



STATE OF TENNESSEE  
DEPARTMENT OF HEALTH

JOHN J. DREYZEHNER, MD, MPH  
COMMISSIONER

BILL HASLAM  
GOVERNOR

Dear Colleague,

Thank you for your ongoing partnership in the well-being and care for people in Tennessee. We wanted you to be aware of improvements in the Tennessee Department of Health Newborn Screening Program that could impact your practice.

**Key Points for You to Know**

- Screening for Severe Combined Immunodeficiency (SCID) is expected to begin in January 2016. It is estimated to affect one in 50,000 to 100,000 live births.
- If SCID is identified in the first few months of life prior to development of significant infections, hematopoietic stem cell transplantation is successful in 95% of cases.
- Tennessee's Newborn Screening Program now screens for more than 50 conditions using a dried blood spot (DBS), for critical congenital heart disease (CCHD) using pulse oximetry and for congenital hearing loss via hearing screening.
- If testing identifies a possible abnormality, nurses in the Department's Division of Family Health and Wellness Newborn Screening Follow-Up contact the infant's physician and advise on the next action steps.
- **Rotavirus vaccination is contraindicated for infants suspected to have SCID.**

**Background**

SCID is a primary immunodeficiency disease and is a group of disorders caused by congenital defects in the development or survival of T-lymphocytes. It often manifests in the first few months of life with failure to thrive and severe, recurrent infections. It is almost universally fatal in the first two years of life unless treated via hematopoietic stem cell transplant. If caught in the first few months of life prior to development of significant infections, transplantation is successful in 95% of cases. It is estimated to affect one in 50,000 to 100,000 live births.

With the addition of SCID Tennessee's Newborn Screening Program now screens for more than 50 conditions using a dried blood spot (DBS), for critical congenital heart disease (CCHD) using pulse oximetry and for congenital hearing loss via hearing screening.

The testing methodology for SCID will be a T-cell Receptor Excision Circle (TREC) assay. This assay detects the presence of TRECs, which are present only in newly developed T-cells, and is thereby a reflection of successful T-cell development and thymic function. A low or absent TREC value indicates that the baby is not making T cells properly. Aside from SCID, the TREC assay can also detect several other forms of primary immunodeficiency and secondary causes of profound lymphopenia. An important consideration when screening for SCID is that babies who are born at less than 37 weeks may have low TREC values simply due to prematurity, especially those with very low or low birth weight.

In Tennessee, the Department of Health's Public Health Laboratory provides testing for specimens sent from the hospitals. If testing identifies a possible abnormality, nurses in the Department's Division of Family Health and Wellness Newborn Screening Follow-Up program contact the physicians and advise of actions needed on your part. The local Genetic Center will also be notified of the abnormality. The Center will arrange confirmatory testing with the local pediatric immunologist, and notify the primary care provider and the parent.

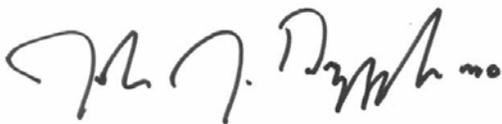
Because of the need for a rapid diagnosis and initiation of therapy for confirmed cases, expedient immunologic evaluation is necessary for any infant with a positive TREC screen in whom SCID is suspected. Please note that pediatric immunologists will join the network of specialists currently involved in evaluation of infants with a positive newborn screen. They will help coordinate and provide confirmatory testing, diagnosis and treatment for babies diagnosed with SCID.

The Centers for Disease Control and the US Food and Drug Administration announced in 2010 that the Rotavirus vaccination is contraindicated for infants with SCID. Therefore, **the Tennessee Newborn Screening Program recommends that all infants with abnormal SCID results not be given the Rotavirus vaccine until the infant is fully evaluated.** It is recommended that all primary care physicians review the newborn screening results to be assured of the SCID testing status prior to administering the vaccine. Until referral, infants with a positive TREC assay should be strictly shielded from any sick contacts. Live vaccines (Rotateq) should be strictly avoided until the infant is fully evaluated.

We appreciate your ongoing support of the Newborn Screening Program and your partnership in helping us to identify and initiate treatment for diseases as early as possible. You play a critical role in this effort to provide every infant born in Tennessee with the best possible start to life. Should you have any questions or concerns, please contact our Newborn Screening Follow-Up Program at (615) 532-8462.

For additional information on SCID please visit <http://tn.gov/health/article/MCH-nbs-providers>

Sincerely,



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Commissioner



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