

Sickle cell disease (SCD): splenic sequestration crisis

What is a splenic sequestration crisis?

Splenic sequestration crisis is a problem that is sometimes seen in children who have sickle cell disease. The name means:

- Splenic – the problem occurs in the spleen.
- Sequestration – the blood in the spleen is separated from the rest of the blood in the body.
- Crisis – the problem is an emergency

The spleen is an organ to the left and slightly above the stomach. It is protected by the ribs. The spleen helps to:

- Fight infection.
- Make and store red blood cells (RBCs).
- Clean the blood.

Splenic sequestration crisis:

- Can happen as early as 6 to 9 months of age.
- Is often seen before 5 years of age.
- Can occur in older children with hemoglobin SC disease and sickle beta+ thalassemia.

What happens with splenic sequestration crisis?

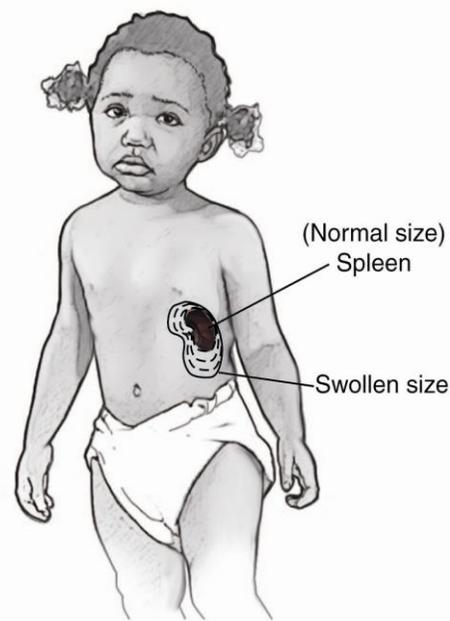
With splenic sequestration crisis, the spleen suddenly gets swollen and big because:

- The spleen is the first organ injured by sickle cells in the blood. The sickle red blood cells block the blood vessels inside the spleen. This prevents the blood from leaving the spleen.
- When this happens, a large amount of blood gets trapped in the spleen. This causes the spleen to get swollen.
- Since the blood cannot get back into the circulation, the hemoglobin level (RBC count) can go down to very low levels.
- There may not be enough blood left in the circulation for vital organs, like the heart and lungs, to work as they should.
- **This can lead to shock and is a medical emergency.**
- This is one of the most common causes of death in children with sickle cell disease.

What are the symptoms?

When the crisis occurs, the spleen gets very big and easy to feel. Your child's care team will teach you how to feel for your child's spleen. When you feel a big spleen, get help for your child **right away**.

Splenic Sequestration



In case of an urgent concern or emergency, call 911 or go to the nearest emergency department right away.

SCD: splenic sequestration crisis, continued

Your child may also have 1 or more of these:

- Pale color of the lips, gums or nails
- Fast breathing and heartbeat
- Weakness
- Pain in the area around the spleen
- Feel more tired than normal
- Not want to play or take part in normal activity
- Sleeping longer
- Not feeding or eating like normal

NOTE: Fever or infection can sometimes happen along with these other symptoms. Your child may or may not have fever when their spleen is swollen.

What should I do if my child has symptoms of a crisis?

If your child has any of these symptoms:

- **Get medical help right away.** Your child needs treatment as soon as possible.
- Call the sickle cell clinic or go to the emergency department (ED).

Call 911 right away if your child is:

- Very limp or pale
- Having trouble breathing
- Not responding well

What tests could my child have?

Your child will need many of these tests:

- A physical exam
- Blood tests (labs), including blood counts
- A blood type and crossmatch in case your child needs a blood transfusion
- An ultrasound test to see how big the spleen is or if there are any other problems

What is the treatment?

The spleen can get big very quickly. This can cause serious problems, so a child with splenic sequestration crisis most often needs treatment in the hospital. Sometimes a splenic sequestration crisis may not be as bad and can get better without treatment.

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SCD: splenic sequestration crisis, continued

If your child needs treatment, it may include:

- Intravenous (I.V.) fluids.
- Blood transfusion if the hemoglobin (RBC count) is very low or if your child has symptoms from the low RBC count.
- Frequent RBC count checks to make sure it does not drop too low. RBC counts will also help check the response to a blood transfusion.
- Treatment with oxygen if oxygen levels are low.
- Frequent checks of the spleen size to make sure it is not getting bigger.
- Antibiotics to help kill germs if needed.

What follow-up care does my child need?

Your child will need frequent clinic visits to check their RBC count and spleen size. Your child's care team will teach you how to feel for the spleen.

After going home from the ED:

Your child may be able to go home if their RBC count is not too low and you know how to feel for the spleen.

- Feel the spleen as you have been taught. Watch closely for any increase in size.
- **Call the sickle cell clinic or return to the ED right away if you feel the spleen getting bigger.**
- Follow-up in the clinic within 1 or 2 days, or as your doctor directs. They will check your child's RBC count and spleen size.

After going home from the hospital:

After your child goes home, take them to their follow-up clinic visit as advised. The follow-up may include:

- A physical exam
- Blood tests (labs)
- Teaching you how to feel for your child's spleen if you do not know how
- A blood transfusion on a regular schedule if your child had severe (very bad) splenic sequestration. This can help prevent a crisis from happening again.

Call the sickle cell clinic or return to the ED right away if you feel the spleen getting bigger.

If your child's spleen stays swollen:

- This is called chronic splenomegaly (enlarged spleen).
- You may need to limit your child's activity to help prevent injury to the spleen. Your child should especially avoid contact sports like football.
- Your child may need a blood transfusion if their spleen gets bigger and their hemoglobin drops too low.

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SCD: splenic sequestration crisis, continued

If your child had severe splenic sequestration crisis or has had it more than 2 times, they may also need:

- Surgery to remove the spleen. This may help to stop it from happening again.
- Blood transfusions on a regular schedule if your child is younger than 2 years old. Children at this age are not old enough to have surgery because they have a higher risk of infection.

How can I help decrease the chance of another splenic sequestration crisis?

A splenic sequestration crisis can happen more than 1 time. A child who has it 1 time is likely to have it again. You can help decrease the chance of it happening when you:

- Keep all of your child's follow-up visits in the sickle cell clinic. This is very important.
- Follow the sickle cell provider's advice for your child's treatment. This includes making sure they get blood transfusions when needed.
- Learn how to feel for your child's spleen so you can tell right away if it is getting bigger.

What should I do if my child is sick?

- Follow your child's sickle cell provider's advice for what you should do.
- Call your child's provider and **get medical care right away** anytime your child has a fever of 101°F (38.3°C) or higher.
- During the time when the sickle cell clinic is closed (between 5 p.m. and 8 a.m. during the week and on weekends and holidays), call the hematologist on call before going to the hospital. They will call the emergency department (ED) and help with your child's care.
- **ALWAYS** tell the ED doctors and staff that your child has sickle cell disease. Also tell them which sickle cell clinic they visit.

For more details on sickle cell disease and services, visit the Children's Healthcare of Atlanta website at choa.org/sicklecell.

This teaching sheet contains general information only. Talk with your child's doctor or a member of your child's healthcare team about specific care of your child.

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