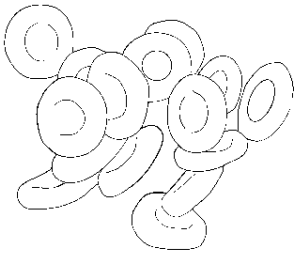
Sickle cell disease (SCD)

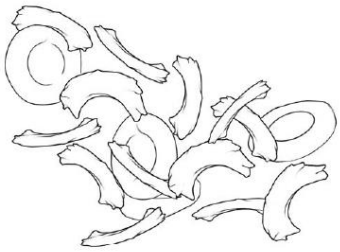
What is sickle cell disease (SCD)?

Sickle cell disease is a blood disorder that affects red blood cells (RBCs) in the body.

- RBCs contain a protein called hemoglobin. Hemoglobin allows the RBCs to carry oxygen from the lungs to all parts of the body.
- Most people have the same type of hemoglobin. It is made up of a certain pattern of building blocks.
- The RBCs of a person with sickle cell disease contain sickle hemoglobin. It is made up of a different pattern of building blocks.



Normal red blood cells are round and smooth



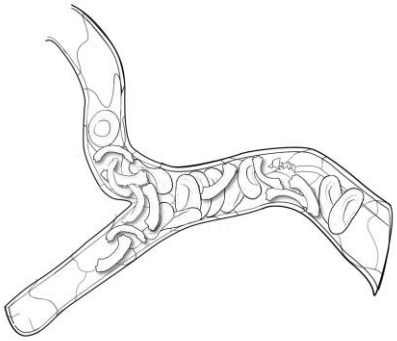
Sickle red blood cells look like a banana or sickle

Why are sickle cells a problem?

Normal RBCs are round and smooth. They flow through blood vessels very easily.

RBCs that carry sickle hemoglobin:

- Change their shape after they deliver oxygen to the body. They look like a banana or a sickle. This is how the disease got its name.
- Sickle cells are hard and sticky. This is because the sickle hemoglobin sticks together inside the blood cell.
- The sickle shape makes it harder for the sickle cells to flow smoothly through the blood vessels.
- The sickle cells may block blood flow to parts of the body.
- Areas of the body that do not receive normal blood flow can become damaged. This is what causes many of the medical problems that people with sickle cell disease have.



Sickle red blood cells stick together and block blood vessels

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

Some of the problems from blocked blood flow happen quickly, such as sudden pain. Other problems happen more slowly and can damage the body's tissues and organs over time.

Another problem with sickle cells is that they do not live as long as normal RBCs.

- Normal RBCs live about 120 days (4 months). Sickle cells live about 16 days (2 to 3 weeks).
- Because of this, sickle cell disease causes anemia. Anemia means a low RBC count or low hemoglobin level.

What symptoms could my child have?

Babies are born with normal RBCs. In a child with sickle cell disease, symptoms start only after enough sickle cells are made. Most often, this takes about 4 to 6 months.

We do not know when or why some children have certain symptoms.

- Sickle cell symptoms change from child to child and from mild to serious.
- Some symptoms tend to stay once they happen. Other symptoms come and go.

Your child's sickle cell provider will talk with you about any problems your child may have.

Sickle cells break down quickly.

Your child may feel tired, feel weak or have low energy. When the cells break open, they release a yellow substance called bilirubin into the blood. This can cause:

- Jaundice – a yellow color of the eyes and skin
- Gallstones – solid clumps in the gallbladder

Blocked blood vessels can cause pain.

Pain happens when sickle cells block the blood vessels.

- This can happen anywhere in the body. It often happens in the arms, legs, chest, back and stomach area.
- This is called a vaso-occlusive episode (VOE) and used to be called a pain crisis. These may last from hours to days. It can happen again in the future.

For severe (very bad) pain, your child may need to be stay in the hospital to get stronger pain medicines.

Blocked blood vessels can cause problems in the body's organs.

Sickle cells can block blood vessels inside organs, such as the spleen, kidneys, brain and lungs.

- When blood vessels in the spleen are blocked, blood fills the spleen instead of flowing through it. This causes the spleen to become very large. This is called splenic sequestration.
- This is a serious problem for children with sickle cell disease and is a medical emergency.

Infections are more common for children with sickle cell disease.

- Infections can happen more often and can be more severe, especially those caused by certain germs.
- Your child may have a fever or other problems. This depends on where the infection is in the body.
- The chance of getting an infection never goes away completely. Most often, it becomes less common as children get older.

Your child's sickle cell provider can talk with you about:

- The type of sickle cell disease your child has.
- Their plan of care.
- Tests, medicines and treatments they need.

Please feel free to ask us any questions you have about your child's care.

What tests could my child have?

All children born in the United States are screened for sickle cell disease at birth. If you do not have your child's test results, check with their primary care doctor (pediatrician).

- If your baby's newborn screening shows sickle cell disease, they will need more blood tests (labs) to make sure.
- If you cannot find out the results of your baby's screening, they may need other tests. One of these tests is called **hemoglobin electrophoresis**.

How can I help my child?

Your child needs your help from the time they are born. Some tips about how you can help your child are listed below.

Keep your child healthy.

- Take your child for regular visits with their primary care doctor and their sickle cell provider.
- Make sure your child gets vaccines to help prevent disease.
- Know and understand the results of your child's blood tests and other tests.
- Learn about sickle cell disease symptoms, treatment and any other problems your child may have.
- Ask your child's doctor about treatments for sickle cell disease.
- Teach your child healthy habits and good nutrition. Make sure they drink lots of fluid.

Help prevent infections.

- Clean your hands often. Wash them well with soap and water for at least 20 seconds, or use an alcohol-based gel or foam. Teach your child and anyone who cares for them to clean their hands often, too.
- Give your child penicillin each day as their doctor directs. Penicillin is an antibiotic medicine used to help prevent or treat infections.

- Dress your child properly for the weather that day. Avoid exposing them to severe weather changes.

Build a good family support.

- Learn how to cope and manage stress. Be a role model for your child.
- Teach other family members and close friends about sickle cell disease and your child's needs. They may be a source of support and help in times of need.
- Find other sources of support you can use through your employer or community agencies.

Will my child need special care?

Your child will need regular care from their primary care doctor and special care from a team of sickle cell providers.

- This may include:
  - Exams, blood tests and X-rays on a regular basis to help prevent or check for problems.
  - Special medicines to help prevent or treat problems.
  - Vaccines to help prevent certain serious infections.
  - Other special treatments based on your child's needs.
- Your child may go to any of the Children's sickle cell clinics and hospitals.

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 <u>right away</u>** any time your child has a fever of 101˚F or higher.
- During the time when the 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.

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