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NCHAM

RISK MONITORING FOR DELAYED – ONSET HEARING LOSS IN YOUNG CHILDREN

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While we wait, if you could give us a little bit of indication about your particular background so that our presenter has an opportunity to have a sense of the perspective that you bring to this webinar, that would be great.

Thank you everybody for responding so quickly to that poll question.

If you've just joined, we haven't started yet. We just posted poll. We will be starting at the bottom of the hour which is just in about two minutes. If you can take a moment to indicate what your perspective is, what roll you bring that prompted you to participate in today's webinar, that gives our presenter a little bit of a context around who it is we have joining us today. So thank you for those of us who are doing that.

I'm going to initiate the record of this meet something that we can get started. Hold on one moment.

>> OPERATOR: Audio recording for this meeting has begun.

>> Good day, everyone. I'd like to welcome you to today's webinar, brought you to the national center for hearing assessment and management, also known as NCHAM, at Utah state university. NCHAM serves as a national resource center on early detection and intervention. And as a part of our goal, we like to provide resources and information. And today's webinar is just one of those. Today's webinar is entitled risk monitoring for delayed-onset hearing loss in young children who will be -- which will be presented by Dr. Jessica Stich Hennen.

Jessica is from Idaho, and she's a pediatric audiologist at the Idaho elks hearing and balance center in Boise. She received her doctorate in Idaho, her AUD, from Idaho State University. She also had a clinical specialty area which includes pediatrics, diagnostics and amplification, auditory evoked potentials, and central auditory processing disorders.

In April of 2011, she achieved specialty certification in pediatric audiology or in the PASK from the American Board of audiology and she is the primary audiologist also for the Idaho cleft palette craniofacial team. So we're really delighted to have you with us here, Jessica. And without further ado, I will turn it over to you.

And just as a reminder for everybody, you will have an opportunity to ask questions once Jessica has completed her presentation and at that point I'll display a text screen into which you'll be able to type your questions.

So Jessica.

>> Yes. Well --

>> We're ready to start.

>> Well good afternoon, good morning, wherever you guys are calling in from. I'd like to thank NHCAM for inviting me back to present. Some of you may have attended a presentation I did back in October on oex toe toxic monitoring, and I've done a similar presentation this past year at EDI, so if you've attempted those, some of this might be repetitive. There's definitely some new information that I've added into the presentation. But I'm hoping I can teach you something and hopefully we can have some good discussion at the end on how we can continue to monitor risk indicators and how to improve our program in Idaho and how to hopefully start some programs across the country. So we'll get started.

Let's see here. There we go. This is just a disclaimer for my presentation.

So the learning objectives today is I want to identify risk indicators that require or should require some kind of monitoring for a delayed-onset hearing loss; and differentiating between ones that need a little bit more or frequent monitoring, more risky risk indicators, and then also give some options for risk monitoring protocols.

With regards to the first ones, I'm really going to present some current research and some publications which give support to why

we should monitor risks, but it also does create some of the publications and research are going to create some confusion, because a lot of the information out there, there's some publications that are going to recommend monitoring for certain risk indicators, where another one might have found that risk indicator to not have been so concerning. So and I'll explain that as we go along.

So to start off, the joint committee on infant hearing, we all know was established in 1969, and it comprises of these three groups. And their goal is not to identify risk indicators but it was really to identify hearing loss in young infants and young children. But as we know, at that time, there really wasn't a good screening method available for hearing screening in young infants and young children, so their goal at that time was really to review research and to do research to find out what was a way to monitor and detect hearing loss in young infants.

So with the publications that came out, starting in the 1970s, they -- the joint committee started identifying high risk criteria, and was what that meant was these are indicators that a child might have or is at risk for developing hearing loss. In their first publication based on research that was available at that time, they identified five risk indicators, including

family history, a fetal information, defects of the nose, ears or throat, low birthweight and high bilirubin levels. At that time, there wasn't really a really good screening technique available, and there wasn't really a good recommendation for what to do with these children if they had these, their statement in that -- in the position statement said if the child's hearing appeared to be normal but the infant fell in one of these categories, they should receive a regular hearing evaluation at their physician's office, well baby clinic or audio logic center, but it wasn't very strict, it didn't say how often, at what age. It just gave a very generic recommendation for these children to follow-up.

A little bit later in 1982, the criteria grew a little bit, because they added in bacterial meningitis and severe asphyxia, and at that time, those were added because of some research that had came out showing hearing loss being a risk in some of those infants that had developed those.

With their screening recommendations, they had said, ideally, they'll have -- the infant would have a hearing evaluation by three months of age, under the supervision of an audiologist but not necessarily by an audiologist, and there could be either behavioral testing or electrophysiologic testing to measure their hearing. And we all know that under three months, a

behavioral evaluation is not very reliable for a child to determine hearing levels.

So, again, at this time, there's there still wasn't really a good way to screen for hearing loss and high risk hearing loss in infants, and we still didn't have newborn hearing screening at this point either.

Again, in a few years later, the list kept growing, and they kept adding risk indicators. At that time, several studies had been coming out about oat toe toxicity and ototoxic medications. Syndromes was added to the list, head traumas, caregiver concern. But at this time, there was a good change where it said that this screening did need to include an ABR measurement, not just behavioral testing, so they recognized that they needed to do a more thorough type of assessment on these kids who were at risk for developing hearing loss in infancy.

In this statement, they also did pose a concern about maternal drug abuse, but they didn't give a clear recommendation, and they didn't specifically add it to their criteria for high risk indicators.

So in 1994, studies have shown at that time, several studies, which are listed there, have shown that 50% of those with

childhood hearing loss could be identified using a high risk register, but we all know, and why newborn screening was developed, is that we're still going to be missing half of the children with a hearing loss discovered in infancy or young childhood. So at this time there was a little bit more research coming out, but it did give a strong indicator that we should be using some kind of a high risk register or there are risk indicators that do need to be monitored in these young children.

In the 2000 statement, all these risk indicators I had mentioned in the previous slides also in the position statement to be monitored. The change in that statement was they gave a pretty strict criteria for what they had recommended for testing, and they had said at that point if a child had a risk indicator, and they list them out in the position statement, audio logic testing should be performed every six months until the age of three years. So which, at that time, when that statement came out, it really did bombard the audiology practices and system with high risk referrals, meaning if a child was identified with ototoxic exposure, the recommendation was to test every six months until the age of three based on that statement. And so it did give us a little bit more strict criteria in that statement, whereas you'll see that in 2007, things started to change a little bit.

So our 2007 statement, which is what we're operating under currently, until the next statement comes out, gave us a new definition of targeted hearing loss to think about. And that's the neural hearing loss in infants, they're more at risk when they are in the NICU population, and then giving us this indication that we do need to screen those babies that are in that higher risk population with ABR in the hospitals and not be screening with otoacoustic emissions.

And then if you think back to the previous slide from the 2000 statement, where I had said audio logical recommendations were to monitor every six months until the age of three, the 2007 statement here, the monitoring of high risk indicators, infants with risk indicators for hearing loss, should have at least one evaluation by 24 to 30 months of age. That's a pretty big change in what the recommendation was. And so I think that that definitely changed a lot of practices and how audiologists thought about monitoring risk indicators.

I just put the other kind of highlight for that position statement with the regard to risk indicators. Babies that are readmitted within the first month of life, that present with a condition associated with hearing loss, or potential hearing loss needed a repeat screening.

Now, in Idaho, we actually also have our hospital ffshgs a child is readmitted and has a risk indicator, which they'd get the screening then, we actually also have our hospitals referring if they pass the hearing screening referring for outpatient follow-up based on the risk indicator present. So we, and I'll explain our program a little bit more, but just so you're aware that we have taken that one in Idaho and said we need to also have our hospital looking at risk indicators that are present for delayed onset hearing loss in these readmission population as well.

And this is just appendix from JCIH2007, our current risk indicator list. You can see that neo natal incentive care really does cover kind of a large range of risk indicators which are typically found in a premature population but can also be found in full term infants. But that's why that one is kind of clumped together there in that statement.

So let's talk about some risk indicator monitoring kind of data. So this is just kind of an interesting number. The incidence of risk factors for hearing loss and how often they're occurring in the population. So this study in 1989 had reported a 10 to 12% of all babies born will have at least one risk indicator, risk factor present, and I'll show new a later slide from Idaho we actually are pretty on track with that number. I believe ours

in is in the 11%. So even 20 years later we're still kind of right on track with that number.

Another study out of NIH in the 1990s had shown that in the NICU population, approximately 33% of the NICU babies in their study had one risk indicator but almost 30% had two or more risk indicators. So that NICU population definitely, in this number, is going to have a higher amount of risk indicators present than a well baby population.

So when we look at risk indicators and look at how often they're occurring in a population, and definitely if any of you are monitoring risk indicators in any of your programs, you'll know that most of the referrals, and I know our program is this way, are coming from the ototoxic medication that is come out of the NICU programs. 70% or greater are coming from -- with ototoxic as the number one risk indicator on their form.

And you can see on that left hand side that those are the one that is are occurring the most frequent. With the other side is the risk indicators that don't occur as often. So we have less kids that are being born with family history of hearing loss as compared to that 70% of risk indicators of receiving ototoxic medications.

Now, if you think about this slide and you can see that difference between which ones occur often and which ones occur less often, then we look at how often does hearing loss occur among these high risk indicators.

And I'm going to go back and forth for just a second here. Cranio facial abnormal as is not occurring as often but as the highest amount of hearing loss associated with it.

Same thing with treatments. It is occurring on the left side of the graph but still not a significant number as compared to ototoxic medications. And so you can see ototoxic medications, it didn't even make the list of how high a percentage of hearing loss among the risk indicators. So these slides I feel like are give us a almost good idea of, you know, we're going to be in a program, you're going to have a lot of referrals for things like ototoxic monitoring and you're going to find less hearing loss but those kids that you're getting with, like a cleft palette, for example, you're probably going to find more hearing loss in that population. And I'll show you some data that we have on just that happening.

So let's talk about ototoxic medications because they are the number one reason that we get referrals in our state, and that's what data has also suggested. There's over 200 known ototoxic

medications. They're used to treat serious infections, cancer, heart disease. You know, we're more concerned about because that's the what's used in the NICU to treat infections that are hard resistant.

And the other thing about aminoglycosides is we don't know, every one that they could be at risk for, there are different effects coming from the medication so, we don't always, you know, it is not necessarily the same as vankamisin but we have to treat them all the same when we're telling a hospital program to make referrals. So I just put this little interesting from Triple A's position statement which kind of gives you some idea of why this having a program is tricky because there's not clear-cut guidelines on risk monitoring, particularly with ototoxic medications. But the Triple A position statement regarding ototoxic monitoring said with regards to aminoglycosides weekly or biweekly monitoring is recommended ideally. And then follow-up testing should also be schedule add few months after the drug has been discontinued.

So I know that if we were having children sent from our hospital programs for weekly or biweekly monitoring, it would be very difficult to get that amount of kids tested in an appropriate amount of time. I do agree with the part that we do need to reach a few months after the discontinuation of the drug, and we

know for longer has effects longer than the -- after the medication has stopped.

So gentamicin was introduced in the 1960s. It is the most common aminoglycoside used in NICU program because of its low cost and the effectiveness against Gram negative bacteria.

All right. So this is a publication that came out of ASHA in 2010. It was a literature review on 20 studies regarding gent my sin and looking at hearing related loss related to this. What they did is re-reviewed these studies in terms of dosage, dosing schedules, routes of administration of the drug, so topical versus IV, and they tried to draw some conclusions on, number one, was the persistence of hearing loss in persons treated with gentamicin and, two, is there evidence of a synergistic effect on hearing loss of multiple ototoxic drugs when they are taken.

Unfortunately what they found is that there was a small number of participants across all of these studies, and a very large -- there was a large variability among the hearing loss definitions in all the studies. So their conclusions were kind of all over the board with some studies reporting hearing loss as low as 0%, and others reporting hearing loss related to gentamicin as high as 58%. And it really was because the

studies were so different in how and what criteria they were using, diagnostic criteria for diagnosing hearing loss, what their definition of hearing loss was, and even the patient populations. Some of the studies were pediatrics, and some were on adults.

There was some good that came out of the study, and they did note some trends, and their trends that they noted were the frequency of administration of gentamicin did not influence the likelihood of hearing loss, and I'll explain why in a little bit. And then the dosing amount also didn't influence the likelihood of their being a hearing loss.

Now, the reason that may be is because of the genetic mutation A1555G that has been identified that may have an amino glycoside deafness associated with it. So dosing and frequency would not make a difference if the theory is true that you receive one dose of gentamicin and it could change your hearing.

Now, this mutation, there's been some studies on this mutation that I'm going to talk about. The first one up there reported on the mutation, and it is associated with amino glycoside deafness. It was found in this particular publication that even one single dose may result in a sensory neural hearing loss.

Where the second study in 1998 had reported that a profound hearing loss would -- in individuals with a mutation that had a profound hearing loss, they didn't have any amino glycoside treatments. So it's kind of two conflicting studies because one is saying if you have it, the mutation, and you receive the medication, you could lose hearing. And the other one is saying you could have this mutation and have a hearing loss and not have had that exposure to the amino glycosides.

Some information on the number of mutations noted in the couple studies. The first study out of the UK in 2002 found that one out of 206 newborns have expressed the mutation, where in a study that came out of Texas had said one in about 1,100 have this mutation. So you can see all of these studies kind of give us different ideas on how to monitor ototoxicity and should we be concerned and, you know, what is the level of concern we should have regarding ototoxic medications specifically regarding gentamicin and the NICU population receiving them.

All right. A little bit more on ototoxicity. There's a um could of studies, a study out of Iowa children's that they have 700 infants, only 1.8 expressed the gene, and none of those had hearing loss.

There is -- they had a theory in that publication regarding amino glycoside and noise exposure presented in the NICU, being together, with more susceptibility to hearing loss. But again, it was just a theory. They didn't have any evidence to support that.

So NICU stay, as a risk indicator. This is, again, one of those high number of referrals we get. A lot of times, though, the NICU stay referral on its own -- or sorry, not on its own. A lot of times NICU referral of greater than five days is accompanied by that ototoxic risk indicator as well.

So the reason that JCIH had add this had to their statement was they found that the children that were discharged by five days of old were considered a more lower risk population for hearing loss versus the children that were in there for greater than five days. And those greater than five days tend to be the very premature, very low birthrate that are in there for much longer.

This study out of -- from 2014 had noted that a NICU stay of greater than five days and exposure to loop diuretics, they found it was not associated with an increased risk of hearing loss. So, again, just conflicting data for if a NICU stay on its own is a good one to monitor or not.

So if he with talk a little bit about me can tal ventilation, these are just another few studies out there. This first one from 2000 had estimated that one in 56 children would have permanent hearing loss by age one if they had received -- or if they had respiratory distress syndrome, bronchial display I can't and they were on me can tal ventilation for greater than 36 days. So these were pretty sick kids and the kids that had severe respiratory distress and respiratory issues, there's a significantly greater chance of them having a hearing loss by age one.

The study from 2002 from Robertson, they found that 50% of severe neo natal respiratory survivors had hearing loss by age four. And many of those patients in that #12udy didn't develop the hearing loss until age two to four years of age. So in that case they're saying that a severe respiratory kid dose are more -- are risk for hearing loss but they're going to develop it much later, two to four years, and they don't -- and in this study they didn't know the true cause of the hearing loss, and the tricky thing is, a child with severe respiratory distress and is in the NICU and receives all of those medications, at that point they're going to have multiple risk indicators that we have to be concerned about for developing hearing loss.

And then the Beswick study in 2013, they also found a correlation between post natal hearing loss and me mechanical ventilation of greater than five days which in the 2007 doesn't have a specific amount of time on me can tal ventilation noted in the physician statement for monitoring purposes.

So ECMO treatments, this is a very aggressive medical treatment that's used for life support in infants who are in respiratory or cardio pulmonary failure. These particular kids are pretty sick, and they're going to have multiple risk indicators not mechanical ventilation. They're potentially going to have oto -- well, likely, ototoxic medication, potentially prematurity, low birthweight, there could be multiple different reasons for why these children are at higher risk for hearing loss. Probably the study we all know of the best is the one out of 2008 that Brian published and it looked at 111 neonates from their research out of Boston children's, and they found that these children were at significant risk for hearing loss. So congenital hernia raced the risk for hearing loss 2.6 times over the regular child. Aminoglycoside antibiotics of 14 days or more raises the risk of 5.5 times. And a child that received ECMO treatment for 160 hours raise it had by 7.1 times. So just a significant risk for these kid dose. One of their findings in that study had said if a child had 14 days or more of amino glycosides over 80% of the children in that study had a sensory

neural hearing loss by age 2.5. So ECMO treatments alone, and, again, these kids are going to be very -- they're going to have multiple risk factors presenting. Which would definitely put them at risk for monitoring.

So syndromes, and I'm not going to go through each individual syndrome because this is just a short list of all the syndromes that are associated with hearing loss. But definitely looking at this list, we see, in Idaho, the most referrals for kids with downs syndrome from this list. But we do see all over the gum mutt of kids on this list. Definitely downs syndrome kids, though, are at higher risk for middle ear, conductive hearing loss but even sensory neural hearing loss as well. So definitely looking at and finding out syndromes and making referrals based on what they find out in the hospital is important.

Looking at infections. Congenital infections such as CMV, are you bell that, her piece, syphilis, toxoplasmosis. We all know that CMV is the most common congenital infection coming at about 40,000 live births per year in the United States. The tricky thing about CMV is about 10% are systemic at birth so we have a lot that are asystemic at birth that may develop hearing loss at a later time.

Cranio facial anomalies, and if you remember back to towards the beginning we talked about how they're not occurring that often in the population but the -- the risk of hearing loss is significant greater in this population, and these are just a few of the cranio facial anomalies that we encourage our hospital toss refer for. The most common one on that list that we get referrals for is ear tags and ear pits, and then cleft palette would be the other one on that list.

So why cleft palette? Why is it concerning? Well, here are several studies that show, at minimum, a 50% incidence of hearing loss with kids and/or adults with cleft palette. So you can see depending on what population they were looking at, there was varying results, but definitely a significant amount of hearing loss, no less than 50%.

Another study that I didn't put on here is and we'll talk about is the Beswick study. And that showed that those children were at significantly higher risk, nearly two times more likely to develop a hearing loss, than those without, if they had a cleft palette.

This is some data from the cleft palette team that I work on. It's a few years old. But just looking at this, showing that we have, again, 50% of our kids having some degree of hearing loss

associated with it. And you can see it's not just conductive. We have mixed hearing loss, sensory neuro hearing loss. Obviously the highest amount is conductive. But we all know that conductive hearing loss can be just as detrimental to speech and language development and education. So super important to monitor those kids as well.

Family history. So when we train our hospital staff and physicians in our state on family history, we train and encourage them to ask the right questions and try and find the family history of congenital hearing loss or acquired hearing loss of a sensory neural nature, not conductive. We, a lot of times, will see those kids that have a family history of conductive any ways if they end up in -- for -- for having conductive hearing loss themselves from middle ear fusion, but we're really looking for the permanent hearing loss kids to make referrals at birth.

Hal had reported in 2007 that family history of hearing loss is the most common risk indicate found in a well baby population. So when we go into a hospital, we train the NICU staff on -- well, we train all the staff on all the risk factors but we really stress the family history one to the well baby nursery, because those -- they're not going to have to focus on did that child receive ototoxic meds, were they on a ventilator.

In a well baby, those children aren't receiving those treatments so we really need to ask those questions about family history. And you can see the Beswick study shows, it's a risk indicator that showed it does need to be monitored, again, two times more likely to develop hearing loss than those without the history.

All right. So head trauma. This is involving a bone fracture that is requires hospitalization, and it may result in facial paralysis, hearing loss, memory perforations.

And then this last one and we're getting close to the end of the risks, and then we'll talk about some other publications. This one, these are one that is we can -- we can talk about with a newborn nursery population, but they're really not going to develop into a little bit later. Oftentimes, in Hunter syndrome usually doesn't develop or isn't diagnosed until two years of age or later, so these are ones we have to teach pediatricians and family practice physicians to be aware of these diagnosis and relation to hearing loss. So it's not just always training hospital staff. It's also training your community doctors on risk indicator monitoring as well.

So I'm just going to talk about a few publication that is have come out recently as regards to risk. With this Beswick study, 40 articles that they reviewed. I think probably the most

important thing I found out of this study was that they, again, had some risk that is they defined as being the most risky for hearing loss which was CMV, ECMO, and congenital diaphragm hernia, and, again, that persistent pulmonary hypertension, so those respiratory kid dose, congenital heart defects, so those being the highest risk for hearing loss.

Another study they came out with about a year later looked at a little over 2,000 children. They showed an incidence of 2.7, with post natal hearing loss, and their findings, they suggest, which we've kind of talked about this a little bit already, that family history, but particularly cane yo facial anomalies, those rings need to be monitored throughout childhood. Syndromes and prolonged ventilation, they also said that those ones had favorable results to be monitored. But they found that low birthweight didn't really have a significant all on its own a significant risk for monitoring purposes.

All right. The Kraft study in 2014. They were trying to estimate the Kraft version of monitoring factors, particularly looking at, you know, does NICU stay and loop diuretics, does the cost of monitoring them do, we find enough hearing loss to justify the cost. And they found in their findings that they were reporting that the NICU stay and exposure to loop diuretics

did not have an increased risk for delayed hearing loss that the cost of monitoring did not benefit by monitoring those children.

And then on this particular one, they -- they were looking at evaluating throughout childhood, so how much would the cost of evaluating throughout childhood benefit.

In Vos in 2015, a literature review, and it looked at findings on, again, risk indicators, and they -- what they did was they had a group of professionals and neuro physiologists, ENTs, pediatricians, and they looked at several studies over a 15-year period that they found, and they were trying to determine which risk indicators were of high concern, of moderate, low, and very low concerns, and this is kind of their information that they found. They feel like family history of hearing loss, syndromes, fetal alcohol syndrome, those were at high risk for developing a delayed onset hearing loss, where if you look at the bottom, the low birthrate, the low APGAR scores, the NICU stay, and they felt like ototoxic medications were a very low or low risk for developing a delayed onset hearing loss.

Now, they were using a list of risk indicators that did not, at that time, it did not include ECMO, it did not include congenital did I photograph hernia, but based on their literature review, they actually determined that they should be

added to a list for monitoring purposes. So they found ECMO and digraph -- congenital diaphragm hernia to be a risk for hearing loss that they feel should be added as a high degree.

All right. So programs. This is, in Idaho, we developed our program, we -- you know, we've been monitoring risk indicators and recommending referrals for quite some time since we developed our new birth screening program in the states. But until the 2007 we really didn't give enough education and didn't push it as hard as we have since 2007. So the goal is to identify the kids that need risk monitoring, to provide diagnostics to those children, and then to our goal of our program is to maintain and monitor a tracking system so that we have evidence of why we're monitoring risks and evidence to present of why we can continue to monitor certain risks.

So for a program, we really need everybody on board, pediatricians --

So what are the roles? The hospital role, and when we go into the hospitals to teach them, we really want them to be to really be making, you know, have a good understanding of the risks and to be making those referrals and to be giving the family the feedback on why they're getting referred. We find that if they don't -- or aren't explaining to the families why they are

getting referred for risk monitoring, the families aren't following up. So but we also, again, there's a fine line. We don't want them to scare the families, but we do want them to have a good understanding of why and we want -- we want them to explain that to the families so the family understands a need for follow-up.

The -- we also need those hospitals to be communicating with the pediatricians, the audio practice they're referring to, and to our state Eddie program.

We provide our hospitals with and hearing screening programs with a script. This is kind of an older one, I believe, but it's kind of the same thing that we're still using. Just telling them that the baby has a risk indicator when, they need to follow-up, and why. So giving, you know, this is our kind of generic statement and then giving them more explanation as parents ask for it.

So the medical home as a primary care provider, being familiar with the risks that are forward to delayed I don't know set hearing loss, and then encouraging follow-up. We have, in Boise here, in our state's geographically, we have a large state, but I can speak specifically about the Boise area, we have a lot of pediatrician practices and family practice that is see these kid

dose and we do have a really good amount of physicians that understand the risk factors and understand when to make those referrals, and obviously we need to always continue to keep training and educating, but we do get a lot of referrals from our pediatrician's office that catch when a child wasn't referred from their hospital or birth program center, that those catch those high risk indicators and make those referrals.

Again, the audiology centers they need to be just as aware of what risk factors there are, why their at risk being. If within audiologist gates referral from a hospital program for a child with down syndrome and they don't know that that's a risk for hearing loss, they might not provide the correct testing, so it's really important that they're aware of the risk factors, and then to also know which ones are more concerning. So if a child came in to your clinic with having a received ECMO treatment and ototoxic medications but they passed their initial assessment with you, that child isn't done. They're still at significant risk for delayed onset hearing loss. So just knowing that some kids need ongoing assessments.

And our state program, really the goal of our state program and what our Eddie program ask so well in Idaho so well is to provide training, to provide support, to, again, hospitals, birthing centers, physicians and audiologists, and then to track

and monitor the data. And that's something that I feel our Eddie program in Idaho has been really good at doing is trying to track the data and then analyze the data.

So here's what we do. We have newborn hearing screening in Idaho. We do not have mandates for it. So we have voluntary programs across our state, with you fortunately, we have all of our birthing hospitals on board. We even have some midwife centers that are also on board with doing hearing screening. But not only do we have hearing screening, and this is our referral form, the smaller screenshot, but then I believe of blew up the risk assessment. So that little box is all the risk indicators from 2007 statement that we ask the hospital to check off with the family, if any of those are present, and then we have a statement at the bottom recommendation the referral. So that's how we collect the data.

This is just to show you, so when we started training hospitals on a 2007 position statement, in about 2007, 2008, after it aim came out, we were only getting about 3 to 4% of kids referred with risk factors reported to the -- to our state Eddie program. And now we are consistently and you can see how the numbers grew after we trained and trained and trained, were consistently in about 11% referral rate for the number of kids with risk factors born in our state.

If we look at this data, this is just showing when I showed in a similar slide at the beginning. Those neo natal indicators, so the NICU stay and the ototoxic medications, those are the highest number of referrals that we receive. You'll see that they are the lowest number of hearing losses that we find, but they're definitely the highest number of referrals we receive.

Family history and everything else is less than 10%.

So we, a few years back, developed a classification system for risk monitoring in Idaho. We recognize that there wasn't based on the JCIH statement, because there wasn't a clear guidelines, it said 24 to 30 months of age is when they should be tested by, and then it says some risk factors need earlier or more frequent monitoring than others but it doesn't say specifically the when. So we decided since we were already referring all of these kids anyway, maybe we need to kind of fine tune when they're coming in and how we're getting these referrals. So after discussions with the neo NICU physicians, we developed this guideline. And I know you can't see and I'm going to try and pull it up here on the next one and it's a little bit bigger and easier to see, and I can definitely e-mail this to people if they'd like to see a copy of it. But we developed a guideline, it took us a few months to kind of fine tune it, and we implement it had in two

of our large birthing hospitals in the state. So here's just a kind of a screenshot of which risk indicators, but what we did is we broke it up into two classifications, a class A risk indicator, and a class B, and the NICU physicians really liked the idea of class A and B because they use those types of labels for other things. So they wanted class A and class B. And what we did is we determined that the class A kid dose, that he they have risk that is are more risky for hearing loss, things like a post natal infection, syndromes associated with hearing loss, cleft palettes, those kid dose are at more risk than a child who has ototoxic exposure, we know that the risk is greater. So we broke those up into two categories. And the children that are being identified with a class A risk indicator, we recommend that they come in for a diagnostic ABR and meet with a pediatric audiologist by three months of age.

Where on the other hand, those class B kid dose who have -- they still have risk for a loss but it's significantly less than the other categories, they're not coming in until -- before their first birthday but typically we see them anywhere from seven to ten months of age, just depending on, you know, when, if the child is able to sit up and do behavioral testing and when they can come in for their appointments. So we kind of these two categories that we're seeing kids.

Now, when we first came out with this, when we were doing, bh we were working with the hospital, they had estimated that this class A category that's they have -- that we'd have about 50 kids a year that fell into that. They didn't feel like there was going to be a ton of kids that fell into that category, where most of the kids are going to be falling into that class B category.

So here's the data from two years we collected. At these two birthing hospitals. And we reviewed this data back in November 2015. We had about 10,000, a little over 10,000 babies that were born at those two hospitals during that timeframe. We had 1.6 which was 175 babies with a class A risk indicator. So it was a little bit more than they thought. Not 50 per year but it was a little bit more than that. That you can see that 11%, that is how many babies were born in those hospitals, and, again, just two hospitals, not even looking at our whole state, with a risk indicator of any kind.

So if we look at that data, this is, just again, showing we have about 11% referral rate of all babies that have a risk indicator.

And this is the occurrence of those risk indicators. So out of those babies that have a risk indicator, those 1,100 babies, 734

had ototoxic medication as a risk indicator. In that class B category, again, that testing before one year of age, for family history, we had 175 babies. And then for the class A risk indicators which, again, is syndromes, meningitis, cleft lip and cleft palette those, again, we had only 175 babies that fell into that category.

So class A babies, they, unfortunately, and you'll see this throughout, we have a high develop rate with any risk we're monitoring. And we hope to continue to improve this with training of staff and explaining the why is important to follow-up, but we do have a high loss of follow-up rates. So looking at this, of the 50% of the class A babies that came in for testing, 6% of them had a sensory neural hearing loss. 19% had conductive. And then 75% had normal hearing test results at that time.

And we look at those one that is have sensory neural hearing loss, two of those five children this had cleft palette, two had syndromes, one had congenital CMV. All five of those children passed their newborn hearing screening but came in later, and all of these kid dose were diagnosed by one year of age at different times just depending on when the hearing actually changed, were all diagnosed with a delayed onset hearing loss.

Now, if you think of those 50% that didn't show up back here, we might have another five kids, so you might have ten kids that we potentially would have with a hearing loss related to a class A risk indicator.

When we look at ototoxic medications, this is the higher end, again, high loss of follow-up rate, 54%. But of those that we did test, we have a small percentage of that sensory neural hearing loss, some conductive which we tend to see a lot, and then at that age particularly, and then significant amount with normal hearing. So because of the bigger N, that 1% was five children out of the 345 tested, and you can see what their kind of history was. And none of them had ototoxic medications really all on their own, except for the first one who had an extended NICU stay. So you can see that the second and the third bullet there, those kids had multiple things present during not just one risk indicator.

So the reason why I put family history in there is because, you know, a couple of the studies had said this is a more risky risk indicator. So I pulled that data out as well. I was just interested in looking at our class A and ototoxic medications, that's what I was most interested in, but then I started digging into this family history, and what I find interesting about this slide is family history is what is o reported by the family, so

these families are in the hospital and when our staffs are asking them, you know, do you have a family history of hearing loss, the families are telling a reported family history of hearing loss but are, by far, this is our highest loss of follow-up which really baffles me that 63% of those who said they had a family history of childhood hearing loss did not come in for testing. So it's really, it's something I think in Idaho we plan to look into a little built more like are we asking the questions wrong or what are we doing incorrectly that's getting this information?

But of those, so 65 babies tested, 5% of those have sensory neural hearing loss, so let's look at that. So 3 out of 65 kid dose that were tested had hearing loss, the delayed onset, they're all born, they all passed their newborn hearing screening but came in later and had a hearing loss. One child had multiple risk indicators, syndrome, family history, kind of -- kind of the works for her. But the other two, the only thing was the family history. So had we not had this program, hopefully, at some point, the pediatrician or the managing audiologist would have recommended that those other kid dose be tested, but it probably wouldn't have been until later, hopefully it would have been early, but these kid dose were diagnosed early enough that they got amplification and they were

on the right track to early intervention earlier than they would have within if we wouldn't have caught them until three or four. So what are the barriers? I'm going to try and wrap this up really quickly so we have time for questions. There are kind of a lot of barriers to have a risk monitoring program and you know, in Idaho, we're still continuing to try and improve our program and try and improve our data collection and trying to present our data so that we can help other states. But really, you can see the first three things, it's accurate reporting, if it we don't get the accurate reporting by the staff, the families, again, regarding family history by the audiologist, we, it's a really tough thing to have a good risk monitoring program.

The other thing is shortage of audiologist. Pediatric audiologist. So if we don't have enough audiologist testing we're not going to have successful programs because families are going to have to wait six months to get in for high risk monitoring, they're probably not going to do it. Hopefully they would at some point, but I high risk -- or sorry, high loss of follow-up rates as you can see in our data, we've got really high loss of follow-up rates which we need to improve our statement.

Lack of support by medical homes. So, you know, we have some pediatricians in our valley that are referring for risk and believe in our program and then we have others who say oh, they're back, they're okay, you don't need to go. So if we can get that support by the medical homes, we'll have a better program.

And then I showed with all of the data that kind of conflicts itself presents, there's lots of data presented, there's no standard. If we could have a standard for monitoring risk, it would be great. But there's not a age of when to start when, to stop, what test to use. There's just not a standard. And that is definitely a barrier in Idaho we have guidelines that we provide for testing and when to test. We don't have a mandate, so we just have to present these to the hospitals and the audiologists and hope that they will follow the guidelines that we've provided.

So really quickly, why do we do this? I had a child who was born past their newborn hearing screening, was only in the NICU for less than five days, received oto toxic medications, she was the fourth girl in their family with no family history of childhood hearing loss. She came in at nine months old, kind of inconsistent testing, so an ABR was recommended. You can see that there's no OAEs in her left ear. And again, she is nine

months old so she passed her newborn screening in her left ear nine months before this, and ABR, we'll kind of skip through these slides a little bit, but this was the hearing loss that she was diagnosed with. And this was at 10 months of age. Now fast forward to three years, this is now her hearing loss so, progressive hearing loss in her left ear only, thank goodness, but her only risk indicator was ototoxic exposure. There's no other, nothing else in her history. And had we not had this program, mom said she probably wouldn't have found out until she failed her kindergarten screening because there's no other reason that she would have been identified. She has good speech and language and she probably would with that good unilateral -- or good ear on the right side, but her mom is so grateful that Idaho has the program and we were able to catch it at ten months of age versus at five years of age.

So for future research, I just wanted to kind of give a little plug. We are, a colleague of mine that I wrote an Eddie chapter with, Dr. Gabe Bargain we're going to be doing a Eddie survey of Eddie coordinators on risk coordinators, and I hope if there's any coordinators on the line that they complete the survey that we send out here shortly. Because we want to continue to do risk monitoring data research and hopefully continue to try and publish or present some of our research.

So thank you. And we can start questions. And I know we don't have a lost time, so I wanted to say, last time hi a presentation, I was -- I'm very welcome to people e-mailing me and asking questions. I know for a lot of people that was easier. So please feel free to e-mail me, and I can send you slides, our hand outs, whatever you guys know. And I know a couple of states, we kind of threw out how to start programs in their states after the last presentation, so feel free to e-mail me at any time.

>> Thank you, Jessica. We've got just a few minutes for a few questions here.

>> Yeah.

>> Which one of them they're asking for your e-mail and that will appear here in just a moment. It's, I just had to remove it from the screen in order to create a space for the Q and A field.

The next question is which hospital employees, the hearing screening, nurse, physician, is reviewing the chart and identifying recording class A and class B risk factors on the hearing screening results form?

>> That is a really good question. And that -- so the hospitals that we started this program in, at the time, were being managed by audiologists, so the screening, the nurses were providing the screenings, but the management was done by an audiologist. Since the data that I presented, the hospital screening program has moved to a contracted program by another group. So it's not managed by an audiologist anymore. So when it was managed by an audiologist and screeners were the nurses, the nurses were going through the charts and consulting with the attending physicians and identifying the risk indicators. So our barrier right now is when we transitioned to a consulting program, so an outside facility coming into the hospital and being the consultant and providing the screenings they're not privy to the same data and the same chart review as a nurse is. So that is actually an interesting and new barrier that we're kind of running into. As of right now, what's happening, though, is the screening program then is consulting with the nurse to try and get all that referral information on the risk indicators from the programs. But it kind of depends who is managing the program in the hospital.

>> Another question that came in is who obtains the risk factors for hearing loss specifically in the NICU?
>>> Okay. Say that again, who obtained?

>> Who obtains the risk factors for hearing loss specifically in the NICU?

>>> I'm not sure I'm understanding how the question is worded. We provide, in our state, we provide, NICU and well babies with the guidelines for risk monitoring. So we provide them all the risks that need to be monitored. Then with all that information, the hospital staff, whether it's -- it's typically in our programs, it has been, the nurses or the -- they're called patient care coordinators, they're the ones who identified the risks and then complete the referral forms on each infant that has a risk indicator. And then once that form is completed, they pass it on to our Eddie program. So with at contracting facility like we have now, the similar thing is happening except for the contracting screeners have to ask for the information from the nurse and then they complete the form, the families have to sign it, because if they don't sign it, our Eddie program cannot contact them, so they sign the form, the family reviews it with the nurse and/or the screener, and then that information is passed on to our Eddie program. I hope I and he that question correctly because I'm not sure I completely understood what they were asking.

>> Okay. We've got three quick other questions.

>> Okay.

>> How do you define severe asphyxia?
>> So that's a tough one because it's defined differently amongst, you know, if we look at all the studies and the publications I presented, it is defined very different across them all. You know, in our state, what we have indicated for the screeners, because it's kind of easier to pick all or none, and, you know, we definitely are always trying to improve this, but we have indicated for the screeners that if a child gets put on assisted ventilation, so not using a CPAP but they're put on assisted ventilation for asphyxia, then they are required to have a risk factor screen testing at a further time. So it's any child that's put on assisted ventilation, that is how we define it in our state in Idaho.

>> Great. Another question is can you briefly comment on the value of not only monitoring risk factors but the value of general periodic hearing screening programs like we see in early head start, head start, and even in part C or part B programs?
>>> Yeah. Absolutely. You know, obviously I am for risk monitoring program and we've been doing this a long time in Idaho and we hope that it spreads because we do find those hearing loss that is I think are really important. As a parent of a child with hearing loss, which is not related to risk

factors, I -- I'm so glad that we can identify these kids and get them the help they need.

So with the early intervention programs in screening, I would love it if, you know, definitely there's going to be kids out there and we find them all the time, but that don't fall into these risk categories but still have a delayed onset hearing loss. So if there's a way that, you know, we can continue to careen the masses at later times, we're going City going to be finding hearing loss. And we all know that if you look at. So research, you know, we know the incidence of hearing loss among newborns is completely different than how the incidence of hearing loss in an 18-year-old population. So I think that before that kindergarten, those early head start programs, early infant toddler programs, you know, doing screening is awesome, and I hope that we can continue to do that.

I know there's a lot of barriers, just like there's barriers to risk monitoring programs. There's costs. There's adequate training of screeners. You know, we, even though we have we provide training of some of our, I'm going to give some examples, we've had mid wiev centers that we've provided training for screenings in our state, often we have parents coming in saying, well, they didn't do the screening because they didn't -- they didn't think the equipment was working. And

so we know that that's it's difficult to train people on doing adequate hearing screenings. So I think that if you have good people who can do the screenings, it's definitely a benefit to all those kid dose.

>> Well, and thank you for saying that, because we offer that at the ECCO initiative.

>> Yes. I know.

>> Kidshearing.org is another resource for all of you to check out for helping to support the development of community based screening problems until their birth until three.

The he last question, Jessica, is can the slides be e-mailed to folks who are interested in them.

>> Yes, absolutely, yes.

>> So I put your e-mail address in the middle of the screen right now so everybody can find that. I also posted information there about the upcoming CMV conference that's going to be held in September in Austin, Texas so, if that's of interest to anybody, there's a resource there, the web link to conference and information.

Anything else that you'd like to say to wrap things up today, Jessica?

>> You know, I would just like to say we are very interested in working with other states either starting programs or improving their programs. We are always trying to improve our program here in Idaho with regards to risk indicators. Like I said, we are going to be sending out a survey to Eddie coordinators across the country on risk indicator monitoring and programs that we would like to get some information back from coordinators on what other states are doing. So that should come out, I would think, in the next hopefully month. I'm going to be optimistic. And then, you know, just yeah, just definitely reaching out to us if you have questions and if you want to collaborate, we would love to do that as well. So thank you very much for having me today.

>> Thank you. And thank you everybody. Again, this webinar was recorded and will be recorded at infanthearing.org within the next week. Thanks for all your time. And again, thank you, Jessica.

>> Thank you.