St. Jude Children's Research Hospital
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Finding cures. Saving children.

Taking Control: Teens with Sickle Cell Disease

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Contents

Welcome to 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.

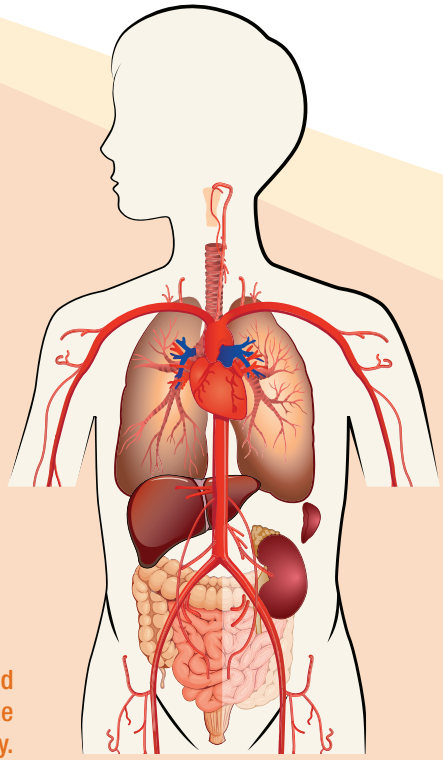
Taking Control: Teens with Sickle Cell Disease offers many details that are crucial to your health. For this reason, it is best to read only one unit of the booklet at a time. If you forget to ask a question during your clinic visit, write it down in the "Notes" section, and bring the booklet to your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

Unit 1: Overview of Sickle Cell Disease

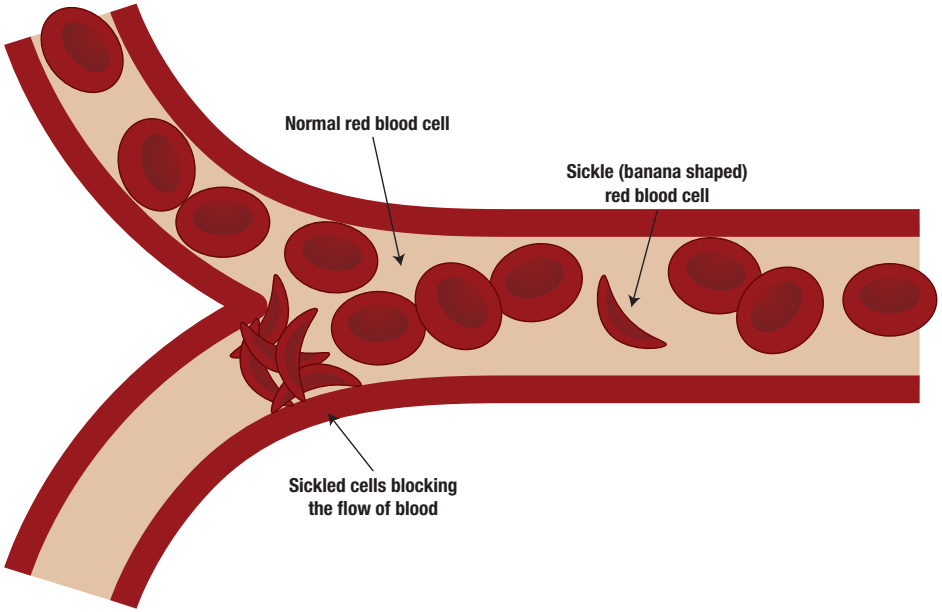
What is sickle cell disease?

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.



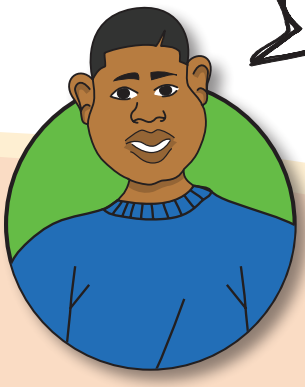
Red Blood cells flow through the lungs and collect oxygen. Then the blood travels to the heart and is pumped to the rest of the body.



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.

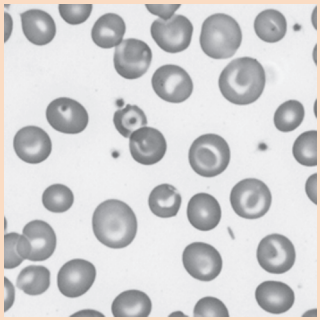
When sickle-shaped cells block small blood vessels, less blood can reach that part of the body. Tissue that does not receive a normal blood flow eventually becomes damaged. This is what causes some of the complications of sickle cell disease.

Sickle-shaped red blood cells block small vessels → Less blood reaches area → Area is damaged!

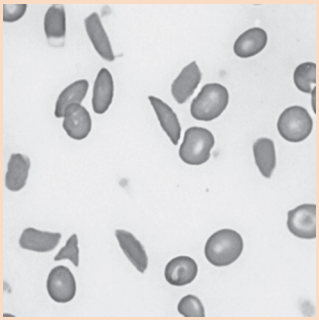


This happens because their blood has less than the normal amount of red blood cells. Sickle cells do not survive as long as normal red blood cells. A normal red blood cell lives about 3 to 4 months. A sickle cell usually survives less than 20 days. When the blood does not have enough red cells and hemoglobin, 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 11–14 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.



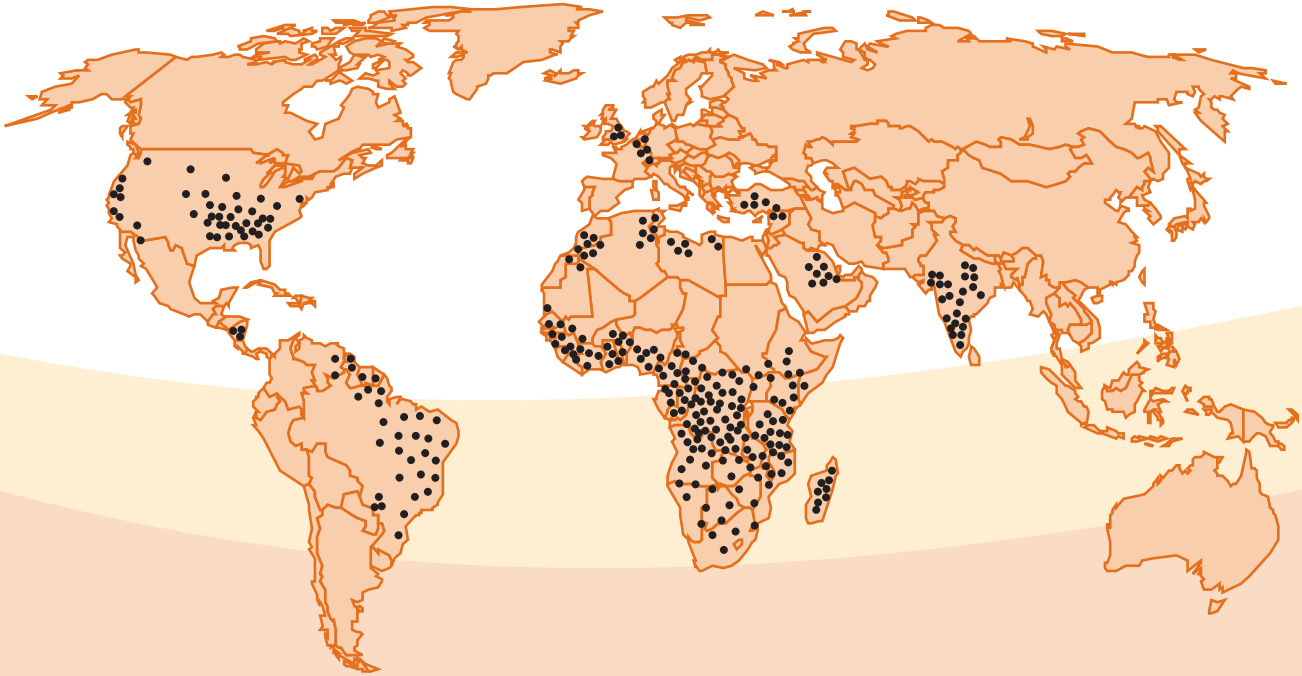
Normal RBC slide



Slide from person with SCD – (anemic)

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, southern Europe, central and southern India, and Africa.



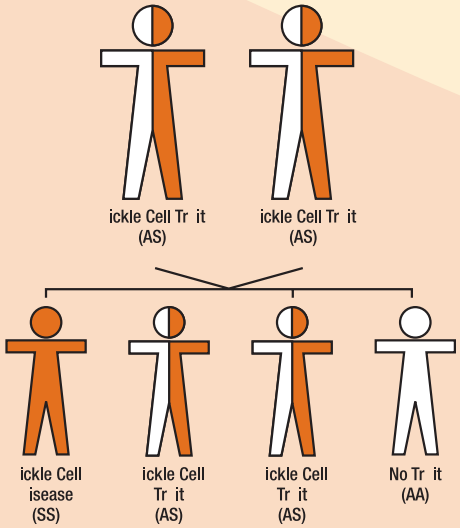
Are there different types of sickle cell disease?

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 hemoglobin S gene from either one or both parents.

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

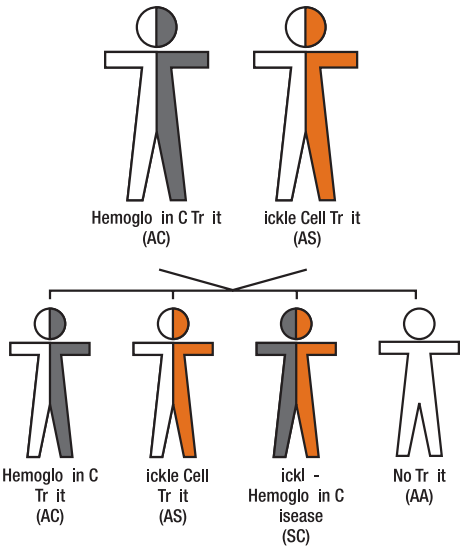
Hemoglobin SS Disease

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



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



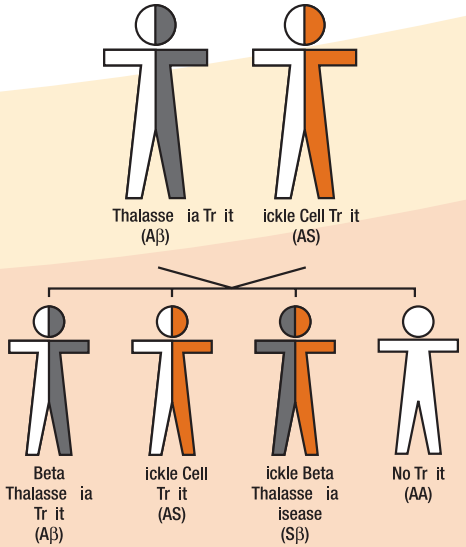
Sickle Beta-Thalassemia Disease

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.

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.

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 disease, and sickle beta thalassemia disease.

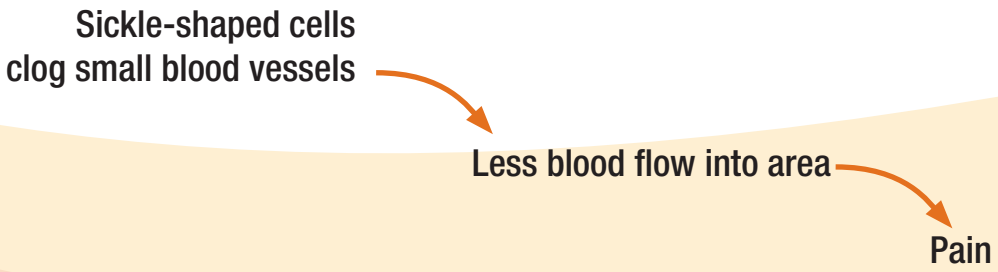
Unit 2: Pain and Infection

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:

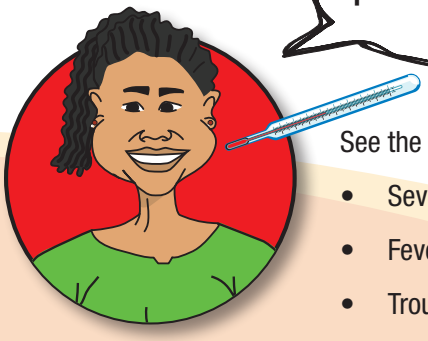
- Lack of sleep—Get at least 8 hours of sleep each night.
- Not drinking enough fluids—Drink at least 8 glasses of liquids each day (water is best). Avoid caffeine drinks such as energy drinks, cola, or coffee. Caffeine causes you to go to the bathroom more often, and it causes your blood vessels to 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 Level	Where:	Treatments	Why	Example
Mild	Can be treated at home	Ibuprofen every 6 hours for 1–2 days.	Prompt treatment of mild pain is important because it may prevent a more severe pain episode.	(Motrin®, Nuprin®, or Advil®)
		Distraction techniques	Helps control pain	Playing video games
		Increase fluids	Makes the blood thinner and increases blood volume. This allows blood to flow more freely through the vessels.	Drink 8 glasses of water. Do not drink caffeinated drinks (coffee, cola). These drinks make you go to the bathroom more often.
		Rest	Allows the body to conserve energy	Sleep, if possible
		Heat—No ice, ice makes the vessels constrict (narrow)	Warm heat causes the blood vessels to dilate (expand).	Use a warm towel or take a warm bath
Moderate	May be treated at home	Oral Opioids—follow directions on the prescription bottle.	Blocks the chemicals that cause pain and reduces the feeling of pain	Tylenol® with codeine
		Increase fluids	Make the blood thinner and increases blood volume. This allows blood to flow more freely through the vessels.	Drink 8 glasses of water. Do not drink caffeinated drinks (coffee, cola). These drinks make you go to the bathroom more often.
		Distraction techniques	Helps control pain	Talking on the telephone
Severe	Treated by a doctor at a medical center	IV Opioids	Reduces the feeling of pain	IV morphine
		NSAIDS	Blocks hormones that cause pain and swelling	Ketorolac (Toradol®)
		Fluids	Helps increase the volume of the blood.	IV Fluids

When does a pain episode require treatment by a doctor?



See the doctor any time you have:

- Severe pain,
- Fever of 100.9 degrees F or above, or
- Trouble breathing.

To learn more about sickle cell pain and what to do, see “Do You Know… Pain and Sickle Cell Disease.”

Severe pain requires treatment by a doctor.

If pain is severe, lasts longer than 24 hours, or the medicine does not help, call the clinic at 901-595-5041 Monday–Friday, 8 a.m. –5 p.m. After 5 p.m. and on weekends, call 901-595-3300 and ask to talk to the hematologist on call.

Infection

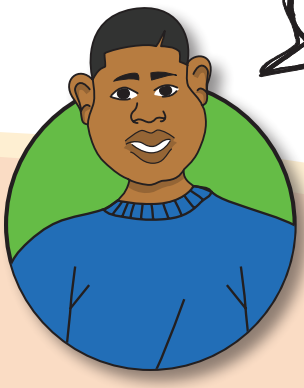
Are people with sickle cell disease more likely to get serious infections?

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?

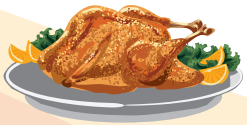
Infection	Common symptoms
Blood	Fever (usually 100.9° F or higher)
Lung (Pneumonia or Acute Chest)	Fever, cough, chest pain, trouble breathing
Bone	Fever, swelling, pain



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.
- To learn more, see "Do You Know… Clean Hands."



Up-to-date immunizations can prevent many of the more serious infections, especially pneumococcal bacteria, which causes pneumonia and other infections.

- All of your childhood immunizations should be up-to-date.
- Get a flu vaccine every year.
- Pneumococcal vaccines: Prevnar® at 2, 4, 6 and 12 months and Pneumovax® at 2, 5, and 15 years old
- Meningococcal vaccine at 24 months, 26 months, 5 years and then every 5 years
- Other vaccines that might be recommended by your medical team, such as hepatitis B.

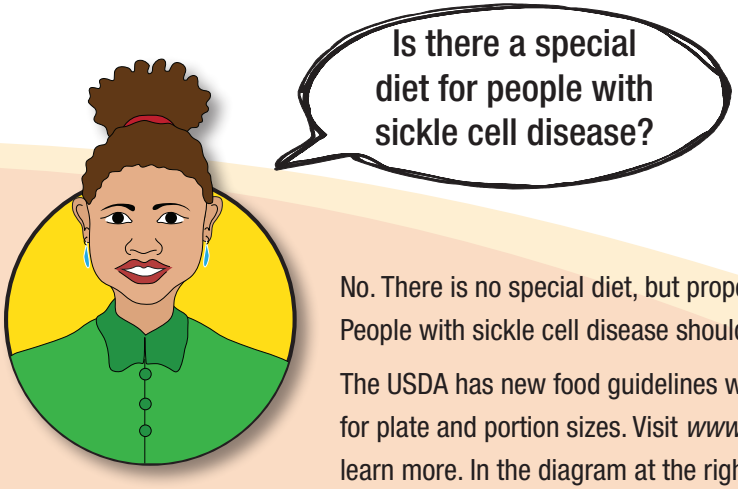
Unit 2: Summary

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 100.9 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



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.

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

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.

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.

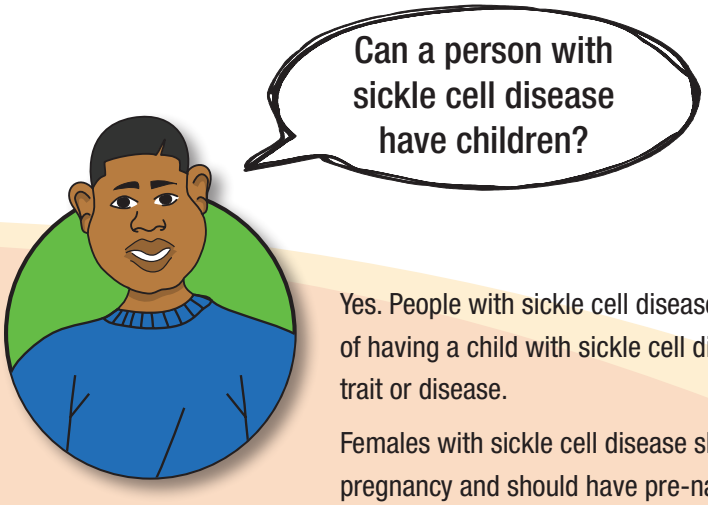
A moderate exercise program 3 to 4 times a week is usually encouraged. However, the best type and amount of physical exercise is different for each person. Most people learn to set their own limits, based on experience.

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.

Should a person with sickle cell disease drink alcohol?

No. Alcoholic beverages cause fluid loss, which can lead to dehydration (too little fluid in the body) and pain.



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. To learn more, see “Do You Know… Priapism and Sickle Cell Disease.”

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.

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.

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 deletes the supply of oxygen to the body.

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. 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.

Unit 3: Summary

A nutritious diet and a good fluid intake (8–10 glasses of liquid per day) are important for maintaining good health.

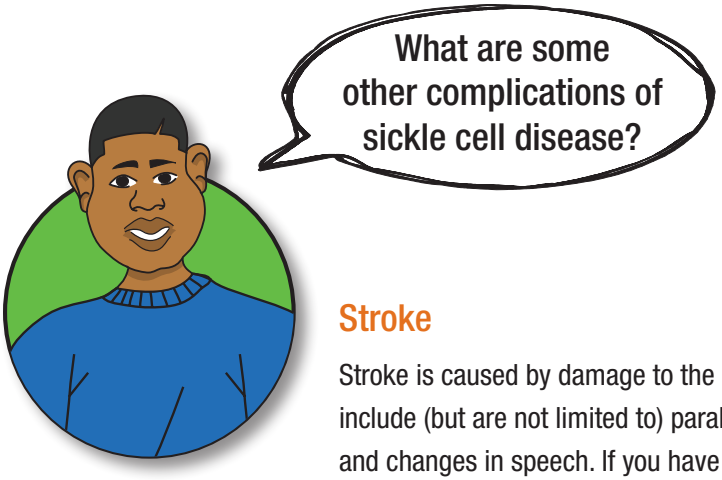
A moderate exercise program, based on your own tolerance, is recommended.

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 Other Sickle Cell Disease Complications



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. One in 10 people with hemoglobin SS disease and sickle beta zero thalassemia disease will have a stroke. 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. This can result in the need for joint replacement. In people with sickle cell disease, AVN normally occurs in the hip joint and the long bones between the shoulder (humerus) and the leg (femur).

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. To learn more, see "Do You Know… Priapism and Sickle Cell Disease."

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

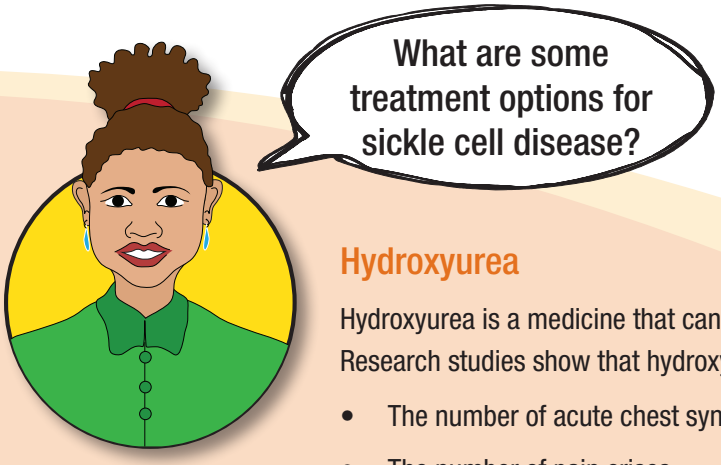
Gallstones are hard rock-like deposits that form in the bile duct or in the gallbladder. In people with sickle cell disease, most gallstones are produced from excess bilirubin, which is caused by the constant breakdown of red blood cells. Biliary sludge is excess bile that settles in the duct. It can 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.

Unit 4: Summary

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

1. Stroke—damage to the brain
2. Avascular necrosis—damage to the bone
3. Acute Chest Syndrome—pneumonia plus other symptoms
4. Retinopathy—damage to the eye
5. Priapism—painful erections
6. Osteomyelitis—an infection in the bone
7. Gallstones- Hard rock-like deposits that form in the gallbladder.

Unit 5: Common Treatment Options for Sickle Cell Disease



Hydroxyurea

Hydroxyurea is a medicine that can help children and adults with sickle cell disease. 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 sickle cell patient might need blood transfusions:

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.

To learn more about transfusions, see the booklet "Red Blood Cell Transfusion for Sickle Cell Disease."

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

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For general information on sickle cell disease, visit our website *www.stjude.org/sicklecell.*

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