Strokes and Silent Strokes

In children with sickle cell disease

Sickle cell disease is a blood disorder you get from your parents (inherited) that affects 1 in 2,400 American children, and 1 in 400 African-American children in the United States.

The best medical care for children with sickle cell disease happens when parents know the signs and symptoms of medical problems, such as stroke, which can happen at a young age.

What is a stroke?

• A stroke happens when blood flow to an area of the brain is blocked. As a result, that part of the brain does not get enough oxygen and is damaged.

• Children and adults with sickle cell disease can have two types of strokes:
  – a stroke has physical signs, such as weakness in an arm or leg
  – a silent stroke has no physical signs, but may affect the patient’s thinking and learning.

What are the warning signs of a stroke?

• sudden weakness or numbness of the face, arm, or leg on one side of the body
• sudden loss of vision or dim vision
• loss of speech, trouble talking, or trouble understanding others
• sudden, very bad headaches with no known cause
• sudden falls, dizziness, or unsteady walking with no known cause.

Call 911 or take your child to the nearest emergency room, if you think your child is having a stroke.
What should I do if I think my child is having a stroke?

A stroke is a serious medical emergency. During a stroke, blood and oxygen cannot get to a part of the brain. The longer that part of the brain goes without oxygen, the greater the chance of injury to the brain.

Call 911 or go to the nearest emergency room, if you think your child is having a stroke.

Can I find out if my child is likely to have a stroke?

The risk of strokes in children with sickle cell disease (hemoglobin SS or S beta thalassemia zero) is higher when blood moves too quickly through blood vessels in the brain. To measure how fast blood is flowing in the brain, doctors use a Transcranial Doppler (TCD) machine. During a TCD test, a device about the size of a big crayon is put on the side of the child’s head to measure blood flow.

A TCD test does not hurt and takes about 30 minutes.

If the speed of the blood flow is too high, a child may be at greater risk for stroke. If the TCD test shows fast blood flow, doctors will do a second TCD to be sure the test was correct. If the second test shows fast blood flow, the child may need to get donated blood. This is called a blood transfusion.

We recommend children with sickle cell anemia get a TCD test at age 2, and every 12 or 18 months each year after, until the age of 16.

How is a stroke treated?

There are two ways a stroke can be treated in children with sickle cell disease:

- blood transfusion therapy and
- bone marrow transplant.

**Blood Transfusion Therapy**

The standard treatment for strokes in patients with sickle cell disease is blood transfusion therapy. The goal of transfusion therapy is to lower the amount of red blood cells that can sickle in the patient’s blood (hemoglobin S) to less than 30 percent.

The level of hemoglobin S in a child with sickle cell disease may be as high as 90 percent. A child who has had a stroke must get a blood transfusion every 3 to 4 weeks for most of his or her childhood and possibly through adulthood to lower the chances that the child will have another stroke or even a third stroke.

With regular blood transfusions, the risk of stroke can be reduced, but will not go away completely.

Without transfusion therapy, 5 out of 10 children who have had one stroke may have a second stroke or silent stroke.

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Possible problems of blood transfusion therapy

High amounts of iron in the body. Blood has iron in it. Extra iron can build up in the body after 1 or 2 years of blood transfusion therapy. Extra iron can damage the liver, heart, and other parts of the body. Doctors measure ferritin, a substance that carries iron, to find out if a patient has too much iron. A medicine called deferasirox (Exjade) or desferal can help remove extra iron in the body.

Infection. An infection from a blood transfusion is rare because all blood is tested for HIV and other viral infections. The risk of second strokes in patients who do not receive blood transfusion therapy is far greater than the risk of getting an HIV infection from blood transfusions.

Rejection of donated red blood cells. Patients who get blood transfusions need blood types similar to their own so that their body doesn’t try to kill off the donated blood (this is called alloimmunization). In some cases, blood transfusion therapy may have to be stopped. To lower the chance of rejection, your hematologist will find the best-matched blood available.

Bone Marrow Transplant

Bone marrow transplant is an experimental treatment option for some children with strokes.

Only 1 in 5 children will have a brother or sister that can donate bone marrow, and the procedure can be life threatening. You should talk with your child’s hematologist before deciding on a bone marrow transplant. For children who have had a stroke and have a matched sibling, a bone marrow transplant is the first treatment choice of preventing a second stroke. For children without a sibling match, talk to your hematologist about other transplant options.

Why are silent strokes a big problem?

A silent stroke is an injury to the brain that does not cause symptoms you can see. Silent strokes happen in about 3 out of every 10 children with sickle cell disease before their 6th birthday.

A silent stroke can cause the child to do poorly in school, to forget a lot, and have a hard time following simple instructions.

Having silent strokes puts children at higher risk for more silent strokes or even strokes. Tests of your child’s thinking, learning, and memory skills can be done to find the impact of stroke on your child’s school work.

The only way to know for sure if a child has had a silent stroke is to take pictures of the brain using a magnetic resonance imaging (MRI) machine. An MRI does not hurt and takes about 30 minutes.

We recommend that school-age children with hemoglobin SS have at least one non-sedated MRI of the brain. An MRI may be done at other times if your child’s hematologist recommends it.

How is a silent stroke treated?

We do not know the best medical treatment for silent strokes. However, if a child has a silent stroke, we will refer him or her to a neurologist for an evaluation and to a psychologist for education testing. Both the evaluation and the testing can help parents and the child’s teachers better understand the child’s strengths and weaknesses, so that the child can get support in the classroom. Sometimes this support is called an Individual Education Plan (IEP) and other times is called a 504 plan.