

Meniere's Disease

A disorder affecting inner ear homeostasis, classically manifested by episodic vertigo lasting hours, fluctuating hearing loss, tinnitus and aural fullness. Rarely sudden drop attacks can develop, but when present are debilitating. It is however heterogenous in both presentation and cause, and thus a single treatment will not be effective for everyone.¹ The reported prevalence varies widely but probably sits around 190 per 100,000. It is almost twice as common in women than men, and most commonly presents in the age range from 20-60 years. Whilst usually affecting only one ear, approximately 15% of cases will involve both ears.² It usually arises de-novo, but can occasionally develop following severe infections or trauma.

Diagnosis however is often not easy, as the classic symptoms are not always present and other conditions can manifest in similar ways. Episodes of vertigo can be triggered in many ways, most commonly stress (physical or emotional) and salt in the diet. Less commonly allergies, food triggers, menstrual fluctuations amongst others can play a role.

In addition to a careful history, documentation of sensorineural (nerve) hearing loss is essential to help confirm the presence of Meniere's Disease. The commonest cause of episodic vertigo is BPPV (benign positional paroxysmal vertigo), but this is characterised by brief head positional vertigo. The major differential diagnosis is vestibular migraine (or MAV, migraine associated vertigo) which essentially cannot directly cause hearing loss. It is not uncommon, however to have coexisting Meniere's Disease and migrainous vertigo.³

There is no definitive test to confirm the diagnosis. An electrophysiologic test called the EChOG has been used in this regard, and if abnormal, can be suggestive but in general terms is not usually helpful. According to the AAO-HNS guidelines a diagnosis of definite Meniere's Disease is only made at autopsy. Audiology and selective balance function tests can assist in assessing severity of disease and ability to compensate to the symptoms.

Initially one of the first steps to be taken is to rule out the presence of intracranial pathology such as an acoustic neuroma (vestibular schwannoma), which requires a MRI scan of the brain.

Traditionally different phases of the disease have been described, but rarely does the disease follow a predictable pattern. In general terms though over time many people suffer from a progressive hearing loss, and eventually recurrent vertiginous attacks fade. Like the permanent nerve damage to the hearing nerve, permanent damage to the vestibular system also occurs. This can lead to permanent imbalance, dizziness, and what is often described as a 'cotton wool' feeling in the head. These symptoms are more pronounced if other components of the balance system are impaired. These include eyesight, joint function (including the neck), and cardiovascular health.

The overwhelming goal of long term management is to minimise the number of vertigo attacks and maintain hearing and balance function, and in the acute phase to truncate the severity of an attack. The recent International Consensus on the management of Meniere's Disease has reinforced the evidence based approach to management outlined below.⁴

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Management of acute vertigo

Usually this involves the use of vestibular suppressants. Commonly used are Stemetil, Phenergen, Lorazepam. Usually ondansetron is used sublingually 10 minutes prior to the oral vestibular suppressant to ensure absorption. An injection or suppository is occasionally required if the ondansetron cannot control severe vomiting. Sometimes patients get a premonitory sensation and can abort an attack with the use of one of the vestibular suppressants, a diuretic or anti-inflammatory steroids.

During a prolonged attack or a series of attacks, these can medications can be taken regularly or the use of intra-tympanic steroids can be used. (This involves an injection through the ear drum under a local anaesthetic in the clinic. ^{5,6}

Prophylactic Management

As the pattern and severity of the disease varies greatly from patient to patient so does their management. Approximately 75-80% of patients can achieve good control with non-interventional treatment. ³ The first step is to control the two major triggers of an attack, salt and stress. ⁷ A low salt diet from a Meniere's perspective requires a significant change in lifestyle, even in those who generally live a healthy life and do not add salt to their food, or eat much in the way of processed foods. There is significant hidden salt in many foods, especially salt and breakfast cereals. Obtaining salt contents in all foods is essential (which we can supply), and the book 'Salt matters' is an excellent resource. The only way to monitor salt intake is to take a 24 hour urine collection. In general terms the first goal is to achieve less than 1200mg/day. If vertigo continues then further tightening to less than 600mg/day is targeted.

Other considerations such as withdrawing caffeine, chocolate, smoking and alcohol are important for some patients.

Management of stress and ensuring adequate sleep is also essential.⁸ Alternative treatments such as massage therapy and acupuncture can be helpful, as can formal psychological treatment in the form of cognitive behavioral therapy, especially in severe cases where consideration of surgical intervention is being made. ⁹

Management of inhalant and food allergies is important in a subset of patients. Allergies are up to three times as prevalent in Meniere's Disease. Interestingly this is especially the case if also suffering from Migraine. The symptoms of MD are generally better controlled, with fewer vertigo attacks and more stable hearing, in those patients with allergy and MD whose underlying allergic disorder is down-regulated with immunotherapy and/or dietary avoidance of reactive food allergens. ¹⁰

When there is inadequate control medications and surgical interventions must then be considered.

Drug Therapy

The commonest drug used is Betahistine (Serc). It is thought to act by increasing the blood supply to the inner ear, acting as a H3-antagonist. Anecdotally betahistine is helpful in some patients, but again there is no overwhelming evidence to support its use. ¹¹ A recent paper has suggested that

it acts on the endolymphatic sac.¹² It has a very low side effect profile, so is worth trying prior to interventional management. The dose can be elevated to 32mg three times a day.

Diuretic therapy, (fluid tablets) have been used for many years in Meniere's. In theory they may alter in the electrolyte balance within the inner ear. There has been controversy over their use, with no evidence to support their use.¹³ Side effects at the low doses used for Meniere's Disease however are minimal, so can their use can be considered. A typical drug is Moduretic: Amiloride HCl 5 mg, hydrochlorothiazide 50 mg; 1-2 tablets per day.

Migraine Prophylaxis: As mentioned it is not uncommon to have co-existing migrainous vertigo, or it is difficult to differentiate. Many of the same triggers exist such as any physiologic or emotional stress and certain foods. There is a low threshold to use migraine preventors, initially non-pharmacologic supplements containing magnesium, riboflavin and feverfew. Pharmacologic options include amitriptyline, dothep, pizotifen, propranolol, topomax, cinnarizine and flunarizine.

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Interventional management (Surgery)

Intervention in Meniere's disease is divided into non-destructive and destructive categories. Again the principal of any intervention is to stop vertigo, prevent ongoing hearing loss and minimize tinnitus and fullness, while minimizing damage to the vestibular system. Unfortunately the procedures with the highest rates of success carry the highest rates of permanent disequilibrium and often place the hearing at risk. Therefore minimal impact interventions are often tried first, even if there is no guarantee of success. The caveat here is in those situations where definite control of vertigo is needed, such as in those patients carrying a heavy vehicle license where their livelihood depends on cure from vertigo. The development of drop attacks is another circumstance where a more aggressive approach is needed.

Another important factor in avoiding destructive procedures if possible, is in the presence of or chance of developing bilateral disease. Bilateral severe vestibular loss is an extremely debilitating condition.

Tympanostomy tube insertion (grommets) are placed and are thought to influence inner ear pressure by stabilizing middle ear pressure. They are predominantly helpful in reducing the sensation of fullness, but can also reduce vertigo. The most important fact is there is no risk to hearing nor of worsening overall balance function. The only precaution is that the ear needs to be kept dry. The placement of a tube also allows the use of the Meniett device. This device emits repeated pressure pulses with amplitude of 12 cm of water and consist of a complex pressure wave composed of static pressure and a 6-Hz sinus modulation. It is an essentially minimally invasive device which in some studies has reasonable rates of control.¹⁷⁻¹⁹ In Australia this is currently not funded by insurance companies.²⁰

Intratympanic steroid injections, (usually dexamethasone, methylpredisone or triamcinolone) have been shown to have good levels of success in reducing vertigo but with with no risk to hearing, as apposed to gentamicin.^{21 22,23} Therefore it is almost always used before any potentially destructive procedure is considered. There is no defined regime at this point, but one protocol is to repeat an injection each week or fortnight for 4 injections. A ventilation tube at the beginning

of the course to make sequential injections/infusions easier, and can also be continued at home, with an appropriate concentration of steroid.

Endolymphatic sac surgery. This involves a mastoidectomy and opening of the endolymphatic sac which in theory stabilizes inner ear homeostasis. There is varying evidence to support its use, although in some hands is reported to have good control rates with acceptable rates of hearing loss.²⁴⁻²⁸

Intratympanic gentamicin²⁹⁻³³ has significantly changed interventional management in Meniere's disease over the last 20 years. Most studies show around 80% good control of vertigo, but with a 10-20% chance of worsening hearing and about 2% chance of complete hearing loss.³⁴ Essentially this is a partial chemical labyrinthectomy, and can make overall balance worse. Balance training in the form of vestibular rehabilitation after this intervention is essential. If the initial treatment fails, normally a repeat injection is performed prior to moving on to more significant interventions. The procedure itself can be either performed under local anaesthetic in the clinic or in the operating theatre. The latter allows visualization of the middle ear to ensure that there is not a thin membrane covering the round window which can impair absorption of the drug.

Vestibular nerve section is a highly effective procedure to control vertigo, with only a 2% chance of hearing loss. It does however involve an intracranial procedure, albeit with a low rate of complications. Essentially this is considered in young patients with good residual hearing, especially if definitive control of vertigo is needed.³⁵

Surgical Labyrinthectomy is a highly effective procedure to control of vertigo, but results in sacrifice of any residual hearing and vestibular function on that side.³⁵ In most cases now a concurrent cochlear implant is placed.³⁶

Hearing and Vestibular Rehabilitation

From a hearing rehabilitation and often control of tinnitus, a hearing aid is recommended. When an ear has no remaining functional hearing, a cross over hearing aid is an option. This can either utilise Bluetooth or bone conducting technology, ie a bone anchored hearing aid [BAHA, Bonebridge] The other option is cochlear implantation, both in unilateral hearing loss, but even more importantly in bilateral disease. In select cases concomitant surgical labyrinthectomy and cochlear implantation can be considered.³⁶

There is some evidence that boosting the hearing can also help to some degree with balance. Gentle to moderate exercise is very important physically and psychologically. Formal vestibular rehabilitation is essential when the disease has caused permanent vestibular impairment and or an ablative procedure is required. It is often much more difficult to compensate if the level of vestibular function is fluctuating.

Psychological Support

This is a very important and much underestimated aspect in the management of Menieres disease. This can help minimize stressors which act as a trigger to acute attacks, and can also help in the management of underlying tinnitus, dizziness and imbalance. A syndrome labeled psychophysiological dizziness plays a large role in many patients with Meniere's Disease. This essentially where an insult to the vestibular system leaves a degree of nerve damage. The brain

needs to compensate for this loss and anxiety, especially anxiety centred on the fear of further attacks or dizziness can further amplify the symptoms of instability.

Recommended Treatment Regime

Step 1

Confirm probable diagnosis, and exclude central pathology
Institute low salt diet, and monitor levels using 24 hour urine collections
Lifestyle modifications, stress reduction with consideration of psychologic therapy if required.
Consider trial of betahistine (Serc), up to a dose of 16mg three times a day
Consider dietary supplements
Hearing and vestibular rehabilitation

Step 2

Consider trial of diuretic, especially if an appropriately low salt level is not reached.
Insertion of ventilation tube, with dexamethasone (steroid infusion) into middle ear, or transtympanic dexamethasone injection. These treatments can be repeated indefinitely if controlling symptoms.

Step 3

Consider trial of Meniett Device
Consider Gentamicin Infusion, Endolymphatic sac surgery or Vestibular Nerve Section in young patients with good residual hearing.

Step 4

Consider repeat Gentamicin Infusion
Consider surgical labyrinthectomy +/- cochlear implantation

Important Contacts and Links

Psychological Management
Cognitive behavioral therapy and Tinnitus Retraining Therapy
Vestibular Rehabilitation
Audiology

Helpful internet resources

www.menieres.org.au

www.saltmatters.org

www.menieresnsw.org.au

www.tinnitus.asn.au

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