Behind the Screens

Missouri Department of Health and Senior Services

Newborn Screening Program

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Featured Disorder

Sickle cell disease (SCD) is an inherited blood disorder that affects the red blood cells' hemoglobin (a protein that carries oxygen). Normal red blood cells are round and flexible and move through tiny blood vessels to carry oxygen to all body parts. In SCD, the red blood cells contain an abnormal type of hemoglobin, which causes the cells to become crescent or "sickle" shaped. These sickle cells become stiff and rigid and can lodge in the blood vessels. The lack of blood flow as a result of these blockages can result in serious medical problems, including anemia, pain episodes, infection and potentially life-threatening complications such as acute chest syndrome, stroke, and organ damage.

Sickle cell conditions are inherited from parents in much the same way as blood type, hair color, eye color, and other physical traits. The types of hemoglobin a person makes in the red blood cells depends upon the hemoglobin genes they inherit from their parents. Like most genes, hemoglobin genes are inherited in two sets, one from each parent.

Sickle cell trait (SCT) is when a person inherits one sickle cell gene and one normal gene. People with SCT do not usually have SCD symptoms but can pass the sickle gene on to their children. Suppose two parents are carriers of SCT. In that case, with each pregnancy, a child has a 25 percent chance of having sickle cell disease, a 25 percent chance of inheriting two normal genes, and a 50 percent chance of being an unaffected carrier like the parents.

There are several forms of SCD. The most common types are hemoglobin SS, also known as sickle cell anemia, which is the most severe form of SCD; hemoglobin SC (Sickle Hemoglobin C disease); sickle beta plus thalassemia and sickle beta-zero thalassemia.

SCD is the most common inherited blood disorder in the United States, affecting approximately 80,000 or more African Americans. Although the disease occurs predominantly among people of African descent, it is also found in individuals with Mediterranean, Middle Eastern, Indian and Latin American ancestry.

It is essential to test babies for SCD. Early diagnosis and treatment of SCD ensures that a program of life-prolonging care begins in the first few months of life to help prevent and manage the symptoms of the disease. Newborn screening for SCD and other hemoglobinopathies (abnormal hemoglobins) began in Missouri in 1989. About 1 in 400 African American newborns in Missouri has the disease, and 1 in 12 are carriers of the sickle cell trait.

Patient Spotlight

Ronald, Rylie and Reese

Erica is a proud mother of three beautiful children - Ronald, Rylie and Reese. Newborn screening played a role in detecting sickle cell disease in all of her children shortly after they were born. Although each child has had a different medical journey, Newborn screening helped to ensure they all received early diagnoses and proper medical treatment necessary to manage their symptoms and promote their growth and development.

"Hello, my name is Erica. I have three children who all have sickle cell disease. When I was about to have my first son, they had me do some last-minute blood tests. That is when I found out I had sickle cell trait. After Ronald was born on June 30, 2019, we were instructed to go to Cardinal Glennon Children's Hospital, where they told me his newborn screening results indicated he had sickle cell disease , and they wanted to confirm it. When I found out, my heart sank. Fast forward to when he was 4 months old. We took him to the ER for a temperature that wouldn't go down. My mom and I were balling our eyes out, but his dad was so strong. From then on, Ronald would be in and out of the hospital.

My middle child, Rylie, was diagnosed with sickle cell. We already knew what to expect, but we were still worried. Thank God she only had one hospital stay, and other than penicillin, she isn't on any other medications.

My baby boy, Reese, has sickle cell anemia as well. He wasn't in and out of the hospital like Ronald, but he had his share. His first hospital stay was at a few months old because of a temperature spike that would not go down. Then he had respiratory issues like Ronald, so Reese and Ronald have inhalers on standby just in case.

I say all of this to say God never makes mistakes. I thank him for the know-how and the ability to do the right thing for my children. I'm just saddened that their aunt, their father's older sister, who died of sickle cell disease, never got a chance to meet them."

Erica,

Mom of Ronald, Rylie and Reese

Did You KNOW?

1. Bright or infrared light, including bilirubin lamps and surgical lights, can affect the accuracy of a pulse ox reading. Ensure the infant is not placed in bright or infrared light while critical congenital heart disease (CCHD) screening is performed. You can cover the pulse ox sensor with a blanket to ensure that extraneous light does not affect the accuracy of your reading.

2. You should never discard a newborn screening (NBS) collection form. If you make a mistake or suspect the specimen will be a poor-quality sample, allow the specimen to dry completely, write VOID across the front of the form, and return it to the Missouri State Public Health Laboratory along with the correct form. The voided form will be replaced free of charge.

Tech Tips

- Report hearing screening results through Missouri Electronic Vital Records (MoEVR) no later than seven days from the screening date.
- When collecting a newborn blood spot screen, including repeat specimens, always document the gestational age and weight at birth on the collection form.
- Accurate date and time of blood spot collection is crucial. This data cannot be amended once the Missouri State Public Health Laboratory receives the specimen.

MISSOURI DEPARTMENT OF
HEALTH & SENIOR SERVICES

Bureau of Genetics and Healthy Childhood
Newborn Blood Spot, Hearing and
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Missouri State Newborn Screening Laboratory
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thank you

for your contribution in ensuring the best possible start for Missouri newborns!