



## Bilateral Meniere's disease in the young, Dilemma's in medical management: A critical review of literature

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### Abstract

It is not very unusual for the ENT personnel to encounter cases of Meniere's disease with bilateral ear involvement. As compared to unilateral cases the patient with bilateral involvement has more significant morbidity and emotional disturbance, particularly in young individuals. The present ambiguity in aetiology of the disease process has leaked to the management too and no definitive treatment options are described till date. The medical management at present is successful in controlling vertigo in the majority of the patients. But the symptoms of hearing loss and tinnitus are relatively less rehabilitated. The options of management like Aminoglycoside infusion and surgery have a limited role in cases with bilateral involvement and this drastically cripples the management protocol. The present review aims to evaluate the efficacy of individual agents and suggest a beneficial protocol based on the current literature.

### Introduction

Meniere's disease or idiopathic endolymphatic hydrops is a syndrome involving the peripheral cochlea-vestibular system.

Patients in addition to the major symptoms like vertigo, fluctuating hearing loss and tinnitus, the patient also has to bear with numerous other minor symptoms causing a lot of emotional disturbance. Most of the patients have a unilateral disease which affects usually in the 4th and 5th decade. The incidence of bilateral cases is around 10-30%, with some literature quoting rates of up to 25-40%. The risk of developing a bilateral disease in a unilateral case is around 14%.

The bilateral involvement is associated with a significant impact on the quality of life of the patient. The lack of specific treatment schedules has further compounded the problem, particularly in young patients with bilateral disease.

Our experience with the case mentioned below has revoked the dilemmas faced in medical management of such patients. This article is our effort to understand and propose a possible treatment plan based on the available literature.

#### Our experience

A 24 year old young male presented to our hospital with history of recurrent episodes of vertigo lasting for 2 to 3 hours associated with nausea and vomiting, fluctuant hearing loss and tinnitus in both ear since last 4 years .The patient had a typical history of ringing sensation in the ear, decrease in hearing followed by severe vertigo that occurs 2 to 3 times a month. The patient was evaluated with a detailed history, examination and investigations were performed. Pure tone audiometry revealed a bilateral severe hearing loss and Glycerol dehydration test was positive. Routine blood investigation and immunological markers were done to check for any immunological disorder.

A MRI scan was done to rule out an intracranial pathology. Based on history and investigations a diagnosis of bilateral Meniere's disease with sensorineural hearing loss was established.

The patient was explained about the nature of the disease and asked to follow a salt restricted diet, with lifestyle modification. Relaxation exercises were added to reinforce life style modification. He was further prescribed tablet Betahistine and diuretic triamterene-hydrochlorothiazide to control recurrent attacks.

Steroids were initiated to prevent further deterioration of hearing. The patient had significant benefit with regards to control of vertigo. But only a moderate degree of improvement was noticed in hearing status.

ent further deterioration of hearing. The patient had significant benefit with regards to control of vertigo. But only a moderate degree of improvement was noticed in hearing status. (Figure 1) The concerns in the patient at present are persisting tinnitus, impairment of hearing and very occasional episodes of vertigo. The young age of the patient with involvement of bilateral labyrinth limits the use of chemical labyrinthectomy and the inherent risk of complications associated with surgery in young patients outweigh the expected improvement in our specific scenario.

#### Discussion

Of the major symptoms the most common symptom is vertigo, followed by tinnitus and hearing loss. Majority of patients present with both vertigo and hearing loss.

Though unilateral to begin with about 72% of the patients develop bilateral involvement over a period of 5 years . Surgical treatment for intractable disease is limited by the fact that in bilateral cases the opposite labyrinth is also hypo functional. This does not solve the issue as the principle is to create a stable hypo functional labyrinth on one side, so that the other side can compensate completely.

The associated risk of hearing loss is a limiting factor in young individuals, for whom the risk might outweigh the gain obtained. So procedures like vestibular nerve section, endolymphatic sac decompression and labyrinthectomy have a guarded use in case scenarios like ours.

The ambiguous nature of the etiology in itself explains the cause for such varied agents being used for treatment of the disease.

The individual medical treatments available and their potency to control all the major symptoms are analyzed.

#### Lifestyle modification:

Life style modification in the form of salt restricted diet (1-2gm/day), avoidance of alcohol, nicotine, stress, allergy and relaxing classes along with psychotherapy always form the 1st line of management. The possible hypothesis for its mechanism of action is by decreasing the endolymph production or decreasing its volume. The other proposed mechanism is activation of renin angiotensin axis with increased production of aldosterone, which in turn improves the stria vascularis ion transport system . There is definitive improvement in vertigo in salt sensitive patients and the natural progression of sensorineural hearing loss is halted . No significant data is available to prove efficacy of solitary salt restriction therapy in the disease process. It is usually prescribed along with diuretics. The literature quotes a vertigo control rate of 79% and hearing loss improvement in about 35% of cases .

#### Diuretics

Diuretics act by decreasing the extracellular fluid volume and altering the salt distribution, thus improving endolymphatic hydrops. They are as mentioned above prescribed along with life style modifications and solitary treatment is a rare occurrence. The commonly used diuretic is hydrochlorothiazide and triamterene, though many classes have been used including carbonic anhydrase inhibitors. The literature quotes an improvement of about 80% in vertigo symptom, but no significant benefit is noted in hearing status or tinnitus.

#### Betahistine

The possible etiology of ischemia of the sac as a cause of hydrops has motivated the usage of bethistine due to its vasodilatory property. It is a H1 receptor agonist resulting in dilatation of blood vessel and H3 receptor antagonist inhibiting the vestibular nuclei. It is generally used in acute episodes, for symptomatic management. The recent MRI based analysis suggests no role in altering the disease process. The literature shows evidence of its utility in controlling vertigo episodes and to some extent tinnitus , . Other vasodilators tried to improve microcirculation are Niacin, Papaverine, Nylidrin, Isosorbide dinitrate and Histamine.

#### Anticholinergics

Anticholinergic medication works by decreasing the action of acetylcholine in the synapses of the neural cells in vestibular nuclei. This inhibits the response of the vestibular system. At a dosage of 2mg of Glycopyrolate it acts as a good vestibular sedative and has a definitive role in acute management of the disease. But the literature does not establish its role for tinnitus or hearing loss.

#### Isosorbide

A hyperosmotic diuretic, it acts in the same principle as does diuretics, urea and glycerol. The sparse literature available suggests a 75% control of vertigo symptoms and 50% of the patients had an improvement in tinnitus. The hearing improvement was only 20%. The continuous oral administration had a better response as opposed to intermittent administration .

## Corticosteroids

An autoimmune cause is proposed as an etiological factor particularly in bilateral cases. The presence of a specific type of HLA in the patients and association of allergy further strengthens this claim. Steroids probably act by its anti inflammatory and immunomodulating property. The steroids also have role on the metabolism of the body and a possible direct action over the sac to regulate the fluid haemostasis. The literature states an insignificant role of intra-tympanic steroids in comparison to placebo in control of symptoms . But few studies have shown a good vertigo control rate of steroids and some propose it to be the only drug available to rehabilitate hearing impairment (30-60% improvement in speech discrimination) . As per the experience of using steroids in pathologies like sudden SNHL, steroids have been considered to have a definite role in bilateral meniere's disease patients for restoration of hearing to some extent. Other agents like, low dose methotrexate, Gamma globulin and Recombinant tumour necrosis factor alpha by the virtue of their immune modulating action are useful in select cases but, further studies are required to prove their significance.

## Aminoglycoside

Chemical labyrinthectomy is indicated in patients who have failed other conservative measures. It aids in the control of vertigo and tinnitus by destroying the dark cells in the inner ear that produce endolymph and impairment of transmission of vestibular response to brain. The systemic administration was associated with a high rate of hearing loss and thus now intra-tympanic titrated drug perfusion is preferred. Gentamicin due to its specific vestibulotoxic action is now used most commonly. The literature quotes a vertigo control rate of up to 91% with a profound SNHL being noted in 3% of patients . But the prerequisite for an uninvolved opposite sided vestibular system makes its usage in bilateral cases as ours a very highly debatable option.

## Antiviral

Some authors postulated that Meniere's disease is a part of polyganglionitis where symptoms appear because of reactivation of virus in the internal auditory canal. Antivirals like gancyclovir were instilled through microwick trans-tympanically and initial control rates were high (80%), but later RCT did not establish a significant role in comparison to placebo .

## Lithium

Lithium is postulated to be helpful by helping in ion transport across the membrane, thus helping in resolution of hydrops. Thomsen J et al in their double blind study of 21 patients didn't establish any significant effect of lithium over placebo . The present literature does not suggest a significant role for the drug in the current scheme of things.

## Picrotoxin

Picrotoxin is a noncompetitive antagonist at GABA-A receptors and the exact mechanism of action in meniere's disease is poorly understood. The literature suggests a role of picrotoxin in medical prophylaxis as it prevents vertigo and also has very minimal side effects.

## Adenosine triphosphate

The opinion that free radicals induce injury and cause hydrops has led to evolution of adenosine triphosphate as a treatment modality. Adenosine triphosphate , acts by decreasing the oxidative stress of the tissue. As with other drugs like picrotoxin there is lack of clear evidence for their regular use in treatment of patients.

## Positive Pressure pulse devices

The menniet's device is FDA approved for positive pressure therapy in patients of meniere's disease. The exact mechanism is not clear, but the positive pressure delivered after placement of a tympanostomy tube helps in better absorption of endolymph and its redistribution. Success rate of around 60 to 70% has been quoted in the literature, particularly in the initial 3 months. Long term results though are not very encouraging when compared to simple tympanostomy tube placement.

The surgical management warrants a small mention. The hearing destructive labyrinthectomy has virtually no role in a bilateral disease, hearing preserving endolymphatic sac decompression and nerve section have a role as the last resort. Particularly nerve section with a low risk of hearing loss is a feasible option.

Medical management	Vertigo	Hearing impairment	Tinnitus
Lifestyle modification	√√	√	√
Diuretics	√√	×	×
Betahistine	√	×	√
Anticholinergics	√√	×	×
Isosorbide	√	×	√
Corticosteroids	√	√√	√
Aminoglycoside	√√	×	√
Antiviral	×	×	×
Lithium	×	×	×
Picrotoxin	√	×	×
Adenosine triphosphate	×	×	×
Positive Pressure pulse devices	√	×	×

## Conclusion

A bilateral involvement in Meiniere's disease presents an additional challenge to the treating physician as the literature does not provide any conclusive answers. The present review suggests a role of Betahistine, vestibular sedatives and Anti-cholinergics in acute episodes with good control of vertigo. In long term the major symptom of vertigo is controlled well by Lifestyle modification, Diuretics and Anti-cholinergic treatment. But with regards to hearing loss and tinnitus, they have a minimal to no role. In patients not responding to regular management Aminoglycoside infusion has delivered good response, but in cases with bilateral involvement its role is very debatable. The relatively less significant symptoms of tinnitus and hearing loss gain tremendous importance with bilateral involvement and their rehabilitation is a major concern. Intra tympanic steroids at present have a role in hearing rehabilitation and decreasing tinnitus. Betahistine is also considered to induce improvement in tinnitus on long run. Surgical treatment is to be selected as per the severity status of the patient. Vestibular nerve section with hearing preservation is a good option to fall back on.

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