- [Christy] At this time, it is my pleasure to introduce Dr. Daniel Zeitler, who is a Board Certified neurotologist and skull base surgeon in the Department of Otolaryngology, Head and Neck Surgery at the Virginia Mason Medical Center in Seattle, Washington. Dr. Zeitler graduated from Northwestern University and earned his MD with honors from NYU. He completed his otolaryngology, head and neck surgery residency at NYU, and a fellowship in otology, neuro-otology and skull base surgery at the University of Miami Ear Institute. He is also a fellow of the American College of Surgeons, the American Academy of Otolaryngology, and the American Neuro-Otology Society.

Thank you so much for joining us Dr. Zeitler, at this time, I’ll hand the mic over to you. Thanks, Christy, it's great to be back. Christy and I used to work together here in the Pacific Northwest, so we go way back, I guess. Everybody, thanks for joining, it's a strange time and I appreciate all of you coming in today. And I think I'd be remiss not to start with a thank you to everybody out there who is doing whatever part they have in this time of need, whether it's seeing patients or just staying at home and being safe.

So thanks for showing up. I want us to spend the time today talking about conductive and mixed hearing loss, and specifically otosclerosis and other causes. I have no financial disclosures, and here are my learning objectives. You do have to pass a quiz, but a little word to the wise, I've seen the questions and it should be pretty easy. Talking about mixed and conductive hearing loss is kind of like drinking from a fire hydrant. And so what I wanna do today is kind of parse down the subject matter. We’re gonna be sort of focusing on physiology, on audiology, as it remains apropos to all of you out there, and we’re gonna spend a little less time talking about the surgical implications of these diseases, but certainly if there is interest, I’d be happy to come back another time and talk specifically how we do these surgeries. The other thing that I wanna say in order to sort of parse down the material is I’m spending the time today talking about those causes of mixed and conductive hearing loss with normal eardrums, normal middle ears, normal ear canal. So what we’re not talking about today are things like cholesteatoma, chronic otitis media with middle ear effusions, tympanic
membrane perforations, things like that. So you all have a very specific objective, when it comes to mixed and conductive hearing loss, you see the patients you make a good diagnosis with a diagnostic audiogram. Hopefully, as we'll talk about today, you do all of the necessary tests that I wanna see. And then you talk to the patients about what their options may be, whether it's surgical, audiological, whether it's amplification through a traditional hearing device or bone conduction. But I have a pretty straightforward objective, if I see a patient with this kind of hearing loss my role is to get an audiogram that looks like this, to look like this. And so, through the talk today, I'm gonna talk a little bit about how I go about thinking about these diagnoses, because the more I know ahead of time, the better equipped I am during surgery to have an outcome that is achievable such as this.

So I think when we look at a hearing test, we see a typical hearing test that we all kind of associate in our minds with otosclerosis, the classic Carhart notch, the low frequency moderate hearing loss, and an ear canal that looks normal, an eardrum and middle ear that look normal. And I think we'd all be pretty comfortable calling this, at least, preliminarily otosclerosis. But it's an ear that looks like this, where the physical exam is identical in my clinic and under the microscope, but the audiogram just doesn't look like a typical otosclerosis picture. You've got this low frequency moderate loss rising to pretty much normal, there's no Carhart notch, there's no high frequency hearing loss as is typical with otosclerosis. So, in my head when I see this, I'm thinking maybe something other than otosclerosis, and I'm gonna hopefully convince you all today, that you in the booth can sort of start making these preliminary diagnosis based on just what you're seeing on the audiograms. But in order to do that, it's important that we first understand middle ear mechanics. The function of the middle ear is pretty straightforward, we have to transform acoustic waves that come in the form of mechanical vibrations, and we have to turn those into waves that stimulate the fluid of the inner ear. And for our distant ancestors who lived in water, that was pretty easy because you are going from fluid to fluid. And as we all know, fluid is a much better conductor of sound than is air. But as we moved from water to land, and we started
walking and using air, we not only had to evolutionarily adapt to getting this
transformative property to occur, but we also needed increased efficiency because
now we're going from air to fluid, rather than fluid to fluid. And to do that, we had
something called exaptation, which is a process in evolution whereby structures that
pre-exist are repurposed for another cause. And so as you can see, in this picture, let
me see if I can get this pointer to work. On the left, you can see here, this sort of
rudimentary middle ear, what we call columella. There is a stapes that formed at the
bottom of this columella, that was a distinct and separate occurrence. And the goal of
this was to get sound directly from the tympanic membrane to the inner ear. But as you
can see, as these animals evolved, you can see that there was a small bone in the
lower mandible called the articular bone, and that became the malleus here in the
middle ear, and there was a bone in the upper jaw called a quadrate bone, which then
became the incus. And so in doing this, we, not we, but in doing this animals created a
much more efficient process of connection, a much more efficient connection between
the tympanic membrane and middle ear, as I'll show you.

So the typical tympanic membrane is about 9 1/2 by 8 1/2 millimeters. It’s not perfectly
circle. I tell my patients when I'm talking about it in the clinic that it looks almost like
the nail on my big finger, and there are both circumferential and radial collagen. This
may or may not be on your quiz. The circumferential collagen provides a softness that
gives the eardrum efficiency in the low frequencies, whereas radial stiffness or the
radial collagen gives the stiffness and allows better efficiency in the higher frequencies.
And so what that allows the tympanic membrane to do is to vibrate different at different
frequencies. So in the low frequencies, the entire tympanic membrane kinda moves like
a sheet, almost like a bedsheet, where the whole thing was up and down together and
the malleus in this case moves as a lever. This is true for frequencies much below 1000
hertz. Whereas once you get much above 2500, or 3000 hertz, the eardrum kind of
becomes a much more complex system. There are different parts of the eardrum that
move differently with maximum displacement around the umbo, and almost looks like
an ED current, if you will. And this in effect uncouples, the malleus from the tympanic
membrane. So it moves in more of an elliptical fashion, almost kind of like this, rather than up and down like this. And this allows not only more flexibility but more mobility. And this again stresses the importance, not only of the eardrum and malleus themselves, but there are several folds within the middle ear, one of which is called the manubrial fold. That allows the malleus to decouple itself in a way to become more efficient. The other thing that these three ossicles allowed the ear to do was to create better efficiency through a lever system. And there are three main levers in the middle here. The first is what we call a catenary lever. I call this basically the hammock lever. If you imagine a hammock hanging between two trees, the weight of that hammock on itself, sort of dipping down is kind of what we call a catenary lever. And in the case of the middle ear, it's the game that's realized by the tympanic membrane resting on the manubrium while supported on both sides rigidly at the annulus and acting upon itself by its own weight.

So this is called the catenary lever. The ossicular lever which is the middle ear bones themselves is the gain that's provided by the length or the difference, I guess I should say, in the length of a manubrium to the length of the malleus or the long process, sorry, the length of the manubrium of the malleus to the long process of the incus. And we'll look at this a little more closely in a second. And lastly, the hydraulic lever, the most important and the most efficient lever. And what this allows is you take sound from a tympanic membrane that's about 20 times larger than the oval window and you directly couple these two things due to direct attachments of the bones. And so you can take sound that hits a eardrum and parse it down to a sound that's much more amplified because it's hitting a much smaller area, again the hydraulic lever. And just to review these, the catenary lever again the lever system provided by the way to the tympanic membrane on the manubrium of the malleus. The ossicular lever which you can see in green there, which is the length difference between the malleus the incus, which is about 1.3 to one. And lastly the hydraulic lever you can see in blue or purple there, which provides a ratio of about 20 or 21 to one between the tympanic membrane and the oval window. And if you sort of add these together, you get a
theoretical gain of about 34 decibels. So sound that hits the tympanic membrane can be amplified about 34 decibels through the middle ear ossicular system. But interestingly, the actual gain is not 34, it's probably closer to 20 or 25 decibels. And the question is why do you lose some of the efficiency of what is supposed to be a very efficient system? Well, first of all, there are different portions of the tympanic membrane that vibrate at different frequencies like I said. So certainly, in certain frequencies, this 34 decibel gain may be more true, however, at other frequencies that 24 or 25 decibel gain may be more true. Second of all, these models all assume that there is perfect frictionless movement of the ossicles and that they move in perfect planes with each other. And so certainly we know that there is not only some slippage of the ossicles around the lever axis, and some friction in the system, but there's also force that's needed to overcome the stiffness and mass of the tympanic membrane itself. And then lastly, as we all know who all of us work in the field of Audiology, there is a reduction in the middle ear space in many patients due to Eustachian tube dysfunction, perhaps they've had some scarring from previous otitis media. So again, the systems that assume perfect function are very rarely perfectly functional. And this is what accounts for that loss of theoretical gain. Let's specifically talk about the ossicular chain itself. So first, the malleoincudal joint.

Again, I learned a lot as I went through the histopathology, and my fellowship as far as these ossicles work, I kind of always just thought of them as three bones that move and amplify sound, but they're much more complex. The malleoincudal joint is actually a diarthrodial joint. So basically this is the kind of joint in our elbow, in our knee, in our shoulder. It is a true diarthrodial joint or a hyaline cartilage on each side with synovial fluid in the middle. And as you might expect, there is primarily a hinge-like motion at the low frequencies so it moves up and down. However, at the higher frequencies, these two bones actually twist around each other slightly, almost like beveled gears. And I always tell my patients, it's these two movements that make getting a perfect outcome, sometimes a little more difficult because we're not only trying to recreate which is pretty easy to do an up and down motion, but we're also trying to recreate a
twisting motion which is very difficult in manmade ossicular reconstruction. Incudostapedial joint is identical, it's also a diarthrodial joint with cartilage on both sides and synovial fluid. And there is a articular disc in the middle with fibrous meniscus which you can see, just here. Here's the distal process of the incus and the head of the stapes. And what’s interesting about the incudostapedial joint is in addition to this disc, this diarthrodial joint. There is also a joint capsule that wraps around the head of the, or the lenticular process of the incus, as you can see here, and sort of envelopes the entire incudostapedial joint. And this serves a very important purpose, which is that it prevents liftoff of the incus from the stapes. So you blow your nose really hard, you're going scuba diving, and you're equalizing pressure, and you pop your ear, that explosive force could theoretically take that incus right off of the stapes as it’s connected to the malleus. And this joint capsule keeps those two bones together. This is also a really nice thing to understand because during stapes surgery when I’m trying to separate the incus from the stapes to do a otosclerosis operation, you can actually lift the incus up very gently and see that joint sort of there, there's a sort of a clear fluid in that joint, and it helps you understand where to make that cut between the two ossicles.

So they incudostapedial joint. The stapes motion, much like the malleoincudal joint has two components, it rocks and it moves up and down. As you might anticipate, it's the piston motion that dominates. This is why we can do stapes surgeries. This is why we can have a stapes piston that recreates hearing. But it’s this rocking motion that has a higher threshold, it's certainly not as important because it has a much lower sensitivity for cochlear activation. But this I think, is the reason why when we do stapes operations, we can get the air-bone gaps to close, in some cases perfectly, but in some cases, we don’t get a perfect closure. And most specifically, the lack of that perfect closure oftentimes is in the high frequencies. And again, I think it has to do with the fact that we really cannot create this rocking motion of the stapes, whereas it’s pretty easy to recreate the piston motion. The stapes join to the annular ligament, you can see here, this is a pathology slide. Here’s the stapes and here’s the annual
ligament. This would be the oval window niche here or the vestibule opening. Unlike the other two joints, this is not a diarthrodial joint, this is a fibrous joint, it is basically has a bursa which you can see there. It is stiffest posterior-inferiorly. And it's this stiffness of the annular ligament that actually works against you in some cases when you're doing ossicular reconstruction and you have an intact stapes footplate, which we'll talk about in a little while. But if you're putting in a TORP, or a PORP and that ossicle reconstruction prosthesis is too long, you can actually load that annular ligament too stiff, and it doesn't vibrate. Imagine if you put someone who's really heavy on a trampoline, it's not gonna vibrate when you jump and the same is true with this annual ligament, if you make it too stiff, you're gonna actually lose efficiency rather than gain efficiency. So these are all the subtleties that we have to think about. So let's start by talking about otosclerosis. This is a patient of mine, you can see that the stapes piston has been placed. You can see here, the round window, here's the oval window, or what would be the oval window, there's a piece of fascia here with my prosthesis over it, here's the long process of the incus ear canal tympanic membrane lifted up. Again, not really gonna talk too much about surgical technique today.

Otosclerosis is certainly one of the most common, if not the most common causes of conductive hearing loss in adults, it has a clinical prevalence of about 0.5 to 1%. But interestingly, if you take temporal bones and you histologically evaluate them the disease prevalence is probably in the neighborhood of 10 to 12%. Whether this has to do with patients who never come to presentation or whether it has to do with subclinical disease, we don't really know. But certainly there is a much higher prevalence anatomically, than there is clinically. But interestingly, the disease prevalence in those who have it, there is a 60 to 90% chance that it's gonna be bilateral. So, again, 10% histological chance, about 1% clinical chance, and in those patients who do have it in that 5.5 to 1%, almost half to even three quarters, and maybe even 90% of them will have bilateral disease. There's a very significant female to male predilection. We'll talk a little more about this in a second. You can see the most common onset is in the third to fifth decade. But there are children who get this
which we'll talk about later, and is exceedingly common in Caucasians. And if you look at Caucasians, compared to African Americans, there's almost a 10 times higher prevalence in whites with otosclerosis. So, just one of those tricks, if you're seeing a patient who has a conductive hearing loss and they are of color, certainly something to think about, that the prevalence of otosclerosis is much lower. Talking about the female to male predilection, some people have proposed that there may be an endocrine etiology, due to this two times more common disease in women. Interestingly, bilateral disease is also more common in women. And a lot of people will say that the clinical onset if not the progression of disease gets worse during pregnancy. Now again, we don’t have true data about this and we don't know whether it’s truly the pregnancy that’s spurning the worsening or onset of the disease, or evolutionarily, women who are pregnant probably have a tuning of their audiological system in preparation for listening for their baby’s cries. But again, we do see and often hear people who will say that their disease either started or got worse during pregnancy. And interestingly, because of this people have investigated this and they have found that there are in some specimens estrogen receptors in the otosclerotic plaques.

Now, certainly males get otosclerosis, so we know that this can’t be the whole story, but certainly it may explain some of the story. What I think is most fascinating about the disease, otosclerosis is that audiometry can actually tell you a lot about the disease itself. And these are some of the most interesting audiograms to look at. The early disease, kind of like the audiogram that I showed you at the very beginning of this talk will typically have a low frequency mild to moderate conductive hearing loss, and oftentimes that will sort of narrow in the mid frequencies at the Carhart notch, we'll talk about in a second. And then again in the higher frequencies, will drop back down to a mild to moderate loss. So that typical sort of inverted V pattern. However, in late disease, oftentimes the hearing loss becomes almost a flat conductive loss through all the frequencies, because you lose the efficiency of the Carhart notch, you lose that vibration as the footplate of the stapes stiffens increasingly, and so the Carhart notch
disappears and so the hearing actually becomes a flat pattern. And then in very severe
disease, which we call labyrinthine or cochlear otosclerosis, or in some cases, you
might have heard it called far advanced otosclerosis, if the otoscope erotic plaque,
which you can see here, penetrates into the cochlea, which you can see here, then
you're gonna end up with cochlear otosclerosis, or far advanced otosclerosis, and you
can actually get a drop in your word recognition score and a sensory neural hearing
loss. And I think it's important here, on this picture, just to show you a little anatomy.
Again, this is the footplate of the stapes, you can see this dense otosclerotic bone with
this yellow arrow. Here's the cochlea. This is the vestibule, and this is important, which
we'll talk about in a second. Here's the saccule of the vestibule and this is why you
want to avoid operating on patients with active Meniere's disease. Here's the facial
nerve coming down. Here are some of the semicircular canals.

So we sort of hinted at the Carhart notch phenomenon. This is a depression in the
bone conduction threshold at around 2000 hertz, sometimes it's described as little
closer to about 1700 hertz, but this can be a pretty significant drop. In fact, you can
see a little drop in the bone line across all frequencies five db, at both 504,000, a little
less at 10,000, sorry at 1000. But at 2000 hertz, you can often see a depression of
about 15 decibels. And while it looks like they have some early cochlear otosclerosis.
In fact, this is an artifact because the primary resonant frequency of the ossicular chain
for bone conduction, as I mentioned is around 1700 hertz. And so you get this
artifactual drop. And in many cases, this disappears after surgery. And in many cases,
the air conduction actually supersedes where the bone conduction line was. And I
always like to joke with my patients that I've given them hearing they never thought
they had. What are the treatment options? So I think we're all familiar with this.
Certainly observation is an option, certainly amplification through either traditional
devices or bone conduction devices. We will not really talk about surgery today. And
then there is some evidence of some medications which we'll talk about right now. For
a while, people thought that fluoride was implicated in the disease, you need fluoride
for bone building because fluoride helps stimulate osteoblasts which are bone building

continued
cells and it decreases osteoclasts, which are bone destruction cells. And so the thought was if you give fluoride you can induce stability in the bone and reduce bone remodeling. And so some people gave it. Now, what we found is certainly that if you look histologically at these patients who had had fluoride once they passed away, there was absolutely no evidence of effect, that should be effectiveness. But interestingly, CT studies over time in some cases have shown that these otosclerotic plaques that you can see on scans, what we call hyperlucencies, or sort of otospongiotic findings on CT scan can actually lessen or disappear. And so this gave some people hope, that fluoride may be beneficial.

Certainly in today's treatment algorithm, in 2020, there is absolutely still significant equipoise, with some proponents still purporting its efficiency, but most of us really don’t use it regularly. Not only because it doesn’t seem to work, but also as you can see there, there are a number of side effects that you just don't wanna deal with, especially in women who may be pregnant. Now, what's interesting about stapes surgery, I put up this guy, Dr. Kessel he was in Germany, and the first successful stapes surgery actually occurred, sort of by happenstance. There was a patient who was riding on, as the story goes, was riding in a horse carriage, the horse carriage tipped over, the patient fell, and when he woke up when he came to consciousness, he said, "Gee, my hearing, in the ear that used to be deaf "is much better." And the idea was that what he had probably had was otosclerosis, and when he fell off the horse carriage he had knocked the footplate loose of his stapes, what we call a stapes mobilization procedure now, and that led to his better hearing. So Dr. Kessel sort of experimented with what was the most early stapes surgery with these sorts of rudimentary instruments, they would put a flame into this device here, which would allow them to use this very rudimentary otoscope, and look in the patient's ears, this would be the light. A little more refined today. When we think about stapedectomy, we typically wanna see a patient who's got a minimum air-bone gap of 15 to 20 dbs at at least two consecutive frequencies. I like to see three, but certainly two can do the job. And perhaps just as importantly, if not more importantly, as we like these patients to
demonstrate what we call a flipping of the fork, which means that if you do the Rinne test, which hopefully we're all familiar with, and you put the tuning fork at 512 hertz on the back of their mastoid, they will hear it better than they do through air conduction. And this demonstrates not only that they have an audiometric chance for recovery, but you wanna make sure that they have a clinical chance for recovery. In other words, if you have a hearing test that shows a conductive hearing loss, but the patient doesn’t appreciate that hearing loss, it’s gonna be hard to make them better. There are some relative contraindications, which are listed here. I did sort of briefly allude to Meniere’s disease in a patient who has demonstrable and active Meniere’s disease, you really want to avoid stapedectomy because if you open up that vestibule, and you accidentally get either your instruments or the laser or your hook into that expanded saccular you can actually cause sensorineural hearing loss. But we never or at least I never do a primary stapedectomy surgery in a perforated tympanic membrane because you’re making a sterile environment unsterile and there is a higher risk for sensorineural hearing loss.

And again, these other things listed here, we'll talk a little more about the inner ear malformations later, and then far advanced otosclerosis is a little controversial, some people think you should go directly to cochlear implant if they have very poor word recognition, whereas others think you should try stapedectomy first. When we talk about surgical outcomes, these are just good numbers to have in your head to talk to your patients about, we typically say that we can get a successful air-bone gap closure, which we describe as an excellent result being less than or equal to 10 dbs, in upwards of 90 to 95% of patients. We will get a partial air-bone gap closure between 10 and 20 dbs and basically the remainder and a very small proportion of 1%, probably even less than that will have an unsuccessful result with a smaller portion of that 1% having actually worsening of their hearing. If you look at these patients over time, and you look at delayed hearing loss, so you compare their operated ear to their non-operated ear to make sure that you’re not accounting for other things like presbycusis, etc. There's really a pretty low risk of worsening of the conductive loss or the
sensorineural loss faster than the other ear having had a stapedectomy. So, patients always ask, is having the stapedectomy gonna put this, sorry I have an itch in my ear. Is having the stapedectomy gonna put my operated ear at risk down the line? The answer by the data seems to be a pretty resounding, no. So moving on, to superior semicircular canal dehiscence. I always like to do this right after stapedectomy and otosclerosis because these are the two things that seem to be most commonly confused, both audiometrically and clinically. So let’s talk a little bit about superior canal dehiscence. Hopefully you’ve all seen this. Hearing in a normal ear, I think we all understand, there’s an inward motion of the stapes, there’s an equal and outward motion of the round window. That fluid wave goes around the helicotrema and vibrates the cochlear partition and this gives us sound. And this is again in a normal ear. Now, what about in an abnormal ear?

So let’s first talk about why you lose air conduction in superior canal dehiscence. So, again, let me get my handy pointer. You can see here, sound goes in the stapes into the vestibule, but now some of that sound is lost through this the dehiscent, what we call third window. And so what you can actually see is that you lose efficiency of that system and therefore you reduce air conduction. So, you have shunting of the fluid volume as I mentioned, this leads to elevation of air conduction thresholds. And what it also does is it decreases the mobility of the round window, because you have this loss of efficiency. And so you can actually measure lower input impedance of the inner ear because of that, as well as umbo hypermobility. So you can actually do electrophysiological studies and find that because all of this air and sorry, all this fluid wave is getting shunted here, the stapes actually moves hyper dynamically, which allows you to measure the umbo of the malleus, which also moves hyper dynamically. So this is why you have increased air conduction thresholds. It’s almost entirely due to loss of efficiency. I think more interesting and more unknown is why we have decreased bone conduction thresholds. So a lot of these patients, as you probably know, will have what we call hyper threshold or super threshold, bone conduction measurements in the negative five to negative 10 area, which really we don’t typically
see. And so let's talk a little bit about this. So, what happens is, you again have sound go into the footplate, you have the vibration of the fluid in the inner ear, you have loss of the efficiency through that exposed, superior canal dehiscence here. And so what you get because of that, is you get an unequal distribution of vibration across the cochlear partition. So you can see that as sound goes in here, you've lost efficiency, but now you can have extra movement around the cochlear partition because it's vibrating a little more dynamically. And so you have super threshold fluid motions in the scale of tympani. And what this does is it increases the responsiveness of the cochlea to this compressional wave, and you get what we call a hyperdynamic bone conduction. And so, very, very typically what you will see in these patients is a bone threshold over or less than zero. So it's very, very important in a patient who has mixed or conductive hearing loss, that you take care, if you get a bone conduction of zero, that you make sure that you haven't gotten to the threshold, because this can often help us determine, again audiometrically, what disease we're talking about.

So we've kind of talked about this first line, the typical symptoms of superior canal dehiscence, vertigo, sound pressure and due sensitivity, which we call Tullio phenomenon, or Hennebert's phenomenon which are sound induced vertigo. And these can often be absent. And the oral symptoms which are typically associated with superior canal dehiscence including aural fullness and autophony, as well as pulsatile tinnitus can also be absent. So these patients can present almost identically to a otosclerosis patient. And there's one key to the diagnosis in addition to those super threshold bone conduction that is absolutely mandatory that you look at. And those are acoustic reflex thresholds. So I get a lot of referrals from audiologists on the outside of our institution, who send for otosclerosis and we very, very often, probably 50 or 60% of the time have to do the acoustic reflexes before the patient sees us because they haven't been done. So if you're gonna do conductive hearing loss measurements and you're gonna identify these types of patients, it is absolutely mandatory that for pure conductive loss, or even mixed loss, you do reflex thresholds, assuming that the tympanic membrane and middle ear are normal. Obviously with a perforation, or
middle ear effusion, or a flattened pentagram, this can't be done. It's very important that we take a detailed symptom review, we talk about vertigo and dizziness, we talk about sound and pressure induced dizziness. Again sound induced dizziness is Tullio phenomenon, pressure induced dizziness is Hennebert's phenomenon. We ask about oral symptoms. We've talked about those bones thresholds. And then lastly, VEMP and CT imaging. And we'll look a little more closely at those right now. So first of all CT imaging. Here you can see, on the left panels, this is a CT of a superior canal, you can see, we call this the arrow sign or the radiology sign. The radiologist as pointed to this pretty significant dehiscence here in the roof of the superior canal. Here's the malleus and incus. Here, just for comparison is what a normal would look like. Here again, is in another view, where you can see, just where this... I'll put this arrow away 'cause it's easier to see, but the arrow is pointing to that dehiscent canal there. And then on the right lower panel, you can see what a normal canal would look like.

So when you have a patient with a conductive or mixed hearing loss and reflex thresholds are present, the first thing I always do is get a CAT scan to make sure this is not superior canal dehiscence. The other thing that you wanna do is a VEMP. So, here you can see this is a patient with superior canal dehiscence in the left ear. You can see, I just wanna highlight there, that was super threshold bone conduction at 250 and 500, again, very, very classic for superior canal dehiscence. And what a VEMP test does is it uses a rudimentary vestibular reflex, we play a loud noise to the ear, and then we watch for a simultaneous response in the sternocleidomastoid muscle on the same side. And what we typically see, the classic findings for superior canal dehiscence would be that the VEMP threshold is substantially lower, which is to say you can continue to get a P1-N1 wave at a threshold much lower than you would in the contralateral ear. So here you can see on the top graph, the left ear, you're getting a nice P1-N1, all the way down to about 80 decibels. Whereas in the right ear, you can see there's a nice P1-N1 at 125, but it's already gone at 90. And again, this is a classic finding. And so classic in fact, that in my practice, I will not do an operation for a superior canal dehiscence without an abnormal VEMP, and certainly without an
abnormal CT scan. The other thing that’s important to know, as we all see these patients is that superior canal repair is not a hearing operation. These are just four studies that showed that basically patients who, had had a surgery for a superior canal dehiscence. And you can see that they basically show that there is no significant improvement in their hearing. Here’s a study we actually did at our institution, my partner, Dr. Seth Schwartz was the lead author. And again, we broke it down MFCA and TMA are two different approaches to the surgery, but the long story short, you can see, there were no statistically significant differences in the pre and postoperative air-bone gap after these patients had surgery, and more importantly, we even broke it down by frequency just to make sure that we weren’t missing those low frequency air-bone gaps. And you can see, even at 500 and 1000, there was no statistically significant difference between pre and postop. So, it’s very important that we tell our patients a superior canal dehiscence surgery is for vestibular symptoms or auditory symptoms, it is not for hearing. Let’s talk about enlarged vestibular aqueduct. This is something you might see in some of your pediatric patients. This is a really cool photo that was taken by one of my friends, Dr. Larry Lustig at Columbia. So I wanna give him credit for that.

So here, on the left, you can see what is a typical vestibular aqueduct. The vestibular aqueduct is a bony channel whereby the endolymphatic duct passes from the posterior fossa into the inner ear, and you can see again, this is a normal caliber. Whereas here again, the radiology sign, or the arrow sign shows you a significantly enlarged vestibular aqueduct. Now there are a lot of different measurements, and tricks, and things that determine whether your vestibular aqueduct is enlarged or not. Here's one that's kind of a classic. They take these measurements and do tangent lines, and this and that, and I've never really been good at geometry, or trigonometry, so I just use a pretty simple one, which is the diameter of your posterior semicircular canal, which is right here. If your vestibular aqueduct is bigger than that canal, then it’s too big. And so not that we’re gonna have you all looking at CT scans, but certainly, if you see them, it’s certainly interesting to just take note of the vestibular aqueduct. This is a really nice

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study that was done about 10 years ago where they looked at patients who had enlarged vestibular aqueduct, keep in mind most of these are children. And you can see that bilateral EVA is far more common than either the right or left ear alone, about twice as common. So typically a patient that has enlarged vestibular aqueduct is going to have mixed or conductive hearing loss in both ears. And these patients will also often as you can see, about 40% of the time, have other malformation, so they may have enlarged vestibular aqueduct plus other cochlear or vestibular anomalies. And lastly, you can see which I think is most interesting, pure conductive loss is extremely infrequent in patients with enlarged vestibular aqueduct. In fact, most of these patients, almost three quarters of them will have a mixed hearing loss, whereas 20% have a sensory neural hearing loss and only a fraction have a conductive hearing loss. And usually the largest air-bone gap in the mixed and conductive patients is in the low frequencies.

Again, this can look very similar to otosclerosis and superior canal dehiscence. In a child who has conductive or mixed hearing loss, I get a CT scan in every case, because these are the kind of things that you want to make sure you find. The physiology of the hearing loss is poorly understood in enlarged vestibular aqueduct. Some people think it may be due to the pressure underneath the stapes, so you actually get, almost kind of like reverse otosclerosis. It’s not the footplate that’s stuck it’s the pressure underneath the footplate that prevents it from moving, due to the increased perilymphatic pressure from that enlarged aqueduct. It could be that there’s a direct communication between the CSF and the inner ear. And perhaps this may explain the link between head trauma and hearing loss in these patients. And you might have heard about that. We’ll talk about that more in a second. And then lastly, this could act like a third window phenomenon much like superior canal dehiscence that we just talked about. Now, I’m not gonna talk too much about enlarged vestibular aqueduct, but the one thing that I wanna make sure that I hit on is head trauma, we’ve all heard or probably all have heard the link, or possible link between head trauma and enlarged vestibular aqueduct. And this is why for many of these patients, we actually
recommend avoidance of contact sports and things like that. And this was a really beautiful meta-analysis that was done about five years ago in Otology Neurotology journal, where they combined 30 studies with about 180 patients who had about 350 enlarged vestibular aqueduct again, sort of that, half of them have bilateral disease idea. And about a third of them had a sudden hearing loss after a head trauma or a noise trauma. But interestingly, when they looked more closely at these patients, the only thing that correlated with a sudden hearing loss after trauma was whether or not their hearing was fluctuating prior to the trauma. And the odds ratio of that was about 8 1/2. So patients who had fluctuating hearing loss prior to trauma, were about 8 1/2 times more likely to have a sudden change in their hearing after trauma as those patients without fluctuating hearing loss. And interestingly, we've all kind of thought about this being linked, but there was no link or no risk for sudden hearing loss after trauma if patients had progressive but non-fluctuating hearing loss, vestibular symptoms or Pendred syndrome, which is a genetic abnormality that causes enlarged vestibular aqueduct, and some thyroid abnormalities.

So it’s really, really important when you see these patients and they're asking whether or not they need to avoid contact sports, you can certainly say probably, it would not be a bad idea. But if we really wanna use the data, the best way to know whether you are or aren’t at risk is to get a hearing test every three to four months. And if we document absence of fluctuation, then they’re probably fairly safe. Certainly you wanna use good clinical judgment, and informed consent, and collaborative decision making as you talk about these things. But there does seem to be a pretty good, evidence-based decision making tool that you can use. Again, enlarged vestibular aqueduct can also mimic otosclerosis in adults, much like superior canal dehiscence did. There are some reports throughout the literature where patients underwent stapedectomy without any noticeable improvements and then had a CT scan and sure enough, had enlarged vestibular aqueduct. I wanna spend a couple of minutes talking about something that’s pretty rare, X-linked stapes gusher syndrome. I forgot I did that neat little animation there. X-linked stapes gusher syndrome is a mutation in the DFN3
locus, genetic focus sorry, on the X chromosome. And this is important because since it's on the X chromosome, we don't see it in women. So if you have one bad copy, and you have one good copy, then you're not gonna have any symptoms, or clinical manifestations. But if you have only one X chromosome to start and you have a mutation, you're gonna have the disease manifest. What do we typically see? We typically see a communication between the subarachnoid space and the perilymphatic space. And that's due to this very, very abnormally formed internal auditory canal that communicates directly with the stapes here through this, what we call an absent crib reform plate. And so what that does is much like I talked about for enlarged vestibular aqueduct, it creates a significant pressure of CSF underneath the stapes.

So it's not that the stapes is fixed, it mimics otosclerosis, but importantly, it's that the stapes can't move because of the back pressure. And in these cases, mixed hearing loss is most common again, this is why I get a CT scan in all of my children with mixed or conductive hearing loss. But certainly my ears are perked up a little more in young males with conductive hearing loss than it is in young females, for this exact reason. And this is important to understand ahead of time because these patients have incredibly bad outcomes after stapedectomy, they often have failure to close the air-bone gap, there is a much higher risk of profound hearing loss. And in these patients you wanna consider a cochlear implant or a hearing aid depending on the severity of the disease, rather than an operation. And again, this diagnosis is going to be made ahead of time with a CT scan. The hearing aid or bone conduction aid can work beautifully in many of these patients. And certainly the cochlear implant can too but from my standpoint, there are some things that are a little more subtle about operating on a patient with X-linked stapes gusher syndrome, a much higher chance for CSF leak during the operation. So you wanna be prepared for that. And because of that abnormal cochlea, you can actually insert the electrode into the IAC. And so these cases are often or can be done with real-time fluoroscopy or real-time X-ray to show that you're putting the electrode where it's supposed to be. What about ossicular abnormality? So not otosclerosis and not stapes abnormalities, but other
abnormalities. So again, sort of my emphasis at the beginning was, you can often use a hearing test to sort of make a determination, preoperatively what you're gonna be dealing with, and this is a perfect example, here you can see in the left ear, a basically flat maximum conductive hearing loss, there is no Carhart notch, there is no sensorineural loss that would be suggestive of otosclerosis. And so, I see this patient in my clinic, and I'm thinking, "Gee, this is probably not otosclerosis, "or not superior canal dehiscence, "or not enlarged vestibular aqueduct syndrome." So you can really do some differential diagnostic elimination, just based on the audiogram. What can cause this maximum conductive hearing loss? Well, anything that causes a ossicular disjunction behind an intact tympanic membrane. And that hearing loss gets worse and worse, as the disjunction becomes more significant and more complete. And you can actually max out your conductive hearing loss at about 55 to 60 db.

Again, you can see there, sort of the list of things that can cause ossicular disjunction. The incus is especially prone to necrosis because it has a watershed blood supply and what that means, you can see those arrows there coming into the incus. Kinda like the Continental Divide that runs through Colorado, there's water that goes to the Pacific on the west side and water that goes to the Atlantic on the east side. But it's that sort of dividing line where you don't really know where the water goes. It always makes for a fun game when you're with your kids on a road trip and you have them pee on the Continental Divide and tell them to guess which ocean it's going into. Or maybe that's just a game I play. But it's especially prone to necrosis because of this watershed blood supply. And so anything that causes inflammation or infection, chronic otitis media, cholesteatoma, recurrent otitis media is going to primarily affect this distal incus. And you can see here a couple of examples. So, here's an ear that I operated on. This is a large perforation. You can see here a large tympanic membrane perforation, here's the chorda tympani nerve. Here's the head of the stapes, here's the distal incus. You can see here, just between the stapes and the incus, sort of what looks like a rudimentary incus. In fact, what that turned out to be, was kind of a fibrous adhesion, where there was sort of erosion of the incus with minimal to no

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communication between the two ossicles and a significant loss of hearing. Now, this patient had a TM perforation, so I could actually see this ahead of time in the clinic. But here’s another example where I could not see it. So here’s a second patient. Again, looking, this is a left ear. Here’s the ear canal, the ear thumb is lifted up here. Here’s the head of the stapes, chorda tympani nerve. Here’s the distal incus, sort of hiding behind the posterior canal wall. And you can see the sort of hypervascular strand of fibrosis, which is interesting. That was just a fibrous adhesion between the remnant of the incus and the stapes, which serve no functional purpose, and this was taken out and replaced. Here’s another example. Here you can see again, left ear incus round window, and here you can see this sort of rudimentary posterior crus of the stapes but there’s no stapes superstructure. Here’s the anterior overwin, anterior footplate, sorry, with this nice shine on it, but there’s no anterior crus. This is a patient again, who had a maximum conductive hearing loss, who I knew ahead of time probably did not have otosclerosis and sure enough, I was right.

And again, this was purely based on the hearing test. The workup, again microscopy, if you have a tympanic membrane perforation in many cases, you can see these abnormalities, but in many cases the eardrum is intact, you often get a deep tympanogram, we call a type AD because there’s no resistance from the ossicular chain, and so the eardrum becomes sort of hypercompliant. The reflexes are typically absent, but again, with absent reflexes and a maximum conductive hearing loss, you’re gonna be thinking less otosclerosis and more ossicular discontinuity. And ultimately, you can always do a CT scan, if your willies feel a little weird, then you can certainly image these patients and try to get a sense ahead of time, but the treatment is still surgery. So in many cases, the imaging is not terribly necessary. In very mild cases of incus necrosis, and there’s a pretty significant communication or the gap between the incus and the stapes is small, you can take little fiber cement, this is that patient I showed you before with that big perforation. Sorry, not fiber cement, bone cement. And you can actually bone cement these two bones together. This is some gel foam in the middle ear, which was in preparation for the eardrum repair that I was gonna do.
next. In other cases... And Kim, I'll take the video, now. In other cases, the distance between the two bones is too significant to do any kind of bone cement. So what you can actually do is what's called an ossicular reconstruction. So the first thing we do is we measure in the middle ear the distance that we need between the stapes and the tympanic membrane. Here you can see a tool that has that measurement, you can see. Difficult to see but just right here, there's a little thing that says three, that's three millimeters. You trim it to the appropriate size, you cut off, here again, you can see a little more clearly that three right there and it has four, five, six markings. You take the ossicular prosthesis, out of its trimming mechanism, and you bring it up to the middle ear. Again, this is a left ear we're operating on. If anybody is squeamish, it's not too bad, but you may wanna look away. There you can see directly down the canal is the head of the stapes. So this is the patient that I showed you before that had that very long fibrous adhesive band between the incus. And you take this prosthesis and you can see it's got a little connection on the bottom which sits right on the head of the stapes. And you very gently push the prosthesis down onto the stapes, like so.

So now you've got your prosthesis, here you can see the stapedial tendon of the stapes. Then you take a little piece of cartilage because you don't want the titanium in direct communication with the backside of the eardrum. You trim a little piece of cartilage, and you put that right on top of the prosthesis there, again, making sure that there's no impingement of that cartilage on the posterior canal wall. And then you very gently take the eardrum and put it back down in place. And if you're lucky, or good, or a little of both, your patient will have significant improvement in hearing. All right, I'll take the slides back. All right, here you can see a couple more examples of ossicular reconstruction prostheses, over the years I've done. This one on the left is with hydroxyapatite. On the right is what we call a TORP. So what I just showed you was a PORP, which the partial ossicular reconstruction prosthesis, the one on the right is a total or TORP ossicular reconstruction prosthesis. This is that patient who had that absence stapes and you can see this piece of titanium. Where'd my arrow go? I don't know where my arrow is. But you can see that the piece of titanium... There it this. You
can see that the piece of titanium sits right here directly on the stapes footplate. I'm gonna skip this. And then just for the sake of time, we'll talk really quickly about tympanosclerosis. We've all seen this in the clinic. Many of us have probably confused this for cholesteatoma but tympanosclerosis is basically a hyaline degeneration of those fibrous layers in the lamina propria of the eardrum, those collagen fibers that we talked about about an hour ago, and they actually calcify. And we don't really know why this happens, we think it can be infectious or inflammatory, but these plaques can often thicken and lead to fixation of the ossicles or the tympanic membrane. And again, sort of highlighting the importance of the audiogram, sort of a fourth very different autogram, sort of a low frequency, mild to moderate conductive hearing loss that rises to normal.

And interestingly, even though you can have extensive disease, the audiometric findings oftentimes don't really connect with the severity of the disease. If you do surgical exploration, you can find this tympanosclerosis at a number of different places in the middle ear. The most common is the anterior ligament of the malleus, so it fixes the malleus. You can also see it at the malleoincudal joint, the staples and pyramidal process, and even down in the round window niche. And when you take the stuff out, it literally looks like cartilage. It has a hardness to it, it's got a three dimensional structure to it. This is not what you would see in that eardrum that I just showed you in the introduction slide for tympanosclerosis. That was just kind of scarring, but this is what you might find the middle ear or underneath the eardrum in cases that are pretty severe. And then just to finish up with pediatric considerations, I don't know how many of you out there do pediatric hearing loss. But we do know that middle ear malformations in the pediatric population are not uncommon. You can see there one in about every 11 to 15,000 kids. The majority of these are some form of stapes ankylosis or fixation. And certainly between stapes isolated alone and state ankylosis with other anomalies, that takes up about two thirds of the patients. There are other ossicular malformations where the stapes is normal. And then you do have a small percentage of these patients that have very severe dysplasia or aplasia of the oval window or round
window. And again, another pretty unique audiometric pattern, you can see a very significant, very severe low frequency conductive hearing loss arising to what may be sort of a mixed hearing loss in the higher frequency is typical of these particular patterns. When we think about conductive hearing loss in the young kids, we think about congenital footplate fixation versus juvenile otosclerosis. This is involving the stapes specifically. There was a nice paper that was put out by the Mayo Clinic where they compared these two. And when you think about congenital footplate fixation versus juvenile otosclerosis, these patients tend to be younger, they tend to have worse hearing, they tend to have a greater incidence of other ossicular abnormalities, and the outcomes after surgery tend to be worse. So congenital footplate fixation is something that we tend to try to avoid operating on if we don’t have to, certainly while they’re young. Typically, I would wait to do a stapes operation in a young kid, at least until their teenage years, and many of these patients do quite well with traditional amplification or bone conduction.

And here again, I just wanna sort of highlight that while we do want to help these patients the overall success rate of air-bone gap closure to an excellent result was much lower. So for adult onset otosclerosis, or typical otosclerosis, we can get that air-bone gap close, like I said, in about 90 to 95% of patients, but in juvenile otosclerosis and congenital stapes fixation, that rate is only about 70. So this is again, things that I talked to the family about ahead of time, and make an informed and collaborative decision. So, in conclusion, otosclerosis is certainly the most common cause of conductive hearing loss in adults so it can be diagnosed without additional testing. Please, please, please remember your acoustic reflexes. But many things can mimic otosclerosis, and for those things, it’s really important again, to do your reflexes, think about your H and P, consider imaging or VEMP testing when necessary. And most importantly, if you’d learned one thing during this whole hour, oftentimes, it’s pretty easy to predict the etiology of hearing loss based on the audiometric patterns. So look at your audiogram carefully and encourage the surgeons you work with to do the same. Knowing that diagnosis before surgery certainly helps me communicate the
safety and outcomes to these patients, we can talk about the risks and benefits. And it allows me too to sort of think ahead of time about what type of reconstruction I might use. And I think most importantly, as we talked about for a few of these things, it’s also important to know when not to operate, not that any of you will be operating, but as surgeons, we need to know that. So with that, I ended perfectly on time and I will be happy to stay for extra minutes if there are any questions.

- [Christy] Zeitler, we have a couple questions here related to vestibular. The first one comes from Christine. Christine asked, how frequently does otosclerosis present with vestibular symptoms and is that only in the advanced cases?

- Yep. So if I have a patient who has conductive hearing loss and vestibular symptoms, I am thinking almost anything except otosclerosis because they don’t tend to go together. So if there are vestibular symptoms, of course not considering BPPV, that obviously can go with anything. But if it’s something that really truly seems vestibular, especially if you’ve done vestibular testing, and you have a unilateral weakness, or other abnormal findings, I strongly, strongly encourage you to think of things other than otosclerosis.

- [Christy] And that should answer Claire’s question. She asked, how often is it that you see vertigo or dizziness symptoms in patients with otosclerosis? So it seems like it’s pretty rare. Is that right, Dr. Zeitler?

- Yeah, so you can see them together, but it’s kind of, correlation does not imply causation. Otosclerosis itself really just doesn’t cause dizziness. So, it can be otosclerosis and dizziness, but it’s probably not otosclerosis causing dizziness. Now one thing I will say, it is not uncommon to get benign positional vertigo after stapedectomy, and that goes to that anatomy that I showed you, the inner ear, the vestibule and the saccule is extremely close to the footplate. And so you can sometimes cause a little irritation or manipulation of the utricle and some of the otoliths
down there. So it's not uncommon to have a patient with some benign positional symptoms after stapes surgery.

- [Christy] Thanks Dr. Zeitler. We have a comment and a question from Peter. Peter, thanks you for this excellent presentation. And then he follows up with the question. Are there any clinical findings or implications of arthritis for middle ear joints?

- That's a great question. So I didn't talk about this kind of stuff. It doesn't seem that typical sort of degenerative arthritis that we see in adults has any implications for middle ear disease. However, there are some things that cause arthritis type diseases such as rheumatoid arthritis, osteogenesis imperfecta, Paget's disease, and those can absolutely have implications for the middle ear and the ossicles. We didn't go into that today because that's kind of a whole 'nother topic. But typical sort of old age arthritis, and even osteoarthritis does not typically affect the--

- [Christy] We'll take one last question here from Erica. Sorry, from Margaret. What type of temp do you expect with otosclerosis? Perfect. Well, thank you Dr. Zeitler, so much for your time and your expertise. We are just so delighted to have you back on in 2020. And we hope to hear from you soon.

- It's my pleasure to be here and I look forward to the next time and if people have questions that we didn't get to, I'm happy to answer them by email or otherwise. So, feel free to give out my email, I think--

- [Christy] Thank you Dr. Zeitler. Have a great day, everyone.