



Hand in Hand

A HANDBOOK ON SICKLE CELL DISEASE
FOR THE CHILD AND FAMILY

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at Texas Children's Hematology Center*



**Texas Children's
Hospital®**

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INTRODUCTION

This handbook was written by the team at the Sickle Cell Center at Texas Children's Hematology Center to offer important information to patients and families on sickle cell disease (SCD).

Serving over 1,000 children each year, the Sickle Cell Center at Texas Children's Hematology Center is the largest in Texas, and one of the largest in the United States. Our multidisciplinary team is made up of board certified pediatric hematologists, hematology-trained nurse practitioners and physician assistants, research staff and social workers. Texas Children's Sickle Cell Center offers ambulatory services, including outpatient blood transfusion and pain treatment programs, a sickle cell pulmonary clinic, sickle cell stroke clinic and genetic counseling, all in one location.

Our program conducts state-of-the-art clinical and laboratory research aimed at gaining a better understanding of the disease, preventing its complications and ultimately finding a cure. The center's research funding is provided by government grants and the philanthropic support of a variety of community-based organizations. This partnership allows us to offer even more novel therapies to our sickle cell patients.

To learn more about the Sickle Cell Center at Texas Children's Hematology Center, please visit txch.org/sickle-cell-program. To learn more about our dedicated team treating sickle cell disease, please visit txch.org/hematology-center/sickle-cell-program/treatment-team.

We hope this book helps answer many questions about the disease including:

- What is sickle cell disease?
- Is it contagious?
- How do you get sickle cell disease?
- How is sickle cell disease treated?
- Is there a cure for sickle cell disease?

In the back of this book, you will find a glossary of medical terms and space for you to add your own words or any questions you may want to ask your medical provider.

Children with SCD have unique medical needs that require ongoing monitoring and management in order for them to maintain optimal health. They may receive multiple treatments prescribed by their hematology providers. Some of the treatments recommended are medications, blood tests, X-rays and/or blood transfusions.

Children with SCD are at risk for many complications related to the disease such as pain crisis, acute chest syndrome, infection, stroke and/or iron overload. This booklet provides information on how to recognize symptoms of common complications, as well as how best to manage the problems.

Children with SCD face many challenges. The comprehensive health care team at Texas Children's Hospital and other community groups can help them meet those challenges through treatment, support, research and education about SCD.

We will be here for you throughout every step of your child's care for sickle cell disease.

WHAT IS SICKLE CELL DISEASE?

Sickle cell disease is an inherited condition that affects a protein inside the red blood cell called hemoglobin. The job of hemoglobin is to carry oxygen to different parts of the body.

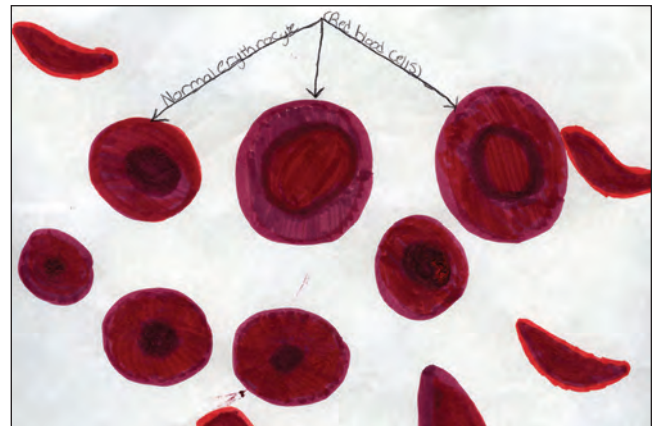
What is the difference between the normal red blood cell and the sickle red blood cell?

The normal red blood cell:

- Has a round shape, like a donut
- Is flexible
- Moves easily through the blood vessels
- Lives 120 days

The sickle red blood cell:

- Has a crescent shape, like a banana
- Is stiff and sticky
- Sticks to the blood vessel walls
- Lives 14 days



Ryland, age 11

When the sickled red blood cells clump together, they block the flow of blood and oxygen through the blood vessels. This causes pain and many other complications.

How does someone get sickle cell disease?

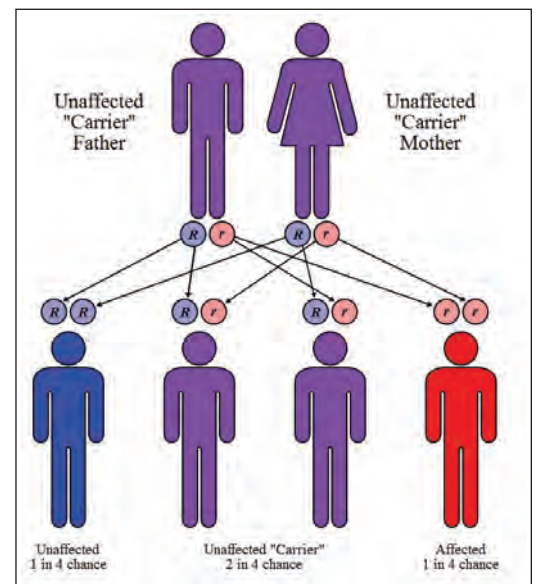
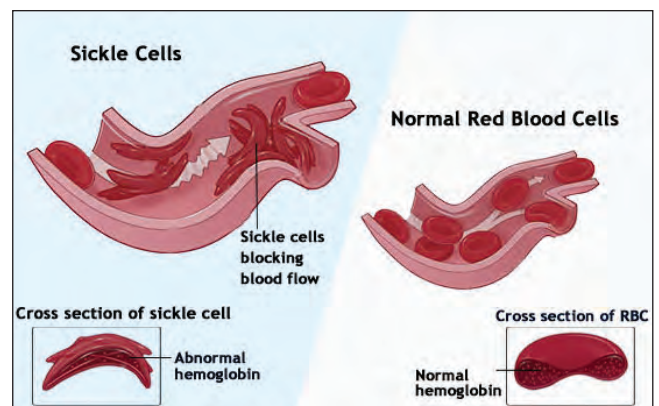
Sickle cell disease is an inherited blood disorder, diagnosed by a blood test in the newborn screening. It is the most common genetic disorder in African Americans, affecting about one in 400. It can also be found in people with origins from the Mediterranean, the Middle East and South and Central America.

Sickle cell disease is passed from parents to their children through their genes. Genes provide the coded information that determines everything about us, including the color of our eyes, how tall we are and what type of hemoglobin we produce inside our bodies. The genes that determine our characteristics are present in pairs. Children get one hemoglobin gene from their mother and one hemoglobin gene from their father. Each parent has two genes for hemoglobin, but only one of these genes from each parent is passed on to the child. The gene that is passed on from each parent happens by chance, like flipping a coin.

To have sickle cell disease, a child inherits a hemoglobin S gene from one parent and a hemoglobin S, hemoglobin C or beta thalassemia gene from the other parent. With each pregnancy from the same mother and father with this genetic make up, there is a chance that the child will have normal hemoglobin, sickle cell trait or sickle cell disease.

What is sickle cell trait?

If a person inherits one normal gene and one sickle cell gene, the person will have sickle cell trait. People with sickle cell trait are healthy and do not have sickle cell disease. However, they can pass the sickle cell gene to their children.



Is there more than one type of sickle cell disease?

The type of sickle cell disease your child has depends on which hemoglobin genes they have inherited. The severity of your child's condition also depends on which type of sickle cell disease the child has. The table below shows the most common types of sickle cell disease and the severity of each type. There are many other types of sickle cell disease, but they are rare. It is important to remember that sickle cell disease affects each person differently.

TYPE OF SICKLE CELL DISEASE	SEVERITY
Hemoglobin SS (Hb SS)	Severe
Hemoglobin SC (Hb SC)	Moderate
Sickle beta zero thalassemia	Severe
Sickle beta plus thalassemia	Mild



COMPLICATIONS OF SICKLE CELL DISEASE

ACUTE CHEST SYNDROME

Acute chest syndrome (ACS) is a complication of sickle cell disease that results in lung injury, breathing difficulty, low oxygen to the rest of the body and possible death. It can be caused by infections. Sickled cells may also cause a blockage of blood flow to the lungs and chest. ACS is one of the most common causes of hospitalization for children with sickle cell disease. ACS is the cause of more than 25 percent of early deaths in sickle cell disease.

How common is ACS in children with sickle cell disease?

About half of all children with sickle cell disease will have at least one episode of ACS. Very often, children who have had ACS will have a repeated episode.

What are the warning signs of ACS?

- Fever
- Cough
- Chest pain
- Fast-paced breathing
- Shortness of breath or grunting
- Wheezing

Why should I worry about ACS?

ACS can create a dangerous shortage of oxygen to important parts of the body such as the lungs, brain, heart, kidneys and other vital organs. The damage to these organs may result in permanent problems speaking, walking, learning and other day-to-day functions. It may even cause death.

What should I do if I suspect my child has ACS?

You should get medical help as quickly as possible by taking your child to the emergency room or calling your doctor immediately. It is important to get treatment right away to reduce the risk of damage to vital organs or possibly death.

How does my child's doctor diagnose ACS?

Your doctor will examine your child, using a stethoscope to listen for breathing difficulty. The doctor will also check your child's oxygen levels, run blood tests and take a chest X-ray.

What is the treatment for ACS?

Your child will most likely be admitted to the hospital, given treatment and watched closely. Treatment for ACS may include antibiotics, oxygen, breathing exercises and a blood transfusion.

In very severe cases, a child will be put on a machine to help him or her breathe. In addition, your child may require a special type of blood transfusion using a pheresis machine to replace sickle cell blood with non-sickle cell blood.

How can ACS be prevented?

The risk of ACS can be reduced by:

- Keeping immunizations up to date, including a yearly flu vaccination.
- If your child has been diagnosed with asthma, using asthma prevention medications. There is a higher risk of developing ACS during an asthma attack.
- Doing breathing exercises using an incentive spirometer. This should be done during all pain episodes at home or while in the hospital.
- Asking your doctor if preventive medications, such as hydroxyurea, are suitable for your child.
- Seeing a lung specialist, if recommended by your doctor.

Children who have repeated episodes of ACS are candidates for a bone marrow transplant to cure sickle cell disease. Ask your medical provider for more information.



COMPLICATIONS OF SICKLE CELL DISEASE

APLASTIC CRISIS

An aplastic crisis happens when the body stops making red blood cells. When the production of red blood cells shuts down, your child's blood count may drop dangerously low. This condition may last up to 10 days.

What causes an aplastic crisis?

In a child with sickle cell disease of any type, an aplastic crisis can happen when the child is exposed to certain viruses. The most common virus that causes an aplastic crisis is the parvovirus B19. This is a common infection in young and school-age children.

What are the symptoms of aplastic crisis?

- Pale color of lips, gums or fingernails
- Fever (temperature of 101°F or greater)
- Extreme tiredness
- Weakness
- Loss of energy
- Rapid heart rate
- Shortness of breath
- Irritability

What is the treatment for aplastic crisis?

It will be necessary to draw blood from your child to check the hemoglobin level. If the hemoglobin level has dropped too low, a blood transfusion will be necessary. The blood transfusion is necessary to give your child blood cells until the body starts making its own red blood cells again. It may be necessary for your child to be hospitalized during the aplastic crisis for additional treatments or tests.

It is very important for your child to follow up with his or her doctor within a week of discharge from the hospital to make sure that the hemoglobin level remains high enough.

Can an aplastic crisis happen more than once?

It is very unlikely that your child will have multiple aplastic crises. Once affected, immunity to parvovirus usually lasts a lifetime.



COMPLICATIONS OF SICKLE CELL DISEASE

AVASCULAR NECROSIS

Avascular necrosis (AVN) is a condition that happens when bone tissue dies due to a lack of blood supply to the bone. This causes the bones to decay and eventually collapse. The condition is most common in the hip or shoulder joints. AVN can begin in the teenage years and occurs in at least 40 percent of adults with sickle cell disease.

What causes AVN?

Sickling red blood cells may block the flow of blood to the bones in the joints. When bones do not get enough oxygen and nutrients from the blood supply, they begin to erode away. The joint eventually collapses.

What are the symptoms of AVN?

- Pain or stiffness in the joint
- Decreased range of motion
- Limping
- Decrease in leg length

How is AVN diagnosed?

Early detection is important to prevent progression. An MRI detects early stages of AVN, often when few symptoms are noticed. An X-ray or CT scan may detect AVN in later stages.

How is AVN treated?

The first lines of treatment recommended are:

- Minimize bearing weight by resting in bed or using crutches
- No impact activities, such as running or jumping
- Physical therapy to increase range of motion, strengthen the surrounding muscles and increase circulation in the joint
- Anti-inflammatory medications, such as ibuprofen (narcotic pain medication can be prescribed for more severe pain)
- Hydroxyurea to decrease sickling of the red blood cells and minimize further bone damage

Surgery is an option, but it is not recommended until a patient has failed the first line of treatment therapy. Surgical options should be discussed with an orthopedic surgeon. Bone core decompression, osteotomy or total joint replacement may be considered at later stages of AVN.



COMPLICATIONS OF SICKLE CELL DISEASE

DELAY IN GROWTH AND PUBERTY

Children with sickle cell disease usually grow and develop more slowly, even reaching puberty later than their peers. This growth delay is caused by having fewer red blood cells. Adults with sickle cell disease are also typically shorter and thinner than the general population.

Is there anything that will help my child grow?

There are things you can do to help your child achieve the best possible health, height and weight:

- Getting plenty of exercise and eating a well balanced diet is important. Children with sickle cell disease need more calories, vitamins and minerals than other children. This helps give them more energy and helps in making blood cells. Eating a variety of meats, vegetables and fruits will provide a good supply of vitamins. Your medical provider or nutritionist will be happy to discuss ways to improve your child's diet.
- A medication called hydroxyurea increases the hemoglobin level in most children with sickle cell disease. A higher hemoglobin level indicates that more red blood cells are circulating in the body. Red blood cells carry oxygen to all the tissues, bones and organs and affect growth. Speak to your medical provider to find out if hydroxyurea is a good therapy for your child.



COMPLICATIONS OF SICKLE CELL DISEASE

RETINOPATHY (EYE DISEASE)

What is sickle cell retinopathy?

Retinopathy is damage to the blood vessels in the retina of the eye, located in the back part of the eye. Retinopathy occurs in 33 percent of people with Hb SC disease and in three percent of people with Hb SS disease. It occurs in older children and adults.

What causes retinopathy?

Sickled red blood cells become trapped in the small vessels of the eye, resulting in decreased blood flow to that area of the eye. This may cause loss of vision. This damage may be permanent and blindness can occur in later stages.

What are the symptoms of retinopathy?

In the early stages of retinopathy, there may be no symptoms. For this reason, it is important to have your child's eyes checked once a year by an eye doctor. The eye exam includes dilation of the pupil in order to see if the vessels appear normal or if changes or scarring are occurring.

In later stages, your child may complain of:

- Seeing dark spots or shadows in his field of vision
- Blurry vision
- Sudden loss of vision
- Pain in the eyes

How is retinopathy treated?

Early treatment by an eye doctor may include more frequent eye check ups to monitor the progression, laser treatment or surgery.

What should I do to prevent retinopathy?

When your child turns 10 years old, he or she should have an eye exam by an ophthalmologist once a year. Ask your hematology provider what is needed to schedule an appointment.



COMPLICATIONS OF SICKLE CELL DISEASE

GALLSTONES

The gall bladder is a small gland between the liver and the stomach. It aids in the digestion of foods, especially those that may be spicy or greasy. Due to sickling blood cells, gallstones may form in the gall bladder.

Are gallstones harmful to the body?

Gallstones are usually not harmful but can lead to serious health problems. Your child might have discomfort when gallstones settle in the bile duct or collect in the gallbladder.

What are the signs and symptoms of gallstones?

- Sudden, sharp pain on the upper right side of the stomach
- Upset stomach or vomiting
- Yellowing of the skin and eyes (also called jaundice)
- Dark urine (tea-colored or brown)

What do I do if my child has these symptoms?

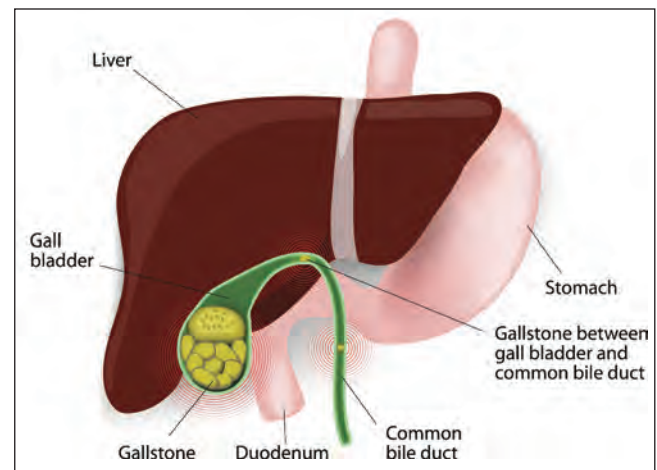
If your child has any of these symptoms, call your doctor immediately.

How are gallstones treated?

If your child is not having problems from the gallstones, his or her doctor may choose to simply monitor the condition.

If your child is having frequent pain, his or her doctor may choose to treat the symptoms in some of these ways:

- IV fluids to prevent dehydration (not enough fluid in the body)
- Anti-nausea and vomiting medicine
- Pain medicine
- Antibiotics (if a fever develops)
- Surgical removal of the gallbladder





COMPLICATIONS OF SICKLE CELL DISEASE

PAIN OR VASO-OCCLUSIVE CRISIS

What causes sickle cell pain?

Pain is one of the most common and well-known complications of sickle cell disease. Pain associated with sickle cell disease is caused when the sickle red blood cells block the flow of blood and oxygen to the body. This is called a pain crisis or vaso-occlusive crisis. Unfortunately, this pain can occur at any time and in any part of the body.

How is pain treated?

Most pain caused by sickle cell disease can be treated at home by:

- Increasing fluid intake to maintain proper hydration
- Rest and quiet play until the pain subsides
- Heating pads, warm/moist towels or hot baths
- Gentle massage to relax tense muscles and increase blood flow
- Using an incentive spirometer to do breathing exercises to prevent further complications such as acute chest syndrome
- Pain medications, as prescribed. Acetaminophen (like Tylenol) or ibuprofen (like Motrin) may be used first for mild pain. If this does not relieve the pain, stronger pain medication (narcotics) such as codeine or hydrocodone may be needed. It is also very effective to alternate narcotics and ibuprofen to provide continuous relief. Talk to your medical team about the best options for alternate medication.

The key is to stay ahead of the pain. It is much easier to keep the pain from getting worse than it is to control severe pain.

If the home management strategies described above do not work, contact your medical team or bring your child to the emergency room.

Your child may need more aggressive treatment such as IV (in the vein) pain medication and fluids. Your child may also need to be admitted to the hospital for further evaluation and to be watched closely.

What causes a pain crisis?

It is often difficult to determine what causes pain to occur. Certain triggers that may cause pain are:

- Dehydration
- Fever
- Physical exhaustion or being very active without resting
- Exposure to extreme temperatures such as very hot or cold weather or swimming
- Stress or anxiety

What if my child has fever and pain at the same time?

Fever is defined as a temperature of 101°F or higher whether taken under the tongue, under the arm or rectally. If your child is experiencing an elevated temperature and pain, it is important to take his or her temperature before giving each dose of pain medication. Both acetaminophen and ibuprofen are fever reducers. Taking the temperature before giving either of these medications allows you to know if your child has fever which may be masked.

If your child's temperature reaches 101°F degrees or greater, call your medical team immediately or bring your child to the emergency room. Fever is a sign of more serious complications, and your child needs to be examined to receive additional medical treatment.

What is dactylitis?

Children with sickle cell disease under 3 years old may first experience pain and swelling in their hands or feet. This is called dactylitis, caused by sickle red blood cells blocking the flow of blood in the hands and feet. You may also notice that your child doesn't want to hold a toy or want to walk.

Treatment is much the same as other pain episodes. It is important to keep your child well hydrated, give pain medication and apply warm compresses or gentle massage to the hands or feet. If your child does not improve, contact your medical team for further instruction.



COMPLICATIONS OF SICKLE CELL DISEASE

PICA

Pica is an eating disorder characterized by a craving or need to eat things that are not food. It can happen to children of all ages. There is a higher occurrence of pica in children with sickle cell disease compared to the general population.

What are the characteristics of pica?

Children with pica may eat items commonly found in and around the home such as ice, foam, powder, paper, sheetrock, fabric, hair, wood, pencils, erasers, dirt, sand, rocks or other things.

Pica behavior is found in children with all types of sickle cell disease, but is most common in children with Hb SS type.

What causes pica behavior?

It is not totally clear why some children have pica. It has been associated with children with low weight or anemia (low blood count). It has also been associated with vitamin deficiencies such as zinc.

What is the treatment for pica?

Removing any items from your home that your child is eating is a good start. Since this is not always possible, distracting or re-directing your child to another activity may help. It is important to let your child know that they need to stop the behavior without scolding or punishing them.

In some cases, children are referred to speak to a psychologist who can help provide more tools for the child and parent that may help.

Why is it important to let your health care provider know if your child has pica?

The longer a child has pica behavior, the more difficult it is to stop. Some of the health problems that may occur are:

- Blockage in the intestines, sometimes requiring surgery
- Infection
- Abdominal pain
- Damage to teeth



COMPLICATIONS OF SICKLE CELL DISEASE

PRIAPISM

One serious complication that occurs in boys with sickle cell disease is priapism. Priapism is a painful erection that will not go away, caused when sickle cells block the blood vessels to the penis. Acute attacks of pain may happen during sleep or following sexual activity, but often there is no identifiable cause. Priapism is more common after puberty but can also occur in young boys.

What should I do if my son has priapism?

There are simple comfort measures to initiate at home with the onset of priapism to help end an episode:

- Increase your child's intake of oral fluids
- Give pain medication, such as ibuprofen (Motrin)
- Encourage your child to empty his bladder as often as possible
- Have your child take a warm bath

Any episode lasting longer than two hours should be considered an emergency and requires prompt medical attention. Go to the emergency room immediately. Priapism that goes untreated may lead to infertility in over 80 percent of cases.

What are the types of priapism?

- Stuttering priapism: an unwanted erection that occurs repeatedly with moments of improvement usually lasting less than two hours. Most episodes of stuttering priapism can be managed at home. Repeated episodes of this type of priapism are common and occur in up to 40 percent of boys with sickle cell disease.
- Prolonged priapism: an erection lasting for more than two hours that may continue for days to weeks.

What is the treatment for priapism?

If your child's episode of priapism does not go away after treatment at home for up to two hours, take him to the emergency room. Your child will be evaluated and given IV fluids and pain medication. He may be admitted to the hospital for further evaluation and treatment. In severe cases, a needle aspiration or surgery may be required.



COMPLICATIONS OF SICKLE CELL DISEASE

RENAL DISEASE AND ALTERED KIDNEY FUNCTION

Sickling blood cells cause damage to the kidneys that may affect your child. Some of the problems caused by sickling damage in the kidneys are:

- Hypertension or high blood pressure
- Enuresis or bed wetting
- Dehydration, which may lead to other complications such as pain, shortness of breath and acute chest syndrome

Why are the kidneys so important?

The kidneys act as a filter by removing waste products and maintaining water balance in the body, almost like a pasta strainer. The kidneys help keep things in your body that you need and get rid of things that you don't. Some of the roles of the kidneys are to:

- Make urine
- Remove waste and extra fluids from the body
- Control the chemical balance of the body
- Regulate blood pressure

What is hypertension?

Hypertension, also known as high blood pressure, is a serious condition which may ultimately lead to heart damage or stroke. The kidneys help keep your child's blood pressure normal. Sickle cells have the ability to cause kidney damage, making it difficult for them to function properly. Therefore, your child may have high blood pressure when his or her vital signs are taken during regular checkups.

Your doctor may refer your child to a renal doctor (also called a nephrologist, a specialist in kidney disease) to further evaluate your child and possibly prescribe medication to keep blood pressure normal.

What is sickle cell enuresis?

Sickle cell enuresis or "bed-wetting" is common in children with sickle cell disease. Sickling red blood cells change the function of the kidneys in ways that are not clearly understood. Due to sickling, the kidneys are unable to concentrate urine effectively, sometimes resulting in larger amounts of urine. Your child may wet the bed even when you do not give him or her a lot to drink before bedtime. Eventually, your child will outgrow this condition.

What are the best ways to treat enuresis?

- Make sure there are no problems like a bladder infection. If your child has fever or burning or pain with urination, call your doctor right away.
- Train your child to feel when his or her bladder is full and empty it.
- Have your child go to the bathroom prior to bedtime and wake him or her up in the night to go to the bathroom.
- Use alarm clocks, night lights and bedwetting alarms.
- Use of medication may also be an option for your child. Talk to your medical provider to learn more.

Bedwetting can be very frustrating for both the child and the parent. It is important to remember that your child is not wetting the bed on purpose. It is not recommended to scold or punish your child for bedwetting.

Why is it important to stay well hydrated?

Dehydration occurs when the body does not get enough fluid or loses too much fluid. Common causes of dehydration are diarrhea, fever, vomiting and sweating from physical activity. Dehydration may lead to a pain crisis or other complications in children with sickle cell disease.

What are signs of dehydration?

- No tears
- Decreased urine (fewer wet diapers)
- Tiredness
- Dry mouth and tongue
- Sunken eyes
- Sunken soft spot on the head (in babies)



Samantha, age 9

How can I ensure my child stays well hydrated?

- Have your child drink before play and call them inside for beverage breaks frequently, especially in hot weather.
- Pack water bottles in your child's backpack and lunch box for school and other activities.
- Make sure school personnel know that children with sickle cell disease need water or other fluids available at all times. A letter for the school can be provided, if needed.

What are the fluid requirements for children with sickle cell disease?

CHILD'S WEIGHT	NUMBER OF 8 OZ. CUPS PER DAY
25 pounds	5
50 pounds	8
75+ pounds	10+



COMPLICATIONS OF SICKLE CELL DISEASE

RISK OF INFECTION AND FEVER

Infection is a major complication in children with sickle cell disease. The risk of getting an infection, especially a severe one, is much greater in someone with sickle cell disease due to a decrease in the immune response or the body's ability to fight germs that cause infections.

One way the body fights germs that cause infection is through an organ called the spleen. The spleen acts as a filter to remove harmful germs, such as bacteria, from the body. It also makes antibodies to fight infection. In someone with sickle cell disease, the spleen does not function properly, therefore it does not protect the body enough from certain bacteria that cause infection.

These bacteria may cause life-threatening infections such as septicemia (infection in the blood), pneumonia (infection in the lungs), osteomyelitis (infection in the bone) or meningitis (infection around the brain). Fever is a sign that your child has an infection. Knowing how to respond when your child has a fever is critical to his or her health care.

What should I do if my child has fever?

Fever is defined as a temperature of 101°F or higher whether taken under the tongue, under the arm or rectally. Many times, fever is the first sign of a serious or life-threatening infection. Infection is a major cause of death in children with sickle cell disease. If your child has fever, you should notify the hematology medical team immediately or take your child to the emergency room.

During the emergency room evaluation, a blood test, chest x-ray, and urine test will be taken to find the cause of the fever. An antibiotic to help fight infection and IV fluids will also be given. Depending on the test results or the condition of your child, he or she may be admitted to the hospital or sent home.

Contact information will be provided to you by your hematology medical team. Someone on the hematology medical team is available 24 hours a day, seven days a week.

What if my child has a low grade temperature?

It is **not** recommended to give medication such as acetaminophen (Tylenol) or ibuprofen (Motrin) if your child has a low grade temperature. These medications will lower the temperature, but the bacteria or virus that is causing the fever will continue to grow. Your child's illness may worsen and become very serious.

If your child's temperature reaches 101°F or greater, notify the hematology medical team or take your child to the emergency room.

What is the role of medication in fighting infection?

Penicillin is a medication that will help prevent serious infections. If your child should become infected with certain bacteria without having this protection, the condition could be very serious and life threatening. Therefore, taking penicillin as recommended is a very crucial part of your child's well being.

Penicillin is started shortly after birth. The starting dose is 125 mg twice daily.

At age 3, the dose is increased to 250 mg twice daily.

Children with sickle beta plus thalassemia do not have to take penicillin because the risk of serious bacterial infections is much lower in these children. If you have questions, discuss it with your medical team.

Will my child always have to take penicillin?

Once your child is around 5 years of age, your medical provider may choose to stop penicillin. It may be okay to stop penicillin if your child has:

- Never had a serious bacterial infection
- Never had the spleen removed
- Had all their immunizations, including specific ones required for children with sickle cell disease

Talk to your medical provider about the best recommendation for your child.

What is the role of immunizations/vaccines in fighting infection?

Immunizations are another way to provide antibodies to help fight serious infections. It is very important for your child to be up to date with all immunizations. Since they are not routinely given, your child's pediatrician or primary care doctor may not give these vaccines during their regular well child check ups. Your hematology medical provider can provide the needed immunizations.

Additionally, children with sickle cell disease should receive two immunizations which are not routinely given to other children. When your child is 2 years of age, it is important that he or she receives a vaccine for the pneumonia bacteria (Pneumovax) and a vaccine for the meningitis bacteria (Menveo or Menactra). Booster vaccines will be needed as your child gets older. It is also very important that your child receive an influenza vaccine every year, starting at 6 months of age.



Julius, age 9



COMPLICATIONS OF SICKLE CELL DISEASE

SPLENIC SEQUESTRATION

What is the function of the spleen?

The spleen is the organ found in the upper left corner of the abdomen, under the edge of the ribs. It is a filter that cleans the blood of waste products and toxins and helps to fight infections.

What is a splenic sequestration?

When sickled red blood cells block the flow of the blood coming out of the spleen, the spleen becomes large. This trapping of the blood cells inside the spleen is called sequestering. As a result, there are fewer red blood cells in circulation to provide oxygen, causing a drop in the blood count.

What are the signs of a splenic sequestration?

- Splenomegaly, meaning the the spleen has become larger and is pressing on the abdomen below the rib cage. In this instance, the spleen will feel like a firm mass and may feel tender.
- Tiredness and lack of energy
- Paleness in the lips or palms of the hands
- Irritability
- Fever

What should I do if my child has signs of splenic sequestration?

If your child is having the symptoms above, you should immediately notify your medical provider or take your child to the emergency room.

What is the treatment for splenic sequestration?

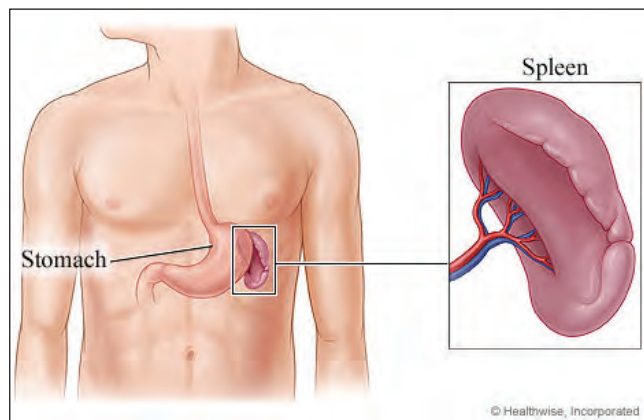
Your child will be examined and receive a blood test. If your child's hemoglobin level has dropped too low, a blood transfusion will be necessary. Your child may require admission to the hospital for further evaluation and observation.

If your child has had a very severe splenic sequestration where the blood count dropped dangerously low or has had more than one episode of splenic sequestration, your child may need to have his or her spleen removed. Your doctor will discuss the option of a splenectomy (surgical removal of the spleen) to prevent another splenic sequestration. Your doctor will discuss the risks and benefits of a splenectomy.

Your child does not need a spleen to live. However, if the spleen is removed, your child will continue to take penicillin twice daily to prevent infection.

Who is at risk of splenic sequestration?

Splenomegaly and/or splenic sequestration usually occurs in young children, less than 5 years of age, with Hb SS or sickle beta zero thalassemia. It usually occurs in children with Hb SC when they are school age. The risk is very low in children with sickle beta plus thalassemia.





COMPLICATIONS OF SICKLE CELL DISEASE

STROKE

What is a stroke?

A stroke is a brain injury caused by a blockage of blood flow to the brain. Two kinds of strokes occur in children with sickle cell disease: obvious strokes and silent strokes. Obvious strokes have very recognizable physical symptoms, while silent strokes have no noticeable signs. However, both cause damage to the brain.

How common are strokes in children with sickle cell disease?

Strokes are common in children with sickle cell disease. Sickle cells may block the flow of blood in the brain in the same way that sickle cells block the flow of blood to muscle or bone.

Historically, about one in 12 children with Hb SS or sickle beta zero thalassemia will have an obvious stroke before they reach 20 years of age. Strokes are rare in children with Hb SC or sickle beta plus thalassemia.

Approximately one in four children with sickle cell disease will have a silent stroke. Silent strokes can only be seen on MRI pictures of the brain. Silent strokes may result in learning difficulty and behavior problems.

What are the warning signs of an obvious stroke?

- Weakness or numbness of a body part
- Facial droop
- Blurry vision, seeing double or vision loss
- Severe headache, dizziness or unsteadiness while walking
- Confusion, trouble speaking or understanding speech
- Seizures
- Excessive sleepiness, difficulty staying awake and alert
- Change in behavior or school performance without obvious cause

Why should I worry about a stroke?

Stroke can result in severe disability with speaking, walking and other daily functions. Children who have had a stroke are more likely to have problems with learning in school and may also have behavior problems. Strokes can also potentially lead to death.

What should I do if I suspect my child may be having a stroke?

Get medical help right away by calling 911. Immediate treatment of a stroke is imperative to potentially reduce the amount of brain damage and save the child's life.

What is the treatment of an obvious stroke?

If your child is having a stroke, he or she will be admitted to the hospital for evaluation, testing and treatment. Your child will receive a blood transfusion, usually with a pheresis machine, where sickle cells are removed from the body and replaced with non-sickle blood cells. After discharge from the hospital, your child will continue to need monthly blood transfusions to reduce the risk of having another stroke.

Is there a way to find out if my child is at risk for having a stroke?

The Transcranial Doppler (TCD) exam can help determine your child's risk for having a stroke. This machine measures how fast blood flows through the brain. Children with fast blood flow through the brain have a higher risk of stroke. The TCD machine is similar to the ultrasound machine that is used to check pregnant women. A wand is pressed to the skin in order to record images inside the brain. The exam is painless and takes about 30 minutes to complete.

This test should be done at least once a year beginning when your child is around 2 years of age and is recommended for children with Hb SS or sickle beta zero thalassemia. A TCD is not recommended for children with Hb SC or sickle beta plus thalassemia because they are not at increased risk of having a stroke.

What is the treatment for an abnormal TCD?

Regular blood transfusions, usually monthly, have been proven to prevent strokes in children who have sickle cell disease. Regular transfusions of healthy blood help by reducing the number of sickle cells in the blood. Other treatment options may also be indicated such as hydroxyurea or a bone marrow transplant. Talk to your doctor to learn more.

TREATMENT OPTIONS FOR SICKLE CELL DISEASE

HYDROXYUREA



What is hydroxyurea?

Hydroxyurea is a medication that has been found to lessen the severity of sickle cell disease. The U.S. Food and Drug Administration has approved hydroxyurea for treatment of sickle cell disease. Hydroxyurea, usually combined with other medicines, was initially used to treat cancer, but now works best for sickle cell as it reduces the frequency of sickle cell-related complications.

It works by increasing the amount of fetal hemoglobin (hemoglobin F) within the red blood cells. Hemoglobin F helps prevent red blood cells from forming the sickle shapes and blocking the flow of blood and oxygen. Hydroxyurea is taken as a pill by mouth once every day. A liquid form is also available.

Who should take hydroxyurea?

Children who have complications due to sickle cell disease should consider taking hydroxyurea. Some of the reasons for taking hydroxyurea include:

- Pain episodes that require hospitalization or interfere with school or play
- Severe acute chest syndrome
- Pneumonia
- Repeated episodes of painful erection of the penis (priapism)
- Abnormal transcranial doppler when blood transfusions are not possible
- Repeated spleen sequestration events

Hematologists are shifting to the opinion that all children with the severe forms of sickle cell disease should take hydroxyurea as early as possible, unless there is a clear reason not to.

What are the side effects of hydroxyurea?

Most patients who take hydroxyurea do not experience side effects. Nausea, increased skin pigmentation, kidney and liver irritation and changes in blood counts (low platelets, low white count and low hemoglobin) occasionally occur. These side effects are typically resolved by decreasing the dose or stopping the medication for a short period of time.

What are the benefits of hydroxyurea?

- Decreased pain crises and other complications of sickle cell disease
- Fewer hospitalizations
- Fewer missed days of school related to sickle cell complications
- Improved energy and feeling of well being
- Decrease in sickling damage to body organs
- Increased lifespan and quality of life

How is hydroxyurea managed?

When your child first begins taking hydroxyurea, he or she will need to be seen often (usually monthly). This is necessary to monitor your child closely while the dose is being gradually increased over a period of months. Your child will be examined and have blood drawn to evaluate how the medication is working and to monitor for side effects.

When female patients have started their menses, a urine pregnancy test will also be done. This is necessary due to the risk of damage to the fetus while on hydroxyurea. It is important to prevent pregnancy while taking hydroxyurea by avoiding sex or using birth control methods. It is necessary to stop taking hydroxyurea during pregnancy.

TREATMENT OPTIONS FOR SICKLE CELL DISEASE

BLOOD TRANSFUSION THERAPY



Oluwatobi, age 16

A blood transfusion may be indicated for your child in the following situations:

- Stroke prevention
- Prior to surgery
- During a hospitalization for a sickle cell complication (such as ACS or splenic sequestration)

When will my child need a blood transfusion?

Stroke prevention

Researchers have determined that children with Hb SS disease have an 11 percent chance of having a stroke by age 20. Blood transfusions are the treatment of choice for any child who has had a stroke or who has an abnormal transcranial doppler (TCD). If a child has already had a stroke or has an abnormal TCD, the risk for having a stroke is very high.

If a child is at risk, it is necessary to start blood transfusions on a regular basis to suppress the body from forming red blood cells with sickle hemoglobin. The child will begin receiving blood transfusions indefinitely about every three to four weeks. The goal is to reduce the hemoglobin S to less than 30 percent in order to prevent a stroke.

Surgery preparation

If a child with sickle cell disease is scheduled for surgery with general anesthesia, preparation is necessary to avoid complications during surgery and recovery. Speak to your hematology provider if your child needs surgery, such as removal of tonsils and adenoids, spleen or gallbladder. Your child should be examined in the hematology clinic the week prior to surgery for evaluation and a possible blood transfusion. This is necessary to:

- Raise the hemoglobin in order to have more cells carrying oxygen to the body
- Lower the sickle hemoglobin to reduce sickling complications

Intravenous (IV) hydration is also recommended on the day of surgery to help reduce sickling complications.

Hospitalization

When a child with sickle cell disease is hospitalized for a pain crisis, acute chest syndrome, aplastic crisis or splenic sequestration, it is often necessary to give the child a blood transfusion to prevent the hemoglobin from dropping too low, a common sickle cell complication. In order to give your child a blood transfusion, an IV catheter in the vein will be placed in your child's hand or arm. The blood will be infused slowly over two to four hours.

Before your child receives a blood transfusion, you will be asked to sign a consent form. The consent states that you understand why the blood transfusion is necessary and what the risks are. Risks associated with blood transfusions are infection or a reaction due to antibodies.

To reduce these risks, the blood your child is given is leuko-reduced (filtered of cells that could pass infection) and matched to your child's blood type, as well as to blood antigens (proteins) most commonly found in African Americans.

Additionally, your child will be monitored very closely by the medical team during a blood transfusion. Vital signs are taken often during the transfusion. If your child develops a reaction, the medical team is immediately notified, the transfusion is stopped and appropriate medication and treatments are given.

The reason your child needs a blood transfusion and the risks will be further explained to you by the medical team.

What is iron overload?

Iron overload is an expected complication of receiving chronic blood transfusion therapy. When your body has too much iron, it can cause serious damage to the liver, pancreas, kidneys, thyroid glands and other organs. Once your child has received about 10-20 transfusions, his or her iron level may become too high.

Your child's iron level is monitored regularly by the ferritin level. A ferritin level is one of the tests performed when your child has blood drawn. A ferritin level greater than 1000 ng/ml is an indication of possible iron overload.

Your child's iron concentration can most accurately be determined by performing an MRI. The MRI will determine the iron concentration in your child's body and which organs are affected.

Once your child has iron overload, a medication to remove the iron from the body will be started. Removing iron from your body with medication is called chelation. Your medical team will discuss which chelation medication is best for your child.

TREATMENT OPTIONS FOR SICKLE CELL DISEASE

BONE MARROW TRANSPLANT

Joscelyn, age 11



Bone marrow transplantation, replacing the abnormal stem cells inside the bone marrow with healthy cells, is currently the only known cure for sickle cell disease. The healthy cells are obtained from an eligible donor, such as a brother or sister who is a close match. As a result of ongoing research, someone who is unrelated, but a close match, may also be donor.

What is bone marrow?

Bone marrow is a sponge-like tissue within the bones. It functions as a factory and produces all the blood cells for the body. The blood cells begin as stem cells. Stem cells grow and develop into red blood cells, white blood cells and platelets.

The bone marrow of people with sickle cell disease produces only abnormal red blood cells which contain defective sickle hemoglobin.

How does a child receive a bone marrow transplant?

In order for a child with sickle cell disease to receive a bone marrow transplant, the body must first be prepared. The purpose for preparing the body is to create a space in the bone marrow for the healthy stem cells to grow. To prepare the space in the bone marrow, a child with sickle cell disease is treated with chemotherapy to eliminate his or her own unhealthy bone marrow.

Then, he or she receives the healthy bone marrow stem cells which were donated either from a brother or sister or an unrelated person. The bone marrow stem cells are given IV (in the vein) similar to a blood transfusion.

How does a brother or sister donate healthy bone marrow stem cells?

The stem cells of the donor are usually collected through a process called a stem cell harvest. There are three different ways to harvest stem cells:

- The first and most common way is to obtain stem cells through the veins. The donor is given medicine to stimulate stem cell growth in the blood, then their blood is drawn from the vein through a pheresis machine. The pheresis machine collects the stem cells from the donor and saves them, then replaces the remaining blood back into the donor.
- If the donor has very small veins, stem cells can be collected from the pelvic bone instead. The donor is put to sleep with anesthesia and a small needle is placed into the pelvic bone. The bone marrow is then pulled into a syringe. Once the bone marrow is collected, the needle is removed and the donor awakens from anesthesia. Once awake, the donor is usually able to return home the same day. The site of the bone marrow harvest may be sore for a few days. The donor's bone marrow quickly grows new cells to replace those taken during the stem cell harvest.
- The third way of obtaining stem cells is by collecting them from a sibling's umbilical cord. The umbilical cord blood would have had to be stored at the time of birth of the sibling donor.

Is my child a candidate for a bone marrow transplant?

Your child is a candidate if he or she has both:

- Complications due to sickle cell disease such as a stroke, frequent pain episodes, multiple episodes of acute chest syndrome or an abnormal MRI (ultrasound of the brain) and
- A closely matched donor
 - A brother or sister who does not have sickle cell disease
 - A sibling who has sickle cell trait
 - A parent, other relative or someone from the general population

Transplantation from a matched unrelated donor is also an option, but is much riskier than transplantation from a relative. This type of bone marrow transplant is performed only as part of research studies.

What are the risks of a bone marrow transplant?

Although the majority of children do well after bone marrow transplantation, there are possible risks.

Once the new stem cells are in the bloodstream of the child with sickle cell disease, it will take several weeks for the new stem cells to grow and develop. During this time, the child is at risk for infection or bleeding. Infection or bleeding may be severe, and, in rare cases, may cause death. Your child will remain in the hospital for approximately two to three weeks while the new stem cells grow in the bone marrow.

In some cases, the transplant may fail. This will lead to the return of sickle cell disease in your child. Other risks are possible, and you should discuss them with your doctor in detail.

CARE IN THE SCHOOL SETTING

Because sickle cell disease places your child at risk for complications at any time, he or she is eligible to receive support services within the school environment. It is very important to keep the teachers and staff updated on your child's health status regularly. It is also important to check on your child's progress in all areas of learning and succeeding in his or her school experience.

Difficulty in school may occur due to an obvious or silent stroke. Your child may also face learning challenges due to anemia and decreased oxygen to the brain as a result of sickle cell disease.

Speak to your child's medical provider or social worker regarding what your child's school may need to know to help him or her have a positive school experience. You may ask your child's medical team to write a letter of medical condition and provide information to the school to help them understand and better provide for your child's needs.



Charlysia, age 11

Children with sickle cell disease may require some school modifications, as detailed below.

How will my child stay hydrated?

It is very important for children with sickle cell disease to drink lots of water and stay well hydrated to help prevent pain episodes and organ damage. It is recommended that they be allowed to carry a water bottle and take restroom breaks when needed.

Can my child participate in sports?

It is important for all children to be physically active. It is especially important for children with sickle cell disease to participate in physical activity and sports teams.

However, it is key that the child know his or her limits and not push his or her body too hard. Extended or strenuous activity could cause or exacerbate a pain crisis. Discuss with your child's coach or physical education teacher the need to allow for rest during activities and the ability to substitute activities that are less strenuous.

Will my child have extra absences?

Children with sickle cell disease may be absent from school for many reasons related to the management of their health such as appointments, checkups, tests, treatment of complications, visits to the emergency room or hospitalizations. Missed days of school contribute to increased stress and challenges for your child to achieve success.

It is important to be in close contact with your child's teacher to make sure he or she is not falling behind in school. It may also be a good idea to ask the teacher to prepare homework or arrange for extra time or tutoring to keep up or catch up with school work. A social worker may be able to assist you in communicating with the school to help achieve these goals.

What is the process for getting help for my child in school?

Your child may receive help either through Section 504 of the Rehabilitation Law of 1973 or through the Individuals with Disabilities Education Act. The law requires public schools to work with parents of children with disabilities to create an Individualized Education Plan to meet the child's needs. To get help, start by requesting a special education evaluation from your school's Admission, Review and Dismissal team. Be prepared to discuss the specific areas of struggle and difficulty for your child.

RESEARCH: WHAT THE FUTURE HOLDS

Texas Children's Hospital is dedicated to finding a cure for sickle cell disease and finding better ways to treat complications of sickle cell disease. Our ongoing participation in many research studies, both nationally and internationally, is aimed at improving your child's overall health and quality of life.

What are the goals of research?

Research studies measure the safety and effects of new treatments and procedures. Volunteers in research studies help doctors and scientists learn how different treatments affect different people. Participation helps improve the health of all people. Federal guidelines and codes of ethics protect research volunteers from harm. Safety and volunteer rights are required legally in any research study.

What should I consider before volunteering my child for a research study?

- Benefits
 - Getting treatment for an illness when no other treatment options exist
 - Receiving high quality, comprehensive care for your child's condition
 - Being among the first to have a new treatment
 - Knowing your child's participation is helping others
- Risks
 - Not being able to choose your child's treatment
 - An unexpected reaction to treatment
 - Unpleasant or serious side effects

Before your child participates in a research study, learn as much as possible. Ask questions until you have a clear understanding of the research study and how it will affect your child. Remember – participation is strictly voluntary and you may remove your child from the study at any time.

What questions should I ask before volunteering my child for a research study?

- What is the purpose of the study?
- What has been learned about the study treatment so far?
- Will I have to pay for anything for my child to participate in the study?
- Will I still be able to see my child's doctor?
- How often will I have to take my child to Texas Children's Hospital while my child is in the study?
- How long will the study last?
- If the treatment works for my child, will he or she be able to continue with the treatment when the study is finished?
- Will anyone else know that my child is in a research study?

To find out more, be sure to talk to your medical team about the sickle cell research studies in which Texas Children's Hospital partakes. Your interest will have a positive effect on the improvement of health care for all children with sickle cell disease.



TRANSITION

The teenage years are an exciting time for you and your child with sickle cell disease. By this time, your teenager has grown, faced many life challenges and changes and achieved many accomplishments.

What is transition?

One of the things your teen will need to prepare for is shifting his or her health care from a pediatric medical provider to an adult medical provider. This is known as transition. Transition is a gradual process involving the teen, parent and entire medical team.

When will my teen be transitioned to an adult doctor?

The age that your teenager will start seeing an adult doctor may vary a little depending on individual health care circumstances. Transition will usually occur around the age of 18 or soon after high school graduation.

How will I know when my teen is ready to transition?

Your medical team will guide you through this process for several years. Focusing on getting ready for transition will be a crucial part of the visit with your teen's health care provider during his or her high school years and even earlier. Often teens have concerns about leaving the pediatric medical team that they are familiar with and moving to an adult medical team that they do not know. This may cause feelings of fear, anxiety and worry. This is a normal adjustment reaction, and these concerns should be addressed during your visit to the medical provider.

What should I do to get my teen ready for transition?

There are many ways to help both you and your teen become ready for transition. The most important thing to remember is to begin early. Your health care team will help you focus on the areas that are necessary for a good transition experience. A large part of getting ready is building a solid knowledge base.

Some of the expectations of your teen prior to transition are:

- A good knowledge base about sickle cell disease. He or she should be able to talk about sickle cell disease and explain his or her own experience. It is important for them to know their medications, baseline blood test results, surgeries, treatments and complications.
- Meet with the provider for at least part of the medical visit without the parent. This helps to build self-confidence.
- Schedule his or her appointments.
- Ask for and call in refills for his or her medication.
- Know about his or her insurance coverage and medical documents.
- Choose an adult doctor and schedule an appointment.

Take time to address your teen's readiness and concerns about transition with his or her medical provider. It is important to work together as a team to achieve the best health care possible.

GLOSSARY OF TERMS

Acute Chest Syndrome (ACS) - pneumonia and sickle cell crisis in the chest

Afebrile - no fever

Analgesic - a drug or medication that relieves pain

Anemia - low hemoglobin level in the red blood cell that decreases the delivery of oxygen to the body resulting in fatigue, paleness and shortness of breath

Antibody - proteins used by the immune system to remove bacteria, viruses and foreign substances from the body

Antigen - a substance found on the cell that can cause the body to form antibodies and trigger an immune response

Aplastic crisis - an episode caused by a viral infection that causes the bone marrow to stop making red blood cells temporarily

Blood vessels - the tube like structures throughout the body that blood flows through

Bone marrow - the place inside the bones where blood cells form and grow

Chelation - therapy to remove iron (or other metals) from the body with special medication

Complete Blood Count (CBC) - a blood test that measures the number of red blood cells, white blood cells and platelets; it also contains other information about the cells such as the size, shape and amount of hemoglobin

Dehydration - lack of adequate fluids in the body; it can result from either not taking in enough fluids or by losing fluids from sweating, diarrhea, vomiting and urinating

Fetal hemoglobin - the most common type of hemoglobin found in the fetus or unborn baby; it is later replaced with other hemoglobin, but some people continue to produce fetal hemoglobin throughout their life

Gene - the basic unit of heredity found inside the cell; genes are passed on by a mother and father and genes determine all the characteristics that make up each person including hemoglobin type

Hematologist - a doctor that specializes in diagnosing and treating blood disorders

Hemoglobin (Hb) - the part of the red blood cell that carries oxygen to all parts of the body

Hemoglobin S beta plus thalassemia - the form of sickle cell disease which occurs when you inherit a hemoglobin S gene from one parent and a beta thalassemia gene from the other parent; a small amount of normal hemoglobin is produced; a milder form of sickle cell disease

Hemoglobin S beta zero thalassemia - the form of sickle cell disease which occurs when you inherit a hemoglobin S gene from one parent and a beta thalassemia gene from the other parent; a severe form of sickle cell disease, similar to Hb SS

Hemoglobin SC (Hb SC) - the second most common form of sickle cell disease which occurs when you inherit a hemoglobin S gene from one parent and a hemoglobin C gene from the other parent; a less severe anemia, but similar symptoms to the person with Hb SS disease

Hemoglobin SS (Hb SS) - the most common and severe form of sickle cell disease which occurs when you inherit a hemoglobin S gene from each parent

Hemolytic anemia - a type of anemia caused by the destruction or breakdown of red blood cells

Hydration - providing adequate fluids, such as water, to the body to maintain fluid balance in the cells and tissues

GLOSSARY OF TERMS (continued)

Infection - invasion of the body by bacteria and viruses causing fever and illness

Inherited - passed on from the father and mother to their child

Jaundice - yellow color to eyes and skin caused from the release of bilirubin from a red blood cell; may also be caused by liver disease

Meningitis - an infection in the brain or spinal fluid due to invasion of the meningococcal bacteria

Ophthalmologist - a doctor who specializes in diseases or disorders of the eye

Osteomyelitis - an infection inside the bone due to a bacterial infection

Pallor - paleness of the skin due to anemia or a low blood count

Pheresis - a procedure in which the blood of a donor is filtered in a machine to remove certain substances and the remainder of the blood is given back

Platelets - a blood cell that helps to form a clot to stop bleeding

Pneumonia - an infection in the lungs caused by bacteria or viruses

Prophylactic - protection or prevention of disease or infection

Sepsis - a potentially life-threatening bacterial infection in the blood

Sickle cells - red blood cells that are shaped like a sickle, crescent or banana

Sickle cell disease - an inherited blood disorder affecting the hemoglobin in the red blood cell; the abnormal form of hemoglobin results in changes in the cell leading to multiple health complications; refers to all types of sickle cell hemoglobin disorders

Sickle cell trait - a normal hemoglobin gene is inherited from one parent and a sickle hemoglobin gene is inherited from the other parent; someone with trait does not have sickle cell disease, but they may pass the trait to their children

Spleen - an organ on the left side of the abdomen that acts as a filter for blood and helps fight infections

Transcranial Doppler (TCD) - an ultrasound to measure the speed of blood flow in the vessels of the brain; the speed of blood flow is related to the risk of having a stroke

Transition - the process of moving health care from one provider to another; such as pediatric to adult care; the goal is to provide health care that is uninterrupted and developmentally appropriate

White blood cells - the cells in the blood that help fight infection

NOTES

WEBSITES

To find out more about sickle cell disease, you may also visit the following websites:

www.SCInfo.org (The Georgia Comprehensive Sickle Cell Center)

www.sicklecelkids.org (an interactive website for kids to learn more about SCD)

www.teenshealth.org (information on health related topics, including SCD)

www.sicklecelldisease.org (The Sickle Cell Disease Association of America, Inc.)

www.ascaa.org (American Sickle Cell Anemia Association)

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