Case Studies in Pediatric Audiology: Wins Worth Sharing, in partnership with Phoenix Children's Hospital

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Thank you for joining today's webinar called Case Studies in Pediatric Winsworth Sharing, which is presented in partnership with the Phoenix Children's Hospital. It's my pleasure to introduce our presenters, Dr. Tanner Robinson, Dr. Robert Fanning, Dr. Alyssa Nickerson, Dr. Ashley Geske, Dr. Ali Sayer, and Dr. Wendy Sterwald. You can find information on each of our presenters on the course page on our website. So welcome, Dr. Sterwald and I will turn the floor over to you. Thank you, Christy.

We are so excited to be presenting for Audiology online today. And just a couple other housekeeping things. All of our disclosures are here on this slide. The limitations and risks of the course and our learning outcomes. I'm not going to go over all of that, but you all can look at it if you want to.

So, as you know, we are all from Phoenix Children's Hospital, and here is a picture of our beautiful hospital out here in Phoenix. We have sunshine and palm trees and it's just a lovely place to be live and work. So a couple things that you may not know about Phoenix Children's is that Phoenix Children's Hospital is the only freestanding children's hospital in the state of Arizona. And our audiology team is comprised of 16 audiologists. We also have a newborn hearing screening side which has 24 newborn hearing screeners.

And we provide newborn hearing screening services at six area birthing hospitals. Along with we see, we, we provide the screens in the NICU and cbicu. Here at Phoenix Children's, we see complicated patients from around the Southwest and international patients. And you're going to hear five thought provoking case studies today. And I want you all to pay attention because you're going to learn a lot.

As you know, the name of our presentation is Winsworth Sharing because these really are wins. They're wins for the patient, they're wins for the audiology staff because we always learn so much from everything we do with the patients and the patients get the value of our audiology expertise along with the expertise of all our collaborative partners here at Phoenix Children's. So you are going to learn a lot today. So I just want to give you just a little bit more information about what you're going to hear today.

The first presentation is by Dr. Tanner Robertson, and Tanner will guide us on how to effectively counsel patients, especially in challenging situations.

Several of us have likely faced moments where we struggle to find the right words or approach when communicating unexpected or sensitive results to a family. Dr. Robinson will share strategies for delivering information in a way that is both collaborative and meaningful and helping to foster trust and understanding in these important conversations. Next, we're going to hear from Dr. Bob Fanning. Bob is our expert on ABRS, otherwise known as Bayer, and the go to resource for any related question. One of the common challenges in Bayer studies is determining when masking is necessary.

Dr. Fanning will explain how to interpret ipsilateral and contralateral recordings to identify signs of crossover and decide when masking should be applied. The Next presenter is Dr. Alyssa Nickerson who will highlight the value of using genetic information as a tool for understanding hearing loss. Genetic evaluations can help determine whether specific genes associated with hearing loss are present. One Such gene is GJB2, which is known to contribute to certain types of hearing loss. Dr. Nickerson will discuss key GJB2 variants and explain how they can influence audiological findings.

Next we have Dr. Ashley Geske who will emphasize the importance of open mindedness and collaboration in reaching an accurate diagnosis. Achieving the correct diagnosis can take time and often requires input from multiple professionals. Dr. Geske will walk us through a real world case highlighting the various assessments and teamwork involved in identifying the patient's condition. Our final presenter is Dr. Ali Sayre who will help us deepen our understanding of microtia and oral atresia and how these conditions impact hearing. Evaluating patients with mixed hearing loss due to microtia and atresia can be complex, especially for less experienced audiologists.

Challenges may include determining how to assess hearing accurately and knowing when masking is or isn't appropriate. Dr. Sayer will present a case study that illustrates these key components and

explores various treatment options. You all are in for a treat today, so first up is Dr. Tanner Robinson. All right, thank you Wendy so much. We are excited to share these cases and be together with you today.

Like Wendy said, Dr. Steuerwald said, I'm going to be talking about uncomfortable counseling moments as part of my case today. My hope is that we can review a case that involves an uncomfortable counseling situation. Then we can explore some potential uncomfortable counseling moments that lots of people can relate to, lots of us as audiologists can relate to, and lastly, review some counseling skills that may help in these difficult situations. My case today is going to be about Trevor. So I saw Trevor first when he was about two months old.

He was born at 37 weeks to a pregnancy complicated by maternal hypertension. He was not in the nicu. He did not have any jaundice or any risk factors otherwise. Of note, the maternal side grandfather has unilateral hearing loss from childhood, reportedly due to ear infections. He referred on his AABR newborn hearing screening once for each ear as inpatient and then when they went and came back twice as outpatient.

When I saw Trevor, he had already had a diagnostic test at another facility. I'll use the word bear or brainstem, auditory evoked response. I'll use that interchangeably as an ABR testing throughout this presentation. This was completed five weeks prior to my visit of seeing him. Per that report, he had a normal 1000 Hz tympanometry, absent OAEs for each ear.

And then based on the ABR responses, they diagnosed a mildly elevated thresholds in the right ear and then normal thresholds for the left ear. They completed bone conduction for the right ear and results suggested a conductive component. Ultimately, the family told me that they were referred to me by the pediatrician for repeat testing. They sent a referral both back to the original facility and to Phoenix Children's and we had earlier availability so they were able to come and see me. Based on this

family's report, they were told that hearing was normal for both of his ears.

Now, I don't know what the conversation was, but that was the family's interpretation of what had happened. It's important that their pediatrician visit led to an additional referral. They had talked to their pediatrician, maybe had some concerns because of his responses or lack of responses. And that's kind of what led to my visit with them. They reported that he had really good eye contact, followed and could track where they were going.

But he didn't seem to startle to loud sounds like a vacuum or door slam. Also of note, the family lives with grandparents and aunts and uncles and things. And so there were some differing opinions about what his responsiveness was like. These were my results today or on that date. As you can see, peaked tympanometry for each of his ears, even if it's a little noisy, not my best work, but still looks pretty good for peaked.

His OAES were absent in each of his ears. And then I picked one wave example. So this was at 100 dB. This is a very different waveform than what I would expect for a normal hearing or near normal child. And there was no response.

This one was particularly for a broadband chirp and this is an NHL. He slept pretty well for me, so I was able to get a lot of really good information. But I got a no response audiogram at the output limits of the machine. I also was able to get bone conduction which had no response at 50 decibels. EHL, which is very different from what the family was kind of expecting or what the other report had said.

So this is an uncomfortable situation. There's a couple of reasons why it's uncomfortable. For one, it's uncomfortable because I'm getting different tests for results than what somebody else got, particularly because it was at a different facility. And I do not want to provider or facility bash another place. I'm also nervous about how the family is going to react.

Getting the results from a normal hearing to a profound hearing loss in each of our ears is a pretty big difference. Can be very devastating for some families, and that makes me nervous or a little uncomfortable. I think it's important for all of us as providers to recognize when we are in these uncomfortable situations, how we feel and how that may impact our counseling skills, how that may impact the family, the family, maybe even our confidence in results or how we go about things. So my hope is that as we talk about some of these next skills and about the situation is that you can one realize and recognize I'm uncomfortable, know that that's okay, and then hopefully have some tools to kind of work through those scenarios. So as I'm going over these results with Trevor's family, what approach do I take?

How do I, quote, unquote, break the news to this family? Initially, what I did with this family was I gave them a lot of information. I brought up the report from the previous audiologist. I brought up all the results that I had for that day. We went over what a normal ABR wave would look like versus what his were looking like that day.

It's a lot of information, and families may feel like it is an information overload. There is research to support that. Families want that and families prefer that. There's lots of ways for you to check in, make sure that they're doing okay and that they're still kind of moving along as we need them to. But it is a great place to just start with the facts and looking at.

Here is what they reported. Here is what mine is. What are things the same? What are things different? So, like I said, reviewing a previous results and comparing to today's and their reaction, and they had a lot of really good questions.

They were very surprised, but also not surprised at the same time because of his lack of responsiveness, lots of tears. But hopefully I was able to kind of help them go through that process with

some of these next skills. So what made this an uncomfortable situation, again, is that there's different results from other providers that can happen within the Same facility, even if for different providers at Phoenix Children's or from other places out in the valley. But let's say the family may disagree with results. They may follow up with a different audiologist.

They may disagree with the things that you had found likely or luckily they stayed with us, they felt very comfortable with us and we'll get to his actual results at the very end. The other thought that I have or some other potential uncomfortable accommodate or situations is pseudo hepacusis patients that can be uncomfortable to kind of talk with families about. And I'm sure you can think of others. One of my favorites for adults is, you know, the wife has brought them in. The husband maybe doesn't necessarily agree as much with the hearing loss and doesn't really want to be there.

There's some uncomfortable counseling situations there. So we as audiologists know that counseling skills are important. It helps us through all of our patients, some more than others. Research has shown that the confidence that audiologists have in their counseling skills varies. Some audiologists feel like they do a really great job.

Other audiologists recognize that I got a little bit of work to do. Our counseling skills have a direct influence on family reactions and their adherence to care. They will be impacted by the way that we counsel them and we have a lot of power that way and we I think should recognize that we have a lot of power and control over that. Ultimately, the way that we counsel families can absolutely change the outcomes for patients and families. For better or for worse.

Poor counseling skills can lead to poor adherence. Great counseling skills can lead to wonderful adherence, whether that's hearing aid use, understanding results, whatever it may be. The last thing is, is there's obviously been a big change kind of in healthcare in general towards family and child centered care. And a lot of that comes with by using our counseling skills. So these are the counseling

tools that I'm going to go over.

One of those is narrative medicine. The next open ended questions, the pregnant pause validation and avoiding projection checking for understanding and informed shared planning. I love narrative medicine and if I can you one tip it is use narrative medicine in uncomfortable counseling situations. How this can be helpful is in a lot of different ways, but one is you get the family talking, you learn a lot of things and then you don't mess up potentially just by saying things that maybe we shouldn't or making assumptions that we shouldn't when we're able to just place the time back in the patient and let them talk. One of the things that I absolutely love about this and this definition is that narrative medicine aims to address the feeling and experiences.

It validates and bears witness to the experience of the patient while encouraging creativity and self reflection in the healthcare practitioner. It gives you an opportunity to see where is the family, where is the patient? And give you information so that you can help come up with a plan that maybe you wouldn't have had otherwise.

One of the most important parts of this, like I said, it gives you a chance to get the information you wouldn't have. An example would be tell me about your last appointment and how you felt afterwards. This was one that I used with the family so that I could really understand understand where they were coming from. That's where I got that piece of information that they thought and were told that he had normal hearing for each of his ears. That was crucial in order for me to understand and know how to navigate this appointment.

The next one that lots of us know is open ended questions. So open ended questions can help be a self check of personal bias or assessments. It helps us to avoid projections on a family or a patient. I would assume that families would be pretty sad when they get the information that their child is deaf in both of their ears. But I don't want to assume or place my personal bias or assessment onto that family.

When I ask open questions such as how are you feeling about today's appointment, today's results? It allows me to gauge where they are feeling and them to share things without me sharing what maybe my own expectations would be. It opens a conversation, it allows them to lead the conversation and what is important for them during that appointment. It's critical in both the case history during the appointment and providing results. We should always be asking open ended questions using the pregnant pause.

This can also be uncomfortable. We should be asking questions, letting the family just sit. Sometimes it's the most or it's most helpful for them to have some time to collect their thoughts and feelings before they share. It's quite easy for us to just jump in, especially in an uncomfortable situation where we want to kind of have the most control that we can to help these families. But I promise you that allowing the family just some time when it's uncomfortable can get you some good information and allows them to feel, listen, listened to and validated, leading into validating the feelings and concerns.

So we always want to validate feelings after the family of the family after we have asked. So I might have asked them how are you feeling? They give a response, response and then I want to validate those feelings and those responses to make sure that they know that I'm listening to them, that I hear their concerns, their feelings, whatever it may be. So that you can move into more of the shared planning. This is an opportunity to include human connection into health care.

These appointments are not just looking at a textbook or getting some data. We are dealing with families and children's lives. And it's important that when we that we use validation as an option for us to connect human to human as their providers, the next thing is you want to check for understanding. So this provides an opportunity to check in and just adjust our counseling as needed. We can ask those questions, such as what questions do you have about the results?

One of the best ways that you can also do this is through a questionnaire. I included one of my most favorite recent favorites that I found that's called the Spicer to really see how families are doing. And you can do that multiple times as you're going throughout the results. The last, I think the last one or second to last one is informed shared planning. So this is when you take all of the information that you've gotten from all of your other counseling skills and use it together with families.

This one was particularly important with this family because I wanted to make sure that we were on the same page and had a plan that the family was comfortable with and that I was comfortable with. For example, in this case I shared with the family based on the results that I'm seeing and how what they were seeing at home, we would recommend a repeat of testing and we were able to do that. But we want to make sure that the families on page with that that they feel comfortable with whatever our next steps are. So where is Trevor now? He had his confirmation test and he is profound in both of his ears.

He was fit with hearing aids and had essentially no reaction with his hearing aids in. And he is the path for to get cochlear implants. He had a normal MRI and I am super excited to see kind of how his journey ends. Just some closing thoughts for you is that I hope you were able to learn some different counseling skills. Use some of them while you are in some uncomfortable counseling situations that we come across fairly often.

All right. And it's my pleasure to introduce Dr. Robert Fanning who will be talking to you about some abr.

Well, thank you, Tanner. My name is Bob Fanning. I'm going to talk to you, I hope, about a case that I that is very interesting to you. Title My presentation is Left, Right or Wrong. I'm going to share with you a case where it was difficult for me to tell what was going on in this little infant.

Also, before I begin, I want to give a shout out to some people I recognize in the audience. There's Amy Magruder and Linda Norris, two women who were just so important in my early training back in many, many years. And it just occurred to me today that I measured my very first ABR test in the clinical environment 30 years ago, 1995 in the summertime. So this is, it's nice to see some familiar faces out there and I hope, I hope I do it justice. So see here, There we go.

So before I start talking about my case, I would like to review some of the basic concepts of measuring an ABR in an infant. This is an example of an unmasked bone conducted ABR measured at 4,000 hertz in a toddler with bilateral conductive hearing loss. It was recorded by an associate of mine. I didn't do this test and she used a standard electrode array, just the two channels. And really when I saw this, I said I need to use this for my presentation because it truly is picture perfect, even for the uninitiated.

If you look at this study, it is very clear what the laterality of the response is here. And let me walk you through it. First of all, again, these are all very well established key points of assessing an abr. And I do have some references here, up, up here that are going to be shared with you in a handout. But the first thing to notice here about the difference between the ipsilateral ABR and the contralateral ABR, most notably is that in the ipsilateral channel, wave 5 tends to be a little earlier and a little larger than the wave 5 measured in the contralateral channel.

And this really just has to do with the differences that it could can occur by placing electrodes in different spots on the scalp. It's nothing too remarkable. I don't want to get in the weeds on this. But hey, just on an intuitive basis, I hope you can appreciate that if you start moving electrodes around, even wave 5 can look a little different. You're dealing with dipoles and differences in latencies.

And really we can take advantages of the differences to make, make assessments and determinations as to which side is responding. So this is the first clue here that, well, this must be the ipsilateral response. Even though I'm using a bone conductance stimulus, again with audiometry, you're always

worrying, well, which ear is it? Which where am I testing? Well, here we have some clear signs.

Wave 5 is earlier and more prominent in the ipsilateral channel, I.e. the test ear, the right ear, than in the contralateral channel. Another way of putting that is I'm testing the right ear, and the ABR measuring in the ipsilateral channel looks like it belongs to the right ear.

Another thing I'll mention quickly that these differences tend to be more pronounced in infants and young children because the ABR is described as a gross response that shouldn't be a surprise to anyone. And they get the. The differences start to narrow in older children. Another thing to note here is that you'll see very clearly that there's a nice prominent wave one in the ipsilateral channel, not in the contralateral channel. It's very hard to measure wave one in the contralateral channel without stimulating that side.

And again, not to get into the weeds here, but it has to do with the proximity of the electrodes that belong to the contralateral channel in relation to the ipsilateral or test ear, auditory nerve. It's just very far away. Also, at those locations, the voltages are positive. And recall that we use common mode rejection, which can wipe out that potential. So really, just to be very basic about it, it's just very difficult to measure.

Never say never. But it's very difficult to measure wave one and a contralateral channel, which serves as a key as a landmark for us to determine which ear is responding. If I did see a wave 1 in the contralateral channel, that might mean that I'm bilaterally stimulating the ear, which can happen with bone conduction, for sure, if you're only seeing it in the contralateral channel. Suppose we saw this in the contralateral channel. Well, then I'd be worried that, all right, I might be crossing over here.

Another thing to make note of. And again, references are up here, but interval attenuation at 4,000 hertz is relatively high. And I'm. We're stimulating at 10 decibels. So the likelihood of crossover here at

10 decibels at 4,000 hertz is very, very low.

Again, never say never, but it's very low. So there is an, there's an additional confidence here. And knowing that you are testing the test ear under this circumstance, which is always, which is always a concern when we're measuring bone conduction, especially with bone conduction to determine the test ear. Here's another thing, too, that I recommend to learners and students is that when you are administering bone conduction test, go ahead and go to the other ear just so you can make some comparisons. Like, well, you know, if this is doing this, I'm seeing this in the right ear and this on the left ear.

If I move over to the other side, all things being equal, everything should flip around. And that's exactly what happens here. The right ear looks like the right ear, the left ear looks like the left ear. And so even to the uninitiated here you can, I think, you know, if you know a little bit about abr, you can see that, wow, those look really different. And the differences are really based on the locations of the electrodes in a two channel recording and intraoral attenuation.

One final note here is that this, these principles apply to any transducer. We talk about it a lot for bone conduction, but it's any transducer air conduction, you'll see these same sorts of behaviors. And again, just finally take a look at these references that will be provided to you in a handout. They're seminal articles and if you are a little hung up on these concepts, I'd really encourage you to read them. I read them when I started out and I found them to be very helpful.

So my presentation is called Left, right or Wrong? Well, here we have wrong. So what I'm doing here, this is the right ear, this is the test ear, and I'm, I'm presenting a 50 decibel click to the test ear. The test ear. The right ear.

Oh, I can't see this very well, but this is, this is a bone conduction test on a right ear with an ear that has, I'm sorry, a profound sensory neural hearing loss with features of auditory neuropathy spectrum disorder. So I finished the air conduction test. The left ear was perfectly normal. The right ear had these features of profound hearing loss loss with ansd. And so I proceeded with a bone conduction test on the right ear.

And what we're seeing here is the exact opposite of what I just showed you in the previous slide. Here I am stimulating the right ear at 50 and what do I see? Well, I see this very anemic looking wave five that's lower and later in the ipsilateral channel. Well, let's look at the contralateral channel, the non test here. Well, I see a wave one, a very nice looking wave one and a very prominent wave five early.

And what I see what's going on here is that I'm actually testing the left ear through the head on the right side of the head. All right, I've made a mistake. Although for your benefit, you're saying that, oh, okay, this is what happens when there's true crossover. You're seeing basically the ipsilateral channel is showing the contralateral response of the left earth and the contralateral response here is showing the ipsilateral response of the left ear. Okay, These are the sorts of things, these are sorts of problems that it can occur too if you mix up your electrodes when you're administering a test that the inverting electrodes, you put the right ear on the left ear and the left ear and the right ear and you're looking at it going what did I do here?

I'm getting the right ear and the left ear, same principle. It's just that in this case everything is set up well. It's just that the degree of the hearing loss in the right ear is such that when I really increase the intensity of the bone conducted signal, it's crossing over. And there's a very effective way to deal with that problem, which is to mask. So let's add some masking here.

85db SPL. And this is what we would expect to see in a right ear with profound hearing loss, with features of auditory neuropathy, empathy spectrum disorder. So again, I hope that this is very clear to

you. Again, these are the types of things that you can rely on to give you some confidence in determining, well which ear is responding here, which is nice. Again, you can't always do that with audiometry.

I mean especially when you're working with kids. Like they'll raise their hand or they'll put a peg down, but like which ear heard it and you have to be. The morphology of the response for the bear study can give you very clear indicators as to which ear is responding. So moving on here, let's jump right into now. Now that I've covered those basics, let's talk about my case.

This is a 6 month old male born at 32 weeks gestation age, admitted to our NICU. I'm not going to. He's got multiple congenital anomalies and just for the sake of time, I'm not going to go through them all here. He's very involved. The thing that is key here, he has a bilateral hearing loss.

He did not undergo a newborn hearing screening because he had a history of otologic malformations. And there was a previous ABR test done at bedside by one of my colleagues. The patient was in the natural sleep state, very fussy, not much was done. But a severe hearing loss was measured in the right ear and a Moderately severe hearing loss in the left ear. And this was the audiogram I was left with.

And again, you know, right ear is poor, left ear is a little bit better. Boy, I'd have to say if there is, whatever the degree of the hearing loss is, it'd be very unlikely to me that it's purely conductive. Maybe in the left here we do see conductive hearing losses to this degree. The right here, I'm not so convinced. There are some otologic malformations, but it's just too early to tell.

I don't know what I'm dealing with here.

And so now I proceed. And when I do this test, this patient is under general anesthesia in a recovery unit and I just dive right in with tympanometry. Again for the, for trained eyes, you can see that the tympanogram in the right ear is not exact. It's abnormal, but it's not exactly abnormal. That is to say, well, it's not flat.

The way we see flat for this is a thousand hertz probe tone. It's not flat in the way that we expect it to see it. With an effusion. There is a bit of a peak here. It's just reduced.

And I can't help but wonder, does this reflect some sort of reduced mobility due to a malformation of the earth? I don't know at this point, but I might expect that there is a conductive disorder on that side. Again, unknown at this time. The left ear looks a lot more normal. Again, a nice high peak.

You'll notice that there's positive pressure in both tympanograms. And this child is placed under general anesthesia. And we often see positive pressure in the middle ear space as a consequence of the induction process for general anesthesia. Especially when nitrous oxide is used. It usually clears up by the end of the evaluation.

So this is not remarkable. These, this is a very looking, very good looking tympanogram. The left ear is a little mis. I'm sorry, the right ear is a little mysterious to me.

Keep pushing the wrong button here. Otoacoustic emissions are absent. That's no surprise. I'm not going to spend any time on that. I hope you can all read that.

And then. So here I am. I'm ready to do the bear study. The clicks have been done before and so I just want to prioritize everything. I'm going to dive right in and start measuring some tonal signals, specifically narrowband chirps at 2000 hertz.

I'm not going to go into detail about the nature of those signals. I hope that, you know, I'd encourage you to read up on them on your own if you're not familiar with them. And so I start with a right ear. And here we have a very nice looking ABR 1, 3, 5. What I've done here is I've shown the rare and contrastings that comprise the alternating ab, the, the ABR that's measured by alternating signals here, just to demonstrate the repeatability.

I think this is smart to do just, you know, to silence the haters. If someone starts looking at your work and saying, well, that I'm not so sure, well, it repeats very nicely. But the key here is that let's just drop down all the way to the contralateral channel and you can see these features that I talked about earlier. No, wave one, no in the contralateral channel, very prominent in the ipsilateral channel. Wave 5 is here in the contralateral channel, but it's later and lower.

The morphology is distinctly different from the ipsilateral recording at 80 decibels. I'm not really expecting any crossover to occur. And I feel very confident this 80 decibel response belongs to the right ear. And you might be wondering, well, why did you look at the contralateral channel? Why did you do all this work?

Or did you, why did you look at all this stuff if you didn't really expect a problem? Because I'm dealing with a complex child. And I really feel that it's beneficial to get started early on assessing the contralateral differences. Because as you get used to them, when things start looking strange, you're prepared for it. You've braced yourself, you've sort of, you've sort of paved the path.

And so everything right now is looking correct. The left here, you can see that I had to turn it up to 100 decibels to see anything, which wasn't what I was expecting. Again, nice repeatability. Looking at the rare and con tracings that comprise the alternating recording up above. And here's something that you'll notice that when you're near threshold, that contralateral channel, again, we're dealing with an infant, so the responses tend to be a little bit more gross and they're more pronounced in the contralateral

channel.

So this wave five that you're seeing in the contralateral channel is just anemic. But again, giving me a lot of confidence here that this is an ipsilateral recording. This one at 100, this recording at 100 decibels belongs to the left ear. Now, I will make a little side comment here. I had been prepared, I've been bracing myself that I might.

There might be a conductive hearing loss in the right ear. Right. Given that abnormal tympanogram. But I don't see any crossover. If it were purely conductive, I would have seen some crossover here.

So I'm can't, I can't help but wondering, is the right ear somewhat sensorineural? I don't know. Yet again, these are the sorts of things that sort of file into my head as I'm moving along.

Oops, let's do this again. All right, and so let's start seeking threshold here again, maintaining this relationship between the rare and, I'm sorry, the ipsilateral and contral lateral responses. Again, I'm continually trying to reinforce what the ear, what the response should look like from both sides of the head. And this looks all very straightforward. Notice that this relationship between IPSI and contra is maintained.

I'm able to measure the 60 decibel, or I should say, I measure the ABR down to 60 decibels. I don't really see anything at 50. And again, this very nice relationship between IPSI and contra is made. Of course, at 60 decibels, we are not expecting any crossover. Why would we be worrying about determining the laterality of the response at this intensity level?

And again, as I said, I think it's useful to look at the contralateral channel just to train your eyes so that when things go sideways, you're ready for it. Like, well, that doesn't look like it did before. You know, I did this part of the test earlier and it looked like this. Now it's looking like that you're prepared for it.

Whereas if you reserve your time to when you need it, you might not be as proficient as interpreting those differences.

Left ear, back to the left ear. As I said before, no evidence of crossover. Very anemic tracing here in the contralateral position. So I might, first of all, you. I must be very close to threshold, the very low sensation level.

And again, no. No evidence of crossover. And I was just think, when I was doing this test, I was thinking, wow, there's going to be some crossover, because I really think there might be some conductive hearing loss in that right ear based on that abnormal tympanogram. I'm not seeing that. So I'm just filing that away in my head as I proceed.

And again, so here we are. I'm going to go ahead and plot the thresholds. I came in at 60, so that's 55 using the correction factors that we borrow from the UK 100 comes to 95. I think we have a good start here. And then moving on to bone conduction and this I already, things start getting interesting here.

Here I am at bone conduction and we've, I've already trained you again if you're not used to looking at these sort of things. We've already discussed some of the, the lateral differences that can occur with these tests. Let's take up the level here. 70 decibels. You know, we see a wave one, it's obscured a little bit by the, by the artifact which I blocked out here so it could see little wave one here, you know.

And again we are seeing these lateral differences here. But boy, that wave five is huge in the contralateral channel and in fact even when I lower it, I lower the stimulus level. That is really strange that that wave five is so prominent again. It is a little bit later at the slightly negative 10 that you know that negative trough to the wave 5 that they call the SN10 a little bit later here. But boy, you all have to, you must, you must all agree that's this wave five is, is awfully prominent compared to the ipsilateral one.

And I can't help but. And then we're able to measure it down to 50, I think. So clearly no air bone gap. I'm not worried about masking. Again, there's no air bone gap for bone conduction.

So we don't have to worry about which ear that bone conduction thresh that threshold belongs to. But what is going on with that contralateral recording? Why does that look so nice? Again, not perfect, but it's a lot more robust than I'd expect. Well, let's take a look.

Well, I'll be. I'm measuring a pretty decent bone conduction threshold in the left ear. I didn't quite expect that. That was the tympanogram that was essentially normal. Again, the child does have otologic malformations.

All bets are off. But here I am measuring wave five down and post and on offline analysis. I am kind of convinced that there is something here. It didn't quite meet our criteria for response. And I'm not going to quibble about 5 decibels, but you can appreciate that not only is this a very nice looking response in the ipsilateral channel, that is the response that belongs to the left ear.

There is just absolutely nothing going on in the contralateral channel which is very common when we are close to threshold Those differences even diminish in that contralateral channel. And another thing too here is that we're able to measure the left ear a little bit lower than the right ear. So I think what's going on here is that I'm getting a little contra. I'm getting a bilateral response here. I certainly am getting an ipsilateral response.

But that, that other side, that left ear is somehow contributing here. Isn't that interesting? So now what I did here, just to keep myself honest, is I did dump some masking at one of the tracings. I didn't think it was necessary, but as one of my old professors once said, it doesn't hurt to wear your belt and suspenders. And so I just tossed a masking in.

It might have not been an effective level of masking, but it should have been sufficient to see a shift. It should have been a sufficient amount to see if the ABR moves at all. Again, not necessarily reaching a plateau, but to shift it. Well, I think I already have, essentially. I think I have my threshold here.

So I'm not worried about plateau. I'm just testing my theory here. I think this is really a mixed hearing loss. Okay, let's speed up a little bit here. I did measure at 4000Hz.

Not spending too much time here. I think now at this point you can appreciate that. Yeah, you know, 60 decibels, 50 decibels. This has got to belong to the right ear. I'm going to plot that.

And again, at 100 decibels in the left here, no evidence of crossover. The ipsilateral and contralateral readings are looking the way they should. I didn't mask here. I just plotted on this estimated audiogram. I, I think I kind of picked up speed here at this part of the evaluation.

And then we get to the very tricky part here, which is 500 Hz. 500 Hz in the right ear. Again, I'm, I'm using the right ear as my anchor because it's more straightforward. I'm not worried about crossover or anything like that. At these levels, everything should be ipsilateral.

And I'm able to measure. In fact, again, here we go again. Offline analysis did really kind of clean up in a way where I think we have a little wave five here. Didn't quite meet our criteria for response. But again, I'm not going to quibble about this.

It is right around 45 on an estimated audiogram. Jumping over to the left ear, again, really having to increase the levels to see anything. No evidence of A response in the contralateral channel, meaning that there is no contribution. Not only am I seeing the. Just the typical contralateral response just from administering a routine ABR test, but these low sensation levels in infants, it is really hard to see.

It can often be difficult to see anything in that contralateral channel. So no evidence of crossover to the right ear. I even added a little masking here to keep myself honest. And I was a little surprised here because again, I was somewhat expecting a conductive hearing loss at 500 hertz because of that tympanogram. Tympanogram wasn't grossly abnormal.

It was a little unusual. But maybe there isn't a sensor. I'm sorry, maybe there isn't a conductive component at 500 Hz in the right ear. Maybe I was wrong. I mean, there should be some crossover if there was this right.

We'd expect at 100 decibels if there was a conductive hearing loss in the right ear. We should be seeing some conduct. We should see some crossover similar to what we saw in those early examples.

So let's do bone conduction at 500 Hz and oh boy, this is crazy. Very clearly some lateral differences. This really does. In the ipsilateral channel. The right ear, this does seem to belong to the right ear, the slightly negative 10.

You know that that trough that follows wave five is bit later. But boy, that is such a big wave 5 in that contralateral channel. So I'm faced with this situation again of thinking, man, is that other ear contributing even here at these low levels by bone conduction in the left ear? Again, these differences are not as pronounced as I had hoped, especially in a child this age. Getting and I'm.

What I'm doing here just for the sake of time is I just reduce the levels continually. I even get down to 10 decibels using chirps. That's why the responses are so early. And I'm just not con. I'm really squinting here and I don't like to squint.

I, you know, sometimes I look at this and say, yeah, that's the, that's the upsliddle response. And sometimes I look at and say I'm not so convinced. But boy, my, my gut is telling me that the left ear is even contributing here, which is maddening. No, it's not maddening. It's exciting.

Let's see what happens when we look. Yep, sure enough, even the left ear wants to respond at very low bone conduction stimulation levels 20 and then going down to 15. Very nice looking wave 5 very attenuated, late contralateral response. This is again what you want to see when you're administering an ABR test and examining the different channels to determine the laterality of the response. Now, the thing is, of course, that the right ear is better than the left ear.

So this is a good opportunity for me to start dumping high levels of masking to see if I can make any changes. So on one of these tracings, I did add 100 decibels of masking. Again, intense enough maybe not to get an effective level of masking, but certainly enough to, you know, to see a shift, just like what we do with audiometry. And I'm not really seeing anything. So then I get, get, I get even more intense here.

I said, well, let me drop 110, 110 decibels of white noise here at 500 hertz and see if anything changes. And again, this wave five, I'm not crossing over here. The wave five in the left ear and the ipsilateral channel is really holding on. There's a lot of mess going on here. And in fact, it's so messy, I decided to clean it up a little bit by calculating grand averages for these.

And if we just look at the lowest threshold here, by calculating grand averages, you can see that even with high intensity masking in the contralateral ear, my wave five holds on in the left ear. Nothing seen in the contralateral channel. Again, I'm going to say this again, which can happen when you're at a very low sensation level. Those differences might be even, even greater, even more diminished when you're at a very low sensation level and you're very near threshold. And this right ear is just not very persuasive to me.

Is there a conductive component here? There might be. I mean, it's tempting to say that if we just look at the audiogram, maybe the bone conduction threshold is a little bit higher. But at this point, I opted not to plot anything. I think that I have.

I think it is sufficient to say that this is a primarily sensory neural hearing loss. The tympanogram is normal. We might find that there's just the slightest 15 or 20 decibels air bone gap at 500 hertz in the grand scheme of things, not critical. And so I begged off. I decided that this would be a time where I don't really need to nail it down.

I think I have enough information to talk to the parents and referring providers about how to care for this child. So to wrap up my assessment in the right ear. The child has a primarily sensory neural hearing loss could be mixed. I'm going to hold off on that. I'm just not convinced.

Of course I can't math task to determine, but there are some. I have some tricks up my sleeve that might clean that up for me. Stay tuned. Left ear is certainly a mixed hearing loss. I do need to retest this child within three months to extend and confirm.

And now that I've blazed the trail, I'll be able to work more efficiently to determine what's going on. This, this child may do very well with a hearing aid in the right ear. Again, just given the degree of the loss, of course I have to consider the otologic malformations, which I did not get a very good opportunity to assess given the test circumstance. So the patient will be coming into the clinic to see me and you know, who knows, maybe there'll be some changes in the thresholds the left ear and maybe that'll make it easier for me to choose a treatment plan. But boy, this is going to be tricky in that left ear given the degree and the malformation to wrap up here.

Think about these things. If there's a key takeaway here, there's something that I'd like you to learn about. This is evaluate both channels. You know, some of you out there are using only single channel recordings. And if you have the ability to measure both, do it.

It's just, it's free, it costs nothing, and it just trains your eye, it edifies you, it educates you. Evaluating both channels can help you determine the laterality of the ABR with and without the use of clinical masking. And it's such an essential tool, especially when you're working with kids. Assessing contralateral recordings, even when they are not, when it is not necessary to do so, can help guide your evaluations when they become murky. Yeah, do it when the kid has normal hearing.

Just get your, get your eyes trained to what contralateral response should look like, especially when you're dealing with a case like that. Start applying these techniques just so that when things get a little muddy, you're ready for it. Oh, that looks different. That doesn't look like what it did before. It trains your eyes and you know that's just free training.

You know, you don't have to pay for it. It doesn't cost any time. You just click other channel and get in the habit of printing them up so that you can look at them later, clean them up and share them with others. Your senior audiologist, who might give you some advice on what's going on here. It's a really a great way to learn more about how to take care of your patients better.

And then finally, finally think dynamically. It is some, it is sometimes all right to seek clarity and follow up evaluations. I find that with learners they want to get everything done at once. In this case, clearly I'm going to have to return to that right here. So I hope I didn't go down too long.

My closing thoughts I really do hope that it was interesting. It was. I was excited to share it with you. I find that these things are very interesting and I hope that if you've never done these before, you're inspired to do them. And if you didn't have confidence in doing them, I hope you have more confidence.

Good luck to you all in measuring these ABRs and and with that I am going to hand the presentation over to Dr. Alyssa Nickerson. Thank you. Well, thank you. That was a great presentation. I am a pediatric audiologist here at Phoenix Children's Hospital and I am involved in both the diagnosis of hearing loss but also the management of hearing loss.

And I'd like to talk to you about something that we see very common here at Phoenix Children's Hospital and that is hearing loss due to Gap Junction Beta 2 or GJB2. If you are an audiologist, you too probably encounter this very frequently.

Hearing loss impacts approximately 1 in 650 newborns. It is the most common congenital sensory impairment at birth and it's one of the most common inherited disorders. Genetics account for more than half of congenital hearing loss, I.e. hearing loss present at birth, and the remainder are due to alternative causes such as environmental factors or infections. Of the genetic hearing loss, approximately 70% is non syndromic and the remaining 30% is syndromic, meaning the patient has a syndrome in which hearing loss is a symptom.

There are a number of inheritance patterns for non syndromic hearing loss, but for the majority they are autosomal recessive. Approximately 80% changes in the GJB2 gene account for nearly half of non syndromic prelingual sensorineural hearing loss in some regions. To be specific, I said prelingual, but in many cases it may very well be congenital. It's difficult in some studies to determine if hearing loss was present at birth or onset in the very early years of life, but very often this hearing loss does occur before the development of language. The relationship between GJB2 and hearing loss was first reported in 1997.

This is a gene that codes for the gap junction protein connexin 26. As audiologists we sometimes hear it referred to as connexin 26 in a casual manner and it's the same thing. The variant prevalence differs

by population and the carrier rate is higher in some regions, including the Mediterranean. Although it's always changing, there are greater than 150 mutations or changes in GJB2.

I'd like to go over a few common audiology findings that we see with patients who have this type of hearing loss. Although not every hearing loss will follow this guideline, very often it's prelingual. In many cases it may be congenital. The hearing loss severity is often moderate or greater. Very frequently you'll see severe and profound hearing losses, although as I said, it can be even mild.

And there can be differences in severity even between family units, between siblings. The severity of hearing loss in some cases is believed to be related to the type of mutation or gene change. Hearing loss is very frequently bilateral, impacting both ears. Generally it's non progressive, meaning it's not getting worse with time and you'll see a flat or slightly down slanting hearing loss. And I'll show you an example today of a patient that follows this configuration.

We will go through two case studies today. Both of these children are children who are diagnosed with hearing loss very early in their lives. This first patient will call case study number one. She was born full term. She referred on her initial newborn hearing screening in both of her ears and then she referred on her outpatient follow up newborn hearing screening in both of her ears.

At that time the parents started to notice, oh, there might be a problem. She was only startling to very loud sounds, but they felt she could hear very loud sounds. There was no prior family history of hearing loss. We saw her initially when she was six weeks of age for a diagnostic BEAR study or a diagnostic ABR study. At that time she had normal middle ear status.

Her 1000 Hz tympanograms were peaked. Her otoacoustic emissions were absent in both of her ears. And the ABR study that we did suggested bilateral, moderate or moderately severe sensorineural hearing loss.

She then underwent a genetic consultation with Phoenix Children's and that revealed a gene change in GJB2. The specific variant that she had is common in individuals of European descent, which she is. She then established with our otolaryngology department, ophthalmology and our speech therapy. She underwent imaging of her brain and her temporal bones and that was unremarkable. We have seen her now for many years.

Her hearing loss is stable. She is a user of listening and spoken language and she's an excellent hearing aid user, averaging about 14 hours a day. On this next slide you'll see a recent audiogram that we've completed for her. And again this has been very stable. She is about 9 years of age today.

This next case is interesting because the hearing loss is a little bit more severe. This patient also was born full term. She underwent an initial newborn hearing screening in both of her ears which she referred on and then she had a subsequent outpatient follow up newborn hearing screening which she referred on in both ears. At the time that we saw her, there was a reported family history of hearing loss with a very distant paternal cousin who was born with hearing loss in both ears. Prior to coming to our facility, this patient underwent an external BEAR study and what I mean by that is a study at an outside facility.

And that facility reported that this patient had severe to profound sensorineural hearing loss in both of her ears. The parents are both medical providers and they were interested in pursuing a second opinion which is how she came to our facility. She came to us at about seven weeks of age and we did this same workup that the other facility did. She had normal middle ear function, she had peaked 1000 Hz tympanograms, she had completely absent otoacoustic emissions. And the BEAR study that we completed at our facility or the ABR study that showed profound sensorineural hearing loss in both of her ears.

And it's worth noting that most of her ABR was a no response ABR meaning she had very little hearing at the outputs of our equipment.

Now this child underwent the same workup she established with ent, our ophthalmology group and speech therapy. She underwent a genetic consult and that revealed not one but two variants in the GJB2 gene. One of the genes is very common in regions of India which is in line with her East Indian descent. She underwent an MRI of her brain and her temporal bones and that was unremarkable. She began using hearing aids quite soon and she was deemed a good candidate for bilateral cochlear implantation which she underwent at seven months of age.

She is now six and a half years old. She uses her cochlear implants about 12 hours a day and she is very successful with her listening and spoken language.

And so just a few follow ups. GJB2 related hearing loss is very common and accounts for nearly half half of non syndromic pre lingual sensorineural hearing loss in some regions of the world. If you're an audiologist working with children, I'm sure you will encounter this very often. Prevalence varies by region and ethnicity, and hearing loss very often is moderate, severe, or profound, impacting both ears. And at this point, I would like to turn it over to Dr. Ashley Geske, who is an audiologist on our staff, and she is going to give a presentation to you now.

Thank you. Awesome. Hi, everyone. I'm Ashley Gueske. I'm another one of the audiologists here at Phoenix Children's Hospital, and I'm going to be talking to you about a patient that had a lengthy journey to get her diagnosis, and we will call her zoo.

So, first, we're going to go back a few years. When she first presented to Phoenix Children's, she was initially referred by her pediatrician to neurology due to concerns about an abnormal gait that she was having. They noted right away that she kind of has a generalized hypotonia. She was already enrolled

in earlier early intervention services. She was getting physical and occupational therapy by this point due to a motor delay.

She did not sit up independently until about a year old, and she did not walk until 21, 22 months. She was also in speech therapy by this point due to a plateau in her speech development around two and a half years old. Her parents just reported that she stopped developing new words at this point. Neurology wanted to just get some more information, some more data about her. So they recommended, you know, evaluations with a bunch, a different specialist, as well as some scans and kind of additional testing.

About a month after she saw neurology, she saw developmental pediatrics. They did not notice any concerns for autism in their evaluation. They noted she's very social. They were also happy that she was, you know, already enrolled in early intervention services, and they recommended just kind of continuing with that and returning in six months to monitor her progress.

She also had an MRI which showed a subcortical T2 flare signal abnormality in the temporal lobes right greater than her left. They noted that this, you know, might be secondary to a prior viral infection, inflammation, or metabolic insult.

Neurology also referred to genetics. At this point, they had a strong suspicion that there was some kind of underlying genetic condition. They also kind of preemptively ordered chromosomal microarray testing and fragile X testing that came back normal.

When she saw genetics, they also recommended some additional testing on top of what neurology already ordered. They requested a metabolic panel and then kind of an expanded autism Panel as well. And all of that came back negative. So they just recommended follow up in a couple years.

By this point it had been about a year, year and a half of kind of follow ups and testing and you know, we didn't have a whole lot of answers. And so they went ahead and recommended audiology and ophthalmology consults, you know, back. I don't think they recommended a hearing test up until this point just because, you know, parents didn't really have any concerns for hearing. You know, she was making progress in her speech therapy. You know, they, she was able to follow directions and converse with them in sessions.

And so that wasn't really at the forefront of their mind.

So she saw one of my colleagues for an audiologic evaluation. At the this point, you know, parents still had a lot of concerns about her symptoms and you know, not a lot of answers and they were just very eager to get down to the bottom of what was going on with their daughter.

So my colleague went ahead and started out with some objective test. As you can see here, she had type a tympanograms, present OEs in the right ear, absent in the left. She had a present kind of screening ipsilateral reflex in the right ear at a thousand hertz and then her reflexes were absent a thousand to four thousand in the left ear. They also did some behavioral testing as well. She was able to do CPA and this is the audiogram they got.

So as you can see, essentially normal in the right and then profound no response in the left ear. She was also able to do a picture pointing SRT that was normal in the right ear. And then it was no Response up to 80dB HL with contralateral masking in the right ear. At this point I think she just kind of fatigued to testing due to her age. But you know, we have a pretty good picture about what's going on here.

Right now it's looking like left single sided deafness. So my colleague went ahead and referred to ENT for a consult and when they saw ent, they recommended proceeding with an MRI and a bear study.

So she previously had an MRI that was ordered by neurology to look at the brain. But the main purpose of this MRI was to look at the inner ear structures so we can, you know, know what her options would be, intervention wise.

So her, she had present cochlear nerves bilaterally, her inner ear anatomy was normal. And they, they repeated the scan of the brain which, you know, still kind of showed that T2 flare in her temporal Lobes. She also had a bear study which lined up with you know, her behavioral testing. So normal in the right and then severe to profound sensorineural hearing loss in that left ear.

The my colleague also recommended a vestibular evaluation since parents initial concern was her abnormal gait.

So over the last couple years PCH has added a vestibular program that kind of does an abbreviated protocol. You know, it's something we hope to add more equipment in the future and kind of expand on our testing. But for now this, this is what we're capable of. And I have this kind of split into two slides. So this is the first part of the testing and then we'll go on to the second part of testing.

Now I am not a vestibular audiologist so kind of bear with me through this. I'm just kind of summarizing what our vestibular audiologist did and the results they got and kind of their, their summary of the results and interpretation.

I also, so I went ahead and bolded the results that came up positive or abnormal. You can see here quite a few of her tests came up abnormal. She was noted to have catch up saccades bilaterally on her head thrust test, her broken man's rotary chair showed absent post rotary nystagmus in both directions. She fell immediately on single leg stance. She also fell in both eyes open and closed conditions of the cat siblings.

They noted that she kind of widens her stance to maintain balance while walking and she would kind of veer to, to both sides.

Whoops.

She also had CVEMP and OVIMP testing as well. And her right ear she had absent CVEMP waveforms and the rest her, her left ear cumps had present waveforms and then her OEMs came up normal in both ears. So overall her results suggest a bilateral peripheral vestibular weakness. She did have absent semps in her right ear which suggests reduced inferior vestibular nerve and or sacule function.

This I found this interesting because it's actually on the opposite side of her hearing loss. So her hearing loss is in the left ear and then these absent waveforms were in the right ear.

So putting all of this information together from her history, vestibular testing, audiologic testing and her MRI results, the top of our differential diagnosis, this is ccmv. I'm sure most of you are familiar with ccmv, but for those of you that need a refresher, Congenital cytomegalovirus is a virus that acts very similar to the common cold in the sense that you know most People have very mild or no symptoms when they have it. But when you get it, when you're pregnant, you can pass the virus to your baby and it can cause issues with development. One of the most common symptoms we see is hearing loss, which is why we see such a high number of these patients. Now, the only, you know, true way to confirm CCMV is either to test the blood spot if the state still has it stored, or complete a PCR test within the first couple weeks of life with either, like a saliva or urine sample.

The state of Arizona does not hold on to the blood spots for more than three months. So, you know, since we had no suspicion or we, you know, didn't really even know her during those first couple months of life, she was never tested for ccmb. So for her specifically, we'll. We'll never know if this is

her, her true etiology. But I wanted to step through the characteristics that we're seeing that make us suspect this to be her, the cause of her symptoms.

So first and foremost, her MRI. Her MRI results showing a subcortical T2 flare in her temporal lobes. It is very common with CCMV to see kind of brain changes on the mri. You know, for example, you can see like, white matter calcifications, subarachnoid cysts, ventriculomegaly, you know, so on and so forth. There's a very long list of possibilities that you can see, but this is something that we see often with these patients.

The second and third characteristic that make me suspicious of CCMV is a rapid progression in hearing loss and an asymmetric progression in hearing loss. So what I've noticed anecdotally, and what's been seen in the research is with patients with asymptomatic CCMV that have assumed normal hearing at birth, many of them start out unilaterally.

When this hearing loss is early onset, it will often progress rather quickly as well. And then just, you know, to note, many of these unilateral early onset hearing losses will progress to bilateral hearing loss. And, you know, while I was doing this presentation and looking into the research, I found a study that was published in 2017 that showed kind of the average time between that unilateral to bilateral progression is about four years. And this is something we'll kind of keep in mind once we get to our intervention. Discussion.

Another characteristic that we saw that make us suspicious of CCMV is her bilateral peripheral vestibular weakness. It is very common to see vestibular dysfunction in patients with ccmb, particularly progressive vestibular dysfunction. So this is a good thing to kind of keep in mind with this patient population, you know, recommending a vestibular evaluation if you have that available to you. And then two final characteristics that we saw, you know, and these ones are nonspecific. You know, when I see these in a patient's case history, I don't automatically think, oh, it's ccmv.

But given all of our other findings, this kind of strengthens, strengthens that suspicion. So the first is intrauterine growth restriction and the second is kind of that low muscle tone or hypotonia.

So the next step for Z was to undergo a cochlear implant evaluation and discuss her intervention options. And this is where I was first introduced to Z. So I went ahead and started out with some testing. I confirmed her audiometric thresholds. Everything lines up with, with previous results.

I also did some word recognition testing, percent in the left ear. You know, with an SSDCI evaluation, I'll typically do additional testing that looks at like head shadow, binaural summation and squelch. But given her age and level of attention, it just wasn't appropriate in this case. So at this point we sat down and had a discussion about her options.

You know, I talked about the possibility of doing a cross as she gets a little bit older, doing a bone conduction hearing aid or just simply monitoring and doing classroom accommodations like preferential seating or like a sound field system. We also discussed about the possibility of proceeding with a cochlear implant. I counseled her family that this was the only option that will like really give her binaural cues.

For this patient specifically, there were two, two kind of main reasons why I was a little bit more excited about the possibility of doing a cochlear implant at and I. And we'll go through those.

So the first reason is that her hearing loss is suspected to have onset around two and a half. You know, given her parents report of kind of that plateau in her speech development around this age, it's reasonable to assume that this is either when her hearing loss onset or when that's. We finally, this is when we saw that kind of final progression in her hearing. You know, from more recent research about outcomes for cochlear implants with ssd, you know, we know that, you know, for congenital and

perilingual hearing loss, the best outcomes is ideally under three years from the time that the hearing loss onset to when they're implanted.

Given, you know, that we suspect her hearing loss onset around two and a half. And the time of this evaluation, she was almost five. She's likely well within that window. So this is really the optimal time to proceed with the cochlear implant, if that's what parents choose.

The second reason is that with ccmv, hearing loss will often start out unilateral, but a large percentage of these patients will progress to bilateral on an average of four years. So she's still kind of within that window of, you know, a higher probability of, like, seeing a progression in her better ear. So, you know, discussing with her parents, we kind of talked about the fact that a cochlear implant could act as, like a safety net if we do see a change.

During our discussion, we also talked about realistic expectations. You know, at this time, her parents did not have any big concerns for a hearing. Their main concerns were her gait.

When this is the case, I always, will always counsel family that, you know, in your eyes, she's already performing at 100%. So after she's implanted, you're likely not going to see this kind of light switch change, you know, because she's already functioning, you know, pretty close to ceiling. You know, we talked about what we can expect from a cochlear implant. You know, it will likely improve her understanding in background noise. In background noise, and potentially improve her ability to localize sound.

So after our discussion, her parents elected to go forward with a cochlear implant for the left ear. She's been activated, her maps were optimized, and she's wearing it 10 to night, 9 to 10 hours a day, and really tolerates it without issue, which is a huge blessing.

So Z is still, you know, very early on in her journey. So we don't know what her outcomes are or what her full potential is with the CI. But given all the stuff that we talked about before now, I am very optimistic that she'll do really well.

So my final thoughts on Z's case are sometimes things are not what they seem. I think her case reiterates the importance of, of completing a basic hearing and vision screenings even when there's not a concern. Hopefully you learned some new signs and symptoms of CCMV so that you can better recognize it in your patients and get them tested, if that's available to you. Thank you so much for your time and attention. I hope you enjoyed my presentation today.

And with that, I'm going to introduce Our next presenter, Dr. Ali Sayer. Thank you, Ashley. That was a great presentation. So I'm Allie Sager. I'm another one of our audiologists here at Phoenix Children's Hospital.

I am also on our cochlear implant team and today's case I have for you is a little bit of a mix of everything. So this is a case of challenging mixed hearing loss. I wanted to start today by briefly reviewing a few concepts that'll be important for my case study. Any one of these could be probably a full presentation on their own. So this is not not by any means a complete review, but due to the mixed hearing loss in this case, the concepts we'll cover are a little bit all over the place.

Today I'll focus on just some of the basics of microtia and atresia amplification options for mixed hearing loss and cochlear implant candidacy for pediatrics to help support understanding for the case to follow. Microtia refers to a range of congenital malformations of the pinna. While there are a few different ways to classify the severity, the references you'll hear today are from the model marks classification shown here. On the mild end of the scale, grade 1 microtia refers to a formed but smaller than typical pinna. On the more severe end of the scale, grade 4 microtia can also be called a nocia or the complete absence of the pinna.

These malformations of the external ear often co occur with malformations of the external auditory canal and the middle ear space. The severity of the microtia is often correlated with the severity of the ear canal and middle ear malformations as as well well while surgical reconstruction can be an option, this is often for cosmetic purposes only, especially when ear canal and middle ear malformations are involved. Hearing restoration is not a common outcome and use of hearing devices is still typically needed following surgery.

In cases where the ear canal is open, traditional behind the ear hearing aids can be a good option depending on the extent of the pin and malformation. There may be difficulties with ear mold fit and feedback, but a patent ear canal makes it a possibility. In more severe cases, the use of bone anchored hearing aids or bahas is required. These hearing aids work to bypass the malformed parts of the ear to stimulate the cochlea through vibration. For young children, these are fit on a soft headband with a connector for the external sound processor which delivers sound through vibration.

Around age 5, children may be eligible for a surgically implanted device device. Historically these were passive devices. The implant was a titanium screw placed into the bone and the external sound processor connects to it either directly through an abutment which is a metal post that sticks through sticks out through the skin and tissue or by way of magnets, one connected to the implant and another connected to the external sound processor. More recently, devices have become available which have active implants. In these systems, the external sound processor does not vibrate it picks up sounds from the microphones and transmits it electronically to the implant which delivers the stimulation.

Middle ear implants are an additional option, though certainly not as common, especially in the pediatric population. Some of these implants can be considered starting around age 5 and typically utilize an external sound processor to deliver a signal to the active implant which connects to the ossicles or is fitted into the round window to deliver that stimulation to the cochlea. There are also totally implantable

systems which place the sound processor and power source in the postauricular area, but this is beyond the scope of my presentation today. They Cochlear implant candidacy in the pediatric population is rapidly evolving. When they were first approved for pediatrics, it was for two and older with bilateral profound hearing loss.

But as time went on, the candidacy criteria has widened to include younger children and those with more residual hearing. Current FDA indications vary by the specific manufacturer, but in general pediatric candidacy now includes children as young as 9 months of age with bilateral profound sensorineural hearing loss loss. For children two and older, those with severe to profound hearing loss can be considered. It's also necessary to demonstrate that the child has poor word understanding scores similar to adult criteria, or that there's limited benefit from the use of hearing aids. For young children, this is done by demonstrate demonstrating poor progress in speech and language development.

There are also indications for single sided deafness in the pediatric population. Now the term off label refers to the use of an FDA approved medication or device in a manner that's not specifically approved by the fda. For cochlear implants, this could apply to a number of different patient categories such as children with asymmetric hearing loss where only one year meets criteria, or those who have better hearing thresholds with poor water understanding scores. This could also apply to early implantation which can be considered as early as 6 months of age here at our center. Off label use of cochlear implants allows us to remain more progressive and inclusive in our candidacy considerations.

To support our patients, patients and their families in achieving their communication goals, we rely heavily on research and recommendations from professional organizations in making these recommendations. There are a few guidelines from the American Cochlear Implant alliance cited in my references, which I highly encourage you to review with those concepts in mind. Let's jump into A case

study for a very unique patient of mine. So Irene is a now 6 year old female first seen at our facility at 5 months of age after failed newborn screening. Her history at that time that we knew of was additionally significant for right sided grade four microtia and oral atresia and left grade one microtia.

An outside ENT provider had ordered an MRI due to her ear anomalies and the inner ear anatomy including the cochlea and auditory nerves was normal. Her initial ABR showed a right profound hearing loss noted to be primarily sensorineural since they didn't see a bone conduction response, but they noted that a conductive component was likely given her microtia and atresia on that side side the left side. They found a moderately severe sensorineural hearing loss.

A few days after her initial abr. She established with ENT here at our facility where cochlear implants were first discussed with the family. They also made a plan for repeat ABR under anesthesia in coordination with imaging for around nine months of age. She also established with plastic surgery who noted concern for bilateral facial nerve paralysis and said that reanimation procedures for that could be considered around six to seven years of age. They also noted that reconstruction for her microtia could be planned around 7 to 8 years of age and that because cochlear implantation could complicate this coordination was necessary.

She returned to audiology for a hearing aid consultation at which time she was not recommended for the use of a hearing aid in the right ear since her anatomy didn't allow for a traditional hearing aid and her bone conduction thresholds precluded precluded her from candidacy for the use of a softband baja. She was recommended for a BTE in the left ear and was fit with her hearing aid around six months of age. At her first follow up they noted constant feedback from the hearing aid and reported that it didn't make any clear improvement in her responses to sounds.

I first Met Irene at 10 months of age for her repeat ABR under anesthesia in coordination with her CT scan. Significant anomalies of the ossicles and middle ear cavities were observed in both ears along

with an abnormal course of the right facial neck nerve. The EBR also had some new findings. The left ear thresholds decreased compared to our initial study now showing overall a severe hearing loss. The right ear still showed no response by air, but we did see some bone conduction thresholds in the mild to moderate range where it was previously absent.

Well, the results for a thousand hertz. If you look at the waveforms at the top of those ABR tracings there were not hugely convincing since it was a very small wave fat. The clear presence of waves 1, 3 and 5 at 4,000 Hz were enough to pause the team's plans which at that point were leaning towards bilateral cochlear implantation.

I saw her for further evaluation of her candidacy for cochlear implants when she was now 11 months of age. We reprogrammed her hearing aid and after those gain increases she widened her eyes quieted in response to speech. Aided testing confirmed this too, showing some response responses to sound in the mild hearing loss range. She also participated well in some unaided testing with a clear bone conduction threshold at 2000 Hz that was consistent with what we measured on her ABR. This was also meant that we did need to pause and change our plans, which were which the surgical team was okay with given the complexity of the surgery.

As they began planning that coordination between plastics and entire the left ear was likely going to be a candidate, but we had more work to do first. She returned shortly after that for fitting of a super power Baja on softband for the right ear and we refit her with a new ear mold for the left ear, planning to extend her trial for a little bit to see if that made any improvement.

She returned at 14 months of age at which time her mom reported she was able to use the BAHA much better than the hearing aid, but retention continued to be an issue. The soft band would slip and prevented the transducer from staying in the the correct spot. They reported that she seemed to hear a little bit better with the BAHA than she did with the hearing aid, but she still wasn't responding normally.

She was babbling but not producing any words. Irene herself, even though she was little, also seemed to notice good benefit from her baha.

During our visit it popped off and she walked over and handed it to me requesting help putting it back on, which to me was a great signal that she was receiving some benefit from the development device. Following this appointment, the team agreed to proceed with left cochlear implantation and to have her continue use of her right baha.

At 16 months of age, Irene underwent surgery for her left cochlear implant. Surgery did not have any complications. Interoperative testing was normal. An X ray was also completed to confirm device placement. Two weeks later, at 17 months of age, she was activated and had a very clear response to the device.

In the eye office she would become upset when it was muted or removed at her first follow up, data logging was already improved over her use of hearing aids at six hours a day on average. At her next follow up, now three weeks post activation, aided testing with the audiogram you see here showed good audibility across frequencies and the family was seeing improved responses at home. She was also noted to be saying her first word, which was dead ass by eight months post activation, where she's now a little over two years of age. The family felt that she was hearing and understanding with ease at home, but her progress with speech was limited due to her facial nerve paralysis. She was communicating with a combination of spoken language, sign language and an AAC device.

The family was also interested in pursuing a cochlear implant for the right ear. They cited that she's upset when the cochlear implant processor is removed, but doesn't seem to care about the BAHA anymore. She was also rejecting use of the BAHA at home and didn't seem to be hearing as well with it alone as she did before she got the cochlear implant. Data logging at that time showed good use of both devices. Because of the parent complaint that she wasn't hearing well with her baja alone.

We repeated some unaided testing to see if there had been a decrease in her unaided hearing thresholds. But remarkably we found the opposite. Her bone conduction thresholds had improved. Now rising up in the higher frequency. I had a really difficult conversation with the family at this point about how Irene is not a candidate for a cochlear implant in her right ear.

We discussed that while I certainly understand why she prefers the cochlear implant to her baja, but the type of hearing loss and the degree of her bone contraction thresholds just doesn't leave that as an option, you know. Additionally, we discussed that she does get some benefit from her baja and we would expect that when she's older and eligible for one of the surgically implanted devices that those benefits benefits could increase. They understood, but were frustrated with it. The biggest impact of that conversation was really not in her audiologic outcomes, but in her follow up. They disappeared on me for a little bit.

So she was a little over two there and next followed up over two years later. Now four years old, only because her baja had broke. It was now out of warranty and mom wanted her to be fit with a smaller baja to see if that improved her tolerance of the device. Device. Her bone conduction pure tone average was at the limits of the newer smaller Baja.

But at their preference, we proceeded with a fitting. I attempted testing in the office that day to confirm her benefit was comparable to what she had with the Superpower device, but she was not having it that day. She did follow up a few months later, at which time she was overall doing well for both devices, although she continued to prefer her cochlear implant over her BAHA and was now actively expressing that she doesn't like her baja. Her speech continued to be poor due to the facial nerve paralys, but she was communicating well with asl attending pdsd, which is our Phoenix Day School for the deaf in Pre K. She did prefer to speak, but her intelligibility was or for unfamiliar listeners.

She was most recently seen last October, now six years old and overall it was more of the same. She strongly dislikes her BAHA and loves her cochlear implant. She wears the cochlear implant processor 13 hours a day and the Baja only 3 hours per day on average. Family was still interested in the right cochlear implant, but given her bone conduction thresholds it would be very hard to justify. There is some literature supporting the use of a cochlear implant in mixed hearing loss, but what's different is that these patients have significantly poorer bone conduction thresholds.

Typically they will exceed the fitting range of available bone conduction devices. In order for me to recommend off label implantation in her right ear here, we need to go off book a little bit and I would need solid evidence to support that and unfortunately at this point I can't find anything to support that for bone conduction thresholds in the mild hearing loss range, even though that is what the family wants. So for some closing thoughts, where do we go from here? The family is hoping that her bone conduction thresholds will miraculously drop so that she qualifies, but I think there are some better options. In the absence of any clear guidance on what to do in these situations, we discussed that prior to considering it.

We'd have to prove that no standard treatments are sufficient, so I recommended that we try a surgical bone conduction device now that she's old enough in order to overcome the reduction of sound of the device going through tissue when she wears it on the soft band. We could also look into her candidacy for middle ear implants, though given the extent of her malformations on the ct, I don't think that's a likely option for her either. Her mother was interested in one of the active bone conduction implants and scheduled a consultation with the surgeon to discuss discuss. They unfortunately canceled and hasn't been rescheduled to date. I'm hopeful that the family will come back in to move forward because I think that could be a great thing for her, and we want to figure out how to maximize her hearing potential for both ears.

She's a unique and challenging patient, so I look forward to seeing how it unfolds from here.

Okay, great.

What wonderful presentations. I learned a lot from listening to all of you guys. Are there any questions from the audience? If there is. If there are, just go ahead and type them into the Q and A.

And while we're waiting, Bob, I have a question for you. I really learned a lot from your presentation about the contralateral measures. And my question is, how long does it take? Like, approximately. And I'm sure it depends on if you're doing, you know, air, bone or, you know.

But. But how. How much time does this add to your evaluation? It adds absolutely no time. The.

Once you have the. Am I on? Yes, I should have checked. Okay, good.

It adds no time because the way that the electrodes are configured, these channels are recording simultaneously. In fact, in. On some units, you can measure a full 1020 complement all at once. I mean, the. The.

The CPU is able to take in recordings from various sites on the scalp. So it adds no time. The only thing that you need to do is click a button to look at the other side. So this is really, really. It's at no expense to any audiologist.

And so I really encourage, when I work with learners and students that just do adds no time. It's happening simultaneously as you're measuring. You know, sometimes they can get in a way. You might want to shut them off or turn them on. But it's.

I really do think it's an essential part of administering a test, especially when things can get a little dicey. So. And if you're not in the habit, you don't do it. And then you realize, I should have done that. But getting in the habit, you're always checking those channels.

Whether you keep them up or not, that's one thing. But just check them. And it's simple. It's as simple as hitting a button. That's very simple.

No problems. That makes a lot of sense. Thank you. Tanner, how do you determine which method of counseling is the one to use for any given situation? Yeah, so that's a great question.

And I think a lot of it comes down to the conversations that you have with the family using narrative medicine. And open ended questions, I think, are the best two ways for you to kind of start that so you can One, figure out where the family is at and two, by using your open ended questions, you can gauge their responsiveness, whether that's technical questions that they have that you need to answer. And we go over as a professional, maybe it's emotional responses that you then need to validate, maybe it's just shock or different other emotions that they may be experiencing. And so I would definitely say the narrative medicine, just getting to hear their story helps you to connect with them and asking open ended questions so then you can know what other counseling skills you need to use and what you need to spend your time on. Makes sense.

Thank you, Ashley. If, if we see a newborn and we suspect ccmv, when's the best time to test the baby and how should the baby be tested or how can the baby be tested?

So either, you know, if, if it's within the first couple weeks of life, you could potentially do like a saliva swab to, to do like a PCR test. If not, then just learning like what your states, how long they keep the blood spots for and seeing if you can obtain the blood spot and have that submitted for the same PCR test. Okay, thank you, Ash. Allie, what are the benefits of an active bone conduction implant? You just

mentioned that Irene was looking at that.

Why is that an option for her? Yeah, so the biggest benefit of that is that we are not going through that skin and tissue any longer with those original bahas. The benefit was that you connected directly to that titanium implant, so we did not have to send that vibration through, through skin, through tissue at all. You went directly into that bone, into the cochlea for the most clear signal. While the ones that have the magnets are a good option, you now are going back through that skin and tissue to deliver it through the magnets.

So these new active systems kind of combine the best of those two systems where we're getting that direct stimulation into the bone because the part that's doing the vibration is underneath the skin and tissue. But we also can use the magnet to put it on rather than having to have, have that abutment sticking out through the skin and tissue, which leads to infection risk, overgrowth issues, and cosmetically is just not the nicest option either. So it kind of gets the best of both worlds there. Makes sense. Alyssa, how does knowing that a patient has a G G B2 variant, how does that impact your evaluation process?

Yeah, absolutely. I think one, it's good for the patient to know for their future, like family planning, when they become older. But also I think one of the questions that I get asked a lot by parents is will their hearing loss get worse? Will it change? And if we know the cause of the hearing loss, sometimes we can answer that question, sometimes we can't.

But in the case of the gap junction base data 2, we know that it's generally non progressive, so we can give the family kind of a more clear roadmap of what to expect moving forward. Perfect. Okay, audience, if you think of any questions that you wish you had asked, here's my contact information. Send me an email and I'll get it to the presenter and we will get back with you. Thank you all for hanging in there and listening and I'd like to thank all of our presenters for their time and expertise today and for

an excellent, excellent webinar.

And I'd also like to thank everyone who participated in today's course. We really look forward to your feedback on the course evaluations and we hope to see you on future courses on audiology online. So this concludes today's course and have a great rest of your day.