

Will Eiserman:

Welcome to Earworm, dialogues on hearing health you can't stop thinking about. Earworm is brought to you by the National Center for Hearing Assessment and Management at Utah State University, known as NCHAM. I'm Will Eiserman and I'm the Associate Director of NCHAM and I'm your host today.

You know, we all make assumptions as we navigate various challenges in life. Many of those well-tested assumptions allow us to function very efficiently, but sometimes our assumptions may be incorrect. Today we're going to be talking about one family's experience that illustrates this point, in the context of a young child's development that took an unexpected and unnoticed turn. It's a story of a child who passed all of her newborn screenings, including hearing screening.

Our guest today is Valerie James Abbott. Let's set the stage. At this point in the story, you and your husband Chris, are parents to one three year old girl whose name is Mary Claire, and now you're going to have a second child. How was that second pregnancy?

Valerie James Abbott:

So the pregnancy itself was pretty uneventful. Bringing her into the world was another story. My first child was born via C-section because she was breach and she was nine pounds. So with Bridie, I really wanted to try and avoid that. It was a very painful recovery. So we decided to try a VBAC, vaginal birth after Cesarean, and that ended up in an unidentified uterine rupture. And eventually when they put me in for an emergency C-section, it wasn't until they opened me up and saw the massive blood loss that they realized that I had had a uterine rupture, which is a very serious medical emergency that had gone on far too long. And so I didn't meet Bridie until several hours after her birth.

Will Eiserman:

And how was she through all of that?

Valerie James Abbott:

So she came out just shy of nine pounds. She was very healthy, super healthy.

Will Eiserman:

How about all those hospital screenings they do on her? How'd she do?

Valerie James Abbott:

She passed everything. She was a hundred percent super healthy, happy, hungry kiddo from the moment that I met her all the way through till we brought her home and in the months that followed.

Will Eiserman:

Yeah. Okay. And so she passed her hearing screening and all the other ones?

Valerie James Abbott:

She sure did. She sure did.

Will Eiserman:

And then in those next weeks and months, you had, I assume, well child visits that you went to, and did you do that pretty consistently like you're supposed to?

Valerie James Abbott:

Oh, yes, with both kids, the well checkups, the vaccinations, everything was on track. I had my little chart and checking everything off, and Bridie was, and still is, my healthiest kid. We rarely went to the pediatrician because she was so healthy. No fevers, no ear infections, no nothing. She wasn't really in the pediatrician's office very often because she was so healthy.

Will Eiserman:

And how about the developmental milestones that everybody's always looking at? How'd she do with those?

Valerie James Abbott:

Physical developmental milestones were all completely on track. Everything was on track, feeding, gross motor skills, fine motor skills, social skills, all of that seemed to be completely on track. And really throughout her toddler-hood, up until the point that we enrolled her in preschool, around two and a half, everything seemed to be completely fine. And her pediatrician had zero concerns and we had zero concerns.

Will Eiserman:

So everybody, the healthcare providers, you, the teachers are making an assumption, Bridie's developing typically. She's developing language, she's developing her speech abilities and repeating words typically as you might expect for that age. And then you enroll her in preschool, and then what?

Valerie James Abbott:

In my mind, everything was still completely... Everybody was developing, including parents exactly as they should be. And when we enrolled her in preschool, we believed that she was developmentally ready for preschool and on track with everything with her peers. That's what we believed. And so that was in September, and it was late January, early February, the parent teacher conference that everybody has mid-year was scheduled. And my husband and I went to preschool to sit in the teeny tiny chairs that they make parents sit in. And the preschool teacher got right to the point and she asked us if we were concerned about Bridie's speech and we said no, because we really weren't. And then she said-

Will Eiserman:

She's about what, two and a half at this point?

Valerie James Abbott:

Two and a half, two and a half, just a little bit older than two and a half. Right.

Will Eiserman:

Okay.

Valerie James Abbott:

And she said, "Do you find her difficult to understand?" And we're like, "Well, yeah, but isn't that normal?" And the preschool teacher said, "We are concerned about her speech. She is not developing the way the rest of the class is." And so I said, "Okay." And she said, "Would you consider having her

evaluated by early intervention?" I had never heard the term before. I didn't know what it was. I said, "Sure." And so she provided that phone number.

Will Eiserman:

Now, had you ever talked with your healthcare provider about Bridie's speech or language development?

Valerie James Abbott:

No. I mean, she was such a healthy kid that the only time we saw her pediatrician was for well checkups and vaccinations, and so those were fairly quick appointments. I do recall a question, something along the lines of, "Do you have any concerns about hearing?" And the answer was no. And that was the end of that. So no, there was no conversation about developmental milestones. He was asking questions and we were answering them, and then we would leave.

Will Eiserman:

So now you have set up the early intervention evaluation, which is in your home?

Valerie James Abbott:

Yes.

Will Eiserman:

What happened?

Valerie James Abbott:

I was so not concerned that I sent my husband off to work. I sent my oldest off to kindergarten. I took the day off of work because they come to the house to do the intake. Bridie was still in her pajamas. And I mean, I gave no thought. I really did not give any thought between making that phone call and saying, "Hey, I'm supposed to call you. And I've been suggested that maybe we need to do a speech evaluation for my child." And they set it up quickly and they came. I gave no thought to any of those things because I thought, "Okay, so she has a speech issue. Okay." But I really was not concerned at all. And maybe that's because I had one in kindergarten and I have fond memories of being pulled out of... Being treated special. I don't know. But it was during that evaluation when it became abundantly clear we were looking at a hearing issue.

Will Eiserman:

And was that because they did actually a physiologic screening on Bridie during that interview, or what happened?

Valerie James Abbott:

Yeah, so they basically were observing me doing whatever I was doing, wiping the counters and telling Bridie to go brush her teeth or whatever I was doing, very normal stuff. And at one point they said, "Okay, Mrs. Abbott, would you please tell Bridie to go into her room and go get three things that she can get safely and bring them to you?" And I said, "Sure." And I said, "Bridie, come close to mommy. Come close to mommy. Bridie, I need you to go to your room. I need you to go to your room and get your hat

and get your gloves. Go to your room and get your hat and your gloves, because after this, we're going outside."

Will Eiserman:

So I should interject here that as you are retelling this story, you are gesturing in a very animated way, signaling, gesturing, and even miming what it is you are requesting of her.

Valerie James Abbott:

And so she goes to her room and she gets the things and she brings them back. And I was like, "There you go. See? She's fine." The lady said, "Great, thank you so much." And she said, "Okay, now can you ask her to go into the kitchen and get two or three things that she can get safely and bring them back to you, but don't use your hands?" And I'm like, "Okay. Okay." So I'm thinking of the things, okay." And so I said, "Bridie, come close to mommy. Come close to mommy. That's right. Bridie. Go to the kitchen. Go to the kitchen, get a spoon."

Will Eiserman:

And you're gesturing again.

Valerie James Abbott:

"A spoon and a napkin. We're going to have some ice cream, go to-

Will Eiserman:

Because your habit of talking to her is accompanied by all of this gesturing and your own made up sign language.

Valerie James Abbott:

And so she got her spoon and her napkin and brought it. And I was once again, like, "See, she's fine." Then they took Bridie and they sat her in the corner of our living room facing the corner, and they put her in the corner of the room and they gave her some blocks. And I'm sitting on the sofa and they sat on the sofa behind me, and one of them took out a bell, that in my memory was like a hand chime like you would see in a church hand bell choir. And they pulled it out and started to ring it, and Bridie did not respond. And then they took out a different bell with a different tone and started to ring that, and Bridie did not turn around or respond at all.

And as you can imagine, I thought every awful feeling a human can have just suddenly I felt like I was both electrocuted and about to throw up. It was this awful, oh my God, what did I just witness moment, and I had a meltdown right there. And the woman from early intervention said, "Mrs. Abbott, this can be so many things, so many things. I think we may be looking at a hearing issue, but that could be, again, so many things. But the next thing we need to do is to line up an audiological evaluation to actually test her hearing and see if in fact, we're looking at a hearing issue. And they did that through their office fairly quickly.

Will Eiserman:

So one of the things that comes to my mind as you share that heart-wrenching experience is that we know that some children with a hearing loss may have turned toward the bell because they might've heard it just enough, or there may have been some signal in the room that cued Bridie to turn toward

the bell, and that if we're only ringing bells, that's not even enough. We really do need a physiologic screening. In that moment that you just described, ideally. So the outcome of that, you started to recognize that Bridie was more delayed than you had realized?

Valerie James Abbott:

Correct. Far more delayed. In fact, we did not think she was delayed at all. So when they were pointing out specifics, it became abundantly clear. Her speech was unintelligible, for the most part. There were some words that were crystal clear and others that were completely a different version of what they should be. She could not understand instructions if she could not see your face. So she had become a very, very successful lip reader. And so if she could see your face she could understand pretty much anything, but without the ability to see your face she couldn't understand anything. And that was something we didn't notice. She adapted so that she could communicate, so that she could understand, she adapted, and we did not notice what she had done to adapt.

Vocabulary in general was she was not on par with where she should be developmentally when it comes to just vocabulary. For example, she did not know the difference between she and he, she did not know the difference between under and over and next to and beneath. Children, by the time they're almost three, have all of that down. You can say, what's beneath the sofa? And they know what you're talking about. None of those words made sense to her. By the age that she was, she should have had a really firm vocabulary for understanding information and instructions, and she didn't have that. She didn't know her colors or her numbers to the extent that she should by that age. Those are just things that we just didn't notice. We really didn't. She was a chatty, chatty kid speaking her own language, and we thought that was developmentally normal, and maybe it was up until a point, but then it became where she was not progressing.

Will Eiserman:

So after the meeting with the early intervention evaluation team, you went on to have the audiological evaluation?

Valerie James Abbott:

Correct. We discovered that Bridie has a moderate sloping to severe bilateral sensory neural hearing loss, and no one could tell us how, when or why that happened.

Will Eiserman:

Yeah, because you're thinking about early on and this language production that was happening where she was communicating with you.

Valerie James Abbott:

Absolutely.

Will Eiserman:

She had learned words, many of them correctly. Right?

Valerie James Abbott:

Some were correct, and some were partially correct, and some were completely not.

Will Eiserman:

Right, like a lot of two-year-olds.

Valerie James Abbott:

Like a lot of two-year-olds. Exactly. Exactly.

Will Eiserman:

So ultimately, you had this diagnosis. What did you do to address it?

Valerie James Abbott:

We did move forward with hearing aids. She picked out her colors and we brought them home. When they finally came, we had ordered custom matching cupcakes, so her hearing aids...

Will Eiserman:

You're talking about the colors of her hearing aids?

Valerie James Abbott:

Yes. So the back was a baby pink and the mold was pink with silver sparkles. And so we got cupcakes with pink frosting and edible silver glitter, and we invited the neighbors over and some of her preschool friends, this is the day that she got them, to show them off and to just, it was a kind of fake it till you make it moment. We were still really upset, really panicky underneath it all. This panic and guilt, very, very heavily weighing on us, but we were a let's go big or go home.

Will Eiserman:

So this is a compelling story, and yet, you know what I'm not hearing about is engagement with your healthcare provider.

Valerie James Abbott:

That's right.

Will Eiserman:

So you're going to the healthcare provider on schedule to all of these check-ins on your child's development. And this significant physiological concern is happening outside of that context.

Valerie James Abbott:

Correct.

Will Eiserman:

So where do they get included in this?

Valerie James Abbott:

Yes. So in our case, Bridie received her hearing aids, I think it was the end of April.

Will Eiserman:

So she's three now.

Valerie James Abbott:

So she's not quite three. She's turned three May 10 and May was when she had her physical, her annual well checkup. And remember, she's so healthy that there's never a time to ever bring her in before that. And I remember the doctor who was older, and we chose him because he was so seasoned and so well-respected and a pinnacle in the neighborhood. And I remember bringing her in and sitting her on the table thing, and he comes in, he's like, "Oh, how are things going?" In his grumbly little voice, and he goes to look in her ears and he goes, "What is this?" And I went, "Oh, they're hearing aids." And he was like, "What?" And he looks in the other ear and he's like, "What?" And then he opens up the folder and he's pulling, he goes, "She passed new newborn hearing screen. "

I was like, "Yeah, uh-huh, she did." And he's looking through the file rigorously and just looking at us with this puzzled look on his face. And we had a couple of months to come to grips with this. And he said, "Well, did she have fever? Did she have...?" I'm like, "No, no." And he's like, "What?" I'm like, "I don't know what to tell you. She has hearing aids. Well, we went through early intervention. We had our speech evaluation." And then I told him kind of what happened and what was wild was every appointment after that, every single appointment after that, when he would pull back her hair to check her ears, it was the surprise. And then the, "Oh, right, this is the kid." My husband and I could feel this sense of, "How did I miss that? How did this happen?" And extra ruffling through the file looking at things, and did we ever figure out what this was?

Will Eiserman:

Because you can't help but wonder, right? If you had not had that preschool teacher who had brought that to your attention, who first of all noticed.

Valerie James Abbott:

That's right.

Will Eiserman:

And second of all noticed and was concerned enough, skillful enough to deliver that concern to you, to motivate you to take it to the next step. If none of that had happened, when would this have been discovered, and by whom?

Valerie James Abbott:

That question has been with me for years, years. That specific question of what would've happened if Ms. Lily hadn't said, "Are you concerned about her speech?" And what would've happened if she didn't tell me, "Early intervention is where you go." What would've happened if I'd gone back? If she'd said, I think you need to talk with a pediatrician. I have no idea what that outcome would've been. What would the outcome have been if she had said, "I think you need to have that checked out." What would I have done? I don't know those answers, but it all started with that question.

Will Eiserman:

And we know that there are a number of stories of children who are in fact referred for speech therapy, who are enrolled in speech therapy, and their hearing status is never determined, especially during

those first three years of life or during the preschool period. So this story could have gone on much longer-

Valerie James Abbott:

It could have gone on.

Will Eiserman:

... And had much more of an impact. So ultimately, the medical community did raise another point of concern about what it really means to be diagnosed. Hearing loss is, in a way, in many instances, a symptom of another condition, which in your case is a genetic condition.

Valerie James Abbott:

At some point, that first year, I believe it was the pediatrician who was just scratching his head and said, "Do we know what this is? You have to have this in your..." All the questions, and mentioned genetic testing. And so we went ahead and had genetic testing done. Part of the reason we wanted to go ahead with genetic testing was my husband and I were feeling very, very guilty for having tried a VBAC delivery. And having had that been such an incredible hot mess. We believed that it was probable that her hearing loss was as a result of the trauma that occurred during her day of birth.

Will Eiserman:

Whoa. That's a really heavy load to be carrying around.

Valerie James Abbott:

Yeah.

Will Eiserman:

Did you ever ask anybody about the likelihood of that being the cause?

Valerie James Abbott:

Yes, it was the geneticist. And so when we were going through the billions of questions that they ask, and I was telling the birth story, at some point, I said, "My husband and I are feel like it was the uterine rupture nightmare that caused this, but we're doing genetic testing so that whatever..." And she said, "I highly doubt, given she was not in a NICU, she came home very healthy. I highly doubt that was the cause of this. But we're going to test for something very, very specific called Connexin 26, and it's a needle in the haystack, but let's just see if the needle's there."

And so she sent off the labs. I get a letter in the mail that says, inconclusive. And I was like, "I don't understand what this letter says. I don't understand what this letter says." So I called and I said, "I need to speak with the doctor because I think I know what this says, but I want to hear it from her." And so she called back and I said, "Okay, can I just paraphrase what I think this letter says?" And she said, "Sure." And I said, "What I think this letter says is that Bridget has inherited connexin 26, but she has only inherited it from one parent, and therefore we cannot say with certainty that connexin 26 was the cause of her hearing loss because you need it from two parents and not just one. And therefore that's why it's inconclusive."

And she said, "You read that correctly." And I said, "Okay, but is it possible that science has not yet determined that maybe you only need one parent with connexin 26 to cause hearing loss?" Because

what are the chances that my child loses her hearing at the timeframe that connexin 26 can cause hearing loss in children, and she has connexin 26, but it's only from one parent. And she said, "Is that possible? Absolutely." And I said, "Okay, well, do you think that if we're balancing between, it was as a result of her birth story or as a result of this connexin 26?"

And she said, "Your child's hearing loss was not as a result of the birth story that you described to me." And that just suddenly, suddenly it was like this giant weight, overnight just disappeared, and the amount of guilt that we were carrying was cut in half. And she said, "However, as a scientist, I can't tell you conclusively that connexin 26 is what caused her hearing loss." And I said to her, "Well, we are choosing to say that is what caused it, because that's what we believe."

Will Eiserman:

So that genetic diagnosis, evaluation and diagnosis of connexin 26, first and foremost helped you in a psychological way, but then it also contributed to an understanding of some of the important things you needed to be paying attention to in terms of Bridie's hearing status when connexin 26 can be progressive or it may be stable. And so in your case, you found out that, you've discovered over time that Bridie's is a stable hearing loss, right?

Valerie James Abbott:

Yes. But because we believed that it was as a result of connexin 26, and we knew that connexin 26 can be progressive, we had her hearing tested for the first year or two every three months, then it was every six months for a couple of years, and then it was every year.

Will Eiserman:

And now she's 17.

Valerie James Abbott:

And now she's 17.

Will Eiserman:

And it hasn't changed.

Valerie James Abbott:

No, it's exactly the same.

Will Eiserman:

Yeah. And when you look back, can you kind of have a guess about...? We are pretty sure right, that she was hearing at birth?

Valerie James Abbott:

Yes. I feel very confident that she was, yes.

Will Eiserman:

Yeah. And that you experienced a fairly adequate hearing level as an infant, and up until about what year, what month, would you guess?

Valerie James Abbott:

Yeah, at some point along the way, someone suggested that her hearing loss may have occurred, that it was likely around 18 months of age. So in the middle of toddler-hood, because some of her words, the ones that a child learns very, very early on, even before they're talking, those were correct. But the words that you might start to acquire after, in later toddler-hood, those were the ones that were all over the place.

Will Eiserman:

So Bridie is now 17.

Valerie James Abbott:

Yes.

Will Eiserman:

How is she doing?

Valerie James Abbott:

She is thriving in all the ways that a 17 year old should thrive, right? So she will be graduating from high school, she has a job. She is involved in extracurricular things. She has friends that she hangs out with. She bought her own car and she just got into her first choice college, which is a big deal. And she's thriving and all kids like her should thrive in whatever way they're supposed to be thriving. So Bridie uses hearing aids, and she is a chatty kid, so she uses listening and spoken language in English. And from the very beginning, that combination of hearing aids and listening and spoken language was what really worked best for her. And so she communicates with the world the way that we do in our own family.

We've seen firsthand what happens when we catch children early, when we get them kindergarten ready, when we invest everything we possibly can into their development and their success because they're capable of anything, these kids, but we have to find them first. We have to find these children with unidentified hearing loss first. We have to find these children sooner because I've seen firsthand what happens to language and literacy when a child's hearing loss goes undetected. And not even for that long. When we think about it, it was maybe a year, it was maybe a year that she did not have full access to the sounds for speech and look at the damage that it did, and just how derailed she became in terms of kindergarten readiness.

Will Eiserman:

Now, since that time, you've become a national advocate for early identification of hearing loss and raising the awareness of monitoring the hearing status of children throughout all of these important developmental stages. And I know you've heard stories, Valerie, about how people don't think of hearing first when a child may be developing in some unusual ways or displaying behaviors which may be accommodating for any number of things that may be going on for them.

Valerie James Abbott:

When a child is presenting with behaviors that are not making sense or seem to be off or whatever terminology we want to use, I always hear people, not just parents, but other family members saying, "I wonder if it's autism. Do you think it could be autism? Or, I wonder if these are the early signs of ADD, or, I bet he has ADHD." No one, ever have I ever heard say, "I wonder if he can hear clearly. I wonder if

there's been a change in the way he hears the world?" Ever. Everyone seems to be going to the conversations about autism and the conversations about ADD or ADHD. So one of the things that I have learned is that we need to be conducting hearing screening before all of the other things that we might be screening for with the child, whether that's screening for autism, screening for a speech delay, and certainly before enrolling a child in speech therapy.

Too often we have children who are identified with autism and then it stops there. Or a child that is identified with a speech delay and it stops there, we enroll them in speech therapy. Well, maybe there's also hearing loss, maybe hearing loss is actually what is the root cause of the speech delay. But too often the hearing screen is at the end of the process, if it even happens at all. And so we need to put the hearing evaluation part on the front end of all of these screening practices. And the other thing we know is that the vast majority of children who are deaf or hard of hearing are born into hearing families, typical hearing parents. And so that's a fact we all need to be aware of, it's the majority.

Will Eiserman:

Valerie, thank you so much for telling your story and for motivating all of us to pay closer attention to monitoring the hearing status of children throughout their early lives, to not rely solely on newborn hearing screening results as a sort of stable, if they pass then that it has to somehow mean they would pass today, result, so that we keep paying attention, especially if changes start to occur in children. Do you have any final parting messages you'd like to give our listeners?

Valerie James Abbott:

Sure. I am convinced that we can find children with postnatal hearing loss sooner, but it takes all of us to be part of this conversation, opening our eyes all the time, adding to the commentary. And I do, I do believe that we can find them sooner, but it's going to require every single one of us.

Will Eiserman:

That was Valerie James Abbott. I'm Will Eiserman at the National Center for Hearing Assessment and Management, and I want to invite you to join us for our other podcasts at earwormpodcast.org for dialogues on hearing health you just can't stop thinking about. Earworm is produced at the National Center for Hearing Assessment and Management, NCHAM, at Utah State University, USU, and is funded in part by a cooperative agreement from the Maternal and Child Health Bureau, MCHB, of the Health Resources and Services Administration, HRSA at the United States Department of Health and Human Services. Any views, thoughts, and opinions expressed by participants in Earworm are solely that of the participants and no endorsement by NCHAM, USU or MCHDB, HRSA is implied or expressed.