Sickle Cell Disease Splenic Sequestration Crisis

Children's Healthcare of Atlanta
Dedicated to All Better

Patient and Family Education

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.

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:

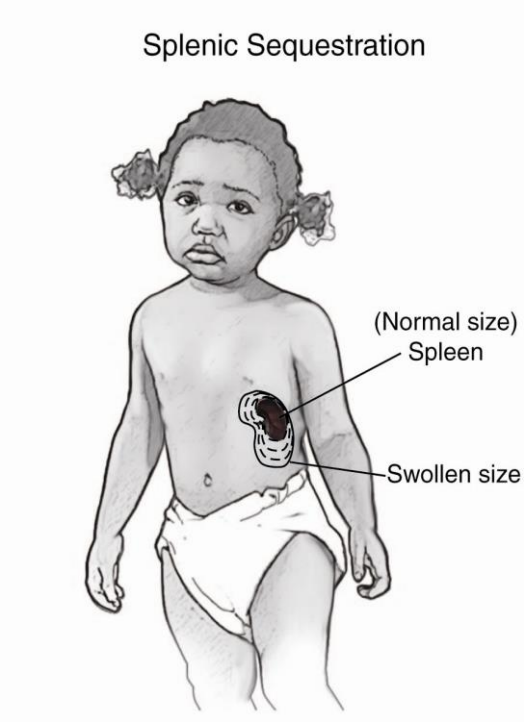
- Splenic – the problem occurs in the spleen
- Sequestration – blood in the spleen is separated from the rest of the blood in the body
- Crisis – the problem is an emergency

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
- 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 sickle red blood cells block the blood vessels inside the spleen. This prevents the blood from leaving the spleen.
- Once this happens, a large amount of blood gets trapped in the spleen. This causes the spleen to get quite swollen.
- Since the blood cannot get back into the circulation, the hemoglobin level (red blood cell count) can fall to very low levels.
- There may not be enough blood left in the circulation for vital organs, like the heart and lungs, to work normally.
- **This can lead to shock and is an emergency.**

What are the symptoms of a splenic sequestration crisis?

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

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

Your child may also have one or more of these:
- Pale color of the lips, gums or nail beds
- Fast breathing and heartbeat
- Weakness
- Pain in the area around the spleen
- More tired than usual, not wanting to play or take part in normal activity, or sleeping for longer periods of time
- Not feeding or eating normally

NOTE: Fever or infection can sometimes occur along with these other symptoms, but fever is not always present. Your child may or may not have fever when his spleen is swollen.

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

If your child has any of the above symptoms:
- **Get medical help right away**. Your child needs to be treated as soon as possible.
- Call the sickle cell clinic or go to the Emergency Room.

Call 911 right away if your child is:

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

What tests are needed to check for splenic sequestration crisis?

Your child will need many of these tests:
- A physical exam
- Blood tests, including a blood count
- A blood type and crossmatch, in case a transfusion is needed
- An ultrasound test to see how big the spleen is or if there are any other problems

How is a splenic sequestration crisis treated?

The spleen can get big very quickly. This can cause serious problems, so a child with splenic sequestration crisis is usually treated in the hospital. Sometimes splenic sequestration may not be as severe and can get better without any treatment. If your child needs treatment, it may include:
- Intravenous (IV) fluids
- Blood transfusion if the hemoglobin (red cell count) is very low or if your child has symptoms from the low blood count
- Blood count checks to make sure it does not drop too low. If it does, he may need 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 is needed?

Your child will need frequent visits to the clinic to check his blood count and size of his spleen. We will teach you how to feel for the spleen.

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

Your child goes home from the emergency room:
If the blood count is not too low and you know how to feel for the spleen, you may be sent home.

- Feel the spleen as you have been taught. Watch closely for any increase in size.
- **Call the clinic or return to the Emergency Department right away if you feel the spleen getting bigger.**
- Follow-up in the clinic within a day or two, or as your doctor directs. Your child will need his blood count and spleen size checked.

After your child goes home from the hospital:
Once your child goes home, bring him back to the clinic for a check-up as advised. The check-up may include:

- A physical exam
- Blood tests
- Teaching you how to feel for your child's spleen if you do not know how
- If your child had a severe case, he may need a blood transfusion on a regular schedule. This can help prevent a crisis from happening again.

Call the clinic or return to the Emergency Department 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 prevent injury to the spleen. This is very true for contact sports like football.
- He may need a blood transfusion if his spleen gets bigger and his hemoglobin drops too low.

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

- Surgery to remove the spleen. This may help to stop it from happening again.
- Blood transfusions on a regular basis if your child is less than 2 years old. Children this young are not yet old enough to have the surgery.

How can I decrease the chance of splenic sequestration crisis from happening again?

Splenic sequestration can happen more than once. A child who has it one 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 appointments in the sickle cell clinic. This is very important.
- Follow your sickle cell provider's advice for your child's treatment. This includes having blood transfusions when they are needed.
- Learn how to feel for your child's spleen so you can tell right away that it is getting bigger.

What should I do if my child is sick?

- Follow your child's doctor's advice for what you should do.
- Call your child's healthcare provider anytime that your child has a fever of 101°F (38.3°C) or higher and is sick. Your child will need to be seen **right away.**

- During the time when the clinic is closed (between 5PM and 8AM during the week, on weekends and holidays), call the hematologist on call. They will notify the Emergency Room and help with your child's care.
- **ALWAYS** tell the Emergency Room doctors and staff that your child has sickle cell disease. Also tell them which sickle cell clinic he visits.

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