

## INFANT HEARING PROGRAM

Maryland Department of Health and Mental Hygiene

Family Health Administration

Office for Genetics and Children with  
Special Health Care Needs

201 West Preston Street, Room 423 A, Baltimore, MD 21201

Martin O'Malley, Governor

Anthony G. Brown, Lt. Governor

John M. Colmers, Secretary



# Infant Hearing News June 2009

## MD EHDI

As part of the legislation that originally established Universal Newborn Hearing Screening in Maryland, an Advisory Council was formed to advise, provide consultation, and make recommendations for the operation of the program. This Council consists of two parents of children with hearing loss, one physician, three educators, one mental health professional, one audiologist and one representative from each of the following organizations: the Maryland Association of the Deaf, the Alexander Graham Bell Association, and the Maryland Department of Health and Mental Hygiene. This spring, the Council underwent some changes in leadership and membership. We would like to gratefully acknowledge the service of our most recent past Chairperson, Mr. Ben Dubin, and Vice-Chairperson, Ms. Louise Colodzin. Their many responsibilities have been turned over to our new Chairperson, Ms. Maryann Swann, and Vice-Chairperson Dr. Steve Seipp. The Council also welcomed a new member, Dr. Mary Mussman.

The Hearing Advisory Council typically meets every other month at the Hearing and Speech Agency in Baltimore. Guests are welcomed at all meetings. If you are interested in attending a meeting, please contact our administrative assistant, Hope Wharton, at 410-767-5803 or [HWharton@dhmh.state.md.us](mailto:HWharton@dhmh.state.md.us) for specific dates and times of the upcoming meetings.

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## In the News

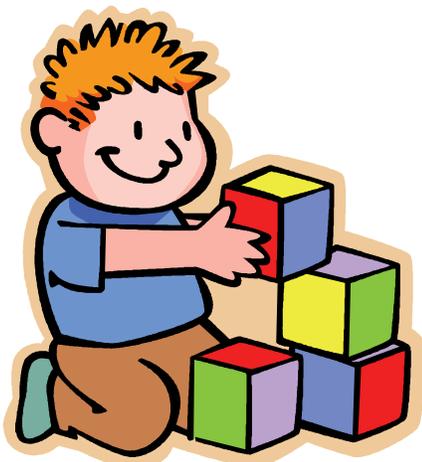
### Scientists find key protein helps people hear

Scientists conducting studies on mice have discovered that the presence of a certain protein in the inner ear is required to help people differentiate between sounds and understand speech. Until now, this function had been attributed to structures called ion channels, which are found in the sensory receptor cells of the cochlea. Ion channels work like a microphone to transform sound into electrical messages to the brain, but they are not responsible for the ability to pick out the correct sounds from a mixture of noises. This study found that mice with intact ion channels did not hear properly in the absence of the protein stereocilin. Stereocilin is required for the production of mechanical and electrical waveform distortions within the cochlea. These distortions allow one tone to suppress another and can also combine to create tones that were not present in the original signal. It is this suppression, or masking, of certain sounds that contributes to the ability to understand speech. The sounds created by the waveform distortions also make up the same otoacoustic emissions that we use in infant hearing screening. The full study can be found in [Nature](#). 2008 Nov 13;456(7219):255-8. Epub 2008 Oct 8.



### Building Blocks of Intervention Webinar Series

The Deaf and Hard of Hearing Program of the Children's Hospital Boston, through a grant from the Verizon Foundation, has created a series of six on-line seminars. The series, Building Blocks of Intervention: Hearing Loss in Children 0-3 Years, highlights how to promote communication and language in children with hearing loss from infancy through early childhood. The modules are presented by an interdisciplinary team of clinicians including pediatric psychologists, audiologists and speech-language pathologists. The series is aimed at both professionals and parents, and includes the topics:



- Assuring Early Access to Language & Understanding Hearing Loss
- Optimizing Access to Sound Through Amplification
- Launching Communication through Sign Language
- Accessing Audition Through Cochlear Implants
- Promoting Communication in the Home
- Understanding the Impact of Hearing Loss on the Family System

To go along with the seminars, the presenters provide downloadable materials, handouts and information sheets on the web site as well. To access the series, go to the Children's Hospital Boston Web site at

<http://www.childrenshospital.org/clinicalservices/Site2143/mainpageS2143P17.html>.

## Summary of 2006 CDC EHDI Data

The Early Hearing Detection and Intervention (EHDI) Program at the Centers for Disease Control and Prevention (CDC) conducts an annual survey to gather data about newborn hearing screening and follow-up activities in states and territories. National data for year 2006 indicated continued progress towards the “1-3-6” EHDI benchmarks. The “1-3-6” benchmarks refer to ensuring newborns are screened for hearing loss by 1 month of age, have a diagnostic audiologic evaluation by 3 months of age (if recommended), and are enrolled in appropriate early intervention services by 6 months of age (for those with confirmed hearing loss). The data show that nationally, over 92% of infants were documented as being screened for hearing loss. A total of 88% of those screenings were completed before 1 month of age, which is nearly a 9% increase from 2005.



Unfortunately, progress related to diagnosis was limited and reflects the need for continued improvements. In 2006, over 46% of infants who were referred for diagnostic testing did not have a documented diagnostic finding (i.e., normal hearing or hearing loss); they were lost to follow-up/lost to documentation (LFU/LTD). While this represents a nearly 13% decrease in the number LFU/LTD compared to 2005, it still means that nearly half of infants were not documented to have received recommended diagnostic testing. Regarding the 3 benchmark, only 55% of infants with a documented hearing loss were diagnosed before three months of age. Enrollment in early intervention showed better progress, with approximately 75% of those infants eligible for Part C (e.g., state-funded) services being enrolled in these intervention services. Of these infants, approximately 61% were enrolled in Part C services before six months of age. Another 8% of those identified with a hearing loss were reported to be receiving intervention services from only private, or non-Part C services.



While results for year 2006 indicate progress has been made in screening and, to a lesser extent, overall enrollment in intervention, additional efforts are needed to ensure infants are documented to receive diagnostic tests and enrolled in intervention services before six months of age. If infants do not receive the recommended diagnostic and early intervention services, especially before the recommended 3 and 6 month benchmarks, the benefits of newborn hearing screening may be severely reduced. Additional information about the 2006 data is available online at [www.cdc.gov/ncbddd/ehdi/data.htm](http://www.cdc.gov/ncbddd/ehdi/data.htm).

### Did you know?

Hearing loss carries a socio-economic cost of over 1 million dollars (in year 2000 \$) over the life-time of each child with pre-lingual onset of hearing loss.

## NICU Infants and Risk Factors for Late-onset Hearing Loss

There were enough questions following the release of the Joint Committee on Infant Hearing (JCIH) Year 2007 position statement that related to NICU infants and infants with risk factors for late onset hearing loss that the following JCIH Clarification Year 2007 statement was released:

### **Clarification for Year 2007 JCIH Position Statement**

#### 2007 JCIH Position Statement Update

Separate hearing screening protocols are recommended for NICU and well-infant nurseries. High risk NICU infants (admitted for more than 5 days) are to have automated auditory brainstem response (ABR) included as part of their hearing screening so that neural hearing loss will not be missed.

#### **Rationale for different protocols:**

The JCIH 2007 recommendation is to identify infants with congenital permanent sensory, conductive and neural (auditory neuropathy/auditory dyssynchrony) hearing loss. ABR technology is needed to identify neural hearing loss. Consequently, the JCIH recommends ABR screening to identify neural hearing loss in the infants most at risk of a neural loss. NICU infants represent 10% of the newborn population or approximately 400,000 infants per year. Infants cared for in the NICU are at increased risk of neural hearing loss. Data from the National Perinatal Research Center (NPIC) (Quality Analytic Services)(QAS) was utilized to make the recommendation of ABR for the subgroup of NICU infants that require NICU care for >5 days. NPIC data indicated that approximately 25% of NICU infants are considered “low” risk (includes infants with diagnoses such as transient respiratory distress, observation for temperature instability, and negative sepsis workup) and are discharged by 5 days of age. High risk NICU or Level 2 infants hospitalized for > 5 days for medical reasons (not social ) were then identified as the target population for ABR to rule out neural hearing loss. Since specific risk factors are often difficult for screeners to identify in the medical record, establishing a time criterion (>5 days) was felt to be easier to implement. Implementation of this recommendation for all NICU and well baby infants was not felt to be indicated based on current evidence. Therefore, a NICU baby who meets this >5 day criterion cannot be screened and passed by OAE alone.

#### **Risk Factor Clarifications:**

**1. Recommendations for audiology follow-up of infants with risk factors.** The prior recommendation for follow-up audiology assessments every 6 months for all screen fails was felt to place a great burden on audiologists and could not be accomplished in most parts of the country. In addition, there are infants with "unknown risk factors" who develop late onset hearing loss. Therefore, the responsibility for surveillance of all infants was shifted to the primary care provider, who will refer to audiologists, as needed, any patient for whom there are concerns or findings consistent with hearing loss. The document lists accepted risk factors for hearing loss, identifies risk factors which are

known to be associated with late onset or progressive hearing loss, and makes recommendations for standard or more frequent follow-up by the audiologist. Therefore, for infants with a risk factor which may be considered low risk, at least one audiology assessment by 24-30 months is the recommendation. This management would change if there is a referral from the primary care provider because of a new concern regarding hearing. In contrast, for an infant with risk factors known to be associated with late onset or progressive hearing loss, such as, CMV or family history, early and more frequent assessment is appropriate. Early and more frequent can be interpreted as every 6 months, or more, depending on the clinical findings and concerns. See following excerpt from document.

The timing and number of hearing re-evaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. Infants who pass the neonatal screening but have a risk factor should have at least 1 diagnostic audiology assessment by 24 to 30 months of age. Early and more frequent assessment may be indicated for children with cytomegalovirus (CMV) infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, or culture-positive postnatal infections associated with sensorineural hearing loss; for children who have received ECMO or chemotherapy; and when there is caregiver concern or a family history of hearing loss.

Appendix 1 in JCIH 2007 lists the risk factors and identifies the risk factors associated with late onset or progressive loss with an asterisk.

## **2. Recommendations regarding ototoxic medications.**

The following are listed as risk factors in Appendix 1 of the JCIH 2007 Statement:

Neonatal intensive care of >5 days, or any of the following regardless of length of stay: ECMO, assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/lasix), and hyperbilirubinemia requiring exchange transfusion.

To be consistent with the intent of simplifying the referral process to NICU >5 days and for clarification the recommendation has been reworded.

All infants with or without risk factors requiring neonatal intensive care for greater than 5 days, including any of the following: ECMO, assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/lasix). In addition, regardless of length of stay: hyperbilirubinemia requiring exchange transfusion.

JCIH

February 13, 2008

Further information can be found at [www.jcih.org](http://www.jcih.org).

### **Did you know?**

Children with unilateral hearing loss have a school grade failure rate that is 10 times higher than their peers with normal-hearing .

## News from the EHDI National Conference

### Early Hearing Detection and Intervention Conference March 9-10 Addison, TX

This year's National EHDI Conference drew attendees from places as far away as Alaska and Samoa. There were several attendees from Maryland, including Cheri Dowling; Mary Ann Richmond; and Maryann Swann, M.S.; who together presented the session "Its Not about Turf or Territory... Its about Listening to a Family." Cheri also presented the session "Choices, Options, Opportunities...How Does a Family Decide?"

There was a great deal of discussion generated by the opening seminar, "Genetic Screening for Usher Syndrome," which was presented by William Kimberling, PhD of Boys Town National Research Hospital. Dr. Kimberling discussed an inexpensive screening test for Usher Syndrome that could be integrated into current U.S. newborn metabolic screening programs. The last session of the conference, "Auditory Neuropathy Spectrum Disorder," was also an interesting session and included the presentation of the guidelines for screening, diagnosis, and early intervention for infants with this diagnosis that were generated at the June 2008 Newborn Hearing Screening Conference in Lake Como, Italy.

Between sessions, there were many things to see in the vendor display room. At one point there was a group from Oz systems, the vendor who designed and supports our online database, who were dressed as the characters from the Wizard of Oz. No one seems to have taken any pictures of this..... or at least none that they are willing to admit to!

Each EHDI conference begins and ends with a state team meeting. This is done to help participants maximize what they learn at the conference and use the knowledge gained to improve their state's EHDI program when they return home. The participants also develop a written action plan for the state EHDI team. At this year's meeting, the Maryland group drafted an action plan that included developing a Deaf Mentor program, organizing a social event for families, and identifying a new Chapter Champion. The Chapter Champion is a member of the American Academy of Pediatrics who agrees to "champion" the cause of Early Hearing Detection and Intervention in the state pediatric community.

The next EDHI National Conference is being planned for February 2010 in Chicago, IL. We hope to see you there!



## Federal Legislation

On Monday, March 30, 2009, the U.S. House of Representatives passed H.R. 1246 the Early Hearing Detection and Intervention (EHDI) Act of 2009. This bill would reauthorize the federal portion of this program for five years. The bill must now be considered by the Senate before it can be signed into law.

Also in March, HR 1646, legislation that would provide a tax credit of up to \$1000 for the purchase of hearing aids, was introduced in the House. A Senate version of the bill is expected to be introduced soon.

## Newborn Hearing Screening Training Curriculum

The National Center for Hearing Assessment and Management has released a Newborn Hearing Screening Training Curriculum DVD. This curriculum was developed to standardize the way that new hearing screeners are trained and to ensure that each screener receives the information that will enable them to provide high-quality, efficient, and effective newborn hearing screening.



If your center would like a free copy of this DVD, please contact Hope Wharton at 410-767-5803 or at [HWharton@dnhmh.state.md.us](mailto:HWharton@dnhmh.state.md.us).

### Did you know?

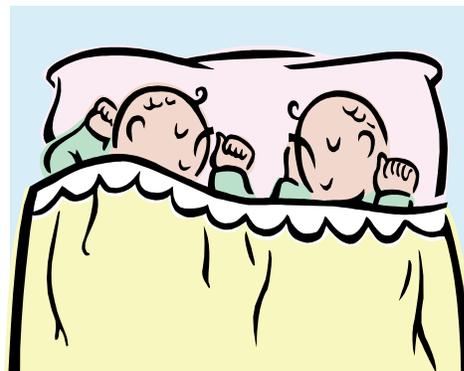
50% of children with permanent hearing loss have no apparent risk indicators

## For our eSP system users

As we continue to add more users to the eSP system, it becomes even more important that the files are as complete and accurate as possible. The feedback we have gathered from various users indicates that there are a few things we may need to review.

While the infants are still inpatient, please be sure to:

- Enter the data with correct capitalization/punctuation**-many letters are generated directly from the database fields, so grammar counts!
- Mark the birth order for multiples**-this helps determine that the file is not a duplicate and also helps ensure that the correct permanent name is assigned to the correct file.
- Mark the appropriate risk factors**-this is how we identify those infants who need to be monitored for late-onset hearing loss.
- Enter the pediatrician information**-without the pediatrician name we are limited in our ability to ensure the infants receive appropriate care.
- Transfer and discharge infants in a timely manner**-remember that many others will need access to the infant's file quickly if we are to be successful in providing Early Hearing Detection and Intervention.

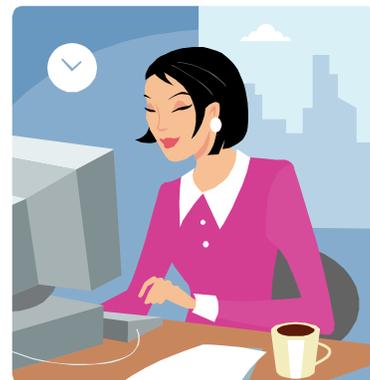


As the infants move through the outpatient centers, please be sure to:

- Edit and update demographic information**-especially note and update changes in the patient's permanent name, and confirm that the address and phone numbers for the parents are complete and correct.
- Edit and update provider contact information**-having the correct pediatrician on file is critical to the follow-up on these infants. It is also very important that you add to the file any provider you refer to—other providers will not be able to see the infant's file unless you add them to it.
- Edit the appointment date fields**-if you make or know of any of the infant's upcoming appointments, be sure to enter that information. It is always best if we

do not have to unnecessarily disturb you, the pediatrician, or the parents for this information.

- Remember to use the "View/Add Case Notes" option**-please make note of anything you believe the state or other care providers should know in following these infants.
- Add your data in a timely manner**-again, many others will need access to the infant's file quickly if we are to be successful in providing Early Hearing Detection and Intervention.



## Resources

We have a number of resources available on our website:

[www.fhamd.org/infant hearing](http://www.fhamd.org/infant_hearing)

Some of these resources include:

- Infant Hearing Newsletters – past and current editions
- Informational pamphlets and brochures – some available in Spanish
- Patient Education forms
- Guidelines and Checklists
- Links to other helpful sites

As always, the staff at Maryland DHMH is happy to assist you in any way we can. The Infant Hearing Program toll free phone number is 800-633-1316, our toll free TTY number is 866-635-4410, and the office fax number is 410-333-5047. Individually, we can be reached by phone or email:

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**This newsletter is intended to serve as a communication vehicle for all UNHS stakeholders. If you have any patient interest stories, photos, announcements, helpful hints, questions, or any information you would like to share with your Maryland colleagues, please email them to Erin Filippone at [EFilippone@dhhm.state.md.us](mailto:EFilippone@dhhm.state.md.us).**

### ***WORKING TOGETHER... EARLY HEARING DETECTION AND INTERVENTION THE KEY TO COMMUNICATION SUCCESS***

The services and facilities of the Maryland Department of Health and Mental Hygiene (DHMH) are operated on a non-discriminatory basis. This policy prohibits discrimination on the basis of race, color, sex, or national origin and applies to the provisions of employment and granting of advantages, privileges, and accommodations.

The Department, in compliance with the Americans With Disabilities Act, ensures that qualified individuals with disabilities are given an opportunity to participate in and benefit from DHMH services, programs, benefits, and employment opportunities.