# THE MENIERE’S TREAMENT LADDER

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After a diagnosis of Meniere’s disease has been established, the family doctor or the specialist will consider the most appropriate treatment. The ‘treatment ladder’ shows steps that are usually taken in Australia from the most benign therapies to the most radical surgery. I will be writing brief explanations about each group of steps along the treatment ladder.



*CI - cochlear Implant*

 *VI – Vestibular Implant*

The medical treatments, introduction

After suffering fearsome attacks of vertigo preventing a normal family and working life, the newly diagnosed sufferer may demand surgical treatments but it should be remembered that the final outcome may be better if surgery is avoided. Meniere’s disease sufferers must remember that the condition will eventually ‘burn out’ leaving some hearing and balance in the affected ear. There is always ‘a light at the end of the tunnel’ although the problem is how long is the tunnel.

‘Burn out’ is the most usual end point of the disease. At this stage the balance organ has lost its re power and has become so weak that it cannot cause severe attacks. The hearing no longer fluctuates and there is a severe hearing loss in the affected ear. The tinnitus remains and can bother some people much more than others especially if they are unable to ignore it. Tinnitus is rather like a noisy refrigerator – some people completely forget it is there while others find it intolerable. The feeling of blockage in the ear usually fades away at burn out. Modern hearing aids are usually effective in restoring some useable hearing and in lessening any tinnitus.

Although sufferers welcome ‘burn out’, it is not a cure just the end point of the disease. The problem is that ‘burn out’ may take many years to occur especially if there are prolonged periods of remission.

Medical treatment can be the alleviation of symptoms during an attack or treatments which aim at preventing further attacks occurring. Alleviation during attacks is usually achieved using antiemetics / antinauseants. In Australia, StemetilTM (prochlorperazine) is usually given either by an injection in the hospital, or by using a suppository (25mg), or by using tablets (each is 5mg). To get an effective oral dose of StemetilTM at least 4 tablets are required and these may have to be chewed or they can be vomited out too easily. StemetilTM should be used cautiously as long term therapy because it will cause Parkinsonian-like symptoms.  A better oral medication is ZofranTM (ondansetron) which is effective in stopping the nausea and vomiting but does not stop the attack. It is placed under the tongue to dissolve avoiding any tendency to vomit it away. To stop the nausea and vomiting during a Meniere’s attack, 8mg is the minimum dose.

**The treatment ladder concerns the measures taken to prevent further attacks occurring.**

STEP ONE - Reassurance

When the first attack occurs, the sufferer is usually rushed to hospital fearing they have suffered a cerebral event or heart attack. Reassurance that there is not a terminal illness is needed.

Once the diagnosis has been reached, reassurance can help prevent further attacks. Stress and anxiety are believed to precipitate attacks both of Meniere’s disease and migraine, although the evidence is only anecdotal. Reassurance should be given that Meniere’s disease will not continue to cause attacks for the rest of the person’s life. In the early stages, prolonged remissions are common and indeed some fortunate people will never have a flare up of the disease again. In the less fortunate sufferers the attacks will continue often in clusters at intervals, but the attacks will get weaker and weaker until the attacks are barely noticeable. There are treatments available to lessen the frequency and severity of the attacks until a natural ‘burn out’ occurs. It is very reassuring for the sufferer to know these facts and by lessening the stress and anxiety, a positive start is made.

STEP TWO - A salt reduced diet

It is widely believed that salt (sodium chloride) increases endolymphatic hydrops and can precipitate attacks of vertigo. The concept was introduced in 1932 by Mygind and Dedering(1). Dedering suffered from Meniere’s herself and documented how increased dietary salt caused her attacks. Over the years many sufferers have given anecdotal evidence that salt exacerbates their condition. Scientific evidence is not possible as the sufferer would easily know if they were being given salt or not. However Harrison and Naftalin(2) did undertake a study when they gave a 5G supplement of salt to the diet of Meniere’s sufferers. They showed that this often was followed by an attack of vertigo and there was an increase in the urinary sodium output at the time of the attack. The study had to be concluded as it distressed many of the participants and nowadays I doubt if ethical approval would be given again.

The author is convinced that controlling dietary salt is important and very effective especially in sufferers who had a high salt intake prior to the onset of the condition. Similarly, scientific evidence for reducing sugar, caffeine and nicotine is lacking but many sufferers have found these extra steps to be helpful.

STEP THREE - Diuretics and Urea

Diuretics are medicines that reduce the amount of water in the body. Most diuretics also reduce sodium levels. Diuretics are commonly used to treat high blood pressure and oedema. Klockhoff and Lindblom in 1967(3) reported success using hydrochlorothiazide. Subsequent studies also reported success but were criticised for poor design and control. The best designed study using Diazide (hydrochlorothiazide and triamterene)(4) showed a statistically favourable effect on the vertigo but did not halt the progression of hearing loss or affect tinnitus.

There are significant side effects caused by diuretics. The commonest side effect is postural hypotension, or on standing the blood pressure can fall significantly making the person dizzy. On starting a diuretic the person should not stand up too quickly and often within a few days this side effect seems to lessen. More seriously prolonged use of diuretics can lower potassium which has effects on the heart. After every few months a blood test is necessary and in some cases a potassium supplement is required.

There is no need to restrict water intake. The Japanese have treated Meniere’s sufferers by giving them 70ml of plain water per kilogram body weight daily as this lowers sodium which decreases intracellular fluid. This is only helpful when there is a bathroom handy!

Urea is an osmotic diuretic which has a very simple chemical formula CO(NH2)2. It is a major constituent of urine and the fertiliser put on the ground is often made from cattle urine. Those who take urea to control their Meniere’s disease will be relieved to know that the urea used medically is made chemically rather than biologically! Urea acts by drawing intracellular fluid into the blood stream and it probably removes some endolymph. The other osmotic diuretics are glycerol which tastes like diesel oil and isosorbide which tastes sickly sweet. Studies of osmotic diuretics show that the hearing improves temporarily both subjectively and objectively (electrocochleography shows a reduction in the summating potential). Hence urea is given as a diagnostic test for Meniere’s because the hearing only improves when endolymphatic hydrops is present.

Usually 30 grams of urea is taken in a minimum amount of fluid as it tastes foul. Excessive urea damages the liver and kidneys so it cannot be given to people with liver or kidney problems. In otherwise healthy people the maximum safe daily dose is 1gram / per kilogram body weight.

A few hardy souls take urea on a daily basis to prevent attacks and to maintain hearing. Most sufferers only use it as a ‘Cinderella medicine’ which guarantees freedom from attacks for 3-4 hours. It can be used to attend important events such as weddings but when the clock strikes the third time, any protection from vertigo is lost!

STEP FOUR - Betahistine

Betahistine dihydrochloride (SercTM) is a vasodilator; a substance which dilates blood vessels. The use of vasodilators is based on a concept that reduced blood flow in the cochlea results in an accumulation of metabolites and a consequent rise in osmotic pressure causing a transfer of fluid into the endolymph compartment. The stria vascularis is the blood vessel within the endolymph compartment of the inner ear and it is hoped that vasodilatation of this vessel decreases the metabolite accumulation and increases the radial absorption of endolymph.

Based on this theory in the 1950’s the cervical sympathetic nerve supply to the ear was destroyed surgically(5). Apart from its effect on the ear, it also caused constriction of the pupil of the eye, loss of sweating on the same side of the face and a dropping eyelid. It was soon abandoned!

Less dramatically, a vasodilator medication called nictotinic acid was given causing flushing of the face. Betahistine hydrochloride replaced nictotinic acid as it did not cause flushing of the skin and some experimental evidence showed it did have an effect on the blood flow(6) through the cochlea. However, the original hypothesis that there is a vascular cause of Meniere’s disease is in doubt.

Betahistine (Serc®) has become the most utilised treatment in Europe. The long term use is supposed to lessen the likelihood of future attacks of vertigo and to halt the loss of hearing. There is no advantage in taking it when an attack is pending or during an attack. The scientific evidence is blurred and the Cochrane review states that ‘there is insufficient evidence to say whether betahistine has any effect on Meniere’s disease(7).

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# THE MENIERE’S TREAMENT LADDER (part two)

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I introduced the Meniere’s treatment ladder in part 1 and explained the first four steps, covering reassurance, the low salt diet, use of diuretics/urea and betahistine.

The ladder climbs from the most benign therapies to the most radical surgery.

In this instalment I will cover the next five steps; oral steroids, grommets overpressure and intratympanic therapies.

The final 4 steps, which look at surgical treatment will be continued in part 3.



*CI - cochlear Implant*

 *VI – Vestibular Implant*

## STEP FIVE -Oral Steroids

In Australia, the usual steroids prescribed are prednisone or prednisolone which can be swallowed (oral medication). Steroids reduce inflammation and are given for various conditions including facial palsy and sudden hearing loss. The use of steroids for the treatment of Meniere’s Disease (MD) is based on the concept that an inflammatory process results in the formation of excess fluid within the membranous labyrinth (endolymphatic hydrops), and fluctuations in the endolymph level cause the attacks of vertigo and other symptoms. The inflammation may result from a reduced immunological response to a virus or to an auto- immune are up. (Auto-immune diseases such as asthma, rheumatic arthritis are when the body falsely recognises its own tissues as foreign and mounts an immunological attack on them). Steroids have been shown to be effective in treating most auto- immune diseases.

Some support for an auto-immune theory is the finding that HLA (human leucocytic antigen) deficiencies are found in some MD sufferers. This deficit is related to an abnormality on chromosome 6 and this probably accounts for MD occurring in family members.

The Cochrane database(1) has given support to the use of intratympanic steroids so it is hoped that giving the steroids orally will have the same effect. The usual dosage is 60-80mg daily for 5-7 days and then a tailing off dosage. Some clinicians prefer to tail off very slowly and a maintenance dose of around 10- 20mg daily for some weeks. Usually the steroids are given when there is a cluster of attacks of vertigo occurring.

A study by well-respected researchers(2) in Los Angeles failed to show any improvement in hearing levels after oral steroids and the Cochrane database review was also inconclusive. Nevertheless many physicians do believe anecdotally that oral steroids help to halt a series of attacks.

The problem with oral steroids is that they affect the entire body and can cause significant side effects. The most serious side effect is bleeding in the stomach and oral steroids should be stopped immediately if significant indigestion occurs. Oral steroids can also adversely affect blood sugar levels in diabetics. Steroids may alter the bodies defence against infections and long term use leads to weight gain and fluid retention. The advantage of intratympanic administration of steroids is that a larger dose is received into the inner ear and virtually none of the steroid leaks into the rest of the body.

## STEP SIX - grommets

Over 70 years ago, Professor Tumarkin in Liverpool, England championed the value of grommets to stop Meniere’s attacks.

Grommets are tiny tubes inserted into the eardrum to equalise the pressure within the middle ear cleft. The concept is that pressure fluctuations within the middle ear can be transmitted to the inner ear and excessive pressure changes can increase the formation of endolymph causing attacks of vertigo. It is also suspected that the feeling of aural fullness is due to an increased sensitivity within the middle ear.

Short term grommets can usually be inserted using local anaesthetic in the ENT surgeons’s office unless there is a narrow ear canal. If there is a benefit, the grommets can be exchanged for longer staying T tube grommets.

There is a risk of infection especially if contaminated water enters the ear. Swimming in public pools is unwise and swimming in the sea only possible if ear plugs fit effectively. Wax can sometimes block the grommet making it ineffective. There is always a risk of a permanent perforation of the ear drum after the grommet extrudes, especially if a long stay grommet is inserted.

The Cochrane database review states that there is insufficient evidence that grommets are beneficial although there are some compelling studies. Montandon and co-workers(3) reported a series of 28 patients in which 82% benefitted with a marked reduction in the vertigo attacks. No placebo group was possible.

## STEP SEVEN - Overpressure (The Meniett device)

The Meniett device delivers small calibrated pulses of pressure through a grommet into the middle ear cleft to ‘milk away’ endolymphatic hydrops.

In Sweden, where MD is a common disorder, studies were undertaken using hyperbaric chambers. These are the decompression chambers into which a diver is placed after a diving accident when ‘bends’ has occurred. Hyperbaric oxygen chambers are used to force oxygen into poorly healing tissues. It was found when a MD sufferer was placed into a hyperbaric chamber, there was often a temporary improvement in symptoms with less fullness in the ear and better hearing. Too great a pressure could actually cause an attack of vertigo to occur so the pressure change had to be very carefully monitored.

In 1980, Barbara Densert and her husband Ove Densert began a series of studies to develop the low pressure unit which became commercialized in 1999 as the MeniettTM device. The concept is that these small pressure pulses gently squeeze the endolymph compartment and disperse the excess fluid either to the endolymphatic duct or back into blood vessels. A number of protype devices were trialled in Sweden and eventually this work led to the commercial development of a machine which is available in Australia and which has been the subject of several scientific trials.

To use the machine, a grommet must be inserted initially into the ear drum to allow the pressure pulses to pass through to the middle ear. The MD sufferer is then asked to wait about a week to ensure the grommet is stable.

The MeniettTM machine should be placed on a level surface. The user must be sitting in an upright position (not lying down). The ear piece is then held in the ear canal and a button to commence the programme is pressed. There are some simple lights on the machine to indicate the progress. The machine is designed to deliver small pressure pulses in a safe manner. First it checks that an airtight seal has been achieved. Then the machine calculates the middle ear pressure, applies a static pressure of 10 cm H2O and then applies pulses of 15 cm H2O. A series of four pulses is presented three times with a gap of 4 seconds, there is then a period of 45 seconds without pulses. This paradigm is repeated three times so the entire treatment period is about 3 minutes. It is recommended that while the MD is active, the machine is used at least 3 times a day.

Gates and co-workers(4) have conducted a controlled trial of 67 patients involving 4 centres in the USA which was carefully designed and showed a positive outcome for the patients when using the device.

Despite this the Cochrane Data base(5) remains uncommitted and suggests that further evidence is needed before they can endorse the treatment.

In conclusion: the MeniettTM device is a mechanical means of controlling the excess endolymph in MD sufferers. It is not expected to be an absolute means of controlling MD. The results can be expected to equal the results seen with salt reduction or the use of diuretics. The advantage is that the treatment can be combined with the other methods of fluid reduction. There are few disadvantages other than those of having a grommet permanently in the ear. Unfortunately the device is expensive and costs over $5000 although the company (Medtronic) will lease the device so individuals can trial it at a lesser cost. Some costs are refunded by some private health insurers on special request but those without private health have to purchase the device for themselves as Medicare does not cover this item.

## STEPS 8 & 9 – Intratympanic therapies

Two medications which are most commonly delivered by intratympanic injection have both been studied and found to have proven benefit. These medications are intratympanic steroids(6) and intratympanic gentamicin(7).

The method for delivery of intratympanic medications requires the patient to lie on a couch and the ear is examined using an operating microscope. The tympanic membrane (eardrum) in the affected ear is anaesthetised. I use a tiny droplet of phenol but other ENT specialists may prefer to use an analgesic ointment. Often the patient is asked to check the labelling to ensure the correct medication is drawn up into a small syringe. Using a long intrathecal needle the medication is delivered through the eardrum into the middle ear cleft. The patient feels little pain but feels the ear is filling up with fluid and some may pass down the Eustachian tube into the throat. The patient then keeps lying down for at least 5 minutes so that the medication can perfuse from the middle ear into the inner ear. There is usually no feeling of dizziness and the patient can go home within a few minutes. The ear should be kept dry for 2 days to prevent infection and the patient should not keep squeezing air up into the ear from the back of the throat so the small perforation caused by the needle can close.

## Intratympanic steroids

Usually 2ml of 40mg/ml of dexamethasone is drawn into the syringe but less than half will fill the middle ear cleft. The amount of steroid entering the inner ear is unknown and variable. The steroid solution will pass along the basal coil of the cochlear before it is removed by the blood vessels within the ear. Some steroid will also pass up the spiral ligament into the vestibular portion of the inner ear.

Steroid injections have a powerful anti-inflammatory effect and have been used in other branches of medicine: for example, to treat inflamed joints. It is suspected that recurrences of Meniere’s symptoms have an inflammatory basis. The intratympanic steroids can halt a series of attacks and have been shown to help preserve hearing.

There are minimal risks involved for the patient. Complications of treatment are exceedingly rare. It is important that the tip of the needle is placed safely through the eardrum without disrupting any of the ear bones. The possibility of a persistent perforation or infection is also very rare.

## Intratympanic gentamicin

The concept is that the affected ear has a faulty vestibular output which suddenly flares up causing attacks of vertigo. It can be likened to a faulty engine on an aeroplane which suddenly flares up spinning the plane around. Surgery can be performed to totally destroy the output of the faulty vestibular labyrinth (labyrinthectomy and vestibular nerve section). However, it needs to be considered that Meniere’s disease will eventually ‘burn out’ and the attacks of vertigo will peter out and eventually cease. After ‘burn out’, approximately 40% of the balance function remains in the ear which is useful to prevent unsteadiness especially for those who develop bilateral disease and older persons. The adage is ‘don’t offer therapies which make the outcome worse than the natural outcome of the condition’. The problem is that it can take years of distressing vertigo before the eventual ‘burn out’ occurs. Gentamicin is a vestibular toxic medication which destroys vestibular function. Several intratympanic doses can be given to completely destroy all the balance function of the ear without the need for surgery. However a major advantage of intratympanic gentamicin is that it is possible to give only limited amounts intratympanically to reduce the balance in the ear sufficiently to reduce the severity of the attacks so that the patient can retain some balance function while waiting for ‘burn out’ to occur. Balance function in the affected ear is gradually lost as the disease progresses. It is hoped that there is little difference in the amount of residual balance function after limited gentamicin treatment compared to the amount of balance remaining after the natural ‘burn out’ of the disease, although several distressing years are avoided.

There is no immediate feeling of dizziness, but usually 2-3 days later the patient may feel some unsteadiness. The unsteadiness will increase if further intratympanic injections are given, especially when the goal is to destroy all the faulty vestibular function in the affected ear. Often a balance test is performed 6 weeks after the last dose to discover the effectiveness of the treatment.

There are the same possible complications as already mentioned for intratympanic steroid therapy. In some patients (5-10%) the gentamicin has little or no effect on the inner ear and sometimes the tympanic membrane is lifted surgically to find out why the medication did not perfuse into the inner ear. Sometimes there are fibrous adhesions blocking the windows of the inner ear and a pledglet of gelatine foam is soaked in gentamicin and applied directly. This can have a marked effect losing a lot of inner ear balance function after just one application. Rarely, intratympanic delivery of gentamicin can have a similar effect (1%). There is a risk to the hearing if several applications are given and typically some of the high frequency hearing can be lost.

Vestibular physiotherapy has a major role in re-establishing balance after gentamicin therapy. Again using the aeroplane analogy, the pilot has to relearn the controls after losing the output of one of the engines.

It is expected that other medications will become available which can be applied by the intratympanic route. For example, trials of OTO-104 are being done in the US and EU by a commercial company called Otonomy. This is steroid gel which is placed into the round window niche and it is hoped that it will have a longer lasting effect.



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# THE MENIERE’S TREAMENT LADDER (part three)

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*CI - cochlear Implant*

 *VI – Vestibular Implant*

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”.

## 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 ES 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 ES 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 ES 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(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.

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