Centers for Disease Control and Prevention
EARLY HEARING DETECTION AND INTERVENTION
Ad Hoc Group

Agenda for July 9, 2002

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CENTERS FOR DISEASE CONTROL AND PREVENTION

EARLY HEARING DETECTION AND INTERVENTION
AD HOC GROUP TELECONFERENCE

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MELANIE GAMBLE: Good afternoon. This is Melanie from CDC. I'd like to welcome all of you to this month's Early Hearing Detection and Intervention Ad Hoc Teleconference on Risk Indicators for Infant and Childhood Hearing Loss. I'd like to remind you that -- to mute your microphones when you're not speaking to help reduce the amount of background noise we'll hear. Today's conference is being recorded and the transcripts will be available on our website in approximately two weeks. Today we'll discuss the JCIH guidelines for risk indicators. And talk about the process several states use to collect information on risk indicators. And then we'll provide an update on the genetics project that's going on here at CDC.

Before we begin, are there any announcements or comments from participants? Okay. Our speakers this afternoon are going to be Pat Brookhouser from the Joint Commission on Infant Hearing, Michelle King from Kentucky's Infant Sound Start, Janet Farrell from the Massachusetts Department of Health, Yusnita Weirather from the Hawaii Department of Health and Aileen Kenneson from the CDC EHDI. Dr. Brookhouser, are you ready to begin?

PATRICK BROOKHOUSER: Yes, I am. I was asked to make some comments here, sort of twofold in nature. Given evolution of the risk indicators that have been included in the several statements made by the Joint Committee on Infant Hearing, and secondly to talk a little bit about, I suppose, why they were left in the 2000 statement and the utility that we feel they may have. I think to begin with it would be good to go back historically to the beginning of the Joint Committee on Infant Hearing. The Joint Committee was really formed in the latter part of the 1960s. If you'll remember back, that was immediately following the last great rubella epidemic in the United States and was at a period where the death of President Kennedy's newborn infant had set a lot of high visibility on efforts to help with the increase in survival rates. So, with both of these efforts moving forward, including the observation at the time of the rubella epidemic, there were a lot of kids who had isolated hearing loss as the sole indicator that they had in fact had prenatal rubella, which was confirmed by either serological or virus isolation techniques. So, we knew that children at that time who appeared otherwise normal were in fact high risk for hearing loss. As we began to see the increased survival rates in NICU in terms of the number of children involved, I think the Joint Committee, which knew at the time that it was dealing with a low prevalence disorder for which no really good test modalities or screening modalities were available which had appropriate levels of sensitivity and specificity, they began to look at how they could somehow stratify the sample in such a way that they at least could screen with less than adequate techniques, behavioral techniques in many cases, the highest risk infants and identify them. So, this really came about in the early seventies.

I think the first indicators were used in about 1972. There were five indicators which were then gradually expanded as we began to observe more and more children who were presenting with delayed -- or with hearing loss being diagnosed, as you remember, basically in the second and sometimes the third year of life for which there was no good obvious explanation as to why they had the hearing loss except for perhaps some high-risk factors in their background. So, the subsequent statements of the Joint Committee's real focus owed on trying to expand in scope the number of high-risk indicators and to become much more specific in the way that they

were described. I think, also, at that time a lot of publications in genetics began to identify a larger number of non-syndromic recessive losses, which as you can imagine have a hard time getting good familial relation. With smaller families today, nobody in the family might know of a relative who had a hearing impairment. So, most of the studies that were being produced in terms of etiology at that point had between 30 and 40 percent unknown or uncertain etiology described in them, which most of us felt, of course, many of which were genetic losses of a non-syndromic variety and unidentifiable.

So, the period up to 1994 in the Joint Committee statements was really characterized by expansion and sort of stratification of the high-risk indicators, beginning first with neonatal indicators and then gradually bringing along indicators for infants 29 days through two years, and then above 29 days through three years who required monitoring. A number of kids didn't have losses at birth but in fact developed losses over periods of time. And we were trying to come up with means of helping to identify a reasonable sample that could be screened with various techniques. We're moving all the time technologically in the direction of a more sensitive and specific screening modality, but we kept trying to respond to that by giving a better indicator and as to which resources should be invested to follow.

The other thing that the Joint Committee is acutely aware of is that our statements have not only had impact in the United States, they're used in many parts of the world where the access to technology to do true universal newborn screening was not present. So, if you compare the '94 statement, which I guess was the final statement prior to our recommendation of universal screening, it gets into great detail with very specific kinds of high-risk indicators during the neonatal period, talking about specific birth weights, talking about hyperbilirubinemia, talking about ototoxic medication, the level of Apgar score, which we felt was high risk, mechanical ventilation, the five days or longer, and stigmata or other findings associated with syndromic changes. At that time parental concern was put in as a high-risk indicator. And I think that's one of our best indicators, a longitudinal observation by parents of a child's behavior in response to auditory stimuli in the environment. Most parents complained bitterly that they were disregarded as they approached their primary caregivers and others with a history of saying my children are not responding adequately to sound. So, that was put in at that point as a high-risk indicator. And we looked at the possibility that otitis media should be included because we were looking at other losses as well, among kids who had recurrent or persistent otitis. This is viewed in many cases as being somewhat related to diffusion of toxins. Also over this period of time that these statements were being issued began to experience the very positive impacts of the vaccine development. So, earlier generations of kids obviously had children with measles, mumps and flu and meningitis. We're now seeing that dropping off.

So, the indicators in the 2000 statement were maintained for two reasons, one of which was that we knew the evolution of universal screening was going to be systematic. Not everybody was going to be able to move rapidly towards universal screening, and therefore the various risk criteria should be left in there for use by hospitals that were in the process of trying to acquire screening equipment or for whatever reason were unable to do the screening, or for our international colleagues who maybe in their particular countries were unable to screen all newborns, and therefore, needed to have an ongoing focus on those children who should be found. Most of you I think from reading the statements realize that the follow-up studies done at the time that specific high-risk screening was being done indicated that at the time children were entering school approximately 50 percent of the children who had educationally significant hearing losses in fact did not have any obvious high-risk factors in their background that would have been missed by any kind of a high-risk registry.

The second reason is that we're acutely aware of is that some of the more important risk indicators; namely, family history of hearing loss, may be difficult to obtain from the small nuclear families we now have, or perhaps the family lore would have been identified hearing losses due to other nongenetic factors in the family and therefore discarded that there was any indication of an inherited hearing loss presents. If you compare the '94 and 2000 statements, I think you will see that the specificity of the risk factors changes quite a bit. And we've gotten more in the 2000 statement to broader definitions of risk, such as instead of all these specific findings that would occur during a stay in the NICU, simply saying an illness or condition that require admission of 48 hours or greater to a NICU should be an indicator for follow-up in screening. And then stigmata and other findings associated with the syndrome known to include sensory neural hearing loss and then family history of hearing loss, all of those fall into the risk indicators in the newborn period. Craniofacial anomalies fall in there, including abnormalities of the ear, and then in utero infections. This basically has telescoped these factors down

to relatively few, but broader in nature than the 10 factors which had been identified in the '94 statement. And I think as you look at the ones for follow-up, age 29 days through two years, it encompasses many of the ones that we're seeing in the newborn period, as well, and adds to it things such as meningitis and other kinds of neurodegenerative findings that would place children at a higher risk for possible problems, as well as at that point recurrent or persistent otitis media comes on board and head trauma comes on board.

Now, who is going to collect all this data? I'm sure you're going to be talking about that. But obviously the primary care physicians with the medical home concept are going to be highly involved with collecting the ongoing information that comes on line about this child after the child leaves the hospital environment and leaving the screening program. And how one begins to assemble that data and apply it meaningfully in the follow-up with these children I think is something that's open to a lot of discussion. Clearly audiologists who are in practice need to be much more involved in feeding this information into the system, and others, as well. And as we learn more about genetics, many of these children may need to be identified as possible candidates for specific genetic screening that might help us a great deal in figuring out exactly what kind of losses they have. We know right now in the workup of kids with hearing loss many of these kids receive what is probably in the future going to be viewed as unnecessary amounts of laboratory and radiological evaluations simply because we don't know where to turn in terms of trying to identify specific etiological factors. But these findings in here are broad enough in nature that they do require the collaborative efforts of a large number of the individuals who are involved in the patient's medical home, in the child's medical home to make sure that data can be meaningfully applied in the follow-up of the child. Now, I can answer questions about any specific risk factors, but that's the philosophy of why they're in here.

We clearly know that a significant number -- the number at this point is not known -- of children are going to develop their hearing losses after they've left the hospital and after their newborn screening period. So, we want to catch those kids as quickly as possible. Some may in fact be after the first year of life. We need to know a lot more about CMV, of course. That's probably one of the great deficiencies in our early identification program is the lack of universal screening for CMV at birth. We don't know what role it plays in many of these children because by the time we find them, we don't have the capability to test definitively at that point. But I think these factors will continue to be a part of our early identification efforts. Granted, they may not be as useful as they were in the past because we have the universal newborn screening going. But I think they're going to continue in future statements of the Joint Committee because they do volunteer utility in the long term follow-up of children who may be at risk or may develop delay onset losses. And they will continue to be of value for our international colleagues, as well. I'm going to stop and ask if there are any questions or comments. Or I'll be happy to amplify anything about the reason why they're in the statement and the utility of them.

UNKOWN SPEAKER: Dr. Brookhouser, this is Minnesota. And we have kind of a crush in a situation we're dealing with here. And this is regarding risk factors for the neonatal period, like the birth to 28 days. If you had to narrow down those risk factors do, you know, which ones are the most important and why -- I'd be interested in hearing that. And why I'm saying that is because at the national level they've come out with a standard for birth certificates nationally.

PATRICK BROOKHOUSER: Yes.

UNKOWN SPEAKER: And as we've reviewed them, there are some risk factors related to hearing loss you get in the neonatal period that are not on that recommended standard, such as the stigmata, craniofacial anomalies, some of the interuteral infections like CMV, Taxol, are not on there. Family history we don't think will ever get on there because that's -- I mean, the challenge is on the birth certificate they need to be risk factors that are readily available to the people that are collecting that information for the birth certificate; i.e., the prenatal record has that information ready. And right now I don't think our obstetricians and family physicians know enough to ask for family history, so they'd probably not find that in the record. But some of the others, the herpes, the rubella, as well as when the baby's born we can get some of that.

PATRICK BROOKHOUSER: Yes.

UNKOWN SPEAKER: So, to back up, if we had to narrow this down, which are the most important risk factors, what would they be? And keep in mind two states right now have added risk factors to their blood spot forum

along with their hearing screening, the state of Washington and Oklahoma. And they vary. Washington has five risk factors. Oklahoma has six. And they're not the same. What would you recommend?

PATRICK BROOKHOUSER: Oh, I hate to choose one from the other, but I can tell you in general terms first of all that the risk factors that you talked about earlier would not be viewed by, say, a neonatologist as evidence of risk during a hospitalization in a NICU simply because their history of hearing loss and stigmata of syndromes that don't pose any life-threatening complication or don't threaten the life of the child. So, they are really difficult to obtain and you're not going to find them in a routine neonatal history.

I think part of that is going to have to be an education of the primary care provider and the parents. If you have a history of hearing loss in your family, you should make sure that your child gets screened and followed up. As we started looking over the years at these, certainly a low Apgar score and anoxia appeared to be significant in nature and were in the risk factors very early. Hyperbilirubinemia was in it one time at a specific level, and then people backed away from it. Now some individuals are saying it may relate fairly directly to this whole condition called auditory neuropathy. So, that may in fact be a factor. But certainly I think the ability of the child to be properly ventilated during early phases of his or her lifetime is probably as important as any because the and knocks I can't and the damage to the central nervous system and the remainder of the nervous system that appears with that can be a factor. Birth weight appears to be not as big a factor as they thought in the past.

But there's a significant increase in the amount of kids found to have hearing loss versus the normal population. I see the Cohen-Wesson data quoted in the statement says that they didn't think it was as important. And there have been some theories that neonates were more tolerant of overtoxic medications than, say, an adult population. I think in terms of trying to narrow it down, if you focus on those factors which are life threatening, anoxia and significant difficulty early on, required ventilation, and then also on the issue regarding hyperbilirubinemia. And of course, general infections. I think it's tragic that we don't screen for CMV because -- now, maybe we can back up now and get blood spots that are kept in libraries or kept in patient records and use those to find the CMV later on. That's going to be a big problem going back to it. But that would be so helpful in terms of separating out the obvious nongenetic causes from the genetic causes.

I think part of our problem with kids that had high-risk histories were that most of these kids, an awful lot of them fell into no obvious etiology. So, we tended to back up and say, oh, this was a high-risk neonate. That must have been the cause. Without necessarily having point for point indications as to the specific cause. But there have been some good histopathological studies done on kernicterus in the hyperbilirubinemia kids that showed you were getting deposition in the auditory nuclei. So, I guess without necessarily saying the other should be eliminated, you might want to look at the indicators in the '94 statement. They're much more specific in nature as to how much mechanical ventilation a child should have, what the Apgar scores should be. But those would be the logical ones. And then, of course, meningitis is always an issue. But I think the obvious infections would impress everybody enough that they'd view that as a high-risk factor. But I think the CMV might not if they didn't know it was there. So, I can't give you 1, 2, 3, but I do think that oxygenation of the central nervous system is a very important issue here.

UNKOWN SPEAKER: Thank you very much, Dr. Brookhouser. And the new standard that's being proposed by the feds for a birth certificate does have the assisted ventilation, it does have the NICU, it does have antibiotics received for neonatal sepsis, so you could probably hit some of those infections. But it does not have hyperbilirubinemia nor CMV.

PATRICK BROOKHOUSER: The studies done -- some of the presentations done over at the international conference in Milan, we need to really look hard at this whole issue of auditory neuropathy. Number 1, how prevalent is it? Number 2, is it permanent or does it resolve? And number 3, what are the antecedent factors? And it appears that hyperbilirubinemia, maybe not even at the level requiring transfusion, is present. Is it causative? That's hard to say. But it may turn out to be a more important factor than we thought it was. We may find that it alters some of the conduction characteristics perhaps in the auditory nerve or the nucleus for a period of time, which may then revert to normal. I just don't know what's going to happen there. We're going to

have a meeting up at the AAP in the fall and talk a lot about auditory neuropathy as being something that needs to be studied much more intensively.

MELANIE GAMBLE: Thank you very much, Dr. Brookhouser. We're going to move on to our next speaker. We have four more to go. Now we're going to hear from individual states. We have Michelle King from Kentucky.

MICHELLE KING: Hello. This is Michelle. I was asked to talk about the letters that we send to families regarding individual risk factors. In Kentucky we track infants if they are referred from the physiological screen or if the hospital reports to the Sound Start surveillance system that one or more of the following risk factors were present: mother exposed to rubella, or CMV or diagnosed with syphilis; did the infant have: sepsis, seizures, or meningitis; hyperbilirubinemia, Craniofacial anomalies; ototoxic meds,; persistent pulmonary history, a family history of permanent childhood hearing loss. The hospitals notify our database by filling out an NCR-copied hearing screen report form. The original comes to the state EHDI office, a copy stays at the hospital; a copy goes to the parent and one to the physician. The forms completed at all 61 of Kentucky hospitals that birth more than 40 babies a year.

If they do not pass their hearing screening or have one of those reported risk factors above, they receive a letter – the hospitals are required to send it to us within 24 hours of hospital discharge. Within 48 hours of receiving it we send a letter out to the families. We also enter the information received from the hospital into a stand-alone access 2000 database, and it generates the letters. The most stringent guideline for follow-up is sent when more than one risk factor is reported with the most stringent follow-up being those that are referred on the physiological screening and secondary meningitis, and the rest are recommended by three months of age to have a diagnostic follow-up.

Parental response has been very positive since specific letters began January of 2002. Several parents reported, and this is probably less than 10, that they didn't recall being told the information that we stated in our letter. And they have called with varying degrees of anxiety. They are asked to contact the hospitals to verify the information. Usually the information was correct. I know of three cases, one that was a hospital nursing reporting area, one was a clerical error on the hospital's part and one was what data entry on our port to which we were able to find the reason and get the information back out. Our largest categories of letters that we do disseminate are as follows: 33 percent on a referral from the physiological screen and 46 percent for family history. And the lowest area in the last six months has been six have gone out regarding meningitis. To date we have identified several infants with hearing loss that passed their newborn screening. In some instances we were able to possibly relate this to the fact that the baby was screened a number of times over the recommended two-stage screen. We have also had progressive hearing losses identified with routine audiological follow-up. And then they were referred for medical evaluation; all have been fit with amplification and were enrolled in early intervention. Any questions?

JUNE HOLSTRUM: Michelle, this is June Holstrum from CDC. What percentage of parents responded to your letter? And what do you do when they don't?

MICHELLE KING: We have three regional coordinators that do follow-up if we don't receive a report from an audiologist that these babies have been rescreened or diagnostically tested. And we contact them, either additional letters or phone calls. I don't know the percentage and my data person had to leave on an emergency this afternoon. So, percentages I don't have.

DEBBIE BARRINGER: Michelle, this is Debbie Barringer calling from Michigan. Can you repeat those percentages again? You said 46 percent had a family history?

MICHELLE KING: 46 percent of the letters we do send out are a positive response or reported to us for family history. We're not always sure how accurate that is. We do instruct our hospital personnel that the history be positive for a ***permanent childhood hearing loss,*** but we do have occasionally families calling in saying, well, no, it was otitis media / tubes type of thing. And then we have 33 percent on the physiological hearing screening itself. But our overall refer rate for infants in KY physiologic screen + risk indicators + missed screening is 9%. Of that 9% - 46% have the family history risk indicator and 33 % have referred from the physiologic screen.

DEBBIE BARRINGER: Is this posted on your website, the stats?

MICHELLE KING: No, they aren't currently.

PAT RICE: Just a question. Of the 46 percent that had a family history, do you know what percent of those did not pass the hearing screening?

MICHELLE KING: 6%

UNKNOWN SPEAKER: Michelle, could you comment on the children who pass the screen but failed the rescreen? What were their characteristics and at what age did you make the diagnosis?

MICHELLE KING: We had one that passed the screen but a risk factor was family history. And they went in for every three months -- well, they went at three months, six months, missed their nine-month, and were diagnosed at 12 months with a hearing loss. And that one has been progressive. The child is now 15 months.

We had the one mom that happened to be a speech pathologist reported that the nurse came in ecstatic that the child *had finally passed* the screening on the fifth time. But the mother decided to follow up anyway, and the child was diagnosed with hearing loss before 3 months of old. So, that was just a referral on the hearing screening itself.

MELANIE GAMBLE: Thank you very much, Michelle. Now we're going to move on and hear from Janet Farrell from Massachusetts.

JANET FARRELL: I'd like to begin with a little bit of the history in Massachusetts. We had the first high-risk law in the nation. That was passed in 1971. So, we have about 30 years' -- a little over 30 years' experience following young children with hearing loss. As you know, only 50 percent of newborns at risk for hearing loss have a known risk indicator. So, many children were not able to be picked up through that system.

And some of the problems include follow-up occurred after discharge and many newborns were lost to follow-up. In Massachusetts hospital newborn hearing screening staff support collection and notification of risk indicators. It's required for our regulations and guidelines and each hospital has a program director assigned in birth centers, as well. They have protocols; they insure compliance with the law, regulations and guidelines.

And there's also an audiologist that has to be assigned to research hospital and birth center screening programs. They consult sometimes their staff at the hospital. They oversee the training and supervision of screening staff, indication of results; including risk indicatation for hearing loss and results of birth certificate contact person, which helps with getting the information into our data system.

Communication with the parents and guardians and the physician is very important. Each birth facility is required to provide written notification of risk indicator to the parents, guardian and physician for each child that has an identified risk indicator. Recommendations are made for diagnostic evaluation. Even if a newborn passed a hearing screening, if they have a known risk indicator and information about major milestones in speech and language development are included as well as links to other available resources and the Department of Public Health 800 number is given for questions or concerns that parents or others can call.

There are financial resources available for follow-up testing after any available health insurance and the Department of Public Health. They accept the cost for any child whose parents do not have coverage. How do we collect and monitor risk indicators in our system? Our system is called the Childhood -- populated through our electronic birth certificates. And we also use information through parents contacted. We have outreach staff contact each family, and we collect other information from our diagnostic centers, which have to become approved from the department. And hospitals are required to refer families to those centers exclusively. We

also receive referrals from early intervention and all sources, parents and professional providers have access to our 800 telephone number for technical assistance.

Our electronic birth certificate collects a lot of the risk indicator information. We get information on NICU admission. Examples would be low birth weight, Apgar scores, gestational age. We get information on congenital anomalies or conditions associated with hearing loss.

Examples would be craniofacial anomalies, at risk factors for pregnancy, including in utero infection, method of delivery. An example of that would be fetal distress and other neonatal procedures. An example of that would be assisted ventilation.

Our diagnostic centers are also someplace that we're able to collect information from families about risk indicators. And families sign an informed consent so that we can get that information. At the diagnostic center they discuss family history with families, in utero congenital infection, herpes, rubella, syphilis, other neonatal indicators, such as prematurity, hyperbilirubinemia, pulmonary hypertension, other conditions such as whether or not ototoxic meds were given, bacterial meningitis and other conditions, head trauma.

And then we have an "other" category so if we don't -- if there's another condition that somebody is concerned may have contributed to the hearing loss, they can fill in the "other" category.

We get a lot of information through parent contact. And that is we are able to put that into our childhood hearing data system. The outreach staff contacts families via telephone, and they are able to provide information.

The staff is able to provide information and referral to diagnostic services. And they discuss risk indicators, particularly information about family history. Early intervention in Massachusetts happens to be in the same division as the universal newborn hearing screening program. So, we have a close collegial relationship.

And all children with speech and language delays or parental or caregiver concern are eligible for follow-up audiological testing. And we receive many referrals through early intervention. It could include children with frequent ear infections, referral of children that move into Massachusetts. And they can help assist in monitoring risk indicators by providing ongoing supports of those developmental milestones to families. In the New England region we've had close contact with other states, as well, who have helped refer children.

In closing remarks, I'd like to say that our system is an integrated system beginning with the electronic birth certificate. We're also integrated with the first link home visiting system and our birth defects system. Eventually we will be connecting to the early intervention database. We have the ability to collect a wide variety of risk conditions via the electronic birth certificate for use in analysis, and we have the potential to report relevancy of risk indicators based on Massachusetts' population over time. Any questions?

UNKNOWN SPEAKER: Did you put all of the risk indicators, such as age recommended in 2000 on your birth certificate?

JANET FARRELL: No. A lot of the information was already collected. And we're collecting it from sorts of a variety of sources. From the electronic birth certificate -- our electronic birth certificate collects, you know, a wide range of conditions. But then we're also collecting some from parents, some from the diagnostic centers and others. Are there any other questions?

MELANIE GAMBLE: Thank you very much. Yusnita Weirather from Hawaii is going to tell us a little bit about their system.

YUSNITA WEIRATHER: In Hawaii we have three different referral sources. The first one is the medical chart review. This is only for special care nursery babies. And referral source number 2 will be from nurseries. If the child has one of these three or any of these conditions, which is family history of hearing loss, in utero infection problems, and also craniofacial anomaly. And the referral source number 3 is pediatrician. For this one in Hawaii all children who are screened and either pass or not pass hearing screening, letter is sent to the

pediatrician for each child. And in the back of the letter is the list of risk indicators that we ask the pediatrician to keep eye on these children in case we miss them.

And then we also send a small brochure to remind the pediatrician from time to time that if this condition exists please give us a call to process a follow-up hearing -- or repeat hearing screening. And the recall system that we have is these children's are risk factors are called back for behavioral hearing test at seven months of ages. And one month prior to that date we send a letter to pediatrician just to confirm that condition exists. I mean was confirmed for this child to make sure that we don't call the parents and the parents do not know what are we talking about, they're not aware of the condition that we think the child has.

And in that letter we include a return-addressed postcard. And in the postcard we put the number of this child. And there's three items that the pediatrician has to check. Number one is whether the child is under this pediatrician's care. Number two is this condition confirmed or positive on this child? And number three that either the pediatrician will let the parents know to return for follow-up hearing test or we need to call the parents for test. And when we receive a postcard from the pediatrician, then we'll act accordingly. We have been doing this since we started the newborn hearing screening program in Hawaii. And so far we have, I believe, four or five children who passed newborn hearing screening and later on developed a hearing loss. And all of them now are using hearing aids. And the hearing loss occurred at the first evaluation at seven months of age. And then when the behavioral test indicates that the child has suspected hearing loss, then we do a sedated APR to confirm that condition. I think that's about it.

Vicky Thompson: I wanted to know if any of the presenters have looked at the cost associated with providing the intense high-risk factors that these states do? It's wonderful and the programs are great. I'm just wondering if they have really looked at the FTE costs involved as well as the supply costs in sending letters and postcards.

YUSNITA WEIRATHER: This is from Hawaii. We were thinking about that just because, like the question earlier, how many percent we were able to bring them back and how many percent did we have to chase them. And the policy in here is used when the pediatrician sends the postcard back, the first follow-up we do is call the parents and tell the parents to make the appointment. And the parents normally don't have any problem with that because the doctor already informed them. And then some of them make an appointment but then cancel and don't follow up, and we have to call them again.

And so, the policy in here is after three attempts we close it with certified mail and then tell them that this is -- and let the pediatrician know that we exhausted our resources to follow this. And looking at this process, the good news is not too many, because Hawaii is small. And so far we have not been, you know, at the point that we don't want to do any more. But we don't know how much, you know, monetary costs we spend on this.

MICHELLE KING: Also in Kentucky there's been no specific cost evaluation of the program. It is something we inherited from our hearing high risks to send out the letters, and we just continued it as we moved to universal newborn hearing screening. We do have currently 6,736 infants that we are following, and we have lost 295 to incorrect addresses and follow-up information. So, that's the status in Kentucky.

PATRICK BROOKHOUSER: How long do you follow them?

MICHELLE KING: We follow them to age 3. That's our proposal.

PATRICK BROOKHOUSER: All right.

UNKOWN SPEAKER: I have a question for Hawaii. What made you choose seven months for the follow-up hearing test if there's a risk factor?

YUSNITA WEIRATHER: Just because we don't feel that we would like to do ABR on this child, or normally we -- I don't know. We just feel like behavioral testing is more valuable on this rather than OAE only or -- and we never even considered using ABR. And more than anything, we would do OAE, but we feel OAE is not sufficient enough to -- is not standard.

UNKOWN SPEAKER: Are you getting any ear-specific information with your behavioral testing at seven months?

YUSNITA WEIRATHER: Yes. We have to do that.

UNKOWN SPEAKER: So, you're doing it with just insert earphones

YUSNITA WEIRATHER: That's correct.

MELANIE GAMBLE: Okay. We're going to move on to our final speaker, who is Aileen Kenneson. She's going to give us an update on the genetics project at CDC.

AILEEN KENNESON: I wanted to update you on the epidemiology study of hearing loss and particularly what we're doing about the risk factors as part of the study. Just to remind everybody, there are four states participating in this study. It's a level 2 project. There's Hawaii, Rhode Island, Utah and parts of Georgia are participating in the study. And we're recruiting both babies that are identified with a hearing loss through the newborn hearing screening program and also recruiting families of babies that pass the screen but are diagnosed with a hearing loss before the age of three.

All of the participants in the study, we collect family history, we collect medical information, both from asking the parents directly and from medical record review with the parents' consent to review the records. And children are seen by a medical geneticist to, you know; look for any of the symptoms that are associated with syndromes that are associated with hearing loss. And then there's a DNA analysis of a couple of genes, particularly the connexin 26 gene and two of the mitochondrial genes. One of the mitochondrial genes we're looking for is the one that causes susceptibility to aminoglycosides.

We used the JCIH risk factors from both the 1994 and the 2000 reports, both of them, combined all the risk indicators, Apgar scores and NICUs, the different infections, the ototoxic drug exposure, the mechanical ventilation and everything like that. And the purpose is to collect more information to help drive the evolution of the risk indicator lists that are being used and also because traditionally we've looked at only environmental factors. We would like to expand the analysis to look at the role of both genetic factors and gene-environment interactions, such as the combination of ototoxic drugs and particular mutations in babies. And then most of our genetic stuff is associated with the connexin 26 mutations which are believed to cause hearing loss that will be detectable at birth, but we're not sure about that. So, we're going to be seeing whether the hearing screening will indeed be picking up the children with connexin 26-caused hearing loss or not. That's where we are right now.

We've already begun the study in Utah and hope to begin collecting samples in the other three states soon. And in the future we hope to be able to expand the study to include more in-depth study of the environmental risk factors. So, instead of just looking in the medical records to see if there was any evidence of infections, maybe we can go back and pull those blood spots and look for evidence of congenital CMV infections or something like that. We haven't figured out exactly what we're going to do yet, but we are going to be putting some more money into expanding the study. So, that's it in a nutshell. And I'd be happy to answer any questions.

ANNE JARRETT: This is Ann Jarrett from the State of Michigan. When you said that you're going to expand the project, are you expanding if within those four states, or are you thinking that you might expand it by adding other states?

AILEEN KENNESON: Right now we have plans to expand it just within those four states. But if Congress would be so kind as to give us some more money, we would love to expand it to additional states.

CAROL HASSLER: This is Carol Hassler from Vermont. And I have a question for Michelle from Kentucky. Did you say sort of in passing that your tracking system uses an access database?

MICHELLE KING: Yes. Currently we have an access 2000 stand-alone database and are looking to move to something that the cabinet for Health Services has, which is -- they call it the computer utilization project, which uses SQL, I believe. But it's not been merged to that or any other state database as of yet.

UNKNOWN SPEAKER: Can we get in touch with you, maybe by e-mail or phone, to ask you a little bit more about your sort of interim database?

MICHELLE KING: That would be fine.

PAT RICE: This is Pat Rice from Minnesota. I'm just curious. With these genetics projects are you developing any sort of parent-friendly materials to talk about what you want to do and accomplish in doing the testing?

AILEEN KENNESON: Yes, we are. We're developing some materials that explain genetic testing in general, what types of things you want to consider before you decide whether or not to have genetic testing. And then also information specific to testing for hearing loss, what the different genes are that can be tested for and how you would interpret the results. It's still in development. And we hope to get it on the web when it's done and then maybe even have -- if we could finds some money, have brochures printed. But whatever we make available on the web would be available for everybody else to use, as well.

JASON MARLAP: This is Jason Marlap from Ohio. I have a question for Janet from Massachusetts. Regarding the electronic birth certificate, first of all, do parents sign any kind of consent to collect the information on that? Second, is the UNHS, the screening results, recorded on the EBC? And third, what kind of data system records all the information which is collected?

JANET FARRELL: The parents do not sign any kind of a consent because we have a comprehensive law in Massachusetts that protects us in receiving the results of the screening. They do sign consent to give us the diagnostic information. So, at the diagnostic center there would be a consent, but not at the birth facility.

Currently our birth certificate is going through sort of a major change, and they're moving towards a web-based system, which will affect all the systems that we use right know. Our childhood hearing data system is currently an access program, but may become web-based once the birth certificate is web-based. We're sort of in a transitional period right now.

MELANIE GAMBLE: Are there any further questions or comments? Okay. Before we close, I just want to open -- are there any announcements that need to be made? Okay. Well, I want to thank all of you for participating, and a special thank you to each of other speakers. The next teleconference will be back on schedule with the first Tuesday in September, which will be the September 3rd. So, thank you all. And we'll talk to you then.