University Hospitals
Seidman Cancer Center

Rainbow
Babies & Children's

PATIENT INFORMATION: Sickle Cell Disease: The Basics

What is sickle cell disease?

Sickle cell disease is a group of inherited blood disorders. The red blood cells contain an abnormal form of hemoglobin, the protein in red blood cells that carries oxygen throughout the body. Red blood cells are normally round, donut-shaped and flexible. In sickle cell disease, the red blood cells become sickle shaped and inflexible. These sickle-shaped cells can keep blood from flowing to parts of the body like organs and tissues. When this happens, patients can develop pain, anemia and other problems. One in 400 African-Americans is born with sickle cell disease. It is estimated that 100,000 people in the United States have sickle cell disease. Sickle cell disorders are common in people who are African, Mediterranean, Indian, Middle Eastern and Caribbean.

How do you get sickle cell disease?

Children with sickle cell disease are born with it. They will have it all their lives and will not outgrow it. It is inherited, which means passed down from parents. If both parents have the sickle cell trait, there is a 25 percent chance that a child will be born with the disease. Sickle cell disease is not contagious.

Effects of sickle cell disease

Effects (symptoms or complications) are different depending on which type of sickle cell disease the child has. Every child is different.

Some of the complications may include pain, infection, acute chest syndrome, fatigue, jaundice, learning disabilities and stroke.

Fatigue is common in patients with sickle cell disease because of their anemia; jaundice or yellowing of the eyes is also common because the red blood cells are breaking down quickly. Learning difficulties are a problem for many students as a result of fatigue, missed school or even brain injury caused by stroke.

Pain is the most common problem. It is caused by sickled cells blocking blood flow to a part of the body. It can come on suddenly and last for days. It is important to learn about triggers for pain including heat, cold, dehydration and infection. Patients may be treated with pain medicine and sometimes require hospitalization.

Infection can be life-threatening. It is important that any child with a fever is evaluated by a doctor and given antibiotics by an IV or shot. Patients are put on antibiotics daily when they are young to prevent infection.

Acute chest syndrome (ACS) causes difficulty breathing and is a serious problem of sickle cell disease. Children can have a cough, fever, chest and abdominal pain, or trouble breathing. Children with symptoms of ACS must go to the hospital immediately.

Stroke is caused by lack of oxygen to the brain. Children with certain types of sickle cell disease are at risk of having a stroke.

Treatment

Treatment for sickle cell disease depends on the complications that each patient experiences. Each child is different.

Medications such as hydroxyurea may be needed to help prevent complications. Some patients require regular blood transfusions to prevent stroke or severe complications. Bone marrow transplant is the only cure for sickle cell disease but requires a very closely matched donor which is hard to find.

It is important to work closely with the sickle cell team and the pediatrician to keep your child healthy. It is important that your child attends all visits and receives immunizations on time.

Rainbow Sickle Cell Anemia Center

If your child has signs or symptoms of illness, please call 216-844-3345 and follow the prompts.

Angie Fowler Adolescent & Young Adult Cancer Institute at University Hospitals

2101 Adelbert Road, Cleveland, Ohio 44106

Have a question? Ask a Rainbow doctor. **216-UH4-KIDS** | **216-844-5437** | Rainbow.org/AngiesInstitute

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