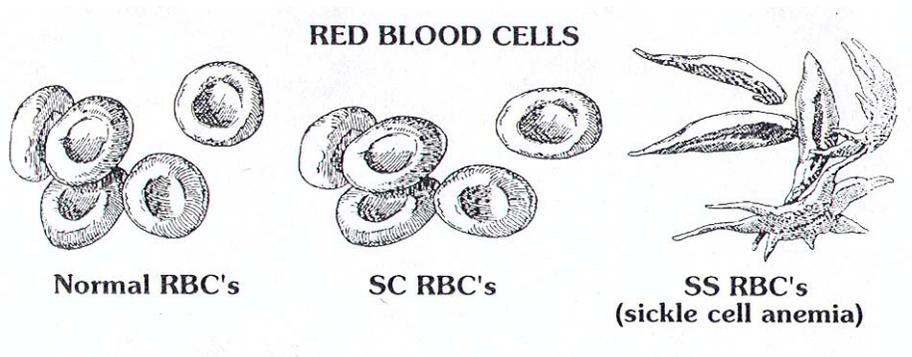


# Sickle-C Disease

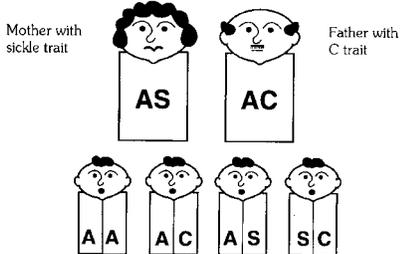
## WHAT IS SICKLE-C DISEASE (Hb SC)

Hemoglobin sickle C disease is a "mild" form of sickle cell anemia. Your child's red blood cells (RBCs) contain two abnormal hemoglobins, called hemoglobin S and hemoglobin C. Hemoglobin is the substance in the RBCs that carries oxygen to all parts of the body. Instead of appearing round (donut-shaped), your child's RBCs are somewhat misshapen, and some even appear like the sickle-shaped cells found in sickle cell anemia. Others are folded or football-shaped.



## HOW HEMOGLOBIN SICKLE C DISEASE IS INHERITED

### HOW HEMOGLOBIN SICKLE C DISEASE IS INHERITED



Babies with sickle-C disease have inherited two (2) abnormal hemoglobins, hemoglobin S and hemoglobin C. If one parent has AS (sickle trait) and the other AC (C trait), there is a 1-in-4 chance (25 percent) the baby will inherit AA (normal), AS, AC, or SC Disease. These chances remain the same for each pregnancy.

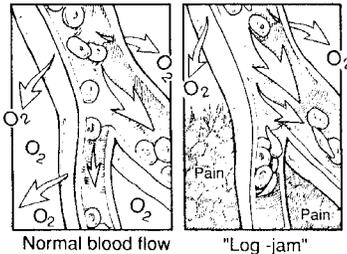
## PROBLEMS SEEN IN CHILDREN WITH SICKLE C DISEASE

### 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 sickled red blood cells in sickle-C disease, somewhat like those in sickle cell anemia, are rigid and stiff and may sometimes cause "log jams" in the small blood vessels in the bones, organs, and other parts of the body. Since oxygen (which is carried by the red blood cells) cannot get past the "log jams" into the bones and organs, this can cause pain. These painful episodes occur most commonly in the back, stomach, arms, and legs. They can last for a few hours to as much as a week or more. Occasionally, the pain is accompanied by swelling. Painful episodes can vary in severity of the pain and the duration. There are medicines to help ease the pain and make your child more comfortable.



### Acute Chest Syndrome/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. This is called acute chest syndrome. Symptoms to watch for include: fever, fast breathing, trouble breathing, retractions (ribs "suck in" when breathing), very congested cough, and chest pain. If these occur, your child needs to see a doctor immediately.

### 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 5 years or older. This big spleen usually does not cause any problems. Occasionally, teen-agers and/or adults may have pain over the spleen and a drop in the blood count - this is called a splenic sequestration.

### Eyes

Older children (over age 10 years) and adults with sickle C disease may develop damage to the retina in the back of the eye. This may cause blindness if it is not treated in time. Regular eye check-ups by an ophthalmologist (medical eye doctor) are necessary to diagnose and treat this problem in its early stages.

### WHAT YOU CAN EXPECT



A child with sickle-C disease can have a normal life-style and life span. Your child should not be considered "sick", and you should treat him or her normally. He/She will need to be seen regularly by your family doctor for regular check-ups and vaccinations. Your child will also need to make several visits a year to see a hematologist or blood specialist.

*Supported in part by Projects # MCJ-481004 and # 2H46 MC00232-02 from the Maternal and Child Health Bureau (Title V, Social Security Act).*

*Adapted from materials by the Texas Department of Public Health Newborn Screening Program.*