Written by Debra A. Vedro, M.S.N., R.N., C.P.N.P., and Becky Morrison, M.S:N., R.N., C.P.N.P. Comprehensive Sickle Cell Disease Program, Children's Medical Center of Dallas, Texas.

Originally Reprinted with permission of Texas Department of Health. Originally supported in part by Texas project #MCJ-481004 from Maternal and Child Health Newborn Screening Program.

Washington State Department of Health

DOH 951-104 10/93

HEMOGLOBIN SICKLE C DISEASE

The Child with Hemoglobin Sickle C Disease

Hemoglobin sickle C disease is a “mild" form of sickle cell anemia. Your child's red blood cells (RBC's) contain two abnormal hemoglobins called hemoglobin S and hemoglobin C.

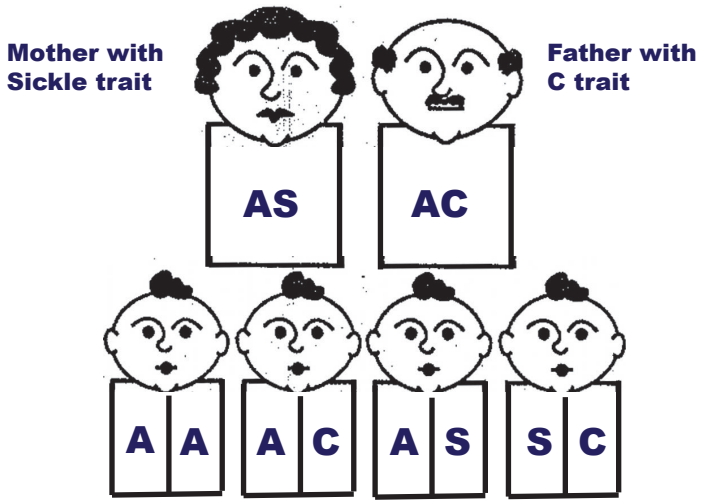
Hemoglobin is the substance in the RBC's which carries oxygen to all parts of the body. Instead of appearing round (donut-shaped), your child's RBC's are somewhat misshapen, some even appear like the sickle-shaped cells found in sickle cell anemia.

Red Blood Cells

Normal RBCs | SC RBCs | SS RBCs (sickle cell anemia)

How Hemoglobin Sickle C Disease is Inherited

When one parent has sickle trait and the other parent has C trait, each baby has a 1 in 4 chance of inheriting Sickle C disease.



Problems Seen in Children With Sickle C Disease

Anemia

Your child will always have a slight decrease in his blood count - this is called anemia. Except for occasionally causing tiredness and or weakness in some children, the mild anemia usually does not cause any problems.

Pain

The red blood cells of Sickle C disease are rigid and stiff and may sometimes "clog up" the small blood vessels in the bones and other parts of the body. Because enough oxygen cannot get into the bones, this can cause pain.

Painful episodes occur most commonly in the arms, legs, stomach and back. These episodes can last for hours, days, or up to a week. The pain can vary from mild to moderate to severe. The location, length, and degree of pain, can vary from episode to episode. How often these crises occur is variable. Some children with SC Disease have no crises at all, but most will have a few each year.

Pneumonia

A child with SC disease has an increased risk of getting certain infections, particularly pneumonia. The abnormal RBCs can "clog up" in the lungs and increase the risk of infection there and cause pneumonia. This is called chest syndrome.
Symptoms to watch for include: fever, fast breathing, trouble breathing, retractions (ribs "suck in" when breathing), very congested cough, and chest pain.

Spleen

The spleen is normally a small organ located on the upper left side of the abdomen up under the rib cage. It acts as part of the body's defense system that fights infection by removing bacteria (germs) from the blood.

Children with SC disease may have an enlarged (big) spleen but this does not happen until they are about age 5 years or older. This big spleen usually does not cause any problems. Occasionally, teenagers and/ or adults may have pain over the spleen and a drop in the blood count - this is called a "spleen crisis."