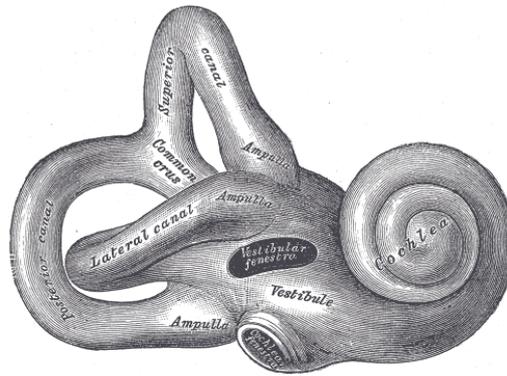


Andrew Hugill
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Ménière's Disease and me.



In 2009, I was diagnosed with Ménière's Disease. MD is an incurable condition that combines four distressing symptoms: vertigo, hearing loss, tinnitus and aural fullness. My MD is bilateral (affects both ears) and is particularly virulent. It has been treated with a chemical labyrinthectomy, which intentionally destroyed the balance function in my right ear, and a range of drugs and dietary modifications. It has also led to severe hearing loss and varying levels of tinnitus. This has had consequences for both my personal and professional life and especially my relationship with music.

The aim of this document is to describe the history and development of my condition. I have to admit that I have kept it mostly secret for many years until now. MD is an invisible disability that is hard for non-sufferers to understand. A person with MD can seem to be either drunk or incapable. The medical profession is often similarly baffled. I was told more than once by doctors that MD is "over-diagnosed" and that my symptoms were more than likely something else. In fact, I have the classic MD profile which has evolved over time in exactly the way that those who know about the condition would have predicted.

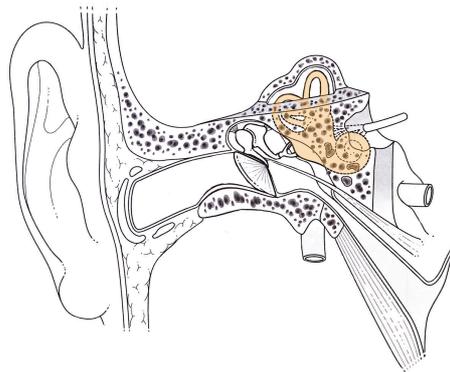
My secrecy was partly a matter of personal pride (not wanting to be perceived as weak) and partly a matter of professional fear. The latter arose from the high-powered position I held at De Montfort University and a strong suspicion that some of my colleagues and managers would take a dim view of a Professor of Music and Director of the Institute Of Creative Technologies with these shortcomings. To what extent my fears were justified I will never know, but I am immensely grateful to my dear wife Louise, my friend Simon Atkinson, and my two personal assistants during this period, Rebekah Harriman and Jos Atkin. They provided the personal support and professional cover without which I would probably have given up work altogether. I also thank my consultant, Professor Peter Rea, of the Leicester Balance Clinic, whose interventions have restored my life to some kind of normality, and Dr Ian Cross, a GP who made the referral to Professor Rea's clinic in the first place.

I have decided to write this document now following my attendance at the ‘Hearing Aids for Music’ conference at the University of Leeds, September 14th/15th 2017. This inspiring event opened my eyes to the possibility that one could admit to these things in public and still maintain a professional career of distinction. In fact, I have been opening up to people about my MD more often in past few years anyway, because the professional environment at Bath Spa University is rather more sympathetic than I found it at De Montfort. Nevertheless, the majority of people I know are unaware that I have this disability, and I think it only right now to present a full account of it, both for their benefit and for the benefit of any other MD sufferers who may be reading this. I found myself desperately searching for other people with a similar condition in the early days, and derived some comfort from the knowledge that I was not alone. I hope this will help other people, and I am always keen to hear from fellow-sufferers.

About Ménière’s Disease

MD is named after Prosper Ménière, a French doctor who first identified the condition in the early 1800s. For those who know me and my work on ‘Pataphysics’, it is quite ironically appropriate that I should have an obscure French condition that affects the only region of the body that contains a spiral (the cochlea)! For those mystified by that comment, my book *‘Pataphysics: A Useless Guide* will provide an explanation.

MD is a disorder of the inner ear, which is shown in brown in the next illustration. Since this contains balance functions *and* hearing mechanisms, both of these are affected. Nobody knows what causes MD, nor how it builds up once it has begun. However, all MD sufferers have endolymphatic hydrops, which is an excess of fluid in the semicircular canals. The fluid flows throughout the inner ear and contains various salts or electrolytes which are critical to the balance function in particular. An excess of this fluid causes a feeling of fullness in the ears and episodes of vertigo, as well as damage to the tiny hairs (cilia) that enable hearing.



MD affects either one or both ears. I am bilateral but, mercifully, the left ear is less affected than the right. MD varies from person to person and even from day to day or hour to hour. The condition always progresses over time, but its severity varies. There is consequently a “ladder” of treatment that is deployed depending on patient need. I have climbed this ladder all the way to the top.

The first step is diet modification. Salt is a major trigger for MD symptoms, so the aim is to reduce salt intake to under 2.5 grams per day (the recommended amount for

a normal adult is 6 grams). There are then various drugs that are taken to relieve symptoms, including diuretics, betahistine and anti-nausea medicine. Early surgical interventions may include the fitting of a grommet to relieve fluid pressure in the inner ear. Transtympanic micropressure pulses may be helpful, and self-administered using a machine. Alternative therapies include acupuncture and herbal remedies, massage and meditation techniques. With every one of these, I experienced some temporary benefits, but not sufficient to substantially alter the progress of the disease.

I didn't have steroid treatment, but in recent years this has become a preferred next step, with steroids being injected or soaked into the ear. I also did not have a saccus decompression operation, which involves surgically releasing fluid from the inner ear. I have heard mixed reports about how effective this might be. Instead, the severity of my condition meant that I went on to the top rung of the ladder with a labyrinthectomy. The idea of this is to remove the labyrinth of the inner ear so that there is nothing for the MD to "work with" in order to create vertigo. It does not cure MD, but it does put a stop to the debilitating dizziness. Unfortunately, it also destroys your balance function, so it is an extreme measure.

There are two ways of performing a labyrinthectomy: physical removal via surgery, or chemical removal with drugs. In my case, I had a series of gentamicin injections into the inner part of my right ear. Once that was completed and I had recovered sufficiently, which took about a year, I needed no more treatment for vertigo. Even so, I do still sometimes get short-lived dizzy episodes. But overall, my focus has changed to managing my other symptoms of hearing loss and tinnitus. Aural fullness has also largely dissipated, thanks to the gentamicin but also the grommet which I had fitted in 2011. I am now deemed to be at a stage called 'burn out', when the MD is still active but has little or no effect on balance. This is just as well: if my left ear were to worsen, then a further labyrinthectomy would be impossible because I would have no balance at all. So, there is little or no further treatment available and my hearing continues to deteriorate. I still maintain a low salt diet, but otherwise I just monitor my progress with my consultant, while trying to intervene with hearing aids and psychological adjustment.

My history with Ménière's Disease

One of the worst aspects of MD is uncertainty. After each attack, I would try to explain why it had happened, blaming food, stress, my prescriptions, etc. In the end, none of these explanations was convincing, and I came to the conclusion that the disease just does what it does and there is no particular rhyme or reason. But it always meant being uncertain whether an attack would occur that day, whether the hearing would get worse, whether this or that treatment is right at a given time. The uncertainty continues to this day, since the condition never goes away.

I first started to get vertigo attacks in about 2007. I don't remember the first one, but I do remember a couple of early ones: one in my studio which completely knocked me over and sent the world spinning around wildly; and another in a café in central Leicester. This occurred just after I had received some Chinese medicine treatment, involving acupuncture and herbs. Naturally, I blamed the treatment, but in retrospect I know that had nothing to do with it. However, it is significant that I had been seeking

treatment in the first case, because I was already aware that there was some kind of problem but did not know what it might be.

At this stage, I was not aware of any hearing loss or tinnitus and, like so many people, put the vertigo down to tiredness, or stress, or a virus, or anything else I could find to blame. The months went by and the vertigo became steadily more frequent and more violent and now tinnitus and hearing loss started to become apparent to me. As a composer and Professor of Music this was, of course, very alarming indeed. Being a researcher, I started to investigate my symptoms myself. It became increasingly clear to me that Ménière's Disease was a real possibility, so I sought advice on that basis, and experienced the scepticism of the medical profession. It was only when I was referred to Professor Rea's practice in Leicester that I finally had confirmation that all four of the necessary symptoms for MD were present in my case.

So, I began climbing the treatment ladder with a low-salt diet. This was an interesting challenge, because the amount of salt that is added to food is unbelievable! The situation has improved in recent years, but back in 2009 it was almost impossible to find meals that did not include large quantities of additional salt. I had to stop eating curries, which was heart-rending, although later on I was able to find one or two curry restaurants in Leicester which were willing to prepare me a salt-free meal. Certain other foods seemed to trigger vertigo attacks, such as caffeine (which I have not taken since 2010), bread and, strangely, rice, to which I seemed to be allergic for a time. These days, I can eat rice without a problem, but back then it would cause an instant attack.

It is hard to describe a full vertigo attack. There is some advance warning, as the ears fill with fluid, the hearing plunges to muffled, and the tinnitus increases to a screech. This preliminary period could last anything from a couple of minutes to half an hour, although it tends to get shorter as the disease progresses. After that, the rotation begins quickly and sustains itself for up to 5 hours. The sensation is of the whole world spinning around you. This is accompanied by violent vomiting. I would be unable to move and could not stand even the slightest motion. I would stare at a fixed point for hours, trying to stop the spinning. My eyes were affected, a bit like in migraine (there is some overlap between MD and migraine), with any bright lights or striations causing distress. I would hold a piece of white card in front of my eyes so that there was nothing to look at. I also could not tolerate noise, or any sound at all.

Some of the worst attacks happened while out and about. Since I do not drive (thank goodness!) it was my darling wife Louise who would be called and would have to come and rescue me. I would go home in the back of the car being violently ill, and then stagger into the house hoping the neighbours would not see me. On one occasion, I was attending a lecture at the Leicester Literary and Philosophical Society at New Walk Museum. Car access is not great there. The lecture was on birds, and I was very interested, so when I felt an attack coming on I stayed in the hall longer than I should have done. The result was that I staggered out in the middle of the lecture and then down the street towards the railway station, texting Louise on the way. I reached the station and had to stand in the pick-up area, swaying perilously while staring at the ground. Anyone who saw me that night would have assumed I was drunk.

On other occasions, an attack would happen while I was at work. I would retreat to my office while my PA held the fort and refused to allow people in to see me. Meanwhile, Louise would be called again to come and surreptitiously take me away. I was determined not to reveal my condition, because the Institute Of Creative Technologies was at an early and fragile stage of development and any sign of weakness on my part could have damaged both my and its future. I'm pleased to say that it still flourishes to this day.

These are just a few examples of the effects of an attack. I had so many that I lost count. As you can tell, having a supportive partner is a wonderful thing. Louise did what was required, but she also would not allow me to sink into self-pity or give up my work. This level-headed approach was vitally important, to keep the thing in perspective and maintain a positive mental attitude. As a result, I did not become too obsessed with what was happening and managed to cope fairly well with the psychological effects of losing my hearing, and the ever-increasing tinnitus.

An attack would usually end with my falling asleep, which was a blessed relief. I would wake up when the vertigo had stopped, but sometimes it continued after I awoke. Either way, the end of vertigo would lead on to "brain fog", a ghastly condition in which the brain seems to be wrapped in cotton wool and refuses to function adequately. It would also be accompanied by screeching tinnitus. This would last several hours, but always cleared in the end, at which point I felt fine and would embrace life and work again with gusto, trying to pack in as much as possible before the next attack.

The treatment ladder led on to SERC, a betahistine that I then took for ten years in varying doses. It certainly does help, by dilating the blood vessels in the inner ear and increasing permeability. It also seems to be a completely benign drug in other respects. Even so, it was insufficient to treat the condition fully, so I then had a grommet fitted in 2011, to relieve aural fullness. This is a common surgical procedure with few risks or side effects. Mine was a T-tube grommet, which stayed in place until I had it removed in 2016. The reason for its removal was that I was getting repeated ear infections, as the eardrum tried to reject the foreign body. Having taken it out, I now have a permanent hole in my eardrum, which has the disadvantage that I must wear protection in the shower or when swimming, but the great advantage that I don't experience any ear pressure problems when flying!

For a time, I had some success with a Meniett device. This is a machine that transmits acoustic pulses through the eardrum via a hand-held tube. The idea is that it stimulates circulation of fluid in the inner ear and so slows build up of the hydrops. This was quite pleasantly comforting, and did relieve symptoms, but in the end did not do so in a sustainable manner.

Having MD is expensive! I was able to have private treatment and I paid for this to happen as quickly as possible. Since the condition is not life-threatening, it tends to get relegated down the priority list in the NHS. I took the view that treating my condition was an investment in the future, and so it has proved, but this option is not available for everyone. I spent a lot of time on the MDUK message board, sympathising with and trying to encourage others. I know from the inside how destructive of lives and jobs this thing can be.

After about three years of worsening symptoms, Professor Rea finally advised gentamicin treatment. This involved a series of injections of this powerful antibiotic into my inner right ear, which chemically destroyed my balance function. At first, this made things worse, as I could barely walk down the road. I tried Vestibular Rehabilitation Therapy, but the exercises just triggered vertigo attacks, so I had to stop. But gradually things improved to the point where life was, to all intents and purposes, normal again. I now balance with my eyes, which means that if I shut them and walk I fall over. I also hate the dark, because I cannot see to balance properly, but fortunately there is so much light pollution in modern life that I do not encounter a problem very often.

Most of the time now, I do not have vertigo at all. I have also opened up a bit more about the condition to various friends. Occasionally I become aware that the MD is still doing its thing and, if I still had a balance function, I should be spinning. But even this is ignorable. I have had some mild attacks, for example when my medication for cholesterol was changed. These seem to be the result of the salts that modern pills are 'cut' with, any change in which affects my whole system. Once I went back to my previous prescription, everything was normal again. Sometimes, there is no obvious cause and I have to lie down and sleep for an hour or so, after which I am fine again.

Hearing Loss and Tinnitus

The most powerful consequences of MD for me today are hearing loss and tinnitus, rather than vertigo and aural fullness. During the vertigo phase, I was somewhat less bothered about these because, frankly, nothing is worse than vertigo. But these days they are increasingly important to me.

Tinnitus is reaching epidemic proportions as a generation has been exposing itself to prolonged loud noises both in daily life and via amplified music. I have always been aware of the dangers and taken precautions, indeed I wrote a warning section about this in my book *The Digital Musician*. It is somewhat ironic, therefore, that I suffer from it today. Tinnitus seems to be a cognitive problem, as the brain tries to hear sounds it cannot hear and consequently generates noise in compensation. But its true pathophysiology is really obscure and unknown. It can be a most debilitating condition, especially for musicians, and has frequently resulted in the depression or even suicide of the sufferer.

Even though my tinnitus is at times very loud, I have always managed to "hear past it" and am not psychologically troubled by it. I will never know silence again, but I accept this as part of my life now. The tinnitus, like everything else in MD, fluctuates wildly in both intensity and pitch, and is worse in my right ear than my left ear. At the time of writing, I have a fluctuating sizzle sound in my left ear and a large whooshing in my right that sounds like a distant aeroplane, combined with a small collection of continuous high pitches. However, this will doubtless have changed by tonight.

To get to sleep, I find human speech is normally sufficient to distract me from the tinnitus, so I listen to the radio, or podcasts, or audiobooks, using an under-the-pillow loudspeaker. I have also on occasions used an app such as WhiteNoise or the Relief app that comes with my hearing aid. I turn to these when the tinnitus is especially

loud, because the frequency masking techniques they use are more effective at cancelling out the tinnitus noises.

Hearing loss is a more challenging problem. My right ear has severe loss, and my left ear mild. This does mean that I can function apparently normally in most situations, but there are nevertheless some limitations which have to be overcome. The hearing aid in my right ear assists the relatively good hearing in my left ear, adding up to a viable hearing system which lets me engage effectively in one-to-one conversation as long as I can see the other person to lip read, and have my left side turned towards them. Group conversation is another matter and is generally extremely challenging. People wonder why I am constantly leaning in towards them, or manoeuvring around them to get as much a sound as possible into my 'good' ear.

The profile of the hearing loss is bizarre, with the low frequencies having gone first (this is typical of MD) and a smorgasbord of peaks and troughs in other frequency bands. Furthermore, this changes on a daily basis and even in real time in response to external stimuli. Therefore, it is quite different from age-related hearing loss, or indeed the loss of someone who is profoundly deaf.

This has naturally had huge consequences for my music. I decided to stop making live music altogether when it became apparent that I could not perform adequately. However, that decision had much to do with vertigo at the time, so it may yet change. My composition has moved more or less exclusively to digital work, where I can control levels effectively and do not cause problems for other musicians. However, this is also not entirely satisfactory. It removes the social dimension of music, which is so important. Also, it still presents real challenges to my ears. Electroacoustic composition, for example, relies on fine and discriminating listening, and is invariably linked to a minimum of two channels and often as many as eight. I have found myself somewhat alienated from this form of music, which I have done so much to create and promote, by its intolerance of anything less than perfect hearing.

Somewhat masochistically, I have worked on some directional compositions and analyses (trying to avoid showing weakness again!) and have ended up relying on PhD students to be my 'ears' and confirm that what I imagine will be heard is accurate. What I actually hear is distressingly weird, with individual tones splitting into a multiple frequencies like a ring modulator, unwanted sonic artefacts coming either from tinnitus or from the ear itself, very limited perception of dynamic range, no ability to localise, and a general tendency to hear all pitches as sharp. My 'good' ear saves me from total disaster, but even this cannot be trusted completely. In earlier years, I was renowned for having accurate 'ears', but now I have to face the fact that this is no longer the case. This is hard to accept.

The situation is equally bad with regards to speech. I really struggle in any conversation where there is any kind of background noise. On very many occasions, people have spoken to me and I have not heard them. I am sure that I have also said the wrong thing many times in situations where I cannot hear someone properly and have chosen to pretend that I can hear, rather than constantly saying 'I beg your pardon'. This is really rude on my part, and I have tried to adopt a policy of explaining the problem to people first, but sometimes this is simply not possible. My job involves talking to people all the time, so many conversations happen "on the fly"

and in uncontrolled environments. Meetings present particular challenges, especially in large rooms with prominent echoes. I am pretty good at lip-reading. I am largely self-taught but did attend some classes at DeafPlus in Bath and also used the LipReader training software by David Smith. And I have my hearing aid (of which more in a moment). Even so I am afraid I miss things. Women's voices, which are softer, are often a big problem.

I got my first hearing aid back in 2012, under the excellent advice of Claire Marshall, my audiologist at the Leicester Hearing Centre. This was a Siemens Tek, which at the time was the bee's knees. It was programmable via a small handheld device and offered special programmes for different situations (outdoors, restaurant, music, etc). It did help quite a bit with these, but was mostly poor for music. Hearing aids are engineered primarily for speech. Music tends to present a much bigger challenge, with wider dynamic and frequency ranges, etc. The compression and noise reduction functions in digital hearing aids actually work against the enjoyment of music, or at least live music. The Tek could not distinguish, for example, between a flute and feedback, so would suppress them both just the same.

For a time, I moved to an NHS hearing aid which is free (or nearly free). Despite the proud boasts of the NHS audiologist that this device was every bit as good as anything I could get privately, it was not very well suited to an MD sufferer. This is because NHS hearing aids are designed for people with age-related hearing loss, and so boost the treble frequencies and tend to ignore the lower end. Since the worst of my hearing loss is the other way around, this resulted in a highly tinny and sibilant sound which was ok for speech but completely hopeless for music. So, in both instances, I would usually take out my hearing aid when making music.

My most recent aid has coincided with a further decline in my hearing, which occurred in late 2016/early 2017. The new HA is a GNResound3D. I am still getting to grips with this system, but it seems to offer better possibilities for music. The speech functions are excellent, and the whole thing is controlled from my iPhone via an app which allows me to adjust directional focus and edit broad frequency bands. It also provides a direct link to my audiologist. I have supplemented this with some free software called EQHearAid, which offers a full graphic equaliser that can be adjusted on the fly. This is great because my hearing changes so much on a daily basis. There are still problems with holes appearing as sound transitions from one frequency band to another, and bass remains an issue because the ear mould I wear has a ventilation shaft which immediately reduces the capacity for low frequency management. Nevertheless, this HA is a great improvement on anything I've encountered so far.

Living with Ménière's Disease

To those who have read this far: thank you and well done! I don't spend my days lamenting and suffering with MD. It is just a condition that I live with and, since the vertigo has more or less ended, is one that does not intrude too much on daily life. Yet there is still plenty to grapple with in the future. In particular, I feel the need to continue to address my hearing issues. In the first instance, I want to be more open and honest about the problem, which is partly why I have written this document. But also I want to try to make a contribution to improving life for other people. For that reason, I am developing a research project that will focus on music and MD. It will

involve a cross-disciplinary team including clinicians, audiologists, psychologists and musicians. I hope to work with a hearing aid manufacturer too. This is an unresearched field at present that would greatly benefit from an increased scientific understanding.

To anyone who has recently been diagnosed with MD, I would say that it is not the end of the world (although at times it might seem so). It will however have a transformative effect on your life, so finding out as much as you can about it is important. There are online resources, but do check their reliability because there is also quite a lot of rubbish on the internet. Various purported “cures” are not to be trusted (and I have tried one or two of them). Join the Ménière’s Society. Every MD sufferer is different and has a different experience, but treatments exist for all levels, as my story demonstrates. In the past year I have travelled abroad frequently to deliver papers at conferences, including trips to Australia, Poland, Belgium and Canada. Life can return to something like normality!

One strange fact is that I now live aboard a narrowboat. During the vertigo phase, I certainly could not have done this (even though I wanted to), but now I am happily living in a house that rocks slightly. Whether it is because I have no balance function on one side, or whether it is because my brain has to work harder at balancing me on a boat, I find it more stable than being on dry land. MD is full of strange paradoxes like this. No doubt, if serious vertigo ever returned I would have to get back to dry land, but for now I am more than happy in my ‘mobile’ home!

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