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OTOTOXICITY MONITORING AS PART OF RISK MONITORING IN THE EHDI SYTEM
OCTOBER 8, 2015
1:30 P.M. ET

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(Writer standing by.)

>> This is an audio tech for today's webinar that is brought to you by NCHAM. Jessica, do you want to do an audio check with us right now as well?

>> Sure, I can. My name is Jessica Stich-Hennen. I am a pediatric audiologist from Boise, Idaho. How does that sound?

>> Great. You sound good. Yep. That's perfect. So, we'll be starting here in about three minutes. Go ahead and adjust the volume to your liking, on your end, on your speakers or headsets. Today, you will be communicating with our presenter through a Q & A text field on your computers, so you don't need to worry about being linked up or being on the phone with us today, and thank you for taking a moment to complete the poll question that is on the screen right now. That's helpful for our presenter to have an opportunity to know a little bit about the perspective that our participants are bringing to the webinar today, so thank you

for that. We'll leave that up there for another minute as people are signing on fairly rapidly right now in these last few minutes before we're getting scheduled to start. Today's webinar is going to be recorded and then posted on Infanthearing.org, so if, for any reason, you have a poor Internet connection today or need to leave the meeting early, you can always come back and stream this webinar live or in the recorded fashion from infanthearing.org. That'll be up there within the next week or less, and our presenter's slides will also be available to those of you who would like them. So, we'll just wait another minute or so and then we'll get started. People are signing on fairly rapidly right now. For those who have just joined us, this is a webinar that is entitled ototoxicity monitoring as part of risk monitoring in the EHDI system, brought to you by NCHAM at Utah State University. You can adjust the volume to your liking on your computer headset, on your computer speakers or your headset, and you'll be communicating with our presenter today through an online text screen that will be displayed when our present is opening up the conversation for questions.

So, um, we'll give it one more minute and then we'll get started. I'm going to disappear this poll question for now, and I'm going to activate the recording for today's meeting, so I will go silent for just a moment while I do that. Well, hello, everybody. My name is Will, I'm the associate director of the National Center for Hearing Assessment and Management, also known as NCHAM at Utah State University, which serves as the national

resource center on early hearing detection and intervention. We are delighted today to offer a webinar entitled ototoxicity monitoring as part of risk monitoring in the EHDI system that will be presented by Dr. Jessica Stich-Hennen, who is an audiologist from Idaho State, and Jessica has received her clinical doctorate in audiology from Idaho State University and has a clinical specialty in areas of pediatric diagnostics and amplification, auditory potentials and central auditory processing disorders. In April of 2011, she achieved specialty certification in pediatric audiology from the American Board of Audiology. She is the primary audiologist for the Idaho Cleft Palate Cranial Facial Association and presented in 2012 at the national EHDI meeting, and we are delighted to have Jessica here today with us to share her information about this important topic. So, before we get started, I just want to let you all know that today's webinar is being recorded, and it will be posted on infanthearing.org within the next week so you can view it or share it with others from that site later, and after Jessica has presented her information today, we'll be opening up a Q & A field on the screen for you to raise any questions or thoughts you'd like to share with today's presenter. So, Jessica, I will turn it over to you.

>> All right, thank you, William. As William said, my name is Jessica Stich-Hennen. I am a pediatric audiologist in the state of Idaho. I work at the largest hospital in the state. I've been here for several years now, also working with the state

EHDI program on the newborn hearing screening process and their high-risk monitoring system. By no means am I in expert in ototoxicity, but I do work very closely with our program here in Idaho and also wrote the EHDI chapter with another audiologist, Dr. Bargain, so I do do a lot of research in this area, and hopefully, I know a little bit about what I'm talking about today, so bare with me here. We're going to get through a lot of basic information first, and then we'll get into the ototoxicity in a few minutes. So, first off, we are going to look at JCIH position statements, because that's really where high-risk monitoring all began. As we all know, in the 1970's is when JCIH first started putting out position statements regarding newborn hearing and childhood hearing loss. In their first mention and first statements, there wasn't really a discussion of ototoxicity or ototoxic medications, it really focused in on trying to establish a way to identify hearing loss in young children, but by the 1990's, the position statement changed a little bit, and they added in a criteria, particularly for neonates from birth to two years of age, and that gave a definition of ototoxic medications, and it particularly read ototoxic medications, including but not limited to the aminoglycosides used for more than five days, and also, loop diuretics used in combinations with these aminoglycosides.

So, when they added this to the position statement, there wasn't any given protocols or recommendations for how to test, when to test, but they did add that in, which definitely sparked

some discussions and brought a lot of research in the area of ototoxic medications. In the 2000 position statement is when they actually gave a guideline or recommendation on how to monitor risk factors in infants, and at that time, they recommended that audiological testing should occur every six months until the age of three years for children having any of the identified risk factors. As we all know, that position statement really sparked newborn hearing screening across the country, but a bigger addition, in my eyes, was that we also recommended this risk monitoring system, and at the time, that was a lot, that was a pretty big burden on the system, recommending six months monitoring until the age of three, but then when the 2007 position statement was then published, there were some changes made at that time. They looked at expanding our targeted hearing loss definition, looking at auditory neuropathy spectrum disorder, and looking at the differences between an NICU and a well-baby nursery. So, we all know that with that statement, they had recommended that ABR screening was the preferred method when looking at an NICU program. So, those are some of the bigger changes in that statement, and then they also looked at readmissions within the first year of life, and anyone readmitted within the hospital within that first 30 days would need a repeat hearing screening if there was a risk associated with hearing loss that presented at that point.

The monitoring of high-risk factors also had changed. As you recall, in 2000, every six months until the age of three was

the recommendation in that position statement, but in 2007, they gave a vague, more vague, I feel like, recommendation for monitoring, and at that time, they recommended that infants with risk indicators for hearing loss should have at least one evaluation by 24 to 30 months of age, and definitely, that, I feel like that big shift from having a very strict criteria recommended to a more loose definition and schedule definitely, I think, is what's caused a lot of confusion and a lot of difficulty when trying to come up with guidelines within hospitals. So, with regards to risk factors, today, I'm going to talk a little bit about multiple risk factors, but the main focus of today is ototoxic medications, but this is just, again, the appendix from 2007 and looking at all the risk indicators for hearing loss in young children and infants. So, one of the ones I wanted to mention today was an extended NICU stay, and the reason I wanted to mention it is because of some information coming out in 2014, and then the other reason is some of the data we have collected here in Idaho. So, the reason this was added, the NICU stay of greater than five days, to this 2007 position statement, it was added because of some information coming out of the National Perinatal Research Center, and they had identified there was kind of these two populations within an NICU program, and approximately 25 percent of NICU infants were in this kind of low-risk category, and those kids were mostly getting discharged by five days old, so they weren't there any longer than five days, but then they found that there was this more high-risk,

or this target population, that they found were the ones that they were trying to rule out some kind of neural hearing problem for, and this was about 75 percent of the NICU infants, and they were staying in the NICU for greater than five days, which is why that recommendation went into that 2007 statement.

What I mentioned earlier is in 2014, they did a publication, and they looked at their program and the NICU stay of greater than five days, they found it not to be associated with an increased risk of hearing loss in the population and the infants that they had evaluated in their study, and we actually, in Idaho, find, we get risk factor referrals all over the board from cranial facial to NICU stay to ototoxic medications, most of the time when we're getting referrals, we have a lot of kids that have multiple risk factors that are getting referred. We definitely have a lower percentage that is just being referred because of an NICU stay greater than five days, and those tend to be premature infants that's in there to be monitored for feeding issues for maybe a week or two, and we do see those kids in our clinic, but we don't have a high number of kids getting identified with hearing loss that have just had an NICU stay of greater than five days. Typically, our kids that we're identifying with a delayed onset hearing loss are those that have multiple risk factors, which we'll talk about a little bit later.

The other part, another risk factor I kind of wanted to talk about before we get into ototoxic monitoring specifically

is ECMO treatments, and that's an aggressive medical treatment. It's used for the life support of infants who are in respiratory or cardiopulmonary failure. It is a very aggressive medical treatment. We don't even have this treatment actually in Idaho, but we have nearby states that do have this treatment, but there was an article published in 2008, and he had presented some information from his hospital in Boston on 111 neonates, and what they found in their research of reviewing cases of kids who had received ECMO was that if a child was receiving ECMO treatments and also received 14 days or more of aminoglycoside therapy during that ECMO treatment, it increased the risk of hearing loss 5.5 six times, but because of having those two treatments coinciding. So, how they broke that down a little bit further was if a child had 14 days more or of aminoglycosides, over 80 percent of the kids who had received 14 days or more during their ECMO treatments had a sensory neural hearing loss by two and a half years of age. This compared to kids who received less than 14 days of aminoglycosides during their ECMO treatment, 30 percent had a sensory neural hearing loss by six years of age. So, pretty big difference in the amount of aminoglycosides used there and increasing the risk significantly, but even those kids who had less than 14 days still had a percentage of kids having sensory neural hearing loss occurring as a delayed onset.

So, let's talk about ototoxicity, why we're here, and what does that mean? It basically is a medication that can damage

the ear, resulting in hearing loss, ringing in the ear and/or a balance disorder, and we're not going to really focus in on ringing in the ear or balance disorders, we're really going to talk more about that hearing loss and the association of hearing loss related to those medications. So, there are different, over 200 known ototoxic medications, and these can vary from anywhere from prescription medications to over-the-counter medications such as aspirin. They can be used, these medications can be used to treat serious infections, cancer, heart disease, and depending on the medication that is given, the damage that can be caused can be temporary or permanent, and the temporary ones that I'm talking about here are things like tinnitus occurring after aspirin use, and when we're talking about permanent, we're going to talk about today more of the permanent damage, which would be from medications, which is a chemotherapy drug, but we're going to focus our time today on gentamycin. So, why are we concerned about ototoxicity with infants?

Why are we having our discussion that we are today? The most frequently occurring, this is a look at some information on over 3,000 NICU babies that they examined to look at how often are ototoxic medications given, and what they were finding was over 70 percent of the infants that they looked at in this, in their research were given an ototoxic medication. So, you can see, when we're looking at how often do risk factors occur in infants, ototoxic medications is the number one risk factor that's

occurring. ECMO treatments, when we talked about, which, obviously, is a very big concern with the hearing loss associated with it, is down there closer to 10 percent, so not occurring near as often as ototoxic medications are in that NICU population. Recently, OHSU published some information that said 600,000 infants in NICU s across the United States, 80 percent of them have received some form of aminoglycoside therapy. So, that's why we're here today. There's a lot of infants out there receiving ototoxic meds, so we need to figure out how to monitor them and what's the best way to identify a hearing loss, if there is hearing loss to be identified. So, least frequently occurring risk factors, things, meaning how often are kids getting referred for these risk factors, definitely lower numbers, cranial facial anomalies, family history of hearing loss. You know, we, with congenital infections and bacterial meningitis, definitely, they are few and far between in our state. We do have kids that get referred for that, but that's definitely not our number one referral for risk monitoring in our state.

So, if we looked at those previous two slides and we looked at how some risk factors occur really frequently, such as ototoxic medications, and some don't occur as often, now this information that she published, it looked at, if we have these risk factors, how often is hearing loss occurring among these risk factors? And right at the top there, which is something near and dear to my heart, working on the cranial facial team, is cranial

facial anomalies with greater than 50 percent, and I've actually done some review of our data in our program, and we definitely have over 50 percent of our kids that have hearing loss, educationally significant hearing loss, whether that be a fluctuating conductive hearing loss or sensory neural, a significant amount of hearing loss, obviously coming in that cranial facial anomaly population, and where we would put ototoxicity in this group would be under the other risk factors. So, that's saying that less than 10 percent of kiddos that receive ototoxic medications would potentially have a hearing loss associated with that risk factor. If we're talking about ototoxic medications all on their own, and as we all know, a lot of these children have multiple risk factors when they present in our clinic, not just one risk factor at a time. So, let's talk about aminoglycosides. What are aminoglycosides? They were introduced, it's an antibiotic that was introduced in the 1940's. They are used to treatment serious infections. They may remain in the hair cells of the cochlea for months after application or after the drug was administered, and this was an interesting finding that I found, some information I found from the ototoxic monitoring guidelines from 2009. When they addressed aminoglycosides, which if anybody has read this position statement from triple AAA about ototoxic monitoring, it really does heavily focus on chemotherapy treatments and chemotherapy drug monitoring schedules, but they did talk about aminoglycosides, and when they talked about them, their recommendation for monitoring was weekly or biweekly monitoring is

recommended, ideally, and then they said that follow-up testing should also be scheduled a few months after the drug has been discontinued, and again, kind of going back to that position statement from 2007, where we said one audiology evaluation before 24 to 38 months of age, this also is a pretty vague definition. It doesn't really tell us how many months after the discontinuation of the drug we need to monitor, it says a few months, and it also recommends this weekly or biweekly monitoring, which I know is pretty unrealistic in an audiology practice, to be monitoring a child that has received gentamycin weekly or biweekly for several months after the drug was given.

So, I'm not sure that statement really helps give us any concrete way of deciding how to monitor infants that received those medications. So, particularly, gentamycin is probably the one that most people know of when we talk about amino glycosides. It was introduced in 1963. It is the most common aminoglycoside used in a neonatal intensive care unit. Why is it used? It tends to be used because of the low cost, but also, its effectiveness against the gram negative bacteria, and I've actually spoken with our neonatologist several times about this, and those are exactly the reasons that they give as well. It's effective, and it works, and it is a low cost, and they feel a really low risk, which, you know, if we're talking about hearing loss, a really low risk drug to be given if we're trying to save an infant's life from an infection. In 2010, they published an evidence-based review of

drug-induced hearing loss from gentamycin, and it's a really great document to read. It's kind of interesting, because basically, they looked at several studies from the early 1990's to the late 2000's about studies that had been published regarding gentamycin youth and gentamycin trials, and they reviewed different things. They looked at the effects of dosing, they looked at dosing schedules, they looked at these studies and evaluated whether the root, or what type of administration of the drug, was it topical versus an IV. They also tried to see, if you use something like gentamycin with another medication, was that showing more or less hearing associated with it. So, the findings from these 20 studies that they looked at, the hearing loss from gentamycin, so hearing loss associated with gentamycin treatments, they were all over the board. They ranged from zero percent reported to 58 percent reported, and I think the hard thing about some of the conclusions from this studies is that they really varied, and so if you look and you read these studies, the studies varied on how much medication they were giving patients, what population they were testing, whether it was pediatric versus adult, what medical problems the patients had, whether they were receiving chemotherapy with gentamycin or what, if they had a specific infection. They definitely varied on diagnostic testing. Some were using high frequency audiometry, some were using traditional audiometry, some used OAE and ABR method, so that also made it hard for them to do a good review on these studies and to come up with a concrete

recommendation.

Then, also, the studies varied on what their criteria was for hearing loss and having a shift of hearing from the medication. So, if they administered, if they obtained a baseline and they administered a drug, some studies would say a 5 decibel shift was considered a hearing loss, and others would say a 15 decibel shift, so it's really hard to examine the effects of gentamycin causing hearing loss, because studies that have been done, they vary so much that it's hard to compare, and not a lot of these studies have a significant participation. So, one of the things in the study that I found interesting was that they said that there was insufficient evidence to draw conclusions on if a hearing loss was occurring because of the treatment of gentamycin, and they also didn't find that there was enough evidence to support multiple ototoxic drugs causing hearing loss either. They just found that, you know, they looked at all this information, but there wasn't really enough to give a concrete answer on how to monitor and when to monitor and if there are really truly ototoxic effects from gentamycin. Some trends in this study and some things that they said that, you know, were trends that they'd like to report, is it concrete enough for them to say this definitively, they felt like it wasn't, but they did find some trends, and they thought that the frequency of administration of the drug, so how often or how much of the drug was given of gentamycin, did not influence the likelihood of there being a hearing loss, and we'll talk about why

that might be, and then the dosing amount, how much did not influence the likelihood of there being a hearing loss. Across the studies, this was the findings that they were noting.

So, why might that be? It may have something to do with this genetic mutation, mitochondrial mutation that we have hopefully all heard of called A-1555G, and this genetic mutation was first reported in 1993, and it was associated with aminoglycoside deafness. This study in 1993, it said that even one single dose may result in sensory neural hearing loss. It's interesting though, if you look, a few years later in 1998, a different study reported that they found profound sensory neural hearing loss in patients who had that mutation, but they didn't receive any aminoglycoside treatments, so that kind of, you know, that information gives us kind of two ideas here, one saying that if we have this mutation, it's associated with deafness, but the other one is saying even if you have this mutation, you might not need to have the aminoglycoside treatments and deafness was found. So, a couple other studies I wanted to talk about a little bit was some information that looked at the prevalence of the mutation, so how often is this mutation occurring. The UK had a study that came out in 2002, and they found that 1 in 206 newborns were expressing this mutation, but some information out of Texas reported that 1 in 1100, approximately, newborn infants had the mutation. If you look at a lot of the research regarding this mutation or publications on it, definitely, hearing loss can occur with or

without aminoglycoside treatments with this mutation.

The mutation is definitely more prevalent in countries outside of the U.S., comparing to the U.S., but we don't have a lot of research being done here, I guess, on this mutation, so is it more prevalent here than we know? Possibly, we just don't have the data to support that at this time. So, ototoxicity in pre-term infants, let's talk a little bit about that. This was some information out of Iowa Children's Hospital in 2011, and there was 700 infants, and only 1.8 of them that they looked at had this mutation. So, you know, again, a very small portion of this population having this mitochondrial mutation, and out of those infants that they found in the study, not one had hearing loss. So, I'm not sure if the mutation is our best predictor, at this point, because even when we have the mutation and we're given aminoglycosides, it's not always resulting in hearing loss, and sometimes, we can have that mutation and not have the treatment and have a hearing loss. So, another good part of this information that they presented on that was interesting, it was loud noise exposure, they presented on several studies, animal studies, that looked at the potential effects of loud noise exposure with aminoglycoside treatments simultaneously, and so there's been several studies looking back into the 1960's that have found that animals that were receiving aminoglycosides, two studies in particular, that when they were receiving the aminoglycosides and they were exposed to loud noise, they were more susceptible to noise-induced hearing

loss.

With that being said, then the theory to consider would be is if we are, you know, if we have a child who maybe is a pre-term, high-risk baby, they receive an aminoglycoside treatment and they're in an NICU that is very noisy, is that putting them at more risk, if they have at mutation? Because we have the mutation, the aminoglycoside treatment and the loud noise exposure, and that was a theory that they posed in this article regarding high-risk aminoglycosides with that mutation. Again, some more recent publications about aminoglycosides. These ones all had to do with looking at aminoglycosides in animals, and a lot of the research is, but the first study here in 2015, their hypothesis was that if they synthesized a new aminoglycoside treatment, could they lower the risk of ototoxic damage, and they found that they could with these synthesized drugs. They were seeing some lower risk of hearing loss associated with the aminoglycoside designer drugs, is what they called them. The second one here looked at ototoxicity in hair cell oblation in adult gerbils, which is one of the models that's researched the most, and what they found, their data suggests that hair cell loss and hearing function is not particularly susceptible to gentamycin or clindamycin all by itself, so all on its own. So, that's kind of the meat of the ototoxic research that I wanted to present to you.

Now I really want to talk about how does this affect our EHDI systems and how is this, what can we do with this

information. If you wouldn't mind, Will, posting the poll, I would like some, I have two questions that I wanted to pose to the audience today about, one is, um, let's see, there it is. Okay, what is your monitoring schedule based on? Is it based on audiology recommendations? Physician recommendations? Do they come from state EHDI programs? I'm interested to see what kind of information. The other one is does your state monitor and track risk factors? We'll give it a second so I can see some of the information here. Looks like there are a significant amount, let's see, more than not are saying that your states do track risks, which is awesome to hear We kind of have a mix here. We have a lot of state EHDI recommendations for monitoring schedules, and we have a lot of audiology recommendations, a few medical home, which is good, that they're involved, so, great. Okay, so now let's, what our program does here and kind of how we have developed our risk monitoring program, and hopefully, that's going to help guide some of you on your programs, and we're always open to hear what other programs are doing too, to see if we need to add, you know, change anything that we're doing here in Idaho.

So, um, what is the goal of a risk monitoring program? These are kind of basic things, but I think we need to know them. We need to figure out how to identify the kids who have risk factors, and we'll talk about how we can do that, but that's really the key. If we don't know that they're out there, we don't know the risk factors, we're not going to be able to find these kiddos. Timely

diagnostic assessments by a pediatric audiologist, and then I really feel a huge part of this is being able to maintain and track the data, because without this, without our data from Idaho, we don't have a back to stand on when I go to the NICU doctor and say we really need to track this risk factor. They ask why, and, I mean, we need the information so that we can tell them why we need to track these kiddos. Okay, so, a risk monitoring program, it really looks at all these components. We need the birthing centers, birthing hospitals, to be involved, we need our medical homes to be involved, the pediatric audiologist in your state and the EHDI program. So, it looks like it's a cycle here, but, actually, these all should intertwine. We really should have them all working together. Let's talk about birthing hospitals first. What is their role? So, I mean, they're huge in this, because they have to be there with these infants and they have to identify them for us, so if our birthing hospitals aren't onboard and if they're not trained, we're not going to be able to get these kids to the right monitoring schedules.

So, first, we need to identify them. Those hospitals need to provide the family with recommendations, meaning, you know, you need to follow-up with an audiologist. They also, though, need to provide them with the why. I see families all the time that come in and say I don't know why we're here, and I always hope that the NICU, or whoever's referring them, has given them that information, and maybe they are, but a lot of times, families just have no idea

why they're coming in, they just say their doctor sent them, which is also good, but we want the families to leave the hospital with an understanding of why they should follow-up, because if they don't have that understanding, why would they follow-up? We need to train our staff in the hospitals to give them basic information on why they need to follow-up. We need the hospitals to provide the medical home with the information so that if the family's not following up, that the hospital has given the information to the pediatrician, that pediatrician can be our backup, they are the backups to say you need to follow-up with this because of this risk indicator. We need those hospitals to report to the EHDI program. That's huge.

So, what do we do here in Idaho and how have we made our program successful? We provide training. We provide training for physicians in the hospitals, the nurse managers in the hospitals, the nurses who are screening, the screening programs. You know, if we have an outside program company coming in to do screenings, we provide them training. We provide midwife trainings. So, we just bombard these entities with training, because if they don't know the risk factors and they don't understand the implications of missing a child, they're not going to make those referrals for us, and we definitely have, since 2007 statement, when we started doing our big pushes on training on the risk factors from 2007, we have so many physicians and practices that have got onboard with us. I have one physician that she, when

she has a new doctor come on, she comes and hangs up all the high-risk information on their desk, so it's so important to get these physicians and hospitals to buy into what we're doing. We train hospital staff on what to say to families. So, for example, this is a script that we provide some of the hospitals, just saying your baby has been identified with a risk indicator, and we would recommend that they follow-up.

This has been edited a little bit since I put this in here, but you know, we just recommend that they have a script available for them so that they're not stumbling on their words and trying to figure out how to explain this to a family, which I think is a lot of, it's difficult for screeners, when they don't know what to say. We provide the hospitals with referral forms so that they can complete a referral form with the family, they can show them this box and say we're checking this off, this is a risk indicator for hearing loss for your child, and we recommend further testing. Another, and we're going to go into this a little bit more in-depth, but I just want to show you, these are just some of the things we provide to those hospitals. Let's talk about medical homes. They're a big part of this, because, again, we get them out of the hospital, but this is the person who they're following up with regularly, so we need the medical home to understand this. We want them to be familiar with the risk factors, we want them to understand, you know, if they pass the screening but they also have received this medication, or they have a cranial facial anomaly,

that there is further testing that we would recommend for them. We want those medical homes to encourage follow-ups, because without them, if the medical home is saying, oh, your child is fine, they can hear just fine, those families aren't coming in for those follow-up testings, and then we really encourage those families, we really want the medical home to encourage those families to follow-up with a pediatric audiologist who is trained in pediatrics. We want them to be seeing somebody who knows how to do testing so that we're identifying the kids correctly.

So, when we train physicians, this is actually what I was talking about, this, I have, one of the physicians I know really well, she hangs, she's actually a pediatrician teaching at the family medicine clinic in our town, and she hangs this up on every new resident's board when they come in the door, you need to know this, because you are going to see that come through our hospital. So, you know, if we can get physicians to buy into this, we definitely are going a long way in that we just have another person helping these kids get into the right places. So, pediatric audiologists, their role is, it's huge. Diagnostic testing, we need them to do appropriate testing, we need them to understand which risk factors are more concerning than others based on the JCIH 2007, and we need the audiologist to provide documentation. So, you know, if they, if all the right stuff happens, the family's referred by the hospital, the medical home refers and then they get to the audiologist and the audiologist diagnoses a hearing loss in

a child who received gentamycin, but that audiologist never reports it to EHDI, we've lost that component of our program, so it's really important that we train audiologists on how to report to us. We have a pretty good follow-up program. If we're not getting the information back at the state EHDI program, the EHDI program is contacting the audiologist to get results, we need that information so we can continue to improve our program for risk monitoring.

So, in an audiology clinic, this is just kind of an example. Our clinic, our name changed a year ago, but this is some data from our clinic, but we have five clinics in the Boise area, so it was across southwest eastern Idaho, and we had 20 audiologists. Kind of what we did there is, at the time, we were tracking at our state EHDI program, but we were also tracking it within our pediatric audiology clinic, and we were looking at how many hospitals for referring and how could we educate the people that were referring to us better, and you can see, what's going to, I'm just going to show you that everything increased. Every couple years, we just got more things onboard. Babies getting referred, 2007, only a couple hundred, to almost a thousand babies getting referred in 2010. That's because of training, and that's because of education, and that's what's huge. If we can educate physicians, hospitals, nurses, on this monitoring, that's how we get those referrals, and that's how we help identify these babies. Obviously, those neonatal indicators, those are the ones we were seeing the most. 2010, the reason those numbers kind of went down

is because this data was pulled halfway through that year. We used a tracking system on an Excel spreadsheet, so it doesn't have to be fancy. You can track data like this, and we just tracked which babies were coming in, which babies we lost to follow-up, which ones had hearing loss, which ones had conductive versus sensory neural, and then we would pull information out of our risk factors and which hearing losses we were identifying. It's all about tracking outcomes, looking at, you know, where our lost follow-ups were, sensory neural versus conductive. We just did a lot of tracking and a lot of education from the data that we collected within our audiology program. Obviously, we still have, and I'll show you later, for our Idaho data, we still have a long way to go for our loss to follow-up. Unfortunately, high-risk is a hard population, but we do diagnose hearing loss in this population.

Now, let's talk about EHDI, and I saw there's lots of EHDI coordinators here, so this is really for you guys. This is what we need you guys to do in your programs, is provide that training and support. Without EHDI programs providing that, that's really how the program's going to work the best. Need to know, um, what, you know, who are we training, are we training hospitals, physicians, you know, use your pediatric audiologist in the state, if they're interested in providing training for hospitals, that's how our EHDI program works. We have audiologists that do trainings for our EHDI program all over the state. We also want those EHDI programs to provide information back. So, we can't

just train them and give them the information just on the front end, we need to tell them how they're doing, we need to tell them, you know, you guys refer this many kids for ototoxic medications, and you diagnosed the hearing loss. That's the stuff that the hospitals and the physicians and the nurses, that's what gets them fired up and gets them onboard with your program, is when they are, they understand that they're making a difference in a child's life by referring them, and then the tracking surveillance. So, our EHDI programs, I showed this a little bit earlier, this is how we collect our data from the hospital.

We also have a form that we collect data from audiology programs, and I'll show you that in a second. Our data on high-risk, so the prevalence of high-risk, we have 3.1 infants reported for high-risk in 2007, and then after training, after we started going out and training on the risk factors from JCIH 2007 position statement, that number jumped up to over 11 percent of infants were getting referred from our births in that year with risk indicators. So, you can see how much training and education can really impact who's getting to the audiology centers and who's getting referred and how the information is getting referred. This just looks at the number of risk factors that were reported in that same time period. Definitely, you know, we use a high track reporting system, so our reporting system kind of change a few years ago, we used to have ototoxic medications kind of listed out on its own, and now it's kind of lumped into this, but so if you look at

the purple, purple is where that ototoxic medications would fall into. So, neonatal indicators typically is an NICU stay and an ototoxic medication given, which is almost 80 percent of the children in this number.

So, if we looked at another date range, 2008, January 2008 through December of 2014, we had about 4700 infants who passed their newborn hearing screening program but received ototoxic medication, and that was the only risk factor they had. So, well, potentially, they had that five-day NICU stay, and unfortunately, we can't rule that one out of there because of how our tracking system is, but they didn't have mechanical ventilation, they didn't have syndromes, they didn't have anything else, just the ototoxic medication, and of those 4700 infants, 2 of them have been diagnosed, thus far, with a delayed onset hearing loss, sensory neural, and without our program, these children would not have been picked up at 9 months old like they were, and both of them are doing awesome today, so it's real exciting that we do find these kids, and you can see, unfortunately, there's going to be a lot of kids who have normal audios, but we do catch these kids, and I think if I could have that, the parents of the severe unilateral talk to you about how appreciative she is that we found out, it's really moving to hear families being so excited about the program and how happy they are that it worked for their kid. So, at our state EHDI program, we also provide recommendations for the audiology practices on appropriate testing. We all know that, you know,

audiologists, we have audiologists who see birth to 100, and so not all of them are seeing children all the time, and some of them may need some guidance on what is appropriate testing for these children, so we provide a best practice protocol for the audiologists in our state.

This is a little bit of information. It kind of supports some information out of JCIH position statement earlier about, we have about 50 percent of the kids that we see that have risk indicators that have hearing loss. Again, there's going to be 50 percent that don't, but 50 percent of the kids we see have some risk indicator. Some more Idaho data, this is why we do what we do, is to collect the data to give information on it, but 2.7 infants per 10,000 were diagnosed with a delayed onset hearing loss, and those infants had multiple risk factors, actually over 70 percent of the infants we examined had multiple risk factors. Just a few infants were only receiving, you know, we have some that have ototoxic meds, but they also had mechanical ventilation and a syndrome associated, but multiple risk factors were reported in this data. Let's see. So, let's look at the guidelines again, and we're going to just look at these. This is that same worksheet, but we're going to single them out. One of the things we did in our state was we looked at, you know, there are risk factors that are more concerning, and JCIH 2007 states that. We put those risk factors and we put those into two categories. The ones that are more concerning, we call them class A, and we actually developed

this with NICU staff who uses class A, B and C system in their NICU, so they said class A to them was always more concerning, so we put our more concerning risk factors as class A, and we have class B indicators, and this was where ototoxic exposure would fall into, and these class B risk indicators, we recommend that they come in for an evaluation before their first birthday.

Now, when we started this, we kind of promoted the nine-month age so we could get behavioral testing and otoacoustic admissions, but we see, depending on the pediatrician and the practice, we see the referrals at all different ages. We just want them to come in before their first birthday, and preferably at an age where we can do some behavioral testing. So, um, when we do get referrals when kids are two months old, we usually contact the pediatrician and ask them to refer back at a later time, and if the pediatrician has concerns, we will see them earlier. So, some of the data from this class A versus class B risk indicators, the reason we did this is because we wanted those class A ones, we want them to come in sooner, because they have more concerning risk factors, and we want to see if we're finding hearing loss in them sooner, so we recommend they come in by three months of age, if they have a cleft palate, if they have a syndrome associated with hearing loss, meningitis, which we would hope that everybody would get referred for anyway, but this is the data we've collected from October 2011 to May 2014. Out of 153 infants, our loss to follow-up rate is, unfortunately, really high, but we did diagnose hearing

loss by three months of age in 16 percent of the kids we saw. If we look at the individual risk factors from this data, post-natal infections, of the 18, 22 percent had hearing loss. We did have some normal hearing, but definitely a high percentage. 50 percent of the kids we tested, and you can see, there's 22 percent we didn't test, had a hearing loss educationally significant. Cranial facial anomalies, which we had 23 kids, which was 20 percent, that had normal hearing, and 12 kiddos that had hearing loss, so 11 percent, and these cranial facial anomalies, they varied from cleft lip and cleft palate with the hearing loss kiddos, and we actually had five kiddos in that hearing loss category that had ear tags and nothing else. Some things to remember is your risk monitoring programs, they need participation from all participating entities, like hospitals, audiologists, medical homes, EHDI programs. Training from the state EHDI programs is huge. Unfortunately, there's not a gold standard for protocols. We use class A and class B, so for our ototoxic medications only, I'm not sure if anybody can see the screen, because I can't see my screen anymore, but I'm going to keep talking, for our ototoxic medications only, we see those kiddos at nine months of age, and we recommend that they come in so we can diagnose hearing loss before their first birthday, if there is a hearing loss.

>> Jessica, tell me what you're seeing on your screen. Did you get booted off, maybe?

>> I'm not sure. My screen went blank. I have a hard

copy, so I can just, can everybody else see my screen?

>> I believe so. Let's see. I moved on to case number one.

>> Okay, I will talk about case number one, and honestly, you know, I can just kind of give brief summaries of this. I can't flip through the slides, but case number one, this is a reason why our program, at the time, didn't work. So, this is in early 2014, before we started tracking ototoxic monitoring and getting these referrals, I saw this child, she was four years old, she was referred for speech delays, her birth history, she had, you know, a prematurity, a NICU stay, and no mechanical ventilation, but the ototoxic meds, and she was referred for testing, oh, nope, I still can't see it. The child ended up having a unilateral high frequency hearing loss. I can't advance the slides, Will, so I'll just talk about it, and we didn't get it identified until much, oh, hang on, here we go. Now my computer's working. This is her audiometry at four years old. You know, CPA, she was a difficult to test child, but here were her OAEs, which are very suspicious. We had these really funky ones in her right ear, so something wasn't quite right, and I recommended that they came back, and unfortunately, so she's already four at this point, has significant speech delays, she wasn't referred for ototoxic meds at the beginning, in the hospital, so three years later, she comes back in, and this is her audiogram.

Now, granted, looking at this, this is not a

substantial amount of hearing loss, but in a child who could we have identified it sooner, potentially, had we had her coming in for ototoxic monitoring at nine months of age, just a thought to pose, could we have caught it sooner? And I'm just going to go forward here a little bit since we lost a little time there. Case number two, this is why we do it, and this is this mom I was talking about earlier. This child passed her newborn hearing screening, she was born premature at 35 weeks, NICU stay for less than five days, so she didn't meet that criteria, she had ototoxic medications, and nothing else in her history. She was seen for an audiology evaluation, wasn't super successful, so they referred for ABR. This was her OAE testing, that's why they were concerned, that left ear, the OAE being absent. This is her ABR. Left ear's on the left side of the screen, right ear's on the right, and, so, this is looking at some tracings, comparing the two sides here. Good morphology from the right ear, poor from the left. This child was identified with this hearing loss at ten months old, and had we not had our referral program and had her physician onboard making that referral for follow-up testing because of the ototoxic meds, and the hospital doing all their right steps, this child wouldn't have been identified probably until kindergarten when they had the kindergarten screening, and I'm just going to skip ahead. This is her hearing loss now. She's three years old, she is on target, she's age appropriate, she wears a hearing aid, and she is doing excellent. So, our program has really given us some successes in

that we feel we'd like to share with our states, that we can hopefully, you know, collaborate with other states, but also give other states some guidance on how we setup our program and how we've made it work in Idaho. So, I apologize for the technical difficulties. I'm not sure what happened to my computer there for a little bit, but if we have some time for questions, now would be the great time. Hopefully, my computer will continue to work.

>> So, I've opened up the Q & A field over on the left-hand corner for our participants to insert their questions, and it occurs to me, while we're waiting for some questions to come in, that one of the things, because I think many people on the phone know that I have a real interest in continuance and periodic screening throughout early childhood, that your experiences and your research here is actually supporting the importance of having multiple ways to continue to monitor children throughout this early childhood period. You know, like, with this last case, if there were preschool programs or healthcare providers who were continuing to do monitoring of hearing, that child wouldn't have had to wait until kindergarten, if this initial follow-up hadn't occurred. So, it's just a great illustration of the value of having multiple nets to catch children. So, let's see, we've got a question here. Somebody's asking about getting slides, and, yes, we'll be sharing those, as well as having the power point being displayed on infanthearing.org. The next question is does your hospital use a contracted in-patient newborn hearing screen vendor?

>> Okay, I'm going to assume you're talking about a program such as, like, pediatrics, is that correct?

>> Well, I think you should take your best guess.

>> Okay, so, um, we, actually, in January, or it might have been November of last year, the hospital that I helped manage the newborn screening program for years, they actually switched to a vendor who came in and took over for the screeners. You know, I had been working in conjunction with the screeners in the NICU for a long time and was up there once a week, it's definitely been a change, but we actually have done lots of training with the new vendor that has come in to the hospital, they are onboard with our program. We have some kinks that we're still trying to work out, because the vendor was not, it wasn't a typical process for them to screen for high-risk and to make those referrals like we were requesting them to do, so we have definitely had to have some work there on trying to get those vendors, or I guess outside, that hospital staff onboard with high-risk monitoring, but we've definitely been successful in several hospitals in our state of getting the vendors trained and making referrals and getting those referrals in.

>> Great. There's another question for you there that you can see.

>> Let's see. Okay, how can we help encourage PCPs and hospitals to use high-risk referral processes in addition to newborn hearing screening? That's a good question. I mean, what

we did is we brought, we setup multiple trainings for physicians and hospital staff on risk monitoring, we presented data from our state, we presented JCIH position statements and gave them information. I think, probably, one of the more powerful things is explaining to them cases. If you can give them a, you know, this child came through your hospital and, you know, unfortunately, sometimes, you have to give them the negative cases for them to understand, you know, when I train on just newborn hearing screening, I always tell them, you know, I have this child that I identified at three with a hearing loss, and he came from your hospital, and he was screened 32 times, and that puts a pretty big impact on that hospital staff, when they recognize something may have went wrong. You know, we don't want to bring up those cases, you know, we just try and give them the more positive examples, but sometimes, that's how we have to get physicians onboard too, you know, is by explaining sometimes things get missed, and if we can do it on the front-end, we might not miss these children.

Okay, let's see, where are we at here? The clarification to the JCIH changes recommendation to not monitor children with ototoxic medications of any amount, okay, I'm not understanding that question. Sorry. It kind of looks like a statement. I'm not sure what's being asked there. We'll go to the next question, and you can e-mail me, if you want to explain your question. I am a pediatric audiologist at a children's hospital. Would you be willing to share some of the documents from your slides

that you give to your physicians in hospitals? Absolutely. We would love to share our information. You know, we, that's why we collect our data, that's why I helped write the risk factors. I feel like the more information we can have to support what we're doing, the better, and if we can get this into other hospitals and you guys can start collecting data, the more information we have, the better. So, yes, just e-mail me and I will send you whatever documentation or slides you would like. Does your facility continue to monitor OAEs when children are able to provide behavioral thresholds? So, here's kind of, you know, I work at one of the bigger audiology facilities in our state, we have several audiologists in our practice, and, you know, we have these recommended guidelines that our state EHDI program provides to audiologists. We still don't have a, we hope that we have a consistent test protocol being performed across the state. I can tell you that from reviewing cases that we've seen in our state, they don't all do behavioral testing, they don't all do OAE testing. Unfortunately, that is a problem. In our practice, we do ototoxic monitoring by way of otoacoustic emissions and behavioral testing to support that. So, if we have a child who has OAEs that aren't looking good and we do testing and it's normal, we get them in the booth for behavioral testing. We continue to monitor the OAEs, so if that child had normal hearing, we'd bring that child back because of the OAEs, and again, this is going to be all case-specific and probably audiologist-specific, depending on which audiologist

we're talking about in the practice.

Okay, sorry. Let's see. With ototoxic meds in the newborn period only in NICU, at what age do you feel safe that hearing is now stable? So, how we have managed that in our state is if we're just talking about an ototoxic medication case, is that if they come in for their before one year check, so the class B indicator referrals that we have, we recommend by one year of age, we typically see them at about nine months of age, depending on their gestational age, it could be closer to twelve months, but when we see them back, if they pass their OAEs, they have robust OAEs bilaterally, we then discharge them, so at that one-year mark, and like I had mentioned from that triple AAA ototoxic, they said within a few months after they've stopped receiving the medication, so we discontinue at a year if there's no concerns from the pediatrician, there's no concerns from the parents, and everything checks out okay at that appointment. Are we still taking questions?

>> You can wrap it up. We weren't able to get all the questions, unfortunately, but there were many, many more coming in, but I think you're open to hearing from folks, if they'd like to discuss or e-mail their thoughts or questions to you, is that correct?

>> Yes. Absolutely. My e-mail, that is a perfect way to get ahold of me. Mm-hmm.

>> Okay, great. Well, Jessica, thank you so much for your time and expertise today. We very much appreciate it, and

thank you, everybody, for attending today's webinar. It will be posted on infanthearing.org within the next week. Thank you.

>> Thanks.