Transcript

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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]

May Wang

Hi everyone, my name is May Wang. For those of you who have listened to season four, you might recognize my voice. I was born with single-sided deafness in my right ear and joined the Hear, Hear podcast team as a lab ambassador at the start of season three with the intention to raise more awareness around hearing loss and support children with similar lived experiences. So it's an absolute honor to be your host for today.

In this episode, we will be focusing the conversation on single-sided deafness, or SSD, with the goal of providing our fellow listeners a greater perspective on what it's like to live with SSD, what sorts of treatments, specifically cochlear implants, are available for those living with SSD. And we'll also get to learn from our guest experts about the latest SSD research that is being conducted both in our very own lab and in the field. And with that, it is my pleasure to begin by welcoming our guests, Dr. Sharon Cushing, Dr. Karen Gordon, Harrison Gao, and Hanne Bartel. and all of whom are strongly involved in the research done here at the Archie's Lab for cochlear implants at the Hospital for Sick Children and have played central roles in the development of the Hear, Hear podcast with both their expertise in hearing loss research and support for the cause.

Dr. Sharon Cushing is an Otolaryngologist at the Hospital for Sick Children, Director of the Cochlear Implant Program and Associate Professor at the University of Toronto. And fun fact, Dr. Cushing was also my doctor at Sick Kids for 10 years, so I'm very excited to see her again. And I couldn't be more grateful for the care that she has provided for me and my family. Dr. Cushing was also featured in our podcast as a speaker in the first two seasons, so be sure to check those out if you haven't already. Hanne Bartels, who joined us in our latest season, season four, is currently a PhD candidate at the University of Toronto. She's actively doing research on single-sided deafness, which makes her a perfect guest speaker for our episode topic today. And Harrison Gao, who is a new guest of our podcast and a medical student at the University of Toronto. And of course, we have our very own Dr. Karen Gordon, who will be joining us as a guest today instead of a host. Dr. Gordon is a senior scientist and director of research of the Archie's Lab for cochlear implants. She is a professor in the Department of Otolaryngology and a graduate faculty member in the Institute of Medical Science at the University of Toronto. And as you all know, Dr. Gordon has been the central driving force behind the Hear, Hear podcast production and We're all so grateful for everything that she has devoted into raising awareness around hearing loss and supporting this community. Would you all like to greet the audience before we begin with the formal discussion?

Karen Gordon

Hi, May.

Sharon Cushing

Hi May, it's so nice to see you again.

Hanne Bartels

Yeah, hi May, it's nice to see you again. I'm very excited about this and thank you for inviting me.

Harrison Gao

Hey May, it's great to meet you and hey everybody in the audience. Really grateful for a chance to be on the podcast and to have this great discussion with everybody.

May Wang

I guess I'd like to first start off this conversation by providing some background context for listeners that maybe aren't very familiar with what single-sided deafness is. And as the name implies, single-sided deafness is a condition where you have normal hearing in one ear and severe to profound hearing loss in the other year. And as an example, I was

diagnosed with SSD due to cochlear nerve aplasia, which means the deafness was caused by the absence of a cochlear nerve in my deaf ear. So This was the etiology in my case, but perhaps one of you could provide some more insight as to how common SSD is in general and what are the most common causes of SSD.

Sharon Cushing

Sure, I can. I did clinic today and I think I looked after three or four patients with single sided deafness. And I think, you know, in terms of the overall Prevalence. I'm not sure. Karen, I don't know if you can help me with that, but there are some very specific causes. And the cause you have, May, is by far in a way the most common. Anywhere from like 35 to 50% of children who have one ear that hears perfectly and another ear that hears very poorly will actually have absence of a cochlear nerve. And there's so much we don't understand about why that happens. And I hope that we learn more because it's a little bit of a black box and it is super, super common. The other common causes of single-sided deafness are from a virus that you can get, and it's a very common virus. It's called cytomegalovirus, or CMV for short. And how it spreads is that a pregnant mom can get exposed to this virus, and it only matters if you get exposed when you're in utero. That's when it can cause hearing loss. People get exposed to this virus all the time when their ex-utero are walking planet, and they might get a cold or things like that. But it can give you hearing loss, and it does often create unilateral hearing loss and progressive hearing loss. So that's what I would suggest is the second most common cause. We do have a group of children who have a nerve, who haven't been exposed to the virus. And some of those may have abnormal anatomy on one side, enlargements of certain part of the cochlea or the inner ear that make it susceptible to hearing loss. Things like it's called an enlarged vestibular aqueduct. You can sort of think of it as like the fluid highway into the ear. is a little bigger and can create hearing loss. And there are some genetic causes of single-sided hearing loss, and we're learning more and more about that. We used to think that it couldn't be genetic if it was unilateral and single-sided, but now we're learning that that's not actually the case anymore. So that's a little synopsis. I feel like it's less than 10%. What do you think, Karen?

Karen Gordon

You know, our rates of hearing loss in children are about one to maybe 3 in 1000. And so a good proportion of that is our children with single-sided hearing loss. We are finding that many children in that group do have this severe to profound loss, like you said, on the one side. And it's a group that's special because we might not have known about them right away before. So hearing screening programs for infants, newborns have really helped us

identify these babies with this significant loss on one side. And this is where we're at, trying to understand what we can do to help.

May Wang

I think this session provides the perfect opportunity to bring more attention to SSD, which I think isn't as well understood as other forms of hearing loss for a few reasons. You know, one, there's probably fewer of us with congenital SSD. So maybe we represent a smaller portion of our community relative to other types of hearing loss. And two, like you said, Dr. Gordon, it's often harder to recognize that someone has SSD because we can still hear without any assistive hearing devices. So it's not uncommon for children with SSD to be diagnosed at a later age, or at least not right away from Earth. In my case, we only started noticing signs of hearing difficulties in my right ear when I was perhaps around three or four years old, and then I got diagnosed with hearing loss shortly after, and then I became a SickKids patient later on in my childhood. Because we can still hear in one ear, I think many people might have this misconception that the hearing loss in people with SSD have less impact on childhood development and quality of life in general. But reality is quite the contrary. In fact, Quite a few studies have shown that children with SSD actually experience challenges in a variety of aspects of life that are more similar to those with bilateral hearing loss compared to their normal hearing peers. And as someone with lived experience of SSD, I can say that, you know, there are definitely additional barriers posed by my hearing loss I had to work around. But I'm wondering, as experts in my field, What are some of the major effects and unique consequences that SSD has on children who are born with it? And are there particular learning patterns and hearing challenges that are consistently observed in SSD patients?

Karen Gordon

May, you said it so well, and that was my experience as a young audiologist too, was, lots of comments from other professionals, like, this child has one good hearing ear, that's enough, and they're going to be fine. And I think what we have learned, as you said, from lots of different groups over the last couple of decades, is that There are significant challenges that children with hearing from only one ear have. And we've been really interested in this because, when we were thinking about treatment for children who have significant loss in two ears, we started with one cochlear implant. And there were lots of reasons for that because we wanted to be careful about that surgical intervention because it is, you do have to have a surgery to put the cochlear implant in.

So we were only doing it in one ear and we started to see that maybe we might be able to actually make things a little better if we actually put it in two ears. So what's the problem

that would come from listening from one ear alone, whether it's a cochlear implant, or one normal hearing ear is really the lost use of those two ears, which are so important for sound localization. So I always talk about this as we think a lot about what is sound, what is the meaning of sound and the language that we get through sound and all of that. But there's also the where part of sound, where is it happening? And that you really do need two ears for. And I think we've been very focused on that in our lab because very few people have been focused on the where pathways. And we think that is a huge contributor to hearing, particularly in children. And your where pathways are going to tell you, well, that sound is in one location that's different from another sound. And then you can start to use your ability to attend to one thing by where it is in space rather than ignored something else.

So when it's all coming in and you really can't dissociate where it's coming from in space, that's harder. So listening in noise becomes a problem. So when you can't localize sound and listening in noise becomes a problem, well, what else deteriorates over time is, you know, all of the other things that you should be doing with your attention. and you're actually concentrating on trying to figure out what and where sound is. And we'd rather kids be focused on, you know, growing, learning language and doing all the other things that they should do with their peers.

Sharon Cushing

I think back to the clinic visits I had today, and it's a really interesting thing to navigate because we do talk out of both sides out of our mouth, 'cause there's gonna be lots of things that baby's gonna do well, and there's gonna be lots of things that are difficult and you, you know what? When I think about how I try and project that to families, I'm trying to give a little bit of both so that families understand that when intervention is available or feasible, that it's a reasonable thing to do or an important thing to do, but also to give them the hope and the crystal ball. And what I love about your contribution to season four and all of the individuals who contributed to season four are you guys are the crystal ball that I need in my clinic. Because these families are sitting there and they're grieving. They are, you know, they just heard this news that they never expected. And, you know, their brains might say, okay, well, at least it's only one ear, but their hearts certainly don't. And what they need in that moment is to hear what you shared in season four, which is that you are an amazing, successful human, but that there are challenges to your day as a result of this. And I think that's the conversation that can be really... hard to get the right balance with.

The other part that resonated with me was this idea that it's not bilateral hearing loss, it's not visible or it doesn't affect. And I say this to families because they'll often come in and say, well, like my mother-in-law thinks the baby hears fine, right? That kind of thing. And

the truth is, what we observe in others' hearing is detection, right? Especially in babies, right? I make a sound, the baby turns their head. And so I'll have that conversation with family and say that, of course, your baby actually doesn't look like they're behaving any differently because the nuance. of hearing with two ears is very difficult to detect in visible behaviors. I'm sure if we thought about it hard enough, and Hanne and Harrison are going to take us through what the science and the data shows, but I think it's really important for people to recognize that their baby is going to look like they hear. And in many ways, sometimes even babies with bilateral hearing loss look like they hear, because as humans, we're so driven to connect. I think their desire for this baby not to have hearing loss also colors that lens with which we observe children. And so it's a really complex dynamic to have these relationships with families. I have not perfected, you know, how to best communicate that double story, right? That there's lots of things they're going to do well and there's lots of reasons to be hopeful, but there's going to be some stuff we need to work on.

Karen Gordon

Yeah, I think, that's how we became interested in this is really from the point of view of children who were only listening on one side. And we wondered, how this would have affected different parts of development and academics and so forth. It's interesting that we often do as part of psychoeducational assessments, tasks which involve space. So, manipulating visual objects in space or remembering where things are in space. It's part of our ability to think about proportions and math and all these kinds of things. And it is interesting when you start probing and kind of pushing, pushing a little that There are these differences in children with hearing loss, both with two ears as well as one ear, in some of these spatial tasks. And that, if you ask children with single-sided deafness how they feel about their hearing, can vary quite a lot. But there is this higher risk for feeling tired, as you talked about last season. And there are a number of different places that have also studied that, interestingly, the listening fatigue is real and it can be measured. And it can be missed too. It might be hard, for example, for teachers to see this in their students amidst all the other students, right? And how much effort they're putting into tasks and other things that require spatial perception.

May Wang

So a lot of these hearing difficulties are very subtle early in age. But I'm also curious, as you know, the child grows up, do we see more obvious differences in their behaviors? So

spatial awareness is one aspect, but maybe there's no sense of direction or imbalance caused by this. oral preference towards their hearing ear? Or are there also difficulties with, let's say, balance or even challenges with learning language?

Karen Gordon

Okay, well, I'm going to leave the balance question to Dr. Christian because that's her expertise and it's a super important question. But I want to just talk a second about balance between the two ears. And I think you raise an amazing point, which is we've studied a lot of how the brain changes. And Hanne was going to talk a little bit more about our current studies looking at that. But we, you know, as I said before, we were starting to get interested in how hearing from only one cochlear implant was changing the brain. And what we found is that it's not surprising that one side that was getting input with a cochlear implant was becoming really, really strong. And the other side, which hadn't got much hearing and wasn't implanted, wasn't getting any input on that side, was really, you know, relatively weak. And it was hard when we provided an implant later on that weaker side if we left it too late in development. And so we find very similar things. in children with SSD that there's this preference for the hearing ear. And maybe we can just talk about that a little bit because we see this change in the brain. What that means functionally is a difference also in the ability to hear speech. Hanne, maybe you can talk a little bit about speech perception with one hearing ear. We can talk about the brain changes maybe a little bit.

Hanne Bartels

Yeah, I'm happy to. I can start with the speech perception. So when all this research was being done in different labs on children with single-sided deafness, so one of the main things, as has been beautifully discussed here before, is that these children don't have spatial hearing, which makes things like speech understanding in noise very hard. Think of a classroom where there's noise coming from multiple directions or a playground. to be able to help these children. We want to try to give them hearing in both ears, so maybe try to get that binaural hearing again, or at least bilateral hearing. So there's many studies on that, on how we can improve speech perception in these children.

But there has been also a lot of other groups or critiques saying, okay, but we have one normal hearing ear and one cochlear implant if we give these children a cochlear implant. So there might still be some asymmetry left. Whereas if you look at children who have asymmetric hearing loss, where they have some hearing loss in their good ear, then there might be less asymmetry compared to the CI. So the first study that I did as part of my PhD, here we looked at all of the speech perception scores in clinic from all of the children who

have a unilateral cochlear implant. So we didn't just look at the ones with single-sided deafness, but also at bimodal device users. So what that means is that these children have a cochlear implant in one ear and then a traditional hearing aid in the other ear. So this is them for a mild to moderately severe hearing loss.

Or in some cases, these children had a moderately severe to profound hearing loss, but still a hearing aid instead of a cochlear implant. So we had these three groups and then we did this study to see how these children perceive speech in one ear, whether this is the acoustic hearing ear or the CI ear or in both ears at the same time. So when you are able to use both. So then we are able to turn the CI on. And what we found then is that yes, in quiet, children who have single-sighted deafness, so they have normal hearing in their good ear, they have better speech in their normal hearing ear than they have in their CI. And they have better speech perception scores as well than the bimodal device users in their normal hearing ear. With the CI, however, all three groups did very well and there was no difference between them. So the children with SSD can use their CI as well as the other groups. But if you didn't go to noise, This is where it gets interesting because now we see that the children with single-sided deafness are indeed struggling. So with their normal hearing ear, they got significantly worse scores. So in noise, it became harder. And then we see that there wasn't that much of a difference anymore with the children who used that device for a mild to moderate severe hearing loss. Whereas the CI, on the other hand, these scores, the speech perception in that ear, remain stable. So This study that, so it's actually recently accepted now for a publication, but this study really beautifully shows that children with single-sighted deafness also have this benefit of the CI. Even though there is some asymmetry towards a normal hearing quiet, in noise, these children really have this benefit and it's helping them perceive speech better. So there was a speech paper that we recently conducted.

May Wang

So who would be able to qualify for receiving a cochlear implant if they have SSD?

Sharon Cushing

What a great question. And, there's part of me that wants to be cheeky in answering this and say, I don't know, actually, and there's, it's very nuanced. And, when we began, we had one set of criteria. And what I'll say is that the criteria are highly variable. So I'll tell you about what we think about here at our program in SickKids. And it doesn't mean that it's, the best way to do it or the only way to do it. Because there are many other centers who are looking at it differently. And I don't know that any of us have truly figured it out. So again, if depending on where listeners are coming from, I don't want you to take this as gospel

because there is different approaches. And any time there's different approaches, it tells you that there's not perfection anywhere, right? The stuff we know how to do perfectly tends to get done the same way everywhere.

The stuff where there's still tons of questions as to, you know, when is it good, who's going to wear it, all of those things, that's when there's variable approaches. So I think that's how you can look at the variability you might see in different programs, the variability you'll certainly read about on the internet. So for us, if we start with the basics, if we think about anatomy, you need to have a nerve. So in our program, and I would say most programs, you need to have a nerve in order to get a cochlear implant for single-sided deafness. The second part relates to the duration of deafness. And I hopefully, I'll give you the surgeon's version of Karen's amazing data. But essentially, we wanted to try and set ourselves up to succeed as best we could. And we felt like based on what we knew, that leaving the brain too long in silence was going to cause irreversible changes that would potentially place limits on the benefits people could get from a cochlear implant.

So certainly early on and still to this day, we like to keep that duration of deafness pretty short. Now, people could talk for hours about what duration of deafness is appropriate for single-sided deafness. In our program, it is typically less than four years, not of age, but of duration of deafness. And again, there's many people who do it differently. This is not the gospel. And When we started, it was only two years of age. And so we expanded that a little bit. What I'd really love to know and be able to predict is who is going to continue to wear it. Because again, we're going to hear about the data, but, and Harrison's going to tell us about who wears it and how often, but if you wear it, will work. The brain will figure it out. I think that's a pretty safe assumption. But you have to wear it. And what we've been incapable of predicting is who is going to wear it. And we do a lot of things. We sit with families. We sit with patients, especially our older patients who have a voice of their own where they've lost their hearing and know what they're missing in order to try and figure that out. But that to me is the holy grail of single-sided deafness and cochlear implantation. And I hope that over my career, we'll be able to do a better job of having a predictive tool for who is actually going to wear it.

Now, the other part is, and then maybe I'll hand it to Harrison to talk about who wears it and who doesn't wear it, is that, you know, we're looking at a snapshot of a lifetime, right? And so that snapshot's getting longer and longer as we follow these kids. And some of them have been followed for over 10 years, right, Karen? in and around. And so what happens is that these kids develop. And so when they're babies, their parents put it on, when their children, children, school-aged children, their parents probably still put it on, but they have more and more capacity to take it off. When they're teenagers, they're in

charge, right, of putting it on. And what we don't know is the kids who stop wearing it or who never started wearing it, are they ever going to start wearing it when they see the value of trying to make it it work, whether that's vocational or family or some kind of incentive. And so this is a lifetime story as we watch it. And I think that, again, we have so much to learn about who's a good candidate. But maybe this is a good time, if it's okay, May, to have Harrison take it away and address that issue about what do we know about the kids who wear and don't wear their devices.

Harrison Gao

Yeah, so Dr. Cushing said it beautifully. And this is some of the work that I had the pleasure of leading during my time doing research with the CI Lab. And we also had the chance to present at the National Otolaryngology Conference in Canada. So it was great going to that conference with Dr. Cushing. And this is the study that I led. And Dr. Cushing, of course, gave many keynotes and great other sessions and other lectures on the topic. So this is one small piece of it. But what I studied with the lab was just looking at usage patterns for children with SSD and who have bimodal devices. So that was a hearing aid and a cochlear implant. So what we really saw was in this specific cohort, so these were children who had cochlear implants and who had really good hearing in their other ear. So these were the children that had such good hearing that they didn't need a hearing aid. So that was the SSD group. And what we saw in their group was that. The ones who experienced the greatest declines in device usage were the ones who were actually implanted later. So these were children who were implanted at a mean age of around 8 years old versus children who were implanted around 2 years old. So these were children who met those qualification criteria or candidature criteria. So they had a nerve, they had a low enough duration of deafness. But perhaps it was a result of getting this device kind of at a school age. But they eventually, over time, would start using their device a little bit less than children who were implanted at a younger age. So that was, I think, one of the more interesting pieces of data there. And I'll let Dr. Gordon and Dr. Cushing chime in, too, about this.

Karen Gordon

Yeah, I think, you know, When we started providing cochlear implants to children with one normal hearing ear, we kind of had to pinch ourselves a little because I think if you had told me when I first started in cochlear implants that we would be giving a cochlear implant to a child with normal hearing on one side, I don't think I would have believed it. was really this understanding of how you know, how many challenges some of these children were experiencing that kind of led us to think about it. And then a lot of the data that we moved, that we were hearing from adults with single-sided deafness was helping us out here to

move forward. And those adults, typically they had hearing for, as they were children, both ears, and they had lost hearing on one side, later in life, and they really missed it. And sometimes they had a lot of noise, so tinnitus, internal noise that really bothered them.

And those adults really found, or were reporting through these studies, some really beautiful benefits of having a cochlear implant. And they were using that with normal hearing. So that's how we kind of got started. And we had, we were cautiously optimistic, I'll say. So I think that all those criteria that Dr. Cushing was talking about, we're really just trying to be careful at the very beginning to just try to give children the best opportunities to use the devices and see what we could get with with development. And I think if we just think about the results that Hanne just talked about in terms of speech perception, like we couldn't be happier from the point of view of how much they're hearing language, spoken language, and understanding spoken language with those devices. It's definitely true, as you've heard before, that that's going to get better the more you use the device. And that's where we got into worries with, what Harrison was talking about is this concern that, well, if there's so much benefit with that cochlear implant, why aren't they using it more? And that's what we're really trying to understand.

We're trying to listen to the children and to the data that we're finding is to understand what are the challenges that they're having that we can't really see. that are telling them not to use it so much. Maybe the asymmetry between their ears, for example, that we're thinking about. And we may have to be, as Dr. Cushing said, very careful about, you know, who we provide these implants to upfront.

Sharon Cushing

I want to get back to one thing that you asked. I think that, you know, fits in here as well, and that was the relationship to balance. because I think that, Karen talked about the wear of sound, right? And if, what was enough for. for human connection. people would read, or not read, they'd listen to this podcast, or maybe they would read it too, through captioning, and they would feel as though they were with us, right? And here we are, we're virtually talking about it. And so we don't feel each other's presence in the room. But Harrison, you know, when we were at that Canadian ENT meeting and we all went out for dinner, that is where the what and the where mattered. And for the wear, that provides that human connection, I think balance is part of that in terms of your spatial surround and the physical component of being in place with other humans, which is such a joyful thing to be. And one of the things that we found early on is that just like in children with bilateral hearing loss, vestibular impairment or the inner ear component related to balance can also be impaired in about 50% of kids. And that can be in children who don't have a nerve. Sometimes they also don't have a nerve that goes to their balance system.

And particularly in our children with the virus, the CMV virus, because that really has a propensity for all parts of the inner ear. And so I think in those kids, there's an amplified impact. And, you know, as we think towards candidacy, you know, there's always been part of me that wonders, oh, perhaps they're better candidates because there's more to help with would sort of be the philosophy there. I don't know that we've had numbers that have allowed us to verify or had this born out, but I do think it's an important thing to continue to look at because so many, you know, hearing is so much more important to balance than we ever recognized. And asymmetric hearing, I think, can do a number on where you feel like you are in space to then have the secondary inner ear issue with the balance part just compounds that. So I think, again, there's a little bit of something there that we need to continue to examine.

Karen Gordon

I think there's a lot of interest in all of this, all, you know, in many parts of the world. So you can see that we are collaborating and thinking about this with lots of different groups. So we, we've put together a whole bunch of different studies that will come out soon in Aaron Hearing. It's a journal that just tries to get a collection of ideas and what we're all thinking about and experiencing. So a couple of the other groups have had successes in different ways that we're learning from. So one of them is a group in Leuven who is in Belgium. who were really, really careful about their candidacy criteria. And so some of the children received cochlear implants at very young ages, and they were followed really, really carefully with very careful therapy. And they're finding that those children are using their devices more with some nice improvements in some language areas. And so, you know, what makes the motivation of those children a little different than the children that we're seeing is an important question. question. And another part is probably around what we're doing with the devices themselves. So how do we properly program those devices? We haven't really figured that out yet. And there are some groups who are really interested in trying to provide the right sounds in the right places in the ear that might match the other ear a little bit better.

So we're learning from them as they do their own studies in parallel. And also learning about therapy itself. maybe these children need to have different kinds of supports than how we were thinking and providing for children who have, maybe bilateral hearing loss, for example. So, whereas We were really trying to make everything quiet for a child who only has cochlear implants to use because it's really, you know, cochlear implants are not normal hearing. They're not even close, right? So they are, they do amazing things. But, you know, we really try to give really good language input by being in quiet and speaking to children with lots of intention. It may, we may need to do things a little differently for

children who have single-sided deafness with or without a cochlear implant. So we're examining things like, for older children, maybe more streaming through their devices that would encourage them to use the device more and to get that understanding of the recognition that they have through that device alone, because otherwise they're really not using it by itself. And also, what are you doing in noise? So if you are in a group, how is it, how is the implant able to help you? And so just using, even for little, little ones where if you have them playing in a group, having them explore what that space is like and listening in that space with them without the implant.

PART 2 POTENTIALLY

May Wang

It sounds like there's a lot of factors that need to be considered when trying to think about, you know, what's the most appropriate treatment for the patient and for the child, which makes, you know, every person's lived experience with SSD unique as well. And I'm also curious, and I'm sure a lot of families, when considering, you know, what they should do for their child would also be wondering, you know, receiving cochlear implantation isn't as straightforward as, you know, just putting something on and then taking it off. Right? So in what circumstances would cochlear implants be recommended to someone with SSD when technically they're still able to hear without it.

Sharon Cushing

You're absolutely right. And, I get the, I have the privilege of having this conversation with families and it is really tough. right, to try and make this decision. It is tougher to think about putting your child through surgery for a cochlear implant for single-sided deafness than it is for a child with bilateral deafness, right? That's still hard and it's an immense gift of trust to place your baby in someone's hands for surgery. But there, if you know they're not going to develop speech and language, then it's a must. do for many families, right? But when, it's a can do when it comes to single-sided deafness. It's not a must do. And I think that can be a really difficult thing for families to navigate. Because, if you said to them, do you want your baby to hear on both sides? Absolutely, right? Like that's absolutely the answer, but at what cost, right? And that's where I think individual variability is going to come into play because that ear is attached to a whole human and that human is attached to a whole family. And there'll be an immense amount of variability within that human and within that family. And risk tolerance will be different as we go through things. And so I think there's some families that, they are very clear, absolutely not. And then there's the other extreme where they're like, yeah, absolutely, let's do this. But I think most families sit in that, I'll call it purgatory, because it is super, super tough to make this decision. All you

want to do is do what's right for your child. And in that moment, many of them, especially the babies, don't have a voice.

They can't help you as a parent. And you think, oh gosh, what if I make this decision and they hate me for it? What if I make this decision and it's the wrong decision? What if I make, I don't make this decision and they wished I would have and now I've burnt a bridge? And so I think it can be super tough. for families to make these decisions. And so certainly if you have a nerve, if you've got a short duration of deafness and you're willing to accept, both the unknown and the risks, then we would offer it. But we do a lot of counseling around this just because it can be really tough to know what you're getting into. But, you know, I see, you know, this is really hard on families making these decisions.

Karen Gordon

The one thing I would also say, and this has been true throughout the history of cochlear implants, is that we're learning with families. So we have a real privilege to have a laboratory in our clinic with us where we really try to do that learning in a clear way. So the families who come into the lab and participate with us are huge part of that because actually, as Dr. Cushing always says, we don't know what we don't know. And there's lots that we didn't know at the beginning of cochlear implantation in general that we know now, lots that we still need to understand. And the same thing is really true in this group of children and their families. we are really trying to provide something that will be helpful. In what ways it's helpful, we're still learning.

In who it's helpful, we're still learning. And how we can support that is still what we're learning. And that's what we really want to do is support learning, right? And to reduce all that effort that you were talking about, I think it's really important for people to understand the effort that comes with hearing loss in childhood. And that's what we've started to really focus on. We know we can get language into children. We just need for them to be able to do that without all that huge amount of work that's being done. Is the cochlear implant the answer? Maybe a little bit. and maybe a little bit more for some children than others. But, that's really what this research is about. Is that little bit enough to make a difference? And how are we going to go forward on that? So maybe what, if you want to talk about some of the new things that we're thinking about, it's just some interesting things about how it might be to listen with one side and how the brain responds to that.
So the effort we can see that the brain has these differences when you're really trying to listen versus when it's not. So we can see these kind of differences in places in the brain that light up or that are become active when we measure them with all those electrodes we put on the head. And I can tell you, there's so many children who've come into the lab and families who are listening will know some children call me the cap lady because that's, I'm

the one who puts that cap of electrodes and makes their hair all goopy. And Hanne knows this very well because she's doing the same thing with children who she's seeing in the lab right now. So we're trying to figure out, you know, what kinds of listening is more effortful, what kinds of listening is better for children who have a left-sided hearing versus a right-sided hearing?

Hanne Bartels

Yes, I think I can comment on that because that is actually what my PhD focuses on. So what is the effect of having a left-sided persistent right-sided deafness on the brain and might that have an effect on specific hearing skills? Now, when I tell that to people, a lot of them ask me, like, why would you even think there is a difference? Why might a left ear have a different effect than a right ear? So a lot of this is based on something that's called a right ear advantage. So there's a lot of studies from different labs all over the world that have shown that normal hearing, we have a right ear advantage for speech.

So we understand speech a little bit better with our right ear than we do with our left. And what we also know is that in the brain, when you look at anatomy of the auditory system, the majority of the fibers will cross the midline, meaning that your right ear has the fastest autonomous direct connection to the left side of the brain. So the left side of the brain has a little bit of a faster connection to this right ear that can process speech a bit better. And then there's other research group who are looking at things like pitch perception or more like amusia and these kind of things. And they have then found that the left ear actually did a little bit better. And the left ear is then connected to the right side of the brain.

So this has led other research groups to look at, well, does that mean that they start with adults or children with late onset deafness, that those who have a deafness in the right ear have more trouble with speech. Those with deafness in the left ear have maybe more problems with music or like prosody in speech where you have to recognize emotion. This is also very pitch heavy. And there are some studies showing that this is indeed the case, but our question was, well, when a little baby is born, and it immediately already has this or very early on has this deafness in one ear, is there still this right ear advantage for speech or left ear, meaning that those who have a right ear deafness would have more trouble with speech, those who have left ear deafness would maybe have more problems with other things like speech perception or maybe more like music things. So that's a little what my PhD is focused on. Because it might be that this is something that they can learn to work with because they have this hearing loss already in the part of their life where they're still very plastic and the brain can still change and adjust. So maybe it doesn't matter to them whether it's a left or a right side deafness. So this is what we are trying to investigate now. So we have all these children coming in now.

So I'm currently in the part of my project where we are doing data collection. So I think right now we have 65 children tested. So these are children who have cochlear implants in either the left ear or the right ear or children who don't. So these are children like me who don't have a nerve, so they couldn't get a cochlear implant or for another reason didn't get an implant. Sometimes it's because it's diagnosed late or another reason. And then we have them do these all the kind of different things. So they come in for a full research day and I'm very grateful that all these parents and all these families are wanting to do that. because I know it is a long day. So they come in and in the morning we do all these kinds of different hearing games. And then instead of just looking at the speech perception in noise that we talked about before and that is done a lot in clinic as routine follow-up appointments, we want to look at a little bit more than that.

So we also want to look at music perception, we want to look at emotion perception, and we even have some memory tests. So we look at like verbal memory, visual working memory. We play some laser games, we call them, where they need to hear specific differences. Some are pitch-based, others are more rhythm-based. And so we do all this bunch of different tests. And in the afternoon, we let them come in. And then now I am the new cap lady. So I put this cap on their heads with 64 discs and we fill them all up with gel, which is not the most fun part. And it's actually easier for children who have never done it before when they were younger because they don't have any memories of it. But they sit in there, we put the gel in and then we look at their brain responses. And so we give them very specific sounds.

We have a very slow sound and we have a very fast sound. And then we look at what side of the brain this goes to. And then we look at whether this is different between our groups. And then the ultimate goal is to see, well, if we find a difference, does that also mean that there is a difference in those hearing tests? Is that correlated? Because maybe they are indeed different, but in real life, in the hearing skills, it doesn't really matter, the deafness versus left or right. And then if there is, might we be able to improve our treatment options this way? So that's what my PG centered on. In terms of results, it's a little bit too early to say. So that will be for next season. But this is what we're currently doing in our lab.

May Wang

I think that's really fascinating. And I think it's pretty clear that a lot of these learning skills can be impacted by hearing loss. And as Harrison also mentioned earlier, it seems like the age of receiving treatment also matters how effective a cochlear implant can be. So, and I think, you know, a lot of parents might be wondering about this too, but why is it important to receive treatments like cochlear implantation at an early age? And specifically for those

with SSD, What are the consequences of delaying treatment? Of course, assuming that there are effective treatment options available and they qualify.

Karen Gordon

Wow, that's such an important question, May. It's so important when, you know, as families are deciding, we're also telling them, you have to hurry up and decide. Time is ticking. And the reason for that is what Hanne was talking about, the brain, is so malleable. We call it plastic in early life and it needs so much as babies are remarkable in how much they're taking in and learning in such a short amount of time. They're really like data collectors. They're just. all of their senses, when they're intact, are there for them to take in the world and try to figure it out.

Language is such an important part of that. Within the first year, years of life, the ability of humans to develop language, and we do it very commonly through hearing, happens from day one, it might even happen really in utero as well as babies can hear it in utero. But as they start to equate the sounds that they're hearing with meaning, that all is happening in the days and months of initial life. And when you miss that, the brain has to do other things to compensate. It's going to use other senses. It's going to change and develop to take other information in, which is okay. But, you know, we need to know. We need to know what the baby's abilities are so that we can provide the right inputs in early life. Getting language in is so crucial that, you know, whatever the mode of language is, we want to provide that. So we need to know that children who have hearing loss, we need to know about them right away. And that's why we have universal newborn hearing programs, screening programs, so that we can, we know which babies have hearing loss and we can try to provide language in whatever mode the family would like, meaning, you know, sign language or visual language or spoken. oral auditory language.

Sharon Cushing

Something you said, May, you know, just about this idea of, there was two things that resonated with me. The fact that absolutely we know that single-sided deafness is hard, right? It's hard on development. It's hard on day-to-day life. And there's, you're right, there's absolutely no doubt about that. And you also use the words assuming effective treatment. And I think that's where we're at, is that we know that cochlear implants can fix some of those things that are on that list. Some of those things you live on a day-to-day basis, but it's not certain whether it can fix them all. It can't is the truth, but I think that's where we're, you know, we don't want to be too... quick to make an assumption that this is the list of problems that you have when you have single-sided deafness and cochlear implants will fix all of them.

And I think that's, again, where we're really looking at how much can they fix and is that balance versus burden worth it? And that's going to be a different balance in every family and every child, right? And having a way to equate that and to sort of have that crystal ball and predict the outcome is really, I think, where we're hopefully heading and all the beautiful work that Hanne and Harrison do under Dr. Gordon's guidance are hopefully really going to give us, you know, that component. I think this question about timing is so important. It's important to single-sided deafness. It's important to any kind of hearing loss. And it is so appropriately on the minds of families because we're also at a stage where we're starting to have other treatments, things like gene therapies, right? And not a day goes by where I don't have a family that says, like, should we save an ear? I heard about these things. should we create a single-sided asymmetric hearing loss for the reason of a hope of a treatment that will not require a device?

And I think that again, Karen says it so eloquently, but the bottom line in my mind is we can save an year, but the brain's not going to wait. And I think that really resonates with family. And I use that information that we got from data in the lab from our patients in order to help other families. And so if there's anybody who's participated in research that's listening, like thank you for that. Like the impact that you sitting in that booth with that cap on and all that gel in your hair, like you have helped a family in my clinic today and in my clinic next week. And so the impact you will never be able to fathom about all that time you spent doing all of the research. So we so appreciate it.

May Wang

For individuals like me who did not use any assistive hearing technologies due to just the limited effective options that were available for, you know, in my case, cochlear nerve aplasia, which wouldn't qualify for, you know, a cochlear implant. What are some consequences that could be anticipated for just having to heavily rely on one ear all the time? And given that this is all we got, is there anything we can do to protect and conserve the residual hearing we have in our normal hearing ear?

Sharon Cushing

I'll make two comments. And the first is that not choosing an implant or an assistive device doesn't mean we're going to do nothing, right? And so I think that there's lots of things that we can do by knowing a child has hearing loss in order to set them up to succeed in the classroom, at home. in terms of preferential seating, in terms of improving the signal to noise ratio, and all of those. And so, no family wants to feel like they're doing nothing. And so I certainly amplify that, not choosing an implant for those families who either can't or it doesn't feel like the right decision for their family. It's not that they're doing nothing, right?

When you say that, it makes people feel like bad parents. And so we do need to think about all of those other things that we can do academically and socially to make the best of the information you have in that super ear, as Karen calls it.

The second part is, again, another one of my favorite conversations to have in clinic. I had it again three times today. There was a lot of teenage eye rolling and a lot of parents in the background going like, yeah, tell them, tell them about their headphones. And so, my general rule is, this is not nagging, it's empowerment, right? Because the only people who can look after your body and your ears are yourself. So I'm going to give you some things that you can choose to do or not. And there's simple rules. Not too loud. And I think that one's a little bit of a no-brainer these days, especially with our fancy apps that tell us, you know, what our listening volume is and our listening time is. But the other one is not too long. And I give very specifics. I say for every hour of listening, regardless of what it is, you want to take a 20-minute break for your eyes and ears. My optometrist tells my son this as well. So I've added eyes just to make sure we get all the senses in there. So 20 minute break, right? And like you got to actually keep track.

You got to set a timer in order to be able to do that. And so that's super important. The other thing that I encourage my teenage patients and I'm going to show my age here is, I grew up and we had a Walkman and we like, you weren't allowed to bring it into the class. And we really didn't have a ton of exposure, but I worry about, this generation that has had sound exposure all the time. Many of them had, sound machines when they were babies, and then now they're growing up again with technology. And so I ask my teenage patients to think about Do they need to be listening to something right now? Because you see it, right? You probably see it in the libraries. You see it on the subway. People have their headphones in and they're listening. They're listening while they're doing their homework. They're listening on their bike ride, on their, everybody's listening to something, right? And I think there are times where we just need to think, do I, does this bring me joy? Is what I'm listening to, am I, you know, present with it? Or is it just habit? Because we all have a tank that we can fill up with listening exposure, right? And when that tank gets full, it's going to be problematic. And if you fill it up by the time you're 40, that's going to be really problematic in terms of age-related hearing loss, which we know is associated with dose, right? It's just like the sun. And so I have these conversations with our teenagers. I don't know if it changes anything. but hopefully at least gives them reason for pause either now or down the road.

Karen Gordon

So maybe listen to this podcast and then take a break, right?

May Wang

And actually, I remember you, Dr. Cushing, telling me the same rule a few years ago at our appointment. So that's something that I've been trying to follow as well. But whether someone qualifies for treatment or a cochlear implant is one thing, but And also, and as we mentioned earlier in our conversation, because of the challenges associated with just identifying that someone has, you know, unilateral hearing loss, you know, like being diagnosed and therefore treatment is often delayed because of that. And as individuals who are heavily invested in this field and committed to supporting this community based on your experiences and perspectives, What measures can we implement to detect SSD earlier so we can provide earlier diagnosis and prevent delays in providing treatment? Or better yet, are there ways to reduce the chances of being born with hearing loss?

Sharon Cushing

You know, I think when it comes to absence of a cochlear nerve, there's so, we have so much to learn in that regard in terms of whether or not there's anything that we can do to change the course. But with CMV, the virus I talked about, it is one of the biggest modifiable risk factors for hearing loss. And we currently do a pretty terrible job of reducing that. And so I think that we're better able to detect single-sided hearing loss, you know, because of newborn hearing screening. We also look for this virus universally in Ontario, which is one of the only jurisdictions that does that. It's pretty cool. We also look for a number of genetic risk factors in Ontario. So I think in Ontario we're pretty avant-garde, so to speak in terms of our doing our best to minimize that risk. But here's what we need to do better.

We need to educate moms and women who are of childbearing age because their actions while pregnant can change their risk of exposure to CMV. And we just don't do a good job of doing this. And so what are they? Essentially, the biggest risk factor is having a toddler at home when you're pregnant. Because what happens is you get the virus by being exposed to new virus in their saliva or in their urine. And so what you want to avoid is those two fluids in your toddler. And probably the most common ways pregnant women get exposed to those other than changing wet diapers, is finishing the chicken fingers on your kid's plate. I know with my son, if he's got leftovers on his plate, they are going to, I'm going to eat them. And that's something that you shouldn't do if you're pregnant. You don't want to drink out of their cup. If their pacifier falls on the floor, you know, moms will put it in their mouth to cleanse it so that their screaming baby can then have, you know, some form of comfort. And those are where women get exposures to virus anew or to viruses they haven't been exposed with. And so if we increase hand washing and avoid those things, avoid, kissing your kid on the lip, there's lots of ways to show affection to your baby.

toddler kisses are not that pleasant anyways. They're all slobbery. It's like kissing a dog, right? Anyways, so these are not hard things to do. But I think in the past, we've been a little patronizing about it that, oh, you know, women are not going to want to do those things. Oh, you bet they are. But we need to let them know. And so I think that is probably the single biggest modifiable risk factor for the reduction in single-sided deafness is letting women who are of childbearing age know about how to reduce their risk.

May Wang

The other thing that I think is really important to emphasize here as well is, like with all forms of clinical research, research is so fundamental when it comes to understanding patient and family needs and how to provide the most effective care for them and think it's really the research efforts, along with supportive staff, that really pioneer the progress in optimizing the patient experience. Having participated in several hearing loss studies myself, I know that hearing loss research, especially on SSD, can be particularly challenging. So my question is, as we wrap up, maybe you could all pitch in with your different experiences and backgrounds. What are some of the main challenges associated with conducting research on SSD?

Hanne Bartels

I think for conducting research, the biggest challenge I faced was getting people to come in. Because I think research can sound scary. Like you don't want to have a parent feel like their child is a guinea pig that I'm going to do some tests on. So a big challenge, that was one challenge. But I think it's important to explain what you're doing and why it's important. And I am very lucky that a lot of the people that I contacted were also very willing to come in. And even some of them even came from 3 hours, 4 hours drive away and they came just for my study.

So I want to give a shout out to all these people who came in from so far. But that's of course a big factor because a lot of people come to SickKids and they come from far away and for research you need to get them to come in. So that's the biggest struggle that we face. in terms of recruitment or getting our study going. And another thing is having children do everything. So they come in and we have this whole list of things we want to do with them. But of course, it really depends on the child, on how they are feeling. Are they scared of the gap? Because the gap is non-invasive. There is nothing bad about it. I've done it many times. I've even done it a lot of times on Harrison here. He was my guinea pig. But it looks scary. And not all children are going to be able to do it. And even though you really want that data, there are points that I say, no, we're not going to do this. You're scared. I

don't want to do anything you're uncomfortable with. So yeah, that's also a little bit of a disappointment because you don't have the data. But of course.

Harrison Gao

Absolutely, and I'll just add on as well, too, is that I think there's not a lot known out there, and I think Doctor Gordon and Doctor Cushing talked about this a little bit, but it's kind of a newer thing to provide implants for SSD and in a lot of countries and a lot of hospitals, even if they are doing... cochlear implants for children with SSD, they're very small cohorts. So when they're, when we do look at the research out there, a lot of it is pilot studies, a lot of it is, it's really, it's an early field and it's still developing. So I think that's one thing I really did. find to be an advantage and something I really enjoyed working with. It's just we have a lot of data at SickKids and there's a lot of children that they treat. It keeps them busy, definitely. But as a result, we're able to ask bigger questions and we're able to answer bigger questions too, because instead of maybe 5, 10, 20, 30 children in a study, we're able to have hundreds or or so. So I think that's something for sure that is a challenge with the research right now.

Sharon Cushing

I think I'll flip it. And I think sometimes there are unexpected benefits from people coming in. And I think of sort of our young our young adult patients where, they're at the stage of their lives where they're looking like for a career, right? And like, what am I going to do with my life? And I love to do science and I like clinical, but I'm not sure that I want to do medicine. And I think that, in our lab and you've got two great examples, well, three great examples here with Hanne, Harrison and May, you know, it exposes these young adults to options in terms of career paths. They can see themselves and the work that you do. And I think it provides an exposure. And I don't know that that's necessarily something they expect when they walk into the lab for research. But I do hope that there have been a few careers inspired by spending the day with all three of you in the lab. I'm sure there have been.

Hanne Bartels

There have. I can confirm that there have been a few participants who have already inquired about the volunteering opportunities and they are waiting for to be graduated from start undergrad so they can come to the lab. So Karen, at some point you will get emails from children who have implants and single sign or don't have an implant who are interested.

Karen Gordon

I love it. That's amazing. What a great experience and feedback. Thank you.

May Wang

That's really exciting to hear. I've learned a lot today, and I'm sure this has been a really valuable discussion for the audience as well. And I think to wrap up, I want to take a moment to express my gratitude to you all and to everyone else in the field for everything you've done to support children and families like mine through multiple channels. You know, the research, the patient experience, And even getting involved in this podcast to raise awareness around hearing loss. It means a lot to me. And I think many of our listeners in the audience will resonate with the same feelings of appreciation when I say this. But it's people like you that helped make my childhood experience with hearing loss. feel more supported and more manageable.

And I couldn't be more grateful for the support team at SickKids, along with my own family, for encouraging me to explore my potential and see my hearing loss as a gift and opportunity to develop resilience and strength rather than serve as my limitation. And the last thing I want to add is for the children like me who are listening to this episode and are also trying to navigate your own life's journeys, Yes, there can be challenging moments, and sometimes it can be frustrating when it feels like, you know, no one around you truly understands all the challenges you have to overcome every day. But something I've come to realize from my own experiences is that every challenge is an opportunity for growth, and every challenge you overcome will bring you closer to excellence. remember telling a friend about my story and how, you know, my hearing loss has shaped my perspective. And the very exact words he said right after was, You're amazing. And when I heard those words, it suddenly became clear to me that you don't have to make others understand what you're going through for others to appreciate just how amazing you are and can be.

And not only do I want to thank that friend for making a lasting impact with those words and for creating that positive memory, which has given me a lot more courage and confidence to share my vulnerabilities. But I also want to take this opportunity and say this to the children who are growing up with similar experiences. Your hearing loss is not a weakness. You can be amazing because of it. And the last little bit I'll share before we wrap up is, you know, as I experience more and more of what life has to offer, I've come to realize that every person's path to excellence will involve some form of obstacles, and we're no exception. But if you give yourself a chance to test your capabilities, you'll discover that there's a lot of hidden potential within you to achieve things beyond what you previously imagined, and we too are no exception to this. So don't let your hearing loss stop you from pursuing your dreams or exploring what you're capable of, because anyone and everyone is capable of living life with flying colors. And with that, we've now come to the end of this

episode. Thank you, Dr. Cushing, Dr. Gordon, Harrison, and Hanne for joining us today and for sharing so much of your expertise and knowledge. I'm very excited for the team and please stay tuned for our next episode.

[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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