Pediatric Grand Rounds: Embracing the Unexpected, in partnership with Phoenix Children's Hospital

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At this time, it's my pleasure to turn the microphone over to DoCTor Wendy Sterwaldd, the direCTor of audiology at Phoenix Children's Hospital. Welcome, Wendy. Thank you, Carolyn, and thank you audiology online. We are so grateful for this kind invitation to present some of our case studies at this pediatric grand round series, and I also want to thank everyone in attendance today and everyone who will be viewing this in the future. We are excited for the opportunity to share some interesting case studies with you.

We will be sharing six case studies of unusual patients, procedures or situations. You will learn about how changes in pediatric cochlear implant guidelines are impaCTing patient care considerations with consent and assent in teens, hearing loss associated with temporal bone fraCTure, benefits of bedside vestibular assessment, branchio otorenal syndrome as a faCTor in hearing loss and tips for effeCTive ABR testing and interpretation. As Carolyn said, my name is Wendy Sterwaldd and I'm direCTor of audiology at Phoenix Children's Hospital. Our case study presenters for today are DoCTor Deborah Flynn, doCTor Ali Sayre, doCTor Christina Dubas, doCTor Rachel Wooster, doCTor Caroline Sabatino, and DoCTor Robert Fanning. Our only disclosure is that we are all employees of Phoenix Children's Hospital.

Please put any questions in the chat and we will answer them at the end of the presentation. You are in for a treat today. Enjoy.

The learner outcomes are after this course, you will be able to discuss the importance of multidisciplinary teamwork, discuss several cases of complex audiology discuss audiology testing methods used to obtain accurate results with complex patients. Just a little bit about Phoenix Children's here in front of you is a map of the state of Arizona and Phoenix Children's Hospital is located in the yellow sort of blob in the middle of the state. We are the only freestanding pediatric hospital in the state of Arizona and we provide services to children who live in Arizona, who live in the surrounding states

and international patients. Many, many of the patients are complicated with more than one condition going on at the same time. Here is a photo of our beautiful hospital.

We have lovely pine trees and lots of sunshine. If you're ever in Phoenix, come by and visit. We would love to take you for a tour. It's my pleasure to introduce doCTor Deborah Flynn, our first presenter of the day. Good afternoon and welcome to our audiology online presentation.

We hope that you obtain some valuable take home information today that will be useful in your clinical praCTice. I think we have some interesting cases to present today, so let's get started. As doCTor Sterwald indicated, my name is Deborah Flynn, and I'm going to start off by discussing changes in pediatric cochlear implant candidacy over the last 15 years or so.

All right. At Phoenix Children's Hospital, we provide families with the options for all modes of communication. But if a family chooses listening and spoken language, then we must assess a child's ability to adequately access the speech speCTrum. As you are all clearly aware, there are limitations to how much gain traditional hearing aids can provide. In addition, not all auditory systems are created equal, and there are children that do not receive benefit from traditional amplification.

Despite appropriately fit hearing aids. The FTA criteria for candidacy is quite stringent and, if striCTly followed, would make it challenging for many children to obtain the goals of listening in spoken language. The American Cochlear Implant Alliance Task Force recently published guidelines in 2022 that provides evidence based research to support a broader criterion for candidacy. This case was challenging at the time, but as you will see, in the end fits well with the new guidelines for those that do not praCTice in a setting that provides cochlear implant services. I hope that this case will provide some insight into when it's time to refer to a refer a pediatric patient for a cochlear implant evaluation.

The kiddo that I'm going to present today came to our clinic at the age of two, and it was about 15 years ago, which means that he would need to follow the FDA guidelines for children between the ages of two and 17. The guidelines are presented here so we have severe to profound sensorineuralural hearing loss in both years, limited benefit from binaural amplification, and MLNT or LNT test scores of less than 30%. I'd like you to keep these in mind as we move through this case study.

Let me introduce Aiden. Aiden is currently an almost 17 year old male who we have followed on and off at our clinic again since the age of two. His history is significant for two failed inpatient newborn hearing screenings, one failed outpatient screening, and a diagnostic ABR at another facility that demonstrated bilateral moderate to severe hearing loss. He was not fit with hearing aids when he came to our clinic at two and records were not available when he established.

He was initially seen by my colleague for a bear study and confirmed a moderate to a moderate severe hearing loss in both years. About a month later, he was fit with oticon hearing aids and continued to wear those for about a year. His care was transferred to me at about age three years and eight months. At that time, behavioral testing indicated a moderate hearing loss in the left ear and a moderate severe hearing loss in the right ear. Speech testing at that time could not be completed due to severe language delay.

In addition, he came from a bilingual home of both English and Arabic. He attended a developmental preschool program, so he was getting intervention services.

Unfortunately, due to insurance reasons, Aiden had to transfer care to another clinic and remained at that clinic for approximately six years. Initially, he was fit with superpower hearing aids, and prior to returning to our clinic, they changed over to power hearing aids because of their recharging ability. So in 2018, he returned for a second opinion. At the time, his power hearing aids, he felt, were not strong enough and that he was not able to hear with them, and he was wearing his previous superpower

hearing aids, which were quite old and not working well.

So his audiologic testing about six and a half years later at age eleven, indicated slight changes in hearing, with results in the moderate severe to severe hearing loss range, right ear greater than left. His primary complaint again was that his hearing aids were not loud enough. The power hearing aids were reprogrammed, but he still felt like he couldn't hear and he was relying on his old hearing aids. I want to point out that his speech discrimination scores were pretty poor at the time. Despite well fit hearing aids, his speech and language continued to be significantly delayed, and academically, he was really struggling.

His parents had switched schools several times in six years, hoping to find a program that could help him catch up. They had tried several public schools, a private school, and a charter school. He attended private speech language therapy and auditory verbal therapy on a weekly basis. At age eleven, he was reading at a first grade level. His articulation was extremely difficult to understand despite full time hearing aid use.

Speech testing was particularly challenging due to his severe speech language delay in articulation errors. So at the time, oftentimes, I had to ask him to either repeat what he was saying or rephrase it or try to describe it to me so that I could determine if it was more of an articulation error or if it was more of an auditory error. Oftentimes I'd rely on the whip b just to get an assessment of full auditory funCTion.

I contaCTed the hearing aid company and they were gracious and agreed to switch out his power hearing aids for superpower hearing aids. And so these test measures just show that we did real measures to meet the DSL targets for his current thresholds and his aided benefit. Overall, his deteCTion levels were not bad, but if you look at his speech scores, even using Ohiopyle, he was still in that fair to poor range. Poor range. In quiet.

We continued to monitor Aiden over the next six months or so, and his hearing loss was stable. He wore his hearing aids on a daily basis for all waking hours. He continued to struggle with speech, language, and his academic performance. He was making slow progress, but his reading level and skills were still in the third grade level. His mother consulted several surgeons in the area regarding cochlear implants, and the family was told that he was not a candidate.

Two years later, at age 13, Aiden returned for biannual testing with a complaint of decreased hearing and hearing aids not being loud enough. Slight decreases were noted, but nothing significant. His discrimination ability continued to be very poor, and academically he was still well below grade level. His mother continued to inquire about cochlear implantation and was pushing for consideration. A CI evaluation was completed and with hesitation, the PCHCI team decided to consider cochlear implantation in one ear.

The deciding faCTor was his poor speech discrimination despite adequately fit hearing aids, as well as his poor academic performance. So this is his CIA evaluation. And so you can see that he had adequately fit hearing aids. His aided deteCTion levels were not horrible. He had access to most of the speech sounds.

But if you look at using a word list that is appropriate for him at his age, his. His percentage of words correCT was extremely poor. So this kind of spawned us onto moving forward with a cochlear implant.

Aiden was implanted in his right ear and aCTivated in September. On day one of aCTivation, he had discrimination of basic ling sounds. Within one month, his aided deteCTion levels were in the mild hearing loss range and his whippy scores were at 92%. By December of the same year, his speech discrimination ability improved immensely and he moved through the pediatric minimum speech test battery very quickly. So you can see here that in OCTober, his LNT scores were at 76, and by December, his CNC scores were also at 76%.

So a big difference between from the 30% that he was previously receiving. A year later, Aiden returned and he and his mother were extremely pleased with his progress. His family had found a private school that had one on one tutoring, and he was making rapid progress in his reading and language skills. In addition, his articulation had improved significantly. They inquired about a second implant.

Aiden indicated that he was struggling to hear with his hearing aid alone and solely dependent on his cochlear implant, although he still wore his hearing aid. We completed a CI evaluation for the other ear and here are results. Overall, his unaided threshold levels were still kind of in that moderate to moderately severe or severe range. His aided deteCTion levels were not bad, but his speech discrimination scores were at 32% compared to his cochlear implant year at 84%. So a big difference and he really noticed.

So Aiden was successfully implanted with his second device. His aided deteCTion improved within a month and his speech discrimination was excellent by three months. His academic performance continued to improve with one on one tutoring and ongoing speech language therapy AVT therapy. Over the past four years, Aiden has significantly closed the gap in regards to his academic equivalency. He is currently enrolled in high school and will be attending college classes during his senior year.

He plans on attending university after graduation and as a side note, he created a successful printing business that now has an online presence.

When Aiden was progressing through this process, he certainly did not meet FDA criteria. Yet he was struggling significantly. At the time, there was limited published consensus on pediatric cochlear implant candidacy, although publications were to emerge. Since that time, the ACIA has published the task force guidelines for determining cochlear implant candidacy in children. This has significantly

improved our CI team's ability to make decisions regarding pediatric cochlear implant candidacy for those off label categories.

So I just want to review the ACI pediatric guidelines and if you find a a patient on your caseload that fits one or all of these criteria, I encourage you to consider referring for a cochlear implant evaluation. So if they have automatic thresholds greater than 70 decibels, or speech recognition scores less than 50% and or poor funCTional performance, limited progress or poor quality of life. Aiden met all of these criteria mentioned in the ACI guidelines. However, at the time of our decision, we did not have these. He significantly benefited from cochlear implants.

Again, we just want to consider the new guidelines with the patients on your caseload. A quick review of Aiden Aiden had a slight bit of progressive hearing loss, nothing significant, but in particular, he had extremely poor aided speech discrimination despite acceptable aided deteCTion levels. He had poor academic performance, poor articulation and at the time, he was really a non traditional CI candidate. However, looking at our new guidelines, he aCTually fit those guidelines quite well.

Thank you for listening to this case on Aiden and I would like to introduce you to doCTor Ali Sayre, who is going to present a case on consent and dissent. Thank you for that introduCTion, Deb. I'm happy to be here to talk with you guys about this case that I aCTually found really, really interesting and helps us explore consent and assent in pediatric audiology, which is something that really doesn't come up very often I do not have slide control. Can somebody help with that?

No. There we go. Thank you. All right, so consent in the pediatric medical setting is complex and from an ethics standpoint, something that we really need to keep in mind as we're making decisions for patients when their expressed desires differ from their parents. You know, there's a difference between consent and assent, and how that applies in the clinical setting can be really important, but also really confusing.

Consent is defined in multiple diCTionaries as to give approval to permit or to agree. The underlying principle behind consent is valuing that individual's autonomy. They are a free, independent person and have rights to decide what happens to their body. Children, though, cannot provide consent in most cases because by definition, they are not yet fully autonomous. They lack the capacity to fully understand the implications of their decisions.

True informed consent, such as an adult could give, requires that not only do the risks, benefits, and alternatives be disclosed, but that person has the capacity to comprehend that information and voluntarily agree to that intervention. In some rare cases, children can consent for their own medical treatment without a parent involved, reproduCTive health care being one of the most common examples. But in general, consent can only be supplied by an adult. In pediatrics, parents provide consent by proxy, or in other cases referred to as parental permission. They're presumed to be able to make more sound decisions than the child and to have their child's best interests at heart.

Children, though, can provide assent, which is defined in diCTionaries as to agree or concur, to approve of something or to give in yield or concede. There's emerging discussion about the moral value of a child's preferences about their own medical treatment and how this can and should be applied. It's a very similar discussion to what happens for adults who are not competent to make their own medical decisions from something like dementia or a traumatic brain injury, among many other examples. Even though children and adults found to not be confident may need someone to oversee their medical care and make decisions for them, they should still be treated as an autonomous person as much as can be possible, and their preferences should be given weight where we can. In some cases, that's easy.

If a parent consents to a recommended procedure and the child also agrees, everybody gets to move forward. But if they dissent, defined as withholding assent or approval or differing an appendix, what happens next is a little less clear. There's a lot of studies and literature available about how to faCTor in

assent from children during participation in research, and in these cases, it's pretty clear that unless the participation is in a trial where there are no standardized treatments available for, like a terminal condition. If a child dissents, you don't proceed. But there's far less research and far fewer guidelines about patient dissent in the clinical setting.

In some cases, it's clear cut, like for life saving procedures. But decisions on eleCTive procedures like a cochlear implant are a little less clear. How much weight does the preference of a pediatric patient hold when compared to parental rights? How do we determine if the patient is competent enough to provide assent? Is there a cutoff by age, a standardized assessment we should give?

There's no clear rule about when a child's descent outweighs parental consent in audiology. I think in some cases it's clear. You know, for example, and it sounds silly if an infant cries when we're taking ear mold impressions after newly diagnosing them with hearing loss. Clearly this dissent by crying shouldn't change our recommendation to proceed with hearing aids. Their parents are obligated to make their best decision possible for them.

And as a newborn can't comprehend the implications of hearing loss and leaving it untreated, it makes sense to defer to parental consent in that case. To contrast that if we have a teenager with single sided deafness who does not want a cochlear implant but their parent wants to proceed, going against their will is a little bit harder to justify. A 17 year old saying no certainly holds more weight than a toddler saying know. But how do we faCTor in if that teenager has developmental delays? Where do we draw that line?

So I'd like to introduce you to my patient Jake, to highlight some of these concepts. He is a 16 year old male who was initially seen at our clinic as a transfer patient in March of 2023. History at that time was reported as significant for a bilateral asymmetric sensorineuralural hearing loss, worse in the right ear. He was only using a hearing aid for the left ear at that time. He also had a reported diagnosis of autism.

He is enrolled in high school and an oral communicator and receiving appropriate services.

Jake was born full term, but his delivery was complicated by meconium aspiration syndrome, which resulted in a NICU stay where he received IV gentamicin, which is known to be ototoxic. He did not receive a newborn hearing screening, as this was not standard in his birth country, but there were some early concerns for his responsiveness to sounds and for speech delay. At two years old, he was diagnosed with autism, so mom reported that his difficulties were attributed to that diagnosis and that he didn't have any hearing testing done, although there were some records that mentioned something about an ABR finding hearing loss from his home country, but there were no results or details or records of that included, and mom couldn't recall that that had been completed. He was first formally diagnosed with hearing loss at four years of age, at which time he had a moderate loss in the left ear and a profound hearing loss with no response at the equipment limits for the right ear. He was fit with a hearing aid for his left ear shortly after diagnosis, which mom reports he adapted to very well.

The first progression of his hearing threshold noted for the left ear was in January 2018, then showing a moderately severe to moderate loss. In May 2019, testing showed further decreases to his hearing loss, now a moderately severe to profound hearing loss, and then he was refit with a more powerful hearing aid in September of 2019 before the family moved to Arizona. So when Jake first came to us, he was seen by one of my colleagues and this was the most recent test results that the parents provided. It was from a local EnT office showing profound no response hearing loss for the right ear with a moderately severe to severe sensorineural or loss for the left ear with good word recognition, but they did not record what type of speech testing materials they used.

Results for his first evaluation with us were generally consistent with the ENT records, but they did show a decrease to his thresholds for the 4000 to 8000 thousand Hertz range in the left ear, and his word recognition scores were significantly decreased when we used an open set test with that PBK.

The audiologist who saw him on this date also completed a wipi since the score differed so much and that wipi score was more similar to what the outside office obtained, but at this point he was a teenager with far more advanced vocabulary and language levels than that test is designed for. Family was interested in getting a new hearing aid for his left ear, but also in considering a cochlear implant. He was refit with a new high power hearing aid for the left ear in May of 2023 and then referred for possible consideration of a left ear cochlear implant Jake established with a cochlear implant surgeon in our ENT department in August. At this point, mom was requesting Jake be evaluated for candidacy, though of a right cochlear implant, not the left ear like we had discussed.

He was referred for full team workup, including evaluation with myself, MRI, speech and psychology evaluations, and for genetic testing. He was also referred to ophthalmology because he had failed a recent vision screening before my first visit with Jake, he had completed most of his additional work up, so results of genetic testing ordered in September showed mutations in the GJB two gene along with to others. This result was interpreted as consistent with him being at least a carrier for autosomal recessive GJB two related sensorineuralural hearing loss, but was not able to establish a diagnosis because a second variant wasn't found. They did note that the result could be possibly associated with an autosomal dominant hearing loss, but they couldn't give us a definitive etiology.

His MRI was completed in January of this year, and for his hearing anatomy was unremarkable, but it did reveal abnormal brain findings. The BPP finding is a neurological disorder which occurs from changes in the grooves and folds of the cerebral cortex during embryonic development, and both the age and onset of symptoms can vary widely. It's not yet clear from Jake's records to what extent he's impaCTed by this diagnosis, but interestingly, there is some research that notes that auditory processing disorder may be a possible symptom of this condition, but sensorineuralural hearing loss is not. Jake was seen by ophthalmology also in January, and there were no significant concerns for his vision. Jake was evaluated by the speech language pathologist on our team in February.

She noted some difficulties getting participation for the assessments, but with what was completed indicated that he had a severe receptive and expressive language disorder. Jake is an oral communicator, but he does rely heavily on visual and other supplemental cues. His mom at this time reported concerned for his participation in therapy and noted that previously telehealth had also been unsuccessful for him. The report also notes that mom was really concerned about how Jake would adapt to a cochlear implant. Jake was also seen by the psychologist on our team for two separate visits, and from these, it was pretty clear that Jake's ability to participate in the cochlear implant process was going to be extremely limited.

He had an extreme dislike of missing a school and having his schedule disrupted and had a hard time adapting to new hearing aids. Each time he'd been fit with a new generation of technology, he was fixated on finding his old hearing aid, which he brought up repeatedly. At visits with every member of our team, mom indicated for our psychologist that she's motivated to get Jake a cochlear implant because she is worried about the progression of his hearing loss and what might happen to him if he's unable to communicate in the future. If his left ear gets worse, she's worried about it not only from a quality of life and day to day communication standpoint, but in her case also from a safety standpoint with his autism diagnosis, if he's unable to hear and speak with people, she was worried that that might open him up to abuse or negleCT in the future when he's no longer under her direCT care. She also reported, though at this point very little understanding of what outcomes could be expeCTed with a cochlear implant for Jake.

At my first visit with Jake, he and his mom pretty quickly confirmed what the team had reported from their visits. Jake had no idea what a cochlear implant was, why he was at our evaluation that day, and mom expressed interest in a right instead of a left cochlear implant. She denied at this point any concerns for how he would adapt to it and for how he would participate in audiology and therapy appointments, which for me was my first red flag given that at other teen visits she had reported

concerns for those things. Jake was very upset throughout my whole visit about missing school to be there, but was able to be convinced to participate in testing. On that day we got type A tympanograms, absent acoustic reflexes at the limits of our equipment for both ears and essentially stable hearing thresholds for air conduCTion as his hearing was stable, long known to be sensorineuralural, I chose to omit bone conduCTion testing on the state so we could move a little bit more quickly through the testing that we needed to get to.

From his speech and language evaluation and from our review of his IEP, we were able to determine that standard lists were appropriate to use with him. So we didn't need to go back to the WIPI or PBK list and we wanted to get the most accurate understanding of how well he's using that hearing. Speech testing showed poor word understanding for the left ear at 64%.

His hearing aid settings for the left ear were verified with on ear speech mapping to DSL pediatric target as family was interested in a right cochlear implant, I also fit a high power BCE for the right ear just using a comply mold for testing to be done in office. Aided testing for the left ear showed pretty good audibility through about 2000 hz with a little bit more reduced deteCTion of the high frequency sounds, like we might expeCT for his hearing loss. For the right ear, only very limited low frequency audibility was measured and it's possible that this might have been just crossing over to the left ear, but at this point he wasn't compliant with masking, was getting a little bit more agitated so I just kind of moved on from there. Speech testing for the right ear showed really no response. He had no awareness or understanding of speech at any level in that year.

For the left ear, an SRT was measured at 25 decibels. Aided and CNC testing at 60 decibels. PL showed a word score of 52% and a phoneme score of 73%.

I had a very long conversation with Jake's mom about his candidacy for a cochlear implant in each year. It's important to note, given the broader theme of consent and assent in this case, that at this

point, Jake chose not to participate and instead went to the lobby to play video games while waiting. His left ear. At this point, we consider to be kind of a borderline candidate for a cochlear implant beyond his thresholds being predominantly in that severe range. His word recognition testing showed significant difficulties, and his aided CNC word and quiet meets criteria for implantation is below 60%.

If Jake were a typically developing child, this might be a case where he and his family were encouraged to consider proceeding with the left implant. However, given his larger concerns, mom was cancelled that for Jake, it may make sense to just continue monitoring. FunCTionally, he does very well with his hearing aid. He's attached to it, and he wears it every waking minute of every day. There's also concern that with his developmental delays and autism, that he might have more difficulty adapting to a cochlear implant than a neurotypical child, which is further complicated by his resistance to change and even attending the appointments.

It's possible that if I had been able to get testing in noise testing with softer speech, we might see more difficulties, and maybe that recommendation would change to, hey, we should proceed now anyway. But we weren't able to get that due to time constraints and his attention. Mom was pretty clear at this point that she does not want a cochlear implant for his left ear. So we just eleCTed to continue monitoring his left ear, and we'll reevaluate if he starts to have more difficulties at home, if the loss progresses more, or if we are seeing poorer scores on our testing here for the right ear, mom was counseled that, well, technically, his right ear may qualify for an implant based on the degree of hearing loss and lack of benefit from a hearing aid. The costs proceeding with a cochlear implant for this ear is expeCTed to far outweigh the little benefit that he might receive his hearing loss.

At this point, we presume to be congenital. And even if we used a more conservative estimate and said that it really did start at about four years of age when he was first diagnosed. He still has a very long duration of prelingual hearing loss with no intervention. Remember, he's never worn a hearing aid.

Given this, we wouldn't expeCT him to develop any word understanding, even with a cochlear implant in that earthen.

And even audibility and awareness of sound with that device. Would really be highly dependent on his ability and willingness to adapt to it and to put in the required work with that oral habilitation. Mom had been hopeful that the cochlear implant in the right ear. Would allow him to communicate orally in the event that his left ear decreased further. So she was pretty shocked and upset when she was given this prognosis.

And she was expeCTing that it would give him an essentially normal or funCTional ear. Even after we reviewed how much time and effort, effort would be required for really minimal benefit. And how difficult this might be for Jake. Given his preferences and resistance to being at these appointments, mom was still insistent that she wanted to proceed with the right cochlear implant. So at this point, I advised mom that the final decision.

Would really be highly dependent on the team review process. And it's not guaranteed that we would proceed at our team meeting. We have a very big multidisciplinary team meeting. Where we review these candidates. We discuss the results from all of our disciplines in detail.

Our concerns were pretty significant, really clearly defined, and well aligned with each other, which we sometimes don't get. Sometimes there's disagreement or less clear cut cases. But in this case, in addition to the poor prognosis, concerns about his ability and willingness to engage in the process, we were also very concerned about mom's poor report of his history. And more importantly, his mom appeared to have very poor understanding. Of realistic expeCTations.

For a cochlear implant in the right ear. She had decided that this is what she wants for him. And was consenting to the procedure despite that. And while Jake didn't really understand what was going on or

what we were talking about, our impression from interaCTing with Jake was that even if we could spend the time helping him understand what an implant was, why his mom wanted it, and what that process would be like, what he could get from it, we didn't think that he would assent. So it was pretty clear to the team that the costs of proceeding.

Far outweighed the expeCTed benefits. And along with Jake not wanting to do it, we ultimately decided that he's not a good candidate for an implant. And we told mom that we'd continue to monitor the left ear and reevaluate for that year if something else came up.

So I learned a lot from Jake. It's absolutely a lot of work in progress still, despite only having met them for that one visit, at this point, this is definitely a case that has stuck with me. The most significant and interesting thing I think I learned from him, though, was that, you know, how do we faCTor in that consent and assent in our day to day work? You know, for us, this is a case that we been able to take that consent and assent and apply it to other populations, like our ongoing discussion of how we handle single sided deafness implant patients. What do we want to consider as we're evaluating these kids for candidacy?

Things like age, developmental delay, cognitive status, parent and patient understanding of outcomes for the procedure, so that as a team, we can grow and ultimately take a little bit more of a well rounded and patient, family centered approach to making these decisions.

That is all that I have for my case. So I'd like to now hand it over to DoCTor Christina Dubas, who will talk to you about one of her patients. Hi. Thank you so much, Allie. I'm Christina Dubas, and today I'm going to be diving into how anything goes after trauma using a patient study here with Bethe.

Before we dive into Beth's story, I just want to review a few concepts so temporal bone fraCTures can be classified a few different ways. The historical way of classifying fraCTures is using either longitudinal

or transverse description. Longitudinal fraCTures account for 70% to 80% of temporal bone fraCTures, and this means that the fraCTure line is parallel to the long axis of the petre's temporal bone. ConduCTive hearing loss is really common with these fraCTures, either due to hemotympanum, ossicular disruption, or tympanic membrane perforations, whereas transverse fraCTures are less common. The fraCTure line is perpendicular to the long axis of the fetish temporal bone, and sensorineuralural hearing loss occurs in 50% of these patients, and that sensory neural hearing loss can come from otic capsule violation, labyrinthine concussions, interruption of the blood supply to the cochlea, or damage to the cochlear nerve.

The classification based on fraCTure line direCTion aCTually comes from research back in the 1950s. Some drawbacks of this classification system are that temporal bone fraCTures are often more complex than just longitudinal or transverse. They can be mixed, they can include both types. The research aCTually shows that categorizing temporal bone fraCTures this way does not help us understand the prognosis of the hearing loss loss and other complications. That could arise from the fraCTure.

So there's been a shift in classifying temporal bone fraCTures by otic capsule status. As I was diving into the research on this topic, there's a lot of support for getting rid of the transverse versus longitudinal classification and really relying on the otic capsule status classification. So, otic capsule sparing temporal bone fraCTures often involve the temporal bone, squamosal portion, and external auditory canal, particularly the mastoid air cells and the middle ear. This has that mixed or conduCTive hearing loss due to hemotympanum or that ossicular chain disruption. Now, odic capsule violating fraCTures.

Force is aCTually transmitted to the capsule through the foramen magnum and the Petris temp Petre's pyramid. This is strongly associated with cerebrospinal fluid leakage, sensorineuralural hearing loss,

and other intracranial pathology. This type of fraCTure rarely affeCTs the ossicular chain or external auditory canal.

With those temporal bone fraCTures, we often see ossicular discontinuity. So ossicular discontinuity can include a few different presentations. ConneCTion between some ossicles can be difficult to aCTually visualize on imaging, especially when there's hemotympanum present. So sometimes we're not able to tell what that dislocation aCTually looks like until the surgeon is in the middle ear space. A complete discontinuity includes no contaCT between the disconneCTed ends and usually results in a large, flat conduCTive hearing loss, maximum conduCTive hearing loss.

A partial discontinuity. On the other hand, there can aCTually be joint contaCT or part of the ossicles replaced by soft tissue, and the ossicles are just holding each other up with the force of one another with that partial discontinuity. The research has shown that this can result in a high frequency conduCTive hearing loss. So it used to be thought that a high frequency conduCTive hearing loss was user error, audiologist error. But now the research has aCTually shown that that can be the result of that partial discontinuity.

Now that we review those terms, let's jump into Beth. Beth is a 15 year old female who was admitted to the hospital following a high speed motor vehicle collision. She was initially comatose, but following the injuries include severe traumatic brain injury, traumatic occlusion of the right internal cardioid artery, and left sided direCT carotid cavernous sinus fistula, left optic neuropathy, bilateral temporal bone fraCTures, and ossicular chain dislocation, right facial paralysis, and right leg paresis.

Her initial imaging results, she had a CT done that showed both longitudinal and transverse bilateral temporal bone fraCTures and ossicular dislocation. I can't say the exaCT words that the otolaryngologist used to describe the ossicles look, but it was not normal. It was very disrupted. So on both sides, that was the case.

ENT aCTually saw the patient following that imaging the day before they were seen for an audiological evaluation. They did bilateral ear canal debridement, and there was persistent bloody odor in both ears, which they noted was likely cerebrospinal fluid leakage. Then both ears were really difficult to evaluate the middle ear space because those eardrums were still healing and thickened, so they couldn't really tell what was going on in that middle ear space face. Ultimately, though, the ENT had seen those imaging reports, which our radiologists are still using the old way of classifying temporal bone fraCTures, and saw that there was the transverse fraCTures on both sides. So the patient was very heavily counseled on what that could mean for their hearing outcome, as they had bilateral transverse fraCTures along with those longitudinal fraCTures.

That brings Beth to my office, where we did the first inpatient audiological evaluation. You can see right off the bat that this is nothing what we expeCTed based off of the imaging. So transverse fraCTure, sensory neural hearing loss. Nope, we don't have that. Longitudinal fraCTure.

Maximum conduCTive hearing loss, possibly with some ossicular change. Dislocation. Nope, we don't have that going on. So it was a really big surprise to see these results. I did every cross check possible to make sure that I was not getting these in error.

The SRT is lined up. She had really good word recognition, even at those elevated levels. But you can see that the right ear has that rounded tympanogram with that hemo tympanum still present in that right ear. So, still that acute injury going on, on.

Ultimately, we were able to fit her with hearing aids as an inpatient, which was a really big blessing to the family. Obviously, the 15 year old female patient, who has had so much going on, was very reluCTant to have one more thing going on. But the parents were very appreciative because they wanted her hearing for her physical therapy sessions. They wanted her hearing for her speech therapy

sessions. They were noticing that she was having a really difficult time understanding her therapies, and we're very grateful that we were able to fit her with those loners.

The next time I saw Beth was a couple months later in the ENT clinic follow up. So, in our ENT clinic, we have a little less time to do the testing that we do in the audiology clinic, but I tried to make it as thorough as possible, given what I knew about her already. And you can see how big of a shift this was. Now the right tympanogram looks a lot better. It's more peaked.

It doesn't have that rounded appearance anymore, following the resolution of the hemotympanum. But, wow, that was a really big shift, especially in that right ear, in those frequencies. So I'm going to go to the next page where we can kind of get a better look at that comparison. So this is a little overlay of the first hearing test versus the second one. There's a lot of noteworthy changes.

There's that huge shift in the right ear that decrease in the low pitches, the improvement in the high pitches, but overall, still pretty conduCTive. But why did that shift even happen? Was the hemotympanum previously providing support for the ossicles? How are there some thresholds so mild still with ossicular dislocation? We had talked about how ossicular dislocation can cause that maximum conduCTive hearing loss.

Flat or a high frequency conduCTive hearing loss. We didn't talk about a rising conduCTive hearing loss. Basically what everyone on her medical team just keeps noting it up to is anything goes after trauma. We might be able to see more once an exploratory surgery happens, but for now, we're just kind of grateful for the hearing that she has. As I said, she was really counseled on the possibility of a lot of sensorineuralural hearing loss with transverse fraCTures.

So this might be a really good case where we can see that maybe we should be shifting to the otic capsule classification of temporal bone fraCTures and not just relying on that transverse versus

longitudinal classification, as that really did to tell us what to expeCT for this patient's hearing loss.

Ent is going to consider middle ear surgery in the next six months or so. There's still more urgent matters for her that need to be addressed. So then she was referred back to audiology clinic for hearing aid management. When I saw her back, we trialed aCTually a more powerful receiver to help account for those low frequencies in the right ear. But interesting enough, the more powerful receiver only increased her SII by one point, and the patient really reported that the sound was muffled and there was a lot of interference of her own voice.

So we went back to the lower strength receiver, and she was happy with the sound quality. She is wearing the hearing aids ten to 11 hours a day. So both the patient and her family are extremely satisfied with the improvement in quality of life with the hearing aids, which, if any of you know, those kind of situations, anytime that we can help bring a little light to otherwise very traumatic experience, it feels really good. So we have Beth here who was in a motor vehicle accident. She had temporal bone fraCTures and ossicular discontinuity.

And despite what we might have thought, with either a maximum conduCTive loss or sensory neural hearing loss, she had a mild, moderate conduCTive hearing loss loss. We had a shift in right thresholds with resolution of hemotympanum, which was interesting. She was able to be fit with hearing aids, and that really improved her quality of life currently. So, overall, a successful story of anything goes after trauma, but we're just all doing the best that we can. So that is all I have for Beth.

I'm going to pass it off to Rachel. Great case, Christina. Thank you so much. I'm going to be talking to you guys about vestibular testing and bedside assessment. So I briefly wanted to walk through our current test battery.

At this time, we don't have a full lab, and we are limited in what we can test, but our whole goal was to at least be able to test some part of the vestibular system to help identify some of these kiddos. So currently, we're able to do bedside screenings and vemp testing. This allows us to monitor their progress and keep them on our radar as we obtain additional equipment. So what are vamps? Vamps are a vestibular evoked myogenic potential.

This type of testing allows us to test the funCTion of the otolith organs and the vestibular nerves. So, just for a quick review, our otolith organs are going to be our saccule and utricle of the inner ear. The saccule helps us to deteCT those vertical movements. So think about it like going up or down in an elevator. Whereas the utricle helps us to deteCT horizontal movements, like driving forward or backwards in a cardinal.

Once these otolith organs sense motion, our brain then sends a message to our different vestibular reflexes. So our VOR is going to kick in to help maintain our eyes focus. The VSR is going to kick in to help maintain our posture. Our VCR is going to kick in to help maintain our head stability. And once the brain receives all of this information, it's going to help us to maintain our body upright to avoid falling.

So, as you can imagine, if one of these systems aren't working well, this can create a lot of problems, especially for a child who's trying to develop gross motor skills.

So C and O vemp are a little bit different. C vamp is going to test the saccule and inferior portion of the nerve, whereas ovemp primarily assesses the utricle and superior portion with a small portion of the saccule. This is just a short list of some of the bedside screeners that we do use with our pediatric population. I am going to be diving in a little bit deeper into the bolded ones on the next few slides, as those were used for my case study. So head thrust test is used to screen the funCTion of the horizontal semicircular canals.

For this screener, you're aCTually going to tilt the patient's head downward about 30 degrees. You're going to have them focus on a target. So that could be your nose, or if you're a pediatric audiologist, a sticker on your nose. You're going to turn their head randomly and rapidly towards the right and left while watching their eyes. Someone with a normal peripheral vestibular system will be able to maintain, maintain their eyes on the target that they're looking at.

But like we talked about earlier, when there's a disconneCT between our vestibular system and the brain, there's also going to be that disconneCT between the other reflexes as well. So someone with an abnormal system, you may see what's called a saccade or a correCTive eye movement. So their eyes are going to move from the target and then jump back to the target. Sometimes these saccades are obvious or overt, where you can see them with a naked eye. Sometimes they're covert and you can't see them without special equipment to record the eye.

So our balance system relies on three different systems, including the somatosensory system, visual system, and the vestibular system. The modified CTSIB screening helps us to assess all three of those systems together and then slowly takes away the somatosensory and visual system to see how our vestibular system is funCTioning when those other systems aren't helping to compensate. So there's a few different conditions for this test that we're looking at, and we're trying to see how long the child is able to maintain balance on all of those conditions.

DVA is a test to assess the funCTion of the voR, which, remember, is the reflex that helps our eyes to maintain focus. The patient has to read the smallest line possible on an eye chart while their head is at rest. And then from there, their head is moved side to side, and they're asked to read the smallest sign they can while their head is moving, if they go three lines above or more. This is considered to be an abnormal response response.

So during gait observation, we're looking for things like swaying or veering while walking. Is the patient falling at all? Are there any abnormal movements with their arms and legs to keep them from falling? And a lot of times these are kind of the observations that parents are reporting as well.

And then lastly, tracking. We're having the patient follow a finger with their eyes. We're looking for really smooth eye movements or for any nystagmus patterns that may be suggestive of a central pathology.

I'm going to get you introduced to Luna. Luna is a four year old female. She has a medical history of epilepsy. She was initially referred to ENt following an MRI of the brain for the epileptic episodes that also revealed abnormal findings of the right inner ear that was later found to be evident. She failed her initial newborn hearing screening in the right ear and was then seen for an outpatient ABR in 2019, which revealed normal hearing sensitivity bilaterally.

She was then seen for a hearing evaluation following her MRI in 2024, where testing was suggestive of a moderately severe to moderate sensory neural hearing loss for her right ear and normal hearing sensitivity for the left year, per her mom's report. She's always been very clumsy, off balance and her mom also did note some delays in her gross motor development as well, with her crawling and walking, and now she's having difficulty learning to ride a bike. So her initial audiogram is going to be on the left hand side. On the right is her confirmation audiogram. In between tests, her mother did report that she had fallen and hit her head off the ground and her mother perceived a decrease in hearing, following which we can see on her audiogram as well.

She was then seen for vestibular testing where her C vent response for the left ear was normal and as to be expeCTed for the right ear, she did have an absent c vent response.

We completed a couple different bedside screenings and all of the screeners that do assess the peripheral vestibular system were positive, revealing abnormal funCTioning of her vestibular system.

Just to summarize everything, she did have that abnormal c vent response for the right ear, which is suggestive of abnormal saccule and inferior funCTion of the right ear. She had the abnormal HID screening, which is suggestive of abnormal horizontal semicircular canal funCTion for the right ear. Her abnormal modified Civ screening is suggestive of vestibular weakness. She did have abnormal gait and frequent falling throughout the appointment.

So all of those results together were most consistent with inferior vestibular nerve and saccule involvement for the right ear and peripheral vestibular dysfunCTion. So next steps for Luna, she will be fit with a hearing aid and receive audiologic monitoring. She was also referred for vestibular rehabilitation with our physical therapy team, and she'll also continue to receive vestibular monitoring as we obtain additional equipment. So why is it important that we screen the vestibular system in our pediatric patients? Well, for starters, children with sensorineuralural hearing loss are more likely to have vestibular weakness, especially those with the hearing sensitivity within the severe to profound range.

Emerging evidence suggests that anywhere between 15% to 80% of children with hearing loss have an associated vestibular impairment. Those same patients that have severe to profound sensorineuralural hearing loss are usually our cochlear implant users as well, and individuals within that sensorineuralural hearing loss range that also have vestibular dysfunCTion are eight times more likely to have a device failure than compared to their peers without vestibular dysfunCTion. Vestibular dysfunCTion can also impaCT their academic performance, so those when sensorineuralural hearing loss and bilateral vestibular loss were found to have worse visual acuity during head movements, which has a direCT relation with reading acuity. Otolith dysfunCTion has been linked to delayed gross motor control and difficulty in walking ability. And just to circle back to our eva kiddos, research shows that about 45% of those with EVA had vestibular signs and symptoms, and 44% of tested patients with EVA had abnormal vestibular testing.

This is very important as abnormal vestibular testing is also correlated with a history of head injury, which we know in Salith Luna can contribute to a progression in their hearing loss as well.

So here are just a few populations that are at a higher risk for vestibular dysfunCTion that you should consider evaluating and monitoring in your clinic.

And, you know, what should we be looking for as providers? What should we be asking during our appointments that are already busy asking about, you know, the hearing side of things? Well, one parental report is going to be critical for these patients. They're going to remember if their child was delayed in any of their gross development milestones. Our true vertigo patients are also very important for us to be seeing, but even things like torticollis in infancy can be linked to a variant of vestibular migraine, also known as benign proximal torticollis of infancy or BTE.

So our current referral guidelines here any child with a hearing loss greater than 66 decibels PTA should automatically be referred for testing, as we know, because they're at a higher risk for those vestibular impairments. Children with hearing loss less than 66 db PTA are really only being referred if there's concerns for dizziness, imbalance, or those delayed milestones. We are seeing our cochlear implant patients pre and post op. Any individuals with peripheral vestibular loss concerns or any inner ear anomaly, such as EVA or SSCD.

For Luna, she started out with those delays with gross motor development where mom was very concerned, and nothing was really kind of pointing to why that was happening. She was later found to have epilepsy, which doesn't really have anything to do with that. But that diagnosis led to her getting an MRI, which revealed the Eva of the right ear. That MRI then led to her having another hearing test, which revealed a change in hearing for that right ear. And then that testing led us to the vestibular evaluation, which gave us a reason for why we were seeing those delays in her gross motor development.

So this testing is very, very important. Even if you can only add, you know, a couple screeners into what you're doing in clinic, I think they can provide a lot of really good information so that we can get some of these kids help.

So with that, I'm going to pass you off to doCTor Caroline. Thank you. Thanks, Rachel. Hello. My name is Caroline Sabatino, and I am excited to talk to you all today about a case that has brought up some unexpeCTed challenges.

And it's a case involving branchial, otorrenal speCTrum disorder.

Let me see about getting this forward.

Sorry, guys.

All right. Went too far. Okay, so, as a refresher, brachioodorenal speCTrum disorder is an autosomal dominant genetic disorder involving the neck, ear, and kidneys. It includes two phenotypes. Brachiootorenal otorrenal arbor syndrome, which consists of abnormalities in the second brachial arch, otologic abnormalities in renal abnormalities.

Abnormalities. And then Brachiootorenal Oto syndrome, or BoS, which is just the abnormalities in the second brachial arch and the otologic abnormalities. These phenotypes really only differ in either the presence or absence of the renal abnormalities. It is important to note that there can be an extreme variability in the abnormalities between the left side and the right side of an affeCTed individual, as well as among individuals from the same family. And then in Boer syndrome and Bos, hearing loss occurs in over 90% of the cases.

In Boer syndrome and Bos, the type of hearing loss can be conduCTive, sensorineuralural, or most often it's mixed. The severity of hearing loss can range from mild to profound, and the hearing losses tend to be non progressive, but they can also be progressive. So it really just runs the whole speCTrum. Abnormalities can be seen in the outer, middle, or inner ears. In a majority of cases, there are preauricular pits, but there can also be preauricular tags or malformed pinna, atresia or stenosis of the external auditory canals can also occur.

Middle ear abnormalities include malformation, malposition, dislocation, or fixation of the ossicle. There can be a reduCTion in the size or malformation of the middle ear space, and then inner ear abnormalities include cochlear hyperplasia, hypoplasia of the lateral semicircular canal, or enlargement of the cochlear and vestibular aqueduCTs. For physicians to make the clinical diagnosis of the birds in the absence of any sort of family history, they have to have either three or more major criteria or two major and two minor criteria that must be present. So now I would like to introduce Marcus. Our department received a referral for a five year, four month old male.

The reason for referral was listed as failed OAE screenings, ear pits, autism, and possible genetic disorder. So, and then the ICD ten codes that were listed on the referral also provided some additional information. They were codes for pervasive developmental disorder, developmental disorder of speech and language, and congenital malformation of the ear, causing impairment of hearing. So I saw Marcus in March of 2023. His foster mother accompanied him to the appointment.

She reported Marcus recently moved from Florida and has only been in her care for the past month. He was adopted by his birth mother's friend, and he has been in and out of foster care. His birth history was unknown, but the medical records that were sent from the pediatrician stated that Marcus was born deaf in his right ear, but he never used a hearing aid. The medical records also stated Marcus has autism. And then Marcus's speech was reported to be and aCTually observed to be very unclear.

He uses some sign language to communicate, but the foster mother doesn't know any sign language. Marcus just started kindergarten, and he's in the process of getting an IEP set up. It was reported he has a history of ear infeCTions and that his left ear was draining last week. And I looked into our scheduling system, and Marcus was scheduled with our ENT department to be seen in January, which at this point in August, Washington, about five months away. So on the initial testing, first I did otoscopy and his right ear had a preauricular pit and a pre auricular ear tag.

His left ear had just a preauricular pit. Both his tympanic membranes appeared dull with some possible effusion. He had bilateral type B tympanograms. DPOAEs were absent bilaterally. Marcus participated really well in behavioral testing using condition play, and test results were consistent with moderate to moderately severe mixed hearing loss in the left ear and a profound sensorineuralural hearing loss in the right ear.

At this point, I counseled the foster mother about the results and that I recommended Mark SC and ENT, you know, much sooner than January. And so I worked with the ENT schedulers and got him an appointment. His appointment moved up to September, and then my recommendations also included to return for repeat testing and discuss amplification at that time. So in September, Marcus was seen by one of our otolaryngologists here at Phoenix children's, and the ENT noted that Marcus had bilateral brachial abnormalities and a history of drainage from his from neck pits. The ENT stated that Marcus's exam was concerning for Bos and the Ent ordered a CT scan in an MRI.

He also ordered a neck ultrasound and a renal ultrasound to assess any renal abnormalities. Given the concern for Boer syndrome, the ENT referred Marcus to genetics, to speech, and to Phoenix children's pcp to establish a medical home for him. He also referred Marcus to a fellow ENT that specializes more in the ears. And lastly, the ENT cleared Marcus for hearing Aidan aid use like immediately, and he stressed to the family the need for hearing aid rehabilitation as soon as possible given the lack of time

he's had without a hearing aid. And the ENT found me in ENT clinic and relayed to me that he wanted him fit with a hearing aid despite the faCT that he had type b tips and a mixed loss.

So I worked with the ENT to get him on my schedule sooner rather than later. So between OCTober and November, Marcus was seen for a renal ultrasound, a neck ultrasound, an MRI, and a CT scan. The major findings from the renal ultrasound were that his kidneys were small for his age. His neck ultrasound revealed thyroglossal duCT cysts. The MRI findings were kind of surprising.

It was noted that the morphology of the cochlea and inner ears were suggestive of Boer syndrome. His right cochlear aperture and right cochlear nerve were both absent. He had effusions in both mastoids and middle ears, and he was found to have microcephaly. And then on the CT of the temporal bone, there was an irregular appearance of the incus of both ears, which they thought might be due to erosion, and the posterior semicircular canals were not seen bilaterally.

I saw, in November, I saw Marcus back for repeat testing in the hearing aid consultation. At that point, Marcus was transitioning to Phoenix day school for the deaf, which could better serve him. His hearing was the same as previous testing, showing the moderate to moderately severe mixed hearing loss in the left ear and the profound hearing loss in the right ear. At this point, I saw him before we had the results that he has an absent nerve. He still had type b tympanograms, and the foster mother participated in the hearing aid consultation.

I counseled her on the different types of hearing aids that we could do for him, what the benefits and limitations were of each. The foster mother was adamant that Marcus is not somebody that would tolerate anything on his head like a headband, and she did not think he would be successful with a bone anchored hearing device on a softband. So she opted to move forward with a traditional behind the ear hearing, a aid with the understanding that his hearing loss would be closely managed and monitored, and if there was any fluCTuation in his hearing, we would have to reprogram the hearing

aids. So the family did not come in for the hearing aid fitting in December, but they were rescheduled, and they did come in in January. Marcus was fit at that time with a bte with a custom ear mold on his left side.

The hearing aid fitting was a huge success. Marcus instantly liked the hearing aid. He asked for one. So the hearing hearing aid was on his left ear. He asked for one for on his right ear.

He was very excited and happy at that point. I discussed with the foster mother that, hey, he doesn't have a nerve on that side, but we can definitely do a device called a bicross, where explained what that was. The foster mother said she wanted to hold off until she knew that he was successful with the left hearing aid, which happened to be wonderful foresight on her part, because Marcus returned in February for his follow up, and the foster mother reported that on the drive home from the fitting appointment, Marcus chewed on the ear mold. She talked with him about how he's not allowed to chew on it, had a discussion with him, and sent him to school the next day with the hearing aid. And while at school, Marcus chewed the ear mold so much that they aCTually broke it in two spots where it was completely unusable.

So he had not worn the hearing aid at all. Since I had fit him, I ordered Marcus a replacement ear mold. And at that point, we kind of used ThE appointment as a brainstorming storming session to figure out possible solutions. So, with his autism, Marcus has an oral fixation, and he likes to chew things. And we thought maybe if he got a one of those sensory chewy necklaces, if he could wear that while he wears the hearing aid.

So we're giving him something that he can chew, maybe he won't chew the hearing aid.

And then in March, Marcus had bilateral ear tubes placed, and at that point, the hearing aid wasn't being used. And so I saw him back for a post op testing and fit him with his new, his new ear mold. And

so at that point, the testing showed a mild sensorineuralural hearing loss for that left ear. The hearing aid was adjusted. And this would be a great place to say that now Marcus is the compliant hearing aid user, and everything is PG.

However, this is an interesting case filled with many unexpeCTed challenges. Marcus does not have a confirmed diagnosis yet. He's not seen nephrology, but he is suspeCTed to have Boer syndrome. With that, he has a very interesting hearing loss. He has the absent cochlear nerve.

He has persistent middle ear disorder and a mixed hearing loss in the left ear that we briefly saw improvement with the placement of an ear tube. But unfortunately, by May, the ear tube quickly extruded. By May, his hearing was back to the moderately severe. And at this point, he is scheduled for a second set of tubes in August. But this has brought up the question of, is this bte the most appropriate for him?

And so we're planning on trialing a bone conneCTion hearing aid at his next appointment. Marcus also has autism, which contributes to his poor compliance and poor use of the hearing aid, and having him have oral fixation of chewing his ear molecules. And then on top of all of this, Marcus is in foster care, which has put a delay in some of his services, and it adds layers to the counseling. When we are counseling the person in the appointment, that's there, and then that information is being relayed to aCTual mom, and there's multiple people have to be on the same page for decisions to be made. It kind of just adds an extra layer.

So Marcus is still a work in progress, and we're trying to embrace all these unexpeCTed obstacles to give him the best care. And now I would like to pass it on to DoCTor Robert Fanning. Well, thank you, Caroline, and thanks for everyone who held in for the end here. I am the last case. I hope it was worth the wait.

I'd like to share with you a case I call which side are you on? A tricky ABR test involving mixed hearing loss. I hope you find something useful about it and informative and interesting.

Let me move on over here to the next slide. First, before I jump into this, let me explain my role here in this case. I'm a collaborator. Here at Phoenix Children's, we often participate in the diagnosis of treatment of others patients due to schedule confliCTs and our availability at certain locations. I am not the managing audiologist in this case, but I have performed two of this patient's ABR tests, one when she was four years old and the other when she was six.

And my work here, I hope, will provide the managing audiologist the information she needs to effeCTively care for the patient.

And without further ado, let me introduce you to Judy. She is a six year, ten month old female diagnosed with trisomy 21. She has also been diagnosed with two q and 13 Q deletion. She has autism, and perhaps is no surprise, she has also been diagnosed with global developmental delays. She is nonverbal.

She does have some manual signs, and she also uses an AAC device. She has been diagnosed with chronic otitis media with a fusion. She had ear tubes placed in both ears at the age of eight months and then had replacements at the age of two and four years. She failed her newborn hearing test, and at the age of eight months she underwent an ABR test following bilateral placement of ear tubes. And in the right ear, she had a mild to moderate, primarily sensory neural hearing loss, and in the left ear, she had a moderate, primarily sensory neural hearing loss, and that's not unusual in cases like that.

There's just a bit of a conduCTive component to the study. Results from subsequent ABR tests in early childhood were generally confirmatory. Judy does not comply or condition for behavioral audiological evaluations, and so we've mostly had to rely on ABR tests to assess her hearing. She was fit with

behind the ear hearing aids around the age of ten months at another facility, but did not tolerate them. She was fit with one and then two powerful bone anchored hearing devices with a soft band here at the age of two years.

She initially accepted them and appeared to benefit from them, but then she rejeCTed them around the age of four years. Her managing audiologist is now trying to have her wear traditional hearing aids again, and she's been met with some success.

Now, before I dive into the case itself, I would like to review some ABR concepts specifically, laterality, excluding the non tester and ABR studies is just as important as it is in clinical audiometry. Appreciating differences between ipsilateral and contralateral recordings can help determine laterality of the age that is, determining the test year versus the non test year. And this is something that was first described by Staples and Rubin back in 1989. And specifically, wave five tends to be later and smaller in contralateral recordings than in ipsilateral ones. These differences can be especially helpful in cases when masking the non test ear is not possible, for example, cases of bilateral microtia and also other masking dilemmas.

And again, before we start with the case, I would like to review the study that I performed on Judy when she was four years old, just to get our bearings here. And on the left side here, what you see is her audiogram and DBNHL. And on the right you see her estimated hearing level audiogram. And in the right ear she has a mild sleep mixed hearing loss. And in the left here she has a severe rising to moderately severe mixed hearing loss.

So there's been a big change since she was diagnosed with those mostly sensory neural hearing loss in the early days. Now, I would like point, I would like to point something out to you here. I didn't mask at 500 hz on the left ear, even though looking at possible risks for crossover here hearing, it might have been a good idea. But I didn't see any evidence of crossover at the time. And I would like to revisit this

concept as I move on to the next study.

So just bear that in mind as we go along. So now let's talk about the test I performed on her at the age of six years. First, otoscopy showed that she has t tubes in place. They're patients. And her tympanometric results show large volumes.

They're not quite as large of what you might expeCT for a child her age, but given the history of down syndrome and other congenital disorders, this is not an unusual finding at all.

And now let's dive into the ABR test itself.

First, let me tell you that I'm using narrow band chirps to do the test, which I've been doing now for three or four years.

But as you can see here, for those of you who are familiar with doing ABR tests, this is a very straightforward subtest here in the right ear, what I have done is I have measured abrs to air conduCTed 2000 hz chirps down to 35 decibels. And in the left here, I've measured them down to 55. Again, these are the ipsilateral recordings only. And this is a very straightforward subtest. There's no risk of crossover stimulation and masking is not needed.

Look, because of the levels of the. I'm sorry, the stimulation levels, the intensity just is not so high that we have to worry about crossover stimulation. And so another thing too here is that I don't really need to worry about any sort of sophisticated analysis of the study itself. But let's move on here. I'd like to look at the same slide but with the addition of the contralateral recordings that are marked with the c.

So for any ipsilateral recording, we have a contralateral recording. And here's what I'm talking about. At 45 decibels here we have a nice wave five early and large. And in faCT, we can even see an emerging

three and one nice and large in the ipsilateral channel. If we look at the contralateral channel, though, the wave five is lower.

And later, again, this is the feature that I'm talking about that gives us some sense of the laterality of the response. And again, as I showed you before, I'm able to track that response down to 35 decibels. And you can see the contralateral wave five is still hanging in there at a lower and later at lower and later in the left here, 55, the contralateral response is lower and later. Again, giving me clues here as to which side I am testing.

Here's the thing. It's not really that important to look at the ABR in this way when you're not really worried about crossover stimulation, but it can really train your eyes. It's something like a process of overlearning, especially when you're working with a case that might be a little tricky. I like to look at the other channel just to get myself set up for when things might get a little confusing. Notice too here that the slightly negative ten or the sn ten is a little bit later here too in the contralateral channel.

So all these little clues to give you a sense of which ear you are testing. And so let's go ahead and plot those quickly on our NHL audiogram. Moving on to 500 hz. I'm going to go ahead and leave the contralateral channels up and we see something very similar to what we saw in the 2000 hz test in the right ear. Again testing at 55, no risk for crossover stimulation.

We see a nice large early wave five in the ipso lateral channel. And in the contralateral channel it is lower and later. Very reassuring in determining which ear. Which ears. The test here.

I am testing the test ear here, the right ear. I track it down. It's really not there at 45, although it sure does seem like it wants to be there at 45 decibels. It doesn't quite meet our criteria for response. Again, 55 is really the threshold here now as we jump over to 95 in the left here.

Well, I did do the test initially and also when she was four years old. I did the test initially when, I'm sorry, without masking because I was very confident the response was ipsilateral. That is, I was testing the test here. Here. Though it could be argued that there was some ambiguity ahead and tested without masking, which I didn't save here, I'm sorry.

But then I tested just by dumping some masking in and see if. To see if there would be a change. And there isn't. As I dump white noise masking in the right ear, I see what I've seen for every panel, for every test so far that the ipsilateral channel shows a wave five that is earlier and larger than it is in the contralateral channel. And so I feel very confident here that even at this high intensity level that I'm really not experiencing any crossover stimulation, that I am truly testing the left ear.

It looks like it wants to be here at 85 two, but again, it's not quite meeting the criteria for a response. So let's go ahead and plot those thresholds on our NHL audiogram this time. Here we are masked at 500, left ear and. All right, so now I hope this was worth waiting for. This is really what I wanted to show you here.

The tested bone conduCTion. I'm sorry, the bone conduCTion test at 500 hz. Let's start with the right ear. This is an unmasked response at a screening level of 25. And really, I can't mask anyway.

Given to the air conduCTion threshold in the left ear, it's just too high. But very clearly, this response belongs to the right ear. Again, as we've been talking about, the response in the ipsilateral channel is earlier and larger than it is in the contralateral channel. As you develop an eye for this, you can see, yes, this response is coming from the right ear. The left ear is different.

This is unusual.

This looks different from anything I've shown you so far with sound to the left ear. With 500 hz chirps delivered to the left ear, I'm aCTually stimulating both ears. If we look at the contralateral channel here, there is an earlier, larger wave five than its counterpart in the ipsilateral channel.

In the ipsilateral channel, there is a very small wave five that belongs to the left ear. And there is a later small wave five that belongs to the right ear. Well, pardon me, I clicked that so it is clear that I am stimulating the right ear at a higher sensation level than I am stimulating the left ear. Because it is so large and it is so early in the left ear, I must be very close to the threshold because of the amplitude of the response. Now, I could poke around and probably proceed with testing this way, but it seems reasonable to me that I should mask just to be safe and to be clear about what I'm testing here.

So let's look at the very same slide. But this time I'm going to add 110 decibels of white noise in SPL to the right ear. I'm not concerned about over masking based on what I measured when I was performing my air conduCTion test at 500. What I know about the effeCTive masking level of white noise at 500, what I see here now at 50 decibels, I see a very nice wave five at 50 decibels and then it's very small later counterpart in the contralateral channel and I'm able to track those down to 45 decibels. Again, very small wave five here in the contralateral channel.

However, I am still able to see a very large wave five that is very late.

This is wave five from the right ear. I am still crossing over to the right ear here, although I have not effeCTively masked the right ear. I'm still getting some crossover stimulation here. At least I have been able to move it out of the, the way so that I can see what is going on here.

And even though it seems to be contaminating the study a little bit, that's okay. I know what the left ear is and I know what the right ear is. This response from the writer continues down to 35 decibels. But it would be a mistake to mark this response as the response for or the threshold for the left ear. It is

aCTually the crossover response from the right ear.

So let's go ahead and plot those on our NIHL audiogram. And just for the sake of time here, what I'm going to do is show you what I measured at 4000. Again, what we're seeing is what we saw even in early childhood that Judy has a, even in the high frequency, she has a partial conduCTive component.

And then if we just compare her EHL audiogram from four years old to six years old. You can see that there's some very good agreement here, and that gives me a lot of confidence that I have an accurate test this time around as well.

Well, I hope you found that interesting and useful and informative and maybe even inspiring and how you approach your ABR tests in the future. I don't get to see something like this very often. Usually there isn't this subtlety in stimulating both ears. And so I thought I'd set this aside and share it with you today. The thing, of course, that is appealing to me, at least, about this case, is that, well, we're dealing with a child here as a complex medical history, and she just did not comply for standard audiometry.

And in cases like that, at least what you can do is hope that an ABR test will bring much clarity. Well, that wasn't quite the case here. Even that the ABR test was pretty tricky, and it requires some poking and prodding in the same way that you might administer standard audiometry to really figure out what was going on. On. Of course, she's.

Her treatment is tricky as well. That is an ongoing process, and I really do hope that what I have done here has been helpful to the managing audiologist. Of course, we might not have seen any of this trickiness had she not had the hearing loss. There are some other challenges. You can encounter neurologic disorders that can make a test difficult, but it was really the hearing loss in this case that really made it difficult to determine which ear was what.

And then finally, what I really wanted to point out here was the importance of teamwork and really working together to figure out and what's going on with a child and helping that child. Well, thank you, everyone, for the great presentations. Anyone in the audience, do you have any questions for us? If you do, just please put them in the chat.

While we're waiting, I'm going to ask a question which came in earlier. Rachel, at what age are we doing oVEMP and cVEMP? So, typically, we're starting cVEMP at six months of age and older, and then oVEMP, we start a little bit later at three years of age and older, just to get more of a robust response. Great. Christina, thinking about vestibular assessment.

Are there concerns for vestibular dysfunCTion for the trauma patient, yeah, for Beth, there was, on the imaging, some note of impaCT to the vestibular struCTures as well. It was a little too much to get into today on this presentation, but we will be bringing her back for vestibular assessment in the future. Well, it'll be interesting to hear what happens next for this young woman. Caroline, what's the prevalence of Boer syndrome? Yeah, so it's not quite known, but it's estimated to be roughly 2% of all profoundly deaf children.

So very, very small percentage. It's good to know about these that don't have a large prevalence, because when they pop up in clinic, we may not be aware of everything that's going on with them. So that makes a lot of sense. That's what this one was, is I had never seen a kid with it before, and this was the first time it popped up for me. Okay, great.

Deb, I know that the FDA criteria has recently changed. What are some of the other new criteria? Absolutely. So we now have implantation down to nine months instead of twelve months. And of course, we have the SSD criteria now for children who are five and older who have normal hearing in one ear and profound hearing loss in the other ear.

Those are the other two kind of newer criteria that have emerged in the last couple of years with SSD. In addition to being severe to profound hearing loss, the CNC percentage needs to be 5%.

Great. Okay, we have a question which was just entered into the chat. Just curious what you all see with third window dizziness thinking about the child who had negative bone after the head trauma. So CVMP is aCTually a great test for this patient population. So we would aCTually decrease the intensity level to see if there's a response present.

If there is, that could be suggestive of a third window disorder. Yeah. We will be curious to see what her vestibular testing when she comes back for. That looks like kind of what we talked about, though, throughout her case presentation was she has a lot still going on. We are in a wheelchair, we are going through lots of therapies, trying to gain some different funCTion back and still some things that won't be gained back.

So kind of the typical, like hearing the ayes or any sort of those complaints that we often hear with the third window were not brought straight forward by her. That was not one of the primary concerns right away, but something we'll be curious to look more into in the future that makes sense. Another comment. This is a comment about the ABR presentation. Presentation appreciated the look at contralateral ABR.

Great job. Despite the technical difficulties, you always have to have some. So I guess it's not a presentation without some challenges.

Bob, is there, what is the best way to teach about contralateral ABR to new clinicians starting out? Well, that is a very complicated question. Just the introduCTion starting out, just the introduCTion in a praCTical mode and then going over it again and again, I think is the most straightforward way. I mean, of course you become very theoretical about it, but I think that in a praCTical environment, it's important

just to go over it again and again and again and ensure that the student understands exaCTly what is going on there. And then you can go from there.

You can get. You can become more theoretical from there, but just really emphasizing the idea that you are looking at the very same abr just from different angles on the head.

That makes sense. Well, if there are not any other questions, we can be done. If you do have any questions, if you go home and things pop into your mind, here's everyone's email address. Just feel free to email the presenter of the case study that you had the question about and we will get back with you. I want to thank all of our presenters.

You all did a fabulous job. And thank you to all the participants for hanging in there with us. An hour and a half is a long time to sit, so have a good day, everyone. Bye.