Semicircular Canal Dehiscence: Lessons for Collaboration, in partnership with the American Academy of Audiology

Presenters: Rebekah F. Cunningham, PhD, Devin L. McCaslin, PhD

Hello and welcome back to the classroom. Before I introduce our presenters for today's presentation, we have a video message from our Academy President, Dr. Tish Gaffney.

I'm Tish Gaffney, President of the American Academy of Audiology. The Academy is the home for audiologists. We are dedicated to achieving recognition of audiologists as the experts in hearing and balance care and the field of audiology as an independent clinical specialty that is integral to the healthcare team. We offer our members the latest resources for practice management and clinical evidence based practice so that they can grow professionally and improve patient care. We advocate for our members in Washington and throughout the country to ensure that the voices of audiologists are heard and we help connect audiologists to one another through valuable networking opportunities.

If you like this webinar, then make sure you join us each year at the annual AAA Conference for Thought Leadership that you won't find anywhere else. Learn more about the conference and how to join the academy@audiology.org thank you. It is my pleasure to welcome Dr. Rebecca Cunningham and Dr. Devin McCaslin today.

This course is in partnership with the American Academy of Audiology. If you'd like to learn more about our guest speakers, you can read their bio on the course registration page, but at this time I'll hand the mic over to you. Hello, My name is Rebecca Cunningham. I'm a pediatric audiologist. I am employed by Natus Medical and below are my disclosures and Dr.

McCasland's disclosures.

Please take a moment to read the limitations and the risk put forth by Audiology Online and the American Academy of Audiology regarding this course.

And finally, I would like to go over the learning outcomes of this presentation, discuss the incidence of superior semicircular canal dehiscence in children, describe subtle symptoms of SSCD in children and and how they vary from those that we see in adults and explain the need for interprofessional collaboration in clinical or school based patient populations.

Our agenda will be as follows. Dr. McCaslan and I will share presenting as I'm a pediatric audiologist who does not do vestibular Dr. McCasin has graciously decided to join and help me kind of understand all the things that we're going going through. So we are going to talk about what is superior semicircular superior semicircular canal dehiscence.

We're going to talk about the symptoms, we're going to talk about demographics for patients. Then we're going to present our case study on at the time a nearly 11 year old girl Treatment options, what can be done, what is typically done, and then some reflections on this particular young lady.

Okay. There will be some polling questions throughout this. So the first one is all about who you are. So which best describes you? I feel like this is very important when we need to learn about the audience that we are speaking with, so that I get a better feel for what you actually do on a daily basis in regards to children, vestibular testing, testing and things like that.

So we're just going to wait a moment here to see some of these results come in.

Okay, so 15% of you see children regularly, 25% see. Do not see children. And about 60% of you. So the majority see children sometimes. So that's wonderful to see.

Thank you.

Next question. Do you do vestibular testing for children? I think in our profession, this has come to light to be something relatively newer. I do believe that it's been going on for a long time, but I don't think it's come to the forefront until probably the last 10 years. So it's very interesting to me to see what's going on out there.

So give us one second here so we can get our results. I'm very interested to see this. Yes, at least once a week was nobody. Yes, less than once a week was about 14%. And about 86% of you said, no, I do not do vestibular testing for children.

And I think those responses are pretty reflective of what is actually going on out in the real audiology world.

Okay. So for the purposes of this presentation, instead of using the big mouthful of superior semicircular canal dehiscence syndrome, we will be abbreviating it to SSCD or scds. Typically, I will use sscd. All right, let's talk a little bit about what it is. Okay.

All right, Dr. McCaslin. Yep. Thank you, Dr. Cunningham.

And thank you, everyone, for a good morning or afternoon based on where you are. It's an honor to present here in the academy putting this on. So excited to do this. Dr. Cunningham asked me to sort of become involved in this patient and as she was working through it.

And I will say we have a pediatric vestibular clinic here at the University of Michigan, and we have audiologists come from our children's hospital over to our main balance lab, and they see kids. And I will tell you that children are some of the most challenging cases we see, not only in the testing, but also the diagnoses they present with and things that you see a lot of in years. Neuritis in adults, but you will see genetic anomalies and all sorts of things that you wouldn't expect. And it's really, you know, one

of the most challenging sort of populations to test with regards to vestibular. Again, not just in how to test, but, you know, also, you know, the presentations that they have.

And this is one of those cases that we're going to take you through and you know, the child will present with a number of diagnoses and, you know, we'll take you through the decision making process. Such an interesting case, such a phenomenal child. And you know, we'll be interested to hear what you think at the end. And so to give you a little background on one of the presentations that diagnoses that the patient came with is superior canal dehiscence, as Dr. Cunningham mentioned, and just a little background on it and in the pediatric population.

So it's only been around since 1998 and that doesn't seem like that long to me. Again, I'm pretty old, but that to me is a relatively new sort of disorder. And Lloyd Minor was at Hopkins at the time, Johns Hopkins. He is a neurotologist and he's now at Stanford where he's a dean. But while he was at Hopkins, he and David Z, who is an otoneurologist and their team presented these eight patients that were patients that if you put pressure in the ear, they would get dizzy, they would have eye movements in the plane of the canal.

They found that these patients, after having a fistula repair or stape indectomy would often have these symptoms. And they really came up with this mobile third window theory. And again, there are three window. Well, there are two windows, right? There is the oval window to get into the cochlea and the membranous labyrinth, and there's the round window.

And you're only supposed to have two windows. But what happens is in cases that I'll go through a little bit, you can develop or, you know, be born with a third window. So now you got three holes instead of two. And that changes the dynamics of the, you know, the vestibular system and the auditory system. And we'll go a little bit through that in how we identify that.

So the symptoms that we get, again, because you have this third window, you've now changed the dynamics of both the hearing mechanism, hearing system and the vestibular system is with hearing, you have autophagy, you have a certain type of hearing loss, which is what some refer to as a pseudo conductive hearing loss. So you have very low bone conduction thresholds and you have elevated air conduction thresholds and not going to go real deep into the background in that other than to say that you bleed off some of the energy. Energy when you have a third window. Now some of the energy is, all of it is supposed to go down the cochlea and the round window is the outlet. If you have a third window in the vestibular system.

Now some of that auditory energy is shunted into the vestibular system where one, it affects the vestibular system and you have symptoms, but it also bleeds off some of that air conduction energy and drops your air conduction thresholds. So again, this third window alters both hearing and balance. They complain of ear pressure and fullness. They have a sensitivity to loud sounds. Right.

And so then when we look at the vestibular, we have sound induced dizziness. Again, there is now a hole in the vestibular system. So as I said, you know, when you stimulate the ear, all the energy is supposed to go down the cochlea because of the round window. But now you've got a third one in the vestibular system. So now energy goes up into there.

It should not, it's completely enclosed. You shouldn't, you know, this system is designed to not be affected by auditory or, you know, sound. But now what happens if sound is allowed to, you know, bleed off into the vestibular system? Now when, when sound comes up, it moves the vestibular system, the organs, the otoliths, the canals and they will get dizzy, pressure induced. Again, if you put pressure in, you're sort of, you know, doing the same thing you're doing with sound.

They complain of dizziness, sometimes vertigo. They will have imbalance and have oscillopsia where the world moves. Because if you're sitting there and you put pressure in and you stimulate the

vestibular system, the world will move and they will complain of what we call oscillopsia. So again, the point being, by adding this third window in this diagnosis in the vestibular system, you now have hearing and vestibular symptoms that co occur not to go real deep into it. You know, people go, why do you have it?

Where does it come from? And when you look at a lot of the data, what happens is people will usually present with it after some sort of accident. And so they will, you know, like a car accident, they'll have some sort of trauma to the head, a mild tbi. All sudden symptoms will start to arise. What we've, I'll Go through a little bit, is that children, often people are born with this.

We. I had a whole series when I was at Vanderbilt of pediatric CTs of kids that had just come in and they happened to identify a superior canal dehiscence. Wasn't really main complaints for the children, but it was just seen now that radiologists knew what to look for. I think one of the coolest emerging sort of ideas, and I'm going to just briefly talk about it, is chronic elevated intracranial pressure. And that has been shown to contribute to the development of superior canal dehiscence.

The idea or the mechanism behind this is that the superior semicircular canal is located near the cranial cavity. It's separated but from the brain and cerebral spinal fluid by this thin layer of bone. Keeps your brain. They're very close. It's called the otic capsule.

And chronic elevated intracranial pressure will continuously exert this mechanical pressure on the bony over covering of that semicircular canal. So that little bony shelf between the superior semicircular canal and the brain. And what will happen is it will actually thin over time. It will say remodel and it can actually create or wear away whole. And so you will often see patients that have been said to have, you know, a thinning of the.

Of it a near dehiscence, or in fact, a true dehiscence, which actually shows a hole in that little bony layer between the brain and the superior canal. And so, you know, that's. That's one sort of emerging theory of why this occurs over time. Right? So people don't have this very young, but over time people will start to develop symptoms.

And it may just be the effect of time and this intracranial pressure from high blood pressure. When the bony capsule overlying a superior semicircular canal is disrupted, as I talked about, what happens is the energy that is going down the cochlea is shunted off into the vestibular system. Now it's like a little chimney. Smoke would go up both, right? And Tullio, who we've all, you know, Tulio syndrome, where you hear a sound and you get dizzy.

What he would do is fenestrate little pigeons. And I've got a picture of him doing it in a rabbit. And what would happen is he. When he would put these third windows in, he would show that he could get in different vestibule in semicircular canals. He could.

When he played a little flute, he had this little pipe. You look at it looks like he's actually smoking the rabbit. But he's actually playing a little flute. And you would play a little flute or the little pipe. And what would happen is he could actually generate different eye movements in different planes based on where he put the hole in which canal.

And so he was sort of the one who sort of described. If you add this third window in, or you fenestrate the vestibular system, you can get the vestibular system to react to sound.

The incidence of superior canal dehiscence, I don't know that we have a true measure of this. What happens is not everybody gets a CT when you're born, so we really don't know exactly what it is. But, you know, we have a 0.5% have a complete dehiscence, is what has been speculated based on. On the research that's been done over time. You can see that there's been, you know, the median age is

40, which is, you know, has some bearing because why do symptoms all of a sudden rise at 40?

Men are equal to women and the right seems to be a little more than the left. Again, you can find anything with statistics. So I will let Dr. Cunningham run the poll at the poll is. Are you.

Oh, I can do the poll, I guess, since she's not running. Are you familiar with the signs and symptoms of sccd? And you can answer here.

Hope you are now, because we just went through them and somebody says no, that's going to be weird.

Yeah, and that's really cool. And I think what's sort of compelling here to talk about is we've got 52% said yes, 7% said no and said yes. But maybe not in children, and I don't know that, you know, maybe it is more in children. Maybe we aren't asking the right questions. Maybe it's hard for them to describe, you know, maybe they don't have the language, you know, that enables them to exactly describe.

Maybe they say, I say feel weird when I hear sound. What you're going to see when Dr. Cunningham goes through Piper's results, very clear and very, very, you know, dead on in terms of what we would expect.

So superior canal symptoms here, this is, you know, some of the key things that we can ask in a case history to get at that when I cough, I get dizzy, I get dizzy when I sneeze. Both of those are increases in intracranial pressure. Loud sounds make me dizzy. Of course, the tulios. And you can see here blurry vision, like the oscillopsia, any of those things sort of increasing intracranial pressure and giving you the perception of movement or dizziness.

So what are the symptoms when we look at people that have superior canal dehiscence? Vestibular signs. 95% with a true dehiscence actually have it. Sound evoked eye movements 82%. So you put in a loud sound and you can actually get it.

I had a patient that could hum and the eyes would move. I just have them hum. We record them under video, nice egmography goggles and just humming would give them oscillopsy and you could actually see the eyes move. Valsalva again, sort of trying to clear your ears and increasing pressure trago pressure sort of bearing down will create it. Valsalva's little more sensitive than the trigle pressure.

And then like I say, sound induced head movement. That is a key. Bekochy really showed that is that when you would do this in humans and you put a loud sound on you, actually the head would tilt to one side or the other. So there are clear vestibular signs that Tulio sort of noted in those animals. Early on.

I just bring this up because when I was at Mayo Clinic and now here at Michigan, we had a very formal superior canal dehiscence protocol that we would do if it was suspected. So if you answered some of the questions early on and it looked like you had superior canal dehiscence, sort of the questions I showed you before tulio was a problem or such, then we would go through and we would do this evaluation here, primarily focusing on vemps and recording eye movements with sound and pressure changes to the ear.

So superior canal dehiscence in children has been reported ad nauseam.

You're starting to see more and more of it now. Here's a couple articles I just gave in terms of reports. So pediatric semicircular canal radiologic histologic prevalence in clinical correlation Semicircular canal dehiscence in the pediatric population. Again, having. You know, I think, like I say we don't.

Not everybody is given a ct. And so we don't know exactly how many people have it, but I suspect that more people have it than we think. I was literally with a physical therapist that teaches. I teach a course with and been teaching with her for like, you know, 20 years. And she goes, I think I have superior canal to hiss.

And so what was kind of interesting is we did all the tests with her sitting and none of them were positive. But when we had her lay down and did them when she was laying supine, all the tests were positive. And so what happens is the brain was actually sitting and sealing up the little dehiscence. And when you lay down, the brain came off the hole and all the tests became positive. We actually published that paper.

But, you know, so there are different ways to try to get at, you know, are you symptomatic or are you not? And you know, if you're in clinic, it's good to be familiar with those. So the incidence in children, it's rare, but I don't know how rare. Guess we don't know. There's limited research in it.

While some people publish it, we really don't. You know, this is not something we're really focusing on and which I think is really interesting because it, this whole case is going to be sort of about this patient's journey with two different diagnoses that affect hearing and then trying to sort out what is the most appropriate treatment.

So the use of vemps is key. Vemps is one of the best. The, the thin cut ct, as I'll talk about in a little bit, is the gold standard for identifying superior canal dehiscence. But a thin cut CT or a CT allows you to, to say that there is a dehiscence or a near dehiscence. What the CT can't tell you is what's happening functionally.

So you can have a dehiscence, like I said, like the person had a dehiscence was sitting, the brain was sitting in that hole, filling it in and didn't have any symptoms. But what the vemp can do, and we rely on

vemps, is to tell us if it's, if it's functionally active. Okay, so we can see the structural issues on a ct, but we can't really say anything about it functionally. The VEMP allow us to see it functionally. And so what, you know, the vemp becomes very handy and is if we identify it and we have low thresholds or large amplitudes, depending if you do an ocular vemp or a cervical vemp, what we can do is then, and then have a repair.

We would hope that then we can measure functionally that it is, you know, that, that it has resolved. So again, doesn't tell us a lot about that.

Again, we sort of went through that the VNG is typically normal. The VEMPs are typically what are abnormal in these cases. This is just a busy slide. It's a wonderful study though, that took 13 children with superior canal dehiscence and did all the head impulse testing, looked at all the canals, looked at ocular motor, looked at vemps. And so it's these sorts of studies that we just sort of illustrating that we need to do to say what is different in them and what is not than perhaps adults.

And lastly, just to finish up a little bit here, these are the typical radiological protocols that you will do to see a superior canal dehiscence. These are poshol or stenvers. You can. I don't know if you can see my cursor, but what you can do is you're looking to see is the superior canal completely encased in bone. And you can use these two different views.

Again, it is a thin cut, so they do very fine cuts. It's like slicing cheese. And so in order not to miss it, what neurotologists or physicians will typically order is a thin cut where it is really slicing cheese really thin so that you can actually determine whether a canal is dehiscent or not.

So I will log off and are not log off, but just. And that's it. And you're all set. It's all you. So I'll let you go from here to Rebecca.

Hello, I'm back and I'm going to tell you about Piper. I kind of want to summarize what Dr. McCaslin had said about doing vemps in children. While it can absolutely be a gold standard and we use it a lot for adult diagnostics for sscd, I think it has limited value in children. And I'm going to prove that point as we go through Piper's story.

I have complete permission from her mother to share photos and stories about this. Matter of fact, Piper's excited that she's kind of famous in the whole world of audiology. Piper is getting ready to turn. I actually think she's going to be 13 now. She is so sweet, so funny and very, very intelligent.

She plays a flute, she's very strong academics. She does have a lot of anxiety. She does take medication for her anxiety and she has since she was much younger. She also has celiac disease, which is very interesting and I think there could be more research to be done on that in the correlation. So let's talk about Piper and her journey here.

So Piper passed her newborn hearing screening in both ears. She was identified at the age of six with a unilateral low frequency mild to moderate hearing loss with a slight conductive component. When this happened, the reason that she was brought in in the first place is because she kept telling her parents, even as a six year old, you know what? Huh? What?

You know. So she was taken to an ear, nose and throat clinic not one that particularly specializes in children. And placed on steroids immediately after this hearing test. So one week after the steroids, her hearing was actually poorer. It got worse, and then she came back three weeks later and it had somewhat stabilized at what you were looking at.

Upon her first identification, they recommended a recheck in six to 12 months and said, well, I guess Piper was born with a congenital hearing loss and recommended a hearing aid for that side.

So I want to know from you guys, after the identification of a hearing loss in an infant or a young child, would you like to see done by hopefully a pediatric ENT or maybe a pediatrician or primary care physician, what would you like to see done? So all of these that are listed here are typical for pediatric ENTs to order or do on newly identified children or infants. But let's see what you guys would like.

Awesome. Okay. Referral to pediatric ENT at 25% and all of these at 75%. I agree. I believe that it should be all of these.

The problem comes in to finding pediatric ENTs who specialize in kiddos and newly diagnosed hearing loss. So obviously that doesn't always happen, and it did not happen with Piper.

So immediately after her identification at six years of age, mom said that the ENT had ordered an MRI to, quote, quote, rule out a tumor. And this came back clear. There was no blood work completed on her. Mom did ask about doing some genetic testing because Piper had some other issues with her celiac disease and now a newly identified hearing loss. And the ENT informed her that no, because that really would not change anything that they were going to do.

They were going to refer her for a hearing aid and then they did so at an outside facility because the practice where mother took her really did not have a pediatric specialist to fit the hearing aid. So she was fit with a Phonak Sky. She was immediately connected with a Roger touchscreen mic for all her classes, in all her specials, and in all of her therapies. As of the time of this presentation recording, which is about 10, nine months ago, Piper was becoming very uncomfortable using the touchscreen mic. Again, we're talking about a 12 year old, right?

Not atypical for children that age to start saying, this is really, you know, calling attention to me. And I actually coincidentally ran into Piper and her mother a couple of months ago and mom informed me that Piper does not want to use the touchscreen mic at all now and barely wants to use the hearing aid.

So we're going to go through some of these audiograms sequentially here. So in May of 2019, which was eight months after her identification, overall, that right ear had dropped a little bit. And then again in 2020, no significant changes. Now remember at this time she's wearing the hearing aid. She did have an auditory processing evaluation, which in and of itself I think is a little bit questionable for a child who has a sensory hearing loss.

But she did have one. And those processing skills came back to be within normal limits for her age. But she definitely had some deficits in her memory. And then in 21, slightly improved 510 decibels. And then in the fall of 22 it was stable.

So we're still having that mild to moderate low frequency rising to typical hearing in that ear. Her hearing loss has really always been sensory. Real slight conductive component at some frequencies, but definitely not a big, big air bone gap.

So because mom was concerned about some of these things that are going on with Piper by this time. Piper is on medication for anxiety. She has been diagnosed with celiac disease. She has been diagnosed with a unilateral hearing loss of unknown origin. And she kind of socially struggled.

Mom took her for a multidisciplinary evaluation. So obviously Piper uses spoken English sometimes. She was very difficult to understand due to being very quiet. And that is still the case with Piper. Piper struggles with some self image issues and she can be super, super quiet and difficult to hear sometime.

She did have typical language skills, but she did have a little bit below average understanding of some things that had like multiple meanings and complex grammar. There were a lot of concerns from the multidisciplinary team about Piper's increased anxiety across environments. She, she had those throughout the evaluation. She has those for school. They were done by teachers, they were done by

parents and family.

Elevated ratings for anxiety and panic system symptoms. So Kiddo has really been struggling with some of that mental. I do actually think that's getting a little bit better her mom's report. But she still is on medication for anxiety. Elevated scores indicated more concerns than are typical for her learning problems and inattention when she was compared to same age peers.

Now again, that's typical hearing, same age peers, less developed executive functioning skills. And we the team gave them a lot of study strategies to try to help her and more resources in how to using strategies for understanding in the classroom or how to self advocate. Because Piper had very little of that mental health supports, I think it was recommended that she go back to see about medication adjustment for her anxiety and her panic, as well as some counseling and then try to Teach Piper a little bit more about her own hearing and how it can fluctuate again. That all feeds into those self advocacy skills that Piper just did not have. So she did get an iep.

This was one year after she was identified. She still has an iep. She gets, you know, preferential seating or extended time on tests. All of those things that a lot of kiddos who are DHH get. At the time, she was identified with three eligibility criteria of DHH ohi, which is other health impairment, taking into account her anxiety and her celiac disease.

And she also had a speech impairment, which is not unexpected for having a hearing loss even in one ear. So I started seeing Piper in the fall of 22. I saw her for several months before she came out to me and said very quietly, of course, that I get dizzy when I go to the cafeteria. Okay? I wrote it down in our log database.

In March of 23, I asked her to try it again. Go back into the cafeteria, but don't wear your hearing aid this time. And it still happened. I offered her some earplugs. She tried that and it still happened.

So it came to the point where Piper would not eat in the cafeteria any longer. She ate in the principal's office in a little side room because it just made her too dizzy. She did tell her nurse and she did tell her teacher that that is what was happening. And it never went any further than that report. When she told me, I of course had immediate red flags, right.

I reached out to the parent and recommended that she be taken back to, I said, an ear, nose and throat physician. Not necessarily the one that she had gone to when she was identified, but they did go back there. And that audiogram revealed a significant decrease in her hearing compared to the year before. So at that time, mom insisted again that we get genetics and balance testing because Piper's getting dizzy with loud sounds. So the question to Piper is, and I asked her, how long has this been going on?

And she had a really difficult time explaining that to me. But the best that I can tell, it had been probably close to a year that this had been going on.

Okay, so In March of 23, you can see her hearing has dropped down into flat, moderate in the low pitches, rising to normal. And then even six days later, we're down into the severe range. So we did have a significant drop. And I had encouraged mom to take her back to the ENT Hearing aid adjustments were made and you can see that left ear stays stable. But In March of 23, she's dropped all the way down into the severe range in that right ear.

But interestingly enough, her hearing loss is still mostly sensory and not conductive. So I'm going to let Dr. McCaslin talk about some of this balance consult and the testing that occurred. Yeah. So just to go through here, we'll go through a little bit of the results that they ordered.

And again, pay attention to that hearing loss because I think that's very interesting. When you look at the overview, they simply coded it as dizziness and asymmetrical hearing loss of the right ear. You can

see here that they did partial views for the superior canal dehiscence. And they also looked at ordering a vemp so completely in line with what the workup would be for a child presenting with this.

When we look at the cervical vemp now, we have two different types of vemps. We have the cervical vemp and we have the ocular vemp. This vemp, which I think is interesting, is normal. Okay. It does not have a reduced threshold.

You can see they ran it at a lower threshold at the bottom. I believe that says 70, which is typical. But the vemps are symmetrical. The latencies are right, but doesn't have a reduced threshold so much. This is a cervical vemp, evaluates the saccule and the sternum, you know, the afferent limbs that go to the sternocleidomastoid muscle.

So it's the vestibulocolic reflex. They didn't do an ocular vemp, which is the utricle, and it's actually an otolith ocular reflex. They didn't measure that. We look at amplitude. Now we usually look at threshold in the C vemp, and again, it's normal in that respect.

The vng, which typically in canal third window disorders are typically considered normal, usually seen to be normal of V and G. Again, just an overview is a collection of tests. It's the caloric test, it's the ocular motor test, it is the positional test. Looking for BPPV. V&G; is a gestalt of tests, but when you look, you can see that the BI thermal caloric irrigations produce robust and symmetrical nystagmus.

And so the caloric exam insight, the VNG was normal. VOR suppression, which is a evaluation of the midline cerebellum and its effect on the vestibular system was completely normal as well.

So the assessment after doing the CTS, after doing the VNGs, after doing the vamps, it's interesting to, you know, sort of see the thought process and how are we going to approach this? So you can see. And it's highlighted in yellow. She has been told she may have Meniere's. And Meniere's disease is not

really a disease.

And you know, it is a collection of symptoms that refers to a homeostatic failure of the ear. So whether your ear is failing because of a hydropic situation, such as you see in Meniere's patients that are like 50s and 60s, or if it's failing for a genetic reason, if it's failing for a vascular reason, if it's failing for a viral reason, what happens is you get the same for symptoms that classify you as Meniere's. Okay, so Meniere's disease is not a disease. They symptom, like I say, a collection of symptoms that the ear is failing and she looks like she has Meniere's and she would fit the criterion if you applied it to her. But it's clear that the ear, the organ of hearing, is failing for some reason, of which there are many reasons, we can speculate on which one.

But it is not a hearing loss that you would expect to go along with superior canal dehiscence. Superior canal dehiscence, as I said, gives you ultra low bone conduction thresholds and mild to moderate loss in the air conduction thresholds, but does not reflect what we see in the loss of function in low frequencies, especially in that child. So again, there are two things going on here. There is the third window identified, as you can see, and the CT scan. I think I'm going to get the radiology which will be abnormal, but also the failure of the end organ of hearing.

So after the normal balance assessment, the mother came back to ent. We were going to get blood work and we were going to get imaging, right? So that temporal bone protocol that will use a thin cut, 0.4 millimeters, 0.5 millimeter radiological test. And what you can see here is that is it has been cut out. There appears to be dehiscence and or thinning sometimes.

You can't always tell of the superior right semicircular canal. So when you're looking, yes, that ear has now been identified as dehiscent, that right earth. But when you look at the hearing loss, that doesn't really look at like what you would expect in a disahison ear. And it may be again, two things going on at the same time, a failure of the ear and the dehiscent ear as well. So then they went on to blood work

and I will hand it off to Dr.

Cunningham.

I wanted to point out the note that came in from the balance assessment physician who said that she had offered to mom to do, to do imaging of the iac, but because of the chance that it really was superior canal dehiscence, she did not think that they really should go ahead and do that. And she wrote that mom agreed with that. And that's not exactly the way the mother reports it back to us. Now, I know there's always stories like this, but mom did go back to the ENT that they had been seeing for over four years, nearly five years, and say, I do want imaging and I do want blood work. And lo and behold, Piper has Connexin 26.

Now, if this had been done when Piper was first identified in, you know, five years ago, I do think that that might have changed the way that they would have managed the case, which is what the physician had said would not. He did not even offer at that time to do any kind of genetic testing or any kind of blood work. So she does have connexin 26. She does have Right superior canal dehiscence. And I also wanted to throw in here that after the balance testing, mom decided she did not want to have Piper's care any longer at that original ENT office and went over to Cincinnati Children's with renowned physician Dr.

Chu for pediatric ENT. And that's where the rest of the story comes is from Cincinnati Children's. So they get this back and mom gives me a call and she's like, I don't even know what this means. So now we have two more diagnoses all coming in the same week for this little girl. As you can imagine, it was pretty chaotic.

So here's what we know about Connexin 26. The GJB2 variants tend to be associated with bilateral hearing loss, right? They tend to be right. 89% of children in most of the studies, even higher,

sometimes no severity difference between ears. More than 90% had it bilateral in another study.

But really, in order for us to know what the prevalence of hearing loss due to Connexin 26 is in children, there needs to be more research. Has it been documented as being unilateral? Yes, but not very often.

So they take mom takes everything over to Cincinnati Children's, and I am in contact with Dr. Chu and the family, of course, and they see Dr. Chu after a very brief waiting time, I will say. And these are some of his notes. Right sided superior canal dehiscence syndrome.

And I mentioned it to her family. That is exceedingly unusual. To see this in the pediatric population. And even when we do see this, it is rare that it causes symptoms. This is from Dr.

Chu. So he went over the intervention options with mom. He was very thorough about this superior canal remodeling, resurfacing, which be done through a middle cranial fossa approach. Also talked about doing a round window patch in order to try to address the third window effect that she is seeing, that dizziness in the cafeteria. So he went over all these options with her.

Well, as you can tell from this picture, Piper was talked to her mom and her family discussed it with her. And she said, I hate the way that I feel. Let's do the surgery. And so they did. So this is Piper post op, six months post op.

And you can see she's got quite the scar there. She has very long hair now, so it is absolutely covered up, but that is quite the scar. Right. And interestingly enough, she was only in the hospital for a couple of days. That is a reflection of our medical system.

And she did very well. And she went home. And now I have some more questions. Right. Why does Piper have a hearing loss?

Right. We know absolutely without a doubt that superior canal dehiscence can cause hearing problems. We know that. Right. Dr.

McCasin went over all of these things. Connexa 26 can cause a hearing loss? Absolutely. Why? More commonly bilateral.

It absolutely can be unilateral. Okay, so I'm going to ask you guys, why do you think Piper has a hearing loss?

Just going to give you a couple minutes here. Not even a couple minutes. Maybe 30 more seconds. Any thoughts you have about why you think Piper has the hearing loss, throw them in there.

Getting some good ones here. I got connexin 26. I got connexin 26 dot I got combination connects in 26. Inner ear disease connects in 26. Connection 26 seems to be the winner.

Ding, ding, ding. So if that. Go ahead, keep putting it in here. I love seeing these 26 secondary do connects in 26 and dehiscence. Yep, we got a combo.

Combo. Yep. I love these. I think. I don't know.

Right? I think. I think that. I don't know. Is it connexin and the superior canal dehiscence is an incidental finding.

Is it a combination? It is an unusual configuration of hearing loss for somebody with superior canal dehiscence. But it is also atypical for somebody with connects in 26. So let's move on here for a question. I will get to all of these Q and A's at the end Here.

Okay. What do you think? Is she better? I think I kind of told you. Besides not wanting to wear her hearing aid and her touchscreen mic, how do you think.

How do you think she's doing? She did tolerate it for a while after this. This was less than a year ago. But as I said, I just coincidentally ran into them recently, and she's done. She doesn't want to do it any longer.

So how do you think she is? So let's get some answers here.

Oh, yeah, she's definitely better. 89% of you said that. Or that she's the same. She doesn't get dizzy anymore. I will say that.

So the patch that Dr. Chew put on there worked amazingly. Right? The hearing loss is still there. It still fluctuates.

It still goes up and down some. Socially, I don't think she's come into her own yet, but she is obviously aware of all of the social stigma related to being different from her peers. And even though she can wear the hearing aid and have it covered up, she's in a very small school district, and everybody knows everybody, and everybody knows that Piper has a hearing loss on one side. But as far as her vestibular symptoms, she is absolutely 100% better and just a delightful young lady. And it has been my experience that preteen middle teens typically stop wanting to be different from their peers, but can come back to their amplification or their remote microphones in late high school, early college.

And I expect that will happen, too. Okay, so this case really, really highlights the need for this interdisciplinary collaboration. So it needed to be other professionals. We needed to have audiologists. We needed to have pediatric audiologists.

We needed to have a pediatric entire. We needed to work with a pediatric surgeon and post op, post therapy, somebody in her school to keep track of her to see how she's actually doing functionally with that hearing loss. And now that, you know, the semicircular canal dehiscence is fixed to keep an eye out in case anything happens, God forbid. Again, so inter. Inter collaboration is of the utmost importance.

And I think that Piper's story could have been a little bit different if that had all happened from the beginning. She did have the tulio effect. All of her balance testing was normal, including her cvemp. Why were neither of these suggested to the family? If either one of them is the cause of her hearing loss or the combination of.

And then this stresses to me the need for education for ENTs and the need for pediatric ENTs and the need to educate other physicians about Semicircular canal dehiscence. And. Or the genetic testing for Connexin 26, because I don't think it's happening enough with newly identified infants and young children.

Okay, so what do you think? Do you think it's a good option for children? You don't have to take into Piper's case into consideration because it's obviously massive. Right? It is brain surgery.

They're opening up the skull. But what do you think? I think that if Piper didn't have Connexin 26, this would be a much easier question to answer.

All right, let's see what you say.

All right, we've got no at 5%. Yes at 74%. And I have some thoughts on 21%. I agree. I think it would have been a much clearer cut and dry case if Piper didn't have Connexin 26.

Kind of confounding the results of this. And there are other options for treating the dehiscence, but her repair surgery was very successful. All right, I'm going to go to the Q and A. It looks like we have some things in here. All right, let me go all the way back here.

What blood work is suggested for a child similar to Piper with a sensorineural hearing loss? All right. Should it be determined through genetic testing? Pediatricians could of course order this. It is a full genetic workup specifically targeting the genes.

And there are hundreds of genes that can cause hearing loss. So that is what would be recommended. I could not tell you personally, I'm not a geneticist or a pediatric entire, but absolutely, those kinds of blood work and genetic workups should be done on any child who is newly identified. I would just add too, for blood work, maybe look at infectious diseases, autoimmune screenings. Right.

You know, torch, cmv, those sorts of things. Maybe metabolic, endocrine disorders. So, yeah.

Next one. Since the GJB2 only showed her to be a carrier. Not home. Oh, somebody else answered this question too.

She actually did get more in depth testing that I didn't put in there that has. That she does have two copies of the gene. So it is possible that she has the hearing loss to that that's not shown. So good catch there. The GJB2 only said she was a carrier, but yeah, she does have two copies of the genes.

Yep. Good. How extensive was the genetic testing? I think I just answered that one too. I don't think I put everything on here because I'm not sure that I got everything.

I got the preliminary results, but I know mom and I know that she went after and had more. Had more done. And that's how we confirmed that she did indeed have Connexin 26, current audiogram. If there

had been a difference in her hearing, I know that mom would have texted me, so I'm going to assume that. So it would have been, what, a year and a half ago was the last one that I had access to.

If there was a difference, I would have known. I do know that she did have her hearing tested. I just don't have those results. But there is no way that if there had been a change, mom would not have reached out to me. So currently, I'm going to assume she still has the moderate low rising into the normals.

Yep. Thinning with the. Okay, if the CT showed thinning at this time, would the expectation for the future be that the bone continues to get thinner or a complete haul? That's you, Devin. Yeah, I don't think that anybody knows for sure.

I think based on what we do know is that that brain rubs against that little shelf, and if it's already thinned, then, yes, you know, you could expect it to wear through again. If you remember what I said is most patients will come in and report symptoms after some sort of head trauma. Whether, like I say, car accidents are a big one. I think when Hopkins went through, it was like a huge percentage of the patients that had. That were in their database symptoms began to arise after an accident.

So I would predict that, you know, this child may not have symptoms until something like that happened. Or again, like I talked about, if, you know, develops, you know, high intracranial pressure for obesity, whatever reason, then that could accelerate it, too. But I expect it to get worse. But again, you don't know, right. Until something happens.

That's a good point. And somebody asked, has she had a workup for autoimmune disorders? Yeah, that's how they identified the celiac disease. But I don't think anything recent. Nothing recent.

Is there a known percentage of the likelihood that the patch will fail?

There is not a known percentage. So where the variability comes in is the approach that's taken, of which there are three for superior canal dehiscence. Actually, there's a fourth where you can actually patch the round window. We won't go into that. That what I will say is the percentage.

The biggest variability in the percentage, in my experience, is the skill of the surgeon. And so, you know, it's not a percentage of, you know, some are going to have higher percentages than others, and some are going to be you know, and the percentages vary on whether you, you know, resurface it, put a little patch over it, or you plug it. So some people, in order to get rid of these symptoms of the superior canal, what happens is the patches seem to fail over time. So some surgeons will decide to just plug the hole. They drill two little holes in it on each side, and they just fill it with bone paste and just take it out of the equation.

The vestibular system compensates, and they never have to worry it again. If you want to keep that canal intact, then what you do is you resurface it and you hope that it doesn't fail. So it's hard to put it in percentages based, you know, based on the technique and the skill of the surgeon. Okay, one more. Somebody's reporting here that a 2010 study, and I do remember this because I'm old, that up to 10% of babies with the GJB2 gene passed their newborn hearing screening, suggesting there could be a.

A progressive component. I have a couple answers to this is Piper's has not progressed. Besides that one dip that she had, it has remained relatively stable. And because of the variations in the genetics of GJB2, I full fully believe that there are some variations of that that will be progressive. I don't think we know it all yet.

I don't think we know it all yet. So that's my thoughts.

She never had an OVANT prior to surgery. She did not. Not to my knowledge. I don't have that information from mom. Okay, guys, I'm sorry.

We're gonna have to wrap this up. This is awesome. Thank you so much. I think Chrissy's gonna come back on with some helpful information here. Yes, thank you.

Dr. Cunningham, can you advance to that very last slide for us? Absolutely. This is just a reminder for all of the members who wish to earn CE credit just to complete the multiple choice exam. We'd like to take this time to thank you, the Academy, and also Dr.

McCasman and Dr. Cunningham for their time and their expertise on this topic. We hope you've enjoyed this presentation. Tune in again. Thank you so much, everyone.