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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.

A REFLECTION FROM CARLOS DURAN, MD, FAAP

All of our state Early Hearing Detection and Intervention (EHDI) programs should be proud of what we accomplish. It is easy to worry about not doing enough, and our high Loss to Follow-up rates. We clearly need to do better, but we should also remember how far we have come nationally and each individual state.

In 1994 the Joint Committee on Infant Hearing (JCIH) recommended universal newborn hearing screening (UNHS), and the AAP endorsed it in 1999. Around that same time—in 1998—Delaware embarked on a formal journey towards UNHS. At that time, 3 neonatal intensive-care units (NICUs) were testing all babies, and one hospital was testing all well newborns. There was no system for tracking these babies. A group of volunteers from all of the hospitals and public health got together and decided to develop a statewide EHDI program. Each hospital started looking at their program. Public health published a white paper and started to search for grant funding to create a state tracking and follow up system. That took a few years. In the meantime we developed a team to partner in this endeavor, and we gathered all the stakeholders around the table. Delaware hospitals gradually implemented UNHS; by 2002, all hospitals statewide had implemented it.

In 2005, Delaware passed its first law mandating all hospitals have a newborn hearing screening program, and requiring the Department of Public Health to track babies who failed the screening at birth. During this time most Delaware hospitals were using OAE as the screening test, and NICUs were using a tandem OAE and AABR screen. In 2008, Delaware passed a hearing aid loaner program extending the age to 18, and also passed HB 355 mandating insurance coverage for hearing aids for children.

Until 2012, the EHDI advisory board was voluntary and acting as a committee of the AAP Delaware chapter, with no true legal standing. We now have a new law (HB 384) and a new Governor's Advisory Board. The Board convened an expert panel which recommended that all well newborns be screened by AABR in order to diagnose babies with auditory neuropathy, as well as to decrease our number of false positives. This is where we stand.

As we continue to move forward, and worry about what's not being done, let's look back and see the long road we've traveled, and how far we have come.

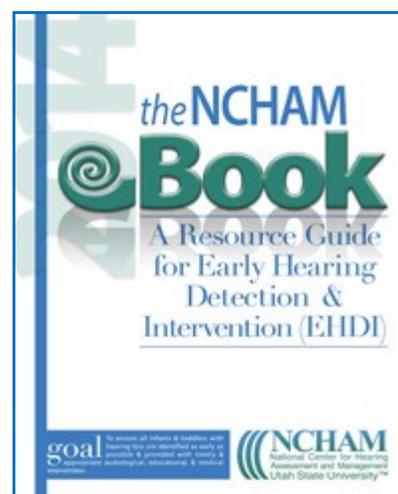
IN MEMORIAM: DR MARION DOWNS

Dr Marion P Downs, an innovator in the field of pediatric audiology and a tireless advocate for the early identification of hearing loss, passed away on November 13, 2014. During her career in audiology, Marion Downs created, developed, and evaluated techniques for testing hearing in children and for fitting them (some as young as a few weeks of age) with hearing aids. Dr Downs created the first national infant hearing screening program in 1963 in Denver, CO and relentlessly pursued making the identification and management of hearing loss in infants and children an important medical and educational consideration and public health issue. For more information about the life and work of Dr Downs or the *Marion Downs Center*, visit [this website](#).

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 (NCHAM), is a 'go to' source for chapter champions and others involved in EHDI.

This month we continue to offer information from the NCHAM e-book, a comprehensive online resource. In Chapter Six, authors N Wendell Todd, MD, MPH and Adrienne M Laury, MD, review the various causes of newborn and childhood hearing impairment. The chapter opens by describing in detail the differences between conductive, sensory, and neural hearing impairments and describes how identifying the type of impairment can help give clues to its etiology. While 3 newborns in 1,000 births have congenital hearing loss, the same number of children suffer delayed onset hearing loss, presenting between 11 and 60 months of age. The chapter describes the different causes of late-onset hearing loss as well as strategies to be able to quickly identify and implement interventions for young children that may have passed newborn hearing screening but have progressively impaired hearing during their first years of life.



The authors use the mnemonic VATICINATE in order to work with and address the needs of the family and child with hearing impairment. The mnemonic stands for: **V**erify that a hearing impairment exists; **A**mplify; **T**ypify; **I**nvestigate the etiology of the hearing loss; **C**onsult with ophthalmologist, radiologist, and geneticist; **I**nitiate discussion about psychological aspects of hearing loss; **N**o harm must be done to remaining hearing; **A**ssure communication in the most appropriate manner for the child and family; **T**est repeatedly as the child grows; and **E**ducate the family. As many pediatricians will encounter relatively few children with hearing loss in their practice, the step-by-step method detailed in this chapter is useful to ensure that each child is identified as early as possible and receives the appropriate interventions in a timely fashion.

UPCOMING EVENTS

Event	Date	Location	Details
NCHAM Move the Needle Webinar: Is Meeting the Needs of Physicians in Support of EHDI	December 11, 2014	Webinar	Web site
2015 National EHDI Meeting	March 8-10, 2014	Louisville, KY	Web site
American Academy of Audiology: AudiologyNOW! Conference	March 25-28, 2014	San Antonio, TX	Web site

PHYSICIAN SURVEY: ADDRESSING EHDI MISCONCEPTIONS

The National Center for Hearing Assessment and Management (NCHAM)—along with support from state EHDI coordinators and the American Academy of Pediatrics (AAP)—conducted a self-report survey with pediatricians and other clinicians who provide care for infants and young children. The purpose of this survey, conducted in 2012, was to:

- Understand the degree to which medical homes are engaged in EHDI activities
- Update our understanding of physician attitudes and knowledge regarding EHDI since the 2005 survey conducted on this topic
- Drive strategies to support physicians in their role within EHDI systems

In upcoming editions of the EHDI E-Mail Express, we will review some of the questions presented in the survey and the results that pediatricians provided. We hope to identify and examine what gaps in understanding and practice, if any, still persist.

Question: What is your best estimate of the earliest age at which a newborn not passing hearing screening should receive additional testing?

Year	<1 mos	1-3 mos	4-6 mos	7-9 mos	10-12 mos	>12 mos
2005	75.7	11.7	7.1	4.2	0.1	1.2
2012	41.0	51.9	6.1	0.3	0.7	0.8

The data reflect that in 2012, only 41% of physicians felt as though newborns who fail a newborn hearing screening should receive additional testing within the first month of life, which is a decrease from 75.7% of physicians reporting in 2005. In addition, nearly 7.1% of physicians who responded to this question in 2012 felt as though follow-up screening should occur at some point after 3 months of age; this is a contradiction to the 1-3-6 message.

Chapter Champions can help improving knowledge among pediatric primary care clinicians about the goal to conduct a follow-up hearing screening within one month of a failed newborn hearing screening. Your efforts can help improve efforts to reduce loss to follow-up/documentation and outcomes for infants and children who are identified with hearing loss.

COCHLEAR IMPLANTATION IN UNILATERAL SUDDEN DEAFNESS IMPROVES TINNITUS AND SPEECH COMPREHENSION

Unilateral sudden deafness can result in decreased speech comprehension and sound localization, even with normal hearing in the unaffected ear. [New research](#) sought to analyze the benefits of cochlear implantation in patients with unilateral severe to profound hearing loss, in comparison to other intervention alternatives such as contralateral routing of signal (CROS) and bone-anchored hearing aids (BAHAs). This meta-analysis reviewed nine different articles in order to determine the effects of cochlear implantation with regard to reduction of tinnitus, and enhancement of speech comprehension and sound localization.

The authors found that tinnitus severity was decreased by 5.29 points on average using the Visual Analog Scale (10 point scale, $p < 0.01$) following cochlear implantation in comparison to other interventions. The study also found that while cochlear implantation reduced signal-to-noise (SNR) ratios slightly using a variety of tests, this reduction was not statistically significant. The study also sought to assess the subjective perception of improvements among patients who have received cochlear implantation. The study showed that patients self-reported improvements in tinnitus, sound localization, and speech comprehension following the intervention. This analysis shows the potential effectiveness of cochlear implantation when considering tinnitus discomfort and sentence understanding in noise for individuals with unilateral sudden sensorineural hearing loss.

Source: Blasco MA, Redleaf MI. Cochlear Implantation in Unilateral Sudden Deafness Improves Tinnitus and Speech Comprehension: Meta-Analysis and Systematic Review. *Otology & Neurotology Inc.* 2014; 35: 1426-1432.

GENETIC INVESTIGATIONS IN CHILDHOOD DEAFNESS

As nearly 60% of congenital deafness has a genetic etiology, recent technological [advancements in genetic testing](#) can potentially lead to increased benefits for children with hearing loss and their families. Non-syndromic sensorineural hearing loss (SNHL) can be caused by a mutation of a number of genes and this can make identification of hearing loss etiology particularly challenging. The authors highlight some of the main genetic anomalies that lead to non-syndromic SNHL, such as GJB3, SLC26A4, OTOF, and TMPRSS3, among others, while also describing the difficulty for diagnosis for each variation as well as the presentation in other family members.

The authors describe *next generation* sequencing (NGS) or massively parallel sequencing and its ability to provide more accurate, faster diagnosis of genetic conditions such as SNHL. These methods allow for large numbers of DNA segments to be analyzed simultaneously in the same reaction, as opposed to previous serial sequencing methods. The authors report that these advances in genetic screening will not only provide clinicians with information on 'target' genes that are known to cause SNHL, but will also help to identify new genetic coding exons that are associated with SNHL.

Source: Parker, M. & Bitner-Glindzicz, M. Genetic Investigations in Childhood Deafness. *Archives of Disease in Childhood*. Published online: October 16, 2014.

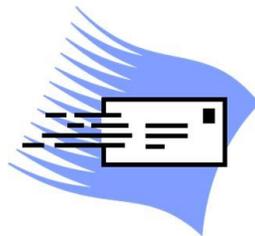
COLLABORATIVE DISCUSSIONS INVOLVING PROFESSIONALS AND PARENTS ON TOPICS RELATED TO DEAF EDUCATION

The Council for Exceptional Children's Division of Communicative Disorders and Deafness (DCDD) has begun holding web-based discussions on topics related to children and students who are Deaf or hard of hearing. Each month a new topic is discussed on Deafed.net; as part of the discussions parents and professionals are given the opportunity to share their frequently encountered challenges and solutions to the topic at hand.

The October 2014 discussion topic was "collaborating with other professionals." Discussion included how to begin collaboration between professionals involved with EHDI, as well as resolving gridlock, and how to promote an environment of collaboration in a field that requires coordination between pediatric primary care providers, specialists, and other clinicians. Notes from the month-long discussion can be found on the [DeafEd website](#). Current and future discussion topics include:

- November 2014: Identifying appropriate augmentative and alternative communication options for students
- February 2015: Providing comprehensive services for students who are deaf/hard of hearing
- March 2015: Providing effective support for parents
- April 2015: Ensuring the safety and success of children with disabilities

These discussions can be a great resource and will likely provide strategies for asking and answering important questions—for both professionals and parents. The goal of bringing all stakeholders to the table in these discussions is to help enhance the care and education of children who are Deaf or hard of hearing. To join the conversation or to register to participate in a future discussion, visit the [DeafEd Topical Conversations web page](#).



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 Stephen Crabbe, scrabbe@aap.org or (847) 434-4738. If you would like to unsubscribe to the update, please notify staff by responding to this e-mail.