

Transcript

[MUSIC]

Karen Gordon

Hello and welcome to the award-winning Hear Here podcast. I'm Karen Gordon. I'm an audiologist and scientist at the Hospital for Sick Children in Toronto, Canada, and I'm delighted to introduce our season five. You'll be hearing episodes hosted by our Hear Here Podcast Team. They are an amazing group of individuals who have experience living with hearing loss themselves or with siblings who have hearing loss. So you'll be hearing from Maria Khan, Nimrat Chani, Sofia Olaizola, Annika Gasse, May Wang, and Maryam Othman. In season five, we recognize the contributions that many families have made to our research conducted at Archie's Cochlear Implant Laboratory here at the Hospital for Sick Children. We discuss the findings of studies that often take many, many years and represent so many hours that children and their caregivers and loved ones spend with us and we really appreciate them and all of this contribution they've made.

[MUSIC]

Hello everyone and welcome to what will be season 5 of the Hear, Hear podcast. I'm Karen Gordon and I am very happy to be directing this season to people who might have been involved in our research at Archie's Cochlear Implant Lab. People often ask us, What did we find? What have we been doing? And we are really happy to be able to talk about what we've been doing in the lab. And we are dividing this into different episodes. So. Each one of these episodes is going to have different guests and different hosts. So I am going to turn this episode over to our host, Maria Khan, who's going to tell us a little more about what we're doing and who we're speaking to.

Maria Khan

All right. Hello audience, hello listeners. As mentioned, my name is Maria. I'm a member of the Hear Here podcast team. I'm also just graduated or just finished my master's in audiology. Woohoo. And for this episode, we're going to be diving into balance in the vestibular system and all that fun, research-associated with that. I believe this is a topic our listeners may not be the most familiar with when it comes to hearing because even within the scope of audiology itself, it's like, oh, audiologists also do balance things I never knew going into my own program. Like I wasn't aware of the extent of the scope we have in terms of balance and vestibular. So I believe it'll be the same for our listeners. So I think before I get into explaining what exactly balance is and what is the vestibular system, we

should introduce our wonderful guests who we have on the call. So maybe I'll hand it off to Melissa to start off.

Melissa Hazen

Hi everyone, I'm Melissa. I am a clinical audiologist at SickKids and I am also a PhD candidate in Archie's Cochlear Implant Lab at the University of Toronto. And so, yes, I do vestibular testing. I'm part of the Infant Hearing Program. A lot of my research is focused on children with hearing loss who also have vestibular issues and how that translates into developmental and spatial abilities. And there's, you know, a big question around all of this and how it's growing in terms of the research as well. Yeah, that's a little bit about me.

Maria Khan

Amazing. And then we can hand it off to Dr. Cushing.

Sharon Cushing

I'm Dr. Sharon Cushing, and I feel like I've grown up in this program. So I first joined as a resident in 2003 and got introduced to the amazing Dr. Karen Gordon and ultimately became her master's student in the midst of my residency. And it's been, you know, a learning journey and the very roots of that initial research were the beginnings of our look beyond just the cochlea and hearing into balance and vestibular function. And so it is really been a journey and we've been able to tell a story and that story is still going. I'm now on staff here and have been for the last 15 years and I'm one of the cochlear implant surgeons.

Maria Khan

Amazing. Now we introduced to our wonderful guests. Before we dive into what is going on with the research, maybe we should define what exactly is balance, what is the vestibular system, what's its role, and how does it work for children, infants, as we know, as they develop, they have different milestones that they'll hit. So who'd like to start us off with explaining all that? A little crash course, if we will, for our listeners before we get into the meat of the research.

Melissa Hazen

So the vestibular system is part of the inner ear. It's attached to the cochlea. The way I like to explain it is it tells your brain where your body and your head is in space. And so you can think of it as like the body's balance control center. And so whether you're standing still or you're turning or you're moving, it helps a child, it helps a person stay upright to balance, to walk, to run. And while you're moving, it keeps your eyes steady and it keeps your visual

field steady. And yeah, sometimes when it's not working properly, what you might see in a child is like clumsiness or falls or feeling dizzy. And the same with adults. If you hear of adults having vertigo, it... And what happens is they start to avoid movement because they feel uncomfortable. that's, my first explanation of the stability in the balance system. Sharon, I don't know if you have anything you'd like to add to that.

Sharon Cushing

I think, there are motion sensors, right? But, that is thinking of them in the simplest of forms in that, what they do and the function that they help us with sort of permeates all aspects of life. balances the core of what we think the vestibular system does, just like hearing is the core of what we think the cochlea does. But then there's all the things in terms of human connection and how we navigate this world that extend beyond those sort of simplistic senses. And so I think that's what we've been learning and where we continue to learn is what does it mean to navigate the world with balance dysfunction, much in the same way that we've asked the questions of what does it mean to navigate the world and not just hear with a cochlear implant. So I think that's where it's really, it's exciting and we don't have all the answers, which in many ways makes it more exciting.

Karen Gordon

Yeah, Maria, I think that what you said about going into audiology and not really thinking about the balance and vestibular system is something that rings true for me too. We almost got to the balance system kind of last. Well, oh, by the way, in your inner ear, you also have these five sensory organs that are going to tell you about your position in space and how you're moving in space, which is a lot of place in the inner ear, many sensory organs dedicated to this important sense. It develops, you know, it's an early developing system. It's been around in evolution. probably before hearing, right? So it's super important to us for how we even get up and move around, right? So you need a vestibular system in order to move. And it's probably pretty important to us as humans as we get onto two legs and then start to walk and so forth.

So for children, it's really important. And Sharon, actually, when she came into the lab and already had a background in vestibular physiology, Really brought this to my attention, and I thought, Oh, that's an interesting part of hearing, and because hearing impairments... are often in the inner ear. We think about them being in the cochlea, but why wouldn't those changes that are occurring in the cochlea also be occurring in the balance organs? There are these hair cells, these sensory cells in both the cochlea as well as the five vestibular end organs that function in similar ways. So if they are compromised by something genetic or an infection, it stands to reason that they could suffer similar changes.

Maria Khan

We did learn the vestibular stuff within our last term of second year and there was so much to learn in that one course. To me, it feels like vestibular and balance is still emerging within the field, even though it is established that within audiology, this is within our scope of practice, but not every audiologist is a vestibular audiologist or has a deeper knowledge set in terms of testing, in terms of explaining to parents and families, your child may have a hearing loss, they may also have a balance disorder and how those two are connected.

Sharon Cushing

I think you've hit the nail on the head, Maria. And that's part of the reason why it's like the lesser-known part of the inner ear, right? Is that when you sort of elicit the words vestibular, whether it's to an audiologist, an ENT resident, a family doctor, everybody feels a little vomitrocious in that moment, right? Because it is like, it is scary. It's complicated. It is way, way, way more complicated than the cochlea. And we don't know how to fix it, right? So we've got some pretty good tools in our toolbox for hearing. We've got way poorer tools for the vestibular system. And that's the other reason it doesn't get the attention that it needs and deserves. But it's complicated, right? And so that degree of complication, because as humans, we don't like what we can't understand, right? What we can't explain to families. We don't like to live in that area of uncertainty. And certainly our families don't like to either when, like to I hear a doctor say, I don't know. Oh my gosh, it's terrifying, right? And so I think that really speaks to why we don't teach it enough, why we don't look for it enough, and also why we don't know what to do with it.

Maria Khan

That makes a great segue into what we know and what the current research says about treatment in terms of diagnoses and all that.

[MUSIC]

Before we get into the rest of the episode, there may be a few terms that you, the listener, may be unfamiliar with, so I am here to break those down so you can better understand the episode. To start off, when we say electrode, what is that referring to? When someone is getting a cochlear implant, the sound signal somehow needs to get to the brain, which is done through a tiny electrode, a silicone covered wire with metal contacts which allow for the electrical stimulation of the hearing nerve, and the person can perceive sound.

In our discussion, you may hear the terms simultaneous and sequential bilateral implantation. What this is referring to one, did you get implanted on one ear (unilateral) or

both ears (bilateral) and two, did you get it done at the same time (simultaneous) or did you go one ear first then the other (sequential). Alright, back to the episode!

[MUSIC]

Sharon Cushing

I think, this is where the origin story, I think, matters, right? And, there have been some pioneers in the field of vestibular physiology in children on the clinical side for like decades and decades and decades. What really led to having some traction actually was this concern that was raised clinically. So, back in the early 2000s, we were at the point of only putting in one implant in children with bilateral hearing loss. And we were starting to think about, well, you know, we can do this safely, should we be doing it in both ears? And that's where the vestibular system got a little spotlight. And so we went from like a few people who were experts and had studied it in schools of the deaf and in children with hearing loss to people being more concerned in the mainstream of clinicians that look after children and adults with hearing loss. It was by no means like everybody.

Again, it was still a very small group, but it added some traction because what we were worried about is that by introducing an electrode as gently as we do it into the inner ear, you know, are we going to affect the vestibular system? And if we do it on one side, okay, not so bad because we've got the other ear and these systems work in paired function. But what if we do in both systems at the same time? What's going to happen to those children? Are we going to render them unable to walk? What's going to happen to adults if we do that? And that was really where we said, okay, we got to start looking at this. And that was certainly the revelation for us and understand, when is it normal? When is it not normal? What does surgery do to it? But that was the question and the worry that led us to care about this. And it's really not been the most interesting part of it. It's the questions that came from that. And that's what, you know, Mel, Karen and I will talk about as we go through it.

Karen Gordon

I would say that the fact that it wasn't interesting was also a great thing, because we were so worried about whether this was going to be a problem. And we found a couple of interesting things

Sharon Cushing

Yeah, we went into this with this worry about how bad is that implant going to be for the balance system. And what we found, in summary, is that deafness is way harder the vestibular system than any implant surgeon. And what I mean by that is that, you know,

whatever is causing the etiology of deafness, whether that's a genetic change or an anatomic anomaly or an exposure to a virus or to medicines, it had a much bigger potential to injure that system than putting in an implant. And so, The first thing that we found was documenting how many kids had vestibular impairment. And the numbers were huge. And by huge, I mean 35 to 40% of our children had an abnormality that was considered significant of the vestibular system. Now, this is a special population.

This isn't all comers with hearing loss. These are children who were being implanted bilaterally or sequentially bilaterally in the 2000s. Those are different candidates than we see today. And it's not every child with hearing loss because these kids had severe to profound. But that was the first like, whoa, whoa, wait a second. You know, these kids are quote unquote walking into the operating room with these deficits already. And so That was ultimately our first finding. And then we wanted to say, okay, how do these kids do, right? To get at that question of like, great, your lateral canal is not working. What does that mean, right? Your otoliths aren't working. What does that mean? And so the first fundamental behavioral result that we went to look at was balance. And so, you know, we measured that and we were curious. So we measured it with the implants on and we measured it with the implants off, you know, thinking perhaps even that maybe off is better 'cause then you can really, really focus. And what we found is that in particular, if you gave sound access on both sides through implants, the balance was better. And man, that was exciting, right? It's like the opposite of the question we were hypothesizing, right? And that's the joy of science, is you go in with one question and you come out with an answer that is completely different. You just have to be open and curious, right? You don't want to miss those opportunities because you're so focused on, well, this is what I thought. And so I think, those were really the two fundamental things that we found, in those first early years of studying in the lab. And so many patients were so willing to help us with that research. it's not always fun to have your vestibular function tested. It's easier now than it was before. But still, we, patients were really the reason we could get that data.

Karen Gordon

Yeah, I just want to reiterate that part is, for those people who were involved in the research that we did, we were so appreciative of them doing these tests. They're clinical tests. Some of them... were just new to us and new to the battery of things that of testing that we had been doing in the past. So, families were so good about just having these additional appointments and going in and this is what has result the knowledge has that. has been taken out from those extra appointments and time that they spent, not only understanding their own particular child, but how we were able to put that together to a better understanding of what children with hearing loss have. The other thing is that we

learned it was kind of fun, right? So there was, you know, this test of balance that Sharon introduced to our program really taught us a lot about, how children stand upright and how they move and how, and we called it the vestibular Olympics because it was actually super fun to do these tasks

Sharon Cushing

Karen, do you remember when we did it at the picnic? We had a cochlear implant picnic and we brought like all the stuff and it was one of the first times where we were trying to recruit as many kids as we could and we had siblings and families and we had a couple of setups and that I can still picture that in my mind. And, just to re-emphasize the impact that families make, I can remember and recognize the patients who participated in my research. Of course, it's all de-identified, but at one point, there is a name to that human, right? And when those names come up, even this, like 20 years later, right, more than 20 years later, I'm like, oh yeah, they were in my master's thesis, right? And like, I will carry those patients and their data with me, like for my entire career. That is how impactful it can be, even just beyond the science.

Maria Khan

And what tasks were in the Vestibular Olympics? Very curious.

Karen Gordon

Sharon called it the Vestibular Olympics because it just looked so fun. And there were kids lined up to like do it, right?

Melissa Hazen

Oh, that's funny. That's funny.

Sharon Cushing

No, for the listeners, it's the Brunus-Oseretsky Test of Motor Proficiency, balance subset. And so like Vestibular Olympics is way funner.

Karen Gordon

And Sharon is the only one who can get through that name. I guess I still say BOT.

Melissa Hazen

Yeah, I mean, the BOT's fun. Even clinically, like we still, do it on children who come through our Dizzy Clinic and they love the test. It's fun, It's comprised of nine different tasks. There's standing on one leg, eyes open, eyes closed, standing in tandem stance,

which is one foot in front of the other, walking on a line, one foot in front of the other, standing on a balance beam. And it's fun for them. So yeah, it's a little bit, I guess, yeah, like Vestibular Olympics, a little bit of gymnastics in there. It's a fun task for the children, for sure.

Sharon Cushing

What I loved was watching the parents watch their kids do this test. And so like there's some easy stuff on this test, like, standing on a line and then it gets hard and then it gets really hard, right? And that's why it's such a beautiful test is that like, depending on your age, like there's going to be parts that are hard enough, especially for our kids with hearing loss. And, so kids managed the easy stuff. But then when it got hard, parents would sit there and go, oh my, how come they can't do this? Is it because it's super hard? And then all of a sudden you'd see the parent at the side, like trying to do the same task to gauge like, how hard is this? And like, should I be worried that my child can't do this standing on a balance beam with 1 foot eyes closed, right? And so I think that when you got into the nitty-gritty of how these children navigated the world, it would come out in the history, but I think it was revelatory to families as well to sort of actually watch the performance of their children in a task where it was difficult enough. And that is the crux of balance dysfunction in children, is that they do great and they struggle, and, not or, and. And so it depends. They, in certain situations, they can manage and they use every bit of sensory information to manage and they fool us. They fool themselves, frankly, right? And if we make it hard enough and if we take away enough of those strategies that they use to succeed in this world, which is so incredible, then we see it fall apart, right? And so that's the part that can be tough because they're succeeding and failing all at once.

Karen Gordon

Yeah, so that came home to me when we focused in a particular study on those children. Some of them, you know, were young adults at the time. We were having them come back into the lab. This was work done by Rebecca Benjamin. we really wanted to know how that function on the vestibular Olympics, the BOT, was going to affect how they could do something while they were standing in tandem stance or while they were doing another balance activity and this was during COVID, we did it not right at our lab in SickKids. We were taking people over to across the street to a very special lab in the Toronto Rehab Institute. And actually, we did a couple of studies there, but in this one, we were putting markers all over the body to see, okay, how are these individuals moving while they were trying to balance? And we actually put them in a big harness so they wouldn't fall and all of that. And then we put them on a treadmill and we like moved the treadmill in certain directions so that they would have to like correct the balance. And it was, interesting to see

they could do it. Nobody actually fell. And so that speaks to this idea that there's, you know, there's such an importance to staying upright that they had developed different strategies maybe to keep them upright and to make the right steps where they needed to stay upright.

Melissa Hazen

I think I think both, you, Karen and Sharon, you touch on a really important point, like a really important point with these children, which is maybe a point of inspiration for me too, where some of these children who have really, really poor vestibular systems can have, you know, very good balance. to a point and then yes, it does start to fall apart, but somehow they find ways to compensate and move in the world and stand on 2 feet and participate in sports and activities. And I think that's, it's really motivating and it is very inspirational. I think of some children who have better balance than I do and they have a super impairments and they've worked hard at developing that skill, which which I think is really amazing.

Sharon Cushing

I've even had, this sort of like talking out of both sides of your mouth in that like they're amazing, but they struggle, like called out, during a scientific presentation where like someone got up to the microphone, they're like, what are you actually saying? Like on one hand you're saying they do great and on another hand you're saying they struggle, like which is it? And I'm like, both, right? And I think that's the super tricky part.

Maria Khan

All right, and that's the end of part one of the vestibular balance episode. The next part, we're going to jump right into Melissa's research and the three parts of her PhD. And then the team will end off the episode with what's next in terms of balance rehabilitation. I hope you enjoyed part one and we'll catch you in the next part. See you then.

Karen Gordon

Thank you for listening. to this episode of the Hear, Hear Podcast. I hope you enjoyed this conversation. I want to give a shout out again to the Hear, Hear Podcast team. Nimrat Chani, May Wang, Maryam Othman, Sofia Olaizola, Maria Khan, Annika Gasee, as well as my colleagues that are involved, Lora Carinci, an audiologist here at the Hospital for Sick Children, Dr. Sharon Cushing, and Dr. Blake Papsin. Also, I hope you enjoy this music, which was performed and composed by Dr. Blake Papsin. Thanks for listening.

Audio file

[S5 Vestib Episode PART 2.mp3](#)

Transcript

Karen Gordon

Hello everyone and welcome to what will be season 5 of the Hear, Hear podcast. I'm Karen Gordon and I am very happy to be directing this season to people who might have been involved in our research at Archie's Cochlear Implant Lab. People often ask us, What did we find? What have we been doing? And we are really happy to be able to talk about what we've been doing in the lab.

Maria Khan

Welcome to part two. If you missed part one, we recapped what exactly is the balance system, reviewed the anatomy and the physiology of the vestibular system. We looked at what it looks like in a child and what can a vestibular deficit do in terms of somebody's day-to-day function. In the next part, we're going to jump right into Melissa's research and the three parts of her PhD. I hope you enjoy.

Karen Gordon

Absolutely. But it's probably a good segue for you, Melissa, to tell us about your present work. Because, you know, we've kind of come so far in defining, you know, what the problems are, where, you know, where the balance issues are happening, what kinds of tasks are harder than others, whether, you know, children can use vision, to help, whether, definitely they're using vision and proprioception, the feeling on the floor to move the feet in the proper ways. So I think that brings us to what you've been studying, and it's been quite some time for, you know, four years, and you've been, and even further, because I guess you could start with even the how you've been looking at the clinical data?

Melissa Hazen

All of it stemmed from this question of, you know, we have these children who come in and they have vestibular impairments and, you know, that translates to these balance impairments, but what can we do for them? How can we help them? And so the thesis was really about stepping back and asking, you know, what else is happening in children with hearing loss? and these vestibular impairments beyond their hearing, just because we know and we focused on for my for the PhD work, we focused on children with cochlear implants, mostly just because of the high correlation of vestibular impairments in this group. But we know that they're at higher risk for things like motor delay, developmental delays, spatial delays. But why, you know, is it the hearing loss? Or is it the vestibular impairment? Or is it something about their environment or their upbringing or their access to resources? And so we looked at this question in a bunch of different ways. And we ran kind of three studies, which I guess I'll explain. And so for the first study, we did a retrospective study of all the children who came into our clinic with a risk of a vestibular impairment. And so that was hearing loss or symptoms of dizziness. And we looked at who actually had vestibular loss and how did that translate to balance function. And we looked at just a little bit over 300 cases just to see how often balance issues showed up in kids with hearing loss. And we, you know, vestibular tests were completed on them and they were split into four groups based off of their hearing status and their vestibular status. And what we found is that children with hearing loss had poorer balance than children with normal hearing and even children with vestibular impairment. And so children with hearing loss, yeah, there's something about hearing and spatial hearing that contributes to poor balance, which, you know, challenges that trilemma of thinking that balance is only comprised of proprioception, vision, and vestibular function. And so I'd like to add, you know, hearing to that. And then in these children with hearing loss, if they had a concurrent vestibular impairment, alongside their hearing loss, then they had even worse balance. And so that was what the first study was about.

Karen Gordon

There's so much that you did over this last four years, but from that first, you know, looking through what we call retrospective data. So these are all the kids who come into the clinic. We always get permission for this, but we get so much important detail from the clinical test too. It just takes, a bit of time to put it together and really look at it. Sharon and you, Mel, put, you know, you guys have this very unique clinic. Of seeing children with complaints of balance problems and dizziness and testing children with hearing loss who with the vestibular test and the balance test that we have, it gives us sort of a responsibility to understand these measures as well in the group. So I think what you did there is really, really important. And one of the things, the findings for me that was so telling was how different the balances by etiology of hearing loss. And I don't know if families have heard that word before. And I think it might be helpful to talk about etiology because when Sharon started in 2003 or when you, yeah, this is not something we talked about. It was, we just have children with hearing loss. And we talked about the degree of loss, but we never talked about this thing called etiology. So maybe, Melissa, you want to talk about that a bit?

Melissa Hazen

How would I describe etiology? Etiology is like the diagnosis or the reason why a child has a certain medical condition. We break it down into, you know, is it something infectious or viral? Is it something to do with genetics? Is it something that happened at birth? Is it something acquired? And then in the case of vestibular function, there can also be some kind of trauma, and I guess with hearing loss as well. And so there's different reasons why a person would have a hearing loss or a vestibular loss. And that is what we define as etiology, is that reason that there is an impairment in the 1st place?

Sharon Cushing

I think that's great, Mel. And, it's one of, my, I had this conversation three times today with families. It's, one of, My favorite things to talk to families about is, there's the why of hearing loss, there's the what are we going to do about it? And those things are sometimes separate questions and sometimes they're aligned. And I always talk about the tools, like what tools do we have in our toolbox to actually define the why? And we have MRI, we've got genetics, we've got some virology tests in Ontario, we've got this amazing ability to screen babies for the why. even before we know they have hearing loss. And as Karen alluded to, the number of children who, despite all the tools we have, that end up in that I don't know why category is getting smaller and smaller. And they're one of my favorite

moments clinically is when I sit with a family and likely a family with an older child who, probably went through the why question 10 years ago or 15 years ago. And for whatever reason, they come back into the fold and we talk about these updated tools. And, you know, when I can tell them the why, I love to savor the moment because shoulders drop. And people breathe more deeply. Because even if it doesn't change what we do, whether that's hearing aids or cochlear implants, you know that those parents have always had that nagging question. Why? does this child have hearing loss, especially when there's no family history, right, which is the case in most. And I can think of those moments and I collect them because there's a gratitude there for taking that weight off, particular mother's shoulders. We're really good as mothers at putting guilt on our shoulders. And so it is immensely important beyond the research component, beyond what it means for us in terms of our treatments. And so I think it's great that those numbers of unknown Y's are getting smaller and smaller. But I would love to hear about what it means for your data. What did you find, you know, for etiology in the vestibular system? Tell me more, Dr. Hazen.

Melissa Hazen

There's specific etiologies that are more at risk of developing vestibular impairment in the presence of hearing loss, namely There's Usher syndrome. There's something called congenital cytomegalovirus, CMV, which we are now screening for in Ontario, I believe, since 2019. So all newborns are screened for CMV. There is Wartenberg. There can be cases of viral infections like meningitis that could put you at risk for vestibular impairments as well. There's a whole host of etiologies that we're starting to learn more about too, right? There's more that are coming up where they're presenting with some kind of vestibular impairment as more and more children are getting tested for their vestibular system, especially at a younger age. Yeah, and that translates in different ways to developmental outcomes as well.

Karen Gordon

Yeah, I think that was something that I've learned through this research too, is that this virus, cytomegalovirus, is pretty common. This virus just likes to be in the inner ear. And it does affect both the cochlea and the vestibular end organs. And that's really what we found from all the testing, from the vestibular point of view and from the balance point of view as well. So knowing that it helps us to look for where and it does help in terms of thinking about a plan for treatment. We may not know exactly what to do, but if we know who needs something a little more from a balanced point of view, then at least we can try to target that. I think ushers is also an important one to talk about. to understand the difference because it's a genetic cause and that's a totally different thing.

Sharon Cushing

Usher is so important and it's where the, and Mel, I know you've got some great data on it, but I'll just give, a little anecdote. And this, again, brings me back to those early years in my masters is that we had an amazing family with two boys who had hearing loss and needed implants. And Mom kept saying, there's something more going on here. both boys, it's got to be genetic. And we were really in the infancy of testing for things like Connexin 26. And, I was doing the testing back then and I was learning and I wasn't so confident from that perspective. But, we identified these boys as having degrees of vestibular impairment. And that was sort of a first domino that led them to moving towards a diagnosis of Usher. And it was very meaningful, I think, for that family, because it's a challenging diagnosis, right? It leads to multiple sensory impairments and ultimately blindness. And so it's a heavy burden. And knowing, though, can you make different choices in life? You make different decisions. You may do things differently. And I think I saw that through this family and that the family remembers that component of it, which I think was so important. And so knowing what the vestibular function is like can also help us with certainty of diagnosis, because it's not always clear, right? Especially in Usher syndrome. And, you know, these boys have been part of our journey. They're like fully grown, amazing adults now, right? And they showed that it is ANT, right? They go and they do amazing things. They do back flips and they do ski jumps. And they persisted at riding a bike and it brought me to tears when they sent me a video at 15 of them finally mastering riding a bike. And so it's how patients in our clinical worlds and research like weave themselves into our hearts. And so again, I think that knowing the etiology again was key for this family? And Mel will tell us a little bit more about why that, why does it matter in terms of how we help these kids?

Karen Gordon

I'm just going to jump in just for a second, because that family really is so important. But to me, it's a part of community building, right? So what I see with different individuals who we've interacted with ushers is this coming together as a community that they have done with people in our area, our program, but with, people all over the world. It's incredible that, first of all, these families are such great advocates for their children and that this connection they have with other people, it creates a community and a strength in understanding what that impacts are going to be in learning from each other. And we learn from that too.

Melissa Hazen

Yeah, I mean, it's pretty remarkable, these children. I'm gonna pivot a little bit and jump into the second part of the PhD, but I'll segue into that with the ushers. So for the second

chapter of the PhD, we looked at almost 100 children, but it was based off of a previous study done by our group that assessed children with unilateral and bilateral hearing loss, and it looked at their outcomes on like working memory, academics, language. And what they found generally is that the children with hearing loss had poor working memory, academics, and language compared to like typically developing children. And so one of the questions was, is that due to an underlying vestibular deficit, right? You'd think that having kind of two hits to your peripheral system, having like hearing loss and a vestibular impairment. And then with that, also a balance impairment, based off of cognitive load and cognitive resources and the brain prioritizing balance over everything else, would that explain some of the variability that we saw in these children with poorer outcomes? And The quick answer is no, the vestibular irvalence didn't come out as anything significant in terms of like explaining why these children don't do as well as typically developing. But we broke it down into etiology as well. And there are a couple groups that came out. So there's Usher's, there's the CMV, Wartenberg, inner ear, anomalies, genetics, unknown. And all of the etiologies across all of these different developmental outcomes had some impairments in one and good function in others. But the Usher's group did well across all of them. So the Usher's group was the only group that didn't fall below typically developing, at least not significantly, across language, working memory, and academics, which I thought was super interesting. And I mean, I don't know if that speaks to, again, that community and the access to resources and help and therapy early on in this group. But they did well.

Sharon Cushing

I think you're right. as I've seen this data and watched you present it so beautifully. And, what just occurred to me in hearing you say that is that, when we look, vestibular dysfunction is front and center in Usher's, right? It's getting there in CMV. It's certainly not there in Wartenberg or other inner ear anomalies. It takes sort of a backseat. And so, you know, I think if, you know, if you Google Usher syndrome, up it comes, right? Hearing loss, vestibular impairment, retinitis, pigmentosa. So I think you're right, there's potentially an eye on, you know, interventions. for to optimize, you know, balance and development and whatnot. And it's hurried in the sense that we only have so much time before vision changes, right? And so I think that that maybe in some families and in some circumstances may help lubricate access to therapies and emphasis on building that skill set early, I wonder. in those families. And so that might be a good set. segue into, what can we do to help, right? Because that's what we love doing. Like we're, there's the research hat and there's the clinical hat. And I think that's what's unique to all of us here, right? Is we wear both of those hats. And so, you know, Mel, I know you tried some therapeutic

interventions. What do you think? How are we going to help these kids in the future? How can we help them now?

Melissa Hazen

Yeah, I mean, it's a good question. And it's something that parents always ask, which I'm sure they ask you as well, which is, okay, you know, if there's a vestibular impairment, there's a balanced parent, what can we do? And usually my first response to these parents, this is kind of outside of the research right now, is anything to really like gently challenge their balance, expose them to different, different balanced environments, different environments in general, activities, sports, really just getting them moving is super good for them. And it's only it's only going to benefit them. And so that's one thing that I like to counsel on is just really getting the children out there, using those other systems that are well intact to learn how to compensate for the impairment. But in terms of what we did for the research, so my final chapter of the PhD was this camp that we did. And we took a bunch of children and we ran a five-day camp looking at spatial abilities. So we had like visual spatial, audio spatial, and kind of this like dynamic balance activity. So we took a rocker board that moved like left to right, and then we turned it and it would move front and back and we got children to balance on it. For the visual spatial task, we had something called the Morris water maze, which they basically, they had this virtual pool. It was on a computer screen and there's this virtual pool and there were four different shapes in four of the cardinal coordinates. And there was a platform that appeared and then it would disappear and the children had to use their memory to basically navigate themselves in this pool to find that platform within a certain amount of time. And then for the audio spatial task, we had something called audio lateralization, where they wore headphones and they heard beeps coming from the left or the right, and they just basically used a mouse to indicate, you know, did they hear it from the left or did they hear it from the right?

Karen Gordon

So I'm just going to jump in and I'll tell you why I think those are important, because there are lots of tasks there that, these kids are at camp, like why did they want to come in and do any of this? But the thinking was that, okay, so you may not have balance or vestibular system, you may not hear that well, but these are things that you can do. You got vision, You've got proprioception, you've got your muscles working for you. You've got some degree of spatial hearing if you've got hearing in both ears through a hearing aid on one side and an implant on the other or two implants. So like, let's use what you have. And these were these tasks that I don't know, they looked kind of fun to me. We did, Melissa went on, I don't know, I'm not even going to say where, but online and found this rocker board. And I

thought it was a beautiful idea because, you know, unlike a treadmill that we did with Rebecca, where we had to set it all up and we had to put a harness, like the rocker board could only go in one, plane either left, right, or we could turn it and do it front back. So I felt like, okay, that's A, it's fun to try to balance on that. And B, that's not, that's pretty safe relative to that treadmill. So I really love those tasks. And the water maze is a video game. I know you worked so hard at camp. I think you may want to tell them, like, why did we even do it at a summer camp?

Melissa Hazen

We wanted to get them outside of the the lab, really, we wanted to mimic like a playground and real life exposure. I know, camp is not necessarily, your every single day, day-to-day, but maybe if you think about recess or the playground, it's participating in activities, it's going out, it's having fun. And we wanted to give these children, an opportunity to one, connect with other peers who also have like hearing loss and cochlear implants, and two, to integrate into these other communities and have a camp experience. And so, that's why we chose the camp setting. It was fun for the children. I mean, they all had a good time. So, most of the day they were participating in just like regular camp activities. And then what we would do is we'd pull them three times a day just to do one, each one of those tasks randomized, but then they did it over 5 days. And we were really just seeing like, what was their capacity to learn in these different spatial domains? Could they learn to improve their ability or, had they hit ceiling and there was no more learning to be had? And so, that was kind of the overall like rationale behind the camp and why we did a camp and over the five days.

Karen Gordon

Yeah, and they could learn. That to me was like the beautiful part. Like, you know, everybody got better. You know, those who had hearing loss, those who didn't, those who had vestibular problems along with their hearing loss, those who didn't, everybody had fun. And, you know, I think we should give a shout out to the camp too, which was Bayview Glen, which really supported this by having us in their space. And we were so happy to see the kids enjoying themselves over that that week.

Sharon Cushing

And I know you have a hard to say, Mal, but I just want to back it up one little bit to emphasize why we need to do this. Because, we can make a diagnosis accurately of vestibular impairment, but then the next two questions that always get asked by families, funders, other scientists, other clinicians is, what am I going to do about it? And what's the evidence for doing that? And, I think you gave a really good synopsis. It's what I say to

families too, in terms of, getting some gray hair and promoting, balance related activities as parents. And we know that works, but there's not a ton of evidence in the literature, right? And so what we're trying to figure out both is the why and how, but to create that evidence base that's going to have some clout in terms of getting rehabilitation funded and those aspects of things. So it is a big task, that you took on in terms of, trying to figure out better answers to those two questions.

Melissa Hazen

Yeah, Sharon, you raise a good point, which is the why, right? And so the first step the reason why we did this camp is really just to see their capacity to learn. And now that we know that they can learn, that's kind of where the research should go in terms of like rehabilitation. And okay, now that we know that they can improve in these different domains, how can we train that to, you know, get better and generalize and like be sustained over time? Yeah, I mean, with the camp, it was pretty remarkable, especially, with the water maze in the visual spatial domain, they improved to the level of typically developing peers. So by the end, by day five, they were at a level that was the same as a typically developing child, which was so cool to see. Yeah, and I mean, even with the audio spatial and then even the rocker board, they improved. Not as much. There was still there was still a little bit of a gap between the children with hearing loss and the typically developing children, but overall they did improve, which is a good sign. It's it means, you know, we can we can take that further.

Melissa Hazen

Each kid had a different favorite game. Each kid had a different favorite game. And I think it actually depended on whether or not, for example, with the water maze, whether or not they played video games at home, right? Or with the rocker board, whether or not they did other kinds of activities. Like I know for some of the younger children who were in gymnastics, they really liked the rocker board. Yeah, it was very interesting. Each child performed a little bit differently on each task, which I mean, goes into a whole other realm of individual, individualized supports and, each child has individual needs. But for some of the children who I knew had some underlying vestibular impairment, they really had a hard time initially on those boards. But over time and over those days, they developed some kind of compensation strategies. And I wish, you know, if I could go back, I wish I would have asked more about, what they felt or what they were doing. Because for some of those children who would fall off initially at the beginning trials, they were able to stay on the board. They wiggled a lot, but they were able to stay on, which was entirely different from the beginning where they would fall every single trial, right? And so they did it. They figured out how to do it, which is, it's a special thing.

Sharon Cushing

It reminds me again of having, one of the two boys on the balance board that we talked about. And, patients and research participants will say these things, right, that inform your questions and your hypotheses, with such remarkable clarity. And, I remember we had them on, we were making it hard, right? We had them on foam and, and we were giving him a little bit of extra information, through his cochlear implants. And after he got off, he said, mama, I didn't know that when I was leaning to the left, in order to not fall, I needed to shift my weight to the right. And he didn't quite use those words exactly, but it was really like, wow, have you ever thought of that? Like, last time you were on the subway, did you think about what you had to do biomechanically to stay upright? And I think that that's what these, you know, learning experiences do, right? Is it provides exposure to what individuals who are born with a functioning vestibular system, like learn how to do instinctively, is that they have to learn how to do it non-instinctively, deliberately, consciously. And I think that's where we can come in with therapy is speeding up that process because, you know, they'll... probably eventually figure it out if they ride the subway enough, right? But I think we can make that easier by backing up and say, what are the things I take for granted in how I navigate my day? And let's deliberately talk about those things, much in the way that we do with auditory verbal therapy or therapy after cochlear implantation.

Maria Khan

I think it's very important for parents and families to understand that if your kiddo has like a vestibular deficit in hearing loss, that work can be done to help them out. Because with hearing loss, like, oh, you can put a hearing aid on, they can get therapy done, hurrah. But they may be a little bit more lost on the vestibular side of things. It's like, oh, okay, they're wobbly, they have balance problems, like now what? And I think what y'all just said is very helpful because there may be parents who haven't gone in and seen like a professional yet for their kiddo, but they're starting to notice like they're not walking in a straight line as well. They're a little wobbly when they're trying to sit and stand. So I think it can also give parents information, like empower them to be like, okay, like, you know, they're not stuck sitting. They're like, what am I supposed to do from here on out?

Sharon Cushing

And I think it also gives a label to what they're seeing, right? Because, these children who have congenital, vestibular, and hearing loss, they're going to walk later, right? They're going to figure it out, but they're going to walk later. It's going to be between 18 and 24 months instead of being between, you know, 9 and 15 months, right? And when you're the parent that's sitting there at, you know, the parent and play group and your kids, the one kid

that's like not moving, like you're worried, right? And then you go see, your family doctor or pediatrician and, they do a beautiful job of, monitoring these kids, but it's a little bit of a wait and see, right? Wait, let's wait and see for it not to develop because the developmental spectrum is so huge. And so families don't know that balance comes from the inner ear, right? And so again, it's like, one of my favorite moments when I'm sitting in clinic and I help families make that connection and they say, so you mean to tell me that this is all related to the hearing loss? And there they thought their child had two separate issues, completely distinct, right? But these are true, true and they're related, right? And so you can collapse that down into, this is a singular issue and we're going to come at it from different ways, but my child doesn't have many things different about them. And I think there's solace in that and in giving it a label because they're seeing it. And I think that part is so, so important to help families come, you know, to that recognition together. And I think that part often, often gets missed.

Maria Khan

And that's what we're hoping to do with these episodes. In our previous season, we had an about hearing fatigue and for parents to kiddos with hearing loss, they might not be aware that, oh, my kiddo may get tired of wearing their CI all day long or their hearing aid all day long. And I feel like this episode also provides another perspective in terms of the distributive side of things.

Sharon Cushing

I recommended season 4 to two parents in clinic today. I'm like, you have to listen to this. And they, you know, they were kids of, or they were parents of young children, right? And I think that it's the best provision of a crystal ball that we can give families is to speak out loud about experiences. So I expect a bump in the listening.

Karen Gordon

Yeah, we all learn together. I think that's what we're trying to show with this podcast is how important it is for us to meld our research with our clinical experiences with families' experiences, because that is the community that we have built, and that is how we're gonna go forward to supporting those children who we haven't even met yet.

Sharon Cushing

I'd like to look to the future. And that so, I think that there's lots more to be done in terms of the rehabilitation component. And we're going to continue to like chip away as to how do we best learn this and incorporate it into, you know, therapies that these kids are already getting, because we don't want to add more to families to do lists, right? The other exciting

part is, we're in the realm of genetic therapy, right? And while genetic therapy is being designed for hearing as the major output, we are a group that's going to continue to be very curious about the impacts on the vestibular system, both positive and negative, just like we did before. And so, you know, I think that, you know, that's more of what you might see come from the lab as well, you know, on those fronts. Again, there are vestibular implants. It's not the same problem as cochlear implantation, and it's not the same solution. So, when and if we'll see that application in children, maybe. Maybe not in my career, but maybe. So I do think, that, we're looking ahead to very exciting things, but having this foundation and this, such motivated patient population that is just so willing to give of their time and of themselves to help us answer these questions, allows us to continue to advance. But, I'm excited for what we're going to learn together in the future.

[MUSIC]

Karen Gordon

Thank you for listening to our award-winning Hear Hear Podcast. Thank you again to children and their families who have spent their time and energy with us in Archie's Cochlear Implant Laboratory. Thank you also to our Hear, Hear podcast team and a special shout out to Dr. Blake Papsin who composed and performed our Hear, Hear podcast theme music.

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