Cochlear Implants and Children with Vestibular Impairments, presented in partnership with Seminars in Hearing

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- [Moderator] Is my great pleasure to introduce today's presenter. Dr. Sharon Cushing is a pediatric otolaryngologist and director of the Cochlear Implant Program at The Hospital of Sick Children in Toronto, Canada.

- [Sharon] What we're gonna talk about today is one of my favorite topics and that really is thinking about vestibular function and the impairments that children with sensorineural hearing loss get, and so this is a subject that's near and dear to my heart so I'm so happy to share it with you. So these are my disclosures and we always start our lectures off, you know, because each one of us has a team behind us and I think that it's really important for us to acknowledge those teams and we have an amazing research and clinical team at The Hospital for Sick Children, and it is really a reflection of their hard work. So this is our lab and these are our clinicians and they really do make it a joy to come to work every day and look after these kids with hearing loss. So we have some learning objectives and again, what we're gonna do is focus on vestibular function in particular, in our children with sensorineural hearing loss. And so at the end of this, I think my hope is that we'll all have a better understanding of who is at risk as well as what are some quick and easy things that we can do to identify these kids 'cause no doubt, we all work in a world where time is at a premium and we really have to be efficient with our time.

So we're gonna start off by looking at the prevalence of vestibular and balance dysfunction and how common it is in sensorineural hearing loss. We're gonna think about what the impact of that dysfunction is, and this is just gonna be the beginning of a conversation that will continue in another lecture later in October, October 28th, I believe, where we really dig deep into what this means for those children. And we're also then gonna start with some practical tools as to how do we screen for dysfunction tomorrow when you see a child. We'll give you some tools so that you can help to identify these children. So I like to start with this slide and you might wonder, well, why is she showing bilateral implants when she's supposed to be talking about vestibular function? And I think this speaks to the story of how we came to really think about vestibular function in children. And so there are experts in vestibular impairment in children and there's a rich, rich literature but it was a pretty niche area of interest in pediatric audiology, pediatric otolaryngology and neurology, for example, however, when the advent of bilateral cochlear implants came along, it really provided an impetus and an increased and broader interest in the topic of vestibular function in children. And the reason why is that as surgeons we were very worried about the possibility of inducing bilateral vestibular injury by the act of either simultaneously or sequentially putting in cochlear implants. And that was certainly the impetus for us here at SickKids to look at the degree of function and dysfunction in this population. And it's very interesting to me because we set out to answer this specific question, so how do cochlear implants impact vestibular function? And while we were able to answer those questions and I wanna talk briefly about what that data demonstrates, we found so many more interesting things. We got answers to questions that we didn't even know we should be asking, and so it really goes to speak for the idea of asking a question and following through with the answer and all the joys and interesting things you might find.

So here is some of the data in the literature looking at the vestibular effects of cochlear implantation. So this was the question that we thought we should be asking. And indeed we know that putting an electrode as gentle as we think we are as implant surgeons into the fragile inner ear can impact vestibular function, and here's some examples. From a microscopic standpoint, we see fibrosis of the vestibule following cochlear implantation, and these are obviously postmortem studies. We see also loss of vestibular function following cochlear implantation in this study. Again, 20 to 100% of individuals lost their saccular response following cochlear implantation. And again, here's another example where you can get distortion of the saccular membrane, which is likely the pathophysiologic correlate of the lost vestibular response. And when we looked at these kids, we began looking at a cross section of our children who had, some of them had implants already, others did not, but this is what we found. So if we break it down into each of the end organs and we think about function in the horizontal canal, in this example, it's being measured by caloric testing. What we saw was that about 50% of children were abnormal and 50% were normal, and you'll see this divide come back again and again. And then when we further broke down this section of kids who were abnormal into specific dysfunction, more than 1/2 of them had complete areflexia, so no response whatsoever from the horizontal canal. 20% had hypofunction and again, this is less worrisome and has less functional impact. When we look at this group of kids with hypofunction, again, they span the spectrum of severity with under 1/2 of them either having bilateral hypofunction or severe unilateral hypofunction and the majority having either mild or moderate dysfunction. And so these children in the red are certainly the ones that we worry about. And so when we look at them as a cohort, what we see is that 38% of them, so just under 40% had significant dysfunction of the horizontal canal in response to caloric testing. Now, we can also measure horizontal canal function in response to rotation, and that's how we measured it when we first began to do this study in the early 2000s. Nowadays, we don't use rotary chairs so much anymore and we'll talk about some of the newer types of testing that we can do that I think are particularly amenable to children, but for this data, we're looking at rotary chair testing.

And again, what we see is that roughly 1/2 of them are abnormal and 1/2 of them are normal. And what I'm showing in this graph here is that we have the capacity in Toronto to look over a much larger frequency spectrum than many rotary chairs do clinically. And what we see is that these shaded blue area is essentially the normative data by frequency and what we can see here, this red line, is essentially where most commercially available rotary chairs stop in terms of their frequency, .6 hertz or some of them will go up to one or two hertz. And what we can see is that all of these children here in this example look normal, all four of them appear normal, but once we begin to increase the frequency, what we see is that some of them only fall off at the very highest of frequencies, these two children here. Whereas others, again, once we get out of that relatively low frequency range, also fall off. And so I think it's very important to have an eye to high-frequency horizontal canal function, and we'll talk about how we measure that and some of the techniques and technology that allow us to do that so much better today than we did when we first started this study. The other part of the inner ear are the otolithic organs and there's two of them, there's the saccule and the utricle, and they are sensors of horizontal acceleration and vertical acceleration so linear plane. To families I call these the elevator receptors. It's the receptor that makes you go whoo in your stomach when you go up and down in the elevator. And again, a very familiar proportion of individuals had abnormal function. So again, we see 50% roughly abnormal, 50% normal. And when we break down this group of abnormal kids, what we saw was again, about 1/2 of those were single-sided and 1/2 of them had bilateral dysfunction.

And so what we see is that these end organs in children are quite impaired. And so this led us to the first aha moment in this research, because I mentioned that much of this testing was done on a cross section. Some of these children had ears that had implants in them, and some of them had ears that didn't and what we realized in that moment is that as much as we were worried about injuring their vestibular function by putting in a cochlear implant, their deafness had won the race in most cases. And so the etiology of their deafness put them at much higher risk of having vestibular impairment than I ever could as a cochlear implant surgeon. Doesn't mean that we shouldn't be mindful of the impact implants can have on the function in children in particular and it's a completely different story when we start talking about adults, especially elderly adults but by far and away, their deafness was their biggest risk factor for their vestibular dysfunction. And so when we look in the literature what we see is that about 70% of children have some degree of dysfunction, this is all comers, this includes the milds, the moderates, the unilaterals and so that's the overall prevalence. But when we really hone in to those that I highlighted and read in the last few slides, those that have severe dysfunction, what we see is that that's 35 to 40% of children have complete non-responsiveness of the vestibular portion of their inner ear. That is very impactful and we'll talk about what that looks like from a day to day life perspective. And so if we put this into context, what we see is that vestibular impairment is the single most common associated feature of sensorineural hearing loss and yet in many cases, children who have hearing impairment will never be assessed for it. And as clinicians we talk about, it's on our exams all the time, what evaluation should you do for a child who comes into your clinic with hearing loss? Should you get an electrocardiogram to look for long QT syndrome? Do you need to get renal function or renal ultrasound for those very few disorders that are associated with renal impairment? And we debate the utility of those tests, but we don't really talk about the fact that we should really be at least screening these children for vestibular and balance impairment. And so why is that the case? Well, I'll tell ya, part of it is that it's easy for me to order an EKG. I just tick a box off in our electronical medical record and it happens, the interpretation is not always so clear, but it doesn't impact our team in terms of manpower and work hours to order that test.

Same thing with renal ultrasound, same thing with urinalysis, but testing vestibular function in children can be a bit daunting. And for many people they just think, oh, how can we possibly develop the expertise and the time that it takes in order to do that? And I'm hoping that that will continue to change, it already has, the degree of interest in looking at vestibular function in these children has increased immensely since we began doing this work in the early 2000s and I really hope that it will continue. And so this next part what we're gonna talk about is children don't really care what their horizontal canal is doing or what their utricle or saccule doing, what they care about is navigating the classroom and the playground. So from very early on we felt it was very important to have a measure of the output of the system, you can consider this sort of the behavioral correlate of the audiogram or the vestibular testing. So looking at balance is our behavioral correlate, just like speech and language is the behavioral correlate for hearing. And so we have a test that we use and we're gonna talk a little bit about it, but I wanted to start by showing you this video. And so let's watch this video together, and this will be of two young boys. And what you can see is that they've been matched for age, gender, and t-shirt color. They're a little bit different in size but as I said, they've been matched, but what you can see very quickly is that they have distinctly different balance skills. The child on this side of the screen, you can see me in the pane, he's needing my help in order to stay upright and you see how he's moving his head all around, you almost think that he's putting on in terms of not being able to do it and his skills are very different than this child over here who doesn't need my help at all. And so this is a really good example of the differences that we can see in children when we use these standardized tests. And so let's go back to the presentation and we'll talk a little bit more about these children and about this testing that we've been doing on these children since we've taken an interest in vestibular function. And just to give you the clinical context of this video, this young boy, just looking for my pointer, I'm gonna master this by the end. This young boy on the left hand side of the screen has Usher's type 1 syndrome and he has bilateral vestibular impairment, bilateral sensorineural hearing loss and bilateral implants and he is going to lose his vision over time to retinitis pigmentosa. And so you can imagine that if his balance skills are already this poor and his vision is still intact, this is gonna be a very difficult thing for this child to have this multi-sensory deficit.

And so we applied this test and the test that I just showed you is called the Bruininks-Oseretsky Test of Motor Proficiency, it's called the BOT-2, and we did this in a number of children who had hearing loss in our cohort as well as repeating a normal hearing cohort. And this test consists of nine different tasks, three of which are performed eyes open and eyes close and what we can see is the results in terms of an age standardized score, where we can compare a single child over time and we can also compare children of different ages. So it's a very useful and powerful tool. I can tell ya that it's simple enough, 'cause I'm not fortunate enough to have a physical therapist who works for me, and so it's simple enough for me as a surgeon, and that's setting the bar pretty low to do in about 10 minutes and all you need is the balance beam and the normative data. I now have the good fortune of working with an amazing vestibular audiologist and this has now come under her umbrella as part of her evaluation. So I think a clinician of any background, doesn't have to physical therapy, can undertake this testing. And so let's look at the results together. So we did repeat a group of normal hearing children, and as I said, we are able to use the standardized scores but we wanted to make sure the test environment had the appropriate fidelity. And then this is a group of all comers in red here of our children with sensorineural hearing loss. Some of these kids have implants, some of them do not, it's really a mixed bag. Some of them will have normal vestibular function, some of them will be in that 40% of the group which has very significantly impaired function. But what you can see is that there is a significant difference between these two groups in terms of their overall function. Now, if we are more selective and we only look at the balance function of children who had bilateral vestibular loss, so again, that's the 40% of kids with very severe impairment, these are their balance scales. And so what you can I see is that they are an order of magnitude worse than both the kids with hearing loss, as well as with the kids with normal hearing.

Now, you saw that young boy who would be in this cohort here and you saw it how difficult it was for him to perform some of those tasks, but if you just watched him come into my clinic, you might not have noticed anything different and that's often what people will comment when they see our data from Toronto is they'll say, "I don't know what you guys are doing up in Canada, but we don't see this in my hearing loss clinic." And the reason is that if you look for non-ambulation as the marker of vestibular impairment in children, you're gonna miss it. A child with bilateral vestibular loss looks very different functionally than an adult with bilateral vestibular impairment. And so we looked at what the age equivalency of these balance skills, and this helps to explain why non-ambulation is not a good marker. And that's because children who have bilateral vestibular loss will develop and plateau a set of balance skills that are on average those of a 4 1/2 year old. Now, if you can think about what the average 4 1/2 year old can do, they can certainly walk into my clinic, in fact, they'll often run into my clinic. They probably don't ride a bike without training wheels. They might ride a scooter, but they certainly can ambulate and so that's why we often tend to miss these kids. We have to make the test hard enough. We have to get them to stand on one foot. We have get them to close their eyes in order to be able to see the deficits. And so that's really one of the most important things that we've learned is that non-ambulation isn't what bilateral vestibular loss looks like in children. Now, we're gonna continue on that theme with another case story, again, where we know that vestibular loss impacts balance, but these balance deficits are not always apparent. And so I'm gonna show you the story of this young man, is a young boy with bilateral cochlear implants. He was deafened early as a result of meningitis and he swam for Canada in the Deaf Olympics. So, you can't quite make out their profile, but he's got his implants on underneath that bathing cap. And so what I'm gonna show you, and we'll watch this next video together, is this dichotomy of balance performance. So here he is on the left. I'm asking him to stand on one foot with his eyes closed and he can't do it. A five-year-old should be able to do this and certainly a 10 year-old can perfect it, whereas he's 15 in this video and he can't do it for very long, but here he is practicing in the pool, beautiful. Watch him flip turn though.

You see how he deviated off the center for just a small fraction of a second? It's because he uses his visual cues in order to track where he is in the pool, and on that flip turn, he temporarily becomes lost in space because he does not have a functioning vestibular system. So what you can see is, again, this dichotomy, if you just think, well, my son's a competitive swimmer, how can he have balance dysfunction? Well, you don't need great balance function to be a competitive swimmer. We have an inordinate, a number of them in our implant population and there's good reason for it. But I look at this as a clinician and I say, well, that's pretty good. Look at this boy who nearly died of meningitis, has bilateral implants and here he is competing for Canada in the Deaf Olympics, that's amazing. It's a success story in my mind, but it's not my perspective that matters, what matters is what he wants and he says to me, "Dr. Cushing, you saw when I did that flip turn and I deviated off of midline, that cost me the milliseconds that I need in order to not only swim in the Deaf Olympics, but to swim in the Olympics and I want to swim in the Olympics. Is there anything you can do about my balance function?" And so it really does show us that it's what these children want and need that matters, not our estimation that they're doing pretty good, and I think we see this both on the auditory side of things and on the vestibular side of things. Now, we'll go onto the next video and this is another, you know, our best stories come from our patients and our families, so this is another such story that taught us a lesson. So this is our young boy with Usher's and here he is at our annual cochlear implant skating party and here he is up on skates. And so we were capturing this video because we thought, look at this, amazing, this child has no functioning vestibular system yet with a lot of practice and a lot of lessons he's up on skates, what a beautiful Canadian success story, and then that happens. And I can imagine every single one of you that's sitting in the audience today cringed and jumped and I've seen this video hundreds of times and every time this child's chin hits the floor or hits the ice I cringe because he took that one on the chin. He did not have a fall response, his arms did not go down to protect him whatsoever.

And we're gonna go back to the presentation and tell you about another aha moment that we had in the moment of capturing that video. And so what we learnt from this is we said to ourselves when he fell on the ice like that so clearly without a fall response, this child had two cochlear implant failures. I wonder if this is why 'cause this is how he navigates the world, as we looked at this example of him on the ice ring. And so what it led us to do was to go back and look at our cochlear implant failure data and when we talk about failure, what we're really gonna talk about is not failure of the device to give linguistic benefit, but rather failure of the device, devices that didn't work, primarily hard failure but at times soft failure as well, so that's what I mean by failure. And we'd looked at our relatively low failure rate and actually published a paper on it and one of the things that we noted was that children who had meningitis had an inordinate number of failures. And at the time, we didn't really make anything of it despite the fact that we were studying vestibular function in these kids but we missed the boat. So we thought, oh, maybe it's inflammation, maybe it's ossification but we missed the fact that what was putting them at risk, just like this boy in the video, was the fact that they universally had vestibular impairment and so we went back and looked at our data with new eyes. And so the first thing that we did is we looked at our group of children who had never had a reimplant and we compare them to standardize norms, and these again is using the test of dynamic balance that I showed you previously, and then we compared these children to children who had undergone reimplantation. And so what you can see is that the red bar demonstrates that these kids had significantly poor balance function than children who had never needed a reimplantation and our standardized norms. Now, many of you in the audience might be saying, well, I'm sure you're a delicate surgeon, however, you've been in that ear a couple of times with an implant and what it will say is that a lot of this data was acquired prior to the children having implants and certainly prior to having bilateral implants. And so we also wanted to look at the end organ function testing to see if it followed the same pattern in our kids who had failure.

And this is what we saw, and I'm gonna present to you only the data for caloric testing of the horizontal canal, but the same held true for rotary chair testing as well as VEMP testing, so a measure of otolithic function. And what we saw was that the proportion of children who had abnormal results, so in this case, those kids who had bilateral areflexia in the reimplanted group were significantly larger than those in the non reimplanted group. And again, the same holds true here. Many of these kids were tested without an implant in the ear and many of them also had etiologies of deafness that were associated with vestibular impairments, so meningitis, Usher's, cochleovestibular anomalies. And so when we re-worked this data, what we found was that vestibular function increase the odds of cochlear implant failure by almost nine times. And I think that really spoke to us, and it certainly speaks to families when we see children with cochlear implants. And so in many ways, if we think about that question that we were asking at the beginning, how do implants hurt the vestibular function? Really this answer that we got is flipping it on its head, vestibular dysfunction puts the implant at risk, so again, an answer to a question that we didn't know that we were asking. So, who is at risk? And so many of us look after a large caseload of children with hearing loss and a large caseload of kids with cochlear implants and it's not possible for us to endeavor to look at all of their function. And so if we look at the prevalence of risk relative to etiology when we do know etiology, this is what we see. So the kids who have abnormalities of the anatomy, so the kids who are in this red box, those with cochleovestibular anomalies, and I've listed a few of them here. Children who've had ototoxic medications as the underlying reason for their acquired hearing loss. Those with Usher's syndrome and universally, those who've had bacterial meningitis, they are all at very high risk of vestibular impairment and so those children definitely need additional evaluation. The children who are in the yellow box are in the moderate category and those include children who have congenital CMV, as well as those kids who have auditory neuropathy spectrum disease. Now, what I'll tell you from experience is that these kids have terrible balance, terrible, terrible balance, terrible knowledge of personal space, and they're awkward essentially, and those of you who look after these kids are probably nodding your heads right now. We put them in the yellow box as opposed to the red box because they have some abnormalities of their end organs, but they're not bilateral at times and they're not as profound. So their dysfunction is a little bit out of proportion with their end organ loss and that's likely because these kids are double whammy kids, where there's a central portion of it, of their dysfunction, as well as a peripheral portion.

And finally, we've got the kids with connexin who generally have pretty intact vestibular function, and I've put mitochondrial and autoimmune kids here and I've put a question mark I think for good reason, because while these kids in our cohort tested normally, there's very small numbers. And certainly from a pathophysiology perspective, we wouldn't be surprised if these children develop dysfunction over time, so that's why they're there with that little caveat. And so this is one way to try and hone in on the group of kids that really are at highest risk of dysfunction. So let's look at a couple of special categories that we've been interested in a little bit more in the last several years. So ototoxicity and congenital CMV. So we do a really great job here at SickKids at screening and following our kids who are receiving ototoxic medical therapies for hearing loss. And so there's a wonderful protocol, the catch is very good and we're really able to follow their hearing function over time, and at times intervene as necessary. We know in the literature that the prevalence of cochleotoxicity from some of these treatments, and I've put a few examples at the bottom of the slide here, and we know that they're high, 25 to 90%. The prevalence of dysfunction is very, very high in these groups but we really don't have a sense of what the prevalence of vestibular impairment is in these groups, and so this is something that with our oncologists we wanted to look at with a little bit more depth. So what we did is we did some screening assessments of end organ function and we used vHIT, and we'll talk about that a little bit more, and dynamic visual acuity as a test of horizontal canal dysfunction. And what we saw was that again, roughly 50% of kids in this category had significant impairment. When we looked at utricular dysfunction and we used a tool called the subjective visual vertical, and we'll talk about that tool specifically in my lecture on October 28th, roughly 60% of kids demonstrated dysfunction. So a significant amount of dysfunction in the vestibular end organs in this group of kids with ototoxicity. Now, what about their balance? So we also did one foot standing as a screening assessment of balance and here are our results, and they've just been input sort of worst result to best result. And this is our cutoff and we'll talk a little bit in a few slides as to why that's the case.

And so what we see is that 70% of kids are unable to stand on one foot for more than eight seconds and so that denotes impairment or an abnormality on that test. If we make it a little bit harder and we have them close their eyes, then we see again that almost 70% of kids are unable to do so for four seconds, which is again our cutoff for abnormal and I'll explain more about why that's the case. And so a good proportion of these children have both balance and vestibular impairment. Now, you'll note that the prevalence of balance impairment is larger than the prevalence of end organ impairment and there's very good reasons for that. So many of these children have had CNS tumors, and again, it's a double whammy for balance impairment. And so now what we're able to say to our oncology patients in particular is that the prevalence in this small cohort of vestibulotoxicity is roughly 50%. It's not always correlated with cochleotoxicity, I'll have you know. And 70% of these kids have balance impairment.

And I think that this really does speak to functionally one of the challenges that these kids have in the longterm, and now that these children are surviving longer and more of them are surviving, it's really important that we at these quality of life issues that are gonna impact their longterm quality of life. So that's ototoxicity and it's something that we are continuing to push from a screening assessment at our institution. Now, the next group of children is a very interesting cohort for lots of reasons and those are the kids who have hearing loss from congenital CMV. And what I've put up here is a scanning electromicrogram of a mouse cochlea. On the left hand side of the screen, the mice cochlea is normal. And on the right hand side of the screen, this is a mouse that has been infected with a model of congenital CMV. And so what you can see is that the vasculature has really been impaired in the mouse that was infected with CMV. And so we know that congenital CMV has significant impact on the inner ear. And what we're used to hearing about in our implant populations is the 20 to 50% of kids in this cohort with congenital CMV who will develop sensorineural hearing loss and we all have these kids in our cohort, but what we wanted to know is, again, what's the prevalence of vestibular impairment in these children?

And there's good reason to think so, so again, this is some data out of Bob Harrison's lab who works here in our department, and what we can see is that there are deposits of the virus in the vestibular portion of the inner ear. And so when we look in the literature to see what is currently reported in terms of the prevalence of vestibular impairment, what we see is that in some cohorts, the prevalence of dysfunction is up to 92%, with many of them, 1/3 having complete bilateral loss. Those are the kids who are gonna have a plateau of balance skills at 4 1/2 years of age. And 50% of them are gonna have progressive loss over time, so very similar to what we see with hearing loss. Now, 50% of the kids will also have dysfunction in the otolithic organs, and so what we can see is that certainly the risk to the vestibular end organs is as high, if not higher, than the risk of sensorineural hearing loss in these children. And certainly what some of these studies have demonstrated is that at times vestibular impairment incurs in the absence of hearing loss in these children, so it's really something that we need to look out for. Now, congenital CMV is such an interesting story and again, we'll talk a little bit more about this in the followup lecture on October 28th, but we're gonna shift gears a little bit so I'll ask you to come with me as I slowly shift gears into another population and that's our kids with single-sided deafness. And one of the things that we learned as we went along and all of these research projects are happening in parallel and so they really do cross pollinate, and what we learn from one informs what we study in another. And so all at the same time we've been studying congenital CMV, we've been looking at children with single-sided deafness, we've also been looking at vestibular and balance impairment, and they're gonna come together beautifully and I'm gonna show you how. So congenital CMV, as we looked at our kids and cohort of children that we were implanting with single-sided deafness, were found to be the most common etiology of implantable single-sided deafness. And so when we look, this is our cohort of kids with SSD who we've assessed for cochlear implantation, and 20% of the children that we see with SSD, the etiology is congenital CMV. When we weed out all the kids who can't get an implant, either because their duration of deafness is too long or they don't have an auditory nerve, for example, and we only look at those children who by our research criteria would be able to get an implant and we look at how many of those have congenital CMV, it's almost 1/2, 43%, and ultimately 40% of the kids who go on to get an implant have congenital CMV.

Okay, so bear with me, it's all gonna come together. And so the other thing that we looked at was the acceptability of implantation in children with single-sided deafness. And what we saw was that even though only 30% of parents who had a child who could be implanted for single-sided deafness, only about 30% of them said yes, the rest of them said, "Oh, no thank you, not my child. You can experiment on someone else's kid." But when we look at the cohort of kids with CMV, what we saw was that 80% of those families said, "Sign my child up for your experimental protocol," and the reason is because those kids are at risk of progressive hearing loss on the other side. And so we were shining a light on these kids who we know have progressive loss, who we know are at risk of poor balance function, who we know are at risk of vestibular impairment. And so we began to think about these kids with SSD and their deficits and really about who should we be implanting, who shouldn't we be implanting? Who is most likely to gain the biggest benefit? And I think when you think about deficits in children with SSD, you always go back to, there are some really amazing seminal work by Dr. Tharpe early on much before we were thinking about implantation and more recently, Dr. Lou, and what we see here is that when Dr. Lou talks about the impact of unilateral hearing loss on the cohort that she looked at, what she demonstrated was that the impact of this was far reaching. Look at this, speech and language delays, quality of life, grade failure, behavioral issues, school performance, need for IEP and speech therapy. So these are not just language based deficits and I think when we think about these kids with SSD, in particular those with CMV, I wonder if we're giving too much credit to the cochlea for these deficits. And so I'm gonna throw out an idea and we'll explore this further in the second lecture is that perhaps what we really need to be also thinking about is, what are these children's balance function and vestibular function? And so that's what we started to do.

This is one of the fancier ways that we measure balance function. We are very fortunate to have this virtual environment that can move and provide us with large force plate data and here's what we saw. This is our group of kids with single-sided deafness and their balance skills. Again, we used the BOT-2 but we used it in this environment. And what we saw was that these are our kids with normal hearing and these are our kids with unilateral hearing loss. And to put it into context of the data that I showed you, while these are statistically significant, this is where all comers with hearing loss, that's the top black bar, and our kids with bilateral vestibular loss are, that's the second black bar. And what we can see is that children with unilateral hearing loss are about twice as good as those with bilateral loss and about as good as all comers with bilateral hearing loss. And they are certainly significantly poorer than normal hearing controls. So perhaps we need to think about this more in our evaluations. Now, is their balance impairment because they lack spatial awareness, because of their unilateral hearing loss, or is it because of end organ dysfunction? The answer is that it's probably a little bit of both, but when we looked at their end organ function, what we saw was that again, 50% had a deficit in horizontal canal dysfunction and almost 60% had otolithic dysfunction. Many of these were on the same side as the hearing loss deficit, whereas others had bilateral deficits despite the fact that they had hearing loss deficits. And so I think now as we go more forward with our group of kids with SSD, we're looking at them more and more in terms of how they compensate for these balance deficits. So these are the tasks of the BOT and they get increasingly difficult over time. So when we look at the really easy tasks, we see that these kids with hearing loss on one side do pretty much the same as our normal hearing kids, but what happens when we make it those are the easy tasks and the mid range tasks? What happens to them when we make it hard, when we get them to close their eyes, when we put them on a balance beam? That again is where you start to see things deteriorate.

So if I told you at the beginning that you have to push kids with bilateral hearing loss to see the deficits, you certainly have to push kids with unilateral hearing loss into difficult tasks in order to uncover their deficits. And again, when we look at how they compensate, there is not surprisingly an over-reliance on vision in these kids compared to the normal hearing controls. And this is the data that I've shown you already. And so I think the take home message is that in these kids with SSD, particularly when we're considering intervention, we need to look at the whole being. We need to look at their deficits in the light that this is potentially labyrinthine loss, not just hearing loss, and I think that that may prove to be very useful in deciding which child with SSD will benefit more from intervention. And I hazard to say that the child who has cochleo and vestibular loss may benefit more, for example, from an implant than a child with cochlear loss alone. So I think we really do need to keep an eye on this space. And I think to really hit home the importance and again, this is a segue into what the majority of the lecture on the 28th is gonna be about, is really seeing how central vestibular function is to learning in children and to development in children. And what we see here in the pyramid of learning is that vestibular function sits at the fundamental base of this triangle. It's actually more important than other peripheral senses like vision or auditory function, and so I think it really speaks to our need to look at this in these children.

So, how are we gonna measure this? And this is the final portion of the lecture, is to give some practical skills so that tomorrow you can go into your clinic and help to identify these children with impairment. And so what I'm gonna design here is, or demonstrate to you is really three easy steps for screening in vestibular function. And so these were designed to be used quickly in clinics where you don't have the ability to do all the fancy stuff. So let's go through them. The first is motor milestones. And so this is a screening assessment that we as physicians do as well as our audiologists do. So any child who does not have head control by four months, who is not sitting by nine months, or who is not walking by 18 months is at high risk of having vestibular impairment. And what I've come to learn over time is that parents don't know that there's two parts to the inner ear. They don't know that balance and hearing can go hand in hand, and so if you don't actively ask these questions, families are not necessarily gonna tell you that their child didn't walk until he was 22 months old. The other thing that we've learned in asking these questions is that when they're normal, most families don't remember, especially if there's many kids, they'll just, you know, they'll scratch their head a little bit and they'll say, "You know what? I'm not sure, I'm pretty sure it was normal." But when it's been abnormal, when you were the one mother who was sitting at the playgroup whose child was still not walking, that is forever gonna be burned in your brain and the minute you ask these questions, the flood gates open. And so remember, parents don't know that both parts of the inner ear are related.

The next thing we want to do is an assessment of motor function. And so the one that we tend to do most frequently is one foot standing because you can do it in the audio booth, you can do it in the clinic, you don't need any special equipment. And so what I've put here is essentially the normative data for children, so how long can they stand on one foot or should be able to stand on one foot? And what you can see is that a 2 1/2 year old should be able to do so very briefly, whereas a four year old can do it for five seconds and a five year old can do it for 10 seconds. And we wanted to know just how good this test was and so what we did is we took all the data that we had, we had a huge database of kids who had done all of the items on our balance tasks as well had had vestibular function testing. And what we saw was that one leg standing, eyes open and eyes closed was really a fabulous test. It's not specific for vestibular impairment so a child with a neurologic disorder is also gonna test poorly, but again, this is screening, this is a quick method in order to decide, okay, this kid, I need to send them to the person who does that testing or I need to have them back to clinic.

So, any child who's over five years of age who cannot stand on one leg for more than eight seconds should be tested for peripheral vestibular dysfunction. Also, if the child is unable to stand on one foot with their eyes closed for more than, is not able to for more than four seconds, then again, that's a child who should be flagged to have additional testing done. Now, the final part is about linking the deficit that we're seeing in the motor milestones and in the motor function to the peripheral vestibular system, so it's some assessment of horizontal canal. Now, you don't have to do all of these three steps. Frankly, many days in clinics, my partners don't get past one and so don't feel like you have to commit to all of them. Sometimes you'll be able to, sometimes you won't but they're individually useful steps. And so number three is assessment of horizontal canal function. And so there are a number of ways to do this and we'll start and look at a video together of the simplest way. So this is the standard clinical head thrust testing. So this is an 18 month old boy and he's sitting on Dad's lap. You have to have some light-up fancy toys and what we're doing, you can see he doesn't really mind me turning his head from side to side. The target is a little bit close so the purist will say, well, you might run into trouble with accommodation but sometimes testing in children is imperfect, but watch as we go left, he has a big saccade back. And so this in that moment in clinic links this child's motor dysfunction and delay in milestones to horizontal canal dysfunction and so we're able to say he has a peripheral vestibular deficit. Now, there are fancier ways to do that and in the next video, we're gonna show you a couple of those fancier ways. So this is video head impulse testing and many of you will be familiar, but it's a test of high-frequency function of the horizontal canal. You can also use it to look at the vertical canals. Here's an example of a young boy who's in our ototoxicity population. So we've got his Captain America goggles on that are shooting out laser beams and we're calibrating the device. Children even if you ask them not to and probably most adults, will move their head even if you tell them to just move their eyes. So we're holding onto their head and in an ideal world, a two tester model goes a long way in getting good data. Now, I didn't edit this out because this is the reality of pediatric testing. So we've just beautifully calibrated that device and then he went and took it off so we had to recalibrate. Now, here we are doing head thrust head impulses and we're measuring the gain of his eye movements over the gain of his head movements.

Now, you're gonna see me moving my hands around a lot and that's because every time I turn this child's head, he says ouch and again, I didn't have the benefit of a second tester. If you look at mom's face, you know that she knows he's joking with me but I don't know that. And so again, it speaks to the challenges at times of pediatric testing, but this provides us with an objective measure of horizontal canal function. And so if there's a way to incorporate a single objective test in your otolaryngology or audiology clinic or hearing loss program, this would be the test that I would add in terms of equipment. And so there are different types of ways that you can test this, this one example. We're gonna go to the next video which shows a different example that we use in the clinic for children because not all children will wear goggles and so we also have a goggle free system, cochlear implants are at a very inconvenient place for head thrust testing. So there is a remote camera for this young boy out here that he's staring at, along with a target that we're trying to entice him with and we're doing some head thrusts and this is a young boy who has fallen a few times and managed to knock out the magnet in his cochlear implant once on each side. So I was waiting to take him for surgery and based on our previous data, we said, okay, let's bring this kid upstairs and test his function and indeed he did have bilateral vestibular impairment. Now, I'm just gonna set up the next video that we're gonna watch so we can go to that, and we can also take advantage of a child's physiology. So under six months of age, children are unable to suppress their vestibular ocular reflex. And so one of the things that we can do in really young babies and what's gonna be demonstrated in this video is the mom powered rotary chair. So you can have the parent spin, and this is my son when he was three months of age, and so we're spinning him around and when we stop, is that not the most beautiful postrotary nystagmus you have ever seen in your life? And so then demonstrates that this young man has intact vestibular function bilaterally, there's not a quantitative component to this, but qualitatively, we know the function is intact. And what you would see in a child who is under six months of age who has bilateral vestibular impairment is you would not see that postrotary nystagmus. So this is a great test in our kids who are coming in with an early diagnosis of hearing loss. So let's sum up now and go back to the presentation and we'll just review what we've learned. I think the most practical part of this presentation really relates to the skill set that we've just talked about and incorporating any one of these three easy steps into your clinical examine.

As I said, our audiologists have become amazing at really picking these kids up and sending them to me. And so if we think about and reflect on what we've learned today, the first take home is that vestibular and balance dysfunction is exceedingly common in sensorineural hearing loss. And the minute you start thinking about it and asking about it, you'll really appreciate that it matters. It matters to these kids, it matters to their parents who have not really understood why their child is different, why they can't ride a bike. And so even though we can't at times make it better, although I hope in my career that we will be able to, just even having a reason does a great deal of help for these children. And the final thing is that it doesn't have to be complicated, you don't have to have the fancy tools I showed you in order to at least screen. So you can identify these kids in the audio booth, in the clinic with relative ease. And so this is where we're gonna stop for today, but it really is the beginning of a bigger conversation about how we holistically look at and look after children with hearing loss, especially as we begin to intervene on the periphery with single-sided implants, for example, and perhaps knowing what vestibular function is will help us lead the way way. And I'm hoping that some of you may join us for that continuation of the conversation as we look at how we can use cochlear implants not only to restore audition and capacity to develop oral language, but really to help these children restore their connections with the world around them in a much broader sense. So we'll stop there and I see there was a few questions that came up throughout the talk. I'm sorry, I'm not coordinated enough to flip back and forth, but perhaps we can go through them now. I think we still have time to do that. So the first one, I'll just start at the top. Okay, the first one was from Doug and what he was asking was that,

- Yes, absolutely. "For the 35 to 40% of children

- Yes, go ahead. "with complete areflexia, "what is the degree of hearing loss in that group?" So it's a great question, Doug, because there is a correlation between severity of loss and probability of dysfunction. And so while you can have everything in between, you're more likely to have vestibular impairment if you have poor hearing function. And for the majority of those kids, they had severe to profound loss that put them in or near the implant range. And so I think as you get to milder forms of hearing loss, the prevalence or the severity of the loss is going to be less, but I think sometimes it is idiosyncratic. So it's still worth at least doing the screening test and deciding very quickly who needs additional attention. So next we're gonna go on to a question from Lauren who was asking if we found in children with SSD who had bilateral vestibular function ultimately have had a higher likelihood of having progressive loss in the normal ear later on. Oh, it's a fantastic question, Lauren, and I hope that over the next decade, I can answer it better than I can today. Our cohort is still pretty small. I think that the group of kids you're describing here are gonna be the kids with CMV who have a high risk of vestibular impairment and also have a high risk of progression, but right now we just don't have enough numbers to predict. But I think these are also a group of kids who have the most functional impact of their sensory deficits and probably who can benefit from intervention more, but stay tuned and hopefully I'll have a better answer for you. Now, Jeanine asked a question about CMV and wondering whether or not it can create vestibular dysfunction with normal hearing. Oh, "Since it can, do we suggest changing infant hearing screening protocols to follow for vestibular testing even if hearing is normal with positive CMV?" Oh, that'd be my dream Janine. So I think the first thing we need to do better is identify congenital CMV, and thankfully where I live in Ontario, over the last year, we've gone to a universal screening program for CMV because so many of these kids, probably more than 1/3 have normal hearing at birth, so they get missed if we only think about hearing loss. There are places in the world where they're screening for vestibular function at birth as well. So in Belgium, there's one jurisdiction that does that and I think that absolutely, but if I had only a few dollars and I could only put it in one place, I'd start off with universal screening for CMV, but absolutely, they are at risk. Now, oh Victoria, thank you Victoria for such nice comments.

Let's see. Leah, "Are there similar results of vestibular disorders in kids with chronic otitis media?" It's a very good question and chronic otitis media really was the model for looking at vestibular disorders in these kids and there's lots of data. I think the disorders are much milder in these kids and they're also temporary much like hearing losses as a result of otitis media. There's a few good studies that have looked at the longterm impact of impairment of vestibular and balance function with OME, like do the deficits continue on long after the OME has gone and the ultimate answer is no, but I think it's really important, even in the updated set of criteria for putting tympanostomy tubes, and one of the things that changed over the last 10 years is the fact that balance disorders and vestibular disorders are now an indication for tubes. And I think the population I see clinically that benefits the most are not necessarily the typically developing child, but rather the child who has other neurologic disorders. And so those are the ones that I think really do benefit from management of their otitis media from a balance and motor development perspective. Okay, I'm gonna keep going answering questions until someone tells me to stop if that's okay. So now we have a question from Aishwarya and she's asking, "Is it possible to develop vestibular dysfunction post CI with no cause or known etiology of hearing loss?" And so absolutely, an implant itself can take out vestibular function, and I think we probably did that more so in decades gone past and a lot of the data that's in the literature were old techniques, larger electrodes, less soft surgery. So I think the things we do for hearing preservation are also good for vestibular preservation, but it is a risk. I love the vestibular system but I'm also still happy to go ahead and do simultaneous bilateral implants because it's all about developmental impact. I think it's a very different question if you're looking after adults, I don't think the same thing applies but I think it's important to know what you're dealing with. So Doug asks, "In regards to your three easy steps, would you recommend that a child with any degree of hearing loss and who is identified by one of those screening questions should be referred for vestibular testing?" In an ideal world, absolutely, but I appreciate, you know especially nowadays we don't live in an ideal world so certainly severity of hearing loss or specific etiologies can help you triage but in an ideal world, absolutely. And what they may be able to access maybe different, for some it might be physical therapy assessment, for others it might be end organ testing. Okay, and I think that's it. Yes.

- [Moderator] I think that covers all the questions. Thank you so much, Dr. Cushing for an amazing course and for your time and expertise and thank you for everybody for the great questions who participated today. We will look forward to your feedback on the course evaluations and hope to see you in Dr. Cushing's future course--

- Thanks so much. Take care, stay well.

- [Moderator] Thanks so much, everybody, have a great day.