Neurotology for Audiologists

Presenter: Daniel Zeitler, MD, FACS

- [Christy] It's my pleasure to welcome back Dr. Daniel Zeitler to AudiologyOnline. Today, he'll be presenting "Neurotology for Audiologists: a Primer." If you don't know much about Dr. Zeitler, he's from the Pacific Northwest out of Virginia Mason Medical Center in Seattle, Washington. If you'd like to learn more about him, you can read his bio on the course registration page, but at this time, I hand the mic over to you, Dr. Zeitler.

- Thanks, Christy. Good morning to everybody, or afternoon, I guess to some, wherever you're tuning in. I appreciate the invitation to come back. I've been here before. It's always a pleasure. And I hope that this is a meaningful talk for everybody. These are my disclosures here. I do receive a small honorarium. I would share it with all of you if I could, and I'm happy to do that, but it's really my pleasure to be here regardless. These are the learning objectives, and I do have a full slate of information to pass along today, so we will not waste any time and jump right in. So let's first talk about vestibular schwannomas. If you think about neurotology, you think about vestibular schwannomas, this is why many of us do a two-year fellowship and probably is kind of the gleaming tower in the sky of neurotology.

These make up about 6 to 10% of all intracranial tumors. You can see here on this sort of faded picture, the cerebellopontine angle is this area right in here between the brainstem and the petrous temporal bone, so this area here, and you can see sort of the outline of a tumor there. People used to talk about these as being sort of one in 100,000. That was kind of the textbook number that we all learned in our training, but the number is clearly increasing. MRI technology is getting much better. People are accessing MRI machines in places, in rural areas that didn't typically have machines in the past. And also there are pretty clear guidelines for screening with MRIs for either asymmetric hearing loss or sudden hearing loss.

And so what we're seeing now is that incidence or the number of new cases per year, is how we define incidence, is probably slightly higher, probably around three to five per 100,000, and the median age is 60. But if you look at the age-specific incidence, because again, we know these tumors are benign and typically slow growing, that incidence gets even higher as you get older. And so in 70-year-olds, there's some new data out that suggest that that incidence is maybe as high as 20 per 100,000. There was a really nice study done many years ago where they looked at about 46,000 patients who had had an MRI. And if you looked at those patients, of those 46,000 with symptoms, 500 of those, or about 1%, had a tumor.

But if you looked at patients without symptoms and just found incidental tumors, that incidence was something significantly less. So symptomatology is clearly an indication to image, and we'll talk more about that. The lifetime prevalence, or the number of, so prevalence as opposed to incidence is the number of people living with the disease. The lifetime prevalence is about one in 500, again, highlighting the fact that these tumors are pretty slow growing, they behave benignly, and people can often live with them for much of their life. In fact, the the thing I typically tell my patients is these are tumors that people die with not of, and only a small percentage of these are related to genetic defects, such as neurofibromatosis two.

Just these numbers you see, besides some of the text here, are the PubMed IDs, if any of you're interested in finding any of these articles subsequently to this lecture. These typically reside on the vestibular division, or by definition, not typically, reside on the vestibular division of cranial nerve eight or the vestibular cochlear nerve. And there's been some research showing pretty equal division between the superior and inferior nerve origins. Originally, they were thought to arise at what was called the Obersteiner-Redlich zone, and this is where the central neuron-covering cells called glial cells interchange with the peripheral nervous system-covering cells called the Schwann cells. But Fred Linthicum, who's a wonderful otopathologist, who used to be

at the House Clinic, actually showed that the likelihood that these arise at something called Scarpa's ganglion is probably much higher.

And this is an area slightly lateral to the ORZ. And what's interesting about this is this is why many of these tumors actually arise somewhere in the mid to lateral portion of the IAC rather than in the CP angle. What's interesting is that there are some tumors that are only in the CP angle, and that throws a little bit of shade on this theory, but this has really become the predominant theory. We think about these tumors as what are called either intracanalicular or extracanalicular, intracanalicular meaning they're contained entirely in the internal auditory canal. And interestingly, these intracanalicular tumors have a growth rate of only about 1/3. So about 30 to 35% of these tumors will grow over 10 years. And even many of these tumors that do grow, you can still allow them to grow because if they start at two or three millimeters and they grow another seven or eight millimeters, you still have a tumor that's not even out of the internal auditory canal, which is about a centimeter.

So only about 15 to 20% of these tumors, depending on the data you read, actually have a failure of observation. Again, even despite some growth, some of these tumors are observed. Contradicting the intracanalicular is the extracanalicular tumors tend to have a much higher rate of growth, up to about 70%, depending on the study you read. And so these are numbers that you don't have to necessarily remember, but what's good to remember is that these extracanalicular tumors do tend to grow more. So we would tend to be a little more aggressive about some of these larger extracanalicular tumors. There is all kinds of data about growth rate of these tumors, but if you kind of parse it out, the growth rate of all tumors combined, somewhere between about half a millimeter to about two millimeters per year.

You can see that the overwhelming majority of those tumors that grow grow less than one millimeter per year. But these numbers include both growing and non-growing

tumors, so if you look at just the growing tumors, they tend to grow somewhere between one and two millimeters per year, but some of them can grow as fast as three to four millimeters per year. So again, you're not gonna be doing the surveillance and the counseling on these patients, but it's important to understand that even those tumors that are, quote, quickly growing are still not growing at a rate where they could become impending or dangerous within the course of a year or 18 months. So that's important to reassure our patients.

What's also interesting, this is a really nice graph that shows intrameatal tumors, or intracanalicular tumors on that solid line and extracanalicular, or extrameatal tumors on that top line. And you can see, again, highlighting, this is just one study, the growth rates are slightly lower than those I just quoted. But what's interesting here is I drew this red line, and why that's interesting is because the growth rate of these tumors tends to asymptote around five years. So typically, these tumors, if they're going to grow, will do so much more aggressively in the first one to two years. They tend to slow down in the first three to four years. And if the tumor hasn't demonstrated any growth after five years, the likelihood that it grows subsequently is much lower.

So this is why people, once they get to that five-year mark, can start to have their MRIs done much less frequently, maybe every three years, maybe every five years. They should never be completely forgotten because there are reports of these tumors not growing for 5 to 10 years and then growing subsequently. But certainly, you can reduce the rate of MRI appointments. I love this slide because this really highlights this sort of trend of these tumors as it relates to the MRI machine. So you can see this is a graph of about 30 years from a wonderful group in in Denmark, the Stangerup group. Denmark is absolutely fantastic. They have universal, or a version of universal healthcare, and they keep some of the most exquisite medical records in the world.

And so they can do these gigantic database polls. And so what they've done is they've taken every patient diagnosed with a tumor over about 30 years. And you can see here, at the beginning of the study, these tumors, when they were finding them, were about 2 1/2 to 3 centimeters big. And over time, as the MRI machines got better and better, those screening protocols got more and more entrenched in practice, you can see the average size of the tumor was was about 1/3, about one centimeter. And so as you would expect, as the tumors get smaller, what else would happen? Well, what else happens is that fewer and fewer of these tumors require an operation. So again, the paradigm has shifted dramatically.

In the past, we used to identify these tumors and operate on every single one of them, whereas now, we can observe many of them. So you can see, again, here in 1975, almost none of the tumors that were diagnosed were allocated to what we call a wait-and-scan or observational protocol, whereas now in 2003, and this line has continued out in the same direction, this group in Denmark, and frankly, all of us all over the world are observing almost every single patient that walks through the door with a tumor. We typically, in our program, we recommend an MRI in six months from diagnosis, from first time diagnosis, and then go from there based on the growth rate.

So this has been a dramatic change in the way that we think about these tumors and a dramatic change in the number of patients who are probably operated on unnecessarily, to be perfectly frank. More apropos to the group here, how do these typically present? Well, the most common symptoms, as you would understand, would be related to that cochlear vestibular nerve, hearing loss, tinnitus, vestibular complaints. The problem is none of these symptoms are specific to a tumor, as we all know. So there has to be a little bit of an index of suspicion as well. You can see here the rates. I highlighted this one here, cranial nerve seven or the facial nerve. It is exceedingly unusual, in fact, almost reportable for vestibular schwannomas to cause facial nerve symptoms, even those tumors that are 3 1/2, 4 centimeters big, which are

gigantic tumors, very, very rarely involve symptomatic signs to the facial nerves, such as paralysis or spasms.

So when we see this, and I highlighted this because these data came from a study that I read, I think it is much, much lower than that. So if you have a patient with facial nerve symptoms and a tumor, I think the suspicion has to be very high that it's actually a facial nerve tumor or something more aggressive, like malignancy, because it's just so rare. I think we're all familiar with what we call a retrocochlear pattern on audiology, which is to say that the word recognition score is far worse than the pure-tone average would suggest. So you know, these patients may come in with a pure-tone average of 45 or 50 and a word recognition score of 30 or 40, and that is highly suggestive, although certainly not predictive, of retrocochlear pathology or a tumor.

Other interesting facts are that about 1% of these patients will present with a sudden sensorineural hearing loss, and then you scan them, as is the recommendation, and you'll find a vestibular schwannoma. However, conversely, patients who have a known vestibular schwannoma, 10% of them will have a sudden sensorineural hearing loss. So from an audiological side, we always counsel our patients that once they know that they have a tumor, it is absolutely vital that if they have any sudden changes in their hearing, they present immediately for an audiogram, because interestingly, even though they have a tumor, the treatment for sudden hearing loss is identical, and many of these patients can actually get much worse or all of their hearing back with the same treatment protocol that we would use for a patient with idiopathic sudden hearing loss.

So counseling is very important to these patients on your end about presenting immediately for any changes. These cisternal symptoms, or what we would call kind of brainstem compression symptoms, only occur in gigantic tumors. These are what people used to present with in the 17 and 1800s when we had no way to image, and these tumors got absolutely gigantic. So these are extremely rare. We see them still,

and I'll show you a couple of scans to demonstrate that, but they're very, very rare. I probably am preaching to the choir here. In fact, probably most of you can can teach me more about this than I can teach you. But just to refresh your memories, what do we see in retrocochlear pathology?

We talked about rollover, or sorry, we talked about retrocochlear pattern. Rollover is when the word recognition score worsens with increasing presentation levels. This can suggest retrocochlear pathology. I'd encourage all of you, if you see a patient with a retrocochlear pattern to do a rollover test. Again, not perfectly predictive, but certainly can be suggestive. We also look at reflex decay, which is where the acoustic reflex thresholds tire out, basically, due to prolonged stimulation, and that's because the auditory nerve is irritated or damaged by the tumor. Again, neither of these tests are perfectly sensitive or specific. They're incredibly useful, and your team will much appreciate it, as will the patient because it'll give them some information that can explain sometimes why when they turn the volume up on their TV, they do worse and not better.

Auditory brainstem response testing is also used. This is really because, again, of the sensitivity of MRI machines falling a bit out of favor. It is certainly good for patients who can't have an MRI, say they have a pacemaker or extreme claustrophobia, or maybe for some of the elderly patients who probably wouldn't want surgery anyway or are too infirm to have surgery, but it can at least help us determine whether there's retrocochlear pathology going on. The pattern most specific for schwannoma would be a wave one that's present with an absent wave three and five and a significant interaural delay. Again, much like standard audiometry, the false positive rates or specificity is not great. The sensitivity's not great either.

So these, again, are tests that are good, but certainly not diagnostic. The one thing that we do know about ABRs is they're much better for larger tumors. So tumors that are

about 1 or 1 1/2 centimeters are going to have a much more sensitive and specific utility for ABR than a small, say, two or three millimeter tumor, in which case it's probably relatively useless. How do these patients do with regards to their hearing? So I just mentioned that many more of these patients we're watching and observing over time, and so the question would inevitably arise from a patient or from you, if I'm gonna watch this tumor, what can I expect with their hearing? And this was a really nice study, again, done by the group in Denmark where they looked at over 1,100 tumors who were allocated to this weight-and-scan approach or the observation approach.

And you can see that a large percentage of them, almost 90% of them had follow-up up to a year, and about 11% of them had follow-up up to 10 years. And what do we see? Well, if you're familiar with the classification, the AAO classification for hearing, we use A, B, C, and D. And so at the beginning of the study, 178 of these patients had class A hearing, which would mean greater than 70% word recognition score and less than 50%, sorry, and better than 50 dB pure-tone average. And if you follow these patients over time, at the end of the study, almost 50% of them had retained class A hearing, and of those, 54% had actually retained it at 10 years, and only about 13% of them went all the way from perfect hearing to non-serviceable hearing.

And so again, very, very useful data. And I think I misspoke, the pure-tone average is less than 30. I think I said 50, so I apologize. But again, you know, you can tell patients, "Hey, if you come in with good hearing, there is a better-than-50% chance you will stay with good hearing." And that is very, very reassuring to patients who have chosen to not operate on their tumors because in some ways, operating on a tumor with hearing preservation techniques gives us about a 50% chance of hearing preservation as well. So you can get the same ROI without a big hole in your head. Same thing is true with word recognition score. If you take out those patients who started the study with 100% word recognition score, so they're about 160 of those patients, and you look at the end

of the study, again, about 50% of those retained that perfect word recognition score, and almost 90% retained a word recognition score better than 70%.

So again, highly, highly useful data. And this is just one study. I don't wanna imply that this is the sort of written in stone. Some studies will show much worse hearing preservation rates. Some will show some better, but it goes to show that you can do very, very well with your hearing without taking out the tumor. I also just love this graph because what these folks did in a different study, again, out of Denmark, is they looked at the rate of decline of word recognition score based on how you presented. So if you come in with a word recognition score of 100%, you have a mean rate of loss of about 2% per year. So if you amortize that over 20 years, you can see why these patients maintain pretty good hearing.

However, if you come with hearing even just a little bit lower, so still within that class A hearing, but now you've gone from 100% to somewhere between 70 and 98%, you can see that your word recognition score declines around four times faster, three times faster. So at a rate of about 6% per year. So again, we don't know why this is the case, but probably any type of damage to the hearing nerve will continue to sort of progress, whereas if the nerve is relatively robust, it can oftentimes stay robust. And interestingly, there's no real correlation between the tumor size and some of these hearing data, although we do know that intracanalicular tumors will have better hearing preservation with observation than large extracanalicular tumors.

We, at our center, do not use vestibular testing routinely for these patients. Some places do, there's nothing wrong with it, we just don't find that it gives us any significant improvement in counseling, and it doesn't really change our plan, but we will use it if patients have significant vestibular complaints or if we're trying to decide how best to approach the patient. We can use VNG, which measures, as you know, the superior vestibular nerve through calorics. Very, very poor specificity. It's not so good

at ruling out the tumor. The sensitivity is okay, 80%, so okay at ruling in a tumor. But again, lots of people can have abnormalities on caloric testing, and lots of people with tumors can have normal caloric testing.

So it's not a perfect test. On physical exam, you'll often see nystagmus, and remember we define nystagmus as the fast phase. So the fast phase will beat away from the lesion. This is a classic pattern of vestibulopathy. And VEMP can also be used, which, as you know, measures the inferior vestibular nerve function through the saccular response. And again, sensitivity and specificity, really no better, certainly no worse but no better. So again, these tests are helpful, but probably not diagnostic and should not be used in that manner. Again, some centers really highly value these, so there's nothing wrong with that. This is a CAT scan, and we typically don't use CAT scans unless we have to, again, a pacemaker or someone who can't have an MRI for whatever reason.

But just to show you, here is the internal auditory canal on the right, you can see it's got a nice sort of pyramid shape. Here on the left, this internal auditory canal is much wider. That is what we call a blown out IAC or a dilated IAC. And that typically implies that there's something filling that space. And it goes to show you how benign these tumors are. They grow so slowly that they just kind of expand the bone. They just remodel the bone rather than breaking it down. So when we see this sort of chronic enlargement, that is very reassuring. If we were to look in and see a lot of damage to this bone here, erosion of the bone, we'd be thinking something much more insidious, something much more malignant or aggressive-behaving.

Here are some patients who I have seen over the years, just to show you how these kind of come in in different flavors. Here is a left vestibular schwannoma, probably measures, I don't know, around 1 1/2, maybe two centimeters, one slightly bigger, or probably around 2 1/2 or so centimeters. Here is what we'd call a purely

intracanalicular tumor, so this one measures about one centimeter. You can see here, this is a absolutely gigantic tumor. This one was a two-day operation that we did actually not too long ago. Gigantic vestibular schwannoma. And then here is sort of a classic presentation of a patient with neurofibromatosis two. So there are lots of definitions of neurofibromatosis two, but one of the definitions, and all you have to have to meet the definition are bilateral vestibular schwannomas.

So there are other ways to diagnose NF two, but bilateral vestibular schwannomas is an absolute diagnosis of neurofibromatosis two. These are incredibly frustrating patients. They are really sick. They are incredibly debilitated by their hearing loss. They often have visual loss, they often have spine lesions. Really, really tough patients. And they're often, as you can see, the question is, well, which one of these tumors do we take out first? Which one do we take out second? Should we take out the whole tumor? Should we leave some behind? Do we put in an auditory brainstem implant? Do we put in a cochlear implant? So these really stress the seams of our multidisciplinary teams. I'm not gonna spend much time on surgery at all with this group.

Obviously, I don't expect any of you to know how to do surgery nor need to know. It's taken me 15 years, and I'm still learning how to do these surgeries. So this slide is really just to show that there are a number of approaches, the suboccipital or the retrosigmoid, which kind of goes behind the IAC, which would be here. There's the translab, which is kind of the workhorse, which goes through the mastoid bone. There's also the middle fossa approach where we open up the skull above the internal auditory canal and approach it from the top side. Again, there are lots of good reasons why one would want to do or not want to do one of these tumors.

Since you have these slides at your convenience, I'll let you read through these. I highlighted kind of some of the basic pros and cons of each of these. But suffice it to

say there is not one of these approaches that is better or worse. It really comes down to whether or not you're trying to save hearing. It comes down to the experience of the surgeon and the comfort level of the surgeon and what he or she likes to use. And then sometimes it comes down to whether there's preserved hearing to try to save, whether the tumor is big or small. So again, not to get into the weeds, just wanted to put this here to close the loop.

So you just learned everything there is to know about vestibular schwannomas. It took me many, many years, and you've done it in about 15 minutes, so I can send you your certificates. Next up is Meniere's disease. This is something probably much more relevant and apropos to the audiology side of things. We know that Meniere's disease is idiopathic. It is multifactorial. And we, because of this, really don't understand the pathophysiology. People have proposed that it is due to an accumulation of endolymph in the scala media and sort of backing up into the vestibular organs with what we call endolymphatic hydrops. The problem with that theory is that you can see a lot of other diseases that, on cadaveric dissection, also manifest endolymphatic hydrops.

Syphilis is a great example. People who used to have syphilitic ear disease on dissection of their temporal bones after death would be shown to have endolymphatic hydrops. So the big question that has kind of come up in the last 10 years or so is whether the endolymphatic hydrops is the cause of the disease or the result of the disease. And if it's the cause of the disease, the treatment that we'll talk about kind of makes sense. If it's the result of the disease, the treatment maybe doesn't make so much sense because we're not treating hydrops, we're trying to treat the result of the disease. We know that the clinical syndrome is episodic vertigo, fluctuating hearing loss, and aural fullness with tinnitus.

We'll talk a little bit more specifically about how to make the diagnosis here in a second. But because there's so much variability, phenotyping of the disease is difficult.

And I think that's one of the reasons why, as we'll see, they've really tried to specify and simplify the diagnostic criteria. The true incidence is unknown for all of the reasons that I've sort of highlighted here, but certainly, we've estimated it to be about 46 to 50 out of 100, 46 to 50, sorry, 46 to 100 out of 100,000 per year. And the prevalence in our country is around just over 600,000, again, estimated by the NIDCD. The onset peaks between 40 and 60 years. It is incredibly rare in children, so much so that if you suspect it, you're probably wrong.

It could be, but it's highly unusual. Much more common in children is migraine if they're dizzy, so that that's something to think about. And about 10 to 12%, and maybe some is even as high as 20%, depending on the study you read, will develop, over time, bilateral disease. So this is the old criteria which was put out by our academy in 1995. And I'm not gonna spend time on this, but just, I think that what I wanna highlight here is that in definite Meniere's disease, we'll forget about certain because that requires an autopsy, but in definite Meniere's disease, you can see that the two sort of highlighting features are the number of episodes of vertigo, the time of those episodes, and hearing loss, okay?

And if you don't have two but you have one, it's probable. If you don't have any or you have episodic vertigo with no time or maybe some hearing loss, then it's possible. But these became incredibly rare, and as you all probably have seen, patients come in and say, "I was diagnosed with Meniere's disease 30 years ago," and they have perfect hearing. And we see that all the time. So a group got together through the International Classification of Vestibular Disorders not too long ago, and they really simplified this to two options. You either have Meniere's, or you probably have Meniere's And if you don't meet either of those things, then you don't have Meniere's. Specifically in red, I've highlighted the big changes.

It is still two or more episodes of vertigo, but there's a time limit, so 20 minutes. So anything less than 20 minutes is probably not Meniere's, and anything greater than 12 hours is probably not Meniere's. That's when you start to get into the vestibular migraine territory. The other thing that they have much more specifically stated now is you have to have low or mid-frequency sensorineural hearing loss before, during, or after an episode, and you can see that they've specified the frequencies and the decibels. So you have to have two contiguous frequencies less than 2,000 hertz, again, those low and mid frequencies, and it has to be at least three decibels. Should you be rigid about this? No, but it just goes to show you that you really need to document hearing loss.

And then the main difference between probable is this 24 hours. So again, once you get beyond 12 hours, Meniere's disease episodes typically don't last that long. So if you get out into the 18, 20-a-day of vertigo, it's much less likely Meniere's. It still could be, probably, but it's less likely. And again, these are very, very good to keep in mind. Vestibular testing is used. We see elevated summating potential because of the displacement of the basilar membrane, again, due to that swelling of the endolymphatic space. Click is not as reliable as tone burst, but even tone burst is not that great, and vestibular testing, electrocochleography, or ECochG, is really not good in early stage disease. It's much better in later stage disease or those patients with hearing loss.

Again, much like vestibular testing for schwannomas, we use this in our center, but it is certainly not diagnostic, it's more confirmatory. This would be the kind of test you might wanna do if you think you have Meniere's but aren't sure, or you know you have Meniere's and you want to track it over time. VEMP can also be used, and that's due to the saccular dysfunction, again, due to the endolymphatic hydrops. We see absent or low-amplitude responses. Again, sensitivity, specificity pretty bad, and again, more sort of as an adjunctive test rather than a diagnostic test. I think the treatment is really

interesting. So again, harking back to what I was talking about before, you know, what is the pathophysiology here?

And because people used to think that it was this swelling inside the inner ear, the historical benchmark for treatment has been sodium restriction. And interestingly, if you look through the data, there is absolutely no data to support sodium restriction in the treatment of Meniere's. It just doesn't work. Similarly, diuretics, for the same reason, people thought, well, hey, if you can pee off the salt, then maybe you can reduce the swelling. They showed in a handful of randomized controlled trials that there was some benefit of diuretics. But 79%, if you look at the data really closely, about 70 to 72% of patients with Meniere's disease get better with no treatment. So the question is, is that real, or is that just the natural history of the disease?

Much, much more commonly in my practice now, I'm using betahistine or Serc. The data, or this is kind of what Europeans have been using in lieu of diuretics for a number of years. The data in the randomized controlled trials is not so good again, but if you look at meta-analyses where they take all of the randomized controlled trials and put them together, you do get an odds ratio that's fairly favorable to Serc, about 3 to 3 1/2 times benefit for vertigo improvement. And anecdotally, I can tell you I haven't done research on my patients, but they do incredibly well with Serc, much better than anything that I've used with sodium restriction or diuretic, so this has become my go-to.

It's a compounded medicine, so you have to find a pharmacy that'll make it for you, but it's worked wonderful, and there's no side effects, which is nice for those patients that have high blood pressure or other issues that preclude them from taking the diuretics. Outside of these, we use steroids. Typically, those steroids can be given orally, but probably much more effective is to give them intratympanically. We don't really know why they work, probably some sort of immune modulation or inflammatory reduction,

but they're nice because they're non-ototoxic, and they don't ablate the vestibular system. And they do work better than placebo, and that's been demonstrated over and over again. On the other hand, gentamicin, which is vestibulotoxic, is ablative.

So you ablate those dark cells in the vestibule, sorry, in the semi-circular canals and the utricle, but this has incredibly good control of vestibular symptoms. But again, it is ototoxic, and there's a chance of hearing loss. If you compare II gentamicin versus IT steroids, right, the question is, well, why would I give something that's ototoxic if I have something that's not ototoxic? And historically, the gentamicin has won the day. So you can see, in one study, 92 versus 61, in another study, 83 versus 48. So historically, if you wanted to get rid of symptoms, you gave gentamicin, but there was a landmark paper in 2016 in "The Lancet." It was done really beautifully. The methodology was wonderful, and these guys showed, their group showed that intratympanic steroids had the same efficacy at vertigo reduction at two years as intratympanic gentamicin.

So many of us now go to steroids first, and we try to keep on steroids until they fail. There are surgeries, you can do what's called an endolymphatic sac surgery. The nice thing in that this is non-ablative, so it doesn't ruin the vestibular system. It is also hearing preserving, but the randomized controlled trials suggest pretty limited efficacy. In fact, there have been some sham studies in the past in the 1970s where they actually went in and did a sham surgery and showed that the patients did no worse and no better than the patients who actually had a endolymphatic sac surgery. That study would be pretty tough to pass through IRB these days, but I think the results are telling.

You can also cut the vestibular nerves. So you can go in, and you can literally cut the vestibular nerve. So you take away the efferent information to the vestibule. You can do this, again, much like tumor surgery, through any of a number of different approaches. Again, comfort level probably is more important than how or why you do which one.

This is just a cartoon of the endolymphatic sac here. So here's the labyrinth, here's the mastoid cavity, here's the facial nerve, here's the external auditory canal. So that endolymphatic sac is a kind of a fold of redundant dura along the posterior fossa. You open that dura. Some people put a little shunt in, some people don't. Some people cauterize it, some people don't.

Again, I think the fact that no one really knows what the right way to do this is kind of telling that no one really knows whether this surgery works or not. But it is something that we do, I would say, twice or three times a year in my practice. And these are just the different approaches. Here's a nice intraoperative view of a retrosigmoid approach to the vestibular nerve. Here's that vestibular nerve. The thing about the vestibular nerve section is you have to be incredibly careful because just under here is the facial nerve, and just under here is the cochlear nerve. So you certainly don't want to cut either of those because that will make your patients not dizzy, but also fairly deaf or paralyzed.

You can see the results of neurectomy are high. They're in the 90s. The problem, again, is that this is an intracranial surgery. You can risk hearing loss, you can risk facial nerve injury. And now with gentamicin, to be perfectly frank, very few of us are doing this operation because the gentamicin has about an 85 to 90% vertigo control rate as well without cutting the a hole into the head. Labyrinthectomy is the gold standard. This is how you treat patients who are at end stage. They cannot be made better with medication. They cannot be made better with intratympanic therapy. And this is what we do in patients who are either so miserable that they are willing to be deaf in that ear or patients who have already become deaf in that ear from their disease, because labyrinthectomy, as you know, is a hearing destructive surgery.

But again, very high vertigo control rates. I'm actually surprised these aren't higher. This is from an old study, as you can see. I think the vertigo control rates with

labyrinthectomy are probably in the high 90s, certainly in my practice, they are. Moving on to autoimmune inner ear disease, kind of a more elusive disease that I think is important to talk about from an audiological perspective. This is called autoimmune inner ear disease or idiopathic progressive bilateral sensorineural hearing loss, which kind of tells you exactly what it is. It's a syndrome of subacute progressive hearing loss that can occur over anywhere between a few weeks and a few months, so a little bit slower than sudden hearing loss.

The definition of this is bilateral. It doesn't have to be. I have patients who absolutely have autoimmune ear disease that don't have bilateral hearing loss, but it is classically bilateral. The suspicion is certainly raised in those patients with other autoimmune ear diseases. So if I see a patient that I'm concerned about autoimmune disease, I always ask about some of these other things, but it's very rare. So, you know, we think in medicine, as you all know, common things are common, right? Horses are more common than zebras. This needs to be thought about and considered. But in all likelihood, whether it is or isn't could ultimately just come down to statistics, right? This is a rare disease, and so if there are two things that are potential, it's much less likely, but you certainly don't wanna not diagnose this because of its rarity.

The incidence is also a little bit controversial because some people think that maybe bilateral progressive hearing loss could just be bilateral Meniere's disease if it's present with dizziness. So again, kind of a dark horse in the race of otologic pathology. There are lots of different theories about why the body would want to attack itself. These kind of come down to the same theories as general autoimmune diseases. Perhaps there's damage to the inner ear that causes an immune response. Perhaps there's something in the inner ear that looks foreign, even though it is not. Perhaps the inner ear releases toxic antigens that cause an immune response. We just don't know. The genetic theories have really also not shed much light on this as well.

We talked about the diagnosis. It's a subacute, typically bilateral hearing loss. There is no pattern to the hearing loss. So unlike Meniere's disease, these do not have to be low-frequency, and steroid responsiveness is the key to diagnosis. So if you have a patient with sudden hearing loss or bilateral subacute, sorry, subacute hearing loss in one or both ears, the test is to give steroids. So what you see, typically, you give steroids, they get better, they stop the steroids, they get worse, you give the steroids again, they get better. And once you start to demonstrate this pattern of steroid responsiveness, that is really the key to diagnosis because other than some of these blood tests, which are there, but don't really tell us much that's specific or sensitive for this disease, there is no other specific test to make this diagnosis.

So it is really a clinical diagnosis. This anti-HP70, or heat shock protein, has classically been used, but the problem is you can see it in lots of other things, including Meniere's disease, again, highlighting the sort of the confusion between the two. So really, it can be ordered, but it shouldn't be relied upon. We talked about sort of the treatment you give, steroids. Once you've demonstrated that responsiveness, you put these patients on a long course of steroids, typically four to six weeks, and probably 2/3 of them will respond. And once you've demonstrated that they respond, you transition them, through the help of rheumatology, to some sort of non-steroid-based treatment. Methotrexate has been the classic agent, but the data is pretty bad.

There are other sort of newer anti-rheumatologic medicines, like Enbrel or Remicade. I feel like every day I turn on the TV to watch a sporting event, I see another new Skyrizi ad. But these also have issues. They are expensive, and there may be some risk of lymphoma on these medications as well. I don't think we'll spend too much time on sudden hearing loss. I think we all are familiar with this. In fact, I think I've even talked about this in past lectures. I just wanna point out here that we cannot be rigid about the diagnosis. So we know that the rule of threes for sudden sensor nerve hearing loss,

30 dBs, three contiguous frequencies, three days, but none of us would not treat a patient who comes in with 20 decibel loss over two contiguous frequencies.

None of us would not treat a patient with, you know, 15 decibels over four contiguous frequencies. So this is the definition, but should not be used to preclude patients who clearly need treatment from getting treatment. You can see the incidence there. Those incidences are estimated from the UK, but they've extrapolated that to the US, so about 60,000 cases of sudden sensorineural hearing loss annually. And in an otology practice, there's been a nice study where, in an otology practice, which is kind of the tertiary referral center, they're gonna see about 1 1/2 to 2 patients out of every hundred per practice with sudden hearing loss. So very, very common in a tertiary care centers, which I'm sure many of you are in.

There are some non-idiopathic causes of sudden sensorineural hearing loss. And these need to be ruled out. Obviously, we know about Meniere's and autoimmune, which we've talked about, a list of just a few of the probably 80 or 100 different causes. These patients often present with tinnitus and dizziness. What's interesting is they'll often say that they have an ear popping, and they'll actually notice that popping first thing in the morning. If you really ask these patients, they'll say, "Yeah, my ear kind of felt like it popped," or they'll tell you their hearing loss was noted when they first woke up. And this kind of leads people to believe that maybe this is a vascular phenomenon, that there's been some kind of ischemic event or insult in the inner ear, which is very common during the early morning hours.

If you ask stroke patients, many of them will tell you they had their stroke first thing in the morning or early in the morning. Many of these patients get better without treatment, and this is probably why that 60,000 per year is highly underestimating the number who actually have this disease because some people will get it, they'll get better by themselves, and they won't ever come to the doctor. And about a 50% to

maybe even 2/3 will regain hearing with or without therapy, but almost as many maybe without therapy. So it's hard to know the true incidence. These people typically get better within two weeks, and that's why we typically say that the treatment protocol really should be initiated within that first two weeks because outside of that first two weeks, probably not a ton of benefit from medication, other than some of those things we'll talk about in a second.

There are some clinical practice guidelines. I'm not gonna harp on this, but I do tell my patients, you're in the wrong office because you may not like what I say, but I wrote these guidelines, and so you have to trust me. And most of them do. It was a fun thing, I don't know how often I wanna do it, but it was really fun to kind of pour through the data and sit around the table with all these people and try to make some recommendations. These guidelines do come with a number of recommendations and a number of recommendations against. I'm not gonna go through this because they are in the paper, but I think for this group, certainly, we want to exclude conductive hearing loss, and we also want to add audiometric confirmation.

I was a little bit surprised that audiometric confirmation wasn't a strong recommendation, but we have to use data to make these recommendations. And interestingly, there's not enough data to suggest that you need to get an audiogram. It seems crazy, but we just didn't write the papers. We do recommend an MRI for any patient who has a sudden hearing loss. Again, you can see here a small vestibular schwannoma in the left inner ear. And what's interesting is we still recommend an MRI for patients who got better, whose hearing got better. And that's because these patients can get better, despite the fact that they have a tumor, and they're treated in the same protocol as typical sudden hearing loss patients.

We give steroids as initial treatment, which is historical. It comes from a really, really bad study from 1980, but almost 100% of ENT doctors in United States gives steroids.

But interestingly, the literature doesn't support it. I kind of looked through the literature a few years ago, and I found three meta-analyses, and you can see here the results of those three meta-analyses, looking at steroids versus no steroids. So unclear benefit, benefit but not significant, unclear benefit. So interestingly, this has been the benchmark, but it's only an option because the data does not support the use of oral steroids for sudden hearing loss. Now, where the data does support treatment is for the use of intratympanic steroids. This is for initial treatment.

This is also an option. And interestingly, what's nice about this study that you can see here is they showed that oral steroids, sorry, intratympanic steroids were not inferior to oral steroids. So for those patients who maybe can't get steroids for mental illness or brittle diabetes, we can give them intratympanic steroids as initial treatment and not lose any benefit. Conversely, if you have been treated and you did not get better, or you're outside of that two-week window, we do recommend, and this is a recommendation based on pretty good data, that you give IT steroids as salvage treatment. There are a number of randomized controlled studies in the literature, all of which showed benefit to IT steroids after systemic therapy failure, and the odds ratio is a six times, so six times likely that you'll get benefit with IT steroids versus placebo.

The problem is the success of that audiometric success wasn't defined. So just because 10 decibels may be a statistically significant difference doesn't mean it's necessarily clinically different. I'm gonna skip through this a little bit quickly. I think I'm getting short on time. Hyperbaric oxygen is also an option. It has, interestingly, the same level of data as oral steroids. So you can see kind of how this becomes a little bit confusing. We do have a hyperbaric chamber here at our center. This is actually the chamber in our center, and this is the Christmas card they send out every year of all the docs and staff in the chamber. So we do use it, but the data is not super-duper good.

And this just goes through our own data where we showed comparing two groups, both on pure-tone averages and on speech recognition that the group who got hyperbaric oxygen did not do any better than the group that didn't, and both groups improved both word recognition score and pure-tone average following hyperbaric therapy. The prognosis, as we talked about, is fairly guarded. It probably gets better in around 2/3 of people with or without treatment. Advanced age certainly is a poor prognosis. The severity of loss, so severe to profound loss has a much worse prognosis than mild loss, the duration of the hearing loss, whether there was vertigo at the time, and interestingly, the shape of the audiogram also has implications on recovery.

So you can see here, those patients with low-frequency hearing loss have much better rates of recovery than those with flat or downsloping. And those, again, patients with profound flat loss have very poor prognosis. All right, I have about 10 minutes left, so I think I can get through most of this. Superior semicircular canal dehiscence. So this is a relatively new diagnosis. It was written about first in 1998 at Johns Hopkins by some folks who've identified a group of patients with chronic disequilibrium. They had sound-induced vertigo, and they had nystagmus in the plane in the superior semicircular canal. And on audiometric testing, they found, interestingly, these patients had bone conduction hyperacusis, and many of them had pulsatile tinnitus.

They went on to show that the CT showed missing bone over the canal. And interestingly, they don't really know whether this is a congenital or acquired disease. They favor congenital because if you look at a temporal bone study, here, you can see that about 0.5% of all the patients that they looked at on that temporal bone study had dehiscence, regardless of their symptoms. And about another 1 1/2% of them had what we call near dehiscence, or very thin bone. Again, in the interest of time, I'm gonna go through this quickly. We all know how a normal hearing ear works. Why do you get loss of air conduction in superior canal dehiscence? Well, you can see if

there's an exit tract of sound, then what happens is that fluid volume becomes shunted out of that.

And then what it causes is an unequal distribution of inward and outward movement of the stapes and round window at the oval and round window along that cochlear lumen. And then what that does is it causes loss of efficiency in the system, and therefore your air conduction gets worse. Conversely, we know that bone conduction can often be hypernormal or above normal in the negative range, and why is that? Well, similarly you have sound that goes in, you have that fluid that gets shunted out, and what that causes is a suprathreshold movement of that fluid across the cochlear partition. And so what that does is it causes an increased response to the cochlea by this compressional wave and almost shakes the bone.

So the bone is actually hyper-attenuated to conductive capabilities. The dehiscence syndrome is bone conduction hyperacusis autophony, or the ability to hear one's sounds in their own head, and pulsatile tinnitus. You often also hear these patients talk about sound or pressure-induced vertigo. You might hear these terms as Tullio and Hennebert phenomenon. Many of them will have conductive hearing loss. And interestingly, there's been some really neat studies looking at the size of the dehiscence, and what they've shown is that the size can be predictive. So in big dehiscences, greater than 2 1/2 millimeters, you may get vestibular and cochlear symptoms, whereas in a small dehiscence, you might get either vestibular or cochlear symptoms. But what's interesting is there's no association between the size of the dehiscence and the hearing loss.

Some people have thought that the size may be correlated to the air-bone gap, but that's not a reproducible result. You can make this exam oftentimes in the audiometric booth or in the clinic. You can ask the patient to valsalva or to hyperventilate, and you'll often see down-beating nystagmus, which is diagnostic. We already talked about

audiological findings, which would include loss of air conduction and suprathreshold bone conduction with an air-bone gap. And I think the important thing here is acoustic reflex thresholds are normal. This is absolutely vital to remember. We have this on our protocol in our audiological clinic here at our center. Any patient who comes in with an outside audiogram that has a conductive hearing loss gets reflexes because they're almost never done.

Again, you have to have a normal working tympanic membrane and no perforations. Those things are obviously not relevant, but if you have a type A tympanogram, oftentimes, these reflexes are not done. And this could be the difference between operating on a patient for presumed otosclerosis and getting in and finding nothing. So have to do acoustic reflex thresholds in any patient with normal tympanogram and air-bone gap. VEMP is the test that makes this diagnosis physiologically. You can use either cVEMP or oVEMP. The oVEMP, if you have those capabilities, has been shown to be much more highly sensitive and specific. And so this is our go-to, but we do both. The ECochG has also been shown to be abnormal, but it is more of a confirmatory test than it is a diagnostic test.

And then obviously the sine qua non is the temporal bone showing that dehiscence. Obviously, treatment is symptomatic. We can do surgery, but interestingly, not a lot of people pursue surgery. Most of them just want to know why they hear the noises in their head, and they want to know that they're not crazy. And once we tell them that they're not crazy, then many of them say, "Well, I'd rather just kind of live with this mild dizziness than have surgery." The surgical intervention has become much less invasive. We've moved from a craniotomy, and now many of us are doing a transmastoid, so you go through the mastoid bone. And many of these patients can go home the same day, or 23 hours later, which is nice.

We have looked at our data here, I didn't present them, but you can find this article in the literature. We actually showed, through the two different approaches, symptomatic control that was similar and actually a higher rate of revision with the more invasive surgery. So we've really transitioned to the minimally invasive. It's important to understand, sorry, here is a low-frequency air-bone gap. You can see here this bone hyperacusis. Here you can see the VEMP on the left. And as you can see, you get a response all the way down to kind of what looks like 80 dB. You can see this N1-P1 here, which is much lower than on the right side, where you can see that you've already lost that wave at 90.

So that's the diagnosis on VEMP, a much reduced threshold and, again, hyperacusis. If you guys want to see a quick video of a transmastoid approach, and I think we are just about done, I'm going to show you this. So this is a transmastoid approach. So you can see here, I'm drilling down in the mastoid. Here's the lateral semicircular canal. I'm opening up here the superior semicircular canal. You can start to see the lumen. You wanna be very careful that you don't enter into the membranous labyrinth. You can open up the bony lumen. Here you can see the top portion of the canal, here, the bottom portion of the canal. I always drill underwater so you don't suck out any of that perilymph.

Now what you can see is I'm taking a small micro-pick and opening up the bone over that canal. And you can see actually here, if you can appreciate on your screen, there's kind of a glimmer here. That is the membranous labyrinth inside. That is what you do not wanna penetrate because that can cause some hearing loss. But you can see here, you just take the bone off, make a nice hole, and then you take a mixture of, you'll see that again in a second, a mixture of, I use bone dust and some fibrin glue. You make kind of like a pate, and you shove it down into that labyrinth. And you can see here that that hole is now closed.

Now I've gone to the posterior limb of the superior canal. Again, the membranous labyrinth. You can see opening up that labyrinth. And if I fast forward, here's that bone pate, and you'll see kind of, I take it, and I just shove it in lightly enough to compress the canal, so it's no longer dehiscent, but not enough to cause hearing loss. And that is a transmastoid approach. And this is nice because the patient has an incision similar to that from a cochlear implant. And they can oftentimes go home the same day. All right, and interestingly, this is not a hearing preservation operation. We don't ever tell patients that their hearing will get better. In this particular person, it did.

And you can see here, now when you go back and do the VEMP once you've occluded the canal, that 90 decibel VEMP response is now absent again, just like it should be. As far as hearing outcomes, again, this is not a hearing loss surgery. We don't ever tell patients their hearing will get better. You can see these studies, they're fairly equivocal. Some patients get better, some don't. The risk of hearing loss is pretty low, but certainly, you have to be very careful that you don't get into that membranous labyrinth. Thank you for staying. And again, the enlarged vestibular aqueduct is pretty straightforward. These slides, you can see. I think this is just the most important slide, if I can just do this for one minute.

There was a really nice study around 10 years ago or so where they looked at enlarged vestibular aqueduct and head trauma. So we all tell our patients, if you have enlarged vestibular aqueduct, you shouldn't play sports, you shouldn't do this or that because there's a risk of hearing loss. And what this study actually showed is that the only risk of hearing loss with, the only significant risk of hearing loss with head trauma are in those patients that had fluctuating hearing loss prior to the trauma. So what we now tell our patients with EVA is we want them to come in every four to six months for hearing testing because if they are demonstrating fluctuating hearing loss, we are going to be much more deliberate about recommending lack of, you know, contact sports.

If they have pretty stable hearing or if their hearing has declined but not fluctuated, we're gonna be a little less deliberate about telling them to avoid contact sports. It is obviously always a risk, and we talk about all of that with the parents, but now we have data to show that it's this fluctuating hearing loss that correlates the most with sudden loss after trauma. So I think that was worth reviewing. Here is my contact information. I say, "Whew!" because I didn't even get through all.

- [Christy] Thank you, Dr. Zeitler. This has been a wonderful presentation. I really appreciate you staying on. We hope you all enjoyed today's course. There will be a recording available on AudiologyOnline in the next coming days. So if you wanted to review any of this information or reach out to Dr. Zeitler, there is his contact information. Have a great day, everyone.

- [Daniel] Thank you.