

## Original Research Article

# Cochlear implantation in Meniere's disease: surgery and outcomes

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## ABSTRACT

**Background:** Sensori-neural hearing loss (SNHL) can be bilateral in 30 to 50% of individuals with Meniere's disease (MD). The incidence of severe to profound SNHL in MD ranges from 1 to 6%. These individuals are candidates for cochlear implantation (CI).

**Methods:** A retrospective study of 12 patients with profound hearing loss due to Meniere's disease was done between January 1998 till June 2018 in a tertiary ENT centre in Chennai, India. The patients experienced symptoms of Meniere's disease for an average of 10 years before CI. All were postlingual candidates. The aim of this study was to determine the effectiveness of cochlear implantation in improving hearing and to assess the impact of CI on vertigo and tinnitus in patients with Meniere's disease.

**Results:** Significant hearing improvement with CI was noted in all patients. Resolution of vertigo and reduction of tinnitus was noted. There was no vestibular dysfunction due to CI. In one patient with disabling vertigo, labyrinthectomy combined with CI helped in complete resolution of vertiginous symptoms.

**Conclusions:** Cochlear implantation is an established option for hearing habilitation in patients with Meniere's disease, with good outcomes. In case there is disabling vertigo, combining labyrinthectomy with CI is recommended.

**Keywords:** Meniere's disease, Profound hearing loss, Cochlear implants

## INTRODUCTION

The indications for cochlear implantation (CI) have expanded in the past few decades. Meniere's disease (MD) is commonly encountered in otolaryngology practice in India. Patients with Meniere's disease may develop bilateral disease. Incidence of severe to profound sensori-neural hearing loss (SNHL) in MD is reported to be 1 to 6%.<sup>1</sup> The hearing loss is usually progressive. Cochlear implants have proved to be an effective management option for profound hearing loss in patients with unilateral or bilateral Meniere's disease. Reduction in tinnitus has also been reported.<sup>2</sup> Labyrinthectomy may be required in patients who have disabling vertigo and

this can be staged or done simultaneously with CI. The aim of this study was to determine the effectiveness of cochlear implantation in improving hearing, vertigo and tinnitus in patients with MD.

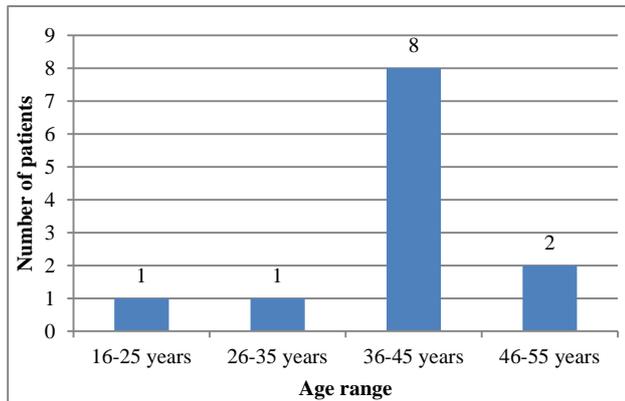
## METHODS

Patients aged 18 years and above with profound hearing loss due to Meniere's disease underwent CI between January 1998 till June 2018 in Madras ENT Research Foundation, a tertiary level ENT centre in Chennai, India. Ethical approval for the study was taken from the Institutional review committee. The inclusion criteria included patients with bilateral profound hearing loss due

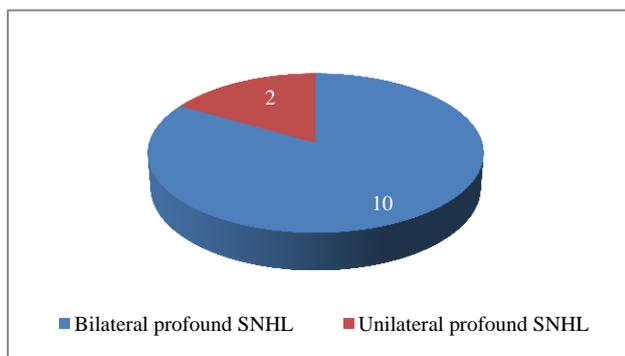
to Meniere's disease. AAO-HNS criteria was used for diagnosis of Meniere's disease.<sup>3</sup> Exclusion criteria included patients with bilateral profound hearing loss due to causes other than Meniere's disease. Profound sensorineural hearing loss was considered an indication for undergoing cochlear implantation. All patients were postlingual candidates. All patients underwent comprehensive audiological and electrophysiological tests, vestibular function tests with videooculography and Vestibular evoked myogenic potentials and CT/MRI of the inner ear. In all patients, hearing aid trial was done and patients did not benefit with amplification. All patients underwent cochlear implantation. Categories of Auditory Performance (CAP) score was used to assess the benefit obtained from cochlear implantation.<sup>4</sup> History of vertigo and tinnitus pre-operatively and post-operatively were recorded. For the statistical analysis, Chi square test and Student's t test were used. Follow up after CI ranged from 12 months to 16 years and the mean follow-up period was 36 months.

**RESULTS**

Twelve patients with profound hearing loss due to Meniere's disease underwent cochlear implantation. There were 9 men and three women with an average age of 41 years and age range from 18 to 50 years (Figure 1).



**Figure 1: Age range of patients with MD who underwent CI (N=12).**



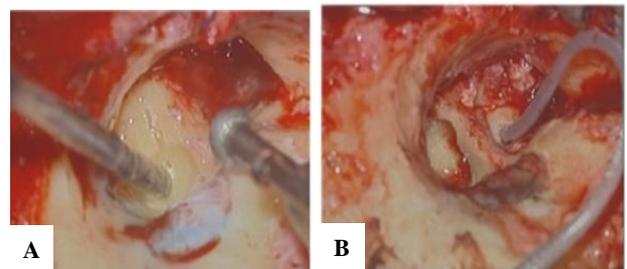
**Figure 2: Number of patients with bilateral / unilateral profound hearing loss due to MD.**

The patients experienced symptoms of Meniere's disease for an average of 10 years before CI. Ten patients had bilateral profound hearing loss and two patients had unilateral profound hearing loss (Figure 2).



**Figure 3: Cochlear implant in a patient with Meniere's disease.**

Vertigo was reported by 3 patients prior to CI; one of whom had incapacitating vertigo. Disabling tinnitus was present in 3 patients pre-operatively. None of the patients had a previous surgical procedure for Meniere's disease. All patients underwent unilateral cochlear implantation (Figure 3) with Med-El cochlear implant. A Med-EL straight electrode array was inserted via the round window in all patients. Complete electrode insertion was achieved in all patients.

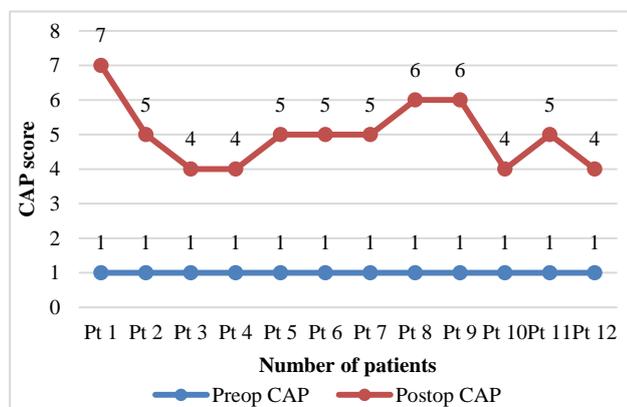


**Figure 4: (A) Simultaneous labyrinthectomy with CI – labyrinthectomy in progress; (B) simultaneous labyrinthectomy with CI – cochlear implant in situ.**

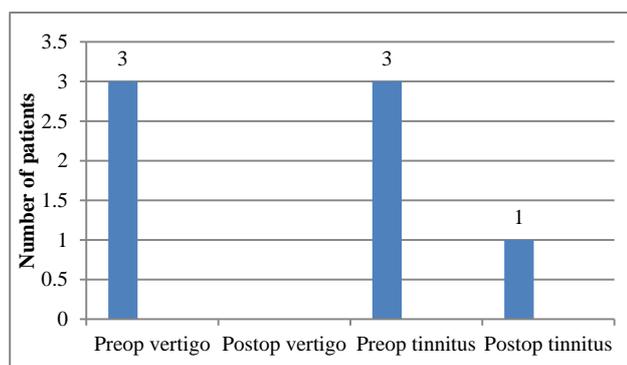
One patient who had disabling vertigo unresponsive to medical management underwent simultaneous labyrinthectomy and cochlear implantation (Figure 4a and b).

No surgical complications were noted. The mean pre-operative pure tone audiometry (PTA) level in our patients was 90 dBHL. Post cochlear implantation, the mean aided PTA was 35 dB HL. The categories of auditory perception (CAP) score were used to assess the benefit obtained from cochlear implantation. The mean preoperative CAP score was 1. The mean post-operative

CAP score value was 2 (range 1-3) at 6 months and improved to 5 (range 4-6) at 12 months which was statistically significant ( $p < 0.00001$ ) and confirmed that CI is beneficial in improving hearing in Meniere's disease (Figure 5).



**Figure 5: Pre-operative and post-operative CAP scores in MD.**



**Figure 6: Pre-operative and post-operative vertigo and tinnitus in MD.**

Vertigo was reported by 3 patients prior to CI. Post CI, none of the patients experienced vertigo ( $p < 0.05$ ). Disabling tinnitus was present in 3 patients pre-operatively. CI helped in resolution of tinnitus in two patients ( $p < 0.05$ ) (Figure 6). The p value as shown above is significant.

## DISCUSSION

Meniere's disease is characterized by fluctuating hearing loss, episodic vertigo, tinnitus and aural fullness. The prevalence of MD is reported to be about 0.2% and bilateral involvement occurs in 30 to 50%.<sup>1</sup> The disease has a deleterious effect on hearing and balance function. The incidence of severe to profound sensori-neural hearing loss in MD is 1 to 6%, and usually the hearing loss is progressive in nature. The etiology of Meniere's disease is unknown; however, pathological examination demonstrates endolymphatic hydrops, in addition to damage of the inner and outer hair cells with preservation of spiral ganglion cells.<sup>5</sup>

Cochlear implantation is an established option for hearing habilitation in patients with profound hearing loss and is effective for patients with Meniere's disease. Significant hearing improvement occurs with CI. Fluctuation in hearing performance during acute attacks has been reported and is seen in approximately one third of patients. Fluctuation in hearing during an acute attack is due to physical changes relative to the implant array with the spiral ganglion neurons or changes in spiral ganglion neurons because of the hydropic state.<sup>6</sup> During an acute attack, the ability to hear with a cochlear implant may decrease, sound may become very high pitched, there may be intolerance to sound, distortion of sound, or tinnitus. The hearing returns to the baseline following resolution of acute attacks. Fluctuations in hearing can be overcome by CI reprogramming.

Very long duration of severe - profound bilateral hearing loss and previous MD treatment (intratympanic gentamicin, endolymphatic sac surgery and labyrinthectomy) are factors influencing CI outcomes in MD. Patients undergoing surgical or ablative procedures for their MD symptoms have statistically significant improvement in their CI hearing outcomes compared with those who do not.<sup>7</sup> Implantation does not seem to adversely alter the natural history of vestibular function.<sup>1</sup> Beneficial effect of CI on tinnitus has been noted in most patients because CI addresses the changes in neural activity caused by reduced auditory input, thought to be responsible for tinnitus.<sup>1</sup> Occasionally, there may be persistent tinnitus after cochlear implantation.

Surgical labyrinthectomy is effective for eliminating vertigo in patients with Meniere's disease. Combined labyrinthectomy and CI are done in patients with intractable vertigo and profound hearing loss due to Meniere's disease.<sup>2</sup> CI is done simultaneously with labyrinthectomy or as a second stage procedure. The benefits of simultaneous labyrinthectomy and cochlear implantation include prevention of implantation of a fibrosed or ossified cochlea, a decrease in the duration of deafness and a single operative procedure.<sup>8</sup> During simultaneous labyrinthectomy and cochlear implantation, the round window is identified and exposed through a posterior tympanotomy before undertaking labyrinthectomy. Good improvement in hearing and reduction of vertigo and tinnitus have been reported.

In our study, there were no surgical difficulties encountered. Hearing outcomes were satisfactory in all 12 patients. Vertigo was controlled in all three patients with pre-operative vertigo. Tinnitus was controlled in two out of three patients with pre-operative symptoms.

In a study by Mukherjee et al, patient age and sex, previous treatment of MD and audiological test results did not significantly affect outcomes.<sup>9</sup> It was reported that CI in MD can yield good hearing outcomes in patients with intact labyrinths, CI with simultaneous labyrinthectomy and in CI sequential to surgical

labyrinthectomy and this is possible even after a long delay after labyrinthectomy. One disadvantage of simultaneous labyrinthectomy and CI is that patients with residual hearing preoperatively will not experience the full consequences of single sided deafness and may not fully appreciate the benefit of the cochlear implant.<sup>10</sup> In a study by Vermeire et al, after CI, the hearing of all seven subjects improved significantly.<sup>11</sup> In a study by Heywood et al, in two patients with simultaneous labyrinthectomy and cochlear implantation, there was complete resolution of vertigo; speech perception in quiet and ability to hear in background noise improved considerably.<sup>7</sup> In a study by Mick et al, at 1 year after CI, improvements in sentence and word understanding in twenty patients with MD did not differ in magnitude from the controls. Tinnitus was reduced significantly in patients with MD. Subjects with MD had significantly more chronic dizziness in the postoperative period than did controls.<sup>12</sup> In a study by MacKeith et al, two patients with bilateral disease were managed by simultaneous surgical labyrinthectomy and cochlear implantation. Both patients achieved control of Meniere's attacks with improved hearing rehabilitation.<sup>13</sup> In a study by Perkins et al, subjects with unilateral Meniere's Disease who underwent simultaneous labyrinthectomy and cochlear implantation experienced improvements in sound localization, speech understanding, tinnitus severity, and quality of life with device use. There was a trend for better performance over the postoperative intervals.<sup>2</sup> In a study by Fife et al, patients who undergo CI for severe to profound SNHL attributed to MD were reported to be capable of achieving significant improvement in their hearing performance, comparable to the gains experienced by patients without MD who undergo CI.<sup>1</sup> No deterioration of otolithic or semicircular function has been observed before or after CI.<sup>14</sup> Proper selection criteria for CI and good pre-operative counselling are important for good outcomes.

## CONCLUSION

Cochlear implantation has proved to be effective in hearing restoration, control of vertigo and reduction of tinnitus in patients with Meniere's disease. A small proportion of patients with intractable vertigo may require labyrinthectomy. Labyrinthectomy may lead to soft tissue scarring or even ossification in the cochlea which could prohibit later implantation. This problem can be avoided by simultaneous labyrinthectomy and CI in patients with disabling vertigo. As Meniere's disease has emerged as an indication for CI, awareness of the issues relating to cochlear implantation in Meniere's disease is vital. In our experience, cochlear implants have proved to be useful management option to restore hearing in Meniere's disease.

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