

# Simultaneous cochlear implantation and labyrinthectomy for advanced Ménière's disease

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## Abstract

**Background:** Patients with Ménière's disease can develop unaidable sensorineural hearing loss. Cochlear implantation has recently been utilised in this group with favourable results. A more challenging group are those with intractable vertigo, and they have traditionally posed a significant management dilemma.

**Case report:** Two female patients with unaidable hearing and recurrent incapacitating vertigo attacks despite conservative management underwent simultaneous labyrinthectomy and cochlear implantation. There was complete resolution of vertigo in both patients. Speech perception in quiet conditions and the ability to hear in background noise improved considerably.

**Conclusion:** Surgical labyrinthectomy is effective for the elimination of vertigo in Ménière's disease patients. The major disadvantage in the past was loss of residual hearing. Cochlear implantation is now an option in these patients. The benefits of simultaneous labyrinthectomy with cochlear implantation include the prevention of implantation of a fibrosed or ossified cochlea, a decrease in the duration of deafness, and a single operative procedure.

**Key words:** Ménière Disease; Cochlear Implants; Vertigo; Hearing Loss

## Introduction

Patients with Ménière's disease can develop unilateral or bilateral unaidable sensorineural hearing loss (SNHL), either as a consequence of the disease itself or its treatment. Cochlear implantation has recently been utilised in this group with favourable results.<sup>1–4</sup> A more challenging group are those with intractable vertigo despite lifestyle measures, medical treatment and simple surgical strategies; these patients have traditionally posed a significant management dilemma.

## Case reports

### Case one

A 63-year-old woman presented with a 3-year history of vertigo attacks with fluctuating, progressive, left-sided hearing loss, aural fullness and tinnitus. The vertigo was unresponsive to a low salt diet, diuretic, betahistine and ventilation tube insertion with a Meniett device. She gained little benefit from her left-sided hearing aid, and a bone-anchored hearing aid trial for single-sided deafness had been subjectively unsuccessful. Neurotological examination was normal apart from an impaired left vestibulo-ocular reflex.

A magnetic resonance imaging (MRI) scan of the temporal bones was normal. Vestibular function tests showed a left canal paresis of 67 per cent and directional preponderance of 46 per cent to the right. Pure tone audiometry showed a moderate-to-severe SNHL in the left ear and a mild high frequency SNHL on the right. Scores for Arthur Boothroyd words, consonant-nucleus-consonant words and

phonemes, and City University of New York ('CUNY') sentences in quiet conditions were all zero in the left ear. Bamford–Kowal–Bench speech-in-noise testing indicated poorer performance in the aided condition compared with that in the unaided condition (+2.5 dB signal-to-noise ratio for 50 per cent correct performance).

Further management strategies were discussed with the patient, including intratympanic gentamicin, vestibular nerve section, labyrinthectomy and cochlear implantation. Simultaneous labyrinthectomy and cochlear implantation (using a Med-El Concerto device with Flex-28 electrode array) were performed. There was complete resolution of vertigo. Speech perception in quiet conditions and the ability to hear in background noise improved considerably post-operatively (consonant-nucleus-consonant words score of 36 per cent and phonemes score of 48 per cent, City University of New York sentences score of 96 per cent, and Bamford–Kowal–Bench speech-in-noise signal-to-noise ratio of –0.5 dB for 50 per cent correct performance, on the left (cochlear implant alone, with the right side masked) at six months).

### Case two

An 84-year-old woman presented with a history of intermittent vertigo episodes spanning many years. The associated bilateral, progressive SNHL had deteriorated significantly in both ears over the previous year. She suffered with intermittent bilateral aural fullness and tinnitus, worse on the right side. Her symptoms were unresponsive to a low salt diet, diuretic and betahistine. She was also visually impaired as

a result of macular degeneration. On examination, the left vestibulo-ocular reflex was reduced. She was able to stand with her eyes closed, but could not perform Unterberger's test. The rest of the neurotological examination findings were normal.

An MRI scan of the temporal bones was normal. Caloric testing indicated symmetrical hypofunction (13° per second bilaterally). Pure tone audiometry demonstrated a moderate-to-severe hearing loss bilaterally. The left ear had deteriorated by 10–15 dB over the previous 18 months. Unaided Arthur Boothroyd word scores were 53 per cent on the right side and 30 per cent on the left side at 100 dB. Consonant-nucleus-consonant word scores in quiet conditions (aided) were 46 per cent on the right side, 5 per cent on the left side and 46 per cent combined. City University of New York sentence scores in quiet conditions (aided) were 81 per cent on the right side, 24 per cent on the left side and 73 per cent combined. Bamford–Kowal–Bench speech-in-noise testing indicated poorer performance in the best-aided condition (+19 dB signal-to-noise ratio for 50 per cent correct performance).

- **Surgical labyrinthectomy in Ménière's disease has traditionally been reserved for patients with non-serviceable hearing because of loss of residual hearing**
- **Cochlear implantation is now an option in these patients**
- **Simultaneous labyrinthectomy with cochlear implantation is rare; it should be advocated in selected Ménière's disease patients**
- **This paper reports two patients who achieved hearing rehabilitation and complete resolution of vertigo**
- **Simultaneous procedure benefits include prevention of fibrosed or ossified cochlear implantation, decreased deafness duration, and a single operative procedure**

Further management strategies were discussed with the patient, including intratympanic gentamicin, vestibular nerve section, labyrinthectomy and cochlear implantation. Simultaneous left labyrinthectomy and cochlear implantation (using a Cochlear® Freedom™ cochlear implant with Contour Advance™ electrode CI24RE (CA)) were performed. The vertigo completely resolved. Speech perception in quiet conditions and the ability to hear in background noise improved considerably post-operatively (consonant-nucleus-consonant words score of 32 per cent and phonemes score of 64 per cent, and City University of New York sentences score of 88 per cent on the left (cochlear implant alone, with the right side masked) at six months).

## Discussion

Hearing in Ménière's disease can be difficult to rehabilitate because of fluctuating hearing loss, unilateral or asymmetrical hearing loss, poor speech discrimination, and a reduced dynamic range. Fortunately, only a small number of Ménière's disease patients go on to develop bilateral severe-to-profound hearing loss; even within a population of those with 'severe' Ménière's disease, only 1 per cent

were considered to be completely deaf.<sup>5</sup> Hearing thresholds 10 years after disease onset were between 61 and 80 dB in 10.9 per cent and worse than 80 dB in only 3.6 per cent of patients in a Japanese study.<sup>6</sup>

The histological effects of Ménière's disease include abnormal stereocilia and outer hair cells. The spiral ganglion cell population (stimulated in cochlear implantation) has not been demonstrated to degenerate.<sup>7,8</sup>

Cochlear implantation has only recently been employed as a method of hearing rehabilitation in patients who develop unaidable SNHL as a consequence of Ménière's disease. Ménière's disease patients may challenge traditional cochlear implantation candidacy criteria because of the fluctuating nature of the hearing loss. Fife *et al.* reviewed 11 cochlear implantations in 10 patients with Ménière's disease and found that audiological outcomes were comparable to those of a non-Ménière's disease population.<sup>2</sup> Mick *et al.* compared cochlear implantation in 20 Ménière's disease patients with a control group.<sup>3</sup> The outcomes were comparable, though the Ménière's disease subjects had significantly more chronic dizziness in the post-operative period. McRacken *et al.* investigated 21 implanted Ménière's disease patients and compared the results to those of 178 implanted adult non-Ménière's disease patients.<sup>1</sup> Implant recipients who had previously undergone ablative procedures for Ménière's disease had significantly better hearing outcomes than those who had received only medical management. Ménière's disease patients in general had worse outcomes after cochlear implantation than the non-Ménière's disease population, but those with active disease had similar outcomes to the non-Ménière's disease population. Lustig *et al.* reviewed nine Ménière's disease patients following cochlear implantation.<sup>4</sup> Hearing outcomes were similar to, or better than, those of a non-Ménière's disease implant population. Some patients experienced fluctuations in their implant performance in association with vestibular symptoms.

Sequential cochlear implantation has been performed following labyrinthectomy in a small number of Ménière's disease patients with reasonable outcomes. Osborn *et al.* reported successful implantation in a patient in whom bilateral labyrinthectomy procedures were performed 21 years previously for intractable Ménière's disease.<sup>9</sup> Her hearing-in-noise test score improved from 0 per cent pre-operatively to 60 per cent at six months. Thedinger *et al.* reported successful implantation in a patient who had undergone an ipsilateral labyrinthectomy 15 years earlier for Ménière's disease and subsequently developed contralateral profound sensorineural deafness due to an acoustic neuroma.<sup>10</sup> As mentioned above, Lustig *et al.* reviewed nine patients with Ménière's disease who had undergone cochlear implantation, one of whom had previously undergone labyrinthectomy.<sup>4</sup> The patient continued to experience fluctuations in hearing associated with aural fullness post-implantation. Audiometric parameters were worse than the average for the group but nevertheless showed improvement.

Surgical labyrinthectomy and vestibular nerve section are the most effective methods for the elimination of vertigo in Ménière's disease patients in whom medical management has failed. Labyrinthectomy obviates the need for an intracranial procedure and requires a shorter operating time than vestibular nerve section. The major disadvantage of labyrinthectomy in the past was loss of residual hearing, and as such this procedure has traditionally been reserved for patients

with non-serviceable hearing. However, cochlear implantation is now an option in these patients. Histological studies demonstrate that the spiral ganglion cell population is reduced but not eliminated by surgical trauma.<sup>11</sup> Three of four post-labyrinthectomy bones studied by Chen *et al.* had ganglion cell populations equal to or greater than the number present in two bones from successful implant users.<sup>12</sup> Kemink *et al.* described post-operative behavioural thresholds similar to intra-operative electrophysiological thresholds for the same stimuli in six post-labyrinthectomy patients, implying the persistence of excitable auditory cells post-labyrinthectomy.<sup>13</sup> A further consideration is the development of cochlear fibrosis and ossification following surgery on the bony labyrinth. In this respect, it seems logical that simultaneous labyrinthectomy with cochlear implantation would pre-empt the difficulties associated with implanting a fibrosed or ossified cochlea.

Simultaneous labyrinthectomy with cochlear implantation has rarely been performed. In 1993, Zwolan *et al.* published a report of simultaneous labyrinthectomy and cochlear implantation in a 27-year-old patient with congenital hearing loss who subsequently developed delayed onset endolymphatic hydrops.<sup>14</sup> The patient was free of vertigo post-operatively, and had improved sound awareness and speech perception one year post cochlear implant activation. Hansen *et al.* recently reported 12 patients who underwent simultaneous labyrinthectomy with cochlear implantation.<sup>15</sup> Ten of these patients had Ménière's disease. Pre- and post-operative audiological data (between 3 and 12 months) were available for 6 patients and showed improvement in 5 of those 6. All patients had complete resolution of vertigo. MacKeith *et al.* described two patients who had undergone simultaneous labyrinthectomy with cochlear implantation for severe second-side Ménière's disease in the only hearing ear.<sup>16</sup> The Ménière's attacks were controlled and hearing was rehabilitated, though both patients noted oscillopsia.

In summary, simultaneous labyrinthectomy with cochlear implantation should be advocated in carefully selected Ménière's disease patients. Here, we report two patients who achieved hearing rehabilitation and complete resolution of vertigo. The benefits of simultaneous procedures include the prevention of implantation of a fibrosed or ossified cochlea and a decrease in the duration of deafness, thereby reducing risk factors for a poorer cochlear implantation outcome. Furthermore, only a single operative procedure is required. Synchronous labyrinthectomy with cochlear implantation may prevent the fluctuations in implant performance associated with vestibular symptoms and enable vestibular rehabilitation of a steady state. Patients with progressive hearing loss and persistent disabling vertigo that is unresponsive to simple measures may therefore benefit from early labyrinthectomy with cochlear implantation, as soon as they meet audiological implant criteria. A

multicentre study would be beneficial in order to assess the long-term outcomes in these complex patients.

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