Volume 6, Issue 2 July 2011

State of Tennessee Department of Health

Newborn Screening Newsletter

NBS Metabolic Follow Up
Ph 615-262-6304
Fax 615-262-6458

NBS Hearing Follow Up
Ph 615-262-6160
Fax 615-262-6159

http://health.state.tn.us/NBS/index.htm

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Special points of interest:

UPCOMING STATE HOLIDAYS

- July 4, 2011 Independence Day
- September 5, 2011 Labor Day

Cystic Fibrosis

Cystic fibrosis is an inherited condition of the mucus glands that affects many body systems. The most common signs and symptoms include progressive damage to the respiratory system and chronic digestive system problems. Mucus lubricates and protects the linings of the airways, digestive system, reproductive system, and other organs and tissues. In people with cystic fibrosis, the body produces mucus that is abnormally thick and sticky.

About one baby in 2,500-3,500 is born with cystic fibrosis in the United States. The condition can be found in all ethnic groups, but is most common in Caucasian (white) population.

The thick mucus can obstruct the airways, leading to problems with breathing and infections in the lungs. These infections cause chronic coughing, wheezing, and inflammation. Over time, mucus buildup and infections result in permanent lung damage, including the formation of scar tissue (fibrosis) and cysts in the lungs.

In people with cystic fibrosis, mucus blocks the ducts of the pancreas. The pancreas is an organ that produces insulin (a hormone that helps control blood sugar levels). It also makes enzymes that help digest food. This blockage prevents these enzymes from reaching the intestines to aid digestion. Problems with digestion can lead to diarrhea, malnutrition, poor growth, and weight loss.

Cystic fibrosis is a genetic condition caused by a change in the cystic fibrosis transmembrane-conductance regulator (CFTR) gene. Cystic fibrosis is inherited in an autosomal recessive pattern, which means two copies of the gene must be changed for a person to be affected with cystic fibrosis. Most often, the parents of a child with an autosomal recessive condition are not affected because they are "carriers", with one copy of the changed gene and one copy of the normal gene.

When both parents are carriers, there is a one-in-four (or 25 percent) chance that both will pass the changed gene on to a child, causing the child to be born with the condition. There also is a one-in-four (or 25%) chance that they will each pass on a normal gene, and the child will be free of the condition. There is a two-in-four (or 50%) chance that a child will inherit a changed gene from one parent and a normal gene from the other, making it a carrier like its parents. These chances are the same in each pregnancy with the same parents.

Babies are tested (newborn screening) for cystic fibrosis before they leave the hospital. The baby's heel is pricked and a few drops of blood are taken. The blood is sent to the state laboratory to look at the immunoreactive trypsinogen (IRT).

The types of treatment your child receives depends on what kinds of health problems the cystic fibrosis is causing and how your child's body responds to different types of treatment. Most people combine medicines, home treatment methods (breathing exercises), high-energy foods, enzyme supplements, and vitamin and mineral supplements. Regular exercise is important to keep the body fit and healthy. Consult with your pulmonologist and /or regional genetic center to determine what follow up and tests are needed.

For more information go to:
http://health.state.tn.us/nbs/PDFs/CF.pdf

Volume 6, Issue 2

Michael D. Warren, MD MPH FAAP

Dr. Michael Warren is the Director of Maternal & Child Health for the Tennessee Department of Health.

Dr. Warren received his undergraduate degree from Wake Forest University and his medical degree from the Brody School of Medicine at East Carolina University. He completed his pediatrics residency, Chief Residency, and fellowship in Academic General Pediatrics at Vanderbilt, where he also obtained a Master's in Public Health. He is certified by the American Board of Pediatrics

Dr. Warren oversees numerous child- and family-serving programs in all 95 counties, including Children's Special Services, Family Planning, Early Childhood Comprehensive Systems, Breast and Cervical Cancer Screening, Injury Prevention, Child Fatality and Fetal/Infant Mortality Review, Lead Poisoning Prevention, Adolescent Health, Asthma, Newborn Metabolic and Hearing Screening, and home visitation services provided through the Department of Health.

Prior to joining the Department of Health, Dr. Warren served as an Assistant Professor in the Department of Pediatrics at Vanderbilt and as Medical Director in the Governor's Office of Children's Care Coordination. He is actively involved in the community and in 2010 was recognized as the Nashville Emerging Leader in the area of Health and Medical Services.

ASK DR. WARREN:

What is your leadership philosophy? In other words what are the qualities important in leading a company, organization, unit, etc. that ultimately motivates everyone to work as a team?

I am a firm believer that everyone on the team has something important to contribute. I learned this early during my Pediatrics residency—I was a July intern in the Neonatal Intensive Care Unit, one of the scariest places for a brand new doctor. Despite having just finished all my medical school coursework and being excited to finally sign "MD" behind my name, I was incredibly green….and knew very little about the practical aspects of caring for premature babies! Fortunately, I had the chance to work with wonderful nurses, respiratory therapists, case managers, and attending physicians who each taught me many important things about caring for those tiny little babies. I learned that I didn't know all the answers (and never would), but with the help of our entire team, we could provide excellent care that saved babies. The same applies to our Maternal and Child Health team. We have an incredibly rich section filled with team members from a variety of backgrounds and with a wealth of public health experience. Each brings unique talents to their work, and our ability to care for the children and families of Tennessee is enriched by their individual strengths. My job as leader is to support each and every individual and make sure that everyone on the team has what they need in order to be maximally successful.

Can you estimate a time line for Newborn Screening results to be web based for access by medical pro providers?

This is a priority for our section. While we do not yet have an estimated date for when this system will be in place, discussions are ongoing about how we move forward with this project. Having practiced in a pediatric primary care clinic before coming to MCH, I know how important it is for providers to be able to access results in a timely and efficient manner, and I am committed to working with our partners at the State Lab to make this a reality.

DR JEWELL WARD

Dr. Jewell Ward currently is medical coordinator of NBS follow-up in W. Tennessee for inborn errors of metabolism. She has served on the Genetic Advisory Committee for the State of Tennessee Department of Health, Maternal and Child Health for several years. She is Professor in the Department of Pediatrics, Division of Medical Genetics, U. Tennessee Health Science Center in Memphis and serves as Division Chief. Additionally she directs an inborn errors of metabolism screening laboratory, primarily evaluating amino acid disorders. At UT she is involved in educational activities for pre-and post–graduate trainees; faculty; and professionals in a variety of disciplines. Her clinical practice includes both patients with inborn errors of metabolism (such as PKU, MCAD, treatment of storage disorders, etc) and general genetic patients (familial disorders, birth defects, genetic counseling, etc). She currently provides Clinical Genetics services at the satellite clinic in Jackson, TN at Jackson Madison County General Hospital, Ayers Clinic.

Dr. Ward graduated from Indiana U. School of Medicine (MD, 1971; PhD, Medical Genetics, 1977). Pediatric Residency training was at Cincinnati Children's Hospital, after which she served as a post-doctoral fellow in Medical Genetics at Johns Hopkins Hospital 1975-79, prior to joining the faculty at the U. Tennessee Health Science Center.

She is a member of the American Board of Medical Genetics, with sub-boards in Clinical Genetics and Biochemical Genetics. She is an elected member of the Society of Inherited Metabolic Disorders. She has been named 'Best Doctors' for several years, most recently 2010. She is on the admitting staff of LeBonheur Children's Hospital and consulting staff of 8 area/regional hospitals. Her knowledge and experience with Medical Genetics has provided an asset to the health and welfare of infants throughout the state of Tennessee.

Her current challenges include incorporating and expanding genetic services with a newly opened LeBonheur Children's hospital.

She lives in Memphis with her husband, Jim Brooks, and two cats, and a son lives in CA. Her weakness is brownies, or anything dark chocolate.

GENETICS ADVISORY COMMITTEE

The Genetics Advisory Committee (GAC) was established in 1985 as an expansion of collaborative efforts between public health, consumers and practitioners. The GAC provides overall direction, policy guidance, monitoring and evaluation to ensure quality comprehensive genetic health care, education, diagnosis, treatment, and support services.

The GAC serves as an advisory group to the Commissioner of the Department of Health. Its members are appointed by the Commissioner for a four

Chief Medical Officer of the state or an appropriate designee chairs the committee that meets twice a year. The committee is comprised of one representative from each the Regional Genetic Center and each Sickle Cell Center and at least 2 members-at-large.

The GAC enhances the expansion of the NBS program by making recommendations to the Commissioner such as the addition of new tests to the screening profile.

(cont on page 5)

ASK DR WARREN *(CONT.)*

What is your vision for MCH and NBS?

The mission of our Maternal and Child Health section is: "to maximize the potential of Tennessee children and families." With that in mind, my vision for the Newborn Screening Program is that we work efficiently to identify children with life-threatening or life-limiting conditions and ensure that every one receives timely follow-up and treatment as appropriate, thereby maximizing their potential. I also think we have a critical role in providing education to parents about the role of newborn screening and also in serving as a support for providers who are caring for children and families in the community.

From your standpoint as a pediatrician, what potential problems do you see with the addition of SCID to the newborn screening panel?

I think that any time we add a test to the screening panel, we need to be mindful of the downstream consequences of positive tests. For every infant who has a positive test, there are parents who are anxious and worried and primary care providers who want to provide the best care possible to the infant with a positive screen. Thus, we need to be very thoughtful about how we educate parents about the newborn screening program, the meaning of a "positive" screen, and the appropriate next steps for ensuring that their child receives the best care possible. We also need to make sure that primary care providers have the necessary information for making clinical decisions, coordinating the care of the infant, and interacting with parents who are eager for information.

Congenital defects, genetic variances, and developmental issues are of great concern to a pediatrician. How do you plan to use your position to educate the physicians?

Community providers play a critical role in our mission of maximizing the potential of Tennessee children and families. Our public health functions provide a critical infrastructure for improving population health and should support the on-the-ground efforts of our community providers. I hope that we can work with providers to identify any needs that they have related to newborn screening and identify opportunities for providing education about the conditions for which we screen, for providing useful information for parents who may have questions about the newborn screening program, and for ensuring a coordinated system of care for babies with positive screens. I want providers to know that we are here as a resource for them and that they should feel free to call upon us at any time.

Do I need to notify someone if a newborn has passed away?

If you are aware of an infant that has expired, please fax the following information to NBS F/U at (615) 262-6458:

- the child's name
- date of birth
- mother's name
- date the infant expired

This is important! In the event a specimen needs to be repeated, NBS F/U will continue to send letters to the mother until the specimen is received. In this case, NBS F/U does **NOT** want to notify the parent and inflict further heartache .

Frequently Asked Questions (FAQs) for Providers

What do I do if an infant was discharged before specimen was collected?
When the nursery or lab forgets to get a specimen on an infant before he/she leaves the hospital, the hospital should submit a newborn screening collection form completely filled in and write, "LEFT BEFORE OBTAINED" on the form. You are responsible for notifying the parent and the physician that the infant was not screened and needs to be tested. Additionally Newborn Screening Follow-Up (NBS F/U) will contact the physician to notify him/her that the infant needs to be screened. When a collection form for these infants is not sent in, NBS F/U is unaware that the infant exists.

What do I do if a parent refuses to let baby be tested?
When a parent refuses the newborn screening test, send in a newborn screening collection form completely filled in and write "REFUSED" on the form and document in patient chart. If a parent refuses the test due to religious reasons, please fill out the Newborn Screening Refusal form. This form should be retained in the medical record for the period of time defined by the hospital or provider per policy.

Can I collect a specimen on a baby who is > 6 months of age?
The test methods used by the Newborn Screening Laboratory are not suitable for infants greater than 6 months of age. If the infant is greater than 6 months of age, contact the Metabolic Center closest to the provider to inquire which tests need to be performed and where to send the specimens.

What is the Hospital's responsibility?
The responsibility of the hospital is to provide the parents with a pamphlet about the Newborn Screening Program, fill out the newborn screening form completely, legibly and accurately and collect a satisfactory specimen on every newborn prior to discharge.

What is the Provider's responsibility?
The responsibility of the provider is to make sure that patient has had a newborn screen, and that it has been reviewed and interpreted with respect to blood transfusion and diet status. Also providers should inform parents/guardian of results. If a specimen is unsatisfactory/abnormal it is the responsibility of parents and providers to obtain a repeat specimen once notified.

Collection Procedure: Visit the Tennessee State Laboratory Website and view the Newborn Screening Directory that provides Collections Procedures and Shipping Information. Also the Newborn Screening Video is available which includes information about the Newborn Screening Program and collection method.

How can I get results for a baby born out of state?
The National Newborn Screening Genetics Resource Center website has a list of all State Laboratory and Newborn Screening Follow Up contact numbers. Please call that State NBS Program for results.

Does the Newborn Screening laboratory run tests everyday?
The regular work hours for the Newborn Screening Lab are 7:00 am to 4:30 pm Monday through Friday. However, mail is picked up from the Post Office daily Monday through Saturday and on holidays. Alternative mail services make daily deliveries to the newborn screening lab Monday through Friday as needed. Alternative mail services can deliver mail to the security guard at front door of the lab on weekends and holidays. Newborn Screening lab does perform screening during the Thanksgiving and Christmas holidays. All mail received on weekends or holidays is dated and run on the next working day in the order of the date received. On any holidays greater than 3 days, lab personnel will come in to perform testing. Specimens will not go longer than 3 days without being tested. Written reports of normal specimens are mailed within 5 to 7 working days after receipt in the laboratory. The nurses from follow-up will notify the Genetic Centers/PCPs for any critical values noted.

GAC

(Cont from page 3)

Currently, they have recommended the formation of two work groups to study the necessity and feasibility of adding testing for Severe Combined Immunodeficiency (SCID) and Critical Congenital Cyanotic Heart Disease (CCCHD) to the screening profile.

They also enhance the integration of NBS and NHS with all aspects of Maternal & Child Health for children with special health care needs by ensuring that a coordinated system of NBS follow-up is maintained between the NBS, the Regional Genetic and Sickle Cell Centers and CSS and Tennessee Early Intervention System. This close collaboration between the centers and Tennessee Department of Health (TDH) has developed a network of genetic and hematological services that offers the latest technology in the field to the rural and urban populations of this state.

State of Tennessee

NEWBORN HEARING SCREENING

Meeting 1-3-6 Goals in Tennessee							
	2007	2008	2009	2010*	State Goal	EHDI Goal	HP 2010 Goal
Total Hearing Screens	92.5%	94.2%	97.8%	97.1%	95%	95%	90%
Before 1 month (of total screens)		98.4%	98.2%	98.4%			
Total Completed Follow-up Received	66.5%	68.9%	69.8%	62.7%	75%	90%	70%
Before 3 months (of total refers)		98.3%	99.3%	93.2%	90%	90%	
Total Enrolled in Early Intervention	12.0%	20.7%	44.4%	47.6%	50%	90%	85%
Before 6 months (of total hearing loss)		76.9%	NA	NA			
Number of infants with Hearing Loss Incidence	35	79 0.90	89 0.83	45 0.50			
* Preliminary data							

Percent Screened	# Hospitals / Birthing Ctrs
100	10
95-99	60
90-94	5
70-89	2
50-69	3
20-49	1

3 of the 4 <69% are small midwifery centers with no equipment.

Test Method	% of Tests
ABR	74.9%
OAE	20.8%
ABR and OAE	3.7%
Unknown	0.6%

Attention Primary Care Providers:

PCPs should ensure infants receive a hearing follow-up evaluation after a referred screen and report results to the NHS program. Training and materials on Newborn Hearing are available at www.aap.org or www.medicalhomeinfo.org

- **Childhood Hearing: A Sound Foundation in the Medical Home**
- **EHDI AAP E-Mail Express – Monthly Newsletter**
- **GAO Report on Deaf and Hard of Hearing Children**
- **Just-In-Time-For PCP Free Material at**

http://www.cdc.gov/ncbddd/hearingloss/freematerials/Just-In-Time-For PCP.pdf

Mark Gaylord, MD, Newborn Hearing, TN AAP Chapter Champion

Follow up services by Tennessee Early Intervention System (TEIS) for Newborn Hearing will be transferred from Kathie Warren at the UT Center on Deafness to Claudia Weber in Nashville, Department of Education TEIS Central Office. Claudia Weber 615-532-3225

Hearing Aid Loaner Bank
Friends of TN's Babies with Special Needs 865-579-2500
www.fotbabies.org

Special Projects :

Teleaudiology – A pilot for diagnostic audiology testing will be conducted in Jackson, TN and submitted by remote upload to Vanderbilt Audiology for interpretation

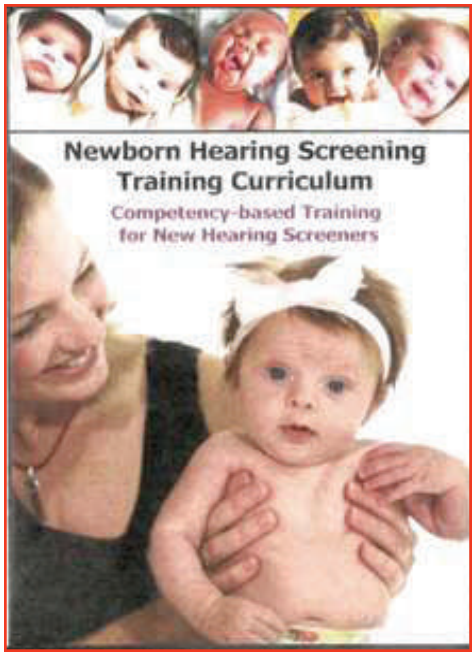
ECHO Project - Early Childhood Hearing Outreach with Early Head Start for hearing screening with 3 centers in TN. www.infanthearing.org/earlychildhood

Hands and Voices is getting a great start in TN. This is a family based organization to offer support to families of children with hearing loss. Call Susie McCamy 865-384-9981 for more Information. www.handsandvoices.org

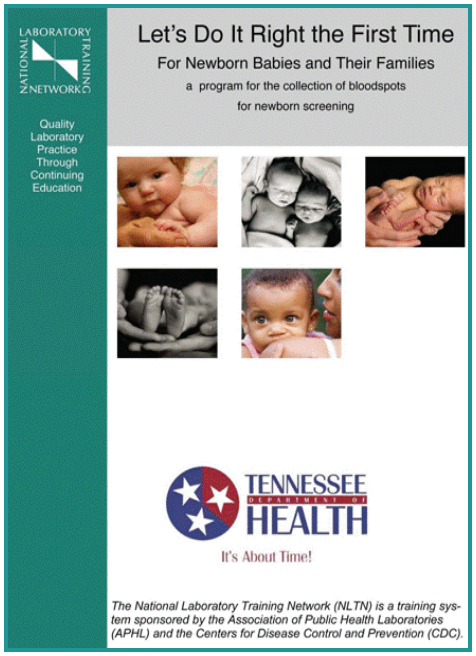
NEW Reporting of Degree of Hearing Loss - Audiologists and ENT's will now report the degree of hearing loss per ASHA standards. These are on the revised TN reporting forms. 615-262-6160

The CARE Project – Parent grief support workshops in 3 areas across TN. Contact Susie McCamy 865-384-9981

State of Tennessee



Hearing DVD available free upon request.
Please Call
615-262-6160



Metabolic DVD available free upon request. Please call
615-262-6304
www.nltn.org/318-10.htm
www.nltn.org/317-10.htm
CEU's available

Important Newborn Screening Phone Numbers at a Glance

NBS Metabolic Follow-Up
Ph 615-262-6304
Fax 615-262-6458

NBS Hearing Follow-up
Ph 615-262-6160
Fax 615-262-6159

Filter Papers
Ph 615-262-6391
Fax 615-262-6455

Pamphlets
Ph 615-262-6304
Fax 615-262-6458

Billing
Ph 615-262-6300
Fax 615-262-6396

ICD9 Codes
Ph 615-262-6300

TENNESSEE DEPARTMENT OF HEALTH
It's About Time!

State of TN Dept of Health
Newborn Screening and Genetics
630 Hart Lane
Nashville, TN 37243

Phone: 615-262-6304
Fax: 615-262-6458
E-mail: Melissa.C.Smith@tn.gov

Let's Do It Right The First Time!

State of Tennessee
Newborn Screening Program
Voice Response System for Test Results

Required:
(1) TN Physician License Number
(2) Personal Identification Number
(3) Mother's SSN
(4) Touch tone phone
(5) Fax machine if you want faxed results

Results Available 24/7

Local (Nashville) 262-3041
Toll Free (866) 355-6132

NOT ON OUR EMAIL LIST?
CONTACT LISA @ 615-262-6304

VRS

Having problems or need to sign up?
Contact Nancy at
615-262-6304