Sickle Cell Disease and Stroke
Sickle cell disease (SCD) is the most common inherited disease among African Americans. In the United States, sickle cell anemia affects about 72,000 people — most of whom are African Americans. The disease occurs in about:

- One in every 600 African-American births.
- One in every 1,000–1,400 Hispanic-American births.

As the parent of a child with SCD, you may know that SCD can lead to pain, anemia, serious infections and damage to major organs. But you may not know that nearly 11 percent of children born with the most severe form of sickle cell disease will have a stroke by age 20.¹

The American Stroke Association and the Sickle Cell Disease Association of America want to help you help your child live a long, healthy life. Knowledge is power. So use this booklet to learn more about sickle cell disease and how to reduce your child’s risk of stroke.

**How does sickle cell disease relate to stroke?**

Sickle cell disease is a red blood cell disorder. Hemoglobin (hee’ moh gloh bin) is the protein in red blood cells that carries oxygen. It underlies the problem in sickle cell disease.

¹Cooperative Study of Sickle Cell Disease
Normal blood cells are round like doughnuts and flexible. They squeeze through small blood vessels to deliver oxygen to the body’s tissues. The sickled hemoglobin sticks together to form long rods in the red cell when it gives away oxygen. These rigid rods can change the red cell from round and flexible to a shape like a sickle (farm tool used to cut wheat). The sickled cells tend to stick to and damage the inner wall of blood vessels. They also clog the flow of the blood. In the brain, this can cause a stroke.

Children with SCD are 200 to 400 times more likely to suffer a stroke, compared to children without SCD. The Cooperative Study of Sickle Cell Disease showed that stroke is a constant threat after age 2, but the incidence is highest in the middle of a child’s first 10 years of life.

What happens when a stroke occurs?

When blood is cut off to part of the brain, cells in that area die. When the brain cells no longer work, the parts of the body they once controlled don’t work either. A stroke can damage your child’s ability to walk, talk and/or understand. It can also affect their emotions and memory. There are two main types of stroke: ischemic and hemorrhagic.

- **Ischemic (iss kem’ ik) strokes** are the most common type of stroke for children with sickle cell disease. They’re caused when narrowed blood vessels leading to the brain become blocked. This stops blood flow so oxygen can’t get to the brain.

- **Hemorrhagic (hem’ or raj ik) strokes** occur when blood vessels in or around the brain break.
Children over age 2 who have sickle cell anemia should be screened for stroke using an ultrasound test called Transcranial Doppler (TCD). TCD is a device that uses painless and harmless sound waves to find areas of abnormal blood flow in the brain’s blood vessels. When sickle cell damage narrows the blood vessels, the blood flows faster through the narrowed area and makes a louder noise. This means that a stroke is more likely.

A large national study called the Stroke Prevention Trial in Sickle Cell Anemia (STOP), performed in the 1990s, showed that children with abnormal TCD who receive regular (about monthly) blood transfusions have a 90 percent lower risk of stroke. So TCD is an important test. Combined with blood transfusion, it can prevent disabling stroke. TCD is recommended yearly during early childhood. But it may not be needed as often, depending on your child’s results. Your child’s provider also may order other tests, such as magnetic resonance (MR) of the brain to better understand the risk of stroke or other problems.

If your child has sickle cell disease, ask your child’s doctor about using TCD and MR screening to test the brain for stroke. TCD is available at most major health centers.
**What do I do if my child has a stroke?**

First, be prepared. Learn the stroke warning signs so you can quickly recognize them. Share this information with everyone in your child’s network — teachers, daycare workers, church youth leaders or anyone else your child may regularly come in contact with. Quick response to the warning signs below will help limit damage — and save lives.

The warning signs are:

- Sudden weakness or numbness of the face, arm or leg, especially on one side of the body.
- Sudden confusion, trouble speaking or understanding.
- Sudden trouble seeing in one or both eyes.
- Sudden trouble walking, dizziness, loss of balance or coordination.
- Sudden, severe headaches with no known cause.

(In very young children, these warning signs may not be this specific. They could include things such as inconsolable crying.)

If you think your child is having a stroke, respond immediately by calling 9-1-1. Don’t wait to see if the symptoms disappear. With every passing
minute, more damage occurs in the brain. Get your child the best treatment possible by quickly responding.

Acute stroke symptoms need to be checked immediately. They’re usually treated with hydration (administering intravenous fluids) and blood transfusion to reduce permanent damage to the brain. A CT scan is also needed to rapidly tell if a person had a hemorrhagic stroke or an ischemic stroke.

**Will my child get better after a stroke?**

Recovery from stroke differs with each child. Prompt medical attention and rehabilitation therapy can maximize recovery. In general, younger people recover more abilities than older people do. Children often recover the use of their arms and legs and their ability to speak after a stroke.

**What treatments prevent recurring strokes?**

Up to two-thirds of children with SCD and stroke will have more strokes unless they’re treated.

Transfusions (to receive donated blood) can prevent a first stroke when the TCD is abnormal, and they can generally prevent second and third strokes in patients with SCD who have already had a stroke. A transfusion will help replace the sickled red blood cells in your child’s body with normal donor cells.

STOP Study. NEJM 1998 reference and Medical College of Georgia website.
When the first stroke occurs, most children get a transfusion to reduce the blocked blood flow caused by the sickle cells. After that, doctors suggest monthly transfusions due to the high risk of another stroke. Transfusions should continue for at least three to five years.\(^3\)

After about two years of monthly blood transfusions, people will have too much iron in their bodies. Iron chelation (removal) therapy with deferoxamine (Desferal) helps keep the body healthy by allowing it to pass excess iron out through the urine.

Transfusions help prevent stroke and reduce other complications of SCD. But there are drawbacks. Talk to your doctor about the risks and benefits for your child. It’s best to talk to healthcare professionals experienced in transfusing people with sickle cell disease and managing iron overload.

**How can I help reduce my child’s risk for stroke?**

The best defense against a first or subsequent stroke in children who have SCD is to effectively manage their disease. Even if you can’t prevent the first stroke, you can help *reduce* your child’s risk of permanent damage from another stroke. Work with your doctor to develop a treatment plan that’s right for your child.

\(^3\)NIH Publication No. 95-2117, Revised December 1995 (3rd Edition) National Institutes of Health, National Heart, Lung, and Blood Institute
Where can I get more information?

Talk to your doctor. Learn all you can about managing your child’s SCD and find out your child’s risk for stroke. Share this information with everyone in your child’s network to get help quickly in an emergency.

For more information on SCD, contact the Sickle Cell Disease Association of America at 1-800-421-8423 or at www.sicklecelldisease.org.

You can find information on the STOP study, use of TCD and sites trained to perform TCD for children with sickle cell disease at the Medical College of Georgia Web site using this link: http://www.mcg.edu/neurology/Research/sicklecell/index.htm

For more information on stroke, call 1-888-4-STROKE (1-888-478-7653) or visit StrokeAssociation.org/power.
Glossary

**CT Scan** — an X-ray test that can show areas of the brain affected by stroke.

**Hemorrhagic Stroke** — death or injury to brain cells caused by a blood vessel that ruptures in or on the brain.

**Ischemic Stroke** — death or injury to brain cells caused by a clot that blocks an artery bringing blood to part of the brain.

**MR (Magnetic resonance)** — an imaging test that uses powerful magnets to form pictures of organs inside the body. It can show how well the brain works.

**SCD** — a genetic blood disorder that mainly affects African Americans. Red blood cells become sickle-shaped, are less able to carry oxygen and are more likely to stick to blood vessel walls, increasing the risk of stroke.

**TCD (Transcranial Doppler)** — an ultrasound examination of blood flow in the brain’s blood vessels.

**Transfusion** — giving a person donated blood.
With branches reaching skyward and roots burrowed deep in the earth, the Tree blends and balances dreams and reality, heaven and earth. As a symbol of life for African Americans, it represents growth and health.

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