

**Centers for Disease Control and Prevention
Early Hearing Detection and Intervention (EHDI)
Special Topics Teleconference
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Presentation: An Update from the Joint Committee on Infant Hearing

<<John Eichwald>>

Dr. Betty Vohr will provide us with a brief historical view of the JCIH and present proposed changes in the next committee statement. Dr. Vohr is professor of pediatrics at Brown medical school and director of the Women's and Infant's Hospital of Rhode Island Follow-up Clinic. Not only is she chairperson of the JCIH, but chairperson of the Rhode Island hearing and screening advisory committee for the Rhode Island Department of Health and Department of Education. The medical director of the Rhode Island hearing assessment program, and a member of the American Academy of Pediatrics Medical Home Task Force on infant hearing. Please hold your questions until the end after Dr. Vohr's presentation. With that I turn the teleconference over to you Betty.

<<Dr. Vohr>>

Thank you so much, John. I want to say I appreciated the invitation to present this topic which had been discussed at the annual EHDI meeting in Washington, D.C., on February 3rd. At that time we had a number of comments and input from the audience. As you noted on my slides it says not for distribution. This is a work in progress. The JCIH is meeting on March 23rd to finalize the position statement. All of the comments and suggestions that you have will be considered. We anticipate publication in '06. A few changes have already been made since the EHDI conference based on input provided and new information we received from the Academy of Pediatrics about the Bright Futures recommendations, which are are going to be coming out in May.

With that, I will start the presentation. The first part is essentially the same as what we presented at the EHDI meeting; it gives you a feel for where we're going as we identify where we've come from. We're going to review the major milestones and the 37-year history of the JCIH and present our proposed changes for '06. It's amazing to me that the JCIH was established in 1969. Currently it is composed of 14 representatives. There are two from each organization. The American Academy of Pediatrics, ASHA, AAA, the American Academy of Otolaryngology, and the Directors of Speech and Hearing Programs in State Health and Welfare Agencies. The original charge of the JCIH was to make recommendations concerning early identification of hearing loss and concerns with newborn screening.

The first document "Position Statement" published in 1971 was a single page document. It concluded that data at that time were inconsistent and therefore universal hearing screening could not be recommended and risk factors at that time were not even mentioned.

It was followed by a position statement recommending screening of infants with the following risk factors: family history of hearing loss, rubella or other nonbacterial infections, defects affecting the ear, nose and throat, very low birth weight (less than 1500 grams) and any direct bilirubin levels considered toxic. If the hearing screen of infants with risk factors was normal, the recommendation was that they should continue to have regular assessments of their hearing. Therefore, in 1973, it was the first recommendation for continued surveillance of infants with risk factors.

The third statement in 1982 expanded the risk factors by two: 1) bacterial meningitis including hemophilus influenza and 2) infants with severe asphyxia, including low Apgar scores or failure to institute spontaneous respirations.

The fourth Position Statement in 1990 had two major changes: 1. It divided the risk factors into neonatal (birth to 28 days) and later onset (29 days to two-years). Two new risk factors were added resulting in a total of 10 risk factors: Prolonged ventilation for greater than or equal to 10 days, and stigmata or other findings related to a specific syndrome associated with hearing loss. An example of that is Ushers.

In 1994 with the 5th JCIH Position Statement, the RIHAP hearing assessment program demonstration project had been implemented demonstrating that hearing screening could be accomplished in well baby nurseries. Also, in 1993, the NIH consensus conference had endorsed universal detection of hearing loss. At about that time, Dr. Yoshinago-Itano from Colorado was able to compare the outcomes of infants with hearing loss who had newborn hearing screens and were born in hospitals with screen programs versus those that were born at hospitals that did not have screening programs, and clearly demonstrated improved outcomes of infants with newborn screens identified before six months of age. The 5th statement, therefore, endorsed universal detection of all infants before three months and intervention by six months. It did not specifically endorse universal newborn screening.

The sixth Position Statement in 2000 endorsed direct physiologic screening of all newborns regardless of their risk status. It expanded the concept of screenings to EHDI (early hearing detection and intervention) systems of care. It concluded that a quality EHDI program in the first year of life was the way to maximize language skills for all children and hearing loss.

For universal newborn screening, the question was asked, "why do we need risk indicators?" Risk indicators were still to be used in regions of the country where universal screening had not yet been implemented. At the same time, data were emerging indicating that a significant percent of hearing loss is not identified by newborn screening. Therefore, risk indicators associated with hearing loss gained importance.

The 6th Position Statement made the recommendation that there should be audiologic monitoring every 6 months until three years of age for all infants with one or more risk factors for late onset hearing loss, but who passed the newborn hearing screen. That recommendation led to quite a burden for EHDI systems.

The current recommendation which you're all familiar with is: screen before one month, diagnosis before three months, and intervention before six months of age. We know that congenital hearing loss involves a significant number of babies annually and is the most common birth defect occurring in 12,000 to 16,000 babies born each year in the United States. The NICU rate, depending on acuity levels in the NICU, averages about 10 per thousand. The well baby rate is about 1 per thousand.

So where are we going with JCIH 2005 now 2006? The new recommendations are based on the experiences that states have had in the last five-years and new evidence that has become available during the past five-years. I'm going to go through each of the recommendations for changes.

The first recommendation relates to the definition of targeted hearing loss. This definition has been expanded. In addition to screening for bilateral or unilateral sensory neural or

conductive hearing loss averaging 40 db or greater, we're now going to be recommending screening for neural hearing loss or auditory neuropathy in infants admitted to a neonatal intensive care unit for greater than 48 hours. Second because of this recommendation, there is a change in newborn screen protocol. NICU babies are to have an ABR included as part of their screening, so that neural hearing loss will not be missed. No change is recommended for well-baby screening. For re-screening, a complete evaluation of both ears is recommended even if only one ear failed the initial screen.

There is a new recommendation for infants readmitted to the hospital within the first month of life. If these babies have conditions present at the time of their readmission which are associated with potential hearing loss; and examples are hyperbilirubinemia requiring an exchange transfusion, or culture positive sepsis; a repeat hearing screen is recommended prior to their discharge from the hospital.

Audiology evaluation recommendations: Audiologists with skills and expertise in evaluating infants with hearing loss should provide audiology diagnostics and habilitation services. The recommendation for the Audiology Diagnostic evaluation is that at least one ABR is included as part of a complete diagnostic audiology evaluation and in conjunction with other measures for validation of hearing loss for children below three years of age for confirmation of permanent hearing loss.

As part of the medical evaluation all families should be offered a genetic consultation and in every case where hearing loss is determined, the infant/child should have at least one examination by an ophthalmologist experienced in evaluating infants. Because children with syndromes or other disabilities often have hearing loss, other specialty consultations may be indicated including nephrology, pulmonology, and a developmental pediatrician.

The list of risk factors has been reorganized again to a single list that focuses on both early and late onset and/or progressive hearing loss. In the next two slides we list the risk factors. The first risk factor listed, because it's been shown to be highly predictive, is caregiver concern regarding hearing, speech, language. The second risk factor is a family history of permanent hearing loss. The asterisks on risk factors listed, indicate risk factors which are also associated with late onset or progressive hearing loss or place the infant at increased risk of hearing loss. The third risk factor is NICU care for ≥ 48 hours. ECMO places the infant at increased risk of late onset hearing loss. The presence of intra uterine infections, particularly cytomegalovirus (CMV) is the next risk factor. The 5th risk factor is cranio-facial anomalies including those involving the pinna, ear canal, ear tags, ear pits, and temporal bone abnormalities. The 6th risk factor is postnatal Infections including bacterial meningitis. The 7th is stigmata or syndromes associated with the congenital or progressive hearing loss. The 8th is degenerative disorders associated with permanent hearing loss. The last two are head trauma requiring hospitalization and chemotherapy.

Recommendations for early intervention:

A new recommendation is that all families of infants with any degree of permanent hearing loss (unilateral, bilateral, severe or profound) should be offered early intervention services.

Recognized points of entry for early intervention for infants with a confirmed hearing loss should be linked to the state's EHDI system and the services should be provided by professionals with expertise in hearing loss, including educators of the deaf, and speech language professionals.

Finally, both home-based and center-based options should be offered as appropriate intervention.

Primary care provider surveillance: Infants with a risk factor for hearing loss should have at least one diagnostic audiology assessment by 30 months of age, although infants with risk factors associated with increased risk of late onset or progressive hearing loss should be followed more frequently. A referral to audiology by the PCP is recommended if the parent expresses concern about hearing or language.

Surveillance and screening: All infants should have regular surveillance consistent with the pediatric periodicity schedule, which is published by the American Academy of Pediatrics. Auditory skills, developmental milestones, parent concerns and middle ear effusions should all be monitored during well child and sick child visits. All infants should have an objective standardized screen of global development with a validated tool at 9, 18 and 30 months of age. It should include receptive and expressive language and gross and fine motor skills. Children who fail the receptive or expressive language components of this screen should then have a second level language screen such as the REEL or ELM, the Early Language Milestone Scale. This can be performed in the primary care provider office or, the child should be referred directly to early intervention and audiology.

Communication:

Information at all stages of the EHDI process should be communicated to the family in a culturally sensitive and understandable format. Hearing screen information, audiology diagnostic and habilitation information should be transmitted to the medical home and the state EHDI coordinator.

Families should be made aware of all communication options and available hearing technologies. We must continue to remember that family choice guides the decision making process in determining what options are chosen for their child.

Finally, information infrastructure:

States must develop adequate data management systems in order to monitor the quality of early hearing detection and intervention services and provide recommendations for improving systems of care. A linkage between health and education is recommended to determine long term outcomes of school aged children with hearing loss. This linkage is important for planning and establishing both public health and education policy.

This completes the summary of the recommendations. We're going to have a shorter abbreviated "user friendly" summary of the statement that will be available for distribution. Publication is anticipated for 2006. This Statement is a work in progress, and we really hope and anticipate that the 2006 position statement will provide recommendations that will further facilitate the development of more effective and seamless EHDI systems in the United States.

I would be happy to respond to questions or suggestions or recommendations at this time.

<<participant>>

I have a question. This is Carrie in Alaska. Regarding the recommendation that the audiologist and the optometrist show evidence of skills and knowledge in testing young children and infants, who would make that determine nation?

<<Dr. Vohr>>

Well, at this point in time states with their EHDI systems are making attempts to identify which audiologists have the most experience. And in some states, this may evolve over time. We're thinking of this more in terms of system development within states. It is consistent with the development of centers of excellence for children. This goal may still be difficult in some states. But at this point in time, it is the responsibility of the primary care

providers, when they're referring their family for further diagnostic evaluations as part of the comprehensive workup to try to identify those professionals who have expertise. I believe that in more remote rural areas, this is going to be more difficult to achieve.

I don't know if anybody from ASHA is present or not but I know there are going to be guidelines identifying audiologists as a pediatric audiologist. Does anyone know if that's been finalized or put out for public review yet?

<<participant>>

Okay. Thank you.

<<John Eichwald>>

Before Dr. Vohr takes more questions, are other members of the JICH on the call here?

Anyone else join us?

Yes, David Savage.

<<John Eichwald>>

Thanks, David.

Albert Mehl

<<John Eichwald>>

Great, hi Al.

You can ask questions for all members of the JCIH committee.

<<participant>>

This is Terese. I'm sure we all want to say thank you to all the members on the committee who are on the call for your efforts on the document. Certainly, it's wonderful to see the progress that you've made and the updates and recommendations and issues that we struggled with in 2000, because we didn't have the data. Specifically, I think that a lot of our states like Texas will be looking at the issue of neuropathy, we love the idea that screening will be done in both ears, it's nice to have that stated. It always seems it's difficult to get it started. I just wanted to say thank you, I know it's a work in progress.

<<Dr. Vohr>>

Hi, Terese. Well, we owe a lot of the progress made to you in the development of the 2000 statement, for developing the framework, moving the recommendations ahead. The early work was much more challenging because of the lack of evidence. So we're working on updating and I appreciate your comments.

<<participant>>

Dr. Vohr, this is Pat Dewey in Virginia. I have a question about the infections. Are you all planning to clarify the specific list as you did with some of the syndromes? We certainly in the past have had a lot of people who just decided that a particular infection should be included and without any specific guidance on our part.

<<Dr. Vohr>>

Okay. We are working on this to some extent. We were really thinking of culture positive sepsis. Are you talking about the TORCH infections?

<<participant>>

No, the other. The readmission.

<<Dr. Vohr>>

We're having a web cast on March 23rd, where we're hoping to finalize a lot of these recommendations and fine tune them somewhat. I will put your comment on our list of making certain that that is clarified in the document.

<<participant>>

Good. Yeah, that would be the post natal infections. Associated with. It would be very helpful if we knew specifically what they were. So that people just couldn't add Willie nilly.

<<Dr. Vohr>>

Thank you.

<<participant>>

I was wondering if you had guidelines to further define what is appropriate in regards to a diagnostic audiology assessment by 36 months of age.

<<Dr. Vohr>>

I was hoping one of our audiologists on JICH was on the call.

<<participant>>

I am an audiologist and was curious if you were going to give some guidelines.

<<Dr. Vohr>>

It's in the document in terms of what should be included and I don't have the statement right in front of me, but it will be clarified, definitely.

<<participant>>

Thank you. This is Michelle. I'm an audiologist that works on that component. I know a battery of tests, specifically OAE, ABR, and behavioral. So I think it includes the whole battery.

<<Dr. Vohr>>

It does. It will be detailed in terms of specific components. I appreciate the question. One thing that we felt was cost effective improvement, is the former recommendation of having audiology diagnostic assessments every six months for children with risk factors has been changed to at least one audiologic diagnostic assessment by 30 months of age. We know some of the larger states, like Texas and California, it was an impossible task with not enough audiologists to be able to do all of the necessary follow-up. The payoff for identification was not that great. So we will identify children who are considered more high risk and who should be monitored more closely by audiologists. Some recent papers have come out on two areas of increased risk: cytomegalovirus and ECMO. Some infants who are in the NICU greater than 48 hours may also have low or high risk. The onus for monitoring is now shifted to the primary care provider who will be monitoring milestones and doing the screening at 9, 18 and 30 months. Should the PCP identify problems as a result of surveillance and screening, referral for further evaluation should be made at any time. Or if there's parent concern, the PCP would address this immediately.

<<John Eichwald>>

Other questions or comments?

<<participant>>

This is Yaoli from Minnesota. At the state's level, the coordinators are always hesitant to refer to early intervention those children with very mild conductive hearing loss. Should all infants with all degrees of hearing loss be offered early intervention? I mean, many audiologists or states have children with minor effusion, for like 6 weeks or 8 weeks. I'm not very clear about that.

<<Dr. Vohr>>

That's an excellent question. The recommendation does not refer to transient middle ear effusion with hearing loss as mandating an immediate referral to intervention. This recommendation includes permanent conductive mild hearing loss. So in permanent hearing loss, there's an anatomical abnormality, so that we know it's not going to resolve. Children with MEE and mild loss might require monitoring by early intervention. But this recommendation does not include immediate referral of all children with middle ear fusion to EI.

<<participant>>

Yeah. So that's word permanent probably should be added into that sentence?

<<Dr. Vohr>>

Right. And you remember the slides were just abstracted. It was my omission, the word permanent should be there.

<<participant>>

Thank you.

<<participant>>

This is Hallie from California. Back on that same question, what about children with prolonged conductive hearing loss? Does the JCIH have any discussion about children who could have a conductive loss going on three months, four months, five months?

<<Dr. Vohr>>

At the moment the recommendation for referral for chronic MEE effusion lasts for greater than three months, having further evaluation by audiology and otolaryngology. Ongoing surveillance of speech and language within the primary caregiver's office would also be in place, so hopefully they will also be identifying language delays that may occur in conjunction with the middle ear effusion.

<<participant>>

Okay. And that would be include early intervention? We actually have developed a protocol in California that if you have a conductive loss for over three months, that would constitute a reason for referrals to early intervention, and I'm just wondering that's been consistent with what you guys are looking at putting out?

<<Dr. Vohr>>

Does someone else on JCIH want to comment? Because I'm in my clinic and I don't have the statement with me. I've taken a note on this, and we will check on this when we have our conference call on March 23rd. But I believe we had a reference of responding to greater than three months of middle over efusion.

<<John Eichwald>>

Any other questions or comments?

<<participant>>

Hi, this is Raina from high lie. I have a question about your risk factors for hearing loss specifically relating to ototoxic medications. Here in Ohio we've had a lot of confusion about which medications are ototoxic, does the new statement give any guidance in terms of which medications are ototoxic?

<<Dr. Vohr>>

At the moment it really is referring to aminoglycosides, et cetera. One of the issues is that since all infants greater than 48 hours in the NICU are considered to have a risk factor, if you're in the NICU you are very likely to receive ototoxic medications (aminoglycosides.

<<Dr. Vohr>>

For some of the children who have late onset, progressive hearing loss, it's unexpected and not related to specific risk factors. The whole story is not out right now and we still have much to learn about risk factors. I know in our hospital, we finally have a database for the last year and a half, where we can now analyze days of medication that an infant receives. Ototoxic medications in Susan in Norton's study didn't turn out to be a predictor of hearing loss. Recently there was a study on ECMO survivors and they showed that number of days of aminoglycosides was an important association with late onset hearing loss for infants receiving ECMO. So this is a challenging part of the statement. And I think it still needs some work, but it's an excellent question.

<<participant>>

Thank you.

<<participant>>

This is Melinda from Louisiana. I have a question about the new inclusion of auditory neuropathy guidelines, are there going to be any further discussion in the statement regarding recommended early intervention and recommended further treatment for the children found with auditory neuropathy?

<<Dr. Vohr>>

Certainly there is a recommendation of providing immediate referral to both audiology and early intervention and appropriate speech language services and audiology management. I think the question of what's the best audiology management of this group still is not clear because it's such a heterogenous population, with those that resolve and those that do not.

<<participant>>

That's why I asked the question. Is it going to be left to the discretion and professional judgment of the audiologist? Will there be any discussions or recommendations in the statement?

<<Dr. Vohr>>

There will be some recommendations, but I can't delineate them at the moment.

<<participant>>

Thank you.

<<participant>>

Julie from Indiana, we have our regional consultants here from around the state. A question has arisen about the physicians who are going to be providing ongoing surveillance. Is there going to be some kind of intense education provided to pediatricians and family practitioners so that they'll know that we're going to be asking them to be doing these ongoing assessments of children?

<<Dr. Vohr>>

Absolutely and our recommendations for this ongoing surveillance are completely in sync with the American Academy of Pediatrics Bright Futures recommendations which will be published in 2006. The Bright Futures recommendations, is an AAP publication of guidelines of what is appropriate in office management of infants and children. The JCIH specific recommendations on surveillance and screening will be consistent with the AAP Bright Futures recommendations. The AAP does have a web site for informing physicians of recommendations and they send out new information to each provider. The Bright Futures document is sent to every primary care provider and pediatrician. So I think there may be a learning curve as there is with anything else. But I'm hopeful.

<<participant>>

This is Louella from Hawaii, the risk factors page that you have in the documents where the stars are, it's just that particular item that is considered to be needing ongoing monitoring periodic monitoring?

<<Dr. Vohr>>

Those risk factors would require more frequent periodic monitoring than the one assessment by 30 months.

<<participant>>

So for instance the NICU care one, it's just the ECMO or is it the entire list?

<<Dr. Vohr>>

No, just the ECMO.

<<participant>>

Okay. Thank you.

<<participant>>

This is Minnesota with a question about the actual recommendation. You have clearly noted these are not for distribution. However we have a significant training going on tomorrow with early interventionists, can we verbally share some of these recommendations plus we have an article that's being published in one of our pediatric journals, a Minnesota one. Also this month and we've been quoting the 2000, so can we kind of use these.

<<Dr. Vohr>>

I can see utilizing some of the information for training, but since we are still modifying the recommendations they should not be in a publication.

<<participant>>

Okay.

<<Dr. Vohr>>

I wouldn't put it in an article that's going to be published.

<<participant>>

Okay. But at least for training, where it's still verbal.

<<Dr. Vohr>>

All right, Minnesota.

<<participant>>
Thank you.

<<participant>>
This is Ruth in New Hampshire. My question is more of a clarification, I guess, when you said for rescreening you wanted to do both ears, did you mean for anybody who's being rescreened? Not just the NICU population?

<<Dr. Vohr>>
That's correct.

<<participant>>
Okay.

<<Dr. Vohr>>
And it specifically addresses the issue of one ear failing in the NICU and only rescreening that ear. Because there's data now suggesting that some infants can start out as failing in one ear and then subsequently fail in both ears. Also sometimes the ear is mixed up. And the wrong ear is retested. This recommendation is made to avoid any problems related to missing an ear that's having a problem.

<<participant>>
Thanks.

<<participant>>
This is Louella from Hawaii again. So the answer is, that it's for all rescreens, not just NICU babies, right?

<<Dr. Vohr>>
That's correct.

<<participant>>
Thank you.

<<participant>>
This is Hallie from California, again. You had said that you have a different protocol now for NICU babies and it appears the difference is doing ABR screening on NICU babies, so then it's okay for NICU babies who don't pass the inpatient screen to go for an outpatient rescreen? Or is the JCIH recommending that they have diagnostic evaluations skipping an outpatient rescreen.

<<Dr. Vohr>>
I realize this is an issue in California and actually, Michelle King and I were discussing this at 7:15 this morning. We do believe that in the statement we're going to have to be a little bit flexible in this recommendation. Even though we feel that going to a diagnostic maybe optimal, we know that depending on fail rates, there may be a lot of false positives. As long as there is a really good system in place for the rescreen this option could be effective. So even though we currently recommend immediate diagnostic assessment for NICU infants, this is one of the issues, where based on the feedback we got both at the EHDI meeting and your comments now, that I think we have to really think about carefully. We want a recommendation that is practical and implementable in states where there may not be enough audiologists to do all these diagnostics, and we don't want

babies waiting for months for a diagnostic when they could go for a rescreen and be discharged.

<<participant>>

Okay. Thanks, Betty.

<<John Eichwald>>

Any other comments or questions?

<<participant>>

This is Louella again from Hawaii. so NICU babies should have a screening ABR, an automated ABR as part of their screen or are you talking about the diagnostic. You want NICU babies to have a screening ABR, an automated ABR as part of their screen or should they have a diagnostic ABR as part of their screen.

<<Dr. Vohr>>

No, still screening ABR in the NICU.

<<participant>>

Thank you.

<<Dr. Vohr>>

One of the concerns is that if babies only have the OAE. Obviously, the issue is that the incidence of auditory neuropathy has been shown to be somewhat higher in the NICU population associated with certain risk factors and these children also have significant associated issues with language development and communication. Since the hit rate for neuropathy will be higher in the NICU, the hope is to avoid missing the subgroup of babies with auditory neuropathy in the NICU. One of the options is to do both AABR and OAE on the babies. The JCIH recommendation is going to be that as part of their screening process that all NICU babies have a screening ABR.

<<participant>>

This is Terese. I just want to say here, your recommendations are now comparable to what they're doing in the UK. They actually do have both most of the time. They do an OAE, and regardless of the results, they do a screening ABR.

<<Dr. Vohr>>

Okay.

<<participant>>

This is Reena from Ohio again. Perhaps I missed this during the course of the presentation. I just wanted some guidance on when this document would be finalized. Is there any guidance on what time of year this would be finalized? We have a big training piece coming up as well.

<<Dr. Vohr>>

Okay. One of the issues with JCIH is that it's composed of all of the different member organizations. I should say that later this month we're having a web cast meeting going over the document and trying to finalize it as much as possible. Once it gets edited and approved, it's going to have to go through each of the member organizations, for instance, ASHA, AAA, American academy of pediatrics. Once it gets the signatures and approval of all the member organizations, we do believe that it will be published pretty quickly in Pediatrics, and be able to get on all of the websites of the member organizations. It's

difficult to give a date right now; sometimes getting those signatures from the different organizations takes time. Our goal is hopefully maybe the end of the summer.

<<participant>>

Great. Thank you.

<<John Eichwald>>

Other comments or questions? Well, with that, I cannot recall a teleconference where we've had as many questions and comments, so this was a very hot topic for us. Betty, I thank you very much for taking time out of your busy schedule to do this for the EHDI community. I know that everyone's very much appreciative. To all the members on the committee for all the hard work that you have been doing to come up with this very important document that will serve us all.

<<Dr. Vohr>>

And I appreciate everyone's input. I did take some notes and we hope that the final document will reflect a lot of the needs of the states and be able to be implemented very easily. So thank you.

<<John Eichwald>>

Thank you, Betty.

<<Dr. Vohr>>

Thank you all. Bye.