

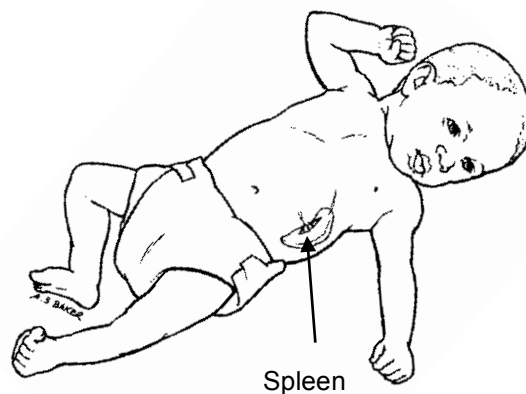
## Sickle Cell Disease and “Spleen Crisis” (Splenic Sequestration)

Sickle cell disease is an inherited blood disorder. Children are born with the condition. It affects a part of the red blood cell called **hemoglobin**. Hemoglobin is the part of the red blood cell that carries oxygen to different parts of the body.

A person with sickle cell disease makes a different kind of hemoglobin called **sickle** hemoglobin. Instead of being round and smooth, cells with sickle hemoglobin are shaped like a banana or a sickle. They become hard and sticky and have trouble moving through small blood vessels. Sometimes they clog up these blood vessels. This keeps the blood from bringing oxygen to the tissues. It can cause pain or damage to the areas that are not getting oxygen.

### The spleen

The spleen is a normally small organ located on the upper left side of the abdomen (belly) under the ribs (**Picture 1**). It helps to fight infection by filtering germs out of the blood. The spleen usually does not work well after 4 to 6 months of age in children who have sickle cell disease. This makes children with sickle cell disease more likely to get infections that can make them very sick. (See Helping Hand HH-I-221, *Sickle Cell Disease and Fever*.)



**Picture 1** Check your child's spleen if you see signs of “spleen crisis.”

### Spleen crisis (Splenic Sequestration)

Sickle cells can block the blood vessels leading out of the spleen. When this happens, blood stays in the spleen instead of flowing through it. This causes the spleen to get bigger, and the blood counts to fall. This is called a splenic sequestration (SPLEN ik seh ques TRAY shun) crisis, or simply “spleen crisis.”

**A spleen crisis can be life threatening if not treated right away!**

## Checking your child's spleen

Some children with sickle cell disease normally have a big spleen. It is very important to know if your child's spleen is normally big, and how big it usually is. If you are not sure if your child's spleen is usually big or not, ask your child's doctor or nurse practitioner at your child's next clinic visit.

## Signs of a spleen crisis

If your child's spleen is larger than usual, call the Hematology Clinic or the hematologist on call right away. You need to check your child's spleen size if your child has any of the signs of a **spleen crisis listed below**.

- pale skin
- fever
- complains of abdominal pain (usually on the left side)
- more tired than usual
- unusually fussy or irritable

## In an emergency call 9-1-1

**The following situations are emergencies. Call 911 if your child:**

- is unresponsive
- is unable to wake after a nap
- suddenly feels weak or loses feeling in a body part
- has trouble breathing
- is unable to move a body part

## How to contact the sickle cell program

The phone number for the Sickle Cell Clinic at Nationwide Children's Hospital is (614) 722-3250. If your child becomes ill during the day, call the Sickle Cell Clinic for treatment recommendations.

On evenings and weekends, call (614) 722-2000 and ask for the hematologist on call. The Sickle Cell nurse is available to answer non-urgent questions between 8:00 a.m. to 4:30 p.m., Monday through Friday at (614) 722- 8914.