THE MENIERE’S TREATMENT LADDER (part three)

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In Part 1 Professor Gibson, introduced the Meniere’s treatment ladder with an explanation of the most benign therapies covering reassurance, diet and the use of diuretics/urea and betahistine. In Part 2 he continued with an explanation on oral steroids, grommets, overpressure and Intratympanic therapies. In this edition he provides an explanation on the final four steps in the Meniere’s Treatment ladder, surgical options.

**Surgical treatment**

Desperation grows when the attacks of vertigo continue to cause distress and ruin family and business life. The sufferer may seek a surgical solution. Nevertheless it should be remembered that the attacks of vertigo will eventually lessen with 30-50% of the balance function retained. This residual balance function can be valuable, especially if the other ear gets involved, or if the person is elderly and needs to retain some balance to prevent disequilibrium and possible falls. The adage is “The treatment should not have an end result which is worse than that of the natural history of the disease”.

CI - cochlear implant
VI – Vestibular implant
STEP TEN - Endolymphatic Sac Surgery

In 1928, George Portmann\(^1\) was the first to recommend endolymphatic sac surgery. The concept at that time was that the endolymphatic sac (ELS) continually drained endolymph from the inner ear and when it became blocked endolymph accumulated in the inner ear (endolymphatic hydrops). Eventually the excess fluid caused ruptures of the inner ear membrane (Reissner’s membrane) mixing the inner ear fluids together and causing an attack of vertigo until the electrolyte balance of the fluids was restored and the membrane rupture was repaired\(^2\). Opening up the ELS was believed to promote drainage and alleviate Meniere’s disease (MD).

This concept has now been shown to be hopelessly wrong.

Research has shown that the endolymphatic sac (ELS) does not continuously drain endolymph but only actively attracts endolymph into its lumen when an emergency situation develops\(^3\). The ELS is the only part of the membranous inner ear which is immunologically competent and can remove viruses and other debris. It consists of a network of tubules which can secrete glycoproteins attracting endolymph. It is also speculated that the ELS can secrete a substance which increases the volume of endolymph to activate the drainage to its lumen and flush debris from the inner ear: it is possible that this action is the cause of the attacks of vertigo.

It is now suspected that ELS surgery causes damage to the delicate mechanism preventing sudden changes in endolymph volume and this stops the attacks of vertigo unless the ELS recovers. The author investigated removing the ELS\(^4\) and showed a similar outcome regarding alleviation of vertigo with less tendency for a recurrence. Recently further studies seem to confirm this research\(^5\).

ELS surgery remains a controversial treatment but it has the advantage that it does not destroy all the remaining balance function in the ear. Most surgeons report a 70% success in preventing vertigo attacks but the hearing results are variable. After ELS removal or blocking the duct to the sac, the hearing usually deteriorates to a 70dBHL loss but recurrences are fewer than drainage procedures. ELS surgery can be recommended for older MD sufferers and when there is a risk the opposite ear will become involved.

STEP ELEVEN - Labyrinthectomy

Surgical labyrinthectomy was first described in 1904\(^6\) and aims to destroy the balance portion of the inner ear, but this has not been possible without also destroying the hearing portion of the inner ear. The usual method is to drill through the mastoid bone behind the pinna and then open each of the semicircular canals to remove the membranous semicircular canals and utricle. After the surgery, the patient is very dizzy and needs to quickly learn to rebalance using the unaffected ear. Physiotherapy is very helpful. Once the balance has been adequately regained, there are no further attacks of vertigo,
There are three major drawbacks: all the hearing in the operated ear is lost; if tinnitus is a problem, it is not possible to mask the tinnitus; if the other ear becomes involved, it is not possible to perform another labyrinthectomy without causing serious balance problems and complete deafness.

Although surgical labyrinthectomy was a popular choice in the past, the drawbacks have limited its indication only to MD sufferers under the age of 70 years, with very poor hearing in the affected ear, and who have absolutely no evidence of MD in the opposite ear. If the MD is causing drop attacks which are preventing driving and employment, surgical labyrinthectomy can be considered.

**STEP TWELVE - Vestibular Nerve Section**
Vestibular nerve section involves entering the space between the inner ear and the brain so that the vestibular (balance) nerve can be sectioned leaving the cochlear (hearing) nerve intact. It is major surgery accomplished by opening the skull above the ear (middle fossa) or behind the ear (retrolabyrinthine or suboccipital approach).

The middle fossa approach\(^{7}\) is surgically difficult and there is a risk of opening inadvertently into the superior semicircular canal and causing a total hearing loss and there is a possibility of facial nerve injury. As part of the brain has been retracted, the patient requires medication to prevent any brain disturbance for a few months after the surgery.

The retrolabyrinthine and suboccipital approaches\(^{8}\) are surgically easier and do not require significant brain retraction. The problem is the nerves are fused together and the cochlear nerve and the facial nerve have to be separated from the vestibular nerve. There is a possibility of leaving some of the vestibular nerve intact although surgeons who undertake this surgery regularly are unlikely to make this error.

As all the balance in the affected ear is removed, there is a period of dizziness after the surgery and physiotherapy is helpful. The main hope in performing vestibular nerve section has been the preservation of hearing but this has not often been achieved in the longer term. The reason for the gradual loss of the remaining hearing may be that the MD within the inner ear has not been controlled. The loss of speech recognition could be because the efferent nerves which control tuning of the cochlear hair cells travel in the vestibular nerve and have been sectioned. The only advantage of the middle fossa approach is that these efferent fibres may be spared.

Vestibular nerve section has most of the drawbacks of surgical labyrinthectomy. It is formidable surgery with possible major complications, so the MD sufferer needs confidence in the choice of surgeon. The indications are similar to surgical labyrinthectomy but the hope of preserving some hearing exists.

**STEP THIRTEEN - Surgical Labyrinthectomy and Cochlear Implantation**
Combining surgical labyrinthectomy and a cochlear implant is a new and exciting option for MD\(^{6}\).
The surgery is much easier than vestibular nerve section and there is a likely possibility that the hearing in the ear will be improved and tinnitus can be controlled. The surgery is performed through the mastoid bone behind the ear and there is no need to open through the skull into the brain area. Surgical complications are unlikely but as the balance function in the ear is totally destroyed, there is a period of dizziness after the surgery and vestibular physiotherapy is helpful.

Until recently, using a cochlear implant to restore hearing when the opposite ear has good hearing was not considered. It was thought that the electronic hearing would not be compatible with the natural hearing and the recipient would dislike the sound. Initially a cochlear implant was inserted into deaf ears which were associated with very severe tinnitus, not only did this often control the tinnitus but many recipients appreciated the recovery of hearing\(^\text{10,11}\). Since then placing a cochlear implant in an ear despite good hearing in the other ear has gained popularity and can now be recommended for babies born with single sided deafness.

Cochlear implants have given good results in MD as the hearing nerve is usually well preserved. The outcome after cochlear implantation in a single sided deaf ear has been shown to be better if the ear has experienced hearing in the past and if the length of time of the deafness is short. It is not surprising that cochlear implantation during surgical labyrinthectomy yields excellent hearing recovery.

Surgical labyrinthectomy and cochlear implantation appears the best option for MD when it is only affecting one ear. Obviously the sufferer should be young enough to learn to rebalance using only one ear and the attacks should be sufficiently distressing to merit surgery.

**Final Remarks**

It is hoped that a vestibular implant will soon be available. A surgical labyrinthectomy combined with both a cochlear implant (restoring hearing) and a vestibular implant (restoring balance) would be suitable for older sufferers and when the MD affects both ears.

But a word of caution regarding surgery for MD. If the sufferer can be treated medically and can await the natural ‘burn out’ of the disease, there will be residual balance and a hearing aid can often restore hearing and subdue tinnitus. Furthermore, if the MD begins to affect both ears, the naturally ‘burnt out’ ear can provide still useful balance. None of the surgical options can offer such a good outcome.
Figure showing the Eighth cranial nerve which is composed of the cochlear nerve and the vestibular nerve

Endolymphatic sac surgery does not always succeed and recurrence of significant vertigo attacks may occur in about 30% after surgery. Surgical labyrinthectomy with cochlear implantation is presently the most certain method of stopping the attacks of vertigo with minimal surgical risk and the hope of restoring hearing and controlling tinnitus but there still remains the risk that the other ear may become affected by MD.

Surgical solutions are really only indicated for MD sufferers who are under the age of 70 years who can learn to rebalance with one ear and are suffering significant attacks of vertigo threatening their employment and family life.
References

1. Portmann G The saccus endolymphaticus and an operation for draining the same for relief of vertigo. J Laryngol 1927; 42 : 809-820


