Getting On The Right Track: Untangling Complex Pediatric Audiology Cases, in partnership with Phoenix Children's Hospital

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At this time, it's my pleasure to introduce Dr. Wendy Steuerwald. She is director of Audiology at Phoenix Children's Hospital. She works with hospital leadership and audiologists to create an environment where exceptional patient care is the norm. We are so appreciative and grateful for all of our presenters today and I'm going to turn it over to Dr. Steuerwald. Thank you, Carolyn, for that wonderful introduction.

We are so happy to be participating with Audiology online for the pediatric grand rounds today. I believe this is the fifth or sixth year we've done this, so we are excited to be doing it again. The topic of today's conversation will be getting on the right track, untangling complex pediatric audiology cases.

So we have some disclosures. Everyone is an employee of Phoenix Children's Hospital. There's other information on there. Go ahead and read it on your own. There are also some limitations and risks.

You can go ahead and read all of those. Also, learning outcomes are presented. Basically after this course, we think you will be able to describe methods to give unexpected results to parents and patients in a clear manner. Explain why using the results of multiple audiology measures is necessary to obtain an accurate picture of the patient's auditory status and to discuss the importance of counseling and follow up in the patient care journey.

This is a picture of our lovely hospital, Phoenix Children's. And Phoenix Children's is the home of the largest pediatric audiology team in Arizona, providing highly specialized evidence based care for infants, children and teens with hearing and balance concerns. Today's presentation highlights the complexity, innovation and clinical reasoning involved in pediatric audiology practice while reminding us that behind every audiogram and every test is a child with a developing future. So our presenters today are Dr. Allie Sayer. Allie is an audiologist level two, Dr. Laura Marchman.

Laura is an audiologist level one. Lynn Eyde is our audiology manager. Paige Petersen is an audiologist level two and Deb Flynn is our lead audiologist. You all are in for a treat today. This is a wonderful collection of case studies.

Thank you. Okay, Allie, you are up for your presentation. Allie's presentation is Ella an unnecessary referral or a candidate for cochlear implants? Thank you for that introduction, Wendy. It's nice to be back again today.

I have enjoyed doing these the last couple times we presented. So Ella was first referred to us by an outside audiologist for a cochlear implant consultation with our ear, nose and throat department when she was seven years old. The concerns prompting her referral at that time were for poor progress in her speech and language despite regular attendance at therapy. Her family also reported concerns for poor performance in school and difficulty communicating at home. Ella is bilingual but prefers English while her parents are primarily Spanish speaking.

Her hearing loss was reported as moderate to moderately severe and was diagnosed at 6 months of age. Genetic testing had been completed but did not reveal a cause and she had not had imaging completed. She was fit with hearing aids just after one year of age.

The outside records sent for review at that evaluation were fairly limited but included this audiogram showing a moderate to moderately severe hearing loss. Bone conduction thresholds were not included, but her loss had been established as sensorineural in nature for many years. They completed functional gain testing using narrow bands of noise with her bilaterally aided thresholds here on the audiogram, noting that she gets good benefit but that she's likely missing high frequency phoneme information. They report that this is despite verification measures showing appropriate match to targets, but they didn't send that data for our review while Ella's speech difficulties were reported to be apparent at that clinical visit. ENT recommended further workup before considering cochlear implants since she

clearly meet candidacy criteria.

Our cochlear implant team has a really big emphasis on multidisciplinary collaboration and works very closely together, so it was recommended that either she returned to her managing audiologist for more testing or that we would be happy to have a second second opinion here and complete some of that testing for them.

Before we get to more about Ella, I wanted to take a moment to review current indications for cochlear implants in the pediatric population because it is rapidly evolving were first introduced in the pediatric population. It was for ages 2 and older with bilateral profound hearing loss, but as time went on candidacy criteria has widened to include younger children and those with more residual hearing. Current FDA indications do vary by the specific manufacturer, but in general candidacy now includes children as young as 7 months of age with bilateral severe to profound sensorineural hearing loss in children older than a year. One of the newest approved indications is for moderately severe to profound hearing loss in young pre linguistic children. Demonstration of poor progress in speech and language development is sufficient to demonstrate that the child has poor sufficient to demonstrate poor benefit from hearing aids.

But for older children with language it's also necessary to demonstrate that the child has poor word understanding scores. This varies by manufacturer as well, but one of the most strict is less than or equal to 20% where adult criteria is typically higher around 50%. 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 or only one earmates criteria or those who have better hearing thresholds with poor word understanding scores. Off label use of cochlear implants allows us to remain more progressive and inclusive in our candidacy considerations to support our patients and their families in

achieving their communication goals, we rely heavily on research and recommendations from professional organizations in making these recommendations.

One of my most frequently referenced sources is the American Cochlear Implant Alliance Task Force Guidelines for Determining Cochlear Implant Candidacy in children published in 2022. These guidelines are somewhat more progressive than the strict FDA criteria. Regardless of age, they recommend referring a child for a cochlear implant evaluation when audiometric thresholds are greater than or equal 70 decibels, word recognition scores are less than or equal to 50% correct and or there is a poor functional performance or limited progress in their speech and language development.

Looking back at the referral for Ella, at first glance the recommendation wasn't too far off base. She has a few thresholds greater than or equal to 70db and her pure tone averages are very near the 70db recommendation. They also clearly note that the patient is struggling both in her speech and language development and academically. However, the biggest factor in this case was the lack of speech testing. For a child who is seven years old and is able to communicate at least some in two languages.

Speech testing to confirm candidacy, both aided and unaided, is an absolute must. Here at our facility eight months later, they followed up an ENT and provided additional reports of a more recent evaluation with their outside audiologist. Unaided and aided thresholds were very similar, but this time they did report completing some speech testing. However, they only evaluated it in the bilateral aided condition with sentences in quiet at a loud, conversational level, nothing ear specific or unaided. They reported a score of 51% but also noted that her responses were difficult to score due to poor speech clarity and they suspected it could be possible that her articulation issues were influencing the poor score, not her actual receptive abilities.

Regardless, they still recommended proceeding with cochlear implant. They also included results of an evaluation with an SLP that were completed around the time of the original referral. They didn't do a

formal evaluation of her expressive and receptive language skills and rather focused only on auditory skills. They reported though a severe delay, noting that she has difficulty answering WH questions and that she would miss keywords and sentences frequently. The outcome of this visit was no different.

Our ENT recommended a more thorough evaluation either with her managing audiologists again or coming to see us for a second opinion.

Unfortunately, Ella was lost to follow up again for about 15 months before she finally came to see me for an evaluation. Now nine years old, parental report was very consistent with what they had told the ENT and what was in the outside records. But when I asked Ella, she denied any hearing difficulties and felt that her hearing aids worked well for her. She was in the fourth grade and was on an IEP with daily use of an FM system in the classroom. She confirmed that English was her preferred language and felt that she actually knows very little Spanish.

She was very cooperative and easy to work with and at least with access to lip reading she was able to follow directions and answer simple questions very well.

On that day, her otoscopic exam was unremarkable. She had type A tympanograms and globally absent ipsilateral acoustic reflexes. Testing showed a moderate to severe sensorineural hearing loss overall, consistent with the most recent outside records that had been reviewed. What I was most interested in though was her speech testing. Her speech was at times difficult to interpret, but she was very compliant and would repeat the words more than once if I asked her to to make sure that I understood her response correctly.

Her errors were generally very close to the target word with no instances of her missing the word in its entirety. Her speech recognition thresholds were attained at 70 decibels for both ears. Testing with PBKLIS was completed at the maximum output intensity for the audiometer, but she had no discomfort

with that volume. Even with the articulation issues. The scores were significantly better than what I would expect for someone coming to see me for a cochlear implant evaluation at 80 and 88%.

Seeing this, I was suspicious that her hearing aids may not be working appropriately or that they weren't programmed appropriately. So before any aided testing we got to work on her hearing aids.

She had not been seen by her audiologist in quite some time, so the hearing aids were in need of a little tlc. Ear molds were loose and the tubing was very hardened. But this was changed in office and we took some new impressions. The audiogram in the fitting software was outdated and showed an overall more mild hearing loss for both ears which was concerning. Her feedback manager had been run and was also very significantly reducing the output of the devices.

Frequency was also lowering was Also enabled where it really didn't need to be.

For all of my transfer patients, I like to get an idea of how their current programming looks. So after measuring an IECD for both of her ears, but before I changed any of her settings, I completed on ear speech mapping to DSL pediatric targets with her current settings to the thresholds that we had just measured that day. We use the VeriFit2 system in our clinic, so her prescriptive targets on this screen for average speech at 65 decibels are the little cross shapes in green and ideally that solid line should run as as closely as possible through those targets. As expected, based on her feedback test, she was programmed well below targets, especially in those higher frequencies. Her aided SII calculations for average conversational speech were only 51 and 54.

So we made some adjustments. A new feedback test was completed first to ensure that if any impact on the test. Sorry. To ensure that if there was any impact of that test on her gain, it would be reflected in the speech mapping. So we knew exactly what she hearing from the devices.

Then we made those programming changes to meet targets. And this is how her programming looked afterwards. She had much better approximation to prescriptive targets, especially in the higher frequencies. And her aided SIIs for average level speech were now 71 and 68. Ella reported that the hearing aids sounded louder and different, but that it was in a good way.

Her mom immediately reported feeling like Ella was hearing and understanding her better when they were talking in the office. We decided to try some aided testing next. Despite having very little time left in the appointment since we were rushing, we were only able to complete bilateral testing. But at this point I was already pretty convinced that she didn't need cochlear implants. So this was okay for the time being.

Her aided detection looked good across frequencies and her speech testing looked really good as well. With a whole word score of 80% and a phoneme score of 89% on the LNT Easy.

I reviewed with mom that at this point Ella is not a candidate for cochlear implants. The family was a little surprised as they had now been told for a few years that she needs cochlear implants. But with how quickly Ella demonstrated improved auditory skills in the office, they agreed to put off consideration for the time being. We plan to bring her back in a few months for monitoring and to expand aided testing to include ear specific testing. At that follow up visit, similar results were obtained for each ear by itself and family was reporting that overall she seemed to be Doing much better both at home and at school.

Ella has followed with me regularly every six months since that time. In general, she has made remarkable progress and her family is happy. There continue to be some communication difficulties at home, primarily due to their language barrier, but her articulation as a less familiar listener is notably improving at each visit, and family has resumed private speech therapy outside of school. School we've worked through speechless with increasing test difficulty, moving to the LNT hard and evaluating at a

softer speech level. Most recently we moved to an adult list with CNC words and she continues to do very well.

We've also added in speech and noise testing while she struggles more in noise than she does in quiet, overall her benefit from hearing aids is good and there's no current consideration for cochlear implantation. We'll continue to monitor her closely and adjust our plan if the need arises, especially since the etiology of her hearing loss is unknown and she still hasn't undergone imaging, so her risk for progression is unclear. But for the time being, Ella has been spared from an unnecessary surgery and family feels much better about how she's performing.

This case for me serves as a really good reminder of several points. First and foremost, the details matter in audiology. If her original audiologist had checked some of these small things sooner, it's possible that Ella would have been hearing better sooner and wouldn't have had such a severe delay in her auditory and speech and language skills in the first place. While I'll never know for certain because the records were incomplete, what I imagine caused the majority of problems for Ella was the feedback test, limiting her gain. Even though they report speech mapping was completed, if the feedback test was run afterwards, her actual gain would be poorer than the speech mapping shows.

This can be a big challenge to stay on top of, especially for young children who frequently outgrow their emails or their ear molds. But is such an important key factor in ensuring that hearing aids are truly set appropriately? Second, never assume, always test it. No test is complete on its own, and we rely heavily on a test battery because that's how we cross check that all of our results are accurate. It's very tempting to look at a child with this degree of hearing loss and assume that because they're not performing well, they may be a candidate for cochlear implants.

Even though unaided thresholds do just barely meet the newer referral criteria, they didn't complete any unaided speech testing. If they had, they may have questioned the need for referral. After seeing

scores of 80% or above. For me, this was an immediate red flag to suspect something was off in her hearing aid programming. Which leads me right to the third point.

Before anything else, double check that you didn't miss anything. Even well seasoned audiologists can make mistakes and it's very easy in a fast paced clinic, especially one full of noise and activity like our pediatric centers are, to miss something by questioning your own work first, you act as your own reviewer and mistakes be caught and rectified much sooner. If you're unsure, ask a colleague. See if there's anything you're missing. Especially in cases where we are considering surgical intervention for a child, it's really important to ensure that things are accurate.

Thank you so much for your time and attention. I'm going to hand things off to Dr. Laura Marchman who is going to talk to you on another other case entitled Cross Checks and Close Monitoring to Optimize Pediatric Outcomes. Thanks Ali, I appreciate it. So like Ali said, my case is really going to be focusing on the cross check principle and close monitoring of ccmv. I think this is a really interesting case where some cross check opportunities were missed, some monitoring protocols weren't followed, and as a result a child lost years of optimal auditory access.

And this particular case is pretty interesting because I think it's a really good learning opportunity for building systems that prevent this from happening to our patients in the future. So to start, I'm going to start talking about the crosscheck principle and what is it? I like to think of it like a puzzle, especially in pediatric audiology. All of our pieces or our tests that we use should fit together in a specific way to create a final product. If we don't use all of these pieces or they're not fitting together in a particular way that we expect our final picture isn't correct.

And in our case the final diagnosis or recommendations may not be correct. So it's important that if things don't align, we continue digging and we don't stop there. The cross check principle is essential and I really think that relying on one test to tell the whole story isn't something that we can do,

especially in pediatric audiology, as tests may be influenced by several different things such as attention, cognitive status, language, personal motive, among many other reasons. So relying on one test leads us, may lead us in the wrong direction. And the standard of care and audiology includes a collection of tests or a test battery using objective behavioral and functional measures can lead us to the right diagnosis, which of course is best practice.

And if things don't make sense. We need to continue to dig to figure out what part is missing, what can we add to the picture to find the direction that we need to go? We can do this in several different ways. Oh my slide's frozen. There we go.

We can do this in several different ways and multiple different tests that can be included in a test battery. And depending on the patient, we may choose some versus others. For example, for a particular patient you may use VRA tympanograms and otoacoustic emissions. But another one you choose tympanograms, otoacoustic emissions and an abr. Or maybe your air conduction results don't make sense with your present OEs.

So you check with inserts or TDH headphones to make things align in the way that you would expect. And there's several other ways that we can include these things with speech testing and verification, questionnaires. You know, the list goes on and on. And I think it's important that we choose a variety of these things to add to that puzzle.

Next, I'm going to talk a little bit about CCMV or congenital cytomegalovirus. This is actually the leading cause of birth defects and developmental disabilities in the United States. Points It's a common viral infection that often causes no harm to people, but when it's passed to the fetus during pregnancy, it can cause pregnancy loss or long term health complications. CCMV affects one in 200 births, with one in five of those to have long term health consequences. And for us as audiologists, especially pediatric audiologists, it's important because this is actually the leading cause of non genetic hearing loss.

The long term effects of CCNV can be several different things. For common examples we have developmental and motor delay, vision loss, seizures, calcifications within the brain, and of course hearing loss. I think it's important to note that hearing loss can be fluctuating or progressive, which is really important for us to think about when we're monitoring these kids with hearing loss.

When we're looking at the recommended monitoring schedule by the American Academy of Audiology, this is what they suggest. Every infant that has known or suspected CCMV should have a diagnostic evaluation by three months of age. This is even if they pass a newborn hearing screening. We need a baseline from there. Every three to six months until age one is recommended, then every six months until age three and then annually after or up until age six and likely past that point.

This is all with a caveat that there is stable hearing if there is a change in Hearing thresholds should be monitored even closer than the typical recommendation.

Some other considerations that we may use when looking at patients with CCMV are the amplification, again with a high likelihood of progression or fluctuation. I think it's important to continue to think ahead for these kids. So it would be beneficial to select hearing technology that fits a child's current thresholds, but could also accommodate a progression in the future. So if you have a hearing loss that is at the end of a fitting range, maybe you consider a technology that allows for progression rather than having to reorder or refit new technology sooner than you would expect for a child with a stable hearing loss.

It's also could be a consideration to start talking about cochlear implantation a little bit earlier than you might. Not necessarily as a recommendation, but as an education for parents sometimes if they start learning what could happen down the road. If you get to that point, they're a little bit more comfortable with moving forward with your recommendations if they know that that was something that could

happen all along. And then also vestibular evaluations for these kids. Lots of kids with CCMB have vestibular concerns, so adding that to our test battery should be considered.

So with that I'm going to move on to my patient Penelope. She is a 16 year old patient that transferred to me earlier this year. She has developmental delays and she is wearing bilateral Phonak M50s hearing aids. When she came to me, the information I had was pretty limited. I was able to see some prior records from Phoenix Children's in 2018 that showed her from a bear study that showed her right ear had mild hearing loss and her left ear had a mild sloping to severe hearing loss.

She was then sent to an outside facility where she was fit with hearing aids and managed up until she returned back to me.

And again, I didn't have any records. When she came back to me at this time, mom was able to share that she attends a school for children with special needs and she has an IEP and she receives speech, occupational and physical therapies. I was able to dig a little bit from the few records that I did have and I was able to see that she had suspected ccmb. So that was an interesting red flag to me. As we continued talking, I brought her back from the waiting room and I noticed immediately that the patient was not talking to me.

She wasn't looking at me and she wasn't able to answer any questions that I had for her directly. So I wasn't sure if what her language abilities were at the time if she was being really shy or how significant the reported developmental delays were. So I talked to mom a little bit more, and she pretty quickly brought to my attention that she felt Penelope had had a recent change in hearing. By recent. She reported about three years prior to when she was seeing me.

She said that when Penelope was home previously, she used to respond with her hearing aids on and even sometimes without devices. But that was not happening and had not been happening for the last

couple years. And now, even with her devices on, family had to talk really loudly or yell to get her attention. But then she did finish up by saying she was doing really well overall, and with support at school, she was doing great.

We began testing, and on this day, I actually had two testers with me, myself and a testicist. And mom was able to report to us when we asked at her prior audiologist office that she would just raise her hand in response to beeps. So with some visual cues, we modeled this for Penelope and she nodded as if she understood and we got to work. Prior to the audiometry, she did have type 8 and pantograms and completely absent OAEs. And as we started testing, she was very inconsistent.

I wasn't able to nail down any air conditioning conduction thresholds when we started at a level about 60 to 70, which with a mild hearing loss from a prior bear, should have been super threshold. And she had kind of a look, a body language look about her that I wasn't confident she was actually hearing the presentation levels that we were using. So we switched to high fives. The same thing wasn't happening. And so I decided to turn up the presentation levels quite a bit.

And quickly she had this aha look of oh, now I hear it. And she became pretty consistent. So once that that was the case, this audiogram here is what we found. And definitely quite a bit different from a mild hearing loss in the right ear and a mild to severe in the left ear. So at some point in time there had been a progression.

Um, I also think it's interesting to look at the word recognition testing that she had. Although she had some thresholds in the right ear, she had 0% word understanding at the levels of the audiometer and she had about 50% word understanding in the left earth. So we moved on to some hearing aid programming. And immediately I noticed that these were the thresholds that were entered into the software from her hearing aids. These were dated back to 2022, so likely the most recent audiogram that had been updated in these devices.

And again, pretty different thresholds, at least for the left ear compared to what I obtained that d. So I decided to run some hearing aid real ear measures initially with this left ear, it is under fit. So I was looking at a couple different things. I changed the probe tube, I checked the placement just in case that we didn't have, you know, a different barrier in the way. But sure enough, this was continued to be the output of the hearing aid. I also noticed at this time that the last time data logging was read on the devices was for the right ear, January of 2025, so one year prior, and for the left ear two years prior.

So my best guess is that the last time these have been connected with these settings was about two years of Penelope listening here. And again, I want to draw your attention back to the word recognition scores that I just highlighted because her right ear, while somewhat close to targets, that was the ear she had zero percent understanding in. And the left ear, which carries the weight for her speech understanding, was severely underfit. So to me I'm kind of thinking that she's essentially wearing an earplug and the one ear that that carries the weight for her language abilities.

So we of course made some programming. Well, first I think we should. Oopsies. We should talk about the consequences to underfit amplification for all patients. Limited access to sound also means limited access to education.

And for this particular patient, it also delays some future recommendations that we have for her, which in her case, or a cochlear implant evaluation. So for at least the last couple years since her hearing aids have been programmed, she's had pretty significant barriers in the way, not just cognitively but also audiologically with her hearing aids. So we made some changes and we were able to get her hearing aids closer to target. Of course, we were a little bit limited due to the severity of her hearing loss. We activated sound recover and immediately she started telling us a story.

She started looking, she started talking and mom said something and she turned and mom even went, oh my gosh. She hasn't turned to me while I've been talking in years. So that was exciting to see that pretty immediately, with some better programming, it made an immediate difference for her.

So what went sideways with this case? Where can we improve? I think there's a few different red flags in her particular case that demand action. First and foremost, mom reported pretty immediately to me that she felt there had been a change in hearing. And so that alone should would have prompted us to do some earlier testing.

In addition, she has a medical history of symptomatic ccmb, which should prompt at least audiologic monitoring annually. And it seems like maybe that had or had not happened. But I will say this case can be a little bit messy. And this is where cross checks really come into play here, because if you're not looking at this case with a really close lens, I could see how there'd be a couple misses. Mom reported a change in hearing, but she also doubled down by saying that Penelope was doing really well overall.

So it might be easy to just write off, oh, mom was concerned she's not listening, but she said she's doing well, so she's probably fine. Had we done an audiologic evaluation with a few different tests, we might have seen that that's not the case. Additionally, Penelope does answer yes or yeah to pretty much anything that is said to her. So it would be pretty easy if you brush past that, to think that she's understanding or maybe even write it off to her developmental delays. But, you know, both can be true.

She might have some delays, but it's also possible that she's compensating by not actually understanding. So that's on us to do our due diligence and do what's best for her. And this is why some of those cross check principles are so important. Had we also taken account her false positives that she was giving us at the beginning of testing, we might have recorded an audiogram that was much more mild and therefore more underfit hearing aids than what she actually needed. So using all of those pieces together helped us lead us to the right conclusion that she's had a pretty big change in hearing.

So we have seen a pretty big change already in the outcomes just by giving her more amplification. But I think we can still do more for her. So we are now in the middle of a cochlear implant evaluation for her family is really on board, and it looks like that might be the next best step for her. And she's also being referred for a vestibular evaluation to address some of those concerns. So the takeaways for me are that each patient is different and our care plans should be created for each individual.

Penelope is getting the correct care that she needs now and more adequate follow up. But it is possible that she would have different outcomes had this been done a little bit differently in the past. And so I think it's important to think about CCNV as a progressive hearing loss until it's proven otherwise. And close monitoring isn't Optional. It's standard of care.

Cross checks also catch what we would have otherwise missed. And I also think it's important to highlight documentation, how important documentation is for us as well for patients. I don't have the prior records to know if this patient came back for follow up or followed recommendations from the prior audiologist or if there was reasons the day that she was present that maybe her hearing aids weren't present and that's why they weren't connected to the software. It's all hard to say. But the more that we document, the more that we can provide better recommendations as well as anyone else that may pick up in this case.

And with that I will move on to Dr. Lynn Eyde and she's going to tell us a little bit more about her interesting case case and hers is titled the Hearing Loss that Lessons from a Pediatric Case. All right, thank you, Laura. I appreciate that. Well, today I'm going to be talking about something that a lot of us here on this team are very familiar with as well as I'm sure a lot of people who are joining today have encountered a patient who may not be reliable in their responses. So let me tell you a little bit about Maria.

Maria is a 12 year old female. I did not meet Maria on her first visit here. She saw a different audiologist and I'll explain that in a little bit. But the initial chart review when we were checking the appointment the day of, we saw that Maria had been referred for failed hearing screenings. From what was gathered from just the pediatrician's note.

There had been several failed hearing screenings across the pediatrician's offices as well as at school of note in there are some of these dots that you see on here. There were some mental health concerns that the mother had brought up along the lines, as well as the mother was concerned about possible adhd, there was a family history of that. So she had been through that before. So that evaluation had not yet been completed. But the mom had brought up those concerns.

The patient had vision impairment. She did not wear glasses or was not compliant with wearing glasses. I think might have been the case. Her performance in school was also poor. She was not doing well.

I don't remember exactly if she had failed out of some grades and they were talking about summer school or holding her back. Also, she did have a significant family history of hearing loss. She had a younger brother who was identified with a mild sensory neural hearing loss. Moss from from an office here in in Phoenix. So Maria's first Phoenix Children's audiology visit.

She was Referred for pediatric hearing evaluation. For us, that's just a diagnostic hearing evaluation. And she was accompanied by her mother and her grandmother. The patient and the parent reported the following things. So we already know her initial case history or.

Or, you know, the review of her medical record showed those things that I just previously talked about. But the mother and Maria both talked about the failed hearing screenings at school and one at the pediatrician, an area that we weren't really able to nail down. And I think it was just lost as far as with

the mom between multiple siblings and appointments. But she may have had a previous hearing test at the same ear, nose, and throat office where the brother was diagnosed. But mom couldn't necessarily recall those results whether they were normal or abnormal because it had been several years.

So the failed hearing screenings was not new for this family. The patient also described an intermittent tinnitus, non bothersome. It sounded like it came and went less than probably 10 seconds. But she did report that dizziness. She reported when she stood up too fast, that she would become dizzy.

But the patient herself actually denied hearing concerns. And for a lot of us here, when the patient is denying it, that kind of seems like, oh, we might be smooth sailing. If the patient isn't experiencing that, then we might be. Okay, so let's look at her first kind of start of hearing. I pushed too many times.

Sorry about that. All right, so right away, and I should have put this in there, but autoscopy was clear. There was nothing in our way as far as us performing the test that day. We got type A tympanograms in both ears, and acoustic reflexes were present at a screening level in the ipsilateral condition for both ears. So we're off to a good start here.

The concerns for Maria having fluid or pressure have gone away. We're not concerned about that. So now we're going to move on and look at her distortion product, otoacoustic emissions. Here we see a lot of different ages of kids and teens, as Wendy was saying earlier. And you've seen with the other cases, typically this is our order of testing, if we can obtain it.

So we want to have the different parts of the test battery to help kind of clue us into what we're going in to before we get into the booth, if we can, because that does help us, you know, figure out the approach that we're going to take. So this patient, again, we're off to a super good start. Our normal temps, present OEs, present reflexes. We're thinking, okay, easy. Patient isn't concerned.

We're not concerned either. So we're going to move on to the diagnostic testing in the booth now. And this is where it starts to become unreliable. And this is the part that many of us are familiar with. And we do see this especially kind of around this age.

You know, a kid hasn't had a failed hearing screening and then for the last couple years they do, and now we start to become unreliable when we go into the booth. So Maria would not reliably respond during speech reception testing. She was giving half word answers and they ended up being at what we determined to be super threshold response levels. She, you know, we were trying to mix it up. At that point.

The audiologist switched it up, went to do tonal stimuli just to see if we could get any other sort of different response from her. She again, was not giving us consistent responses. They were quite loud and she was not responding. And of note, I mean, she carried on a normal conversation all the way back from the waiting room and throughout the beginning of the appointment. And then once she was into the more the hearing test that I think she might have been more familiar with, she was not giving us reliable responses.

She when the examiner or the audiologist would go into the booth and talk with her, whether she had inserts on or super oral, the patient would answer the questions inside the booth with the headphones on, talking to the audiologist face to face. But when we would go out to the other side of the booth to actually try to like administer the test, she wouldn't give us a consistent response. We tried all the different strategies to try to obtain consistent responses. Giving her an out, switching the headphones, asking her to count the beeps, saying yes or no. She would not budge.

And at that point, you know, we're starting to think now, and Laura talked about this too, the crosscheck principle, we rely very heavily on that, especially working with pediatrics. I know it pertains to all ages, but no single test should be used alone. If we used her behavioral results alone, we would have thought

that she had a pretty significant hearing loss or auditor neuropathy or something to that regard. With her inconsistency, this crosscheck principle, as we know, will allow us the ability to compare subjective and objective results. We really should be using this for adults and pediatrics to help prevent misdiagnoses.

And we see that as we continue on with this, that using OAS is a really powerful tool, especially in these cases where you're getting inconsistent responses because you can suss out who maybe has a hearing loss, and maybe who doesn't?

Maria's subjective responses were very inconsistent, and we couldn't get, you know, they. They weren't consistent with what we had for the objective results. So the testing at that point was discontinued because we're not going to write down results that are abnormal, that are unreliable, because that just is confusing to the family, the next audiologist, the ear, nose, and throat, whoever they're going to see next. So the testing at that point was discontinued.

So the audiologist reviewed the results, you know, and counseled Maria, her mother, and her grandmother. There were several inconsistencies with the testing and her and her responses, and that was reviewed. You know, each part of the test was reviewed with the mom and the grandmother and the patient. The mother and grandmother were both skeptical. The grandmother had made a comment like, if Maria can't hear, if says she can't hear, then she can't hear.

So there are some skepticism there about maybe the testing. Or, you know, if the patient says it's true, then it's true. So we had two options. The mother was given and the patient ultimately were given the option. We could come back on another day, try again, see if that works better, or a different time for Maria when she's maybe feeling more up for it.

Or we could do the brainstem auditory evoked response study or a bear. And that's the same as an ABR if we just call them bears here. So we gave them the option rather than going to under anesthesia, let's have her come back for a natural sleep. She's at an age where she'll likely participate in that, and it should be fairly easy. The mother did elect to return for the ABR in the office, and that is when I met the patient.

So the audiologist who, you know, did the testing, we talked about this case after because she thought that the moment would likely be calling to be upset that they weren't able to get the test done. So we decided that if we could get them in quickly and that I could touch base with the family and do the abr, that we could hopefully avoid me having to call them after because I could just handle it there in the office rather than having a parent who is upset about results or the lack of results that they may have gotten. So we saw them back two weeks later. It was just Maria and her mother at that point. They came back for that ABR in the office.

Since the visit two weeks prior, she had actually seen an educational audiologist within her school district. The mom did have the results on her phone and did allow me to look at them. The results were inconclusive. And looking at the report again, it was a similar, it was a similar pattern that we saw half word answers or rhyming words. I'm making up words now, but if the word was baseball, she said football or if the word was mike, she said bike.

So those are the examples that the mom had provided me at that point. Looking at the note, there were some, they had obtained some behavioral thresholds, but again, they didn't. Nothing really added up. And the audiologist was definitely clued in like, hey, there's something, something strange here. So that audiologist had recommended that she return to see us for further evaluation and also to see in your nose and throat for her tinnitus because at that point she had reported that the tinnitus was now most of the time in both ears and it was more noticeable while at school.

The other things were still true. The the mother had talked about the ADHD evaluation as well as talked about her school performance. I didn't put those in there because they were, you know, there's nothing. I just put the things that were new in her case history which was an increase or a notice, more noticeability of her tinnitus. I took a lot of time, more so than I probably do, for an ABR to explain how the test works, what was going to be done to try to get me some support for after.

If the test doesn't indicate normal hearing sensitivity. I wanted to be sure that I had really explained the test and that they understood how it worked, what I was looking for, why she had to be relaxed, all of the above. Just trying to counsel ahead, to prepare for the counseling after to buy me some support or credibility. I'm not sure which of those what I was looking for, but really trying to make sure that they were prepared. So the hearing test results were this.

Now again, she had been seen a couple weeks prior. So I didn't repeat tympanometry and the room I was in and didn't have a tympanometer, I could have gone out and done that. The distortion product acoustic emissions were present bilaterally. Unfortunately, 8,000 Hz did look reduced or absent in comparison to the previous results. I think it was a fit issue.

I tried several different tips. I even changed the probe TIF of the actual OAE cable, but unfortunately it didn't get better. So that is kind of frustrating because you really want it to be tied up with a nice bow and everything to line up. But we Will, we will follow her back on that. So here I'm just going to show you her ABR results.

So we will see here that. I'm so sorry, my computer went minimized. We will see here that I did or I did a click in both ears, the right and the left ear. In this case, that click that I did at 20 corrected to 15 decibels estimated. So that was nice.

That was beautiful. It was robust. No concerns there. As far as right away I'm like, great. So I switched over to do the ASSR just for the sake of time and I, I tested her at a level that is lower than we normally would take the ASSR kind of to prove the point home.

The ASSR was really fast. It did not take me much time at all, which is really encouraging for the fact that she does have very normal hearing. So this is, this is one of those cases that probably most of us have been faced with where sometimes telling families that hearing is normal is not what they want to hear because they've been waiting for you to tell them that something is abnormal with their hearing because it's going to explain everything and then it's going to fix the other things that they're concerned about.

So I did review the results with Maria and her, her and her mother. The test results again were consistent with normal hearing sensitivity in both ears. Right away, Maria, you could tell, was disappointed. Her shoulders dropped, her head dropped. She was, I could tell, maybe frustrated.

The mother definitely her expression somewhat changed too. She expressed understanding of the results, yet she brought up the confusion of the inconsistencies from the behavioral responses, specifically the one from when we saw her the first time as well as with the educational audiologist. She didn't understand how she could have normal hearing sensitivity and that not translate to the behavioral testing that we did. So she wanted to know why that didn't line up. And so one of the things that we talked about was her confusion that she explained about the inconsistent results.

And I, this is where I sort of started to put my foot in my mouth because I, you know, I hadn't seen her for the behavioral testing, which makes it a little bit challenging because I didn't see, you know, I can read the notes but if you're not in the room, it's hard to know. And so I talked to the mom about, you know, generally when we see a patient who is giving half word answers or they're giving inconsistent responses that we think that they may not be fully participating to their full, you know, capabilities and

right away the mom was very. I could tell that was not the thing to say, because she was like, she would never fake that she would not. And so then I quickly had to say, you know, I. I understand. I said I wasn't there to see it, and so I can't speak to it necessarily.

But when I see that in a note, that does bring some concern that the patient may not be either understanding the test or that they're not willing to fully participate. And so I said, the good news that this will sort of help us is that she does have normal hearing, and then we can focus on the other things to try to help her and to support her. The mom brought up the school, you know, the school concerns about the poor school performance, also about how she has not necessarily got the ADHD evaluation completed, but she was kind of on the hunt for how to get that done. So I talked to her at that point. You know, I asked if we ever had a neuropsychology evaluation.

And I explained to her what that would help with or support her with. And so I kind of encouraged her to talk to the pediatrician about the neuropsychology evaluation as well as I put that in my note. One thing she sort of mentioned, as we were passing or we were at the end of the appointment, she said, well, what about auditory processing disorder? And I said, well, that is something that we do here. But I said, in order for us to do that type of testing, Maria has to participate reliably and consistently.

We can't until we can get a behavioral test on her in the booth. That's not something that I would recommend at this point. So the mom understood that. And again, with that 8,000 hertz and the family history of hearing loss, I did say that they should come back in a year or sooner if there's concerns for her. So Maria was.

Was a fun one. So I just want to say thank you, and I'm going to turn it over to Dr. Paige Petersen, who is going to talk to you about a wonderful, straightforward case or not, promoting hearing aid adherence in pediatric patients. All right, thank you, Lynn, for the introduction. And like she said, I'll present. I'll be presenting what I thought was going to be a straightforward case.

Let's see here. All right, so you can read through the objectives here for some background on this case. When looking over the past year and trying to decide on a case to present, one topic kept coming back up for me. And it's because I could think of this one case example of a situation that I've been in multiple times over my career that, although is seemingly straightforward, can be the most difficult to manage and that's when your patient just won't wear their hearing aids. And so today I wanted to present a case example of a patient with low hearing aid adherence and the ways that we as providers can tackle the problem.

So hearing aid adherence is defined as the patient and family's active and empowered choice to integrate hearing aids into their lifestyle. It includes consistent daily use of the hearing aids, attending follow up appointments, maintaining the devices and the ability to troubleshoot them. Well, general patient or patient and parent engagement, and other factors like participating in early intervention services. We can measure it in several ways, most commonly where we look at data logging in programming software. But assessing other factors like whether or not patients are attending their follow up appointments, are they maintaining the device as well, and whether they're enrolled and continue to receive their EI services can tell us a little bit about patient and families adherence in general.

So hearing aid adherence matters for several reasons. We know that early auditory access is crucial for speech and language development, academic outcomes, and social and emotional development. Inconsistent hearing aid use reduces overall benefit and loss to follow up delays intervention and monitoring. Studies show that especially before school age, children are often not wearing their hearing aids as much as recommended.

The reasons that patients don't wear their hearing aids consistently are often multifactorial. Studies have shown several common themes. It's important when we see patients that we use big picture

thinking when considering the reasons that our patients might not be wearing their hearing aids. At least for me, I don't think it's ever been the case that a patient or family just doesn't want to follow recommendations. There's often something bigger going on.

Low hearing aid adherence is often more of a symptom of a family experiencing some sort of barrier. Common reasons for low hearing aid adherence include hearing aid retention and comfort problems. This includes things like behavioral issues with children just pulling the hearing aids out constantly, as well as physical comfort issues and sound related aversions. School and day care factors include things like whether or not the children have good support from daycare providers and teachers during the day and do these professionals know what to do with the hearing aids. Parents also report difficulty with the emotional adjustment after receiving a diagnosis of hearing loss in their child and this can be a big barrier to seeking prompt follow up.

Something we often see as a barrier, especially in children with lesser degrees of hearing loss, are problems related to the parental beliefs or perceptions of benefits from the hearing aids. And I Think this really speaks to the fact that especially with lesser degrees of hearing loss, it can be hard for families to really see the deficit at home. And then there are also barriers to adherence related to socioeconomic, economic and health equity factors that you can see listed. And these are often more systemic challenges that can be very difficult to address.

So that brings us to our case example. This is Mateo. Mateo presented following failing his newborn hearing screening. He had an uncomplicated birth history, no family history of hearing loss. We attempted multiple bear studies on him in infancy, but they were all typically complicated by poor sleep.

And ultimately he required a bear under anesthesia, which occurred at 8 months of age and revealed this estimated audiogram. You can see it had a mild to moderate sensorineural hearing loss. We recommended bilateral hearing aids and ENT console, ophthalmology and genetics consultants and a

referral to the Arizona Early Intervention Program. Now, seeing this diagnosis, I would typically expect a fairly straightforward path to intervention and a favorable clinical outcome. But unfortunately, as we will see, that was not the case for this kiddo.

So this is where the path becomes windy for Mateo. You can see that he received his bear studies in infancy. He did return for the hearing aid fitting. We typically recommend a one month follow up up after the fitting and unfortunately he missed that appointment and then was lost to follow up for three and a half years. that time, his family called the clinic because the hearing aids were both broken.

He was seen for repair and for ear mold impressions. And we recommended a fuller appointment to be able to monitor his hearing and verify the devices.

So he did end up coming back for that recommended follow follow up, but the family forgot to bring the hearing aids to that appointment. Unfortunately, at that time, significant speech and language delays were noted and Mateo had reportedly never received any early intervention services or speech therapy services. He continued to show a stable, mild to moderate sensorineural hearing loss. We then saw him a few months later with the hearing aids to verify the devices, at which time his data logging was only 0.1 hours a day. We wanted to monitor him closely, so we saw him back a few months later.

And at that time he was wearing the hearing aids at school, per their report. He still didn't have any support services at school, but his data logging had increased to 3.3 hours a day. Unfortunately, he missed the next follow up appointment, which was intended to check his data logging again and provide further counseling. And then he was lost to follow up for another two years. So he finally showed back up at the age of seven after being referred by his pediatrician and reporting broken hearing aids.

Again, this was my first visit with Mateo. At this point he's a 7 year old first grade student with essentially no previous meaningful hearing aid use. His mom reported that he doesn't like the hearing

aids and they stopped forcing him to wear them at home. He's in the care of his mom, grandma, and sometimes his dad. His dad or his grandma have never been to any of his audiology appointments and they believe he hears fine.

They also see him reacting to sounds at home and so they believe he hears fine. He performs at grade level. Per his mom's report, she had some speech concerns and she couldn't really understand his speech. I couldn't understand his speech really either. But otherwise she felt that he was doing pretty well overall.

He reported that he doesn't like the hearing aids because they were annoying, but that he can't hear his teachers or his friends or his parents. He had no IEP or 504 accommodations and his teachers at this point didn't even really know that he needed hearing aids at all. So this is the point where you kind of really wonder, like, how did we get here and what can we do moving forward to get him back on track?

So studies show that parents do report several common facilitators to hearing aid adherence. These include support from professionals, particularly home visit professionals. So that would be essentially our early intervention providers. They also want information on options for hearing aid retention, want support in understanding why their child needs to wear hearing aids, and support in finding ways to get hearing aids in and keep them in, as well as practical device management strategies. They also want to be more connected to other parents of children with hearing loss for advice and encouragement.

So there are many things that we can do as providers to address barriers. We can provide ongoing counseling and parent coaching. I often have families sit in the booth and perform aided and unaided testing in the sound field or do aided versus unaided speech perception testing to objectively show differences in performance to families. You can also use hearing loss simulators. We can provide help with emotional adjustment by normalizing a family's response to a hearing loss diagnosis and modify the ways in which we're giving information to families to acknowledge the fact that they often have a

hard time soaking in information when faced with an emotional response from a new diagnosis.

We can address retention issues by offering things like pilot caps and toupee tape and also make sure to address issues like acoustic feedback by making sure that we're staying on top of timely ear molds and using feedback management when appropriate to increase wear time, we can encourage families to use positive reinforcement systems, have short wear time goals that gradually increase increase starting with predictable routines and have frequent early follow up visits. One of our biggest tools that we use is data logging, and data logging can be very helpful if used constructively. I've had my share fair a fair share of families that come in with almost the sense of shame because they know that the data logging isn't going to show what I want it to. But I like to encourage families that data logging gives us a starting point to talk about what the barriers to hearing aid use are when those barriers are happening, and to talk about how we can problem solve habits and routines to get to a point of increased use. It's also one tangible way that we can really celebrate progress with families.

To address school barriers, it's really important that we open up the channel of communication between us as the clinical audiologist and the educational audiology or special education team. Socioeconomic and health equity factors are often more systemic require often more systemic changes, but things like reminder systems, providing phone calls especially early on, making sure that we have interpreter access, transportation assistance programs and social work resources can be helpful.

So one of the biggest ways that we can move the needle on improving hearing aid adherence is by partnering with community programs in schools. Our early intervention providers and school based teams can make all the difference in ensuring good adherence. Early intervention providers can help reinforce daily use, provide consistent caregiver coaching, and they often see barriers that we don't get to see as the clinical audiologist. Similarly, our school based teams can help encourage daily device use, support classroom listening, build advocacy in students, and communicate any device related

problems that may need some troubleshooting. From our end, our role in all of this as the clinical audiologist is to make sure that we communicate well with our early intervention providers and school teams.

Teams share what information that we have about the family's communication goals and device information as well as provide troubleshooting support. Families also greatly benefit from being connected with community support resources like Hands and Voices and other parent to parent mentoring programs. In addition, there have been times when having continuity of care with a patient's pediatrician has been really helpful, especially for patients who are just lost to follow up.

So the role of counseling the point of this slide really is that, you know, figuring out these barriers and addressing them is really impossible unless we are using good counseling techniques. It's hard to really know what the barriers are without asking open ended questions and using some of these counseling micro skills. It's Also difficult to get patient buy in or create behavior change in families if they don't really feel like they're getting a say in the intervention. So using shared decision making can especially be helpful when working on increasing wear time with kids and teens.

So going back to our friend Mateo, he really had a lot of barriers spanning various domains. He didn't meet his 1, 2, 3 or 1 3, 6 goals for intervention. He had a mild to moderate degree of hearing loss that was really difficult for his parents to see subjectively. Even though they did note his speech and language concerns, they didn't really put two and two together. There seemed to be a significant significant parental like knowledge gap and a lack of awareness of the long term language risk to poor hearing aid adherence.

He had extensive periods of loss to follow up and significant speech and language delays. He said that the hearing aids were annoying, the family reported transportation issues as being a barrier and we also just had an overall lack of community partners. He hadn't ever received early intervention services

and school was completely unaware that he needed the hearing hearing aids.

So here is what we did to address some of Mateo's barriers. So his hearing aids are out of warranty and broken so we updated them. He was also way overdue for ear molds so we updated those and it ended up that what was annoying to him was actually acoustic feedback and this was easily addressed with new secure fitting ear molds as well as a feedback test temporarily until we got the new ear molds. We decided to monitor his device use closely over the next weeks few year and I was able to check in with his mom a few times in between our appointments. We use data logging constructively and even got to the point where he was excited to connect the devices to the software at our follow ups because he wanted to see if his hard work was paying off in loggable use time.

He has several caregivers at home and so and they seem to have like various perceptions of the importance of hearing aid use and so I used the hearing hearing loss simulator in the verifit and had his mom record our conversation to share with his family members. We also used unaided and aided speech intelligibility index values as a way to help his family better understand his audibility with and without the hearing aids. I also recommended a speech and language evaluation as a way for his family to get a more objective, realistic picture of his current speech and language skills. I had the family sign a release of information to communicate with school and recommended that the family request evaluation for a 504 plan or IEP. Matteo also reported that he didn't know any other kids with hearing loss or kids who had hearing aids.

And so I offered to connect the family with another family of a child with hearing loss and gave them resources for hands and voices. I also offered them a social work consult to assess for barriers to care and discuss transportation resources which they were open to.

So this is where we are currently. Contact with the school has been ongoing about every month or so. We'll email back and forth. A 504 plan has been established and speech and language evaluations are

in progress. School reports that Mateo is wearing his hearing aids full time at school.

He did lose one of his hearing aids but the family promptly contacted clinic for the one time replacement basement. He hasn't missed any scheduled follow ups. Data logging recent Most recently showed 7.8 hours a day and when he found that out he was so excited. Of course it's not quite where we would want it to be, but it's really great to see him demonstrating like such a growth mindset. Mateo's mom reports that Mateo is wearing the hearing aids more even at home and that she notices positive behavioral changes.

And Mateo is now saying that he likes his hearing aids and is hearing his friends better.

So Key Takeaways Takeaways from this case are that there are many barriers to consistent hearing aid use in pediatric patients. Some barriers are easy to modify and others are more challenging to solve and more ongoing. Having a close partnership between providers, families and community partners can lead to the best outcomes. And it's also important that we acknowledge that each family has unique barriers to hearing aid adherence and it's crucial to use evidence based counseling techniques to really find out what those barriers are and identify solutions. So with that I would just like to say thank you for the opportunity to present today and I will hand it off to Dr. Deb Flynn here who will be presenting Starting Over Navigating Bimodal Care with Without a Map.

Thank you, thank you Paige for that lovely presentation in this next case titled Starting Over Navigating Bimodal Care Without a Map. The topic for today is getting on the right track in untangling complex pediatric cases. This next case is going to highlight the complexity of a bimodal patient that's coming to our clinic without current medical record records. So let's take a look at this kiddo.

And untangle some of the situations or obstacles that arose during her first two appointments. To begin, she is a 14 year old female with bilateral sensorineural hearing loss who has a hearing aid for her right

ear and a cochlear implant for her left ear. Her primary mode of communication is listening and spoken language. Language. She has severe articulation errors and at times is difficult to understand.

A little Background History she was diagnosed at our clinic but had to transfer care to another facility due to insurance. She then reappeared 10 years later or more than 10 years later and there's a lot of missing information that needed to obtain in a short period of time.

In situ.

In situations like this I like to start the appointment off by taking by talking to both the parent and the child if appropriate in order to obtain a case history. The first obstacle that I ran into was that she did not want to discuss her history nor did she want her mother to discuss it. She became quite upset early the in on and so her mother and I agreed to discuss it at a later date by phone. So obstacle number two. Her mother provided very limited neonatal history and was fairly brief in her answers regarding the last 10 years of audiological history.

Much of her neonatal history was obtained from the limited records that we had. Records from our facility indicated that she was born full time term, but there were significant complications. There was a placental abruption that resulted in emergency C section at a regional hospital. Her care was quickly transferred to a level 3 NICU. She was Diag diagnosed with HIE and required a cool cap.

She had an umbilical venal venous catheter placed that leaked into her abdomen causing some cardiac issues due to to fluid built up around her lungs. She had an MRI that revealed an ischemic infarct in the left temporal occipital cortex and prior to discharge she did not pass a newborn hearing screening.

Her journey with us began at 2 months of age when she was sent for a diagnostic abr. Results from this evaluation were limited, limited but demonstrated bilateral sensorineural hearing loss. She was

scheduled immediately for a follow up ABR but unfortunately did not return for another nine months. Behavioral testing at that time could not rule out a mild precipitously sloping to severe hearing loss. Another ABR was recommended and the hearing loss was confirmed.

Following this test she was scheduled for hearing aid fit.

Due to insurance, her care was transferred to another facility. Her mother indicated that she was indeed fit with hearing aids sometime after diagnosis, but she was unable to recall exactly when. Her mother reported that she wore her hearing aids consistently until about five years ago when her hearing decreased and she was implanted in the left earth. She and her mother indicated that she does not Wear her cochlear implant and data logging confirmed this.

On the day we met, I completed testing or as much as I could. Results indicated type A tympanograms, bilaterally absent ipsilateral acoustic reflexes except for at 500 Hz in the right ear and a normal precipitously sloping to profound hearing loss. Left greater than right. Note that she has significant low frequency residual hearing in her left ear. Even after cochlear implantation.

Her speech discrimination ability for the right ear was 36% and testing could not be completed in her left ear because she came upset by the fact that she couldn't understand the words.

Obstacle number three. The big question what's best to treat this type of hearing loss? As all of us know, there's no easy way to provide exceptional treatment for this type or degree of hearing loss. Providing appropriate amplification for precipitously sloping loss is always a challenge. Prior to the new cochlear implant guidelines, there was little we could do for children with this degree of hearing loss other than provide gain at 550 and 1,000 Hertz and then add some sound recover or speech rescue.

More to come on the upcoming slides regarding CI guidelines. So Aida testing was completed with the incoming settings from her previous facility and demonstrated fair benefit with her cochlear implant, which is actually quite surprising given that she was not working wearing it and poor benefit with her hearing aid. Obstacle number four. Aided speech testing was was not completed because once again she became upset by the fact that she could not understand many of the words that were presented. It was more important for me to gain her trust and to push her into getting the information needed.

There's always another day.

Obstacle number number five. She is not wearing her cochlear implant. Overall, if she wore her cochlear implant, her combined aided detection is fairly good. She obtained high frequency gains from her cochlear implant and the more natural pitch, porosity, intonation and sound quality from the low frequencies in her hearing aids. So the question is, how do we convince her to wear her cochlear implant?

All right, back to our appointment. Due to her emotional state, I pivoted and decided to go check her hearing aid given that this is her preferred device. Incoming settings indicated gain over targets in the low frequencies and under targets in the mid and high frequencies. Note that her SII score is 34. Obstacle number six.

I attempted to complete RECDS, but she was emotionally drained and would not allow placement of the Probe tube adjustments were made to her hearing aids using averaged recds and sound recover was enabled. Unfortunately, she did not tolerate the changes. It was noted that her current ear molds did not fit well and did not have a vent. Given the normal low frequency thresholds. We decided to remake her ear mold and make the changes to her hearing aids at the next appointment.

On this date, her hearing aids were set back to the incoming settings due to tolerance.

At our follow up appointment, I fit her ear mold and reprogrammed her hearing aid. Obtaining RACDs was not an issue on this day. I'm not sure if it was event or just a new day, but she tolerated the changes as well and her SII scores slightly improved to 43.

So I checked her CI map which we did not get to at her last appointment.

Data logging was extremely low and she confessed that she takes the cochlear implant to school school but removes it in the morning session and places in her backpack. She had a Cochlear N7 nucleus sound processor and it had an acoustic receiver and a double dome. It was noted that the receiver was too short and did not reach her ear canal. In addition, the tip of the implant that holds the acoustic component was cracked, thus making it difficult to replace that component with a more appropriate option. To our benefit, her cochlear implant was out of warranty.

This particular processor is now obsolete and cannot be repaired and parts are no longer available. This gave us an opportunity to discuss options.

Her primary complaints with her cochlear implant were size and not being able to hide it. Second was sound quality. She said it sounded like a robot. And third, retention. She plays sports and does not like how floppy it is.

Given her residual hearing, we have a couple of options. We could replace her current Device with an N8 and use an electrical signal only. We could replace it with an N8 and get a better fitting acoustic component or we could try a Kanso. Each option was discussed based on her three complaints. We also had a very long discussion about Roger connectivity as it is a little bit more cumbersome connecting Roger to a Kanso processor.

After a long discussion and weeding through the emotional turmoil, we decided as a team, team, mother, child and provider that we would rather her select a device that she feels most comfortable wearing than a device that'll sit in her backpack as it currently does. In the end, she elected to order a Kanso with the knowledge that she could exchange it within 90 days if it does not work out for her.

Interestingly, despite the fact that she does not wear her cochlear implant. Her mother inquired about a second cochlear implant for the other ear. Her previous audiologist had discussed this prior to the transfer, but her mother was hesitant due to the fact that she didn't wear her first device. I reviewed the CI pediatric CI criteria with her which included audiometric thresholds greater than 70, speech discrimination poorer than 50 and challenges that affect quality of life. She currently meets all of these criteria criteria.

However, we decided to table the conversation until she started to wear her left processor. This case is ongoing. The upgrade process took several months and she just received her KANSO after a three month wait. Her follow up appointment for monitoring has been rescheduled three times. So unfortunately I don't have any audiological data around the Kanzo.

But during a phone conversation with her mother she indicated that she is wearing it more than she ever wore her N7 prostate I'll take this as a win for now, but I also know that we most likely have a lot of ongoing work to do to optimize her hearing experience.

So some of the take homes from this case the importance of managing patient emotions, being flexible, bringing patients back on a different day and being sensitive to how they're feeling. Number two, thinking outside the box, switching processor styles in an attempt to improve device use use and number three counseling. Lots of counseling, particularly on device use.

Thank you for your attention and we'll all join now for any questions that you have.

Okay. Okay. Hello everyone. We have a couple questions. If there are any others of you out there who has a question to ask, just please put it in the Q and A tab tab.

First question we have is from Jesse and Allie. I think this question is for you. The question is how do you decide what word list to use when completed? Aided speech testing as best we can, we try to follow the pediatric minimum speech test battery that pretty clearly walks you through progressions of tests, what you need to do, what should be included and when to move on to the next list. You know there are of course times where we have to deviate a little bit based on the kids language or abilities, but we try to stick to that when we can.

That makes sense. Thank you. Okay, the next question is from Chris and Paige. I think this one is for you. The question is how much use time do you typically recommend for pediatric patients?

That's a good question. Of course we would like our patients to be wearing their devices during all of their waking hours except when they can get wet or if they're around loud noises. But a Lot of times for families, they want something that's more concrete than that. And so I most often will cite the outcomes of children with hearing loss study and talk to families about the importance of getting to at least 10 hours of day of use for the best speech and language outcomes. That is great.

Thank you. Okay, next question is from Joe and Lynn. I think this one is for you. The question is what testing methods are used when you suspect someone is not responding reliably?

Yeah, there's a lot of different things that you can do. Some of the methods that we use here that I know, it would be like count the, the beeps. So you'll have ask the patient to, you know, count the beeps when they hear. And oftentimes someone who's malingering or faking will say zero every time they hear a string of pulsed warble tones. So that's usually your tell that they, they heard you.

Same with like the yes or no. You know, say yes when you hear it, no when you don't hear it. Again, I know that each of us probably have our own spin, you know, spin on those things. But usually those things things are usually we can get someone to start responding or, or, or get them on their mgr with those tricks. Okay, that makes sense to the rest of the group.

Do you guys have any things which, what are your go tos? If they were not mentioned by Lynn, I think she pretty much nailed them. Totally agree.

Okay, Deb, are you thinking anything? I use the count the beeps. Okay, great. Okay, the next question is from Pat and Deb. This question is for you.

What do you think the biggest challenge to overcome at her next visit will be?

There are many, actually. I would say probably her emotional state. She is extremely anxious and she wants to do well on testing, but it's difficult for her. So I think, you know, being sensitive to that and making her feel as comfortable as possible so that we can get the information that we need. So I think, you know, detection levels with cochlear implants are truly just detection levels.

It doesn't mean that she's actually understanding with her cochlear implants implant and so focusing on, you know, speech discrimination and of course, counseling. Counseling on the importance of device use and really trying to get her over the hump of wearing it and learning to hear with it because she's really, she really hasn't learned to hear with it. So I'd say those are probably the main tasks. That is a lot. I have a question.

Is she showing up for her Appointment? Treatments? No, unfortunately. Lots of calls to mom, but. But they haven't actually come back to the clinic, so we hope to be persistent.

We have a cochlear implant coordinator who is absolutely fantastic. And every once in a while I send her a message saying, hey, can you check in with the family again? I happen to have a cancellation or an opening. Can we see if we can get her back in? So hopefully we'll be able to do that.

I. I suspect that this might be an ongoing issue, but we'll keep trying. Hopefully she will. Hopefully her attendance will improve. Okay, the next question is from Alex. And I think this is for you, Laura.

Question is, if a child with CCMV has stable hearing for two years, do you still follow the same monitoring scheme, schedule? Yeah, that's a good question. I do. Especially for the case that I shared today. She didn't really have a dramatic change until she was suspected to be around 12 or 13.

And so I think it's a prime example to show that that progression doesn't happen in the first couple years or doesn't have to happen in the first couple years. It may happen at three months old, it may happen at five or at 16. And so maybe a child's been stable. So we see them once a year, but I think once a year is definitely the minimum and sooner if. If there is a change that makes sense.

Okay, well, we don't have any other questions unless anyone in the audience wants to add something real quick.

If not, I want to thank all the participants for hanging in there. I know an hour and a half is a long time to sit still. Thank you for hanging in there with us. And thank you again to all, all the presenters. I'm so honored to work with you guys, and I am in awe every day of what you do.

And it's so nice just to hear the case studies, just to have those tangible examples of how you spend your time. So thank you all and thank you to Carolyn and Kimberly for keeping us on track today. This was so much fun. Thank you. Appreciate you all.

That's a wrap for today's course. Thank you to everybody from Phoenix Children's and everybody who logged in today. Have a great afternoon.