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Restoring Connection with Cochlear Implants, presented in partnership with the American Auditory Society
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- [Elizabeth] It is my pleasure to introduce today's presenter. Dr. Sharon Cushing is a pediatric otolaryngologist at The Hospital for Sick Children in Toronto Canada, and an associate professor and clinical investigator in the Department of Otolaryngology Head and Neck Surgery and the director of the Cochlear Implant Program at the University of Toronto. With no further ado, I'll turn it over to you Dr. Cushing.

- [Sharon] Thanks very much Elizabeth. It's great to be here again today and I hope you're all comfy wherever you are working from home or working in your offices and have something good to eat and I'm really looking forward to taking you through some of the work that we've been accumulating over the years here in Toronto. Now about a month or so ago we had another session with Audiology Online you know, where we talked about the importance of vestibular assessment in our kids with hearing loss and this session while it wasn't planned to do so actually builds on that concept. And so I'm mostly gonna focus on the importance or why it matters to our kids with hearing loss. Now you'll notice that this title slide is a little bit different than what you signed up for, but I promise you're gonna get what you signed up for but it just, you know, I think we've all had quite a year with the pandemic and I had the very good fortune of being able to present at the American Auditory Society and it's the last conference I went to. And I wonder how long it's gonna be before we have that luxury again. And it reminds me, this was the title of the talk that I gave there and the intention was to replicate or build on that specific topic. And even though the data has not necessarily changed between now and then, although we've been busily working hard with respect to research as much as we can in the interval, our perspective has. And so I cannot look at the data the same way as I did before and that's where the new title comes from, is really so much about what's gone on in the last while has been about connection and I think we all feel very pained for the lack of connections that we've been able to have and to make a new in the setting of the pandemic. And so I think that this talk is now got new perspective since I gave it at the American Auditory Society and I'll try and communicate that to you over the course of this talk. So these are my disclosures and some learning outcomes. Now I'm gonna start off by acknowledging the amazing team

continued

we have here at The Hospital for Sick Children. And so we used to have different pictures but those have even changed in this setting 'cause this is what our research team meeting looks like these days and so this is our heart at work research team trying to do whatever they can amidst the current restrictions up here in Canada with respect to the pandemic. And again, a lot of what we've been doing is sort of to look at old data with new light the stuff that's been sitting on the shelf and gathering a bit of dust waiting for us to have the time which has been a joy in and of itself. We also have an amazing cochlear implant research team and here's a pic just leading up to this talk, we were in our cochlear implant weekly implant team meeting which again has gone mostly virtual and again, this is what we've all gotten used to over the last many, many months, but this is a group of hardworking individuals who care immensely about the population of children and the families that we look after.

Now, many of you might've seen this picture and it first came out when they were going back to schools and daycare in France. And I had an emotional reaction to this picture and I'm sure many of you, if you had seen it or if you're seeing it for the first time do as well because this is not what childhood is supposed to be like, this is not what a playground is meant to be like. And when we see these boxes drawn around the individual children and essentially eliminating their capacity to interact fully with their environment and more importantly with their peers, you know, what I saw here was lines that were drawn around their development. No much in the way that hearing loss or other sensory deprivation like vestibular dysfunction might also draw lines around the capacity to develop. And I began to think about this and it made me very sad and this viewing this picture was then followed by a conversation I had in the operating room with one of our OR nurses who you know, had to drop her child off to a daycare much like this in order to come to work, in order to be available to put a cochlear implant in the child that we at that moment had on the table. And in that moment that, you know, witnessing of that individual's experience changed my perspective on this photo because if I told you that this little girl here in the jean jacket, well, you know that was the daughter of that OR nurse that came that day. And over here, the little girl in the yellow, well, her mom's an audiologist and she came in that day so that she could turn on devices that we implanted prior to the pandemic. Then all of a sudden how we feel about this picture and this world changes. And I thought what a remarkably powerful thing perspective is that the world hasn't changed, the picture hasn't changed, but our perspective on it has. And I think that that's again what a lot of us are experiencing during this pandemic at times for good and at times for bad. Now we all exist to interact with our environment. That's what we do so beautifully as humans, it's the reason we have senses.

And I love this photograph of this individual who is in one of the ravine systems in Toronto and the interposition of sort of the urban and jungle that's put together and you can just, you know, perhaps even imagine what's going through her mind as she's breathing in the somewhat polluted air in Toronto and looking up at the trees and hearing the sounds of the highway and the subway train above her. And what a beautiful embodiment of what our senses give us that this picture is. But what's missing from this picture is the individual witnessing, the person taking the picture. And that is just as important as the individual in there. And I think that a lot of us have felt lonely over the last while or unable to connect in a way that is meaningful for us. And if I think about some of the themes that come up for our kids, with hearing loss, and in particular we run a teen group along with our social worker, a lot of the themes that come up is this challenge in connecting. This move for our kids with hearing loss towards a more electronic form of communication, because the fidelity is better for them. There's more flexibility in terms of responses and how many of them do lack those meaningful connections with peers in particular and so enjoy coming together at the teen group because they find that connection and individuals who have the same challenges as them. And so when we think about what these children are going through, we might be experiencing a little bit of that during this time in the pandemic. And this is a slide that I've borrowed from Blake Papsin, one of my surgical partners and he often will say that, you know, as humans we're predators of language. And so we will use all of our senses and our senses have been honed in order to be able to communicate with language. And I think that this pandemic has made this all the more apparent and for me, you know, the perspective of the pandemic makes me take it one step further because language is only one of the building blocks. So, you know, we're connecting in this moment through language, but it's not the same as being in a room together where I can look in your eyes, I can, you know, you can see enthusiasm on my face and feel it in a way that's just not possible to do virtually. And so here's how I would take this one step further by saying that humans are predators of connection. And that can be through language but it can be through all other different modalities and so humans need to connect.

And so, unfortunately during a pandemic, this is what connecting looks like. We have Zoom happy hour and Zoom social events and we meet up with our friends and relatives separated by a screen and it really can have an impact. I think, you know, many of us are, you know, there's advantages to it, but there's also significant disadvantages. You're not able to use your senses fully. The joy of interacting individually has been compressed down to this two-dimensional world. And so if we think about it and go back to the physiology, which is what the, you know, the individuals who make the up American Auditory Society do so well is let's think about what is the underlying physiology of connection. And I think a good example of this is mirror neurons. And I'm not an expert in this, but I'm gonna apply this topic for the use of this talk. And so essentially what these neurons do is, you know here's our monkey sitting here and the mirror neuron is in a resting state. He grasps for a ball and that excites the ball grasping mirror neuron, but then he watches somebody else grasp the ball and we get the same excitation pattern. There's some people who use the example of I yawn, you yawn mirror neurons may be responsible for that reflexive interaction. But what I wanna know in this time of the pandemic is what happens to that mirror neuron if we put a screen and a Zoom meeting in between the monkey and the grasper, do we still get excitation? Do we or does it have to be physically present with the individual? And so what are we losing? What are we missing out when we impair all of our senses capacity to fully interact, and whether the impediment to that is an inability to be together in the same room because of a pandemic or an inability to understand and connect fully as a result of a hearing loss? How are those two things similar? And so that is really where we come to the new title of this talk, which again is the same data I presented before where we're talking about restoring those connections with cochlear implants. What is it that we're missing for these kids who come to our teen clinic and say, you know what I struggle to connect with my peers. I have a few good friends, but what I'm really searching for is that village and, and that personal connection.

So how we're gonna think about this is gonna be in the context of cochlear implants. And, you know, since the very first cochlear implant and I think we as clinicians and as scientists have really been struggling to catch up, and essentially this is a technology that is far surpassed what we ever thought it could do and in addition to that, it has given us a platform for looking at, in particular in children development in the presence of an auditory deprivation that then gets restored. And this has been an amazing scientific platform to help us learn what happens when we restore it and what happens when we don't restore it. And so let's move into that concept a little bit more. So there's tons of fascinating literature coming out with respect to what happens to the brain when we lose our senses. So what happens to our elderly population and cognitive decline when these individuals have hearing loss, when they have vestibular impairment and how excitingly can we slow that progress when we rehabilitate these individuals. And for me I'll probably never see another adult in my clinical practice and so while I find it fascinating to look at the impact of sensory loss on cognitive decline, what we work in every day is looking at how we build a brain.

So I look at the other end of the clinical spectrum and cognitive incline in terms of what happens to a brain and how does children build a brain in the presence of a sensory depravation that is partially restored for example with cochlear implants. And so that again is the framework that we're gonna use throughout this talk. And so here's an example or another type of framework that looks at building a brain. And what we can see here is that audition is one of the fundamental building blocks of cognition and intellect. It is super, super important, and we know bad things happen when we don't have access to sound in terms of cognition and ultimately development. And thankfully, because of cochlear implants and hearing aids, we have the ability to restore that connection when it wasn't present either at birth or was acquired through a loss. And so we know how a brain gets built in the presence of normal audition. And we develop with something called aural preference and simply put that means that the right ear speaks to the left cortex and the left ear speaks to the right cortex. And here's a beautiful example of 64 lead EEG showing that relationship between cortex and periphery. And we're gonna come back to this technique repeatedly 'cause it's one of the tools that we use in our lab, but we know what happens when hearing is normal on both sides. And so in keeping with this, we also know that the brain doesn't like sensory deprivation and here's a beautiful example of what happens to the brain in the presence of sensory deprivation from Lee in the early 2000s.

And so these blue parts of the brain are essentially the brain doing nothing. And what we can see is as the duration of deafness increases there is less of the brain doing nothing. And the simple reason for that is essentially the brain does not like to do nothing and so as time and duration of deprivation has increased, the brain that is responsible for receiving those incoming auditory signals says, you know what nothing's coming, I'm gonna pack it up and I'm gonna go do something more interesting 'cause I'm tired of waiting. And so essentially those areas of the cortex then get lent to other sensory modalities, vision for example, perhaps even proprioception and the part of the brain that was meant to be responsive to hearing is just no longer there in a functional sense. And so when we look at aural preference in the context of cochlear implantation, this is what we see and I'll take you through this slide. So here again is our representation of normal aural preference and so we've got right ear speaking to left cortex and left ear speaking to right cortex. And that's what a normal pattern looks like. Now we can create a normal pattern which is pretty remarkable with simultaneous cochlear implantation. So again, we see a pattern of aural preference that is nearly identical to normal hearing. And what if we don't get them in at the same time, what happens if there's a period of asymmetry between those ears? Well, as long as it's short enough, and in this study the definition of short enough was less than a year and a half of significant asymmetry between the ears, we were able again, to restore normal aural preference. Now these are children who are bilaterally profoundly deaf and the final group I'm gonna talk about is a group of kids who came at a time where we were not performing bilateral implants. And so they got one implant and then many, many years later at least more than two, but for some as long as 10 or 15 years later, they got their second implant. And you can see that all of the preference goes to the left cortex.

Now that's because all these children, their initial implant was on the right 'cause we thought that that was better and when we only put one in, we put it on the right. But what you can see is that even when we implant the contralateral left ear, the aural preference still goes to the left cortex. We are no longer able to restore the brain as it was meant to be built in the presence of bilateral hearing. And so there are limits to the system as flexible and as plastic as it is and it informs our decisions as to who may benefit from cochlear implantation. Now we haven't always done bilateral implants. I just showed you some data that was prior to the times when we were thinking about doing simultaneous implants and I've just told you that it's a good thing. It's the best way in a bilaterally deaf child to restore the brain and its capacity to be built in a typical fashion. And there were lots of things that got in the way of us initially doing bilateral implants until really the early 2000s. And one of the biggest concerns and questions as clinicians and surgeons was well there's not just the hearing in the inner ear, there's the vestibular and balance apparatus. And so what would we potentially be doing to that system if we put in bilateral implants and that's for us where the story about vestibular function and the importance of measuring it came in. And so we really wanted to know in the early 2000s as to what was that risk and was it reasonable to consider that risk in the context of giving these children bilateral audition? And like many people at that stage, we were very worried about the negative impacts of implantation on the vestibular system. And certainly we know that there is a risk to the vestibular portion of the inner ear by putting in a cochlear implant, no matter how gentle we are. And there's been documented histologic evidence of injury, fibrosis in the vestibular, distortion of the saccular membrane and there's also been functional loss in terms of for example, loss of vestibular evoked myogenic potentials which are a measure of otolithic function that then become absent following cochlear implantation. So it's not to undermine this risk, and it was the question that we were trying to decipher. However, this line of questioning led us to something way more interesting that has continued really in terms of our research over the last 10 to 15 years.

And so one of the first and most important things we found was that vestibular dysfunction is really common. And so if we look at the literature about 70% of children who present in the cochlear implant candidacy range will have a degree of vestibular dysfunction. For some it's partial or it's mild in nature, but for 35 to 40%, of children will have complete areflexia and that is essentially, you can consider that deafness of the vestibular portion of the inner ear. And this tells us that vestibular impairment is the single most common associated feature of sensory neural hearing loss. However, I would hazard to say that only a portion of children with hearing loss and probably a pretty small proportion have screening or evaluation for vestibular impairment. And we really addressed how to do that and what tools are available to us in a screening assessment in the prior talk that I gave, so I'm not gonna focus on that today. Other than to say that it's important when we're thinking about these children and in particular their outcomes with the therapies that we're delivering to them that we think about not only the hearing part of the inner ear, but also the vestibular portion. Now it's important to also look at the behavioral outcome, we've known that in the hearing world for a long time. We can look at an audiogram but we really wanna know behaviorly how does that child do? And the same is true of the vestibular system and so while it's always been important for us to measure the function of the periphery, it's also been important to measure the output and for that we've used tests of balance function. Now, this is an example of such a test called the Bruininks-Oseretsky test of motor proficiency, and I'll call it the BOT2 for the rest of the talk for obvious reasons and it's a pretty simple test. You know, I don't have a physiotherapist that works with me and so it had to be simple enough for me to deliver this test in the clinical context as a surgeon, and this can really be done in 10 minutes. We now have an amazing vestibular audiologist who does this as part of her vestibular and organ evaluation, she monitors this test. And so we're gonna go to a video next which is really gonna highlight the benefit of this test. So if we can have the video please. So what you're gonna see here are, you know, two young boys who have been matched for age, gender, and t-shirt color. And I don't see it moving on my screen, but I'm hoping that it's moving on yours.

Let's see. And so essentially these children are going through, there we go, are going through the items of the BOT and you can see that they have very different balance skills. The little boy on the left, you know, I'm in the frame and he really is unable to do most of these tasks, whereas the little boy on the right has no trouble, he doesn't need my help. Now this little boy on the left has Usher's type 1 syndrome. So he has bilateral profound deafness and wears bilateral implants. He has bilateral vestibular impairment and he progressively lose his vision as a result of retinitis pigmentosa. And so you can imagine that this child's ability to navigate the world is going to become very difficult particularly as time goes on. And look at this boy, look at what it says on his shirt, surf tour. Do you think this young boy is ever gonna be able to get up on a surfboard with balance skills like this? I would hazard to say absolutely not. And so it really emphasizes the impact that these children have when they come in with vestibular dysfunction. So we can go back to the lecture. And so oftentimes, you know, people will say both clinicians that when we travel we'll say, you know, we don't see this in our program and there's certainly is a dichotomy between at times what we see from an end organ perspective and what we see in terms of balance function, but that's because a child who has bilateral vestibular dysfunction walks. In fact they usually run, and so we cannot look at non-ambulation as the marker for vestibular impairment. It's essentially we have to put them in difficult tasks or tasks where we limit their sensory input just like we do in this BOT2 test in order to really have these surface. We also need to talk to the parents because they are intimately aware at what their child is able and not able to do relative to the peer. So we do have to look for it. And let's go back now to sort of what happens for this young boy for example, as he's trying to build a brain in the presence of a vestibular deficit. And what you can see is as important as audition was in this pyramid of cognition and intellect, vestibular function is at the base. It is actually even more than important than audition yet we don't really even factor it into our children's outcomes. And it makes sense, if you can't stand upright then how are you gonna learn to read? If you have complete visual blurring any time you make the slightest of movements, then how are you gonna read what's on the whiteboard or in the book in front of you?

And so intact vestibular function is highly important for children to develop the cognitive and intellectual skills they need for adequate development. Yet we don't measure it, yet we don't recognize it in a way that we can provide them therapy for it. The other important thing about the vestibular system is that it projects everywhere. It projects to so many different areas of the cortex and that is very, very important in terms of its overall function. The auditory system is pretty specific in terms of where it projects, but the central projections the vestibular system are pretty immense and amazing. And so oftentimes the impact of that sensory deficit is broadly based and somewhat non-specific such that we don't recognize it or pin it on the inner ear. And so let's think about what that looks like for a child. And so this is essentially the world of the child. You know they interact and they navigate all at the same time and so when you think about how we communicate, you know, prior to the pandemic is that, you know, if we were at a dinner or at a party, you know, I would look at you, you would look at me and we would have a conversation. However, when you watch children interact with their environment, they're moving, they're talking, they're not looking at the auditory source. And so they're so much more dynamic than we are in our adult world and that's very important and why we need to think about both balance and navigation when we think about, again, them building the cognitive and intellectual skills they need. And so let's think about how that all comes together as they build a brain. And so balance and navigation requires inherent knowledge, spatial knowledge, where you are in space but also where are the things around you in spatial memory? Have I been in this room before? How far is the door? What's outside that door? All of that comes into play and requires input from our senses as well as input from our muscles and from our joints. It's a very complex thing. What we're gonna focus on for today's lecture is not all of it, but we're gonna focus on the impact of vision, hearing and vestibular and function on how children acquire the spatial knowledge they need to successfully navigate and balance in a three-dimensional world. So let's start with hearing and I've written it in red.

So we're gonna start with what happens when hearing just isn't working? And so for that we're gonna look at some of the data that we obtained through the BOT2. So here we have two groups, we have a group of normal hearing children that we retested and we have our group of all comers with hearing loss. Being that we're an implant center, these are mostly kids with profound deafness who are bilateral implant users or who are becoming bilateral implant users. And what you can see is that on a whole they have an order of magnitude worse balance skills than their normal hearing controls. And some of these children have intact vestibular systems, not all but some. And so next what I'm gonna do is and we'll start the video is I'm gonna tell you a bit of an anecdotal story about what when we change the auditory environment in order to emphasize the importance of the auditory environment to balance in our children. So this is a young girl, and I'll start the video in a sec, who is a competitive gymnast, she has bilateral hearing loss and bilateral cochlear implants but normal vestibular function. And so we did a little bit of an anecdotal experiment and we had her take off her implant in this scenario. So she's in the unilateral configuration and here she's got her usual bilateral configuration. And what you can see is that she does fall in both of the occasions, however, she does so more so in the unilateral locations. And this fit with a report from her mom that she could always tell if one of her processors went down during practice or worse yet had a competition, she would witness an acute change in her balance function. And so a change in this young girls auditory environment, and in particular a change in the electrical milieu of her inner ear caused a change in balance in the presence of normal vestibular function. So hearing in and of itself, and we can go back to the presentation is an important variable in terms of children being able to maintain a right posture. We wanted to take this further and look at the effects of poor spatial hearing. And so we did this with three groups that we had access to.

First, we took a group of kids with normal binaural hearing. So these are normal hearing children who have no asymmetry between their ears. We compared this to a second group of children with abnormal binaural hearing. So these were our bilaterally implanted children who had a long duration of use and who had symmetric caring, essentially they were the simultaneously implanted group. And then we have a group of children who we know do not have access to any binaural hearing. These are a group of children with untreated single-sided deafness. And I think at the end we're gonna finish with a little bit on single-sided deafness. So here's partly where it ties in, okay? And so we put these children through a number of different memory and learning and navigation tasks and I'm just gonna present a small portion of that data. What we found when it came to memory and learning deficits is that these children with hearing loss and abnormal binaural hearing had them. And interestingly our children with untreated single-sided deafness were as bad as our children who were profoundly deaf but received bilateral implants. And so for us, that was a really surprising finding. This is a group of children who have normal hearing ear but their memory and learning deficits were similar to those who were profoundly deaf but received simultaneous implants at an early age. So the best possible scenario, okay? And what was even more interesting is if we looked at what kind of memory and learning deficits we had, we found that they were visual spatial in nature. And these were the tasks that we used in order to put them to the test. So this is a two-dimensional visual spatial task where the dot appears in a number of areas and the children have to remember the sequence of the dots. And this is a three-dimensional visual spatial task where the tester taps on the blocks in the specific series and then the child has to remember that. And these is where we saw these deficits in both of the groups with binaural hearing impairment. So what if both hearing and vestibular function is impaired? So this is the 35 to 40% of kids that I spoke about at the beginning of the talk, what happens with these kids? Well, let's first look at their balance skills. And so again, here's our group of normal hearing children, in red is all comers with hearing loss, many of which are in the cochlear implant range and this is what happens if we take out that 35 to 40% of children who have bilateral vestibular loss. What we see is that their impairments are an order of magnitude worse both than the whole group with hearing loss, as well as the normal hearing group.

And, you know, we talked about the idea that sometimes these kids are difficult to decipher because they do walk and they do run and they do do activities that may make us miss or under-appreciate these impact of their sensory deprivation. And the reason for that is if we look at, you know what does it mean to have a BOT score of around five? You know, what age child would we expect that to be normal for? Well, who it's normal for is your average four and a half year old. And if you think of any of the four and a half year olds in your life and what they can do, they certainly walk, they frequently run, they can jump, they are able to stand on one foot for a period of time, they're probably not able to yet ride a bike without trainers, or if they do, they've just learned. And so the four and a half year old can do lots of things and that's why we miss the impact on these children. The trouble is that a balance age of four and a half isn't gonna serve you well throughout your lifetime, especially when you're at the other end of that spectrum and undergoing a cognitive decline. If you're only starting out with the balance scale of a four and a half year old, you're gonna be impaired very, very quickly as cognitive and motor decline occurs with older age. And certainly if we think about what we're willing to accept in the speech and language domain, we wouldn't be happy if the outcomes of our kids with cochlear implants or with hearing aids was a language age of four and a half. And so I use this data to suggest that we shouldn't accept this from a motor perspective either. And so let's think about our young boy with Usher's, okay? So he essentially is gonna plateau at a balance age of four and a half. Now what if he wants to learn to ski or take a ski jump for example? Do you think an average four and a half year old can do that? Probably not. And so this is what it looks like, and we'll go to the video of a child with a balance age of four and a half taking a ski jump, here we go. And of course that's gonna happen. We'll go back to the presentation and you know, this is a child who doesn't know where they are vertically in space and that has got to be important when you're trying to learn to ski jump. And so this young boy's physiology and his lack of vestibular function is making it very difficult for him to do something that he wants to do. Now, let's talk about vision?

So let's get out of the inner ear for a second and talk about vision. And the reason we wanted to look at vision is that there's generally this concept in vestibular physiology that thankfully unlike hearing for the balance system we have all kinds of different inputs. We have visual input, we have proprioceptive input, we have vestibular function, and we have all of this capacity to integrate these senses in order to deal with conflict or less than ideal situations. And the idea in adults or older children is that when an individual acquires a loss in one of these systems, then the other senses pick up the pieces. But the reliance and the capacity for them to do that is based on the foundation of a framework that allows this integration. So what if you're born without a sense, does that not mean that you'll never be able to develop the framework to allow those senses to integrate? So how could you possibly map onto them if you then lose a sense because that framework never existed. So we went to vision in order to try and see what we could decipher in this regard. And so we looked at a group of children who have developmental visual anomalies. And so this was a group of kids with strabismus and amblyopia. And what we demonstrated by having them do the BOT2, so these kids have normal hearing, they have normal vestibular function. And what we see is that compared to the controls they have significantly poor balance skills. Now vestibular physiologists will tell you two out of three ain't bad but it's certainly not good enough for these kids and that's because these are congenital sensory impairments, just like many of our kids that we've been talking about in the hearing domain. And if we compare this to the graphs I've shown you previously, this is what we see. So they are not as good as our all comers with bilateral hearing loss, they're worse than that group and they're nearly just as bad as kids with bilateral cochlear vestibular loss. And so this tells us the important of the idea that we have to look at these kids with congenital loss in a different way. They're not gonna be able to tap into the compensatory strategies that we expect of an individual who acquires this loss. Not surprising, we see the same thing in the hearing world. And so let's go back to our framework and think about what is it that we have the capacity to do and to help these children with? So, first of all, we do have great ability to help restore hearing through hearing aids and cochlear implants. So how does that help us? Well, this is some of our very early data which demonstrates improvements in balance skills in the presence of bilateral access to sound.

Now I'll point out that these differences while significant are the effect size is pretty small. So what we see is that this is BOT2 scale score that improves and again, it's a pretty small improvement about one point on the BOT2 scale score which is defined as being significant but they do improve in the presence of bilateral hearing. This is individual data which demonstrates that sort of hearing access to sound doesn't generally hurt balance and certainly in many kids improves balance score. Although again, the effect size for this is pretty small but sometimes we'll take whatever we can get. Now, what about plasticity? How far can we push the system? And so we'll start the video and this is the 64 lead EEG that I mentioned earlier. And it's a technique that was designed in our lab by one of our PhD students, Dan Wong. And what you can see is it's similar to an FMRI, but it's looking at activation over time and you can see different areas of the brain light up during tasks. And we can use it in our kids with cochlear implants in a way that we're unable to use other functional imaging modalities because of the magnet. And so this next bit of data I'm gonna show you utilizes this technique so we can try and figure out you know, what plasticity there is and how we can potentially use it to extend the performance of kids with cochlear vestibular loss. And so here's an example of some data that uses this technique. And what we did is we took a group of teenagers who were normal hearing, and teenagers who experienced cochlear implant users. And we had them do a specific listening task. And then we took the 64 lead EEG data of the two groups and we subtracted from the group of kids with cochlear implant, the data of the normal hearing group. So we wanted to take out everything that was similar and see what we were left with. And what you see in this graphic here are all the parts of the brain that light up in the kids with cochlear implants that did not need to light up in the kids with normal hearing. And so these are the novel pathways or the extra work that these children needed in order to accomplish the same task. So they got to the same goal, how they did it was different and chances are how they did it required more effort 'cause that's the thing about plasticity is that while it's possible, it often occurs through mechanisms that require more cognitive effort. And so that's the other tool that we use is looking at measures of effort of processing and so reaction time is the classic one and what we've been using more and more is pupil diameter.

So for those vestibular diehards we actually used our eng goggles to look at pupil diameter as a proxy for effort of processing for these kids. And so what we see is yes, they can do it, they can get to the same goal, they use different pathways and it costs them more effort. And so if a child in a listening task can with effort and with practice get to the same goal, what about a kid with vestibular impairment? If he practices enough, can he land that ski jump? If he puts enough effort in, can he defy his physiology in order to do what is so developmentally important to him? So let's go to the next video and we'll see what happens in that context. So we can go to the video, here it comes. Now this is a feel good story. So the answer to that question is, of course he can. There he goes, our child with Usher's, it's so beautiful, let's watch it again. Defying what his vestibular system is telling him that he should not be able to do and he's landing that ski jump. So let's go, you know, as we go back to the presentation I'll tell you, you know, we had a conversation with this young boy's mom because these kids do all sorts of amazing things. And what I'll tell you is that his mom said, yeah it's pretty amazing that he did this but he spent entire two weekends, that's the only run he took on the ski hill. He didn't interact with his friends, he didn't do his homework, he didn't make his bed, he was exhausted, but he did what he set out to do. And it speaks to both the plasticity, but the cost of that plasticity, which is effort. And so what we then wanna think about is can we make it easier? We have the capacity to restore hearing, but what about giving some more information to that hungry brain that just wants some vestibular input? And so this brought us to one of the questions that we had early on in our studying which is can electrical current from a cochlear implant spread to the vestibular system? So that was the simple question and we'll talk about that answer next.

So the short answer is yes. And how we demonstrated that is we went back to some of the evoked potentials that we can get from the vestibular system in particular the vestibular evoked myogenic potentials which are a measure of otolithic function. How you elicit this for those that aren't familiar is with an acoustic stimulus that you see here. And this is a beautiful VEMP response in response to an acoustic stimulus. Now, what you have here is the same ear that has a cochlear implant in it and instead of putting an acoustic stimulus, we are turning on specific electrodes at relatively high current levels and we're able to evoke that same myogenic response. You know, this is pretty cool, but what's even cooler however, is the fact that we're also able to do so in an ear that no longer has an acoustic response. Either because it's been obliterated by the implant surgeon or it's been obliterated by the etiology of the deafness. And so we're able to use an electrical stimulus to restore a response in an ear that was no longer able to do so in the natural way. Now, for us that's pretty exciting. But the question remains will this help that young boy land the ski jump better, with less effort? Hard to know. And so this is where the behavioral correlate comes in. And so part of again, we've been talking about spatial awareness and we like to also measure the functional deficits in terms of assessment of verticality. And so what we did also in these children is we looked at something called the subjective visual vertical. And this is a test again of otolithic function and this is how we measure it using a bucket and a iPod app that's been validated in pediatrics. And so here's our data and we'll go through it slowly. So when you have an abnormal SVV, you either lean to the left or you lean to the right. And when we turn on the cochlear implant at the same level and current level that we did in order to obtain a myogenic response what we see is that the left leaners shifted right and the right leaners shifted left. We were able to correct an abnormal perceptive verticality by providing an electrical stimulus to the inner ear. Now that I would imagine may help this young boy land his ski jump. You can imagine that the perception of verticality would help you in terms of landing that jump. And so this gave us the possibility that perhaps even with the tools that are already implanted we could make these children's lives easier, but how far could we push it? And that is this part of the experiment that's been going on for a number of years where we've been looking at stabilization of balance using signals from the cochlear implant. And what you can see here is the latest version of this where we're converting cochlear implant stimulation into a head referenced signals.

So there's an accelerometer and gyroscope in here that is measuring the deviation signal of the head, and that then gets transferred electrically to the child via the implant. And we've tested a number of patients and we have a second cohort going through testing right now. We'll go to the video for this and this was one of our first attempts at this with our original prototype which was actually an iPod that was attached to the child's head. And this is our young boy again with Usher syndrome. And what you can see is that on the left side of the screen is that he's unable to to stand upright and on the right hand of the screen he's got enough cognitive reserve left to clap his hands, he's able to stay upright, there's way less sway and he also learned quite a bit about how he had to move his body in order to maintain the stability of that center of gravity. And so while the video is convincing, we need to look at some data, so let's go back to the presentation and we'll look at a group of 16 children and what their performance was like. And so what we saw again using the BOT2 as a measure were significant although relatively small but meaningful improvements in balance when they were provided with this head reference stimulation. When we look at time to fall because we know falls are important as they induce injury and put the implant at risk, this is what we saw. So this is our group of normal hearing children going through the items of the BOT from easy to difficult. This is what we see in blue is our kids with bilateral cochlear vestibular loss and this is what happens when we give them head reference stimulation is that it seems to improve significantly these mid-range tasks, although we still struggle out here with these relatively impossible harder range tasks. And so this was encouraging data. When we look at posturography data and here for those of you who aren't familiar, a reduction in a number is an improvement, so we're looking at center of pressure at the level of the feet in terms of root mean square, duration and velocity. And in each of these domains, we see significant improvements in function. And so this has given us a really encouraging view into how we can help these children with bilateral implants not only hear better, but also balance better. But what we know is that, you know, if we were where we needed to be, there'd be no more research questions.

However, we know as I described at the beginning that so many of these children still have functional deficits in their day-to-day life. There are things about how we are rehabilitating them that falls short in terms of their capacity to connect in the world with their peers. And so part of this final chapter of this talk is really looking at a population who may be the key to us figuring out what's missing. And remember I said we'd talk a little bit about single-sided deafness. Well, this is where it comes in. 'Cause children was single-sided deafness have a normal hearing ear. They don't need a cochlear implant to develop speech and language like our kids with bilateral deafness do. They're for the most part gonna develop that based on their single auditory input, but their lives are not normal. There are things that they would like to do better with less effort. And so if we can figure out what we can make better in those children with cochlear implants for example, or with amplification then perhaps that will unlock the key to restoring connection in our kids with bilateral implants. It might be the what we are missing. And so let's take it this final chapter a little detour and look at our kids with single-sided deafness. And so, again, this is the auditory world of these children. You know, and if we think about this young girl if she has hearing loss in this year, again, her access to sound to what's happening on in behind her may be very difficult. And we know that sensory deprivation is bad, it's how we started this talk but there is a burden of evidence to suggest that unilateral hearing loss has immense impact and this dates back initially to Rosemary Tharpe and Bess who looked at grade failure and a need for additional resources. But in 1991, there was no way that we were ready to put a cochlear implant in a child with single-sided deafness. We would have thought we were absurd for even suggesting it. And this data continues to be updated and the conclusions continue to hold true, which is that unilateral hearing loss does not set children up to succeed and there can be variable impact depending on these children's environment, their baseline cognitive ability, and that the impact of unilateral hearing loss extends much beyond speech and language to quality of life, to school performance, to their behavior, to their need for additional resources.

And so it really is important that we recognize our capacity to intervene, but in whom should we intervene? Now at the beginning we talked about the idea that we have learned so much as a result of cochlear implantation. And this is another example where we have learned a lot because we're asking the question. And so this is one of my favorite slides and it's looking at the etiology of single-sided deafness in 1990, back when that data from Bess and Tharpe came out. And what you can see here is 60% unknown, we had no idea why they didn't have hearing in one ear. But now we know this because we've been looking at these kids because we've been thinking about implanting in them and so what we've learned is that this 60% is made up of two groups, children with absent cochlear nerves and children with asymptomatic CMV. These are groups of children that were underappreciated but that we now know about because we're investigating them for cochlear implantation. And the kids was CMV and we're going to talk about them 'cause I think there are quite a special group, they've always been in our implant programs. This is them 2%, these are the kids who were severely affected by symptomatic CMV, but there's this whole cohort of kids with asymptomatic CMV walking around often with dual sensory loss and we'll talk about that. So let's sidetrack a little bit to CMV. And so this question really came through as we were investigating, who we should be implanting from a single-sided deafness perspective. And what we found is that one of the most common etiologies in the groups that were presenting with SSD were kids with congenital CMV. It made up roughly 20% of all referrals, but when we took out all the kids that couldn't have an implant, those with no nerve for example, what we found is that more than 40% of those kids that were actually implant candidates had congenital CMV. And frankly 43% of the kids that we ended up implanting were CMV. Fascinating. So let's look into this a little bit more. This is what happens in the setting of congenital CMV to the stria vascularis.

So these are from Dr. Harrison in his lab here at SickKids. And what we see is that this is the stria vascularis through corrosion casting in a normal mouse cochlea. And this is what happens when that mouse is exposed to congenital CMV. It gets decimated. And so this certainly accounts for some of the 20 to 50% of kids with congenital CMV who develop sensory neural hearing loss. But this is a talk about how do we fix this? And so what we see is that single implantation for single-sided deafness in kids with congenital CMV is hugely acceptable. Now any of you who have counseled families on single-sided deafness know that it is not an acceptable intervention for a lot of families, probably only 30% of kids that we see who are candidates will actually say yes to implantation but 80% of those with CMV say yes to implantation. And this is the reason why? When one ear has early profound or congenital loss, the probability of developing bilateral loss is 75%. And so that's the risk that speaks to these family and that we can think of implanting in CMV as a long delay sequential, keeping the input symmetric until the other ear goes in 75% of cases. Now the other reason we consider these kids is that it's hugely common. It's actually the most common thing that we screen for now universally in Ontario. Here are some of the numbers. And the trouble with it is that it happens early and it progresses early but many of these kids are born with normal hearing, so our normal screening mechanisms miss them. A lot of them will lose their hearing in that ear between three months and a year and not get recognized until they're school age which is probably too late to rehabilitate them well with a cochlear implant based on our data from previously. The other thing is that many of these kids have dual sensory impairment. So here we have accumulation of the virus in the vestibular system. And what has been shown in the literature is that a good proportion, at least 50% of kids with congenital CMV will have impairment in the vestibular part of the inner ear. So most of these are gonna be in the category that we've talked about. When we look at balance in our kids with SSD, what we see is that they have significant balance impairment compared to normal hearing and here they are relative to the other groups we've been talking about, all comers with hearing loss and bilateral vestibular loss. And so again when we look at their end organ function, 50% of them have vestibular dysfunction.

So this really brings us back to the consequences of SSD, because if we're gonna look at outcomes from our interventions, we have to suggest that we not only have to look at the fact that they have unilateral hearing loss but that they have unilateral labyrinthine loss and that the vestibular component of this is probably responsible for some of these deficits. And perhaps the child with combined cochlear vestibular loss is a better candidate for a cochlear implant than a child with hearing loss alone. And so that is the question for the future that I pose. Now we know that we can improve hearing through the use of cochlear implants in single-sided deafness and we're gonna go back to the beginning in terms of the data looking at aural preference. And so this is what we see. So what we see is that over time with a cochlear implant we can restore normal aural preference in the setting of SSD. So we're able to get the brain to be built as it normally was. And this is a measure again of plasticity. So certainly in summary, sensory deficits are a big problem for these children and we have great tools in the form of implants, but there's limits to this with respect to the effortful compensation these children have. Perhaps we can push the limits and get more benefits than just hearing alone, but outcomes are complicated and we need to know really what it is that we're dealing with in terms of sensory deficits when we're measuring outcome. And so I'm gonna leave you with one last video, and that is just to remind us and we can go to that video now about what drives plasticity in children. And we're gonna go back to our, if we can start the video, we're gonna go back to our young boy with Usher's. And so his dad is from the Dominican Republic and what greater motivator is there for a young boy than to do what his dad does. And so here is that young boy, despite his absent vestibular system, getting up on a surfboard. And what this reminds us of as clinicians is what an amazing role we can have because humans are designed to defy their limitations. We all have obstacles, we're certainly having lots right now in the midst of this pandemic but as humans we're designed not to break. So scientifically and clinically let's tap into that design and figure out how we can get these children to achieve their goals with less effort. Thank you very much.

- [Elizabeth] Thank you so much Dr. Cushing for a fantastic presentation and very hopeful and I love that little boy getting up on that surfboard should be the new meme for 2020. Thank you so much for the time and expertise. Thanks to everybody who participated today. We really look forward to feedback on this course evaluation and seeing you in future courses on Audiology Online. Thanks so much Dr. Cushing, bye bye.

- [Sharon] Bye.