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EHDI: Vestibular Monitoring Protocol for Cochlear Implant and cCMV Patients

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>> Captioner is standing by.

Vestibular Monitoring Protocol for Cochlear Implant and cCMV Patients

>> I'm here, and I am the room monitor for today and I'm going to make sure everybody is good and comfortable. The host and attendees, the fire escape is behind us. The topic for the presentation is vestibular monitoring protocol for cochlear implant and cCMV patients and I'm going to let them introduce themselves.

>> KAREN HENDRICK: Thank you, everyone for joining us this afternoon. My name is Karen Hendrick, I'm team lead for our vestibular program at Children's hospital Colorado and with me presenting today is Jennifer Drohosky our team lead for our cochlear implant program so thanks for choosing to join us for this talk.

So for our learning objectives today we're going to discuss on ‑‑ we're going to look at the specifically the cochlear implant population, what those risks can entail.

And we will address children with CMV, their risk of vestibular function and what we recommend for monitoring we will also end with a couple of case studies. So first of all, let's talk about the inner ear and why balance concerns are a common issue with people who have sensorineural hearing loss. I like this diagram because it shows the complexities of the inner ear and especially looking at all those little branches that go to different parts of the inner ear. Today we're really focusing on the left part, the part that looks like a roller' coaster that houses our FBI vestibular organs we have in each ear. So we actually have ten vestibular organs between each year. These are sensing movement and gravity in all the different plains so each individual organ has a direction it focuses on perceiving that movement so up and down, forward, backward, and side to side, and then all the angular planes with head turning. And then there is a branch of that eighth cranial nerve that goes to each individual organ. It combines with the part of the nerve that is receiving the sound input from the cochlear and that comprises the eighth cranial nerve.

So why is it more common for someone with sensorineural hearing loss? There are a few things you can tell the inner ear beyond just being complex it's really close together where the hearing organ and the vestibular organs are. They share the endolymph fluid that flows throughout those labyrinths and that's what stimulates our different organs and they also share a similar blood supply. Together they create the eighth cranial nerve and then of course if there are any cranial malformations that can affect both sides, hearing and balance.

We're going to talk about a few different studies today. But all are kind of finding that about half of children with severe to profound hearing loss will have some degree of vestibular function and I'll share couple more stats in a little bit on that. But very, very prevalent in this group.

Here I do want to just say a disclaimer with all of the really fantastic research that has gone into looking at these numbers. It's still pretty limited, and you can see that there are ranges here with the different etiologies of hearing loss. We will be going into depth about the congenital CMV diagnosis and looking at those numbers more closely, but the point I'm trying to make here is that vestibular dysfunction is common with sensorineural hearing loss, and especially with certain types of etiologies so if you are a family member, a provider, a therapist, and you have a patient with one of these etiologies of hearing loss there is a really high chance there could also be vestibular dysfunction occurring as well.

Now let's think about our balance system because it's more than just our ears. I like to use this three‑legged stool analogy to give a good representation. So we think about our visual system, everything that our eyes are seeing is part of our balance. Our proprioceptive system, so how does the ground feel beneath our feet, that provides a lot of input and information. And then also our vestibular system, so how do our ears sense the movement that our body is experiencing. So these three systems are continuously measuring movement or different stimuli, sending that information up to our brain where we have our central vestibular system. Our brain then has to decode that information, make sense of it, and then send instructions back down to the rest of our body so that we don't lose our balance. So just because someone has a vestibular loss doesn't mean they can't balance, but it does mean that we're putting more pressure on those other systems to perform. I'll talk a little bit about vestibular rehab and how these systems play a role because we're really going to try to maximize those other systems to help the child with their balance. But this is just kind of a good analogy to kind of picture, keep in the back of your mind, when you're talking with families about the balance system, it's something easy to kind of interpret.

I want to get into something specific within vestibular dysfunction, and so there can be a large range. You could have unilateral hypofunction, it could be bilateral, it could be complete bilateral areflexia, meaning that none of the organs are perceiving any kind of movement. Or it could be bilateral partial. We're going to talk a little bit about this vestibulo‑ocural reflex. I'll be talk about it throughout today's presentation. So the VOR is a reflex that connects the movement of our ears to the movement of our eyes. If you put your thumb out, stare at it, move your head side to side, you can keep your eyes locked on your thumb because your eyes are actually moving opposite. If you do not have a functioning VOR, and your peripheral vestibular system your ears are not providing your brain with that information. So as your body moves, the brain is not being aware of that from the inner ears, and so they can't give those instructions to the ayes on how to react. The result is something that is called oscillopsia where things in your visual field might look ‑‑ appear as though they're bouncing. I had a patient one time who said on a walk just in her neighborhood she looked at houses down the street and it looked like houses were bouncing with every step she took.

And that's because that VOR wasn't active.

So when we think about this VOR and how do we stabilize the gaze and that's where vestibular rehab can come in and play a role in that, we are not necessarily fixing the problem but providing them more tools and skills to help with that.

Between a quarter about a third of children who have severe to profound hearing loss will have bilateral areflexia, so that complete lack of vestibular input. So remember about half of those kids will have some degree of vestibular dysfunction, and about a quarter to a third of them will have a dysfunctional VOR.

And so we'll be getting to that next point here in a bit but I wanted to take at least a side bar to talk a little bit about vestibular rehab because it plays such a role in the treatment we are providing for our patients.

So vestibular testing, the diagnostic workup is in the necessary to start vestibular rehab but that information really helps provide the framework for the therapist to help them choose the correct treatment strategy. So like I mentioned, depending on if it's unilateral, bilateral, partial, or complete, that does make a difference in the treatment that the therapist is providing. I also want to take a moment to really acknowledge the role of early intervention physical therapists because a lot of these kids that we're seeing, especially what we'll be talking about today, are in that population, and the early intervention therapist plays such an important role in the treatment and help for these children, and we do ‑‑ we do not ever want our families to get the impression that they should stop doing their early intervention PT and start seeing a vestibular rehabilitation specialist because the early intervention therapist is probably working on a lot of things that are very important for the overall development of that young child.

So it is possible sometimes for families to receive both early intervention and private therapy. It does depend a bit on insurance, but as long as they are not using the same treatment strategies and using different codes, that can sometimes work out for the family.

What we recommend is that they always start with going to their early intervention physical therapist and having that conversation about what that might look like. Maybe they have the training to provide balance treatment as well. That would be ideal so they can just kind of stay working with the same therapist and add on that treatment strategy. Sometimes though they say, yes, I really want to work on these goals but we do feel like that's another avenue that they can explore through private PT, and that's where the early intervention and private therapist can kind of work together to ensure that that plan is going well for the child. We don't know all of the ins and outs as the audiologist in that diagnostic appointment so it's really important to keep in mind the overall treatment and therapy that these children are receiving.

So this brings us to our call to action for our team at Children's Colorado is that we want to be providing the vestibular testing to our highest risk patients, those will be our children with cochlear implants and children with congenital CMV, and then helping them get connected with the vestibular rehabilitation early. So I'm going to hand it over to Jennifer to talk about the cochlear implant population.

>> JENNIFER DROHOSKY: Thanks, Karen. So as Karen was mentioning earlier the prevalence of children with vestibular disorders with children with severe hearing loss is eye opening.

I think to us and for someone who has been in this field for 30 years, this is a really large number, and it really can impact the patients that we're seeing day to day for cochlear implantation. So as she mentioned, it's up to about half of the children that can have a vestibular loss and it can be anywhere from a third to a quarter or a quarter to a third of children who have the complete bilateral loss which is really, really impactful. This was one of our earlier studies that discusses these findings, and then there are a few more that have followed up with very similar results. It's exciting to see more research emerging about this population that's been something that's relatively new, so we're excited to continue to see what the research shows and also contribute to it ourselves. The study by Cushing was one of the earlier studies that did point this out and we're going to talk about this in a moment in terms of the impact you could have on a potential device failure for children with a cochlear implant because of these vestibular issues. Jack Cohen also did a study that had similar results in terms of the prevalence of 50%, and again with up to 30% with that bilateral areflexia; which can really, really impact development past a certain point. it can affect reading skills.

>> [off mic]

>> I would, I could e‑mail you the answer to that question later.

>> I don't think, these are not necessarily a lot of itty‑bitties, but they are children. this study is looking at a functional BOT2, and it's looking at how balance is used in day‑to‑day life and how much at risk you are of having a fall. Based on the results of that test. You can see typically developing children or children who have also normal hearing sensitivity are at a lot less risk of having a fall than children with severe hearing loss or children with a cochlear implant and the graph or the bar on the right‑hand side as you're looking at it shows children that have a potential device or have had a device failure because of this vestibular loss. This is really, really interesting stuff, and it's really impactful in terms of how we're counseling families and what they should be looking for. Obviously we never, ever want to have a cochlear implant device failure, and if there is anything we can do to prevent that for a family, that's something that we would want to do. So in a nut shell, the poorer that the score is for their BOT2, the more at risk they are for a fall or a trauma. Now, we sometimes different ways that you can have trauma to a cochlear implant. One is several small hits to the head or falls where kids are maybe knocking their head on the ground. We always talk about toddlers who are learning how to walk and exploring their world, and sometimes they're falling down a lot. You know, we always think that those internal devices are very sturdy. They are, but when they're hit repeatedly, there is a potential for that to fail. The other way that a cochlear implant could be damaged is by one really large blow to the head or fall.

So the takeaway for me and the thing that got me really interested and had we join the vestibular team is children with vestibular dysfunction are eight times more likely to have a cochlear implant failure, and this is due to a risk of injury from falls. That's large. Really, really impactful.

So the takeaway is that children who have cochlear implants are at higher risk for vestibular hypofunction, and these children specifically, they are more at risk of having a device failure because of their falls. However, we can stay ahead of that by identifying those children earlier as well as getting ahead with some vestibular rehab. If it's not treated or if we don't move forward with that vestibular rehab, there does come a point where their skills will plateau especially if they have areflexia, and they will maybe not have as much development in terms of their daily activities. They may not be able to do as many activities as we would like them to do. And we want every child to reach their potential.

Treatment is effective once it is identified. It's actually surprisingly short. There's lots of things that parents can do at home, but it's also really, really effective for those kids, so seems like a big win.

And so with all of this research emerging and with Karen's leadership, we have developed some protocols at our hospital for children who are the cochlear implant candidacy process to undergo vestibular testing so we can rule things out before their surgery. We are also going to be monitoring these kids after their surgeries, but we are hoping to get a baseline before whenever possible. And this is what that looks like. We are going to do a test called a VEMP, and I should just ‑‑ I do want to say that all of these tests are not invasive, not painful. The VEMP test you're going to see an example of that in a little bit with Karen, but just involves just some stickers and lots of dancing by adults in the room to keep the kids focused. And also a HIT test, a head impulse test where we're gendly moving the child's head from side to side and watching their eyes. The results of that testing say no, you can't get a cochlear implant or yes, you do. It's just another piece of information so that parents have everything that they need going forward.

So if our children are implanted before the age of 3 years, we don't do the balance testing after their cochlear implant right away. We wait until they're 3 years of age so we can do a more full and complete test battery. At that time, it would include the vestibular testing as well as the physical therapy evaluation. However, if their pre‑op testing was abnormal, the VEMP or HIT test were abnormal we would bring them back sooner.

When children are implanted three years of age or older, we do see them back in three months. Generally they're a little bit better able to follow directions and maybe have acclimated to their cochlear implants faster, and at that time we do vestibular testing and then a physical therapy evaluation if the results are abnormal.

So Karen's going to walk us through what this looks like. She may not brag, but I think her vestibular lab is one of the coolest things I've ever seen. They've made it very, very child friendly, and so she's going to walk us through what these tests look like and how the children react while they're doing them.

>> KAREN HENDRICK: Hey, I realized I should've had a photo that really showed the whole lab, but you get a little bit of a view here in this video. I'll set the scene a bit before I start play, and we might ‑‑ it's a short video, so we can watch it more than once. Behind us that big red thing that's wrapped to look like a gondola, we are in Colorado, so we want to kind of be on theme there. Inside there is our rotary chair. I have a video in a couple of slides that shows how that operates. And then they also wrapped our walls to make it look like you're in the rocky mountains, so that's a really big component of doing this testing with young kids because it's uncomfortable. It's scary. It's really new. They've never experienced something like this before. so putting a kid into that kind of situation and hoping they do everything you need them to do is not always easy, but there's a lot of modifications you can do to that same test that you would do on any other adult and still get a reliable result. So in this video we're doing a cervical VEMP, cervical evoked myogenic potential. I'm on the left in the clinician role and one of my colleagues Alyssa she is on the right playing our assistant role. She's also doubling as mom in this video that's her 12 month old son and her husband who's playing the parent. And we are, the way that the VEMP is set up is with sticker electrodes on the forehead, on the sternum and on either side of the neck on these sternocleidomastoid muscles that run on either side of your neck. We stimulate one of the organs in the inner ear by playing a loud sound stimulus. You can use air conduction or bone conduction. We find that bone conduction in the pediatric population is really useful because these kiddos are more likely to have middle ear problems, more likely to have tympanostomy tubes, and kiddos with a cochlear implant can have a very, very slight air bone gap at the exact frequency that we use to test. We use in this test. So there's lots of reasons why a bone conduction can be a helpful stimulus for these kiddos. It's worn on a head band so in my right hand you see this white washcloth. That's just there to provide some padding and support for a smaller head, and in my left hand there is going to be the little black box, the oscillator that plays the sound. So here I put the oscillator on. I'm going to try to get the infant to contract his neck muscle by looking over at the iPad, and I'm just kind of gently bracing his head to get a stronger reaction of that neck muscle. The test runs for about 15 to 20 seconds per run. So you can see it's already done there, and normally if I was really doing this test, I probably I'll just play it again while I'm talking, probably would've gone right into my repeat since he was so engaged in the iPad. We want to always repeat our response, make sure it's a true response and not some artifact that might've come in. If the child is a little upset or not really engaging with the iPad, we can always takes a many breaks as we need to in between those trials.

The next slide shows just a slightly different seated position. I'm still doing bone conduction but I'm hand holding it, and the dad is actually kind of leaning the infant back, and the infant is forced to then use their neck muscles to keep their head up. Oops. let me go back and play. There we go. He doesn't get a lot of screen time at home, so he was really into the iPad in this video. But you can see it's relatively noninvasive. It's just using a sound stimulus and electrodes don't really feel like anything, they don't get hot or shock the person. They just measure the input in the body.

So this test we can do quite young. There are some centers that are going six months or even younger. We just need them to be able to contract their neck muscle and this guy in the video is only 12 months old.

So I mentioned the rotary chair. Now usually the testing is done with the door closed, so this is just kind of an example to see how fast the chair goes. The reason for, and this is, I'll say this is the fastest the chair ever goes with the different frequencies, it just changes how many rotations you're going in each direction. But it's nothing wild. This isn't Disney land. We tell our patients there is no flippin', there is no floppin', there is no going upside down. The thing that's kind of scary about it is it is done in the dark. We always will have a test assistant in there with the child. If they want. And younger kids we always do. But the door, when the door closes and we turn that light off, it's a pitch black environment, and that is why we have the enclosure. We need to have a blacked out room. I'm wearing V&G goggles which are actually recording my eye movements. I'll just keep playing it since it's such a short video, and that's ideal, but those goggles, we do have a pediatric size, but they don't really fit faces that younger than three years of age all that well. So the other option we have is ENG, electronystagmography, so we have a couple different ways we could do that test. We could have them in the car seat, or on a parent's lap or the assistant's lap. We always worn the parent that you're doing the test too if you are the lap in there, so if you have motion sensitivity or anything like that, we might want to have our assistant give it a try. Obviously we wouldn't have any screen in front of the child while we're doing the test. We do need them in that pitch black environment, and then the electrodes instead of having video of their eyes, we can measure that nystagmus response through the electrodes. So it's a great modification. We can test virtually all ages. We're going down to about eight months is about when we start seeing these kiddos for testing, and this information is really, really helpful for looking at that VOR. We'll show you in our cases in a little bit what an abnormal rotary chair looks like, but if we are not seeing those eyes moving in response, which keep in mind as our body moves, our eyes are going to move opposite to stabilize our vision, in this pitched out environment, when we're spinning in a circle, our eyes can't go opposite in a full circle, so we see them kind of go to their limits and bounce back. And that's that nystagmus response. And that's exactly what we're measuring. There's other details about how fast the nystagmus is occurring, how far it's going, at what point does it start changing in the other direction, that will provide us further diagnostic information but overall this rotary chair is a really helpful piece of testing that gives us an idea of how that VOR is working and also how their brain is compensating if there is a unilateral weakness.

So we're shifting gears here to talk about our CMV group. I know there have been fantastic groups already and there is more to go today and tomorrow about this population. It's a very hot topic right now. There's lot of legislation going on currently to get either targeted or universal screens added but we're just really focusing on the vestibular side of things in this talk. Just to give a little bit of background, I'm sure you guys are all familiar with some of these stats here, but CMV is a common virus that we've probably all had some kind of interaction with in our life. It's exposed and passed through bodily fluids, most commonly through saliva. If a pregnant woman contracts a novel strain of the CMV virus, they can pass it to their unborn child. It occurs ‑‑ and this study is fresh off the press.

I love getting the current year studies in there. They found that about 1% of all live births, all live births, have congenital CMV. That turns out to be about one in 200 births. Approximately 11% of those born with CMV are symptomatic at birth. And the other 89% are asymptomatic.

In addition to hearing loss and vision loss, there is quite a range of other possible impacts of congenital CMV on the brain, liver, spleen, lungs. It can cause ‑‑ anywhere from no impact at all to very concerning life limiting impacts. It is the leading cause of hearing loss and neurologic disabilities worldwide.

So we knew that this was a group we really needed to target. We really needed our attention on them, and to see how can we best serve these children. It is the cause of 25% of all congenital sensorineural hearing loss and can be progressive. This is a massive amount of kids we are seeing and the progressive nature of it. So with cochlear implants, yes we are seeing them before their surgery. We're seeing them at some point after their surgery. But this is a slightly different group where there's also the need for monitoring even if it does look normal.

So here are some studies. I'm just going to pull them all up here. I love systematic reviews too because they do a lot of the heavy lifting in a literature review but kind of looking at the original researchers for these study. Bernard et al. was looking at the symptomatic children. I will say these numbers may be slightly inflated because their population was primarily those with balance concerns, so I will be really curious to see some other studies try to replicate these numbers, but 92%, I mean, that is a staggering number of these children with symptomatic CMV also having vestibular problems. Pinninti found with asymptomatic children 45% of them were showing signs of vestibular dysfunction. So even if these children are not showing hearing loss or other symptoms of this disease, there is still some underlying vestibular dysfunction going on and we won't know until we test them.

The other really key thing from some of these studies was looking at the progression. So one study looked over, they did serial testing over 10 months and showed 10% of the kids shows a progression in their results. And Bernard found in little over two years time, half of them progressed. So we definitely know that this isn't a one and done test, we're good. It's something that we are going to have to monitor.

So when we were putting this protocol together, we had lot of considerations. Now let's go back to thinking about that VOR with that bilateral areflexia. It's not just balance. It's not just the risk for an internal device failure if they have a CI. But we start thinking about other things like academics, learning to read. Can you imagine how hard it is to learn to read if every time you get bumped or you move your head, you lose your spot on the page? That oscillopsia is going to be a really hard thing to combat when you're trying to learn as a child. Looking down at your desk, looking up at your teacher, any kind of body movement, those things are going to create an added layer of challenge for those kiddos. So we have those kinds of things in mind. We also are thinking about the research, the studies showing the prevalence, showing the progression. So there was a lot of thought that was put in when developing this. We know that vestibular hypofunction is common in this group. We know that it can occur even if there are no symptoms of the disease. We know that it can be progressive. We know it can cause problems in reading and academics. It leads to that risk of an internal device failure if they do have a cochlear implant. And that it can improved with vestibular rehabilitation. It cannot be cured. Same as with hearing loss. Putting an implant or a hearing aid on is not restoring the function of those inner ear hair cells. Similar with vestibular dysfunction. Rehab is not going to fix the problem. It's going to give them more strategies and more skills to be able to maintain their balance and maintain their gaze stabilization.

So similarly, let's look at what we have put together for these kiddos. So we want to see children with congenital CMV at 12 months of age, and we will do that cervical VEMP test, the one from the video with the 12‑month‑old and the head impulse test. And I apologize I don't have a video of the HIT test, but let me just describe it to you. Basically the infant is going to look at a sticker or something interesting on your nose, oftentimes you're switching out stickers in between to get something novel and more exciting. You are actually facing the child with your hands on either side of their head and doing a short, fast unpredictable head movement side to side, so a short, fast movement. Parents joke that we're like a chiropractor or something but we're not doing anything near to the amount of head turning that it would take to actually crack your neck or anything. A normal, healthy VOR, the infant will be watching your nose the whole time if they're engaged with your target. If they have a, an absent VOR, then every time you move their head one way or the other, you're going to see their eyes drift off and come back on the target. It's called a catch‑up saccade and it's very obvious when it's bilateral. It can be hard to see when it's unilateral, but bilateral kids, it sticks out pretty obviously.

So one reason why we kept it to those two tests and keep good access for families. Our rotary chair is in our ‑‑ location in Aurora at our main Children's hospital. And we will probably never have another one of those. It's a very expensive piece of equipment that we're very lucky to have one of them. It's the only enclosed rotary chair in the state of Colorado as far as I know at this moment. So we have expanded services into Colorado springs and into Broomfield for these partial screening appointments where they can get VEMP, vHIT or HIT testing going. We're able to see the kids, keep access good and if we're seeing abnormalities that's when we would recommend they come down to do the rotary chair testing. So assuming that at 12 months, everything looked great, no abnormalities observed, we will recommend that they come back around three years of age.

Now if something changes during that tige, if they have a big change in hearing, if they start to plateau or regress in their gross motor milestones, these are things we would want them to come back sooner for. Simultaneously, we also are monitoring their hearing pretty closely every three months until they're a year of age, every six months until they're three years of age, and annually until six. And so all of our 47 audiologists across our network of care are trained in asking gross motor milestone questions so that they can be having these ongoing conversations with families during these follow‑up monitoring appointments, and if any concerns are emerging, that's when they can make that referral for more testing. At three years of age, they're older and bigger our video test goggles can fit kids 3 and up. We can do an occular VEMP which is similar to the cervical VEMP. We just put the electrodes under their eyes and measure ‑‑ the way we flex eye muscles is by having them look up at a target actually. And then we can do the rotary chair test: they are also aging out of early intervention at that time, so it's a great time for them to get a PT evaluation as well. We do have the ability to coordinate those with families, so especially if they're coming from the western slope or another state or just anywhere that's farther away, we can put those together for them.

And then assuming at three years of age it all looked great, we still want to see them back one more time at seven. At seven years of age, they're now old enough to do our complete vestibular battery. That would include the VNG battery that has oculomotor testing, positional, calorics, high frequency head shake test as well. The reason we don't do those on younger kiddos is it takes active participation on their part to get reliable results. They need to be able to follow directions, ensure we make it fun instead of a red dot they're looking at a Mr. Potato head or a sunflower or something a little more exciting for them, and we still have many other modifications we're doing throughout testing to make it more fun and less scary. But this is the age where finally we can get it all. And we know with CMV there is that risk of central problems as well, so it's important to get this last piece that really kind of wraps up function altogether.

And then to add the caveat if things are looking abnormal we might have recommendations for more testing or follow‑up outside of these kind of age ranges.

So from here, this is our starting point. I do suspect that over time, as we do more research, and we learn more about the evolution of this disease in these kiddos, that we may have more recommendations for monitoring. I highly suspect that will be coming at some point, but we knew we needed to start somewhere. We knew that this was a big problem and we needed to get families into us, and this felt like it would work well in terms of the access we can provide families and the information that we can give them throughout these early years of development.

I mentioned that our audiologists are asking gross motor milestone questions. We have a few very pointed questions that we ask. When are they sitting and when are they walking by? If they're not sitting by about 8 months, if they're not walking by about 15 months, those are some red flags on top of having the CMV or sensorineural hearing loss diagnosis. So it's really good to just have kind of an idea of gross motor milestones, early ones for these kiddos because that's a really easy thing you can ask in your audiology appointments. If they have CMV or sensorineural hearing loss and they're delayed it's good to consider a vestibular evaluation.

And I hinted at it in the previous slide of we hope that we'll have more information soon, so we're working with a couple other colleagues at Cincinnati Children's, at Children's hospital Philadelphia, and the university of Michigan who have similar test batteries and philosophies on monitoring these kiddos. We're combining all of our CMV data so we can have the largest in as possible. From this, we are hoping that we'll have better data to kind of direct these monitoring recommendations. Hopefully by this time next year, we've got some really solid data to share out with you all. On that note, I'm going to hand it off to Jen to do our first case, and then we have one more after that.

>> JENNIFER DROHOSKY: So this is a case that combines both of these diagnoses, someone who was a cochlear implant candidate but also he is diagnosis of gentle CMV. We're going to call her Cricket, and she was born early due to pre‑eclampsia and she had a very low birth weight which fortunately for her led to CMV testing. It's not standard in our state yet. And because of that, she started on an antiviral medication, was seen by her infectious disease department, and was monitored very closely, mostly for her hearing as well as her vision. I'm not going to go through each of these steps, but basically her hearing loss did progress despite being on the antiviral medication. It started in one ear and then the other ear caught up. And by 15 months, she was fit with hearing aids in both ears.

Cricket however is just a delight and was developing speech and language just amazingly well. She has a lovely family who is very, very on top of her overall development, and they were noticing that she was starting to fall behind. Because she had been on such a great trajectory. So around the age of 3 they went through a cochlear implant evaluation. This was going to be for one ear only because she had nearly normal hearing in the other ear. A mild hearing loss in the other ear. It's interesting because at that time she had age appropriate speech and language skills for receptive and expressive language. But her family was noticing that she was not able to localize as well, and she was also having some gross motor delays. I will say that this was about five years ago before we had all of these protocols in place, so when you look back now, I think oh, gosh, if I only knew then.

But ultimately she did bet a cochlear implant in the left ear, she has a mildly high frequency hearing loss in her left ear that she has had since she was a toddler but we do tend to keep a good eye on that as far.

As far as her gross motor milestones are concerned, she was very behind. She was in PT from the get go. She had early intervention for physical therapy as well as her hearing. She was not sitting up on her own at 12 months, and we're hoping to see that by at least 8. At 14 months, she still wasn't really sitting unsupported, and at the age of over 2 years, she started walking. But bear in mind, her speech and language is way ahead of the game, so just a few, what's the word I'm looking for. Discrepancies in terms of her development.

So recapping her physical therapy and motor milestones, she did have a PT evaluation outside of early intervention before her cochlear implant surgery, and it was shown that she had pretty poor endurance as well as muscle strength for her age. And really she was as a stubborn little nugget and pretty unwilling to do anything that really challenges her balance. That was then and that is still the case today. So after her cochlear implant surgery, which was smack‑dab in the middle of COVID, she started physical therapy through the hospital. Family was continuing on to do exercises at home. Like they were really, really working hard on her balance. She would make some small gains but really plateaued after a year and a half of therapy, and this led her physical therapist to think hmm maybe we should do something different and they recommended an evaluation with our vestibular PT. Meanwhile the family was enrolling in dance class, karate classes and gymnastics to keep her challenged and moving forward with her balance, but along with her CMV diagnosis she was very small in her size and really strength, physical strength was not a strength of hers.

We won't necessarily go through all of these tests, but in a nut shell, her vestibular system is not great. She has bilateral areflexia, which is probably a big reason or is the reason that she was plateauing in her progress. That's a lot of times what we will see when kids have that diagnosis, they'll make good progress for the first couple of years, but there does come a time when they plateau in those skills. And we don't want that, so cricket did get enrolled in PT vestibular therapy. She did intensives of that. And when she was five years of age, she also was able to do some of our at the testing with Karen and her colleagues in the lab, and this is not Cricket. This is another colleague we borrowed for the day. But basically in a nut shell, all of Cricket's tests came back abnormal, specifically her VEMPs. These are not normal results. Nothing is repeatable. Her video had impulse tests, so again where we're moving her head very low gain, and that showed that for all six of her semicircular canals, so her balance organs are not working in any direction. She also completed the rotary chair, which looks at angular function, and all of those were absent and abnormal, so Cricket has areflexia, which definitely impacted her development, and I have to say, this was probably my biggest a‑ha moment as a clinician. I thought we were doing all the right things. We were monitoring her hearing. We had a cochlear implant. She was in physical therapy, in early intervention. She was doing all the right things but there really did come a point where it needed to be something else and I would never have known that had the vestibular team had not come along. So huge learning moment for me.

At this point, this is her most recent test. She's doing very well. Her hearing has stayed stable for the most part in her right ear since she was a toddler. She has nice benefit from her cochlear implants. She wears both her hearing aid and implant full‑time, and she dances. She loves to stream music from her mom's phone and will dance. And she just I think has really thrived because of this testing and because of the specialty care that she was able to get but also because of her family. They are just beyond phenomenal in how they work with her. And given her everything that they can for her to succeed. So turn it back over to Karen.

>> KAREN HENDRICK: All right, we're getting close to the end of our time, so I'll go through this case kind of quickly here. We have little Molly. So her start was a little bit different from Cricket. She was born to a young mother, induced at 37 weeks due to severe intrauterine growth restriction which is one of the those red flags for congenital CMV. And big kudos to that doctor who decided to test for CMV based off of that red flag. And it did come back positive. She actually passed her newborn hearing screening in our NICU where she was for about one week, but due to that positive CMV diagnosis we did recommend a diagnostic hearing test done at 22 days of age. Although her hearing was normal, she had absent distortion product otoacoustic emissions in her right ear. So when you see that with a normal tympanogram that makes us a little suspect that there could be something going on in the inner ear that's affecting those inner ear hair cells. The PCP did refer the family to infectious disease, but unfortunately, she was lost to follow‑up at that point. You can imagine there are probably some challenges that family was navigating.

She did return at 20 months of age for a repeat hearing test, and at that point, they found a mild hearing loss in the right ear, so those absent OAEs did progress to some mild hearing loss. The left ear still has normal hearing, but we're starting to see a change in those OAEs.

At that 20‑month hearing test, our audiologist also asked about gross motor concerns, anything there, which there were concerns. At that point, she was still not walking, so she's pretty late. She's already 20 months. And the family did have concerns about her balance. So even if we didn't have the CMV recommendations, we would've recommended the testing based off of some of those red flags that the audiologist identified.

We got her in right away, so I think we tested her a week or two later in our lab, and as far as her cervical VEMPs, it was absent in her right ear but present in her left ear. It's a, the wave forms we see on our screens. We did the head impulse test and found the catch‑up saccades in both directions. And then the rotary chair test, you can see all those numbers at the bottom of the screen there, that's telling us that all the frequencies we tested, we were not really seeing a nystagmus response. So despite still having normal hearing in her left ear, this configuration of the rotary chair is something you would only see if it was bilateral.

It's not areflexia because she still has a cervical VEMP in her left ear. That's giving you information on up and down movement. But the VOR is affected. The semicircular canals are not responsive.

So from here, she is already in early intervention PT, so we recommended that they talk first with that provider to see what their options were, and they are awesome. They did so much background work and connected with the family and insurance and found they can seek additional vestibular rehab as long as they're working on different treatment strategies and using different codes. So she is in the process of ‑‑ or she already has had her evaluation actually. That was done at 22 months of age. They found doing Peabody developmental motor skills although she is 22 months of age she has the balance equivalence skills of a 13 months old so very, very delayed and they listed out a number of different interventions they are going to be working on with her. You can see anything from the gait training and to the balance training. They also are going to look at gait stabilization exercises. She's got fitted for foot inserts to kind of help with that foot and ankle stability. So a lot of really great things that they're able to add on in addition to her early intervention PT.

This is a more recent case, so she's actually scheduled next month for a sedated ABR to really get comprehensive hearing evaluation. And we will have her come back for additional testing when she is a little older and we can do more of our vestibular test battery as part of our monitoring protocol. So we might have time for a couple questions but I also wanted to let you know that we do have a booth table down in the vendor hall on the left side, and we'll be there this afternoon, so if we're not able to connect or you're not able to ask your question, please come visit us there. We will also be there tomorrow, and I did put some business cards on the back table back there so if you want to snag one and shoot me an e‑mail at any point, that's totally fine. All right. Any questions. Thank you.

>> You had mentioned follow‑up with, if they had changes in their hearing. How often do you suggest those follow‑ups?

>> Oh, that's a great question. So if there is a big change in their hearing, we probably would want to consider repeat testing, and then from there, it kind of depends on what the results look like. We also keep in mind there are certain ages where we can and cannot do testing, so one reason why we might be holding off is because we've already done that test and it was absent. You can't get more abnormal than something being absent, and so we might also just have to wait for them to be older to do more tests.

>> Have you guys seen a trend in approvals, denials, or in between in the time since you started?

>> That's a fantastic question and I don't have enough data on that yet. We just rolled this out in August of last year, so now we're starting to really see them coming in is more but that will definitely be something we're paying attention to.

>> So this is obviously amazing and you guys have a great setup.

>> Thank you.

>> But most people seems like don't have this. What, what do families do who are in areas, I mean, I'm in a main city, and they told me if I want vestibular testing, I've got to drive three hours and even if we got those results, and they said yes, you need something, the providers locally don't know what to do about this. So what do we do for those families?

>> Yeah. That's a, such a great question. So one thing I say is you don't have to have diagnostic vestibular results to start rehab. Yes, they do help the therapist develop something more customized but it's not completely necessary. There's a lot of functional tests the therapists can do to identify areas of weakness, so if you're not in an area where there's testing and it is true, there is very few centers across the United States right now that can test this young with children. You don't have to have that to get started. Now, for therapists, there are vestibular competencies and trainings that they can do to get that certification or get the knowledge for that, so that would be something that I would start encouraging the physical therapists in that area. There is going to be more of these kids getting identified, a bigger need for that more specialty treatment, and so on top of having their physical therapy degrees, the competency or the certification takes it a step further in really honing in on that. And there are building more resources online. There is a vestibular special interest group that does an amazing podcast that really hit a lot of great topics, so I'm, my hope is that over time, knowledge and awareness spreads, and that it becomes something that is more accessible to families. Yeah, great question. Do we have time for one more or? Might be it. But yeah we will be downstairs at our Children's booth, so please come find us. Oh, there is one ‑‑ if anyone had one last question. In the back.

>> Hi. So a number of the kids I follow with CMV have ended up having a cortical visual impairment so how do you maneuver around that?

>> Yeah, that is an added layer to some of that as well. So we do think about that, and that can be an added component thinking about that three‑legged stool, our vision is a big part of our balance as well, with testing too though some of our tests they have to look at a target, and so we have bigger targets maybe like that size rather than the little tiny sticker that they might use in an adult evaluation, those, and then we can find like the biggest, brightest, the Cocomelon head is actually a really great one. (Laughter). And then we have a white magnetic background that we can put that and we have a little magnetic strip on the back of that laminated target that we can put at whichever height we need, and it's this bright pink and green kind of circular face that's really obvious for those that have some cortical visual impairment. Other tests though like the rotary chair test don't require any kind of vision, so we can actually do that test on children who are legally blind and still look for that nystagmus response, but that's definitely one of the many considerations in testing them. Awesome. Great questions, guys. Thank you for attending and find us if you have more.