Audiological management of patients with Meniere's disease

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Meniere's is not a single entity but several different diseases that may cause the combination of symptoms known as Meniere's disease (MD). Prosper Meniere was a French doctor who first associated the symptoms of fluctuating hearing loss, tinnitus, ear fullness and vertigo with a disorder of the inner ear in 1861. It is now understood that these symptoms are associated with excess endolymph (endolymphatic hydrops) in the inner ear. Research points to several possible causes such as a viral or bacterial infection causing inflammation in the inner ear, allergies affecting the upper respiratory tract, hormonal disorders, genetics, trauma and injuries to the ear as well as auto immune diseases.

The classic presentation of MD is a sensation of ear fullness, tinnitus and a sensorineural hearing loss affecting the lower frequencies that precedes a severe vertigo attack associated with nausea, vomiting and even diarrhoea lasting from 20 minutes to 6 hours. The vertigo attacks occur in clusters followed by periods of remission. The symptoms settle down spontaneously only to return a few days, weeks or months later. The order of appearance of the symptoms can vary amongst patients although it is possible that the initial cochlear symptoms go unnoticed until after the first vertigo episode.

The frequency and severity of these symptoms varies. For the majority of patients the disorder progressively destroys the inner ear's delicate structures causing permanent loss of hearing and loss of vestibular function.

Vertigo is the most distressing of the symptoms leading to anxiety and significant life disruption. Hearing loss is usually the last symptom to be acknowledged and addressed by patients and doctors alike. Tinnitus is more likely to bring a patient to the audiology clinic rather than the hearing loss itself.

Misdiagnosis is very common when the symptoms are assessed in isolation. Vertigo and vomiting may be misdiagnosed as "food poisoning", labyrinthitis or vestibular neuritis while ear fullness may be attributed to a middle ear or Eustachian tube dysfunction.

Diagnosis is based on the clinical history after exclusion of other conditions using tests such as CT and MRI scans. Audiometric documentation of a fluctuating hearing loss and electrocochleography showing enlarged summating potentials helps to confirm the diagnosis of Meniere's disease. (Gibson, 2019).

There is no known cure for MD and treatment aims to manage individual symptoms to improve quality of life. Relief from the attacks of vertigo is the most sought after treatment but unfortunately, there is no effective method to completely resolve the vertigo without destroying the residual vestibular function.

Low salt diet is a very popular treatment option with only anecdotal evidence to support it. Diuretics are also widely prescribed together with medication to reduce vomit as well as pharmacological to improve blood circulation in the inner ear. The evidence to support these treatments is still weak. Changes to a healthier lifestyle seem to provide good results for many patients.

Trans-tympanic injections of steroid to reduce inflammation may halt a cluster of attacks. Gentamicin intratympanic injections may be used to partially or completely destroy vestibular function but may also damage cochlear hair cells. Different surgical techniques, such as insertion of grommets and endolymphatic sac surgery are utilised but their effectiveness is controversial. The most radical interventions are labyrinthectomy and vestibular nerve section. It has recently become common practice to perform both labyrinthectomy and cochlear implantation simultaneously to address both vertigo and hearing loss in patients with intractable vertigo and severe hearing loss due to MD (Mukherjee et al. 2016).

MD is usually unilateral but it has been estimated that up to 40% of patients can develop the condition in the contra-lateral ear over time (Stahle et al, 1991). Hearing difficulties are more obvious for those with bilateral loss but the challenges of unilateral loss are well known amongst audiologists. Hearing loss in one ear is likely to affect the ability to lateralize and localize sounds, interferes with speech understanding in the presence of background noise and reduces tolerance to loud sounds.

Even a mild unilateral hearing loss may have adverse consequences and negatively impact on stress levels. Stress in known to aggravate the symptoms of MD, hence treating the hearing loss may help patients to feel more relaxed, minimizing the severity of symptoms and improving their quality of life.

The hearing loss in MD has distinct characteristics. It starts as a mild sensorineural hearing loss in the low frequencies which returns back to normal levels intermittently for a period of time. Vertigo attacks tend to occur when low frequency hearing is at its lowest point. Fluctuating low frequency sensorineural hearing loss with recovery to normal audiometric levels is the characteristic of the first stage of MD (fig 1).

At the second stage of MD, the low frequency hearing no longer recovers back to normal and the mid and high frequencies become affected. Hearing fluctuation continues to occur but in the mid frequencies only (fig 2). Vertigo attacks are also intermittent during this stage but less severe.

The third stage, also known as “burnt-out MD”, is when vertigo attacks subside due to a significant loss of vestibular function. At this stage, sudden brief attacks of vertigo may cause drop attacks. The hearing loss becomes severe and flatter affecting all frequencies. Our studies showed that even at this stage hearing continues to fluctuate (McNeill et al 2009) but at a lesser degree than previously, and the audiogram maintains a flatter configuration (fig 3).

Some patient still progress to a fourth stage when the hearing loss is so severe and quality so poor that hearing aids are no longer helpful. These patients are good candidates for cochlear implantation and usually obtain very good hearing results.

For a long time hearing aids were not considered a viable option for patients with MD. The reasons for contra-indication were mostly based on old fashion criteria such as poor speech recognition (discrimination) scores as measured during audiological assessment, unilateral hearing loss not considered as disruptive enough to warrant amplification, recruitment leading to poor tolerance of amplified sounds and hearing fluctuation (Koefoed-Nielsen and Courtois 1995; Valent et al 2006).

Our experience shows that hearing fluctuation is the only real challenge when dealing with this population. In fact, research conducted by the author showed that poor speech recognition scores should not be a contra-indicator to amplification as it has been demonstrated that acclimatisation

with properly fitted hearing aids improve speech recognition overtime providing the residual hearing levels are within an aidable range (McNeill et al, 2002). We speculate that poor speech recognition scores found in audiology tests of patients with MD may be caused by inappropriate loudness of presented speech test materials which trigger recruitment, hence reducing tolerance and creating distortion. Our experience shows that an increase of 2dB instead of traditional 5dB steps in the presentation level of speech test via the audiometer resulted in significant change in recognition scores of patients with severe recruitment. We also found that the levels of sound tolerance of these patients tend to improve overtime as one of the reasons for intolerance is caused by auditory deprivation. Gradual acclimatisation with amplification improves speech recognition and decreases sound intolerance.

Patients with hearing loss due to MD benefit from hearing rehabilitation but audiological intervention and hearing aid selection will vary according to the stage of MD.

The greatest complicating factor for successful hearing rehabilitation is the presence of hearing fluctuations that occur in the first and second stages of the disease. The problem is aggravated further because the fluctuations are unpredictable in respect to time of occurrence. The audiogram configurations are shown in fig 2.

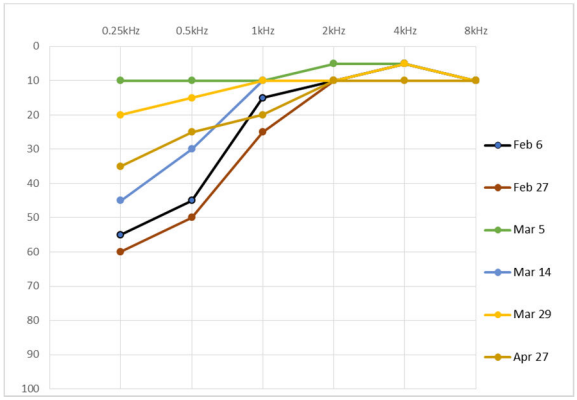

Fig 1: Hearing fluctuation over a period of 12 weeks (stage 1)

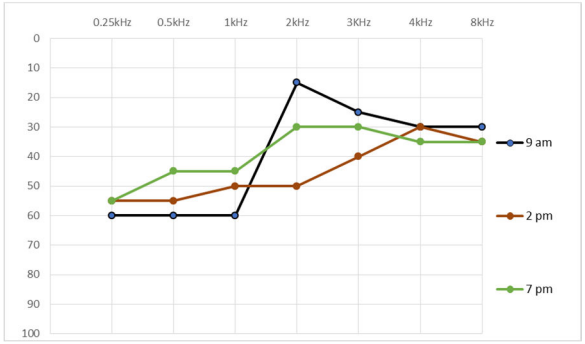

Fig 2: Hearing fluctuation over a period of 10 hours in one single day (MD stage 2)

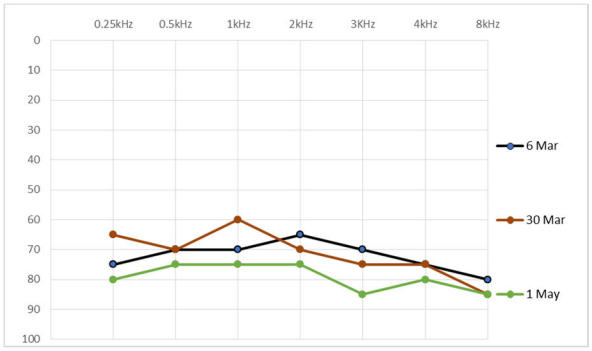

Fig. 3: Hearing Fluctuation over 8 weeks (MD stage 3)

It is challenging to ascertain the stage of MD and the extent of hearing fluctuations based on clinical tests. An audiogram is only a snap shot of the hearing levels at one point in time. Successive audiograms performed several times a day are needed to be able to demonstrate hearing fluctuations. Self-hearing tests using a mobile phone app is an effective method of assessing hearing fluctuation as the patients are able to test their own hearing any time whenever changes are noticed.

At my clinic it is protocol to instruct MD patients to test their own hearing up to 3 times a day for a period of at least 4 weeks during the active stages of the condition to verify occurrences of hearing fluctuation. Hearing rehabilitation is recommended once the stage has been determined according to the following criteria:

Stage 1: Hearing aids are not recommended unless hearing loss is bilateral. In cases of unilateral fluctuating hearing loss in the low frequencies, as in stage 1, hearing tactics may be sufficient and the patients are also advised to monitor their hearing changes as it may be a warning signal that a vertigo attack is on its way. Such approach also helps patients to better understand their condition giving them a greater sense of control.

Stage 2: As the disorder progresses to the second stage the hearing loss becomes more evident and disruptive. Hearing aids with programming software that have in-situ audiometry features are recommended as the best option to address unpredictable hearing fluctuation. We provide the patients with the hardware and software to program their own hearing aids when a hearing fluctuation occurs. The patients are instructed how to test their own hearing levels via the software that automatically adjusts the hearing aid frequency response when changes occur.

Multi-memory hearing aids have been attempted by the author with no success and this has been attributed to the lack of pattern and unpredictability of hearing fluctuation. Our novel protocol has proved more successful than multi-memory hearing aids and avoids dissatisfied patients constantly returning to the clinic for hearing aid adjustments (McNeill 2005, McNeill et al 2008).

Stage 3: Also known as the burnt-out stage is when patients are more likely to seek audiological intervention because the tinnitus becomes more intrusive and the hearing loss more disruptive. At this stage hearing fluctuation becomes more linear and less dramatic hence more predictable, so that a hearing aid with a volume control is sufficient to address hearing changes. Unfortunately, even in this day and age, many patients on stage 3 are denied hearing aids by medical professionals under the false belief that hearing aids will not help them. Our experience has proved them wrong. Patients on stage 3 are usually very satisfied hearing aid users as amplification not only improve their hearing ability but also reduces tinnitus perception (McNeill & Taylor, 2010).

Stage 4: As the cochlear membranes become more and more distorted due to hydrops, residual hearing becomes more distorted with a much reduced dynamic range, limiting the positive effects of amplification. Cochlear implant is a viable option which has proved beneficial to many patients with MD as it by-passes the damaged cochlea stimulating the auditory nerve directly. Nevertheless, recent evidence shows that the electrical hearing levels may still continue to fluctuate. It is likely that endolymphatic hydrops varies affecting the cochlear implant electrode impedances (McNeill & Eykamp 2016). Such changes mean that the cochlear implant of a patient with MD may need to be re-mapped more frequently than those with hearing losses of other aetiologies.

Our clinical experience with this population shows that early audiological intervention is beneficial to patients with MD as it contributes to better understanding of their symptoms giving them a greater sense of control. Hearing rehabilitation by means of hearing aids and cochlear implants not only improves overall hearing ability but also reduces tinnitus perception in this population.

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