)**	**Device type**	**Sound coding strategy**	**Mean dynamic range (in stimulation units)**
S1	63	2	16	Nucleus CI512	ACE/900Hz	48 (±10)
S2	57	2	25	Nucleus CI512	ACE/900Hz	58 (±7)
S3	71	5	19	Nucleus CI24RE(CA)	MP3000	57 (±10)
S4	69	5	19	Nucleus CI24RE(CA)	ACE/900Hz	79 (±7)
S5	70	6	16	AB HiRes90K	HiRes S Fidelity120	204 (±24)
S6	72	6	6	AB HiRes90K	HiRes S Fidelity120	103 (±3)
S7	68	3	23	AB HiRes90K	HiRes S Fidelity120	229 (±59)

Table 2

Audiologic data for speech recognition pre- and post-cochlear implantation (reported as % correct)

Subject	Monosyllabic words				Sentences in quiet				Sentences in noise			
	Pre	6-mo	1-y	2-y	Pre	6-mo	1-y	2-y	Pre	6-mo	1-y	2-y
S1	13	77	76	74				88				30
S2	27	47	58	61				100				30
S3	0		100	85			100				0	
S4	20	35	55	52			50	88				84
S5	30	87		69		100				90		
S6	0	70				80						
S7	16	75		84								

implantation was assessed using the Nijmegen Cochlear Implant Questionnaire (NCIQ).[12] The NCIQ is a self-assessment health-related quality of life (QoL) instrument for use in CI recipients. There are six subdomains: basic sound perception, advanced sound perception, speech production, self-esteem, activity limitations, and social interactions.

Results from the speech reception tests were analyzed using the Wilcoxon matched pairs signed-rank test. Spearman correlation was used to look for correlations between tests.

Results

Hearing outcomes

Patients experienced the symptoms of Menière's disease for an average of 18 years before implantation (range: 6-25 years). Follow-up after CI ranged from 6 months to 2 years (Table 2).

Mean preoperative hearing loss, the mean preoperative Pure Tone Average (PTA; calculated as the average pure tone frequencies of 0.5, 1, 2 and 4 kHz) was 99 dB HL for the implanted ear (range: 88-106 dB HL) and 85 dB HL for the unimplanted ear (range: 66-120 dB). After CI, pure-tone audiometric tests showed a mean aided PTA of 25 dB HL (range: 19-29 dB HL). The improvement in hearing was significant ($p<0.001$). Speech recognition, as measured by means of the monosyllabic NVA word lists at an intensity of 60 dB SPL, before implantation ranged from 0 to 30% (mean: 15%). After at least 6 months of implant use, the speech recognition improved to a mean of 71% (range: 52-85%). The improvement is significant ($p=0.018$). Speech recognition in noise (at +10 dB SNR), measured after implantation (minimum 6 months of implant use), showed a mean result of 47%, with a range between 0 and 90%. The results of the speech in noise tests correlated with the results of the speech recognition test results in quiet, but the correlation was not significant ($p=0.203$).

Results for the NCIQ and its six different subdomains are reported in Table 3. Overall,

Table 3

NCIQ results (reported as %; maximum score, 100%)

Subject	Sound Perception Basic	Speech Production	Sound Perception Advanced	Self-esteem	Activity	Social Interaction	Total score
S1	60	67.5	62.5	67.5	67.5	57.5	63.7
S2	40	95	40	72.5	57.5	62.5	61.2
S3	57.5	62.5	35	45	60	72.5	55.4
S4	32.5	40	15	17.5	17.5	25	24.6
S5	45	72.5	40	47.5	50	35	48.3
S6	42.5	47.5	15	57.5	30	25	36.2
S7							
MEAN	**46.2**	**64.2**	**34.6**	**51.2**	**47.1**	**46.2**	**48.3**

subjects reported a mean quality of life after CI of 48.3%. When analyzing the correlation between the results of the NCIQ and the speech recognition results (i.e. the monosyllabic word test and the sentences in noise test), the correlation was not significant (p=0.329 and p=0.322, respectively).

None of the patients showed fluctuating hearing in the nonimplanted ear. Of the five patients who reported tinnitus before implantation, only one patient (S5) reported persistent tinnitus post-implantation.

Vestibular outcomes

Vestibular outcomes are shown in Table 4. Four of the seven patients were free of vertigo, while two patients reported that they still suffered from vestibular attacks. One patient (S1) reported relief from vestibular problems as long as she continued taking anti-vertigo drugs. One patient (S5) was incapacitated by the vestibular attacks that surfaced after implantation. This patient was lost for longitudinal follow-up.

Discussion

This study clearly demonstrates that CI is a good treatment option for subjects with Menière's disease who develop bilateral severe-to-profound sensorineural hearing loss. Our results are in accordance with those found in other reports on CI in subjects with Menière's disease. Lustig *et al*.[8] reported a mean monosyllabic word score of 53% and a speech recognition in noise score of 68% 1 year post-implantation. These results are not significantly different from those obtained in this study: t-test, p=0.113 for monosyllabic words and p=0.722 for sentences in noise. Other reports in the literature also agree with our finding that CI represents a viable option for patients who have lost their hearing due to Menière's disease.[7-9] Histopathological analysis of the temporal bones in subjects with Menière's disease demonstrates that extensive neuronal degeneration in the spiral ganglion is rare.[13] This finding is supported by the electric DR data in our subjects (Table 1). These DRs are not different from the DRs reported in the literature.[14] These histological findings strongly support the underlying concept of using CI to treat patients with Menière's disease who develop bilateral deafness.

The audiological results in these subjects are also in agreement with the results of CI in groups with different pathological causes. Two studies have shown the beneficial results of CI in late-onset progressive deafness.[15,16] Hildebrand *et al*.[15] reported excellent results in five individuals with DFNA17, and Vermeire *et al*.[16] reported good outcomes in eleven CI users suffering from DFNA9. Otosclerosis is another cause of severe hearing loss for which CI has been shown to be beneficial.[17,18] Muñoz-Fernández and colleagues[17] conducted a retrospective study of 13 CI recipients who suffered from otosclerosis. One year after implantation, the average score on bisyllabic word testing was 80%, while the average score on an open sentence test was 85%. Rotteveel *et al*.[18] analyzed the speech perception of 53 CI recipients with otosclerosis. They found a wide variation in the speech perception scores. The difference between the poor performers and the good performers was due to factors directly related to the disease (e.g. the extent of otosclerosis on the CT scan) and factors indirectly related to the disease (e.g. fewer electrodes due to partial insertion).

Table 4

Menière's symptoms pre- and post-cochlear implantation

Subject	Pre-implantation		Post-implantation (> 6-mo)		
	Vertigo spells	Tinnitus	Vertigo spells	Tinnitus	Hearing fluctuation
S1	Yes	No	Yes	No	No
S2	No	Yes	No	No	No
S3	No	Yes	No	No	No
S4	Yes	No	No	No	No
S5	No	Yes	Yes	Yes	No
S6	No	Yes	No	No	No
S7	Yes	Yes	Yes	No	No

Our NCIQ results show that the subjects reported a satisfactory quality of life after CI, although the NCIQ scores in this study tend to be lower than those reported by other studies.[19-21] The cause of this difference may be the negative effect that Menière's disease has on overall quality of life. Several publications have described the daunting limitations faced by subjects with Menière's disorder.[22-24] Some patients reported that they continued to have vestibular attacks after implantation. Notably, this patient population has significant psychological problems associated with feeling ill due to the nausea that is secondary to the vertiginous attacks and due to the anxiety that stems from the unpredictable nature of the attacks. This finding is in accordance with a study by Neuhauser *et al*.[25] that found that vertigo is associated with a considerable personal burden.

Statistically, there was no correlation between the speech recognition scores and the NCIQ results. This is not in accordance with other studies.[19-21] The lower quality of life scores found in this study may explain the lack of correlation.

Conclusion

In conclusion, patients with bilateral Menière's disease who develop profound bilateral hearing loss or deafness are good candidates for CI. Menière's patients can be expected to perform as well on speech recognition tests as other post-lingually deafened implantees. The results were less clear in terms of the effect of CI on Menière's-associated vertigo, as only four of the seven patients had no complaints about vertigo post-implantation. One patient even complained that incapacitating vertigo began post-implantation.

Because of the discrepancy between the results of the speech recognition tests and the reported quality of life, assessment of CI outcomes should include QOL measures in order to provide a more holistic picture of the results, especially in more challenging patient populations.

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Katrien Vermeire, Ph.D.
Thomas More University College
Speech-Language Therapy and Audiology
Molenstraat 8
2018 Antwerpen, België
Tel: + 32 (0)3 241 08 29
E-mail: Katrien.vermeire@thomasmore.be