

Dr. Jason Barnes:

Hey there. Welcome to another episode of ENT In A Nutshell. My name's Jason Barnes. And today, we're joined by neurotologist Dr. Alex Sweeney, and we'll be discussing adult sensorineural hearing loss. Dr. Sweeney, thanks so much for being here.

Dr. Alex Sweeney:

It's an honor to be here, Jason. Thank you.

Dr. Jason Barnes:

It's in the title, adult sensorineural hearing loss, but could we start with just what the typical presentation is for an adult who presents to your clinic with hearing loss?

Dr. Alex Sweeney:

Sure. There is, I think, a very stereotypical presentation for adult onset sensorineural hearing loss, but there is also a very wide variety of what we see. I think after the pediatric years, hearing loss can be a problematic symptom for a variety of different people and a variety of different ways. But most typically, I think what we see is somebody who's older, who's suffering from what we would call presbycusis or literally elder ears who is presenting, perhaps at the suggestion of a loved one in the form of what we would call spousal surveillance, where somebody is actually saying, "Well, you're not hearing me as well. You've been complaining about this tinnitus for a long time. You need to go see a doctor."

We probably see it more commonly in men than women, but again, it's something that has a tremendous amount of variability in how it presents.

Dr. Jason Barnes:

And when you first meet these patients, how do you break it down in your mind when you're thinking about different symptoms and onset such as time course, fluctuation, laterality those kinds of symptoms?

Dr. Alex Sweeney:

There are a variety of things that we would consider in terms of the history and assessment of somebody with adults sensorineural hearing loss. I think in terms of associated symptoms or maybe time course, what I would say is, the things that we're most interested in are, were there any sudden aspects to the hearing loss, or was it just a gradual progressive almost not noticeable progression towards the symptoms that led you to come see me or the doctor to begin with? I think that in terms of laterality, we're always curious to know whether there are any asymmetric or symptoms that localized to one ear as opposed to the other.

So somebody who can grossly determine that one ear is their worst ear is always something that would be a key point in the history for us. The same goes for associated symptoms, such as dizziness or tinnitus, the latter of which could also be a very important asymmetric symptom that somebody could have. But overall, I would say that the thing that we are probably most interested in or the things that we are most interested in are the presence of asymmetry, as well as the presence of a sudden episode of hearing loss, durable or not, that has been noted by the patient and their history of present illness.

Dr. Jason Barnes:

And from a resident side of things, we're often coached to ask questions about dizziness, tinnitus, how do those symptoms play a role in this presentation? And what does that tell you about possible etiology of the hearing loss?

Dr. Alex Sweeney:

And frankly, they're both very common things that we hear in association with a hearing loss, probably the tinnitus more than the dizziness. And it makes sense, I mean, as much as we don't understand tinnitus as a phenomenon, there are a variety of things that we actually understand fairly well both in terms of pathophysiology and just in terms of the pattern recognition of tinnitus's association with hearing loss. So the tinnitus is something that we believe in most cases is associated with hearing loss, and so it's a very common complaint that we would see in the adult patient presenting with hearing loss. As I was mentioning before, the presence of asymmetry in the tinnitus is a very important thing to note.

If you look into your American Academy of Otolaryngology guidelines on the management and diagnosis of tinnitus, there are a couple of sections that deal specifically with that, but the presence of asymmetry or just unilateral tinnitus are things that pique our interest in terms of the possibility that somebody would also have a causative ideology that maybe we can't see externally, but if we were to investigate a little bit more with, let's say an MRI, we might find something there that would be a game changer in terms of how it is that we would approach that situation from a management perspective.

Dizziness is something that we see a little bit less commonly, but in the world of neurotology, I would say that there's a pretty rich history of how it is that dizziness, tinnitus and hearing loss could all relate to each other. And I would say most commonly that falls under the auspices of what we would refer to as Ménière's disease. There are also other things like autoimmune inner ear disease that could play a role in that, and some of the things that perhaps we'll talk about later, but the presence of concurrent dizziness and specifically, I would say true vertigo, so the actual sense that the world is turning, is something that would pique our interest in terms of considering a diagnosis like Ménière's disease,

Dr. Jason Barnes:

And when getting your history, what other questions might you ask in terms of risk factors or other historical aspects that might be contributing to this hearing loss?

Dr. Alex Sweeney:

The history is one of the most important aspects of how we assess hearing loss. In some cases, the history is fairly bland and there's not a whole lot to go on in terms of what could be the possible etiology beyond just what we would consider to be presbycusis. But it doesn't mean that there's not an importance to us making sure that we're asking all the appropriate questions. And so when considering the actual breakdown of the history in a more granular sense, there's the past medical history, so did somebody have a history of an infection that led them to possibly receive ototoxic medications? Did somebody have a previous trauma that conceivably could have affected their temporal bone and otic capsule?

We talked about some of the concurrent symptoms that would be important to consider, but maybe in terms of the social history, it would be important to consider, did somebody work in a manner that would have exposed them to loud noise that could be associated with noise-induced hearing loss? Is there a history that is suggestive of a potential genetic etiology to the hearing loss? Maybe radiation exposure that could have gone into a previous malignancy that was treated with radiation as a part of their medical history as well. So there are, there are a variety of different things to consider.

Dr. Jason Barnes:

And from the resident's standpoint, again, when we evaluate these patients with hearing loss, we lean a lot on the audiogram to tell us about the hearing loss and maybe sometimes overlook important aspects of the physical exam. Could you tell us what we should be specifically paying attention to when we first evaluate these patients on physical exam?

Dr. Alex Sweeney:

Just like the history, the physical exam is truly an important part of our assessment of these patients. I think on the external aspects of the ear, we want to see is just in terms of the formation of, and growth of the ear. From conception on, is that you're well-formed, is it not, is that suggestive of potential middle ear etiologies that could be part of what is the causation of the hearing loss? And then working down from the external ear, is there anything in the ear canal that would impede the progression or conduction of sound to the eardrum? Are there eardrum anomalies? Could we see acicular anomalies through the eardrum? Is there something like Schwartz sign like we would see an otosclerosis where the cochlear promontory perhaps has a reddish hue?

All of those things are things that could indicate that perhaps there's a clearly identifiable distinct etiology of the hearing loss, perhaps even in addition to something like presbycusis. There are other things that could be indicative of perhaps some of what we've talked about before, and so the idea that somebody could have had radiation in the past. You might see some of the external changes around the ear to indicate that the temporal bone and the otic capsule has been affected by radiation changes. We can see skin diseases that would be indicative of a possible systemic problem that would also be causative in terms of a hearing loss or their café au lait spots. Are there other cranial facial anomalies that are visible? So there's a lot of things really to consider.

But in addition to everything that I've just said, I think one of the most important things to your point about leaning on the hearing test is performing a tuning fork exam. And so sometimes I feel like I look at the tuning forks and even get a laugh out of the patients on occasion about what we're doing with those, but I truly feel like the tuning fork exam, if nothing else, is a great way to validate what you're seeing on a hearing test. Sometimes the history and the hearing test don't match up and like any diagnostic tests, there's some wiggle room for error, that's built into what we accept a hearing test to be, so performing a good tuning fork exam I think also is an incredibly important thing.

Dr. Jason Barnes:

And next I wanted to move on to pathophysiology and get into more of the details of what causes this process. But before we do, could you give us a quick overview of the anatomy, a little bit of the physiology of what we should understand when we start to think about sensorineural hearing loss?

Dr. Alex Sweeney:

Sure. So I think that it's important to consider as always in our own minds and that of the patients who we're seeing and treating and educating, the distinction between conductive and sensorineural hearing loss is there's a variety of different things that I mentioned previously that we would expect to because in and of themselves a conductive hearing loss, not a sensorineural hearing loss. And so I often find that that's a very important point to make, but when I think about the junction point between where the conductive mechanism is tapering off and the sensorineural mechanism is becoming more important, I'm thinking of the cochlea itself, which is a remarkably elegant and intricate structure that is fluid filled, and that is taking those vibrations from the conductive component and then converting them into a signal that can be understood by the brain.

So when considering the cochlea, the sound is going into the scale of this tubule, the sound is coming out in a sense of the scale of timpani. And then the scale of media is where a lot of the magic happens in terms of the inner hair cells producing that neuro signal or being a part of the creation of that neural signal that then goes into the cochlear nerve, which then takes a remarkably complex pathway through the brainstem and into the brain. I'll oftentimes discuss with people, particularly those who have symptoms of tinnitus and hearing loss that are curious to know more about the true pathophysiology of it, the pathway of auditory input going from the cochlea to the brain, which is something that is again, probably not perfectly well understood, but something that truly is phenomenal.

And I'll use the analogy that it's like a local train as opposed to an express train, because not only is that signal being created and going to the brain, but there's a variety of different stops along the way at different places where there's integration of that sensory input that's combining what's happening in the right ear versus the left here with the and so on and so forth as the sound signal goes to the brain. So in terms of sensorineural hearing loss, there's a lot of different possible mechanisms, but hopefully that provides a bit of a framework for where it is that the problems could occur.

Dr. Jason Barnes:

And with that framework, I wanted to move on to the most common causes of sensorineural hearing loss in this patient population. Could you walk us through these three most common causes and give us a little bit of background and specifics around each?

Dr. Alex Sweeney:

So three causes that I think are fairly common in our world are presbycusis, noise-induced hearing loss and ototoxicity. Those are three things that I feel like I see fairly commonly in my world, in the adult patients with sensorineural hearing loss. And so in terms of presbycusis, there's a very rich amount of literature that's been written on trying to understand this subject, understand specifically where it is in the inner ear that we're seeing this problem arise. Is it something that's happening in the stria vascularis? Is that something that's happening intrinsically in the inner or outer hair cells? And so there's a lot of great literature that's worth looking into, and even some I'll talk with our residents.

Sometimes there's some great literature where people have looked at the histopathology of the temporal bone and tried to correlate that with the actual appearance of an audiogram, whereas the classic presentation of presbycusis would be a high frequency sensorineural hearing loss. There have been thoughts over time that there are different forms of presbycusis. But that being said, what we commonly see, we most commonly see as presbycusis itself. And so presbycusis being something that most classically is a bilateral symmetric high-frequency sensorineural hearing loss. We can see varying degrees of difficulty with speech discrimination.

In a sense, presbycusis is a diagnosis of exclusion, and so I think it's very easy to assume that somebody comes with that hearing tests that they have in the history suggestive of presbycusis, that they have it, but it's something that we always should be arriving at in terms of a diagnosis after in a sense, excluding other potential identifiable causes of hearing loss. But in terms of the impact of presbycusis, it's tremendous. And so the percentages of patients that have presbycusis is quite high when you get up towards the end of the human lifespan. And so it's thought that maybe over half of people by age 75 have some degree of identifiable sensorineural hearing loss.

In terms of the pathophysiology behind this, we talked a little bit about what happens in the inner ear or what we presume happens in the inner ear and have documented happening in the inner ear. From a genetic standpoint, there's been some discussion about whether or not there's a genetic

predisposition for the phenomenon that we call presbycusis there's the group of genes, the DFN group, though that's something that still is a work in progress in terms of our ability to identify a clear genetic predisposition. But presbycusis, I think simply put to me is a combination of two things: it's what it is that you bring to the table genetically, and what it is that you're exposed to from an environmental perspective throughout the course of your life.

And so is it possible that from the environmental standpoint that there is some atherosclerotic component, is there we talked about noise exposure, which is a little bit of a distinct thing, but are there dietary or medical comorbidity considerations in terms of why it is that some people lose hearing faster than others or present at a relatively similar age with very different degrees of sensorineural hearing loss. And so in terms of noise-induced hearing loss, that I would say is a topic in and of itself, but the idea of noise-induced hearing loss is that you've had some acoustic trauma, so a signal that has passed into your inner ear with such energy that it actually has caused an inflammatory response, so actual damage and then the body's innate response to that damage.

I'd say what we believe in regards to noise-induced hearing loss is that there can be a predisposition to damage in the outer hair cells, in the outer hair cells being the amplifier and fine tuner of the sound that makes it into our inner ear. It's not uncommon, I would say in the practice of a neurotologist to see patients who have noise-induced hearing loss, particularly I think depending on where you work in the United States or thereabout, you might find patients more commonly who work in a factory or in some sort of setting where they've been exposed to noise for a very long period of time.

I oftentimes will talk with people about the OSHA guidelines for noise-induced hearing loss, which I actually find difficult to remember off the top of my head, but generally, it's a pretty easy resource to look up to see exactly what those guidelines are. But the thing, I think, that's a key point that I reinforce to our residents is that it's not just a simple threshold of, if you hear a noise that's beyond this intensity that you're going to lose hearing, but it's actually a relationship between how loud the noise is and how long the duration is of exposure. But frequently, as I was saying, we'll see people with different histories of exposure, whether it be machine work, whether it be gun use or chainsaw use or things of that sort.

I think over time, one of the positive things that we've seen in this world is the implementation of noise protection. And so we'll see people perhaps more commonly now than in years past that have your plugs that they're being required to use as an industry standard. So in terms of ear protection, we can see people that are using earplugs. We can see people that are using earmuffs, and we generally expect that somebody can get about 30 decibels of protection. Interestingly though, there's a thought that those things aren't necessarily additive so that you wouldn't necessarily see additional benefit by wearing both the earplugs and the earmuffs together.

In terms of the OSHA guidelines, there's not, I find, a perfect way to remember this, but it is something important to consider. And as I was saying before, there should be a very easy way for this information to be accessed and looked up. But probably if you were going to remember one thing, I think one way that you could create a mnemonic for yourself is that the duration is something that halves, and so it goes from eight hours to 15 minutes where eight hours is the first point in time of exposure timewise that you would expect to potentially see noise-induced hearing loss, or you could consider a hearing loss to be noise-induced. And so it goes from eight hours, four hours, two hours, one hour, or half hour and 15 minutes.

And then in terms of the actual noise intensity that you would hear, it starts at 90, and just with each increasing interval, it goes up by five decibels. So at eight hours, the noise intensity that could go

along with a noise-induced hearing loss is 90 decibels, four hours is 95 decibels, two hours is 100 decibels, one hour is 105 decibels, one half hour is 110 decibels, and 15 minutes is 115 decibels.

Dr. Jason Barnes:

And finally, could you tell us some specifics about ototoxicity? What are some of the commonly encountered ototoxic drugs. And maybe also in the world of ENT, what are some medications that we use that could be ototoxic?

Dr. Alex Sweeney:

Ototoxicity is a fascinating subject, and one just like the ones before that could almost be a talk in and of itself. And so ototoxicity is a general term for whenever something that we are putting into the body generally with a medicinal effect is actually damaging the inner ear. And so what we classically see with ototoxicity are aminoglycosides, platinum-based antineoplastic agents, salicylates, loop diuretics. Those are things that I think most commonly historically, we have good evidence to support that those things are potentially ototoxic. What we're talking about when we talk about ototoxicity, most specifically is probably damaged to the outer hair cells and specifically damage that starts in the basal cochlea, the high frequency area of the cochlea.

As I had mentioned before, the outer hair cells are incredibly important for the function of the cochlea, the frequency tuners of the sound that comes into the inner ear, and in a sense, the amplifiers that allow the phase transition from the air of the middle ear to not be a dampening effect any more than it has to be as moving into the fluid-filled inner ear. Whenever we're seeing ototoxicity, it's those outer hair cells that we think are most commonly affected. There are some medications that could have an ototoxic effect that's transient. I think salicylates are one of the ones that... nothing is so binary that it's always transient, always not, but we could expect that the functional injury that happens with the outer hair cells in aspirin use is something that perhaps could be transient in some cases.

Whereas there are others where we would expect a higher probability that there would be a permanent damage to those outer hair cells. In terms of what it is that we use, I think that very commonly, I worry about ototoxicity in the setting of putting an ototopical agent into an ear canal where there's a large tympanic membrane perforation. Some of the smaller perforations, some of the smaller tubes, I would hope that breaking the surface tension of that medication and it going through the tube or going through the hole would be something that's hard enough to happen to where you might not be likely to see that ototoxic effect.

But I'm generally very cautious, and in fact, avoidant when it comes to the use of things that are ototoxic in the setting of a potential route into the middle ear. And the reason being that we don't want to see the possibility that something gets into to the inner ear and causes those damaging effects. But when considering the treatment of infections, gentian violet is something that's used very commonly. And gentian violet is something that generally, at least in a variety of animal studies, has been demonstrated to be fairly ototoxic. We see the use of streptomycin still in ototopical preparations, and that's something that clearly can be ototoxic.

There are some ototoxic things that are actually preferable or at least ototoxic effects that are preferable also are expected. And have we talked about Ménière's disease previously, but there are some medications that have an ototoxic effect, like gentamycin, which is preferentially vestibular toxic, which is an important point. Not all ototoxic things are just generally ototoxic to the inner ear, but through a lot of history and evaluation, we have a great sense that there are some medications that are more vestibular toxic than cochlea-toxic or vice versa. And gentamycin is one that's actually fairly



commonly used as a vestibular ablation agent in the setting of Ménière's disease which is, which is vestibular-toxic that we use frequently to try to get that effect in our Ménière's patients.

Dr. Jason Barnes:

And with these three more common causes in mind, could you tell us what else you keep towards the top of your differential diagnosis when folks present with hearing loss?

Dr. Alex Sweeney:

So of course there are a lot of things that we should be considering whenever we have a patient who's an adult with sensorineural hearing loss. As I talked about a little bit before, there are things that should jump out at us or that we would want to be on the lookout for that are more historical in nature that can indicate that Ménière's disease is present or autoimmune and inner ear disease is present. And in addition to that, and we were talking about asymmetry previously, there are a variety of different so-called retro cochlear pathologies that could be considered among them, the vestibular schwannoma which is a benign tumor that affects the vestibular nerve, but also affects the eighth nerve in general and also affects cochlear function.

So those are things that I would say that we're generally on the lookout for in terms of distinct etiologies than the more common ones.

Dr. Jason Barnes:

And one question that I like to ask is about the natural history of the disease, meaning what happens if this goes untreated? And at face value, I feel like you can say people continue to struggle to hear, but what are some of the other effects of untreated hearing loss?

Dr. Alex Sweeney:

I like to tell people that hearing loss is in a sense part of the deal. And I say that as somebody who has hearing loss myself. And so in terms of natural history, on some level, given the wear and tear that affects our body everywhere, we might all expect that just if nothing else, the environmental exposures of the world are enough to cause us at some point in time to experience some degree of hearing dysfunction. And if not that, then perhaps something more on a processing level in terms of the pathways that we were talking about before that could create a sense of hearing loss that maybe isn't clearly demonstrated on a hearing test. But in terms of the natural history of hearing loss, I think generally when we see it, there's a pretty decent chance, at least in the world of presbycusis that it would be gradually progressive in things that have a more distinct history of exposure or a history of a sudden loss.

It's a little bit trickier to speculate what's going to happen with that hearing loss, but the hope would be that at least in terms of the actual effect of an exposure, say to an ototoxic medicine, that there's a point in time whenever the hearing loss occurs and that as time goes on, the probability that there's going to be additional damage from that exposure is pretty low. So in terms of the effects of that, they're tremendous. And so people suffer with hearing loss. It's I think one of the things that makes cochlear implants as wonderful as they are is that when you consider the sensory deprivations that we can have throughout the five principles senses that we have.

The hearing loss is a tremendous burden on the people that suffer it, and so to know that we have something at the end of that, just philosophically, I think it's a comforting thought hopefully to every human in existence that might suffer from hearing loss. But in terms of the primary effects of what we might see, I think first and foremost are safety issues. And so just from our own personal

experience, we actually have natural disasters where I'm located that can involve hurricanes and tornadoes, and so when you consider how it is that somebody could be awoken in the middle of the night and to tell them of an impending disaster as it relates to a natural disaster or a weather related phenomenon, very often there's an auditory alarm that there's some of sense of noise that alerts you to the fact that something is happening.

And so if there's a fire, if there's a doorbell, if there's an alarm, those are things that you wouldn't necessarily be able to hear as well as you should with hearing loss. I think in terms of what we would consider to be secondary effects of hearing loss, one of the biggest concerns that people will have when they come in is employability, which hopefully is something that as time goes on, we'll get better at mitigating as I was saying, in terms of the rehabilitative options that we have, as hearing aids and cochlear implants have evolved, I think there's a lot of great things that we can offer to people. But employability can be a problem where people have lost so much of their hearing that they actually are unable to maintain the level of performance that's expected of them at their job.

Independence is another big one, isolation is probably one of the most devastating ones, both for patient and provider. When you consider our role as otolaryngologist, as communication specialists, the fact that somebody loses their ability to effectively communicate is truly at right smack dab in the middle of the heart of what it is that we do. So that's a particularly terrible one to see. And one of the things that is becoming a hot topic or not even becoming, is a hot topic in our world of the relationships between hearing loss and cognitive decline, and so one of the things that I think a lot of research is focused on in that world now is how it is that the cognitive effects of hearing loss could be potentially mitigated by the use of some hearing rehabilitation, being a hearing aid, being a cochlear implant.

And so I think the verdict is still out on just how much that we can affect that potential relationship between cognitive decline and hearing loss, but I think that what I'll say to people is that out of all the factors that would influence cognitive decline in terms of when it happens and how it happens, hearing loss might be one of the biggest modifiable factors of cognitive decline. So it's something that we clearly have to be very tuned into.

Dr. Jason Barnes:

Once a patient presents to your clinic, they probably already are presenting with an audiogram or maybe it's pending, but say, they don't have one, what is your workup for this patient? And we can start with the audiogram. What are you looking for on the audiogram and what are some key aspects of the audiogram?

Dr. Alex Sweeney:

The audiogram, I think, is a critical part of our evaluation. There are things that as I've mentioned before, that we should be doing in our own end to go along with and evaluate the history of somebody's hearing loss or the physical exam, but the audiogram provides us with one of the most important objective findings that we have that can help us in the assessment of somebody who presents with hearing loss. And so in terms of the audiogram, I generally will break this down to patients as there are being three principle components, which is an oversimplification, there's a lot of very important things that are happening in an audiogram, but one of the first things that actually jumps out to a patient with most audiograms is the big graph that's on the front. And that is an assessment of truly sound perception.

So if meaningful hearing is both sound perception and sound understanding, that graph that you see, which can be two graphs, if it's divided into the right and the left ear like RSR, but there are still a lot of folks that will have it all on one graph that are testing the perception threshold of pure tone sounds.



And so looking at it and looking at the little lines you might see on the vertical sense. And so numbers on the X axis are associated with the vertical lines, it's like keys on a piano keyboard. Again, this might be a little bit of an oversimplification, but it's just, I think, a helpful way to think about it, and it's something that I explain to our patients.

And so what we're doing is we're using pure tones of sound, and by pure tones, I mean, a specific frequency of sound, and then when you look at the horizontal lines on the audiogram, those correlate with a certain sound intensity. And so we take a pure tone, let's say, 250 Hertz, which is a very low frequency sound, and then we play it louder and louder until the sound is perceived at which point in time a mark is placed. And we do that across not the entirety of what we would expect the human ear to be able to perceive, but quite a bit of it. And that gives us a sense of whether or not sound perception is impaired.

And then the second part of what we're doing is getting to the second point of what makes up meaningful hearing, which is sound understanding. And so we're using validated lists of words, and in most cases and depending on what the evaluation is for, there can also be sentences that we're using, which provide a little bit of context, but in essence, what we're trying to do is to see beyond our ability to perceive a noise, is it possible that you don't understand the noise or that there's something that's happening in the transmission of that signal from the ear to the brain that's garbling the signal enough to where it becomes more on the nonsensical end of the spectrum.

And then the last thing in terms of breaking down the hearing test into three principle parts that I think are very important, is the tympanogram. And so that gets to more of what we see on the physical exam, but there are aspects of how it is that the conductive mechanism of hearing can be rendered less functional than they should be, that are things that would show up on a tympanometry. And so in terms of our measuring emittance with an eye, the process of a tympanogram can give us a sense of pressure dysregulation under the eardrum, in the middle ear

It can give us a sense of in some cases, a secular dysfunction where there is enough stiffness and the obstacles that we don't see as much movement in the tympanic membrane. And we can actually see the opposite where there's a secular instability or discontinuity, which leads the tympanic membrane to be more movable, I guess, it's probably not the best word. We can see a secular discontinuity or instability that makes it so that the tympanic membrane is less stiff than what we would expect.

Dr. Jason Barnes:

And are there any specific audiogram patterns that might clue you in to the etiology of hearing loss?

Dr. Alex Sweeney:

Absolutely. I think as I've mentioned before, one of the first things to focus on is the distinction between a conductive and sensorineural hearing loss or the presence of both of those things, but when considering sensorineural hearing loss, I think historically, and to the present day, we consider presbycusis to be something that can have a variety of different effects, but probably the most common thing that we see is high-frequency sensorineural hearing loss. So the so-called ski-slope appearance of the pure tone thresholds where low frequencies are relatively preserved to the high frequencies, which are more effected.

So in addition to that, what we can see is a notch shed 4,000 Hertz or the so called 4k notch that can be indicative of noise-induced hearing loss. There's the so called cookie-bite hearing loss, which is actually more of a middle frequency in terms of the spectrum of what we test in an audiogram hearing loss. And so, relative normalcy or at least thresholds that are less effected at low and high frequencies and more effected at mid-range frequencies, which can be indicative of a congenital potentially even

infectious etiology and congenital sense. We can see up-sloping low frequency hearing losses most commonly in Ménière's disease.

And so ones in which the lower frequencies of sound have been disproportionately affected relative to the higher frequencies of sounds, sort of the opposite of the classic ski-slope audiogram that I referred to in presbycusis.

Dr. Jason Barnes:

And do you regularly obtain imaging in this kind of classically presenting elderly patient with sensorineural hearing loss?

Dr. Alex Sweeney:

I do not, but there are some fairly good guidelines on how we should approach that question. I think that it's not uncommon just from a philosophical perspective, from an emotional perspective, even when people come in with hearing loss who are say, very bothered by tinnitus, that they really want us to get imaging to evaluate. But the fact of the matter is, we are in a wonderful place when considering the past 100 years with what we're able to identify, but that being said, we're also not so sophisticated that we can identify with simple diagnostic tests, the etiology of many forms of sensorineural hearing loss.

In other words, we would actually expect in many cases that imaging might be negative, which in a sense is reassuring if we don't find any clear causative etiology, but I would say that there are certain patterns of hearing loss that might make us more likely to pursue imaging. And generally, in terms of conductive hearing losses, which is maybe a little bit apart from this a CAT scan or a computed tomography is something that's very valuable to us. But when we're talking about sensorineural hearing losses, I would say that the MRI is generally what we're considering.

And when considering the MRI and the patterns of hearing loss that would maybe be a more indicative of something we would see on an MRI, probably the two most common things are a sudden hearing loss or an asymmetric hearing loss. The definition of a sudden hearing loss that is classically used is a 30 decibels of loss at three frequencies occurring over about a three-day period. And so that's something that is worth evaluating for sure with a MRI. And what we're looking for, as I said before, is something that fits into the general category of retro-cochlear pathology, like the vestibular schwannoma.

In terms of asymmetry, that's a little bit trickier. And so asymmetry, there's actually a variety of different definitions for asymmetry, and I think depending on whether or not you're interested in finding every possible evidence of retro-cochlear pathology, or you're more interested in just having a higher probability of finding something whenever you're doing the MRI, or whenever you're ordering an MRI, you can have two different guidelines. But the general guideline, I think, that's perhaps most commonly accepted is having two consecutive frequencies with the 15-decibel asymmetry when comparing one ear to the other.

But those are, I would say the most common reasons that we would order an MRI, but as I referenced the American Academy of Otolaryngology guidelines as well, there's also evidence to support that we would get an MRI in the setting of asymmetric tinnitus, though from a probability perspective, I would say that it's less likely that that is going to yield a positive finding or something that's clearly the etiology of the hearing loss.

Dr. Jason Barnes:

And now considering treatment, there are two main treatment options for hearing rehabilitation, including hearing aids and potentially cochlear implantation. Could you start with describing to us how you counsel patients on hearing aids, who is generally considered a good candidate for hearing aids and what are some pros and cons to pursuing that treatment option?

Dr. Alex Sweeney:

Of course. Hearing aids are wonderful devices, I've mentioned cochlear implants before because I think that they're wonderful and I'll talk about them in a second, but I think that the progress that's been made in the cochlear implant world shouldn't overshadow the progress that's also been made in the world of hearing aids. And so hearing aids are wonderful options for probably the majority of people that experience adult-onset sensorineural hearing loss. And I think historically, the conception of a hearing aid is that it's one of those big cones that you put up to your ear, and so it's very ostentatious and it's just going to make all sound that you hear louder, and that's going to be a terrible thing because maybe you want to hear sound X better, but then you're going to also hear sound Y better, and that's going to be bothersome to you.

But in reality, I would say that hearing aids have evolved so much that they almost, I joke with people, give them an unfair advantage in today's world, they're smart, they're discreet. The hearing aids can interface with a variety of different electronic aspects, there are a lot of different electronic devices that we have with us. When considering the appropriate patient for a hearing aid, I think there can be a little bit of or at least in my world, I am not infrequently surprised by the people who benefit from hearing aids and want hearing aids, and then people who don't benefit from hearing aids or don't want hearing aids.

But the general sense is that there's a consideration of what's technically aidable, and historically speaking, the definition that I have gone by is if you are able to score above 50 or 60% in terms of your word recognition, which was one of the aspects of the audiogram, and there would be a thought that putting an amplifier on your ear would make it so that you're amplifying enough good signal to where you would get benefit from it. Now, I think that that's a generality, it's not necessarily true for every patient, because we do see people that actually have less word recognition who are very happy with their hearing aids.

So I think you have to take it on a case by case basis, but I think that considering the pure tones of what's aidable, 75 decibels and low frequencies, 90 decibels and high frequency. Some of the cons of a hearing aid, I think that in today's world, many forms of the sophisticated hearing aids that I'm talking about are very expensive, and so it might be problematic in terms of access and some of the physiologic cons of a hearing aid. And so when you're considering some of the hearing aids that actually include the ear canal, there can be a problem with what's called the occlusion effect, where you're actually blocking some sound out while amplifying other sounds. And so, that can be problematic.

We see people really, actually fairly routinely in our clinic who have such a hearing aid who also have infectious issues where the actual environment of the ear canal has changed by the constant use of a hearing aid that blocks the ear canal, and so they're developing bacterial and fungal infections as a result. So those are all things that I think are important to consider in terms of the pros and cons of hearing aids. There are people still, despite all of that, and despite the counseling that we provide, that have such a negative reaction to the hearing aid, once they get them that they become non-users, which we try everything we possibly can to avoid in terms of how we structure our counseling between the neurotologist and the audiologists.

I'd like to think with appropriate counseling, the rate of non-use is actually fairly low. And I'm very hopeful as I imagine many people are, that as we evolve as a society, the stigma associated with

hearing aids, where people would assume that a hearing aid means that you're elderly and nobody wants to have that particular stigma is perhaps diluting a little bit over time and becoming less significant, but we still see that as an issue as well.

Dr. Jason Barnes:

And you mentioned cochlear implantation, could you tell us some of the pros and cons of cochlear implantation and who is a candidate for this procedure?

Dr. Alex Sweeney:

Yeah. Cochlear implants, they're very biased, I would consider to be the most incredible, sophisticated and successful neural prosthesis device ever discovered, invented, created by humanity. And so, as I referenced before, I think one of the best parts about cochlear implants is that when you consider the end result, the end of the line for sensory deprivation in any sense, we actually have an answer whenever people have lost hearing, whereas with the sight, with taste, smell, there are definitely great answers and wonderful things happening in those worlds, but cochlear implants for a variety of reasons have really gotten off the ground in terms of neural dysfunction in that sense.

And so, the person that I think in a general sense is a possible candidate for a cochlear implant is the person who comes into you who's tried a hearing aid, maybe tried multiple hearing aids, that just isn't happy, isn't satisfied, and it's for reasons that have to do with their hearing acuity. And so, we can see a lot of people that are dissatisfied for a lot of reasons, but the person who's struggling with what you would consider to be the best fit hearing aid is the person that at least in my world, should go to be evaluated for a cochlear implant. And so when you consider that evaluation, it's a fairly systematic thing.

And so per the FDA at this point in time, cochlear implantation is considered whenever somebody has presented in the world of let's say, presbycusis with less than 50% understanding, and that ear, that's the worst ear, so the ear that's potentially to be implanted, which in our world, I would say in a general sense is the worst ear. And then less than 60% understanding and what we would consider to be their better ear. And again, this is done in the best aided condition. So it's actually a test to be distinct from a traditional, basic audiogram hearing tests, which can involve little inserts that are put into the ear or headphones, the evaluation for a cochlear implant is something that involves the patient wearing the best possible fit of hearing aids on their ear.

The FDA is creating a labeling for the cochlear implant, but an important thing I think to consider in our world is that beyond that, there's the question of, does your insurance company follow that labeling? Does your insurance approve the use of cochlear implant or in other words, will they be willing to pay for the implant? Because the issue with costs, even though hearing aids is a unique world and how insurances cover it and don't cover it, perhaps more commonly, don't cover it. Cochlear implants, if it were to be not covered, if somebody were paying cash is really the overwhelming majority of cases, a prohibitive enterprise.

So, it's very important that the insurance company buys into it and that you're meeting criteria for it. And so one of the notable deviations from what I said that the FDA does is actually CMS. And so Medicare, it's a lot less in terms of what somebody can hear. At least the residual auditory function has to be less. And so what Medicare's criteria are at this point in time is that patients have to have a moderate to profound hearing loss in terms of their pure tones, and they have to be able to understand less than 40% of what they hear in the best aided condition. Cochlear implants are in general, they're wonderful device, I think that in terms of pros and cons, the biggest pros I was saying before, is that it's providing an option for people that historically have had no option.

And the option is pretty good, we expect there to be very good function with the cochlear implant. In the general sense, we expect people to resume a lot of their normal activities. But the cons with it is I'll be very distinct with people, is that it requires a surgery. And while that surgery has become something that we can do in less than an hour, and it's an outpatient procedure in most people, it still is a surgery. And so not everyone is a candidate for a surgery from just a general medical and physiologic perspective. It's not a light switch. And so even though it's using electricity to stimulate the auditory system, it's not something that you just put into somebody, and then all of a sudden they wake up and their hearing is just like it was when they were younger and it presumably was better.

So it requires a tremendous amount of work and effort on the part of the cochlear implant team, which in our world is a cochlear implant surgeon, a cochlear implant audiologists, a speech pathologist potentially. There's a variety of different people that contribute for the whole spectrum of auditory rehabilitation from hearing a noise to having that noise be tuned, to the oral rehabilitation that ultimately make the cochlear implant as successful as it can possibly be, which is different than perhaps what somebody would expect with a surgery in which maybe we do the surgery and shortly after, their hearing improves.

So we expect some degree of improvement, but there also is a process that we would expect people to go through.

Dr. Jason Barnes:

And finally, could you speak a little bit to "hidden" hearing loss, a patient who presents and doesn't objectively have hearing loss on an audiogram, but is really bothered by this, hearing loss that they perceive.

Dr. Alex Sweeney:

Yeah. It's both fascinating and troublesome phenomenon that we see in our world because I would say increasingly, it's more common than I'm seeing these patients. As you said, the hidden hearing loss is somebody who presents with, in most of their, if not all of their objective evaluation indicating no discernible hearing loss in terms of their pure tone appreciation or word discrimination or sound understanding. And the thought process behind this is perhaps more aligned with what I was saying before about the complexity of that auditory signal, going from the inner ear to the brain.

It's not just the express train that goes right there, and so as long as you hear it in your ear, it's interpreted by your brain and that's that, but there's a variety of different processing things that happen in route. And so in the brainstem, in terms of how it is that sound is integrated with a sound from one ear with the other ear. And so it's possible that in that fashion, that there are people that don't have a measurable hearing loss in terms of how we measure it, but are still suffering in some sense with regards to their appreciation of sound.

I'd say most commonly where I get this complaint from patients or this concern from patients is with regard to sound and noise. And so folks that seem like they're doing fine in a quiet room, but as soon as they go into their favorite restaurant with all of their friends, they become more or they have a greater degree of difficulty in terms of their ability to hear and understand sound.

Dr. Jason Barnes:

Well, Dr. Sweeney, thanks so much for this discussion regarding adult sensorineural hearing loss. I wanted to move on to the summary, but before I do, is there anything we didn't talk about that's worth mentioning or anything you wanted to eliminate?

Dr. Alex Sweeney:

No. I think that hearing loss is at the core of who we are as neurotologists in so many different ways. And I think that hearing loss is a tremendous burden to the people that suffer from hearing loss. And I think that there's quite a few of those people, especially when considering the percentages that are associated with hearing loss as we age, I think that as I referenced before, we really have wonderful rehabilitative things to help folks with hearing loss. And I truly believe that as time goes on, that we'll only get better in our abilities to rehabilitate hearing loss.

And so I think that as much as hearing loss is a dramatic burden on the people that suffer from it, I feel like the message that I try to give people whenever I see them is one of hope. I think that there's a lot out there that we can do to help, and we work with our patients, we work with our referring doctors, we work with our friends and families to make sure that nobody is suffering unnecessarily whenever they're experiencing a hearing loss.

Dr. Jason Barnes:

Well, thank you so much. In summary, adults who present with hearing loss often present with gradual-onset multifactorial disease, which includes presbycusis and possibly a history of noise exposure. The most common causes of adult-onset sensorineural hearing loss are presbycusis, noise-induced hearing loss and ototoxicity. Hearing loss is most commonly due to injury of the inner or outer hair cells, depending on the mechanism of injury. And workup mainly includes an audiogram, but CT or MRI might be warranted depending on the clinical situation.

Treatment options include hearing aids and cochlear implantation if hearing aids do not provide enough benefit. And treatment of hearing loss helps in many ways, including from a safety standpoint, improving social function and employability, as well as possibly deterring cognitive impairment in a subset of patients. Dr. Sweeney, anything else you'd like to add?

Dr. Alex Sweeney:

No. Thank you so much for having me, it was really an honor to be here.

Dr. Jason Barnes:

Appreciate it. I'll now move on to the question-asking portion of our time. As a reminder, I'll ask a question, wait for a few seconds to give you the opportunity to think or press pause, and then give the answer. So the first question is, what are some of the effects of untreated hearing loss? As we discussed, there are some primary effects, including safety issues, such as not being able to hear a fire alarm, a doorbell, a telephone or other things that could be a warning of a possible disaster or something going on in your surroundings.

In terms of secondary effects, there's reduced employability, reduced independence and social isolation. And we also talked about the possible link between hearing loss and cognitive impairment. For our next question, what are the OSHA guidelines for noise exposure? As Dr. Sweeney said, this might not be the easiest to remember, but might be worth trying to come up with an easy mnemonic. And that's by starting at eight hours of duration with a 90-decibel noise exposure. So at eight hours, it's 90 decibels, half that is four hours, and increased five is 95 decibels, two hours, 100 decibels, one hour, 105 decibels and so on.

And for our next question, describe some commonly seen audiometric patterns and what they typically represent. For presbycusis, we will often see sloping, high-frequency hearing loss, or the ski-slope pattern, a 4k notch is usually noise-induced hearing loss. The cookie-bite pattern is usually



congenital hearing loss, and the up-sloping hearing loss is more associated with Ménière's disease. And for our final question, what is a good rule of thumb when considering what patient would be a good candidate for a hearing aid?

When considering offering a hearing aid to a patient or considering who would be a good candidate for hearing aid, word recognition scores of 50 or 60% are generally considered better for hearing aids, where if it's lower than that, they might not gain as much benefit from hearing aid. Thanks so much, and we'll see you next time.