

# Splenic Sequestration in Sickle Cell Disease

## **WHAT IS THE SPLEEN AND WHAT DOES IT DO?**

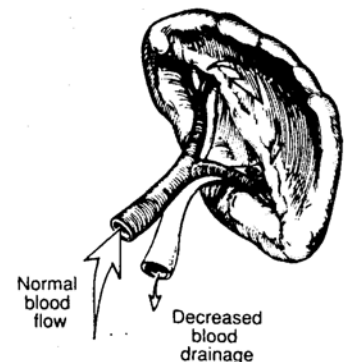
The spleen is normally a small organ located on the upper left side of the abdomen under the rib cage. It functions as part of the body's defense system that fights infection by removing bacteria (germs) from the blood. Basically, it serves as a filter in the bloodstream.

In sickle cell disease the spleen usually does not work after 4-6 months of age. It has been damaged by the sickled red blood cells (RBC's) and is not able to remove bacteria from the blood. This means that bacteria can grow in the blood and cause septicemia or blood poisoning (infection of the blood).

Some children with sickle cell anemia normally have enlarged (big) spleen. This is usually seen up to age 5 years. Spleen enlargement is rare after age 5 - by this age, the spleen is usually shriveled up and therefore you are not able to feel it. Children with Hemoglobin Sickle C disease (SC disease - another form of sickle cell disease) may have a big spleen, but this does not happen until they are about 4 years of age or older. Also, the spleen functions more normally in Sickle C disease.

## **WHAT IS SPLENIC SEQUESTRATION? (SPLEEN CRISIS)**

When sickled cells block the blood vessels leading out of the spleen, blood stays in the spleen instead of flowing through it. This causes the spleen to get bigger. When this happens the blood count (hemoglobin and hematocrit) falls and the spleen gets very large and easy to feel. This is called splenic sequestration crisis (or "spleen crisis"). Splenic sequestration can sometimes be painful.



## **WHO CAN GET SPLENIC SEQUESTRATION?**

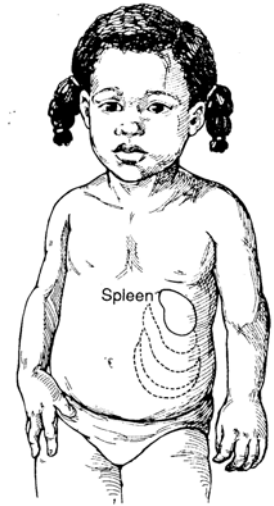
Infants and young children with sickle cell anemia who are between the ages of 2 months and 4 years are at greatest risk of splenic sequestration and blood poisoning. Sequestration crisis can occur in older children with SC disease and Sickle Beta-Plus Thalassemia. In these cases, the spleen remains enlarged or has the capability to enlarge.

## WHAT ARE THE SYMPTOMS OF SPLENIC SEQUESTRATION?

Your child may experience any of the following symptoms:

- Weakness
- Irritability
- Unusual sleepiness
- Paleness
- Big spleen
- Fast heart beat
- Pain in the left side of the abdomen

*A child can have a seriously low blood count without many symptoms. Sometimes the only symptom is that he/she is not as active as usual.*



## IS SPLENIC SEQUESTRATION SERIOUS?

Any enlargement of the spleen must be monitored.

Parents should be taught how to feel for their child's spleen at their regular check-up visit. They need to know how their child's spleen usually feels so that when he/she seems sick, they can feel the spleen to see if it is bigger.

Acute splenic sequestration crisis can be serious and a potentially life-threatening problem if the spleen suddenly enlarges with a large drop in the blood count. The child needs to see a doctor immediately.

When the spleen gradually gets bigger over several weeks, the blood count does not change much and therefore it may not be as serious.

## WHAT IS THE TREATMENT FOR SPLENIC SEQUESTRATION?

A blood transfusion is given if the blood count is dangerously low.

Minor episodes of splenic sequestration are common. You will notice moderate increases in spleen size associated with a decrease in hemoglobin levels. These minor episodes usually resolve spontaneously, but require monitoring of spleen size and blood counts.

If a child experiences several episodes of splenic sequestration, surgery to remove the spleen may be considered.

## WHAT CAN BE DONE TO HELP CHILDREN WITH SICKLE CELL ANEMIA FIGHT INFECTION SINCE THE SPLEEN DOES NOT WORK?

Prevention and early treatment of infection is the best defense against serious complications. The child who is sick should be carefully watched for symptoms of serious infection. A fever of 101 degrees or higher should always be considered a symptom of possible septicemia or bacteria in the blood (see fever section). Penicillin is given twice daily to prevent infection. Pneumococcal vaccines (a shot) are given to boost immunity to harmful infection.

## CAN SPLENIC SEQUESTRATION HAPPEN MORE THAN ONCE?

Yes. An infant or child that has had one episode of splenic sequestration is likely to have other episodes.