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

Agenda for November 6, 2001

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JUNE HOLSTRUM: Hello, everyone. This is June Holstrum from the Centers for Disease Control and Prevention. Welcome to the November Teleconference on Early Hearing Detection and Intervention. Thank you for joining us.
Before we begin our scheduled program, are there any announcements or comments from any of our participants?
(NO RESPONSE)

JUNE HOLSTRUM: Just to let you know, we are trying out a new technology today to facilitate the participation of persons who are deaf and hard of hearing. Marcus Gaffney will explain that a little bit more.

MARCUS GAFFNEY: Hello, everyone. I would like to extend a special welcome to our users who are using the Caption First service today. I'd like to just take a moment and remind our speakers on this call to try and speak as slowly as possible because there is somebody actually captioning the presentations form this call and then typing those comments into a website where users will be able to read the a text version of the presentations. Please keep this in mind while you are giving your presentations. There may be a delay in comments from users using this captioning service, so we may have moved onto another subject and a question may arise about the previous subject. That's all I have, thanks

JUNE HOLSTRUM: Thank you, Marcus. Are there any questions about the Caption First?
(NO RESPONSE)

JUNE HOLSTRUM: If not, we'll get right on with our first speaker who is Marin Allen from NIDCD to talk about the "Working Group on Communicating the Need for Follow-Up to Improve Outcomes of Newborn Hearing Screening." Go ahead, Marin.

MARIN ALLEN: Okay, thanks. I just wanted to update the group. We had, as you remember, the Indian Health and NIDCD third workshop on neonatal hearing screening. And one of the recommendations that came out of that meeting was a need to examine the health care delivery systems, issues and related communication issues, with the goal of determining methods to resolve the problems of infants who were not returned for follow-up evaluation after an initial assessment of hearing impairment has been made. Let me just ask the captioner if that pace is all right for you.

CAPTIONER: The pace is just fine, thank you. You do not have to speak slowly. I am able to catch it.

MARIN ALLEN: Okay. I just wanted to be sure you were okay because this is -- this is pretty word-heavy. These infants are considered, quote, "lost to follow-up." We planned the meeting along with the help from the National Institute on Child Health and Human Development and the National Institute for Nursing Research and the National Library of Medicine and prepared a literature search on practices from other medical settings, including issues related to medical home, practices of follow-through for other diseases and disorders, and issues that eliminated problems related to geography, communication, and the establishment of need for parents in terms of -- need to the parents in terms of follow-up.

The working group was convened July 23rd, 2001. Both Amy Donahue and I served as co-chairs, [inaudible] many of you who are on this line today who are experts in pediatrics, otolaryngology, audiology, speech and language, ethics, newborn screening programs, health communication, nursing, state health education, special population communication, child health, telemedicine, and epidemiology. We were particularly pleased that we had the Directors of Communication, overall communication, for both NIH and the CDC as participants in the panel. And just so that I am sure to acknowledge the folks that were here, they included: Dr. Thomas Tonniges of the American Academy of Pediatrics; Dr. Patrick Brookhouser from Boys Town; Dr. Judy Gravel from Albert Einstein School of Medicine; Dr. Jackie Jones from the Cornell University Medical Center; Dr. John Lorenz from the Children's Hospital of New York; Carolyn Lund, who is the Director of Nursing for the Children's Hospital of Oakland; Mary Pat Moeller of Boys Town, soon to be doctor; Dr. Isaac Montoya, who is the CEO at Allied Systems in Houston and is member of the NIH, Director of [inaudible] public representative; Dr. Hallie Morrow of the California Department of Health Services and Friends to All Newborns; and Dr. Dena Puskin, Advancement of Telehealth at HRSA; Howard Hoffman and Judith Cooper from the NIDCD -- that's the epidemiology and speech language areas; and we're also grateful for the participation of Irene Forsman of Maternal and Child Health, June Holstrum -- June, your

participation -- and Bobbi Stettner-Eaton of the U.S. Department of Education.

The report is going to cover descriptions of current practices, practical issues of screening and follow-up decision-making, differences in hospitals as compared to and compared with newborn nursery systems of other types, a description of what parents need to know, how to communicate that information, what impedes successful follow-through, what aids [inaudible] communication or best practices [inaudible], and improved [inaudible] professional associations in hospitals.

Health communication professionals [inaudible] for parents [inaudible] and communication strategies and tactics and they're going to be outlined in the final report. This report also includes a summary of potential models for communicating follow-through from various programs in the country and what we're calling a living bibliography of materials. In other words, we're starting with key resources that were helpful to the panel, but we will be adding at the website new resources as they become available, as well as links to all of our traditional groups in terms of being able to reach the resources.

We are making this as an approach to a sort of general federal document. We're trying to make this a much more user-friendly piece that will be bulleted and [inaudible] from without too much additional description, and we expect to have it available within the month. I think that's all unless there are questions, June.

JUNE HOLSTRUM: Any questions for Marin?

CONSTANCE PAGE: This is Constance Page from Anchorage, Alaska. Where would we access that? How will we know when it's available?

MARIN ALLEN: Oh, okay. I was just going to say to June, the second that we've got it available both by web and on paper, June, can I count -- can I ask for your help in disseminating the website address?

JUNE HOLSTRUM: Sure. Just send it to us and we'll send it out to everyone.

MARIN ALLEN: I think it would be easier to do it from your point from the way that the listserve works.

JUNE HOLSTRUM: That will be fine.

MARIN ALLEN: Thanks so much. We appreciate that.

JUNE HOLSTRUM: We'll look forward to getting it.

MARIN ALLEN: Amy, did you have anything you wanted to add?

AMY DONAHUE: No, thanks. It was good.

MARIN ALLEN: We're in different buildings. Well, thanks so much.

JUNE HOLSTRUM: Thank you. Amy Donahue is going to talk about the "Auditory/Perceptual Processing by Infants with Hearing Loss: Issues in Assessment and Management," which was another of the NIDCD discussion groups. Go ahead, Amy.

AMY DONAHUE: Thanks, June. Hello, everyone. This is a new initiative that NIDCD hopes to undertake in the near future. We have spent the last several months considering what we might do in this area. This initiative is, of course, driven by real-time clinical issues that are with us here and now.

Now that we are identifying these infants with hearing impairment at birth, the question is how do we assess and manage these infants. While a lot of programs have spent their efforts and energy getting the hearing screening portion of the program up and running, the next step is the assessment and management.

The clinical decision-making is very difficult. Current practice is not really driven by evidence-based medicine, and the decisions regarding the management of the infants with hearing loss are crucial. They have to be made at a very

early time period. We don't have tools and techniques that are really sensitive enough to measure and evaluate progress of various types of intervention programs.

So Marin had mentioned, I think, that we had a research workshop last September, in September of 2000, and those minutes are up on the web. They had several recommendations and a couple of them NIDCD has acted upon. But one, in particular, was this issue of better characterizing the auditory perceptual performance of infants with hearing impairments. In order to get a better handle on that and really address it in a more multi-disciplinary way, we had a smaller discussion group, and we included not only speech and language and auditory scientists, but we included cognition and memory and learning. The intent is to examine each infant as a whole and each infant as an individual and to try to understand exactly the relationship of hearing loss to auditory and perceptual processing.

The group had considerable discussion. We identified some research needs that are clearly urgent and we would love to see the community send grant applications in. Some of those, for example, are developing techniques and methods for looking at not only threshold but supra-threshold auditory/linguistic abilities, the understanding of multi-modal perception, and its impact on linguistic function in infants with hearing loss, devising and developing better tools to evaluate progress with various strategies, trying to understand the individual variability that we see in all of these infants, and examining and determining ways to evaluate the appropriateness of some of our technologies or strategies or habilitation approaches that we're taking.

We'll be talking to other NIH institutes -- for example, the Institute of Child Health, maybe Mental Health -- and hopefully, we will have some sort of an initiative or request for applications from the research community in the near future to help us address some of these research questions. So let me stop there if there's any questions.

JUNE HOLSTRUM: Any questions for Amy?
(NO RESPONSE)

JUNE HOLSTRUM: And Amy, is this report on the internet yet?

AMY DONAHUE: No, it's not.

JUNE HOLSTRUM: Okay. Good. Because I looked and I couldn't find it. So I didn't just miss it. It wasn't there, huh?

AMY DONAHUE: No.

JUNE HOLSTRUM: Okay. Our next speaker is Aileen Kenneson and she's going to give us an update on the CDC genetics project.

AILEEN KENNESON: Okay. Good afternoon, everybody. I'm glad to have the opportunity to update everybody on the details of this project. Three of our Level 2 states are working on this project. The goal is to describe the etiology of hearing loss in infants and young children. These states are Hawaii, Rhode Island, and Utah. And in addition, we're working with Georgia to try to pilot the project in two of their counties. Utah was funded at Level 2 last year and has developed the protocol that we hope will be used in the other states as well. The protocol has been through the CDC IRB, and after we make a few changes to the informed consent forms, they'll be getting underway in Utah. And then once we get the protocol through the IRB's in the other states, we'll be ready to begin there as well.

Families of infants identified with hearing loss through the universal newborn hearing screen will be eligible for participation in the project. In addition, children identified with a hearing loss before the age of three will also be included. Participation in the study includes a visit to a clinical geneticist to try to establish the cause of the hearing loss. Information about family history, risk factors, and medical history will be collected, and a physical exam will help determine whether the child has a syndromic or a nonsyndromic hearing loss. Referrals will be made to specialists as appropriate, such as ophthalmologists and cardiologists. Families will also be offered the opportunity to participate in the second part of the study which looks specifically at genetic factors involved in hearing loss. Specifically, we're studying the connexin 26 gene and two mitochondrial mutations. All three of these genes are involved in nonsyndromic hearing loss. Results of these tests will be given back to the participants, along with appropriate genetic counseling services. We are also asking for permission to store DNA samples for future

investigations of the contribution of other genes to hearing loss. So let me just take a few minutes to tell you a little bit about each of the three genes that we have chosen for the study. The connexin 26 gene, which is also called the GJB2 gene, encodes for a protein that's --

UNIDENTIFIED SPEAKER: Aileen --

AILEEN KENNESON: Yes?

UNIDENTIFIED SPEAKER: -- we keep losing you on phrases and sentences.

AILEEN KENNESON: Oh, okay.

UNIDENTIFIED SPEAKER: At least we do in Minnesota.

AILEEN KENNESON: Well, let me know if you missed anything that sounded really pertinent.

JUNE HOLSTRUM: Or you can read the transcript.

AILEEN KENNESON: I'll keep going and just interrupt me if there's a big gap. The prevalence of the connexin 26 mutation varies greatly from population to population. For example, mutations in this gene account for about eight percent of cases of nonsyndromic sensorineural hearing loss in Korea –

CAPTIONER: This is the captioner. I am also having a lot of trouble with you. Are you on the speakerphone?

AILEEN KENNESON: Yes.

JUNE HOLSTRUM: Why don't you try talking directly into this.

AILEEN KENNESON: Okay.

JUNE HOLSTRUM: We don't have a -- We're on the bat phone.

AILEEN KENNESON: AILEEN KENNESON: Okay. So mutations in this gene account for eight percent of cases of nonsyndromic sensorineural –

FRANK BOWE: Excuse me, Aileen. This is Frank.

AILEEN KENNESON: Yes.

FRANK BOWE: Are you talking about chromosome 6? What is a --

AILEEN KENNESON: Connexin --

FRANK BOWE: What is a section 26 gene? Where can we read plain English versions of your CDC genetics project?

AILEEN KENNESON: It's the connexin 26 gene. That's the name of the gene, and I think we have some information on our website. And if we don't, I'll try to put some on there.

FRANK BOWE: This is also from Frank Bowe. If you say it's on the web, it would be helpful if you would say what URL it is. Thanks.

AILEEN KENNESON: Okay. I have no idea what that is right now.

JUNE HOLSTRUM: The URL is www.cdc.gov/ncbddd -- which stands for National Center for Birth Defects on Developmental Disabilities -- then /ehdi. And then on that, just look down the list for the genetics project.

AILEEN KENNESON: I know we have some information about, in general, genetics of hearing loss on that website. And if that doesn't answer your questions, then you can feel free to e-mail me. I would be glad to answer any questions. Getting back to the connexin 26 gene, there are over 90 different mutations in this gene that are associated with hearing loss. Two mutations are common in Caucasians in the Ashkenazi Jewish populations. And because they're common in these populations, they've been very well studied and we really understand them pretty well. They're associated with moderate to profound hearing loss.

However, of the over 90 other mutations that have been described, we're really lacking data on most of them. So that's one of the goals of the study. We also need data on the contribution of connexin 26 mutations to hearing loss in diverse American populations.

And in addition, most of the current data is related to hearing loss identified in childhood. So we need data that's specific to hearing loss that's identified at birth. Those are just, in general, the connexin 26 gene issues that we're hoping to address in the study.

The second gene that we're looking at is the mitochondrial mutation. It's specifically the A1555G mutation, if anybody is on that's interested in that kind of detail. This is the mutation that's been implicated in hearing loss that results from the use of aminoglycosides, such as streptomycin. There is also recent data that suggests that this mutation, in combination with mutations of other genes, can cause hearing loss without streptomycin use. So we'll be looking at the contribution of this mutation to hearing loss in infants and young children.

The third gene that we're looking at is also mitochondrial mutation. We're including it because it's an easy-to-test-for mutation and there's really very little data on it. A few studies have been done in China and Japan, but I think the only study that's been done in the U.S. looked at about 60 people. So there's really very little information on that mutation. Because it's easy to test for, we're including it.

This project is unique in that it's a population-based study of hearing loss done in conjunction with the state-wide newborn hearing screening programs. Previous studies have generally included children whose age of onset could only be guessed at because they hadn't been tested at birth. So, in addition to newborns, we're also including children who passed the newborn hearing screen but identified with a hearing loss before their third birthday. This way, we can determine which genes and risk factors are associated with hearing loss that is present at birth versus hearing loss that develops later in infancy.

This is important, for example, in hearing loss caused by connexin 26 mutations because connexin 26 mutations cause a large proportion of childhood hearing loss, but we don't know how often this hearing loss is present at birth or if it develops later in infancy. So we don't know how often -- how many of these cases will be picked up by the newborn screen.

A second limitation of previous studies is that they generally excluded syndromic cases and cases of hearing loss attributed to environmental factors such as infections and antibiotic use. It's possible that mutations in genes such as connexin 26 might increase the susceptibility to other genetic and environmental factors, and this will be the first study that's ever assessed this possibility.

Furthermore, previous studies have often failed to meet complete ascertainment of all individuals with hearing loss, especially mild and unilateral cases. So by including all the cases in this project, we'll be able to fill in some of the gaps in the literature on the association between various factors and mild hearing loss.

So just to wrap up, this study will look at the causes of hearing loss in infants and young children, including genetic causes. The study has been designed to fill in many gaps in the literature related to genetic causes of hearing loss, particularly in reference to the connexin 26 gene. The project should begin in Utah in the next month or so, and we hope to have the other states on board shortly thereafter. The project will collect data for three years and will also be creating a DNA bank that can be used in future studies. So are there any questions?
(NO RESPONSE)

JUNE HOLSTRUM: All right. If there are no other questions, then we'll go into our last topic. Most of you have read the Preventive Services Task Force report on universal newborn hearing screening and many of you have voiced your concerns about this report. So we have invited representatives of five different agencies and

organizations to give the response to the report, but before I call on the first speaker, sort of to put things in perspective, or maybe to confuse things more, I would like to read a quote from Diane Thompson, who is one of the members of the Task Force. This quote is taken from HealthScout, which is an on-line news report. In this, Diane is quoted as saying: "We are concerned that the media will interpret our report to say we are not in favor of universal screening and that is not true. It's just that the evidence to show whether universal screening can improve language skills for these children is not clear." So with that little introduction, I'll turn it over to Scott Grosse from CDC to give

SCOTT GROSSE: Thank you, June. I'm sure most of you, if not all of you, have heard that the U.S. Preventive Services Task Force has issued a new report on newborn hearing screening. While the Task Force reports clear evidence that universal newborn screening is a reliable test that results in substantially earlier identification of children with hearing loss, the report concludes there is no clear evidence that newborn hearing screening results in improved long-term language outcomes. Consequently, the Task Force report concludes there's no clear evidence that newborn hearing screening results in improved long-term language outcomes. Consequently, the Task Force concluded that the data are insufficient to recommend for or against routine newborn hearing screening. On behalf of CDC, I would like to offer some clarification and explanation of what the Task Force report means for EHDI programs in the states.

First, the Task Force is an independent panel of experts in primary care and prevention that reviews evidence of effectiveness and develops recommendations for clinical preventive services. The Task Force is sponsored by the Department of Health and Human Services through the Agency for Healthcare Research and Quality, or AHRQ. The systematic review of evidence on newborn hearing screening was actually prepared by staff of the Oregon Health Sciences University working in collaboration with the Task Force.

It is important for EHDI programs to understand the criteria used by the Task Force to evaluate evidence of improvements in language development resulting from early identification. The Task Force concluded that the types of studies needed to demonstrate long-term benefit have not yet been done. The Task Force applies very strict criteria in evaluating research findings. The primary standard used to assess evidence is whether randomized controlled trials have been conducted. No randomized controlled trial, or RCT, of hearing screening to evaluate language outcomes has been conducted. The only RCT of any newborn screening test that has looked at long-term outcomes was a trial of screening for cystic fibrosis in Wisconsin from 1988 to 1994. That RCT involved screening all newborns but not telling half the parents the result of the screening test. All children who screened positive were followed up years later and outcomes were assessed. Such a study design would not be considered ethically or legally acceptable for hearing screening, at least in this country. The traditional and widely-accepted newborn screening tests, such as for PKU and hypothyroidism, have never had a RCT prior to their recommendation and general use in public health.

The alternative to a RCT is analysis of observational data to assess how language outcomes are influenced by early identification to newborn hearing screening. It is well-known that deaf and hard-of-hearing children typically have serious delays in language development which affects school performance and occupational success. Several studies have been published that report dramatic improvements in language development of children with hearing loss that is identified in infancy and followed by early intervention. Children with hearing loss who receive appropriate treatment before six months of age are reported to have language development approximately one standard deviation better than those with comparable hearing loss not identified until two years of age or later. The Task Force concludes that these observational studies all have serious methodological limitations that limit their conclusions. CDC agrees with the Task Force that the studies each have limitations. While inability to control for potential bias can lead to overstatement of benefit in observational studies, the strength of the association observed across studies raises the probability that a true effect is present.

CDC agrees with the Task Force that more rigorous research needs to be done to establish the long-term impacts of early identification on language development. As children identified through universal newborn hearing screening programs are now reaching school age, it will be possible to conduct studies of language development in school-age children, as recommended by the Task Force. We welcome input from the Task Force and from AHRQ, as well as collaboration from state EHDI programs, in developing new research protocols using observational data from EHDI programs that would meet the evidence-based standards set by the Task Force.

While additional studies are being conducted, CDC recommends that states continue working to ensure that children

receive hearing screenings, appropriate diagnosis, and effective intervention services. The development by states of data systems and registries of children with hearing loss is essential to ensure that children receive intervention services and to allow for population-based evaluation research to be conducted. The role of state EHDI programs is critical in resolving the challenges posed by the Task Force.

In terms of how the report may affect EHDI stakeholders, recommendations of the Task Force are primarily directed to clinicians. If health care providers raise the issue of this report with state EHDI programs, they can be directed to statements from professional organizations that support early hearing detection and intervention. Links to these statements can be found on our website as well as the NCHAM website.

Third-party payers may also consider Task Force recommendations in making reimbursement decisions. It is unclear whether the new Task Force report will affect future reimbursement decisions regarding newborn hearing screening.

Finally, the Department of Health and Human Services continues to support the Healthy People 2010 goals of ensuring children are screened for hearing loss by the age of one month, are diagnosed by three months, and enter into intervention by six months. CDC remains firmly committed to supporting state-based EHDI programs and research. Thank you.

JUNE HOLSTRUM: Are there any questions for Scott?
(NO RESPONSE)

JUNE HOLSTRUM: If not, we'll go on to Irene Forsman who is with Maternal and Child Health at HRSA.

IRENE FORSMAN: Scott, thanks for that very comprehensive response. We prepared a statement that would be released if we got any calls from the press. The statement said simply that we agreed with the Task Force, that the long-term research wasn't there, and it was a good thing that the EHDI programs are being implemented because that would provide the population on which to carry out the long-term research. We did not refer to the fact that it was unethical to do controlled studies in this country at this time. Our general approach is to not make much of the report.

JUNE HOLSTRUM: Thank you, Irene. Let's go onto the National Association of the Deaf and Frank Bowe.
(NO RESPONSE)
JUNE HOLSTRUM: Frank, are you still with us?
(NO RESPONSE)
JUNE HOLSTRUM: Okay. Maybe he'll join us in a little bit. Let's go on then to Jim Potter who will be speaking for ASHA.

JIM POTTER: Thank you, June. And I want to just thank the work done by Scott on this issue and by Irene. I am very indebted to both of your work on this issue and very much appreciate your stand. However, while they're both federal agencies and must put things in a more politically-correct context, I'm a little freer to speak our mind. I think that we have issued a press statement and it's available on our website, and if you would like, I can send it to folks who are interested in it.

Essentially, I would like to take my short report and talk a little bit more about some of the process. I think ASHA is very disappointed not only in the process, but I would term it as somewhat reckless recommendations that went beyond the scope of the research review that was done. I can describe from a [inaudible] standpoint an individual from the CDC and ASHA went to the Task Force meeting this summer when this was being considered and were not allowed to testify at that meeting. Also, the issue is not quite as straightforward, and perhaps Scott can correct me if I'm wrong here.

But the committee that was -- that had taken this on, the subcommittee -- or the committee of the entire Task Force voted in favor, a majority vote in favor. However, to pass approval by the Task Force to get a B recommendation, it needs a two-thirds majority vote and that we did not get. Is that correct, Scott?

SCOTT GROSSE: No. No, that's not quite correct.

JIM POTTER: Okay, thank you.

SCOTT GROSSE: The final vote on the recommendation was taken on September 30th. I do not know what that vote was. I was not present at that meeting. There were a number of intermediate votes. And so it's -- I can't say what the decision -- how many voted for or against at the final vote. The group that met in June was inconclusive.

JIM POTTER: We lost part of what you said, Scott.

SCOTT GROSSE: The final vote on the recommendation was taken on September 30th. I do not know what that vote was. I was not present at that meeting. There were a number of intermediate votes. And so it's -- I can't say what the decision -- how many voted for or against at the final vote. The group that met in June was inconclusive.

JIM POTTER: Thanks. However, this is an improvement over the last recommendation which I believe was a C rating, somewhat of an improvement to the I category. I would probably echo many of the comments made about the unethical ability to do a prospective double-blinded clinical trial for children in the U.S. In fact, this same group, just to give you some perspective, was until recently reluctant to endorse mammography screening. And the only way that they won that recommendation was an analysis of clinical trials work being done in Europe and some of the western European countries formerly in the Soviet Union. I'll also let you know that I did an interview with Reuters yesterday and that is published -- was published last night. That takes a pretty hard line against the Task Force. And I think that we believe, as an organization, that someone needed to push back a little against the agency -- or I should say the Task Force just so that if different payers or other employers or insurers were thinking about making moves against -- or minimizing EHDI screening or the intervention services that at least they would have a little bit of a fight on their hands. So that's, in a nutshell, our perspective, and again, I'm happy to forward on some of these articles or the press release, if so desired.

JUNE HOLSTRUM: Any questions for Jim?

(NO RESPONSE)

JUNE HOLSTRUM: If not, we'll move on to the American Academy of Pediatrics perspective with Michelle Esquivel.

FRANK BOWE: Excuse me, this is Frank Bowe.

JUNE HOLSTRUM: Oh, yes, Frank.

FRANK BOWE: The National Association of the Deaf is concerned that all the publicity over this Task Force report may slow down or even squelch newborn hearing screening programs. We think it is very important that everyone on this call do all you can to make it clear that this Task Force did not, in fact, recommend against newborn hearing screening. Let me add that a recent conference on this topic co-sponsored by the National Association of the Deaf strongly recommended that physicians -- This is the interpreter and that's where it has stopped -- strongly recommend that physicians, hospital personnel, and others tell families about local adults who are deaf. We think that it is urgent that family members know that adults who are deaf, like me of course, are happy, well-adjusted campers. Thank you.

JUNE HOLSTRUM: Do we have any questions for Frank, realizing that you can ask them now and they may be answered later?

(NO RESPONSE)

JUNE HOLSTRUM: If not, we'll go on to the American Academy of Pediatrics. Michelle, are you with us?

(NO RESPONSE)

JUNE HOLSTRUM: If not, we'll try Patrick Brookhouser with the Joint Committee on Infant Hearing. Pat?

PAT BROOKHOUSER: Yes. The Joint Committee on Infant Hearing is going to have a conference call on this issue but, obviously, there's a high level of concern expressed by various members in e-mails to me that this in some way might derail what has been sort of a long-term process by the Committee to help promote screening nationally. I personally, as a physician, am very concerned about misdiagnosis at birth and the fact that these parents, if they're not given an opportunity to have their babies' hearing screening, may go on for a fair period of time without an opportunity to obtain a diagnosis. And I think, obviously, this committee -- I've worked on other projects in terms -- that have involved the U.S. Preventive Services Task Force and the level of evidence that they demand in terms of

double-blind studies are not possible in this area. They should have spoken to that. They should have spoken to the ethical issues that would be involved in withholding intervention for children with educationally-significant hearing loss. That was not, to my knowledge, addressed at all. I also have Mary Pat Moeller and Michael Gorga with me who may wish to make an additional comment.

MICHAEL GORGA: This is Michael Gorga. I'm sort of new to this. I do screenings, but I've not been involved in what appears to me to be a very political process. However, in reading this report, there are some things that I thought were a little confusing and that I did not know exactly how to respond to.

One of the problems -- one of the things the report gets right is that there are not a lot of good data out there to talk to test efficiency. That is because that's an impossible task to do. I was part of one project that attempted to do that kind of a study. It was extremely difficult. With heroic efforts, we were able to find, from seven different sites, 56 infants, 86 years, with hearing loss. That's the basis upon which you build your statistics regarding test sensitivity. That's an extremely small number and, as a consequence, one needs to be careful in interpreting those data. But even so, looking back at those data, it seems to me that there are things in the report that don't particularly match up. For example, the refer rates that they describe in here, who are two-stage screenings, are much higher than we obtained in our sample. Our two-stage referral rates were between about two percent and about one and a half percent, not the seven, 10, 12 percent that seems to be referred to in this document. In addition, the goal that is stated in this document is to identify moderate, severe, or profound hearing loss. In point of fact, we do reasonably well identifying moderate, severe, and profound hearing loss even in the sample -- the paper they seem to like, the Norton, et al., paper in which the sample of hearing-impaired children was very, very small. In a larger project that we performed where we had about 800 years with hearing loss -- of course, these were older subjects, but we have no reason to suspect that test performance would differ between young and old subjects -- when we looked at our statistics for that group for moderate, severe, or profound hearing loss -- and that's a loss exceeding 40 dB -- we had hit rates on the order of 99 to 100 percent. Yes, the error rate does jump up when you have mild hearing losses. That's a matter of fact of all of these measurements. And in the paper on older people, as well as this paper on -- that describe results from infants who graduated from a newborn hearing screening program, yes, the error rate is higher among babies with mild hearing loss, but I don't believe that, in and of itself, is a major criticism of newborn hearing screening.

So my problem is with some of the statistics that are described relative to test sensitivity and test specificity. We do ourselves a disservice if we say that there is no false negative rate. Of course, that's untrue and we should be careful not to say that. And in point of fact, we do miss babies sometimes with mild hearing loss. Not all. We catch a good number of those babies, but when we make mistakes, they are far more likely to occur with babies having mild hearing loss. But I think the statistics are not quite as unfavorable in the data that I'm familiar with as compared to this report.

As a final notion, I think -- I heard this from the first speaker -- Scott Grosse and Dr. Brookhouser also reiterated this -- it is simply impossible to do the kind of study that I think is being requested by this service. You simply cannot deny service to children with hearing loss because you want to see if they're going to develop speech and language differently than the children you identify early in life who do have hearing loss and you do provide service to. It is simply a morally and ethically impossible thing to do. You cannot do prospective research.

MARY PAT MOELLER: This is Mary Pat Moeller. The only comment I would add is I'm in agreement with both Dr. Brookhouser and Dr. Gorga. I think that it's relevant for us to consider what a low-incidence population we're working with in relation to the need to design well-controlled studies. I would advocate for better science certainly, but the ruler that they're holding us up against, as has been stated, is an impossible one. I think we need good science that might involve collaboration among centers because I know I'm currently involved in a prospective longitudinal study and the numbers of children we're enrolling are quite small. And that's going to happen with a low-incidence population with a great deal of heterogeneity within that population.

So it would have been good for them to acknowledge the difficulties in doing research on this topic and not hold quite as high a standard. And I echo the concern, I hope that this does not derail or deflate enthusiasm about this issue because as someone in the trenches, we are every day seeing the benefit of universal newborn screening in terms of developmental outcomes for children.

SCOTT GROSSE: This is Scott Grosse. I would like to clarify a few points that Dr. Gorga raised.

First, the Task Force did not insist that randomized control trials be done. They recommended that additional studies of observational data be conducted. And we are going to try to work with them to design studies with other collaborators -- some of you who are on the phone, no doubt -- to conduct studies that will meet their criteria. The primary standard, or the gold standard, the Task Force uses is the randomized controlled trial. If you want an A rating, you need to have a randomized control trial. They will recommend interventions on the basis of observational data, but that is a less strong recommendation and requires more cumulative evidence.

In terms of the test characteristics, we chose not to emphasize that because the Task Force did acknowledge that the testing is highly reliable. The statistics they report are confusing because the definitions they used are idiosyncratic. The definition of "screening" the Task Force chose to use is that screening is only done before discharge from the hospital. Anything that occurs after discharge is not considered screening in their definition. So that when they say two-stage screening, they mean two screening tests done in sequence before discharge. It is not the kind of two-stage screening that EHDI programs necessarily talk about.

Further, when they talk about sensitivity, they are not talking about the actual sensitivity of the screening test, even though they state that. The sensitivity is what proportion of children who are later identified at one year of age to have hearing loss are picked up as a result of screening. Those children who were never screened or never followed up for further testing or who had late onset hearing loss are considered missed cases. So the sensitivity is calculated by taking all children who were not picked up through the screening program and subtracting that from unity.

MICHAEL GORGA: Well, if I could just respond to them. Obviously, there's a last point. One cannot identify a hearing loss before it exists, so late-onset hearing loss -- some of the genetic causes of hearing loss which can be progressive, there's some thought that cytomegalovirus has a late onset. It's hard to think of how a screening measure in the neonatal period would be able to pick those up.

But the second clarification is, I think that is what -- your characterization of a two-stage screening program is not exactly in agreement with what I think many two-stage -- what many screening programs do. For example, in our screening program, we do a two-stage screening program and we do it prior to hospital discharge, and our referral rate is around one percent right now. So I think we're not unique in this. I think other hospitals are moving toward plans in which they'll do, for example, an OAE screening first. Any child who passes that is not referred on, but any child who fails that is not automatically referred or not asked to come back at two or four weeks later but, rather, moves to, say, an ABR screening or a second OAE screening in some cases.

So I think a lot of programs are doing a two-stage screen and are doing them before hospital discharge. I think the follow-up problems are so significant that most people agree that if you could do that second stage prior to hospital discharge, you would have a much better success rate.

FRANK BOWE: This is Frank Bowe. Could anyone here estimate the proportion of infants and toddlers lost to follow-up so far? Is that a really serious problem? We screen, but families never get the follow-up test. How big is this problem?

MICHAEL GORGA: I think -- This is Michael Gorga again. I think it's a very serious problem. In the report that I was looking at, it was 30 percent. I would put it 30 to 50 percent. And yes, indeed, that is a serious concern. And one of the concerns I have with this report is not necessarily how this report -- what this report says, but more how it's interpreted. If people start saying, gee, let's not have those kids return, then it's only going to get worse. But, yes, it is a very serious problem and the follow-up is not what those of us who do this would like it to be.

PAT BROOKHOUSER: This is Pat Brookhouser. I think Marin Allen's point early on was that her efforts are being devoted to try to take care of that. Simply because we're not getting called doesn't mean the program is bad, it means that we need to add other interventions to the program to make sure they come back.

JUNE HOLSTRUM: This is June at CDC. And our data has been showing that about 50 percent are lost to follow-up, but keep in mind that the data that we get is from the states. We would expect that those who -- those states that are able to send us data are probably the better states. So, at best, we're probably getting about 50 percent.

UNIDENTIFIED SPEAKER: I agree. I also agree with Marin. You know, we get better on another front.

PAT BROOKHOUSER: Does anyone have any comments about the stress issue? It's interesting if there's a problem with returning, if people are stressed worrying about the possibility of a hearing loss and then they fail to return, somehow that doesn't compute unless they're afraid to find out for sure, which I don't think is the case. I think that that -- an awful lot is made of that. And yet, when we have the Joint Committee hearings in Washington, the parents said we would want to know. I mean, there's no way that they wouldn't want to know what was going on as early as possible, and I think to -- I mean, all of us are stressed by having to go in for all kinds of tests, but the fact is I think that's a bad argument to use for not doing it.

JUNE HOLSTRUM: Was that Pat just speaking? Who was our last speaker?

PAT BROOKHOUSER: Pat Brookhouser.

JUNE HOLSTRUM: Okay. I thought that was you, Pat. Okay, thanks.
Are there any other comments from any agencies, or groups, or individuals that would like to get in a few words before we close here?

BETH BENEDICT: Oh, I'm sorry. This is Beth Benedict. Yes, I'm always concerned about the follow-up. I have two deaf children. After the screening, no one checked with me what I was going to do with them although I knew what to do with them. It's scary, especially for hearing parents with no knowledge in deafness.

JIM POTTER: June, this is Jim Potter. I would just make the comment that there are known problems in the Part C -- IDEA Part C early identification program, zero to three. We feel like that and we have had complaints from a number of states to that regard. That program has been woefully under-funded. However, we have made some headway this year. The House Appropriation bills moves that funding level up from about an average of about eight million dollars over the last three to four years per year to 46 million dollars this year. I mean, we'll be working with the Senate conferees to help ensure that that substantial level increase is hopefully obtained this year. Because the lack of funding to that problem is a great contributor to this follow-up problem.

JOY O'NEILL: This is Joy O'Neill in Texas. Because the Department of Health in Texas has a mandate to ensure that babies do get into follow-up services, we have a series of letters and phone calls that we make, and it seems to be working. We're just beginning this process and maybe at a later time we can tell you how it's impacting our follow-up rates, but we are getting really positive response from families who get our letters and we're getting a lot of phone calls back to say, "We had our baby tested and everything is okay." And we do a two-stage screening prior to discharge, also.

MARTHA CARMEN: Hi. This is Martha Carmen in Congressman Walsh's office. I wanted to thank Jim, first of all, for the word on the appropriations side because I was going to say something about IDEA, but I'll pass over that and say that possibly, Irene and June, maybe we need to re-look at the follow-up mechanism as the states are developing their programs. Maybe that component needs to be re-evaluated since the numbers are still so high. I do get some complaints also along the same lines here sort of at the ground level.

JUNE HOLSTRUM: Thank you, Martha, and that is a major concern for us here at CDC and we are looking into some various mechanisms, looking into that and seeing if we can come with some strategies that could help the problem.

MARTHA CARMEN: And can I -- one question for Jim Potter, if I may. Jim, what is the next step for ASHA in terms of this report?

JIM POTTER: The Task Force report?

MARTHA CARMEN: Correct.

JIM POTTER: At this point, I think we'll monitor it. I think some of these reports that have been issued by the Agency or in conjunction with the Agency at times tend to fall on deaf ears. So it really, I think, goes back to the point -- The Task Force did raise, I think as Scott and others have mentioned, some good points that need to be addressed in perhaps the research. However, if that starts to impact the policy-making deliberations of state legislatures or employers in their health benefit plans or insurers and things like that, then we will probably step up

our advocacy efforts to counter some of the information. Again, as was stated, our concern is that the information will be misconstrued and give someone the opportunity to deny these programs for benefits to those who need it.

PENNY HATCHER: This is Penny in Minnesota and just a comment about the follow-up. Anecdotally, what we hear in Minnesota is it is the providers, the physicians and nurse practitioners, that are also kind of poo-pooing the need for rescreening, that it's not alarming, and now I'm even more concerned I'm not hearing what AAP had to say. It is the physicians in already in Minnesota that are raising the red flag with the JAMA report and, you know, wondering what are we going to do here in Minnesota, are we going to keep going forward. So June and Irene, did the AAP say anything?

IRENE FORSMAN: I haven't heard that they've made a statement. I heard that they were preparing one. But sometimes getting things through their hierarchy takes some time.

PENNY HATCHER: Okay, thanks.

KARL WHITE: Hi, this is Karl White in Utah. I know that Michelle and Lou Cooper are working on a statement from the AAP and I would expect we'll see it within the next few days.

FRANK BOWE: This is Frank Bowe. On IDEA, Part C, I still only see eight million dollars, no jump in funding.

JIM POTTER: Frank, then you're not looking at the right report. The House bill, in fact, did a 46-million-dollar increase. The Senate was level-funded. Those were the final bills passed. The Senate hasn't passed its yet. So that still may change on the Senate floor, but I believe that they were debating it today. So I can't give you any final numbers on that.

JUNE HOLSTRUM: Any other last comments?

UNIDENTIFIED SPEAKER: Thanks. This was so invaluable.

JUNE HOLSTRUM: Well, thank --

KARL WHITE: June, this is Karl White. I have a question for Jim. Jim, do you know what's happening on the Senate side with respect to the Walsh Bill appropriation? Is that in process?

JIM POTTER: That's still in process and we have some meetings arranged with some of the conferees to discuss that and flush it out a little bit more. We do have some concerns about that and it is one of the issues that we'll be raising besides the IDEA Part C funds. But since this is the last year of the authorization, we will be making -- please be assured that we will be making a very dedicated push to make sure that we can get as much funding as possible for both the agencies in question.

KARL WHITE: Now, when you said conferees, has the Senate side passed a version of the appropriations bill that would impact on the EHDI programs?

JIM POTTER: It's being considered today --

KARL WHITE: Okay.

JIM POTTER: -- I believe.

KARL WHITE: Okay.

FRANK BOWE: This is Frank Bowe. I'm looking at ed.gov, latest numbers as of yesterday.

JIM POTTER: Frank, this is Jim Potter. If you would like to have a discussion about this off-line, I would be happy to give you my number, which is 301/897-0125. That way we can go into specifics of where the funding is.

JUNE HOLSTRUM: I believe our time is up. I want to thank you all for joining us today. Our next meeting will

be in January. And in January, we usually have the second Tuesday to allow people to get back from their Christmas vacations. So, again, thank you for joining us and we'll talk to you in January. [Whereupon, the teleconference was concluded at approximately 3:04pm]