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EHDI E-MAIL EXPRESS

The monthly newsletter of AAP Early Hearing Detection & Intervention Program

This is an e-mail communication from the American Academy of Pediatrics (AAP) "Improving the Effectiveness of Newborn Hearing Screening, Diagnosis and Intervention through the Medical Home" project funded through cooperative agreements with the Maternal and Child Health Bureau (MCHB), Health Resources and Services Administration (HRSA) and the Centers for Disease Control and Prevention (CDC), National Center of Birth Defects and Developmental Disabilities (NCBDDD). It is designed to provide AAP Early Hearing Detection and Intervention (EHDI) Chapter Champions with resources and current clinical and other information. The EHDI E-Mail Express is sent on a monthly basis. Please feel free to share the EHDI E-Mail Express with colleagues working on or interested in childhood hearing detection and intervention issues. Distribution information appears on the last page.

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FROM YOUR CHAIRPERSON

Friends,

I'm old enough to remember measles. I'm old enough to remember German measles (rubella), and mumps, and H. flu meningitis, and a host of the other medical conditions that, thanks to vaccinations, are mostly a thing of the past, at least in the United States.

And I'm old enough to remember when newborn screening meant blood spot screening. Period. I'm even old enough to remember when blood spot screening meant testing for only one disease, phenylketonuria or "PKU." We called it the "PKU" test, even after gradually adding more tests, including a test for congenital hypothyroidism, and then a test for abnormal hemoglobin molecules such as "sickle cell" hemoglobin, and eventually a host of other tests. Despite this expanded panel of additional dried blood spot tests, we still called it the "PKU test" for many years (and sometimes we still do). But gradually we came to realize the potential of testing for rare disorders when early identification could facilitate early treatment, resulting in the avoidance of serious, even life-threatening complications.

And then, in concert with all of you who are reading this column, together... we broke the mold. We proposed a newborn screening test that was not performed on a sample of dried blood, but was instead performed with newly available technologies, a physiologic test of newborn hearing. And we discovered that congenital hearing loss was much more common than all of the blood-spot-screened diseases combined!

No, it wasn't always an easy journey. But today thousands of families around the world would eagerly step up to say, "Not easy... but worth it!" Despite the bumps along the way, and despite our continuing struggles to assure timely follow-up after a baby does not pass the initial hearing screen at the hospital, so many children born deaf or hard of hearing, and their families, now have the opportunity for early identification and early effective intervention.

During the past couple of years, I have followed the widespread introduction of another newborn screening test. The test isn't done on dried blood spots. Instead, it is a measurement of blood oxygenation, a test to assess if the baby who looks pink might actually be just slightly blue, a test to screen for critical congenital heart disease. I am not involved with any of the planning or implementation of this recent newborn screening initiative. But I remember all the steps involved. I remember the research grants and early trials. I remember grinding the data to assess the frequency of the condition in question. I remember calculating the costs of universal screening to determine if a mass screening program could be justified and estimating the benefits of early intervention over the morbidity associated with delayed recognition. I remember the struggles to get the medical world to accept a new initiative and the gradual but rewarding progress, one state at a time, as screening programs demonstrated effectiveness. I remember thinking that the establishment of universal screening in hospitals would be the hard part of the work, and I remember eventually learning that ensuring timely follow-up and arranging for high quality intervention turned out to be the more difficult task. I remember all of these puzzle pieces on the path to a mature newborn screening initiative.

Yes, I'm old enough to remember that we've come a long way, baby! And I'm young enough to remember where I put my car keys. That's not a bad place to hang out for a while...

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UPCOMING EVENTS

Event	Date	Location	Details
Beyond Hearing Loss Identification: Audiological Management for Children	August 1, 2014	Omaha, Nebraska	Web site

MESSAGE FROM AAP DEPARTMENT OF FEDERAL AFFAIRS: *NEWBORN SCREENING SAVES LIVES ACT*

On Tuesday, June 24, the *Newborn Screening Saves Lives Act* (H.R. 2181), legislation that renews federal support for programs that support testing of newborns for specific conditions at birth, was approved in the House by voice vote. The Senate passed their version of the bill on January 29, 2014. Because the newly passed House bill is slightly different from the version that the Senate passed in January, it is expected that the House bill will be considered by the Senate for final passage before it moves to the president's desk.



NIDCD INFOGRAPHICS ON HEARING LOSS

Put a Plug in the Noise



Properly inserted earplugs help prevent noise-induced hearing loss.

www.noisyplanet.nidcd.nih.gov



Two infographics—visual representations of information or data—are available from the National Institute on Deafness and Other Communication Disorders. The infographics on newborn hearing screening and proper use of earplugs to prevent noise induced hearing loss, respectively, can be found [here](#).

SAVE THE DATE: 14TH ANNUAL EHDI MEETING IN 2015

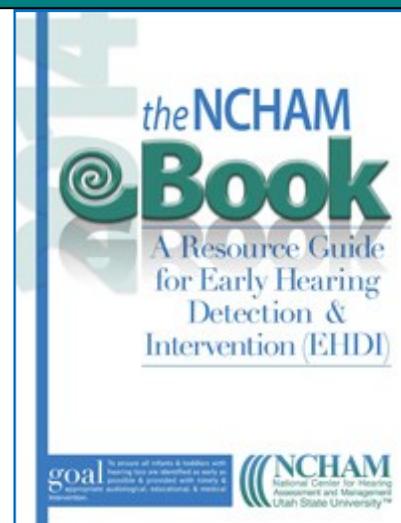


The 14th Annual EHDI Meeting will take place March 8-10, 2015 in Louisville, Kentucky. Abstract submission opens July 25, 2014. For more information, visit www.ehdimeeting.org.

EARLY HEARING DETECTION & INTERVENTION E-BOOK FROM NCHAM

The EHDI e-book, *A Resource Guide for Early Hearing Detection and Intervention*, from the National Center on Hearing Assessment and Management, is a 'go to' source for chapter champions and others involved in EHDI. For the next several months, the EHDI E-Mail Express will feature brief excerpts from the e-book in an effort to stimulate your interest in same. We know you will like it and will begin to consult the e-book going forward to support your EHDI efforts locally and in your state.

For those of you who are newer to EHDI, and for those of you who have been involved in EHDI for some time, you will find [Chapter One](#) to be quite informative. It includes a summary of the principles of effective screening programs, an historical overview of efforts to identify permanent hearing loss among infants and young children, information on the status of newborn hearing screening, and an overview of the degree to which each of the National Goals for Early Hearing Detection and Intervention (EHDI) programs, established by the federal government, are being achieved. Although almost all newborns are now being screened for hearing loss prior to hospital discharge, significant improvement is needed with respect to the availability of pediatric audiologists, implementation of effective tracking and data management systems, program evaluation and quality assurance, availability of appropriate early intervention programs, and linkages with health care providers who are well-informed about permanent hearing loss among infants and young children.



NEUROCOGNITIVE RISK IN CHILDREN WITH COCHLEAR IMPLANTS

Children who receive a cochlear implant (CI) for early severe to profound sensorineural hearing loss may achieve age-appropriate spoken language skills not possible before implantation. Despite these advances, reduced access to auditory experience may have downstream effects on fundamental neurocognitive processes for some children with CIs. The objective of this study was to determine the relative risk (RR) of clinically significant executive functioning deficits in children with CIs compared to children with normal hearing (NH). In this prospective, cross-sectional study, 73 children at a hospital-based clinic who received their CIs before 7 years of age and 78 children with NH, with average to above average mean nonverbal IQ scores, were recruited in 2 age groups: preschool age (age range, 3-5 years) and school age (age range, 7-17 years). No children presented with other developmental, cognitive, or neurologic diagnoses. Parent-reported checklist measures of executive functioning were completed during psychological testing sessions.

Estimates of the RR of clinically significant deficits in executive functioning (≥ 1 SDs above the mean) for children with CIs compared with children with NH were obtained based on 2 parent-reported child behavior checklists of everyday problems with executive functioning. In most domains of executive functioning, children with CIs were at 2 to 5 times greater risk of clinically significant deficits compared with children with NH. The RRs for preschoolers and school-aged children, respectively, were greatest in the areas of comprehension and conceptual learning, factual memory, sequential processing, working memory, and novel problem-solving. No difference between the CI and NH samples was found for visual-spatial organization on one checklist and for school-aged children on the other checklist.

A large proportion of children with CIs are at risk for clinically significant deficits across multiple domains of executive functioning, a rate averaging 2 to 5 times that of children with NH for most domains. Screening for risk of executive functioning deficits should be a routine part of the clinical evaluation of all children with deafness and CIs.

Source: Kronenberger WG, Beer J, et al. Neurocognitive Risk in Children With Cochlear Implants. JAMA Otolaryngology - Head & Neck Surgery 2014; May 22. doi: 10.1001/jamaoto.2014.757. [Epub ahead of print]

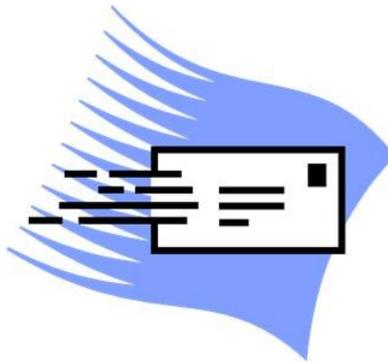
NEWLY DISCOVERED GENE MUTATION IS LINKED TO HEREDITARY DEAFNESS

Researchers led by geneticists have discovered a new gene mutation that causes hearing loss. Their study, which focused on a large Turkish family in which six individuals have been affected by hereditary deafness, identified a mutated form of the gene FAM65B as a cause of sensorineural hearing loss.

The research also demonstrates that FAM65B is a previously unrecognized component of the inner ear that is required for hearing. Their report, "FAM65B is a membrane-associated protein of hair cell stereocilia required for hearing," was published online in June 2014 by the Proceedings of the National Academy of Sciences. The study was led by Mustafa Tekin, MD, professor of human genetics at the Dr John T Macdonald Foundation Department of Human Genetics. Oscar Diaz-Horta, PhD, a postdoctoral fellow at the John P Hussman Institute for Human Genomics, was first author of the study.

Researchers in this study, who conducted a genetic analysis of the subject family, identified a mutated form of FAM65B — a protein previously unassociated with hearing — as the cause. Further characterization of the protein product of FAM65B in rodents and zebra fish has confirmed the findings of the family study.

Source: Newly Discovered Gene Mutation is Linked to Hereditary Deafness. University of Miami School of Medicine Web site. <http://med.miami.edu/news/newly-discovered-gene-mutation-is-linked-to-hereditary-deafness/>. Published 2014. Accessed June 28, 2014.



Distribution Information:

The AAP EHDI Program implementation staff send this e-mail update to the Academy's EHDI Chapter Champions, other interested AAP members, staff and state EHDI coordinators. For additional information on hearing screening and to access previous editions of the EHDI E-mail Express, click on the following link <http://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Early-Hearing-Detection-and-Intervention.aspx>. Previous e-mail updates are available upon request from Hollis Russinof, hrussinof@aap.org or (847) 434-4983. If you would like to unsubscribe to the update, please notify staff by responding to this e-mail.