

TAKING CONTROL:
Teens with
Sickle Cell Disease



St. Jude Children's
Research Hospital

Finding cures. Saving children.

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This document is not intended to take the place of the care and attention of your personal doctor. Our aim is to promote active participation in your care and treatment by providing information and education. Questions about individual health concerns or specific treatment options should be discussed with your doctor.

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Welcome to the St. Jude Teen Clinic

The teen clinic treats teens from 12 to 18 years old, who have sickle cell disease. Our goal is to prepare you for transition to adult care by 18 years of age. In the Merriam Webster Dictionary, the definition of transition is “a passage of one state, stage, subject, or place to another: CHANGE.”

In the St. Jude Hematology clinic, transition means changing from your pediatric (childhood) doctor to an adult doctor. In the teen clinic, preparing for transition is an ongoing process.

As a person with sickle cell disease, one of your greatest challenges will be achieving self-management. To successfully make the transition to adult care you and your health care team must work as partners. Self-management will help you prepare for transition.

The St. Jude Hematology staff created this booklet to offer complete information and easy to follow guidelines. This process is interactive, and you are a partner. Please discuss the information in each unit with your health care team.

Taking Control: Teens with Sickle Cell Disease offers many details that are crucial to your health. For this reason, it is best to read only one unit of the booklet at a time. If you forget to ask a question during your clinic visit, write it down in the “Notes” section, and bring the booklet to your next visit.

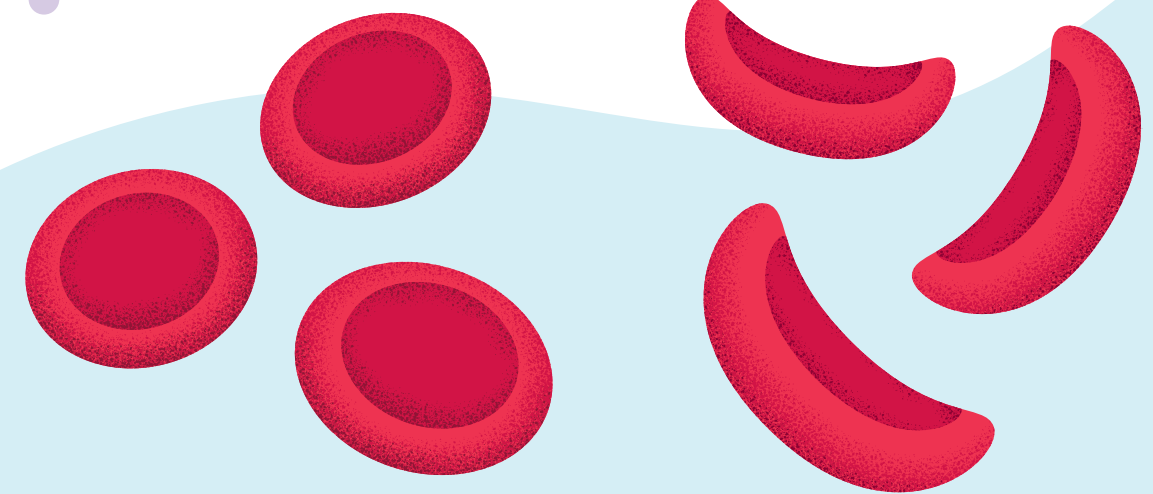
UNIT 1: Overview of Sickle Cell Disease

What is sickle cell disease?

Sickle cell disease is a blood disorder that affects **red blood cells** (RBC). The main purpose of red blood cells is to deliver oxygen to the body. Blood delivers oxygen to all of the tissues in the body.

The part of the blood that carries oxygen is called hemoglobin. Sickle cell disease is a blood disorder that affects the **hemoglobin** (HEE-muh-glow-bin) within the red blood cells. The main role of hemoglobin is to deliver oxygen to the cells of the body.

People with sickle cell disease have red blood cells that contain mostly hemoglobin S, an abnormal type of hemoglobin. Sometimes these red blood cells become **sickle (banana) shaped**, and have trouble passing through small blood vessels.



Normal Red Blood Cell

Sickle-shaped Red Blood Cell

Normal red blood cells are shaped like discs and are flexible. They flow easily through the body's blood vessels.

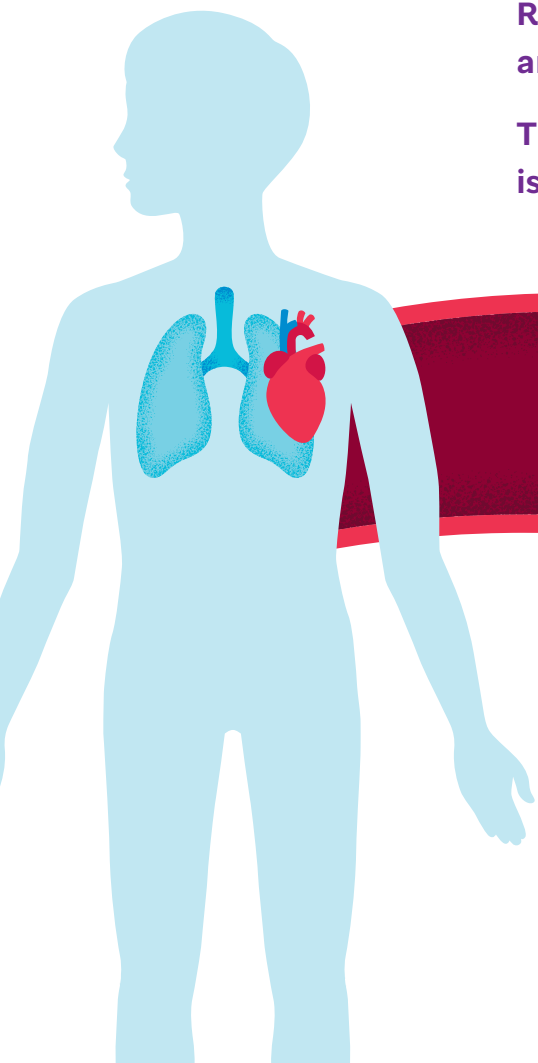
Sickle-shaped red blood cells are sticky, stiff, and rigid. They clog the body's small blood vessels.

When sickle-shaped cells block small blood vessels, less blood can reach that part of the body. Tissue that does not receive a normal blood flow eventually becomes damaged. This is what causes some of the complications of sickle cell disease.

Red Blood cells flow through the lungs and collect oxygen.

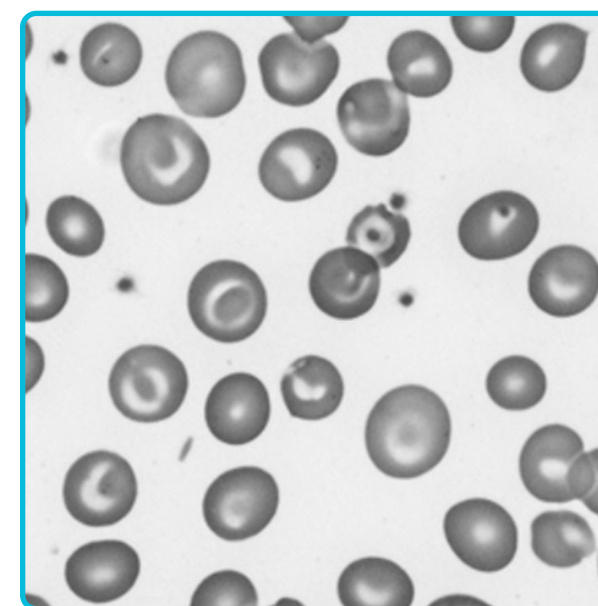
Then the blood travels to the heart and is pumped to the rest of the body.

Sickle-shaped red blood cells block small vessels and eventually the area becomes damaged.

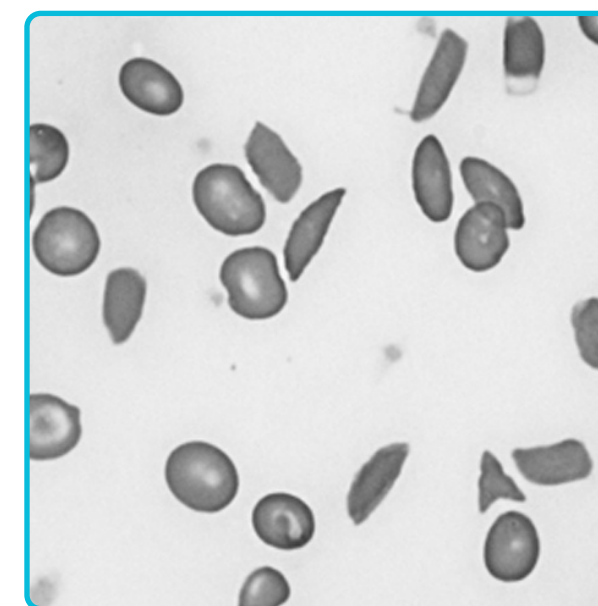


Why are people with sickle cell disease anemic?

This happens because their blood has less than the normal amount of red blood cells. Sickle cells do not survive as long as normal red blood cells. A normal red blood cell lives about 3 to 4 months. A sickle cell usually survives less than 20 days.



Slide of Normal Red Blood Cells



Slide from person with SCD - (anemic)

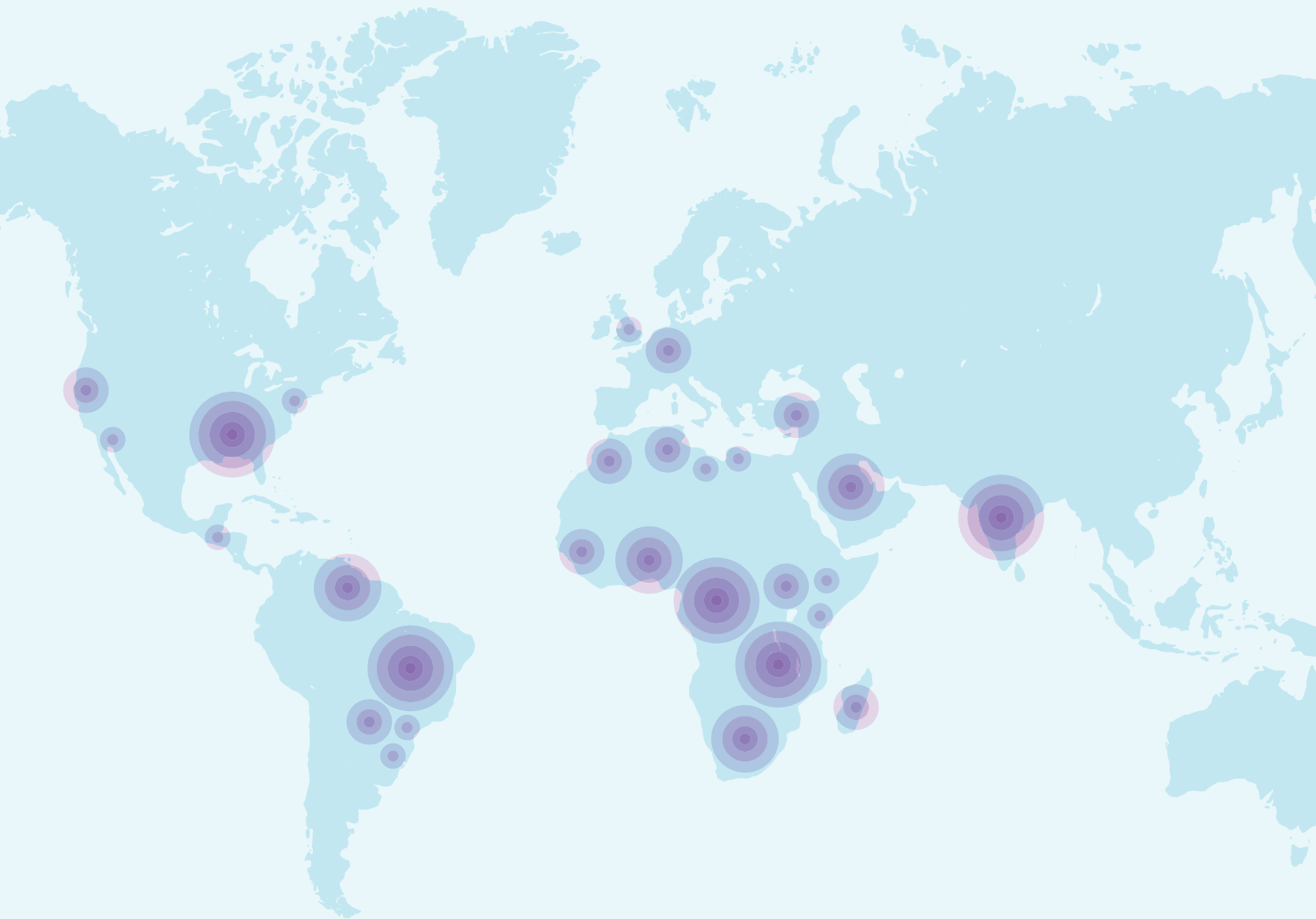
When the blood does not have enough red blood cells and hemoglobin, a person has **anemia**.

The hemoglobin level and the hematocrit are part of a lab test called the complete blood count (CBC). These levels are tested to see if a person is anemic. *To learn more, see "Do You Know...Blood Counts and Sickle Cell Disease."*

A normal hemoglobin level is about 11-15 g/dL (depending on your age and gender), and a normal hematocrit is about 34-42 percent. A person with a value that is below normal is said to be anemic. The anemia from sickle cell disease cannot be corrected by taking iron.

Who is affected by sickle cell disease?

About one of every 375 African-American babies is born with sickle cell disease. Sickle cell disease affects about 100,000 people in the United States and more than a million people worldwide. Around the world, the disease affects people of all races and is heavily concentrated in Central and South America, Middle Eastern, Mediterranean, India, Asia, and Africa.



Are there different types of sickle cell disease?

Yes, sickle cell disease is the name for a group of disorders. There are many different types of sickle cell disease. People are born with the disease. Sickle cell disease is inherited like hair color or eye color. Each person has two hemoglobin genes—one from the mother and one from the father. People with sickle cell disease receive a hemoglobin S gene from either one or both parents.

The three most common types of sickle cell disease in the United States:

Hemoglobin SS Disease

This is the most common type of sickle cell disease. People with sickle cell anemia have mostly hemoglobin S in their red blood cells. They do not have any normal hemoglobin A.

Sickle-Hemoglobin C Disease (Hemoglobin SC Disease)

People with sickle-hemoglobin C disease have both hemoglobin S and hemoglobin C in their red blood cells. Hemoglobin C is another type of abnormal hemoglobin. Normally hemoglobin C does not cause many problems, but when a person has hemoglobin C and hemoglobin S together, they do not have any normal hemoglobin and can have many medical complications.

Sickle Beta-Thalassemia Disease

There are two types of sickle beta thalassemia disease:

- **Sickle Beta Zero Thalassemia Disease**

People with sickle beta zero thalassemia disease have mostly hemoglobin S like a person with hemoglobin SS disease. The symptoms are the same as a person with hemoglobin SS disease. People with sickle beta zero thalassemia disease can have severe medical complications.

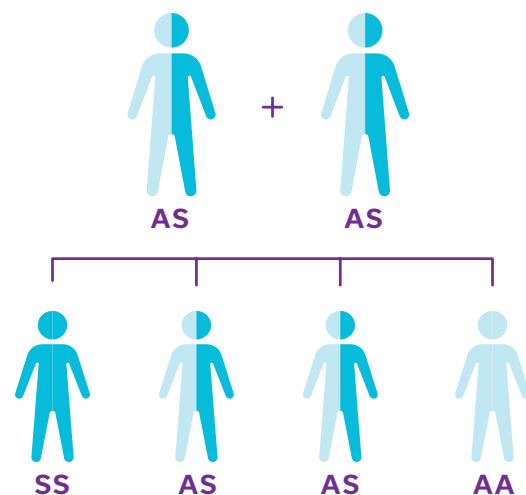
- **Sickle Beta Plus Thalassemia Disease**

People with sickle beta plus thalassemia disease have mostly hemoglobin S, but they also produce a very small amount of normal hemoglobin A. People with sickle beta plus thalassemia can have medical complications, but the hemoglobin A helps prevent some of the complications from sickle cell disease. For instance, people with sickle beta plus thalassemia normally do not have strokes as a complication of the disease.

Hemoglobin SS

Both parents must have sickle cell trait in order for a child to have hemoglobin SS disease.

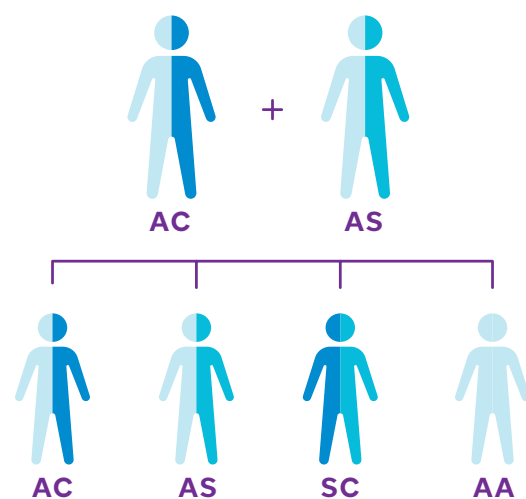
- AS** Sickle Cell Trait
- SS** Sickle Cell Disease
- AA** No Trait



Sickle-Hemoglobin C (Hemoglobin SC)

One parent must have sickle cell trait and the other parent must have Hemoglobin C trait for a child to have Hemoglobin SC disease.

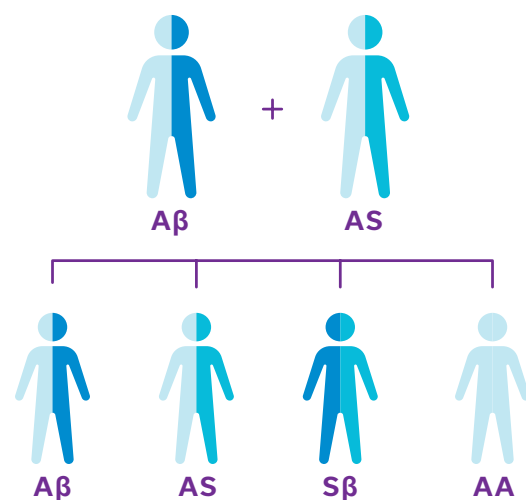
- AS** Sickle Cell Trait
- AC** Hemoglobin C Trait
- SC** Sickle Hemoglobin C Disease
- AA** No Trait



Sickle Beta-Thalassemia

One parent must have sickle cell trait and the other parent must have beta thalassemia trait in order to have a child with sickle beta thalassemia disease.

- AS** Sickle Cell Trait
- Aβ** Beta Thalassemia Trait
- Sβ** Sickle Beta Thalassemia Disease
- AA** No Trait



Unit 1 Summary

- People with sickle cell disease have red blood cells that contain mostly hemoglobin S (sickle hemoglobin). These red blood cells can become sickle (banana) shaped and block normal blood flow. Many of the complications of sickle cell disease are a result of the blood vessels getting blocked.
- Sickle cell disease is inherited. It affects about 100,000 people in the United States.
- The most common types of sickle cell disease are hemoglobin SS, hemoglobin SC, and hemoglobin Sβ thalassemia.

UNIT 2: Infection and Pain

What causes sickle cell pain?

Pain can have many different causes. The most common cause of pain in people with sickle cell disease is due to the sickling of red blood cells. When sickle cells clog a small blood vessel, less blood gets to the area supplied by that blood vessel. This area of the body does not receive enough oxygen, and this causes pain. Pain can occur anywhere in the body and often occurs in the arms, legs, chest, back, and abdomen.

Can anything be done to help prevent a pain episode?

Know your triggers and try to avoid them. For instance, you may notice you have a pain episode after being out in cold weather. Try to dress for the weather to avoid these pain episodes. These might be other triggers for you:

- **Lack of sleep**—Get at least 8 hours of sleep each night.
- **Not drinking enough fluids**—Drink at least 8 glasses of liquids each day (water is best). Avoid caffeine drinks such as energy drinks, cola, or coffee. Caffeine causes you to go to the bathroom more often, and it causes your blood vessels to constrict (become smaller).
- **Stress can cause a pain crisis**—plan ahead when you think an event might cause stress, and try to prevent as much stress as you can.
- **Avoid tobacco**—Do not smoke.
- **Avoid alcohol**—Do not drink wine, beer, nor liquor.



What is the best treatment for a pain episode?

Pain Score 0-2

For mild pain:

- Your child should take all daily medicines (such as hydroxyurea)
- Drink more water and other fluids
- Rest
- Apply heat to the painful area
- Massage the painful area
- Avoid painful triggers, such as very cold temperatures
- Try treating your pain without medicines, using techniques, such as distraction (with music, videogames, or drawing), relaxation, meditation, and sleep.

Pain Score 3-6

For moderate pain:

- Continue everything listed in the **Blue Zone**
- Start taking an anti-inflammatory medicine, such as ibuprofen (Motrin®, Advil®) and naproxen (Aleve®).
- If your pain is not improved in 12 to 24 hours of treatment with an anti-inflammatory, move to the **Red Zone**.

Pain Score greater than 7

For severe pain:

- Continue everything listed in the **Blue Zone** and **Purple Zone**
- Continue using an anti-inflammatory and start treatment with an opioid pain medicine (Tylenol with codeine, Lortab).
- To prevent constipation, start taking polyethylene glycol (Miralax®).

If pain has not improved in 24 to 48 hours, please contact St. Jude Hematology at 901-595-3300. If calling after 5 p.m., on a weeknight or on the weekend, ask for the hematologist on-call.

What else can I do when having sickle cell pain?

Other things to do in addition to pain medications, rest, water, and heat include:

- Relaxation technique:** Deep breathing or soothing music
- Distraction technique:** Singing, praying, or watching tv
- Thought stopping technique:** Speak positiveness to self

Are people with sickle cell disease more likely to get serious infections?

Yes. People with sickle cell disease have an increased risk of developing certain types of infections, especially in the blood, bone, and lungs. Fever is the most common symptom of an infection.

In people with sickle cell disease, the spleen does not work correctly. The spleen is an organ in the abdomen that helps protect against infection by filtering bacteria from the bloodstream and by producing antibodies. Early in life, sickle cells clog the blood vessels in the spleen leading to damage and poor protection against infection.

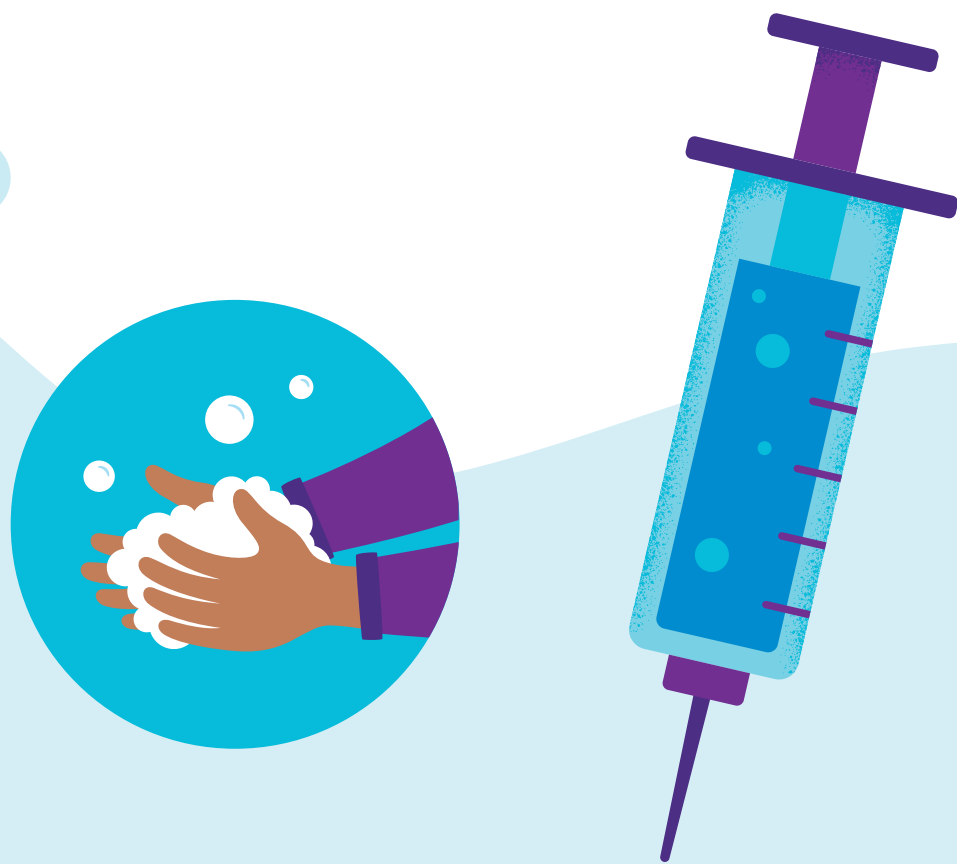
What are the symptoms of an infection?

Infection	Common symptoms
Blood	Fever (usually 100.4° F or higher)
Lung (Pneumonia or Acute Chest)	Fever, cough, chest pain, trouble breathing
Bone (Osteomyelitis)	Fever, swelling, pain, area warm to the touch

Can anything be done to prevent an infection?

Yes. The best prevention against infection is keeping your hands clean.

- Wash your hands with soap and water at least 20 seconds.
- Wash when preparing food and before eating.
- Wash after using the restroom, after sneezing or coughing, shaking hands, or touching things that might have germs, such as a baby's diaper.
- All of your childhood immunizations from your primary care physician should be up-to-date.
- Get a flu vaccine every year.
- Pneumococcal vaccines: Prevents pneumococcal sepsis and is given every 5 years
- Meningococcal vaccine: Prevents Meningitis and is given every 5 years.
- Other vaccines might be recommended by your medical team, such as hepatitis B, human papillomavirus (HPV), and coronavirus vaccine (COVID-19).



Unit 2 Summary

- Pain is the result of blockage of blood flow to the tissues. Various stresses to the body can trigger a pain episode. Good health practices can help prevent pain.
- Pain episodes can often be managed at home. But severe pain (unrelieved by medicines or with a fever of 100.4 degrees F or higher) always requires treatment by a doctor.
- Sickle cell disease causes an increased risk for certain types of infection because the spleen does not work correctly. The most common symptom of an infection is fever.

UNIT 3: General Health Issues

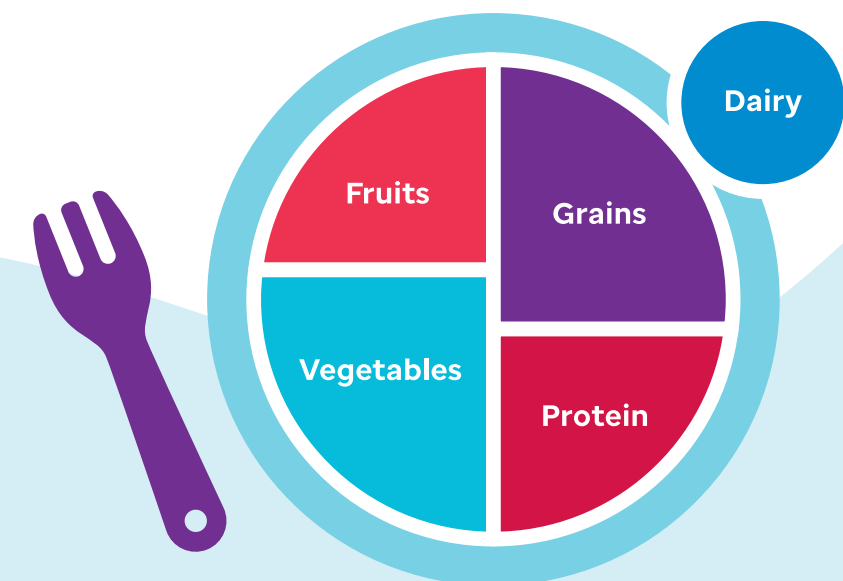
What are some general health issues with sickle cell disease?

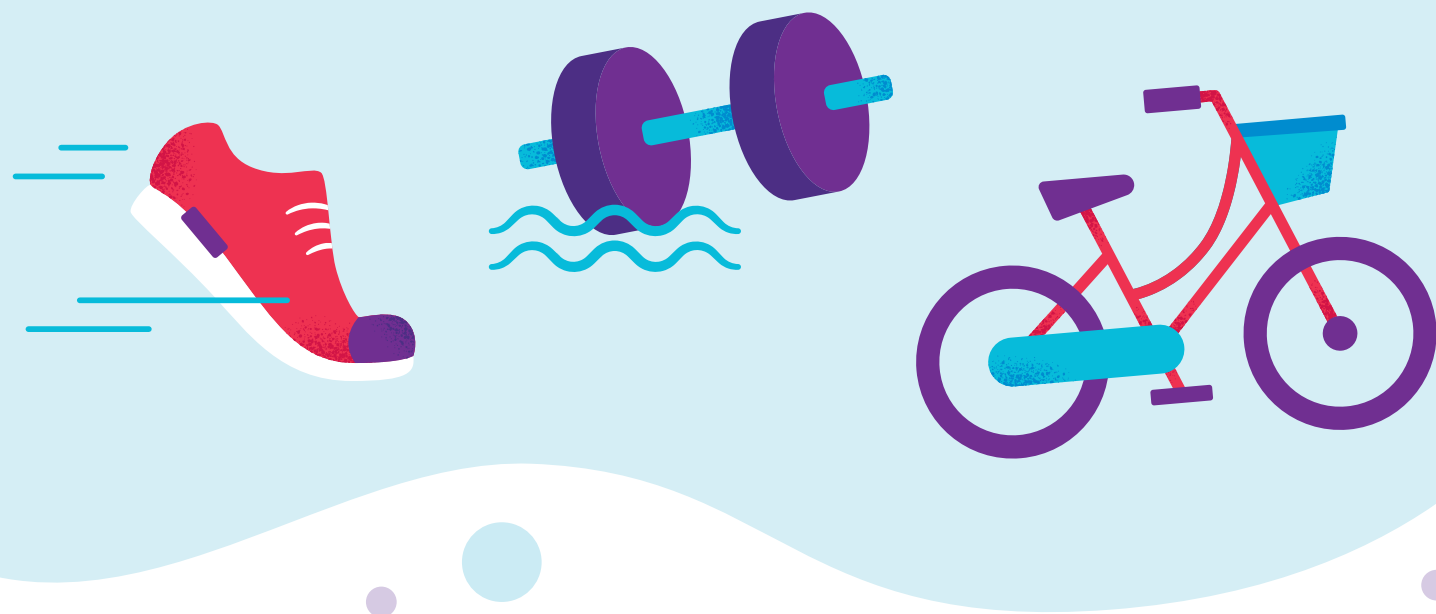
Is there a special diet for people with sickle cell disease?

No. There is no special diet, but proper nutrition is important for maintaining good health. People with sickle cell disease should eat a nutritious and well-balanced diet.

Water is very important for people with sickle cell disease. Drink at least 8-10 glasses per day and avoid sugary drinks and drinks with caffeine.

The USDA has food guidelines with recommendations for plate and portion sizes. In the diagram below, notice that one-half of the plate should be fruit and vegetables. Visit www.choosemyplate.gov to learn more.





Can a person with sickle cell disease take part in exercise?

Yes. A moderate exercise routine is recommended. Regular exercise promotes a healthy body and mind. A moderate exercise program 3 to 4 times a week is usually encouraged. However, the best type and amount of physical exercise is different for each person. Most people learn to set their own limits, based on experience.

When starting a new activity or exercise program, it is important to start slowly and gradually increase the time and effort spent. Rest and drink plenty of water while exercising. If you plan to take part in strenuous exercise or sports, talk to your doctor or nurse case manager first.

Is it normal for a person with sickle cell disease to feel sad and worried?

Yes. It is normal for anyone with a chronic illness to feel sad and worried at times. But if these feelings do not go away and prevent you from functioning normally or you have thoughts of harming yourself, you should talk to someone on your medical team.

Will taking pain medicine cause me to become addicted?

No. Addiction or drug dependency is rare. Strong pain medicines such as Tylenol® with codeine and morphine can cause physical and mental dependency, but this is rare when the medicines are used as ordered by your doctor. Therefore only take medications that are prescribed to you. Ibuprofen does not cause dependency and should be used to treat mild pain.

What about street drugs? Can they help with sickle cell pain?

No. Street drugs are harmful to everyone. Some street drugs can cause addiction and permanent damage to vital body organs. For people with sickle cell disease, even occasional use of street drugs can lead to severe sickle cell complications.

- Marijuana causes blood vessels to constrict (become smaller). Smaller blood vessels can lead to increased pain. Marijuana also has cancer-causing agents.
- Cocaine is a stimulant and can increase the heart rate, breathing rate, and blood pressure.
- Crack is a solid (rock) form of cocaine and causes the same symptoms as cocaine listed above.

Should a person with sickle cell disease smoke?

No. Smoking is harmful to everyone because of the cancer causing agents in cigarettes. But for people with sickle cell disease, it is even more harmful because nicotine causes the blood vessels to constrict (become smaller). This can cause increased pain crises. Also, nicotine results in reduced oxygen to the lungs. Since many people with sickle cell disease are anemic, this further depletes the supply of oxygen to the body.

Does sickle cell disease affect puberty?

Yes. Puberty is a time of rapid physical change. Growth and sexual development are more noticeable at this time than at any other period in your life.

Here are some of the ways boys and girls change at puberty:

- Growing several inches in height in a short period of time
- More changes of mood
- Growth of hair under the arms and in the genital area Breast development and the onset of menstruation in girls
- Lowering of the voice, growth of facial hair and growth of the penis and testes in boys

Many teens with sickle cell disease experience puberty later than others. The average delay is about 2 years.

Some people may have a delay in puberty but others may not. It is important to remember that even if it is late, puberty will still occur.

Can a person with sickle cell disease have children?

Yes. People with sickle cell disease can have children. They should be aware of the risk of having a child with sickle cell disease by knowing if their partner has a hemoglobin trait or disease.

Females with sickle cell disease should be treated by a high-risk OB-GYN doctor during pregnancy and should have pre-natal care throughout the pregnancy. A female with sickle cell disease has a higher risk for health problems during pregnancy.

Sickle cell disease does not cause any reproductive problems for males. However, males can have problems with painful erections (priapism). If an erection lasts more than 2 hours, seek medical attention.

See *Unit 4* for more information.

Unit 3 Summary

- A nutritious diet and a good fluid intake (8–10 glasses of liquid per day) are important for maintaining good health. A moderate exercise program, based on your own tolerance, is recommended.
- The use of alcohol, street drugs, and tobacco can greatly increase a person's risk of developing serious complications of sickle cell disease.
- The onset of puberty is delayed for some people with sickle cell disease but will still occur.
- If you feel sad or worried and these feelings do not go away, please talk to your medical team about a referral to a counselor or other mental health professional.

UNIT 4:

Brief Review of Sickle Cell Disease Complications

What are some other complications of sickle cell disease?

Stroke

Stroke is caused by damage to the brain due to lack of oxygen supply. Symptoms can include (but are not limited to) paralysis in part of the body, changes in vision, seizures, and changes in speech. If you have any of these symptoms, you should seek medical attention right away. The incidence of having a stroke is more common with individuals who have hemoglobin SS disease and sickle beta zero thalassemia disease. If a person has a stroke, long-term treatment requires red blood cell transfusions about once a month to prevent a second stroke.

Avascular necrosis (AVN)

AVN is caused by a temporary or permanent loss of blood supply (oxygen) to the bone. Without oxygen, the bone tissue dies and can cause painful bone to bone contact. AVN normally occurs in the hip joint and the long bones between the shoulder (humerus) and the leg (femur), but can happen in any joint. This can result in the need for joint replacement.

Acute Chest Syndrome (ACS)

ACS is unique to people with sickle cell disease. It is pneumonia (an infiltrate on an X-ray) along with one or more of these symptoms: fever, chest pain, respiratory symptoms (coughing, wheezing, shortness of breath), and hypoxemia (reduced oxygen in the blood). This can be treated with oxygen, antibiotics, or blood transfusions depending on the symptoms.

Retinopathy

Retinopathy is caused by blockage of vessels in the eyes. The eye has very small blood vessels, and a blockage can cause eye damage and lead to blindness. Surgery may be required if the condition is severe. An eye exam is required every year or every 2 years (depending on the type of sickle cell disease) to ensure the eye is not damaged. *To learn more, see "Do You Know... Retinopathy and Sickle Cell Disease."*

Priapism

Priapism is a painful erection of the penis that lasts for an extended time caused by sickling of red blood cells. The erection is not related to sexual stimulus or arousal. Priapism can begin at a very young age. If it is not treated, it can cause impotency (not able to have an erection), infertility (not able to have children), scarring, or permanent damage to the penis. If an erection lasts more than 2 hours seek medical treatment.

Osteomyelitis

Osteomyelitis is an infection in the bone. In people with sickle cell disease, bacteria carried by the blood can settle in bone marrow and cause an infection. Certain types of bacteria are more likely to settle in the bone, such as salmonella. Osteomyelitis requires treatment with antibiotics that can last many weeks. If left untreated, the infection can damage the bone or spread throughout the body. Symptoms can include fever, swelling, soreness, and heat in the area. If you notice any of these symptoms, contact your doctor or nurse case manager.

Gallstones

Gallstones are hard rock-like deposits that form in the bile duct or in the gallbladder. In people with sickle cell disease, most gallstones are produced from excess bilirubin, which is caused by the constant breakdown of red blood cells. Biliary sludge is excess bile that settles in the duct. It can also lead to gallstones forming in people with sickle cell disease. Symptoms can include abdominal pain, fever, nausea, vomiting, dark urine, clay colored stools, and yellowing of the skin and eyes (jaundice). Gallstones can be treated with IV fluids, pain medicines, antibiotics (for fever), and possible removal of the gallbladder, depending on the symptoms, blood tests, physical exam, and ultrasound results.

Sickle Cell Nephropathy

Your kidneys normally remove waste from your blood. They have many small blood vessels that help do this. The waste becomes part of your urine (pee) and goes out of your body.

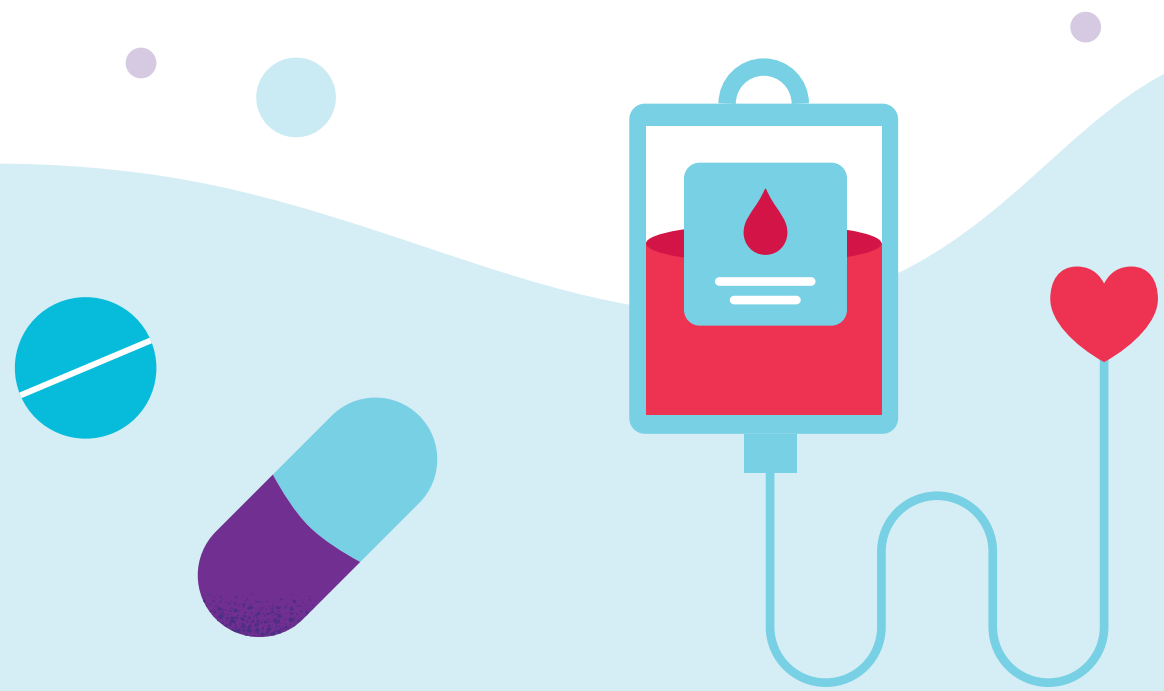
If sickle cell disease blocks normal blood flow to the kidneys, they do not get enough oxygen. This keeps them from working well. This is called kidney disease or “renal disease.” Another name for sickle cell kidney disease is “sickle cell nephropathy” (say “neff-RAW-puh-thee”).

To test for sickle cell nephropathy urine is collected to test. A laboratory tests the urine for protein, blood, and a substance called “creatinine” (say “kree-AT-uh-neen”). Too much of these substances can mean the kidneys are not working normally.

If sickle cell disease affects the kidneys, you could have some or all of the conditions below.

- Protein in the urine – Protein does not normally leave the blood through the kidneys. Having a lot of protein in the urine is a sign of sickle cell kidney disease.
- Urine with too much water in it – Normal urine has a certain amount of waste in it. Urine that is mostly water, with less waste than usual, could be a sign of sickle cell kidney disease.
- Blood in the urine - Your urine might look bloody or brown. The blood in your urine might only be visible under a microscope.
- High blood pressure - People with sickle cell usually have low blood pressure. Sickle cell kidney disease can cause high blood pressure.

Blood pressure medicines are the usual treatment. They help the kidneys stop losing protein your child’s body needs. They also lower your child’s blood pressure.



Unit 4 Summary

There are many complications of sickle cell disease. The most common complication is pain. Some other complications include the following:

- **Stroke**—damage to the brain
- **Avascular necrosis**—damage to the bone
- **Acute Chest Syndrome**—pneumonia plus other symptoms
- **Retinopathy**—damage to the eye
- **Priapism**—painful erections
- **Osteomyelitis**— an infection in the bone
- **Gallstones**—hard rock-like deposits that form in the gallbladder.
- **Sickle Nephropathy**—damage to the kidneys

UNIT 5: Common Treatment Options

What are some treatment options for sickle cell disease?

Sickle cell is a lifelong disease. Common treatment options for individuals with sickle cell disease include hydroxyurea, red blood cell transfusions, and bone marrow transplant.

Some medicines approved for sickle cell disease include Oxbryta, which helps increase hemoglobin levels. Adakveo is a medication that helps prevent pain, and Endari helps reduce pain crises.

Curative therapies under research include gene therapy, which uses the person's own treated cells. Another type of curative therapy is Haplo-identical bone marrow transplant, which uses a mother or father as a transplant donor. If these therapies are shown to be effective, they will give everyone the option for a curative therapy. This section will discuss hydroxyurea, red blood cell transfusions, and bone marrow transplant in more detail.

Hydroxyurea

Research studies show that hydroxyurea lowers the following:

- The number of acute chest syndrome (Pneumonia) events
- The number of pain crises
- The need for blood transfusions
- The number of trips to the hospital
- Hydroxyurea is given by mouth one (1) time each day. It comes in liquid or capsule form. To learn more, see the booklet "Hydroxyurea Treatment for Sickle Cell Disease."

Red Blood Cell Transfusions

Red blood cell transfusion—involves the transfer of red blood cells from one (1) person to another. Red blood cells are obtained when a person donates blood. Transfusions are given for different health problems caused by sickle cell disease. Sometimes only a single transfusion is needed. Other times, patients need long-term transfusions, which could mean receiving blood one (1) time a month. These are some reasons a person with sickle cell disease might need a blood transfusion:

- **Stroke:** When a stroke occurs, the brain suffers damage because blood flow is blocked to a portion of the brain. Chronic transfusions are used to prevent further strokes and brain damage, and they are usually given for the rest of a patient's life.
- **Acute chest syndrome (pneumonia):** When anemia is at its worse, breathing is hard and often the oxygen level in the body is lower than it should be. At these times, a transfusion may be needed.

Bone Marrow (Stem Cell) Transplant

In a person with sickle cell disease, the bone marrow produces red blood cells that contain hemoglobin S. This leads to the complications of sickle cell disease.

During a bone marrow transplant, the bone marrow of a person with sickle cell disease is replaced with blood-forming stem cells from a donor who does not have sickle cell disease. This can result in a cure for the disease.

The first bone marrow transplant on a person with sickle cell disease was performed in 1984 on a St. Jude Patient. Two major requirements must be met for a transplant to proceed.

- Identify the person who is the best match (donor).
- After the donor is chosen, both the donor and the patient will have pre-transplant evaluations of the heart, lungs, kidneys, etc.

These requirements limit bone marrow transplants for people with sickle cell disease because of the small chance of having a matched donor. To learn more about transplants, see the booklet “Bone Marrow (Stem Cell) Transplant for Sickle Cell Disease.”

Unit 5 Summary

These are the three main treatment options for sickle cell disease:

- Hydroxyurea:
reduces pain and acute chest syndrome
- Red Blood Cell Transfusions:
can be one time or long-term
- Bone Marrow Transplant:
A cure for sickle cell disease



**St. Jude Children's
Research Hospital**

Finding cures. Saving children.

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For general information on
sickle cell disease, visit our website:

www.stjude.org/sicklecell