

Dr. Jason Barnes:

Hey there, welcome to another episode of ENT in a Nutshell. My name's Jason Barnes and today I'm joined by neurotologist Doctor Brian Neff, and we will be discussing Meniere's disease. Dr. Neff, thanks so much for being here.

Dr. Brian Neff:

Thank you.

Dr. Jason Barnes:

I first just wanted to start with presentation because I feel like this is something that can be hard to nail down, especially as a resident sometimes. Can you tell us how patients with Meniere's disease will present to your clinic?

Dr. Brian Neff:

I think it's most important to kind of think of it as how do I talk to a person with a chief complaint of vertigo? Really, the first thing you're going to want to ask is have them describe their symptom. And I know it's cliché, but without using the word vertigo or anything they've been coached with, so they need to describe, is it flipping over, is it spinning, is it rocking? Most of my Meniere's disease patients will come in with vertigo. Occasionally there'll be a few that'll come in with hearing chief complaints, but most are complaining of dizziness.

Dr. Jason Barnes:

And what kind of dizziness are you trying to tease out here?

Dr. Brian Neff:

When you're seeing somebody with Meniere's disease, you want to establish what their dizziness feels like to them. Vertigo is essentially a sensation of environmental movement in their visual surround when they're still, so it can be flipping over, feeling like you're falling to one side or the other, rocking, spinning is the obvious one, but it shouldn't be things like lightheadedness or I'm graying out or presyncope. I've heard that my brain is spinning before, that's not vertigo. So those are what you want a description of for your dizziness.

Dr. Jason Barnes:

And when you see these patients, Meniere's is classically described with hearing loss and tinnitus, how much do you pay attention to that when they first present?

Dr. Brian Neff:

So when you've asked about dizziness, one of the things that you want to establish is, is there any associated symptoms? So, hearing loss is usually the first thing I ask about, and you want to know the timing of it. Is it during, before, or after? Sometimes with Meniere's disease though, it doesn't have good timing. And the patient just says that my hearing's been going down over the last two years. That doesn't necessarily mean it's not Meniere's disease, but maybe you're going to ask about other associated symptoms or not at some point, but.

Dr. Jason Barnes:

Fill us in. What else would you expect?

Dr. Brian Neff:

The other things you want to ask about I think are very important because they help you lean one way or the other. Classically, Meniere's is the hearing loss and the tinnitus and the ear fullness. And the ear fullness really should be ear specific, it shouldn't be bilateral or head fullness. You also want to talk about or ask about, nausea, vomiting, but then neurological symptoms such as vestibular migraine. You want to ask about photophobia, osmophobia, headache. The description of the headache, whether it's focal, pulsatile. As far as vertebrobasilar insufficiency or stroke symptoms, I think those are very important, so you want to ask about confusion, disorientation, dysphasia, diplopia, dysphonia, they call them I think the three D's, any focal or sensory weakness. So there's a long list of things you should ask about as far as associated symptoms.

Dr. Jason Barnes:

Mm-hmm (affirmative). And with those associated symptoms, you started to get into the differential diagnosis. What else do you consider here when you have folks presenting in this manner?

Dr. Brian Neff:

The differential is huge and that's part of the problem. I think from an ENT standpoint, it's helpful. If you can come up with the list that you're constantly thinking about, you're not going to remember 250 things that it could possibly be, but if you can at least have a go-to list of the most common things in your differential in your mind, I think that's important. So I think in kind of an order of frequency and importance, the number one thing is differential for Meniere's disease is vestibular migraine. It has all of the same symptoms. There are some subtle differences and patients can have both at the same time. BPPV or benign paroxysmal positional vertigo, very different description of the vertigo, but it's in my list of things I'm thinking about. Benign recurrent vertigo is essentially vestibular migraine that doesn't meet the strict neurological criteria.

They could be missing timing, they could be missing headache, but yet have vertigo spells that are very similar to vestibular migraine. Persistent perceptual postural dizziness is really a common thing that we see. It tends to not have vertigo, so there's quite a few differences, but it's a large number of people that I see. Vestibular neuritis, health anxiety and panic, labyrinthitis, auto-immune disease. Can be just purely autoimmune inner ear disease or secondary to a known systemic disease or it can even be things like Cogan syndrome which has visual and ear complaints. Vertebrobasilar insufficiency, posterior fossa infarcts, acoustic neuroma or vestibular schwannoma, and to throw in to make it 15 even will be tertiary syphilis.

Dr. Jason Barnes:

Okay. And when we talk about Meniere's disease, can you tell us what the pathophysiology of Meniere's disease is? I know this might be hard to nail down, but what is Meniere's disease?

Dr. Brian Neff:

Meniere's disease basically has a histological finding on post-mortem studies. So endolymphatic hydrops is essentially overproduction of endolymph with stretching of the membranes of the endolymph compartment. So that is the sine qua non of Meniere's disease. However, everybody that has Meniere's disease usually has the finding of hydrops or endolymphatic hydrops, but there's a lot of people that have endolymphatic hydrops that may not have had Meniere's disease, so there's not a one-to-one

correlation and it's really poorly understood why endolymphatic hydrops occurs. There's theories out there and I guess the two theories that I think about that have a little bit of credibility could be a channelopathy. We just don't know is it a potassium-calcium channelopathy, but it has a lot of hallmarks of channelopathy diseases elsewhere in the body. And then the second would be repetitive vascular insult, whether it be from a migraine or some systemic vasculitis, those are the two things. But I will tell you that I am very open to any well done study that establishes this and will be willing to discard those rather quickly because the evidence behind them is poor.

Dr. Jason Barnes:

And are there any risk factors for Meniere's?

Dr. Brian Neff:

There's essentially epidemiological risk factors. I guess about 10% of patients have a family history, so there is a genetic component. Gene tests to date really have not isolated a single gene, so it'll be more a multifactorial genetic disposition with environmental causes like diabetes that I think eventually will be found. But the only clear comorbid condition that is very common, I think is a history of migraine headache and that's what makes distinguishing vestibular migraine so difficult.

Dr. Jason Barnes:

So you see someone in your clinic and they're describing the symptoms that we've talked about. What's your initial workup when you suspect Meniere's disease?

Dr. Brian Neff:

Essentially, if you're suspecting Meniere's disease, the only thing that I think is absolutely essential is an audiogram. It's part of the diagnostic criteria. Essentially, you want to see hearing loss in the symptomatic ear. Typically, it should be low-frequency hearing loss. I personally believe there should be an effect on the discrimination score, but that's not in the official criteria. If you go to the 1995 AAO criteria and there's some newer papers out too, they actually give you the criteria that you need to see for the hearing loss to be enough. But essentially those things come down to there should be a low frequency, sensorineural hearing loss in the affected ear. And I think that that is really all that's essential. Now we end up practically getting an MRI in most of these patients because they meet criteria for asymmetric hearing loss.

So we want to rule out of a vestibular schwannoma. We also usually get vestibular testing, which I think the important thing with that is, is that it does not establish your diagnosis. You shouldn't be like this isn't Meniere's disease or this is unlikely Meniere's disease and then you look at your vestibular testing and say, "Oh, now I think it's been Meniere's disease." It should be that you have a patient that you think has been Meniere's disease and then the vestibular testing is just added circumstantial evidence I guess. And so I get a VNG, a rotary chair, and [inaudible 00:10:18] testing.

Dr. Jason Barnes:

And going back to the audiogram, how often do you see this bilaterally?

Dr. Brian Neff:

The bilateral disease basically depends on when you develop your Meniere's disease. So, if you develop it at 60 or 70, I think your chance of getting bilateral disease is probably 10% or less. If you develop

Meniere's disease when you're 30, I think you're probably closer to 50%. So, the incidence goes up the longer you follow it.

Dr. Jason Barnes:

Sure. And you spoke about MRI and we obtain it not to diagnose Meniere's, but to make sure there isn't another disease process going on, such as a tumor, but what is found on MRI and folks with Meniere's disease and do you hang your hat on much of that?

Dr. Brian Neff:

MRI is typically normal. And so there's a couple things that I'm looking for. I didn't mention in the differential, but multiple sclerosis can cause dizziness, so you're looking for lesions that would be consistent with that. Intracranial hypertension can have dizziness symptoms with it, so you're looking for clues to that diagnosis such as empty sella. Chronic meningitis, various forms can have dizziness associated with it, so you're looking for thickening of the dura and other things, but usually almost all of the time it's normal. Now that being said, there's studies being done mostly in Asia that look at intertympanic gadolinium installation into the middle ear that will then with contrast sequences, show hydrops. These are usually pretty high-resolution MRI scans. They're not really clinically available however, for most physicians in the United States. So you're going to probably see a normal MRI.

Dr. Jason Barnes:

Any role for CT scan here?

Dr. Brian Neff:

I don't think so. Again, superior canal dehiscence was a dizziness differential that I didn't list, but it really is very different in its description and its symptoms. And so, I think the only time I would consider a CT scan and seeing a dizzy patient is if I'm getting symptoms that suggest that possibility, but Meniere's disease tends to be very different than that. So it's very rarely that I am getting a CT scan.

Dr. Jason Barnes:

Yeah. And finally, do you have any set of labs that you routinely get for these folks?

Dr. Brian Neff:

Again, it comes down to, I think the answer is no. And a short answer, a little bit expanding on that I think that if you suspect somebody that has autoimmune inner ear disease or has a systemic autoimmune disease, then you may consider getting some labs in those areas such as a vasculitis or an ANCA panel, I guess it's called a PR3 and MPO now. Tertiary syphilis is very rare in this part of the country, but I think depending on what kind of practice you have and where you're practicing, that should be considered and maybe be routine, but I don't routinely get that.

Dr. Jason Barnes:

So we've talked about presentation, differential diagnosis, pathophysiology, and workup. So how do you make the official diagnosis?

Dr. Brian Neff:

It essentially is a list of symptoms. So the criteria are definite Meniere's disease. Requires two or more episodes of what you've established to be spontaneous vertigo lasting more than 20 minutes and less than 24 hours. You need the documented audiometric change and at least one occasion you need tinnitus and ear fullness in the suspected ear. Probable Meniere's disease is just basically the exact same criteria, except you only need one episode of dizziness. There are possible and I don't find those very helpful. That being said, you have a lot of people that don't clearly fit those definitions. And I saw one study that said it sometimes takes a couple of years for people to actually meet the diagnostic criteria. And so it's a clinical criteria, doesn't the vestibular testing, MRI scan, it just requires symptoms and an audiogram.

Dr. Jason Barnes:

Mm-hmm (affirmative). And can you speak again to the vertigo? You said greater than 20 minutes and less than 24 hours, is that the typical range for what's known as a Meniere's attack for example?

Dr. Brian Neff:

It's usually hours. So that being said, there's a great variety or kind of distribution. I think basically when I am getting reports of vertigo that's lasting less than five minutes, I really don't think that is Meniere's-like. The other end of the scale, 24 hours is again, a bit arbitrary, I guess, but that came about as basically one of the subtle differences that you can see, don't have to see, but can see between Meniere's and vestibular migraine is that when you start getting people saying they're having vertigo for two to three days, that becomes much less likely to be Meniere's disease and you have to start thinking of vestibular migraine, vestibular neuritis, labyrinthitis.

Dr. Jason Barnes:

Yeah. So once you make the official diagnosis of Meniere's disease, I next want to talk about treatment. Can we start with medical therapy for folks you suspect of having Meniere's disease?

Dr. Brian Neff:

As far as long as I can remember in my practice at least, the starting point is usually a diuretic. We usually use potassium sparing diuretics, which is a mixture of hydrochlorothiazide and triamterene, trade name Dyazide. It's a once a day medication. There's a lot of controversy on how that works. There's been some pretty good studies showing that it actually does not change the serum osmolality or the perilymph osmolality and that it probably doesn't work classically by diuresing the inner ear essentially, but it's still is the first line therapy in North America. It's usually combined with a low salt diet, so it's very variable on what people ... how much they limit it. We use 1,500 milligrams a day, others use more or less.

I think in my mind it's very important to evenly distribute that through the day, so that you're not salt loading during one meal, but those are the two standard medical therapy approaches. Now there's a lot of lifestyle stuff that comes in that we don't have very good studies for or against, but some of the things that I think are frequently told to people is to avoid caffeine, smoking and nicotine, alcohol, stress, get good sleep, exercise regularly, I think those are all just general healthy things that I'm not against, but I don't have good data to support their efficacy.

Dr. Jason Barnes:

Mm-hmm (affirmative). And can you talk about betahistine?

Dr. Brian Neff:

Betahistine is a first-line therapy in Europe. It's an older anti-histamine, there's animal studies that show that the thought of how it works is to improve cochlear blood flow. And by doing that, maybe you diurese the inner ear. But there's been several reviews, including a Cochrane review saying that they haven't shown any benefit of betahistine. We can get it and use it here at Mayo. My typical prescription is eight milligrams, three times a day, so it's kind of hard for compliance and they also can't get it generally anywhere else, except maybe some mail out pharmacies or Canadian pharmacies, so.

Dr. Jason Barnes:

And how much do you talk with patients about supportive medications, maybe for nausea that kind of thing?

Dr. Brian Neff:

I think those are important because quite frankly, we're not going to do something specific. If somebody calls up, "Hey, I'm in the midst of a vertigo spell." They just have to ride it out and so the supportive medications I think help them with that. So in general, I use benzodiazepines, I have a lot of elderly patients, so you really have to be careful with that, use low doses. I like shorter half-life benzodiazepines like Ativan. You can also use a lot of anti-nausea medications. For whatever reasons, Zofran, seemingly anecdotally doesn't work very well for patients with nausea associated with vertigo, so I use things like Compazine, Phenergan.

Dr. Jason Barnes:

And moving on to some other therapies, before we get to surgical intervention, maybe the next thing that we can talk about is procedural, I guess. So can you tell us about transtympanic medication?

Dr. Brian Neff:

There's two medications that we use for Meniere's disease therapy, period. I look at it as kind of one is ablative and the other is non-ablative. So, the non-ablative therapy is a intertympanic steroid injection, it's controversial, the dose and the type of medication. I use dexamethasone at a concentration of 24 milligrams per CC and I usually inject until the middle ear is full, which is around 0.4, 0.5 CCs. That medication, the dose delivery, how often you do it, how many you do is really not very standardized. What I do is I usually give three injections and I try to space them one or at most two weeks apart and then I give a period of time of, let's say, six weeks afterwards to see if they've had an impact on their vertigo control. And that gets very complicated on patients that may have longer in between their spells or they have clusters that only come every six months. So that's just the general approach I use. The second medication is an ablative therapy, which is gentamicin. There's lots again ways to give that, some people try to give it daily until there's complete vestibular ablation or 0% function on ice water calorics.

We tend to not do that here. We do a more of a titrated approach to symptoms. So I do an injection and then usually give six weeks and do another injection if they're still having spontaneous vertigo. I think you have to be very careful though that when they come back for that first appointment, most of them are complaining of various dizziness spells that are not spontaneous vertigo. They're having head motion, dizziness, or they're having imbalance, or something from the gentamicin, so it's actually counterproductive to give more. But, the reason I do the way I do, it was six, eight weeks in between is that I think you can always give more, but you can't take injections back. And I think you increase your side effect profile if you give it to complete vestibular ablation, your hearing loss rate goes way up and the risk of kind of permanent worsen imbalance it goes up.

Dr. Jason Barnes:

And what are some surgical options for these patients?

Dr. Brian Neff:

There's really three options. The non-ablative least invasive option is an endolymphatic sac decompression or shunt. It's highly controversial, whether this works or not, but most otologists in North America are still employing this as an option. The reason I think it's attractive is that it has a low side effect profile or risk profile. It can be done in both ears in case of bilateral disease.

It does show about a 70% effectiveness at controlling vertigo, and it's trying to save the inner ear function, so preserve the hearing level at its current level and preserve whatever balance function they have in the ear. The other two options are ablative. A vestibular nerve section can be done in a lot of different ways. We typically use a retrosigmoid craniotomy and section the nerve within the cerebellopontine angle. I've only done a handful of those in my career because gentamicin is so effective, but it's for our patient that has continued spontaneous vertigo and yet has very serviceable or good hearing and is young and healthy enough or healthy enough to undergo a craniotomy. Lastly, there's a labyrinthectomy, which to me is really kind of the gold standard therapy for Meniere's disease it's highly effective. Its downside is, is that it sacrifices remaining hearing in the ear, so we tend to do that in patients that have poor or non-serviceable hearing with continued vertigo spells. The new thing I think that we're thinking about much more is it can be combined with cochlear implantation for single-sided deafness in patients with unilateral Meniere's disease.

Dr. Jason Barnes:

Mm-hmm (affirmative). And can you briefly go over how you teach these patients about expectations for different treatments and what they should expect moving forward with their disease treatment?

Dr. Brian Neff:

I typically tell them that this is unfortunately a very chronic disease kind of like high blood pressure. So we don't cure it, we try to control it. That's one of the things I typically say. I'm not surprised if they have relapses in the future and we can respond or treat to those as they happen. I see a lot of my patients I think that I saw at the beginning of my career, I'm still seeing, so it's kind of a lifelong relationship. I think that in general, effectiveness of the different therapies I discuss with them side effect wise, it depends on what you're talking about, but the ablative procedures specifically, I think need to be talked about quite a bit because they do risk hearing loss. Gentamicin, we see about a 20% hearing loss rate, the vestibular nerve section, very similar. And then I think anything that you're doing ablative, basically you're damaging the ear that's involved, you're decreasing the vestibular function on that side. You have to talk about the expected dizziness that comes with that and then the compensatory timeframe and sometimes vestibular therapy that's needed to kind of get back to baseline walking and imbalance status. I tell people there's about a 5% chance that it'll never get back to normal, hopefully minus the vertigo, but I talk about those two things I think mostly with ablative procedures.

Dr. Jason Barnes:

Well, thank you so much. This has been a really helpful discussion. Before I move into our summary, is there anything you'd like to add?

Dr. Brian Neff:

I think from a history standpoint, we talked a lot about what is vertigo-associated symptoms, but you really want to get an idea of how long the dizziness is. Is there any triggers such as foods, Valsalva, is it provoked by anything? And I think the key thing with Meniere's disease is there's the concept of spontaneous vertigo. Basically you're not doing anything and it hits you out of the blue. That's what you want to hear for Meniere's disease. If they're consistently being provoked with head movement or something else, that is not what you hear with Meniere's disease. And I think that's a very important concept, especially as we talked about when you're dealing with somebody that just had gentamicin, they're going to have dizziness. They'll feel that their eyes don't track right for a few weeks after the gentamicin injection, just with head movements. And that is not spontaneous vertigo, that's not continued Meniere's disease, that's a gentamicin side effects.

So I think those are a couple of the important things on the history. You want to know the frequency because a lot of these interventions you're basing success on reduction of frequency and severity and duration. So you're not going to get a lot of these patients perfect and so you have to have an idea where you're starting. And then lastly, I try to get an idea of ... you'll have somebody coming in and saying they have 50 spells of vertigo a month and there's severely affected by it, but then when you start asking them about, "Well, are you bedridden during this episode?" And they're like, "No," they're up and around. And it's just a way of trying to ... People will tell you various things about the severity and how much difficulty they're having, but I really like to hear that they're prostrate or in bed or can't function at all during the vertigo spells because well, sometimes when I'm not hearing that, I start to wonder about some of their descriptions, so.

Dr. Jason Barnes:

Yeah. Well thank you so much. I'll move into our summary now. Meniere's disease is a disease that presents with fluctuating, usually low-tone sensorineural hearing loss, and repetitive vertigo. And as Dr. Neff said, this vertigo is usually, or almost always spontaneous and pretty debilitating, lasting greater than 20 minutes and usually less than 24 hours. This can be accompanied by aural fullness and tinnitus. The differential diagnosis is quite long, including vestibular migraine towards the top of the list, BPPV, vestibular neuritis, autoimmune inner ear disease, and many others. The pathophysiology is a little bit tough to nail down, but the pathology includes hydrops though here isn't a great etiology, so to speak for exactly what's causing this. In terms of workup, the audiogram tends to be the most helpful thing showing low-tone hearing loss. Imaging is often obtained due to asymmetric hearing loss, but the MRI and a CT, if obtained, are usually normal. Lab workup can be performed to rule out any sort of autoimmune inner ear disease or auto-immune issue going on.

And it can also help maybe with more syncopal-type episodes. There is a diagnostic criteria that divides it into definite and probable. With definite Meniere's disease being two episodes of vertigo lasting greater than 20 minutes, an audiogram showing hearing loss and tinnitus or aural fullness. There are a lot of different treatments. non-ablative and ablative. Usually we start with a low salt diet and diuretics and also encourage healthy living. Transtympanic steroid treatments are also provided, but might not be of benefit from a surgical side or also from a transtympanic side. Gentamicin is offered as an ablative therapy and then from a surgical side, we have endolymphatic sac decompression, as well as vestibular nerve section and labyrinthectomy. Dr. Neff, is there anything you'd like to add?

Dr. Brian Neff:

I think that's an excellent summary. And that's it in a nutshell.

Dr. Jason Barnes:



Well, thank you so much. Appreciate you being here. It's now time to bring this episode to a close, but before I do, I wanted to finish with a few questions. As usual, I'll ask a question, wait a few seconds to give you time to pause or think about the answer and then give the answer. So the first question is what is the classic presentation of Meniere's disease and how do patients describe their vertigo? Meniere's is classically described as repetitive vertigo lasting more than 20 minutes and usually less than 24 hours. Vertigo can be described as the room spinning or a rocking, but there should be some sort of movement outside the body that is being perceived. This is accompanied by fluctuating, low-tone sensorineural hearing loss, and tinnitus.

The next question is what is the appropriate workup for a patient in whom you expect Meniere's disease? There are a lot of things that can be considered for workup in Meniere's disease. An audiogram tends to be the most helpful, however, an MRI is usually obtained due to asymmetric hearing loss, but this is not required for the diagnosis and is often times normal. A CT scan is also normal. Lab workup can include things like a CBC, ESR, and CRP to rule out inflammatory conditions or anemia or other causes of syncopal-type episodes.

And finally, describe the ablative and non ablative therapies that are offered for Meniere's disease. To start, the most common initial treatment is a diuretic with a low salt diet. Healthy living is also encouraged. From a non-ablative standpoint, you can offer transtympanic steroids. From a non-surgical standpoint and from a surgical standpoint, you can offer endolymphatic sac decompression. For ablative procedures, you can offer a transtympanic gentamicin injection which comes with a 20% risk of sensorineural hearing loss. You can offer a vestibular nerve section as well as labyrinthectomy, which of course would cause definitive hearing loss.

Thanks so much for joining us and we'll see you next time.