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CLINICAL RECORD

Contralateral cochlear implantation prior to vestibular nerve section for 'drop attacks' in the only hearing ear

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Abstract

Background: A dilemma occurs in the treatment of second-sided Ménière's disease in the only hearing ear, particularly in patients with severe symptoms such as 'drop attacks'. This paper describes a patient treated with contralateral cochlear implantation prior to vestibular nerve section of the symptomatic ear.

Case report: A 53-year-old man, with second-sided Ménière's disease and drop attacks in the only serviceable right ear, underwent successful left cochlear implantation 30 years after hearing loss, followed by right vestibular nerve section. The patient achieved control of Ménière's attacks and improved hearing. Although the patient experienced oscillopsia post-operatively, he was satisfied with his improved everyday functioning.

Conclusion: Patients with severe second-sided Ménière's disease in the only hearing ear are a small but difficult treatment group. In those that are suitable for cochlear implantation in the non-serviceable ear, it is suggested that this be employed prior to surgical treatment of the Ménière's symptoms, even if the implanted ear has had no auditory stimulation for many years.

Key words: Meniere Disease; Syncope; Vestibular Nerve; Cochlear Implant; Hearing Loss

Introduction

Ménière's disease is an idiopathic inner-ear disorder characterised by episodic vertigo, fluctuating sensorineural hearing loss, tinnitus and aural fullness.[1] About 3–7 per cent of patients with Ménière's disease develop 'drop attacks', characterised by sudden falls that occur without warning, loss of consciousness and other neurological symptoms.[2–4] In the majority of cases, Ménière's disease is limited to only one ear; however, bilaterality increases with duration of the disease, with up to 35 per cent of patients affected within 10 years and up to 47 per cent within 20 years.[5]

Drop attacks are potentially life threatening, and are frequently treated with labyrinthectomy or vestibular nerve section.[6] A management dilemma occurs when affected patients who have already lost hearing in one ear through their disease or previous treatment develop bilateral Ménière's disease. The potential for bilateral deafness has traditionally precluded destructive surgical intervention.

Cochlear implantation has been used for rehabilitation of severe to profound sensorineural hearing loss for 30 years, and its application has continued to expand.[7] In patients with Ménière's disease, cochlear implantation has been shown to improve receptive communication without adversely affecting vestibular function.[7,8] In addition, there is emerging evidence that delayed cochlear implantation, even that which takes place decades after hearing loss, can be beneficial for some patients.[9,10]

We present a case of successful delayed cochlear implantation of the contralateral ear, prior to vestibular nerve sectioning for the treatment of bilateral Ménière's disease coinciding with disabling drop attacks in the only hearing ear. The case report illustrates a management option in this challenging group of patients.

Case report

A 53-year-old gentleman was initially referred to the senior author's practice with right-sided Ménière's disease, causing fluctuating tinnitus, aural fullness and frequent rotatory vertigo. Despite medical management with high-dose betahistine, amiloride hydrochloride and hydrochlorothiazide, there was no resolution of his symptoms. Four and a half years after the initial presentation, he developed drop attacks. He experienced more than 10 drop attacks over 3 months, and was experiencing attacks of rotatory vertigo daily. He avoided social situations and stopped driving as a result of his illness.

There was a background of long-standing left-sided Ménière's disease and the patient had undergone left endolymphatic sac surgery in 1983; this was complicated by a post-operative 'dead ear'.

Absent left-sided vestibular function was confirmed on vestibular-evoked myogenic potential and caloric testing. Audiometry showed a profound sensorineural hearing loss

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on the left, and a severe and progressive sensorineural loss on the right with a pure tone threshold of 70 dB (Figure 1).

To mitigate the risk of operating on the patient's only hearing ear, the patient was assessed for left cochlear implantation, 30 years after his hearing loss. The audiometric results met the implantation criteria, and the magnetic resonance imaging scan showed normal left cochlear morphology and a normal left cochlear nerve. His pre-operative Bamford–Kowal–Bench score for open-set sentences (using pre-recorded speech) with right aided hearing was 36 per cent.

The patient underwent left cochlear implantation. At the one-month review, with a left cochlear implant and a right hearing aid, he scored 72 per cent on the bimodal Bamford–Kowal–Bench test (conducted using pre-recorded, open-set sentences). His scores for the Bamford–Kowal–Bench test (with open-set sentences) using a live voice with lip reading were 28 per cent for visual cues unaided, 86 per cent for the left cochlear implant alone with visual cues, and 96 per cent for the right hearing aid alone with visual cues.

Two months after cochlear implantation, the patient underwent right vestibular nerve section via a retrolabyrinthine approach. No intra-operative complications were encountered.

Audiometry at one month post vestibular nerve section showed no worsening of his right-sided hearing (Figure 1). The patient reported better hearing in noisy environments and more stable hearing overall. After one month of adjustment to his bilateral vestibular dysfunction and bobbing oscillopsia, he was able to return to work, and recommence driving and travelling overseas. His only functional limitation currently is disequilibrium in the dark. In the eight months post vestibular nerve section, he had no further drop attacks or other Ménière's symptoms.

Discussion

The majority of patients with Ménière's disease are managed conservatively with lifestyle changes and/or medical therapy. If incapacitating symptoms are present despite these measures, invasive treatment options are considered.

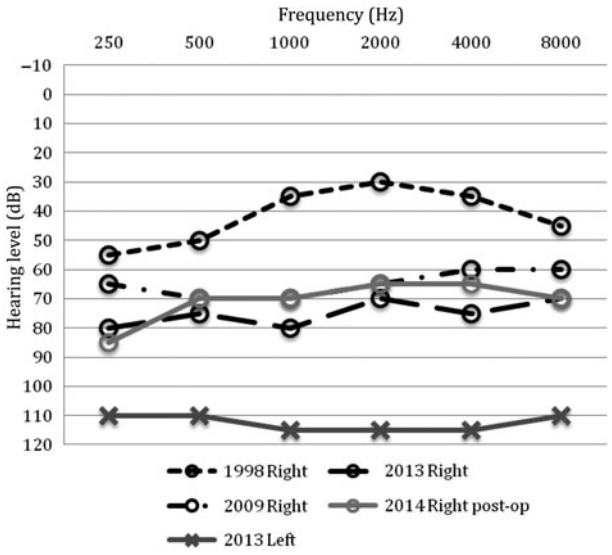

FIG. 1
Serial audiometry results. Post-op = post-operative

However, destructive treatment of an only hearing ear has traditionally been contraindicated.

Drop attacks are potentially life threatening, and are often treated with labyrinthectomy or vestibular nerve section.[6] Recently, intratympanic gentamicin has also been shown to be effective in the treatment of drop attacks.[6] Labyrinthectomy destroys all hearing on the treatment side and is thus reserved for patients who have a serviceable contralateral ear. Although gentamicin is predominantly a vestibulotoxic drug, there is still a real risk to the cochlea, with reports of substantial worsening of hearing in 5–25 per cent of patients. Sensitivity to aminoglycoside-related vestibule or cochlear toxicity varies between patients and is unpredictable.[11,12] Vestibular nerve section, however, is not absolutely contraindicated in the only hearing ear, as the risk of substantial hearing loss is low, at 0–7 per cent.[13–15] However, few would perform it under these circumstances because of the risk of damage to the cochlear nerve, causing deafness that cannot be rehabilitated by cochlear implantation.

With advances in cochlear implant technology, traditional restrictions on surgical management of the only serviceable ear in bilateral Ménière's disease may be lifted. Second-sided Ménière's disease often develops after many years, and there is emerging evidence that a cochlear implant can return hearing in patients, even if carried out decades after hearing loss.[8–10]

Our case report adds to the relevant literature, describing one of the longest reported delays between hearing loss and successful cochlear implantation in Ménière's patients. The findings confirm that delayed cochlear implantation, even when conducted three decades after hearing loss, can be of substantial benefit in selected patients. When combined with contralateral cochlear implantation, vestibular nerve section can be a viable treatment option for patients with Ménière's disease in the only hearing ear.

- **A dilemma occurs in treating severe second-sided Ménière's disease in an only hearing ear**
- **Vestibular nerve section effectively controls 'drop attacks' and vertigo in Ménière's disease, and has the advantage of potential hearing preservation**
- **Advances in cochlear implantation enable the return of hearing after decades of loss**
- **This paper presents a successful case of cochlear implantation in a 'dead ear' prior to vestibular nerve section of the symptomatic ear**

The disadvantage of this treatment strategy is the resulting bilateral vestibular failure, with patients experiencing bobbing oscillopsia and disequilibrium, particularly in the dark.[16] In patients with adequate visual and proprioceptive input, this largely compensates for the vestibular deficits in spatial orientation, posture and balance, and ocular motor control. Our patient was able to largely compensate for the bobbing oscillopsia. Compared to the impairment he experienced from Ménière's disease and drop attacks, he was very satisfied with his functional status post-operatively.

Conclusion

Treatment of severe symptomatic second-sided Ménière's disease in the only serviceable ear is a difficult dilemma.

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For those in whom conservative management has failed, destructive surgical procedures are required to control debilitating symptoms such as potentially life-threatening drop attacks. This is, however, at the expense of significant risk of bilateral hearing loss. We propose that cochlear implantation in the non-serviceable ear prior to vestibular nerve section in the symptomatic ear is a viable treatment option for symptomatic relief and hearing preservation.

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