SICKLE CELL DISEASE RAISES A CHILD’S STROKE RISK. CLOSE EVALUATION AND TREATMENT MAY HELP PREVENT STROKE.

KEY FACTS

- Sickle cell disease (SCD) is a blood disorder where misshaped or “sickled” red blood cells can clog blood vessels and cause complications including stroke and blood vessel damage (including to the brain).
- All babies in the U.S. are screened for sickle cell disease, which affects 1 in 2,400 American children, and 1 in 400 African-American children.
- Stroke risk is high in all children with sickle cell disease. The stroke risk is increased over 100-fold in children with SCD compared with other children without sickle cell disease.

The most severe form of the disease, called sickle cell anemia, occurs when a person inherits two abnormal copies of the hemoglobin-Beta gene. Sickle cell trait describes the presence of one abnormal and one normal copy of the gene. Sickle cell trait does not cause symptoms, but people with this condition are “carriers” — meaning they’re at risk of having children with sickle cell disease.

In 1998 it was estimated that before age 20, 11% of patients with sickle cell anemia had a stroke with physical signs, such as weakness in an arm or leg, sometimes referred to as an “overt stroke.” Today, with appropriate screening, called the Sickle Stroke Screen or transcranial doppler ultrasound (TCD), and use of stroke prevention therapies in high-risk patients (usually chronic red blood cell transfusions), the risk of stroke can be reduced significantly.

The common stroke presentation in children with sickle cell disease are “silent strokes,” or “silent” injury to the brain. These may occur in up to 39% of children by age 18. Silent strokes do not have any outward physical signs such as arm or leg weakness but can be seen on an MRI of the brain. Silent strokes may cause problems in thinking, learning, and decision-making. Silent strokes are a risk factor for future strokes.

There are two main types of strokes: Ischemic (caused by a blockage in blood flow) and hemorrhagic (bleeding in or around the brain). Ischemic strokes are more common in children and hemorrhagic more common in adults with SCD. However, both types of strokes may occur at any age.

Learn more at stroke.org.
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WHAT YOU CAN DO

• Guidelines recommend all children with sickle cell anemia have an MRI screening of the brain when they are old enough to hold still for the test without sedation.

• Work with your child’s health care team to determine if annual stroke screening is appropriate for your child. The Sickle Stroke Screen — also called transcranial doppler (TCD) ultrasound — is a painless annual test for stroke risk that measures the speed of blood flow in the brain. Experts recommend this test for children ages 2 to 16 with sickle cell anemia. Despite this test being recommended every year, fewer than half of children who qualify received it and are screened.

• If the Sickle Stroke Screen detects any problems, work with your child’s hematologist to review and discuss treatment to prevent stroke, which may involve approximately monthly blood transfusions or a medicine called hydroxyurea.

• All children with sickle cell disease should be seen by their sickle cell doctor (a hematologist) every year for routine screening, even if they feel well.

• If your child with sickle cell has a stroke, continue to work with their health care team to discuss post-stroke management that may include: seeing a neurologist, rehabilitation therapy, extra help at school and possible monthly blood transfusions. Other treatments might be offered depending on the exact issues and situation.

LEARN THE SIGNS OF STROKE

Symptoms of stroke are similar in people with and without sickle cell disease. Use the acronym F.A.S.T. to remember the warning signs of stroke: Face drooping. Arm weakness. Speech difficulty. Time to call 911.

If your child has one or more of these signs, don’t delay — call 911 or your local emergency number.

If it looks or feels like a stroke, it may be one.

Other stroke symptoms include sudden onset of:

Confusion  Trouble walking

Headache  Vision problems

Numbness

Learn more at stroke.org.

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