

INFANT HEARING PROGRAM

Maryland Department of Health and Mental Hygiene

Family Health Administration

Office of 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*;

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Infant Hearing News September 2007

It has been a busy spring and summer for the Infant Hearing Program. We attended the Early Hearing Detection and Intervention conference in Salt Lake City in March. Then we got busy planning our own Infant Hearing meeting for June. It was a pleasure to have met so many of you there! Our thanks again to all of our speakers for providing such an interesting program. We have also appreciated the opportunity to meet many of you during our recent hospital site visits; hopefully you have found them to be as helpful and informative as we have. If we haven't gotten to you yet, don't worry, we are coming!

Infant Hearing in the News

This quarter our newsletter focuses on new developments related to Universal Newborn Hearing Screening. There is new technology that allows ABR results to be obtained in the presence of "noisy" environments and high patient activity, new tests that can detect the most common genetic causes of hearing loss, and there has even been a study suggesting that newborn OAE test results may be of great value in identifying infants at risk for sudden infant death syndrome.

Please note that while some of the following articles relate to commercial products and services, these articles are presented strictly on an informational basis. The Maryland DHMH Infant Hearing Program does not specifically endorse any products or services.

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OAE and Sudden Infant Death Syndrome

One of the most exciting recent developments may be that some of the work done by Universal Newborn Hearing Screening programs could lead to a reduction in sudden infant death syndrome (SIDS). SIDS is the most common cause of death in infancy. Although it is not definitively known what causes SIDS, a respiratory control problem is believed to be a primary factor. The link between SIDS and otoacoustic emissions (OAEs) is the inner ear vestibular system.



A study by Rubens DD, et al., Newborn otoacoustic emission hearing screening tests, *Early Hum Dev*(2007), doi:10.1016/j.earlhumdev.2007.06.01, was based on the knowledge that stimulation of the vestibular system affects the firing of neurons responsible for central respiratory patterns. The study was designed to determine if damage to the inner ear, resulting in a disruption of vestibular

function, played a role in an infant's predisposition to SIDS.

Since any damage to the vestibular system would likely also affect the cochlear system, these researchers studied the newborn hearing screening results of infants who subsequently died of SIDs and compared them to matched controls.

The results of the study indicated significant decreases in the signal to noise ratio (SNR) of the right ear transient evoked OAE (TEOAE) responses at 2000, 3000, and 4000 Hz in all of the 31 infants who later died of SIDS. (Typically, the right ear OAE responses of infants are greater than left ear responses.) The researchers hypothesize that this finding may be due to some damage to the right inner ear during delivery. They believe this damage may be caused by the significant pressure of transfused placental blood on the right ear. The left ear does not show this damage as it is protected by the angle of the left innominate vein. This pressure is thought to damage the hair cells of the inner ear, resulting in a disruption of the vestibular/respiratory control interface and the decreased signal to noise ratios of the OAE response. Supporting this theory is the fact of increased incidence of SIDS with long labors and prima gravida pregnancies. It is also supported by the statistic that a second delivered twin has more than double the risk of SIDS than a first twin. In all these circumstances, blood accumulating in the placenta has a greater pressure than in standard circumstances, and the affected infants show a concurrent increase in the rate of SIDS.

The researchers are recommending that inclusion of the frequencies above 4000Hz become a standard part of the newborn screen as those frequencies are more sensitive to pressure damage. They also recommend a large prospective trial and comprehensive analysis of the hearing screens of infants with SIDS to determine if their findings can be definitively determined to be a valid tool for identifying infants at risk for SIDS.

Identifying the Cause of Hearing Loss

Pediatrix Screening recently introduced SoundGene™, a screening panel that detects the most common genetic and environmental factors responsible for hearing loss. The premise of this development was the idea that learning the cause of a hearing loss can help the healthcare provider better direct the medical management and care of a newborn or child.

The SoundGene test requires only a few drops of blood applied to filter paper for analysis, much like the current Maryland newborn metabolic screening. The sample is analyzed and the results of the screen are reported back to the healthcare provider approximately 72 hours after the sample arrives at the laboratory. Physicians are contacted by phone with abnormal results, and genetic counseling staff are available to physicians 24 hours per day, seven days per week.

“SoundGene provides a method for physicians to identify some of the most common causes of hearing loss in newborns and children. Knowing the causes will help health care providers facilitate timely care and intervention for a child that is affected by hearing loss,” says Gail Lim, Vice President, Program Development, Pediatrix Screening. “Using dried bloodspots on a filter paper, we are able to screen for some of the most common genetic and environmental causes of hearing loss, including mutations in the Connexin 26 gene, which is one of the most common causes of hearing loss.” Pediatrix Medical Group is also conducting a research study to demonstrate the utility of universal SoundGene testing in identification of some late-onset hearing loss causes.

Congenital Cytomegalovirus (CMV) infection is included in the SoundGene panel. The congenital CMV screen is based on a qualitative test for the detection of the presence or absence of CMV viral DNA in the blood. The sample collection time limit is two weeks after birth in order to qualify the results as congenital CMV infection. Samples can be collected at a later time, but it cannot be determined whether the infection was congenital or acquired.

For parents, the decisions that need to be made regarding rehabilitation strategies for their hearing-impaired child can be overwhelming. Knowing the cause of the hearing loss can give insight into the expected progression and impact of the loss. This can make some strategy options, especially surgical ones like cochlear implants, easier to make.

More information about SoundGene, can be found on the Pediatrix Screening website at www.pediatrixscreening.com.

SoundGene Screening Panel

- Connexin 26 (Cx26 GJB2 mutations)
- Connexin 30 (Cx30) GJB6 large deletion
- Mitochondrial mutations
- Pendred SLC26A4 mutations
- (CMV)

Did you know?

The Infant Hearing Program's new web address is: <http://www.fhamd.org/infanthearing>

New technology for ABRs



Wouldn't it be great if your patient didn't have to be asleep or sedated for an ABR? Vivosonic Inc., a Canadian corporation, has developed the Integrity™ system, an auditory evoked potential (AEP), ASSR (Auditory Steady State Response) and otoacoustic emissions (OAE) analyzer that is able to record results on awake, active patients.

The Integrity™ uses special software, a new type of amplifier and a wireless Bluetooth computer connection that enables AEP testing even in the presence of patient activity, thereby removing the need for the patient to be asleep or sedated. During the test, parents are able to feed or walk with their baby in a stroller up to 30 ft away. Young children and toddlers can quietly play games or watch TV at low levels, and clear ABR results can still be obtained. In addition to reducing patient risk, eliminating the need for sedation significantly lessens the cost of administering an ABR and significantly increases the population eligible for the test. Another improvement that is particularly beneficial for Universal Newborn Hearing Screening programs is that this new technology allows data to be obtained in environments that are electromagnetically “noisy” like the NICU or the OR.

Denise DeMonte, Au.D., Audiologist at the Hearing and Speech Agency (HASA) in Baltimore currently uses the Integrity™ system and says, “You really can get results on a moving child. I have been able to get several repeatable waveforms down to 20dBnHL while the child remained alert and active. However, it should be noted, the more active the child, the longer it takes to get results” HASA purchased the system last year to better serve their infant and toddler population. As with any new technology, there were some start-up glitches and issues to be resolved, but with the current software updates and more complete training, HASA is having increasing success using the system. It generally takes about one hour to obtain two click and four toneburst thresholds. The system is extremely useful ruling out hearing loss or confirming thresholds on hard-to-test patients, but is not as well equipped for neurological ABRs. For HASA, where sedation is not an option and there is a large pediatric population, it is ideal. Unfortunately, even this new technology is still no match for a real screamer.



Did you know?

The next Early Hearing Detection and Intervention (EHDI) meeting will be held February 25-26 at the Astor Crowne Plaza Hotel in New Orleans, Louisiana .

Risk Factors for Hearing Loss

Don't forget to report any risk factors for the babies you test. The current risk factors reportable on the Metabolic Disorder slip are:

Apgar scores of 0-3 at five minutes

Head/Neck defects

+ TORCH

Ototoxic Drugs

Bacterial/Viral meningitis

Family history of hearing loss

Did you know?

The Joint Committee on Infant Hearing is expected to release the "2005 Infant Hearing Screening Guidelines" in September. The new guidelines will be featured in our next issue.

Updates at DHMH

Do you hate filling out paperwork? We have good news for you! The Infant Hearing Program is expecting to launch a live online data management system in the Spring of 2008. With this new system, hospital screening personnel will be able to input demographic and screening result data directly into the system. This new system is expected to dramatically improve timeliness and accurate communication of test data information. It will also cut down on the paperwork required for programs to report results to DHMH. Watch for more information early in the new year.

Maryland Universal Newborn Hearing Screening Protocols

Maryland's Universal Newborn Hearing Screening Advisory Council have developed protocols for newborn hearing screening and diagnostic evaluation which are available on our website:

http://www.fha.state.md.us/genetics/pdf/UNHS_protocol_FINAL04SEPT06.pdf

The protocols support the recommendations of the Joint Committee on Infant Hearing (2000) and the American Academy of Pediatrics endorsement of the "1-3-6" process which outlines the timeline of identification and intervention:

All newborns will be screened for hearing loss before **1 month of age** and preferably before hospital discharge.

Diagnostic audiologic assessment for infants who screen positive will take place before **3 months of age**.

All infants identified by diagnostic assessment will receive intervention services - audiologic, medical and early intervention- before **6 months of age**.

Just in time...1-3-6

Resources

We have a number of resources available to assist you on our website:

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

Here you will find:

Infant Hearing Newsletters – past and current editions

Informational pamphlets and brochures – some available in Spanish

Patient Education forms

Level 2 and 3 screening forms

Guidelines and Checklists

Links to other helpful sites

Please also note our toll free phone number 800-633-1316 and our toll free TTY number 866-635-4410.

As always, the staff at Maryland DHMH would be happy to assist you in any way we can. We can be reached by phone:

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This newsletter is 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

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

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